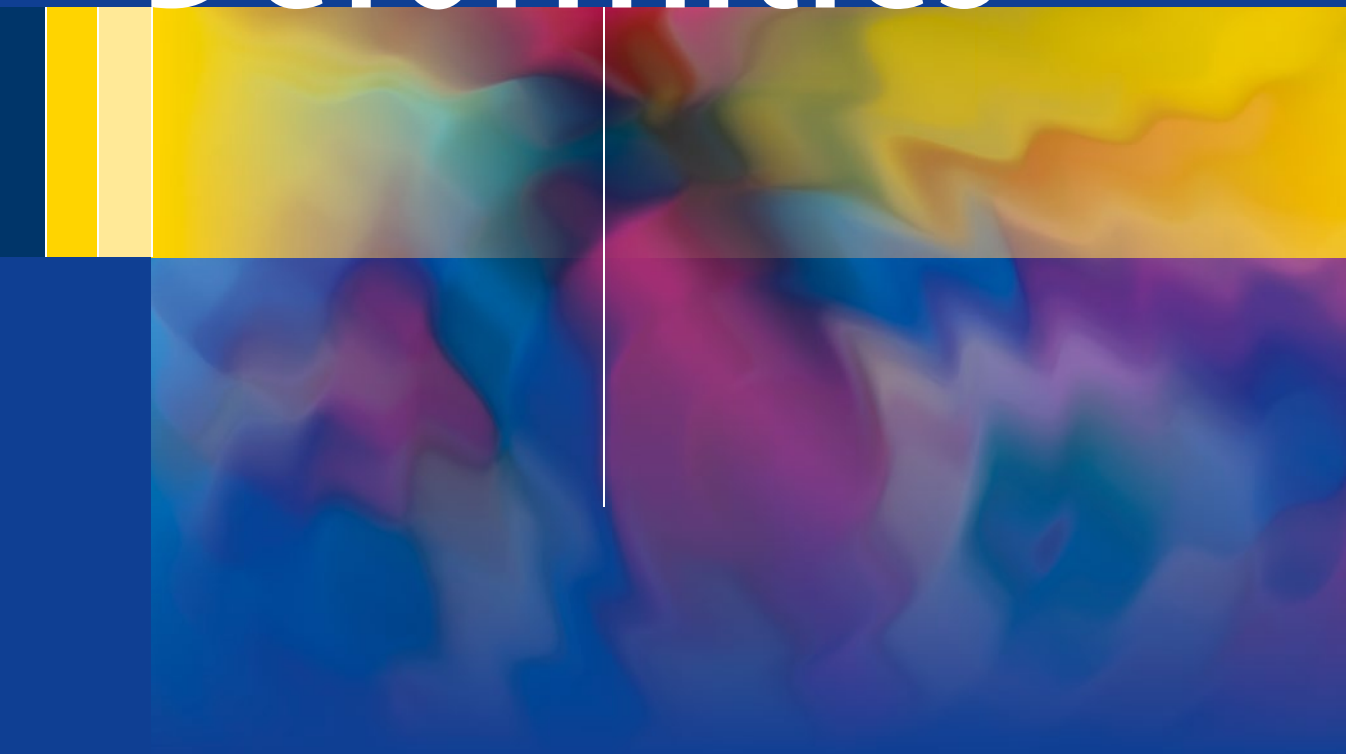
Amulya K. Saxena
Editor

Chest Wall Deformities



Springer

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“The performance of surgery is art, the rest is all science”

This book is dedicated to the memory of my father Dr. Gopal Krishna Saxena, a person of principles and immense virtues and talents who dedicated himself to support my education and guided me throughout.

Preface

Chest wall deformities present a wide spectrum of anterior thoracic cage anomalies, and depending on the specific type of deformity the most appropriate approach must be employed to achieve optimal results.

The approach towards management of patients with chest wall deformities has undergone enormous transformations in the past century. In the beginning of the last century, with the advent of initial successes in open procedures for treatment of various thoracic organ pathologies largely attributed to the advances in anesthesia and assisted ventilation, correction of the deformities of the anterior chest wall was attempted. These pioneers who performed chest wall deformity repairs have to be commended for the development of their respective techniques bearing in mind the unavailability of high quality commercially manufactured instruments in that period. However, when compared to contemporary times, slow communication and exchanges between centers at that time contributed towards the emergence of variations in techniques for the surgical management of chest wall deformities. Furthermore, at that time the evolution of novel techniques were known either through presentation at National Congresses or through publications in local languages, which again played a major role in the lack of popularity or precise understanding of these techniques. Also, with the limited use of photography, it was difficult to picture the results of corrective surgery and to understand the analysis of results. Towards the end of the last century, larger series were published using the few open techniques that gained popularity, but these reports were limited to centers that gained surgical experience after management of large number of patients.

The introduction of a minimal *access* repair of *pectus excavatum* in the beginning of this century was the first technique that had a global impact in the acceptance of a single concept towards the management of the funnel chest type of chest wall deformities. The minimal *access* repair technique has been frequently referred incorrectly to minimal *invasive* repair, bearing in mind that this technique is extremely invasive but is only performed through small-size incisions. The popularity of minimal *access* repair in the present era of mass communications has allowed comparison of the outcomes between centers worldwide and towards the development of safer standards in performing these procedures. The introduction of thoracoscopy in the minimal *access* repair technique was an important addition to this technique to improve safety during surgery. The success of minimal *access* repairs of funnel chest was then modified and employed for the treatment of pigeon chest

type of chest wall deformities. In addition to the widespread practice of minimal *access* repairs, another aspect in the treatment of chest wall deformities was the utilization of noninvasive techniques such as the application of suction devices for the treatment of certain types of funnel chest deformities and the use of braces for the correction of specific types of pigeon chest deformities.

This monograph is a compilation of the various interrelated subjects associated with chest wall deformities. The introductory sections address topics that are related to classification, diagnostics, and associations, knowledge of subjects that are necessary when managing patients with these deformities. This is followed by chapters that focus on the management of funnel chest deformities and surgical techniques for repairs. The chapters on techniques have been presented with schematic illustrations to guide the reader through the important steps of the procedures. Dedicated sections focus on the management of pigeon chest deformities and their operative repairs. The final section's emphasis is on other conditions that present as chest wall deformities as well as options to their respective management. This comprehensive work is a concise collection of conventional surgical procedures and contemporary techniques for surgeons that manage patients with chest wall deformities.

London, UK Amulya K. Saxena

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Part I

General

History of Surgical Repairs of Chest Wall Deformities

1

Amulya K. Saxena

Introduction

Chest wall deformities represent a spectrum of musculo-skeletal disorders of the anterior chest wall. Most of these deformities fortunately are not life-threatening and do not manifest in severe functional pathophysiology of the thoracic organs. The management of these deformities has gained more importance in the past decade because of the increased participation in competitive school sports as well as athletics, and the changing trends in the world of fashion which expose more of the chest to the public view. It is important to note that, although thoracic wall deformities do not arouse the sympathy generated by limb or cranial anomalies, children with such deformities are viewed with curiosity by their classmates and are often confronted by teasing remarks.

Except for severe chest wall deformities, it is generally extremely difficult to predict the course of progression after the deformities have been discovered in early infancy. Therefore, mild forms of abnormalities warrant the *wait- and-watch* approach during the first 4–5 years before operative management can be considered. In mild cases, the dormant deformity sometimes becomes accentuated during adolescent growth. Since, the deformities can also manifest frequently primarily during the pubertal spurt, often with rapid progression, operative correction in young adults is more favorable in mild cases. The correction techniques offered today have evolved over the last century and development in surgical finesse with comparison of results has now set the trends in the approaches towards treatment of chest wall deformities.

Historical Background on Chest Wall Deformities

Pectus excavatum was recognized as early as the 16th century Johannes Schenck von Grafenberg (1530–1598) who collected literature on the subject [1]. Schenck von Grafenberg was a German physician who student studied at Tübingen, and was later a physician to the city of Freiburg im Breisgau (Fig. 1.1). He was one of the more influential authorities on medicine during the late Renaissance Era. Schenck von Grafenberg's best-known written works were the *Observationum medicarum rariorum*, which was a seven-volume compendium that described pathological conditions concerning all the parts of the human body (Fig. 1.2). Information in these books was derived from medical experiences described by Schenck and his

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Fig. 1.1 Johannes Schenk von Grafenberg (1530–1598) (Printed with permission: Bibliothèque Interuniversitaire de Santé, Paris, France)

OBSERVATIONVM
MEDICARVM,
RARARVM, NOVARVM,
ADMIRABILIVM, ET MONSTROSARVM,
LIBER SECVNDVS.
DE PARTIBVS VITALIBVS,
Thorace contentis.
In quo, quæ Medici doctiſſimi & exercitatiſſimi, abdita, vulgò incognita, gravia, periculoſaq; , in harum partium Conformationibus, earundemq; Morborum Cauſis, Signis, Eventibus, & Curationibus accidere compererunt, Exemplis vt plurimum, & Hiſtorijs propoſita exhibentur:
Studio atq; Opera
IO. SCHENCKII A GRAFENBERG,
apud Friburgum Briſgavorum Medici, multa & varia lectione conquiſita.
Continentur præterea hoc libro, illuſtrium ætatis noſtræ Medicorum ANECDOTA complura, & nunquam publicata Exempla memorabilia, quorum Autores ſingulari Catalogo traduntur.
FRIBVRGI BRISGOIÆ,
Ex OFFICINA MARTINI BECKLERI:
Anno cIↃ. IↃ. XCIV.

Fig. 1.2 Volume 2 of Observationum medicarum rariorum liber, Johannes Schenck von Grafenberg published in 1594 (Printed with permission: Bavarian State Library, Munich, Germany)

contemporaries, and well as medical observations taken from existing sources to that date.

This was followed by classic paper by the Swiss anatomist Bauhinus in 1594, who described the clinical features of pectus excavatum in a patient that suffered from pulmonary compression with dyspnea (shortness of breath) and paroxysmal cough as a result of a severe pectus excavatum [2, 3]. Johannes Bauhinus (1541–1613) was actually a Swiss-French botanist and the son of Jean Bauhin (1511–1582) a French physician to Johanna of Albret (Queen of Navarre) who moved to Basel (Switzerland) from Amiens (France) after converting to Protestantism. He studied botany at Tübingen under Leonhart Fuchs (1501–1566, traveled and collected plants with Conrad von Gessner (1516–1565). Later, he practiced medicine at Basel and was the elected professor of rhetoric in Basel (1566). He became physician to Duke Frederick I of Württemberg at Montbéliard (1570) and remained so until 1613 [4].

Another documented description of a deformity of the thorax could be found in 1860 by Woillez in Paris [5]. In 1863, von Luschka [6] reported about a 6 cm deep depression in the thorax wall in a 24-year-old man. In 1870, Eggel [7] published the first comprehensive case report of a patient with a funnel-formed thorax depression calling it a '*miraculum naturae*'. He presumed that the deformity was a weakness and an abnormal flexibility of the sternum caused by nutritional disturbance or by developmental failure. Individual case reports were then followed by Williams [8], Flesch [9] and Hagmann [10].

The topic of pectus deformities regained limelight in 1882 when attention was again directed to the deformity by Wilhelm Ebstein [11], discoverer of Ebstein's disease (Fig. 1.3). In his report, which heralded the commencement of modern investigations of the deformity, Ebstein presented the theory that the anomaly was caused by arrested growth of the sternum. He also gave the deformity its popular German name of "*Trichterbrust*" (funnel chest). Wilhelm Ebstein was a German physician. He studied medicine at the Universities of Breslau and Berlin, graduating from the latter in 1859. In that year he was appointed as physician at the Allerheiligen Hospital, Breslau. Later on, in 1868, he was appointed as the chief physician at the municipal poorhouse and in 1874 as Professor in Göttingen University. In 1877, he became the director of the University hospital. Wilhelm Ebstein's interest in pectus excavatum was shared by his son Erich, who seventeen years later also wrote a detailed study on the subject [12]. Treatment at this time was limited to fresh air and exercises [13, 14].

The subject of the first clinical report in the modern medical literature was from a Viennese university student, Heinrich Wojaczek [15]. Wojaczek then traveled widely through Europe, displaying his "sunken" chest at different Universities which further increased reporting of the condition [16, 17].

Fig. 1.3 Wilhelm Ebstein (1810–1902) (Source: US National Library of Medicine)

Historical Reports on Etiology of Chest Wall Deformities

The genetic predisposition of the deformity was first noticed by Coulson in 1820, who reported a family of three brothers with pectus excavatum [18]. It was later Williams in 1872, who described a 17-year-old patient born with a pectus excavatum whose father and brother also presented the same deformity [8]. Reports also emerged on the trauma related etiology of chest wall deformities by Alexander [19]. Although most of the cases identified and reported were congenital, occupational etiology was also presumed to be responsible for the development of these deformities with the introduction of nomenclature such as "*zapatero de torax*" (Shoemaker's chest) in the Spanish. In 1873, the theory that chest wall deformities resulted from an overgrowth of cartilages was reported by Flesch, a medical student who quoted the suggestions made by his chief Schiffer [20].

Lester correlated the development of chest wall deformities has also been to various intrauterine compressive forces such as pressure by the chin, knee or elbow, and also to postnatal conditions such as obstructive diseases of the respiratory tract, rickets, tuberculosis or congenital syphilis [21, 22]. Raubitschek suspected latent mediastinitis as the possible cause of this condition. With regards to the embryonic association of chest wall deformities, Müller reported that the depression of the sternum is a expected developmental finding in the normal embryo and is not a result of non-directional growth [23]. This depression may persist at birth and could increase in later life in certain individuals. The first association between pectus deformities and collagen diseases was published by Curschman in 1935, who reported a patient with pectus excavatum, arachnodactylia and ectopic lenses [24].

There was a shift in ideology during the 1900s in which the diaphragm was implicated in the etiology of chest wall deformities based on earlier

reports of Bauhinus and Woillez [2, 5] which implicated the centripetal force of the *centrum tendinosum* of the diaphragm. The role of diaphragm was accepted in the etiology of chest wall deformities and surgical procedures were conceived by Lincoln Brown bearing in mind the role of the diaphragm [25]. However, the diaphragm theory, together with the proposed Brown operation that utilized the dissecting of the sterno-diaphragmatic attachments did not receive acceptance for the treatment of pectus excavtum.

Paradoxical breathing, a common symptom in patients with pectus excavatum, was first mentioned by Nageonette-Wilbourschewitz in 1925 [26]. Compression of the heart was recognized and radiologically documented by Pohl in 1928 [27].

Historical Advances in Thoracic Surgery

In 1883, Block, a surgeon in Dazing, East Prussia, now Gdansk, Poland, performed the first surgical procedure on the thorax – a pulmonary resection on a female relative, after performing a series of animal experiments that were presented to the German Society of Surgery meeting [28]. Soon after he opened the chest, the atmospheric pressure not only collapsed the lung, but also put pressure on the opposite lung, and without the means to counteract the atmospheric pressure the patient's vital signs gradually declined and the patient died. Block was severely criticized and admonished all other doctors never to repeat such a procedure. This unfortunate outcome retarded the development of chest surgery for half of century. The story has a more tragic end, for not only was the patient a relative of Block, but he committed suicide through a self-inflicted shot to the head after his failed procedure [29]. A contemporary lecturer in 1883 indicated that: "The first attempt of this kind had such an exceedingly tragic ending that every sensible surgeon should be warned to resist the temptation to make any future trial of the method". Ernest Dieffenbach's admonition to "stop at the pleura" was obeyed by all prudent surgeons for the next half of the century [30]. Surgeons had yet to understand and find methods to prevent the lungs from collapsing when the chest was opened.

In 1904, two new anesthesia techniques were suggested to solve the open chest problem. Ferdinand Sauerbruch (1875–1951) (Fig. 1.4a, b), from the surgical clinic of the famous von Mikulicz-Radecki (1850–1905) in Germany, introduced his method of "*Unterdruck*" (low pressure) ventilation, where the lung was maintained expanded during thoracotomy by keeping the patient's entire body inside a negative pressure chamber (at −15 cm H2O), while the patient's head remained outside the chamber at atmospheric pressure (Fig. 1.5) [31, 32]. The other approach, "*Uberdruck*" (high pressure), was to keep the lung expanded by placing the patient's head in a positive pressure chamber was proposed by Ludolph Breuer (Fig. 1.6) [33]. Naturally, unterdruck method took the early lead, and was advanced by the eminent Sauerbruch who proceeded to build negative pressure whole operation theaters large enough to accommodate the entire surgical team dressed in suits and helmets connected to outside atmospheric pressure while successfully performing open chest operations. Sauerbruch became the acknowledged leader of thoracic surgery in Europe, and the Surgeon-General of the German army during World War II.

In 1908, Sauerbruch visited the USA to present his negative pressure chamber at the meeting of the American Medical Association. At the end of his visit he did not take the bulky theater back to Berlin, but left it with the prominent New York thoracic surgeon, Wilhelm Meyer (1858–1922), an unconditional advocate of the concept. Meyer and his brother Julius, an engineer, continued research and designed what they called a "universal chamber", allowing either negative pressure with the surgeon inside, or positive pressure by a small box for the patient's head, along with the surgical team working on the open chest outside at atmospheric pressure. In 1911 this highly complex construction was installed, and used for a series of operations at the thoracic surgical service of the Lenox Hill – Hospital during World War I. His textbook "*Die Chirurgie der*

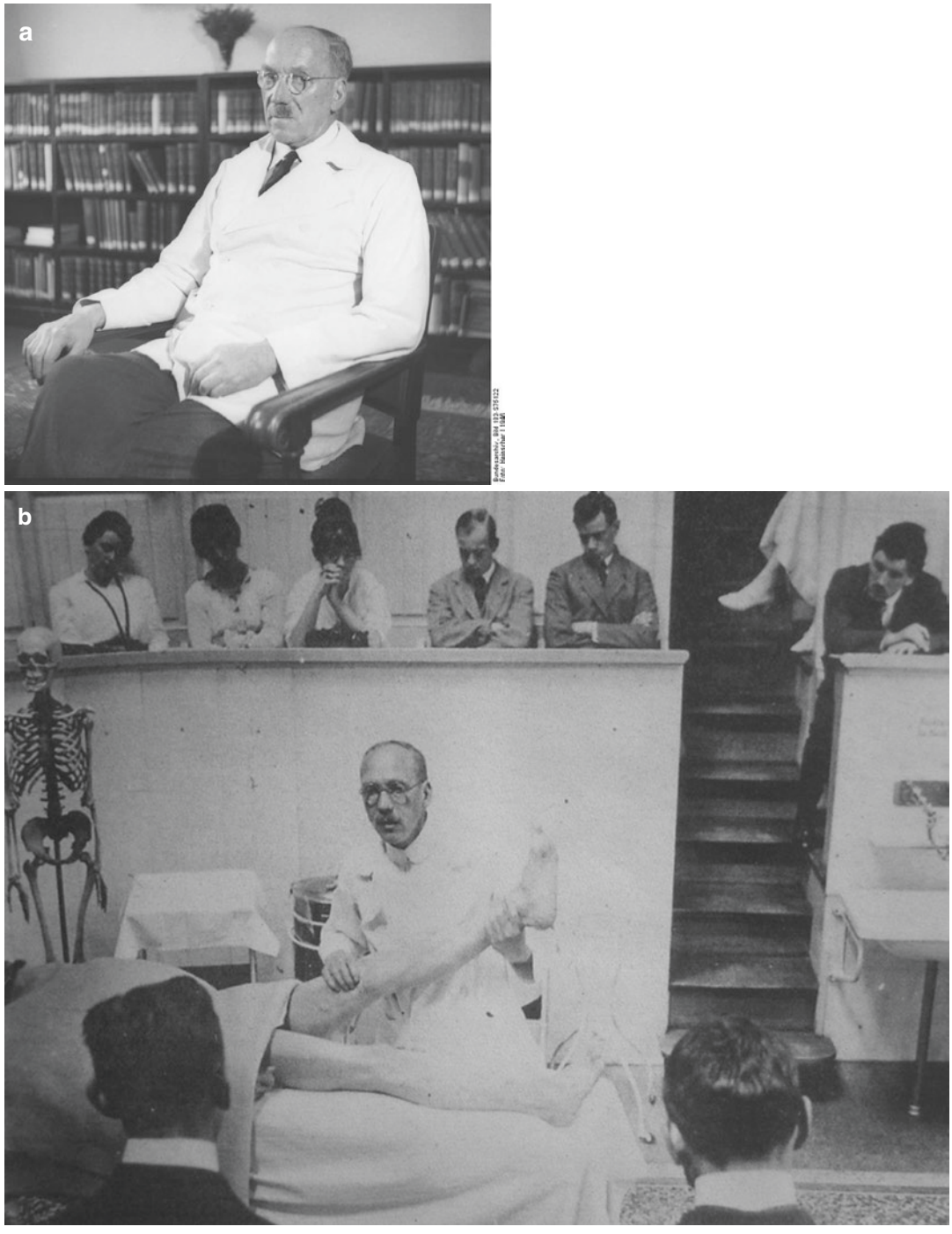

Fig. 1.4 (**a**) Ferdinand Sauerbruch (1875–1951) (Source: German Federal Archives, Picture 183-S75122/CC-BY-SA [http://creativecommons.org/licenses/by-sa/3.0/de/deed.en], via Wikimedia Commons). (**b**) Ferdinand Sauerbruch (1875–1951) giving medicine lectures at the University of Zurich where he worked between 1910 and 1917 (Source: Wikipedia Public domain)

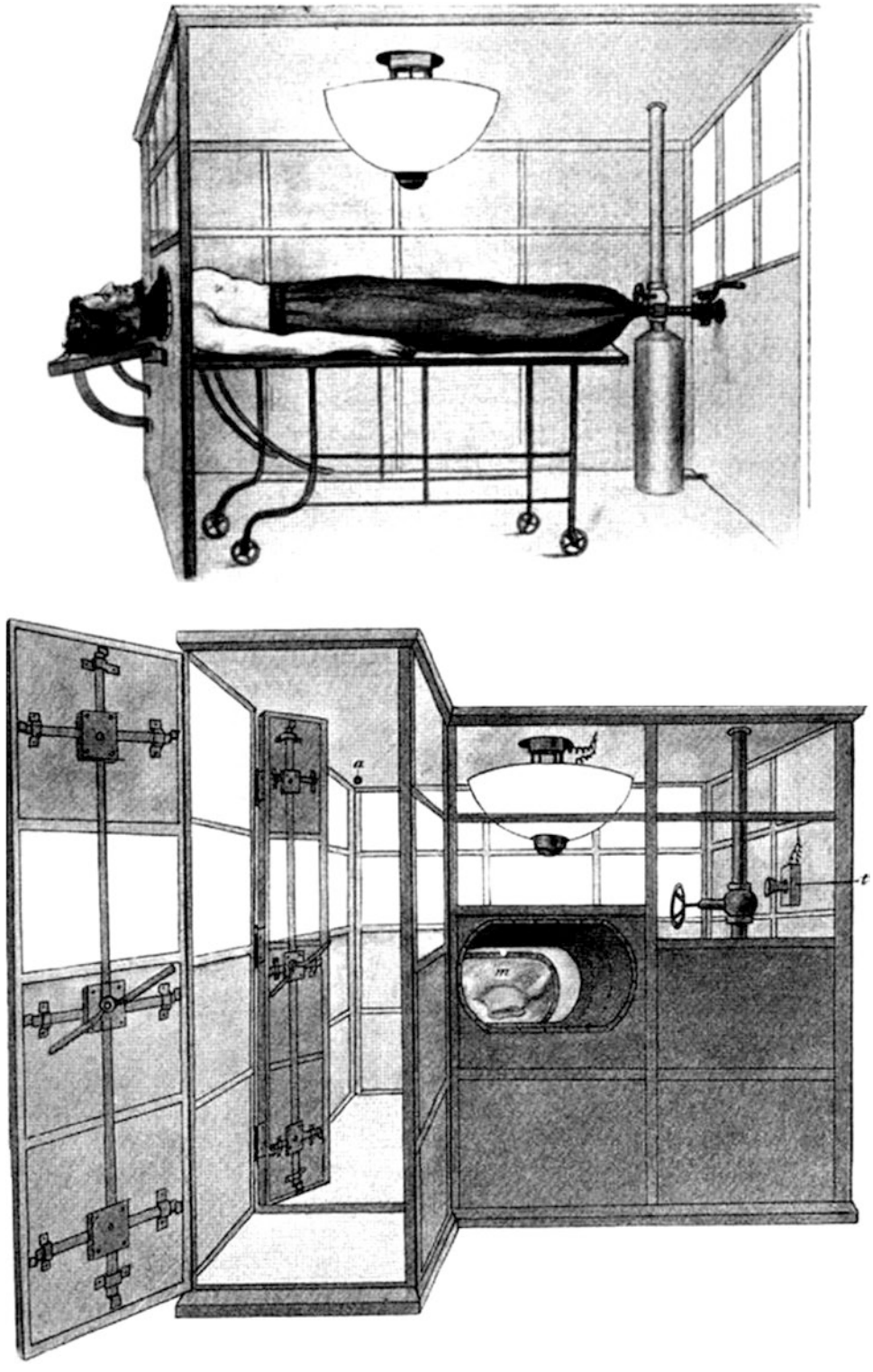

Fig. 1.5 Drawing of a "*Unterdruck*" chamber designed by Sauerbruch for thoracic surgical procedures (Source: Wikipedia public domain)

Fig. 1.6 Ludolph Breuer who presented his "Überdruck" concept for performing of thoracic surgical procedures

Brustorgane", first published in 1918, remained the classical bible of thoracic surgery until 1930.

History of Chest Wall Deformity Repairs

Wilhelm Meyer in 1911 performed the first surgical intervention for the repair of pectus excavatum [34]. Meyer resected short segments of the right 2nd and 3rd costal cartilages with the intention of relieving cardiac compression. As reported by him, the patient, who was significantly dyspneic before the intervention, showed significant improvements afterwards. The non-invasive nature of this procedure and the success achieved by it to relieve cardiac compression was put into question by Sauerbruch, who performed a more radical procedure in 1913 by resecting segments from the 5th down to the 9th costal cartilage, along with the corresponding left hemisternum [35]. After his surgery, though the heart was seen to pulsate under the muscle flap, the first patient operated was able to work without dyspnea and married three years later.

Lexer in 1927 intended to treated a patient with severe pectus excavatum with the aim of resecting the sternum and replacing it in a turned-over position. But during surgery, the resulting gap between the turned-over sternum and the cartilages was found to be too large, which resulted in the removal of the depressed sternal segment [36]. In 1929, Alexander operated two patients with pectus excavatum acquired after traumatic injuries. In the first patient, the sternum was divided transversely at the level of the third rib, then axially down to the xiphoid process. In the second patient, the entire sternum was removed, but the manubrium was secured to the neighboring costal cartilages. He observed that the lack of covering over the heart did not have clinical consequences [19]. He further used external traction after open repair and maintained it for various intervals with the assumption that enough tissue fixation might take place. Ochsner and DeBakey then reviewed the early experiences with repair and reported them in 1938 [37]. In 1943 Nissen explanted the deformed portion of the sternum, flipping it over and reattaching it horizontally to the costal cartilages [38]. Judet and Judet in 1954 performed a sternal turn-over procedure by reattaching the resected sternochondral apron into the resulting gap in the anterior chest wall (Fig. 1.7) [39].

Over the next few years, pectus excavatum surgery was dominated by the theory proposed by Lincoln Brown (Fig. 1.8), that the deformity was caused by undue traction exerted upon the sternum by the diaphragmatico-sternal ligament [25]. Accordingly, in 1940 he performed an operation in which all the tissue attaching the diaphragm with the lower sternum was dissected. While the operation seemingly worked in the very young, it was not found to be successful in older individuals. In 1943 Sweet performed a procedure, consisting of a transverse sternal osteotomy and bilateral cartilaginous resection, quite similar to what later became

Fig. 1.7 Robert Judet at Hôpital Raymond Poincaré – Garches, France (Mostofi, S. *Who's Who in Orthopedics*. London: Springer. 2005)

Fig. 1.8 Abraham Lincoln Brown at the UCSF Medical Center, Mount Zion, California, USA

known as the "classic" Ravitch operation [40]. Following this trend, the principles of modern pectus excavatum surgery were laid down by Brodkin in 1948 [41], and by Ravitch in 1949 (Fig. 1.9) [42]. Ravitch accepted Brown's theory and as a result advocated even more radical mobilization of the sternum, with transection of all sternal attachments, including the intercostal bundles, rectus muscles, diaphragmatic attachments, and excision of the xiphisternum. He published his experience in 1947 with eight patients using this radically extended modification of Sauerbruch's technique of bilateral cartilage resection and sternal osteotomy. Since the sternum was cut loose from all its attachments, Ravitch believed the sternum would no longer sink back into the chest and eliminated the use of external traction.

In 1956, Wallgreen and Sulamaa in Finland introduced the concept of internal support by use of a slightly curved stainless steel bar (Fig. 1.10) [43]. This steel bar was pushed through the caudal end of the sternum from side to side, bridging the newly created gap between the sternum and ribs. This introduced the era of osteosythetic stabilization in chest wall surgery. Fritz Rehbein (1911–1991) in 1957 developed a procedure that involved presternal sternum fixation after mobilization of the concavity, in which the elevated sternum was held in a position of slight overcorrection by steel splints and metal bands that were left in place for 3 years (Fig. 1.11) [44]. This procedure was evaluated to have good results in 69.2 %, satisfactory in 18.3 % and poor in 12.5 % of cases [45]. As early as 1958, Welch advocated a less radical approach as Ravitch with excellent results in 75 cases without cutting through all the intercostal bundles and not cutting through the rectus muscle attachments [46]. In 1961, Adkins and Blades modified the concept of internal bracing by passing a straight stainless steel bar behind the sternum rather than through the sternum [47].

Gerd Hegemann's report on surgical management with metal struts to stabilize the sternum in 1965 from Erlangen-Nürnberg, laid the foundation for the present techniques practiced

Fig. 1.9 Mark Ravitch (1934–1989) from John Hopkins Baltimore, Maryland, USA (Printed with permission: The Alan Mason Chesney Medical Archives of The Johns Hopkins Medical Institutions, Baltimore, Maryland, USA)

Fig. 1.10 Matti Sulamaa (1912–1978) from Finland introduced the concept of application of metal struts for repair of chest wall deformities (Courtesy of Prof. Risto Rintala, Department of Pediatric Surgery, University of Helsinki, Finland)

worldwide (Fig. 1.12) [48]. Hegemann opted for a radical approach with (a) bilateral parasternal chondrotomy, (b) partial or wedge sternotomy, (c) employment of a transsternal strut and (d) fixation of struts with mutiple wires. Günter Willital from Münster who was a student of Hegemann modified the technique and employed (a) bilateral serial rib osteostomies, (b) additional longitudinal struts and (c) the use of 2 metals wires in general (Fig. 1.13) [49]. Willital and the author presented the largest series of 1262 chest wall deformities operated with this method with good outcomes (Fig. 1.14 a/b, Fig. 1.15) [50].

Robert Shamberger and Kenneth Welch reported a large series in 1988 and reported an experience with corrections on patients from 1958 to 1987 on 704 patients that had corrections for pectus excavatum (Fig. 1.16) [51]. Surgical correction was performed using a uniform technique for bilateral subperichondrial resection of the deformed costal cartilages and sternal osteotomy

Fig. 1.11 Fritz Rehbein (1911–1991) was the Chief Surgeon in Klinikum Bremen-Mitte, Bremen, Germany from 1951 to 1976 (Courtesy of Prof. Christian Lorenz, Department of Pediatric Surgery and Urology, Klinikum Bremen-Mitte GmbH, Bremen, Germany)

Fig. 1.12 Gerd Hegemann (1912–1999) at the Friedrich-Alexander-Universität Erlangen-Nürnberg, Germany

resecting a wedge of the anterior cortex and fracturing the posterior cortex. Anterior displacement was maintained with silk sutures closing the osteotomy defect. In 28 early cases, the sternum was secured by intramedullary fixation with a Steinman pin.

The age at which open repair were to be offered to children was influenced by 2 important publications. Initially, Hougaard and Arendrup in 1983 reported on female breast deformities after surgery for funnel chest in young girls who did not show demarcation of their breast which were incised at the time of surgery [52]. Later, Alexander Haller in 1996 drew attention to the risk of "Acquired Asphyxiating Chondrodystrophy" in a report in 1996 (Fig. 1.17), as a result which most surgeons ceased to offer the open pectus repair in young children, preferring to wait until after puberty. This also decreased the amount of cartilage resected in open procedures [53].

The author introduced the "Pectus Less Invsaive Extrapleural Repair" (PLIER) technique which is a open correction method with excellent outcomes reported in 2007 [54]. Among the advantages of PLIER technique are the relatively smaller size of skin incision, extrapleural repair and the selective use to struts based on the (a) severity, (b) area (presence or absence of platythorax) and (c) rigidity of the thorax (depending on the age of the patient). PLIER has been found to be an excellent technique for severe asymmetric pectus as well as for repairs failed from other techniques.

Fig. 1.13 View of the iconic University Hospital in Münster, Germany

Fig. 1.14 Amulya Saxena (left) and Günter Willital (right) evaluated and reported the largest experience of pectus repairs using the Willital-Hegemann technique

Fig. 1.15 Pediatric Surgical team in Münster (1998) that performed repairs using the standard Willital-Hegemann technique (left to right) Amulya Saxena, Andreas Kolberg-Schwerdt, Klaus Schaarschmidt, Andreas Hebestreit, Jose Morcate, Günter Willital, Christopher Becker, Jean Tsokas, Jürgen Schleef and Hermann Stenchly

Fig. 1.16 Robert Shamberger from Boston Children's Hospital reported a large series of pectus excavatum patients in 1988

Fig. 1.17 Alexander Haller John Hopkins (Printed with permission: The Alan Mason Chesney Medical Archives of The Johns Hopkins Medical Institutions, Baltimore, Maryland, USA)

Minimal Access Surgery Era of Repair

The focus of attention in pectus excavatum repair revolved primarily on the issues of achieving good long-term results and the elimination of perioperative complications. With the developing trend towards "minimal access surgery" in the 1990s, a new approach to the management of pectus excavtaum was set by Donald Nuss in 1998 (Fig. 1.18) [55]. Nuss decided to make the standard skin incision across the chest, but decided and not to remove the skin and muscles off the chest – a necessary requirement in order to gain access to the rib cartilages and sternum. He also did not remove the rib cartilages and sternum. Instead, he created a small access between the ribs, in line with the deepest point of the depression, after which a long,

Fig. 1.18 Donald Nuss in 1998 pioneered the minimal access approach at the Children's Hospital of the King's Daughters, Norfolk, Virginia, USA

curved clamp was then inserted into the chest and a tunnel under the sternum was slowly created. The tip of the clamp was then advanced out of the chest on the other side of the sternum. Surgical tape was attached to the clamp, and pulled through the substernal tunnel. The tape was then used to guide a convex titanium bar through the tunnel with the convexity facing posteriorly. Once the bar was in position, it was turned over 180° to correct the pectus excavatum.

Nuss initially used a titanium bar for the repairs, but realized the the bar proved too soft for the minimal access procedure. The initial excellent result achieved directly after the procedure was not evident six months after the procedure. Nuss then sought the modification of the bar from titanium to stainless steel. The next modification was to change the access from the long anterior chest incision to two small incisions on each side of the chest. One of the most important additions to this procedure was the introduction of thoracoscopy as it allowed visualization within the thoracic cavity, greatly decreasing the risk of the procedure. Protocols were also developed for proper patient selection, pain management and postoperative management.

Since its introduction, thousands of patients have undergone this procedure and there is an abundance of publications and presentations describing the technical details and modifications of this technique [56, 57]. Due to this extensive publicity as well as to the fact that the operation is now performed not only by thoracic surgeons, but by surgeons from other specialties as well, the number of patients operated for pectus excavatum has more than tripled in the last few years. While good early and mid-term results are observed with this method, the lack of prospective and randomized studies and the different patient characteristics have made a systematic comparison between the Nuss procedure and conventional open repair difficult.

Pectus Carinatum

Historical Background of Surgical Repair

Pectus carinatum (pigeon chest), a protrusion deformity is the second most common malformation of the anterior chest wall. Mark Ravitch in 1952 was the first to suggest that surgical correction was the only effective method for the treatment of this deformity. He surgically corrected the chondromanubrial prominence by resecting the multiple deformed costal cartilages and performing a double osteotomy [58]. Later on, Lester in 1953 performed a repair involving resection of the anterior part of the sternum, but abandoned this approach due to unsatisfactory results. He then reported a second less radical technique which involved subperiosteal resection of the lower body of the sternum and sternal ends of the costal cartilages [59]. Chin in 1957 and Brodkin in 1958 employed an operative procedure which used the traction effect of the rectus muscles to relocate and maintain the sternum in a corrected position [60, 61]. This method was further modified by Howard who preferred the radical resection of cartilages and a sternal osteotomy [62].

Later, in 1960, Ravitch reported another surgical procedure that left the sternum alone, and involved the resection of the affected costal cartilages along with the shortening of perichondrial strips with reefing sutures [63]. In 1963, Ramsay utilized a rectal muscle flap to fill the lateral defects resulting from the protrusion without altering the position of the sternum or resecting the deformed costal cartilages [64]. Also in 1963, Robicsek reported a technique that involved subperichondrial resection of the deformed lateral asymmetric costal cartilages, transverse sternal displacement, and resection of the protruding lower portion of the sternum along with the reattachment of the xiphoid and rectus muscles to the new lower margin of the sternum (Fig. 1.19) [65]. Welch in 1973 and Pickard in 1979 reported techniques which were similar and involved costal cartilage resection and sternal osteotomy [66, 67]. Horacio Abramson in 2005 initially reported on a

Fig. 1.19 Francis Robiscek at the Carolinas Heart Institute, Carolinas Medical Center, Charlotte, North Carolina, USA

Fig. 1.20 Horacio Abramson from Antonio Cetrangolo Hospital, Buenos Aires, Argentina pioneered in the minimal access surgery of pectus carinatum

minimal access surgical procedure to correct pectus carinatum followed by a 5-year follow up of his results and set the trend for minimal access surgery in pectus carinatum (Fig. 1.20) [68, 69].

Sternal Defects

These defects constitute a broad spectrum of rather rare deformities of the sternum and heart. While the specific cause is yet unexplained, a partial or complete failure of fusion is responsible the defects that occur. Cardiac pulsations are quite prominent with each type of cleft sternum, suggesting that the heart is partially outside the chest wall (ectopia cordis) in true cases. Associated cardiac defects and multiple anomalies in other body regions usually preclude survival.

In 1818 Weese provide the first anatomic classification of the defects [70]. Weese divided sternal defects into *ectopia cordis cum sterni fissure*, *ectopia suprathroracica*, *and ectopia subthoracica*. Breschet in 1826 provided a similar classification [71]. Later on Roth in 1939 divided these lesions into *ectopia cordis thoracalis extrathoracica* (manubrium defect, sternal body defects and *ectopia cordis pectoralis sternoepigastrica*) *ectopia cordis ventralis and ectropia cordis suprathoracica* [72]. Shao-tsu in 1957 divided the sternal defects into *ectopia cordis cervicalis*, *ectopia cordis thoracalis*, *ectopia cordis thoracoabdominalis*, *and ectopia cordis abdominalis* [73]. Cantrell in 1958 provided further classifications of sternal defects in which he presented patients with *ectopia subthoracica* or *l'ectopia abdominale* associated with defects of the diaphragm, pericardium, abdominal wall, and heart which was subsequently referred to as Cantrell's pentalogy [74].

Ectopia Cordis

The first attempted repair of ectopia cordis was performed in 1925 by Cutler and Wilens, however, in 1975 it was Knoop who performed the

first successful repair [74, 75]. Skin flap coverage had been since used in a two stage reconstruction for the management of thoracic ectopia cordis. However, successful one staged correction has been reported by Amato who performed a construction of the partial anterior thoracic cavity surrounding the heart, but however avoided the return of the heart to its orthotopic location [76].

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Classification of Chest Wall Deformities

2

Amulya K. Saxena

Introduction

Chest wall deformities (CWDs) encompass a wide range of anomalies which extend from the sternum to the vertebral column. Few patients with CWDs present clinically with functional respiratory impairment and noticeable symptoms, but these patients with deformities seek medical due to psychosocial concerns and their aversion to sports and public exposure. The variations and the complexities in the presentation and occurrence of CWDs, often leads to their misdiagnosis or neglect by medical care specialists, that results in inappropriate diagnosis which in turn delays the therapeutic management. Surgical repair using radical procedures and inconsistent reporting of outcomes has deterred the medical community, not directly involved in repairs, to opt for a conservative "wait and see" approach. However, with the introduction of the minimal access repair techniques as well as evolving options in conservative treatment for pliable CWDs, interest has been revived both in the scientific community and the general public with regards to treatment of the most common forms of CWDs- Pectus excavatum and Pectus carinatum and their combinations.

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The exact description of the various types of CWDs is challenging as some forms are well defined, whereas others are part of a larger spectrum of deformities or syndromes. Classification is of CWDs is important as correct diagnosis has direct implication on the treatment options. Complex classifications are difficult to understand; furthermore, since the management of CWDs involves multiple specialties a clear and concise classification for CWDs is of paramount importance to offer suitable therapy options.

Classification and Guidelines

The classification of CWDs is based on the morphological site of deformities extrending from the sternum to the vertebrae. This "Chest Wall Deformities Morphological Classification" divides CWDs in to 5 types based on the anatomic topography of the chest wall: Type I- sternum, Type 2- costal cartilage, Type 3- rib, Type 4- combined costal cartilage and rib, and Type 5- Costovertebral junction (Table 2.1).

Pectus excavtum, Pectus carinatum and their combinations represent the majority of the patients who are referred for evaluation to medical practitioners worldwide [1]. The trend with regards to these deformities (see Chap. 1) was initially associated with reporting of the deformities for over five centuries with the advent towards open surgical corrections in the 1930s. This was followed by introduction of the era of minimal

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Table 2.1 Chest wall deformities morphological classification: deformities are classified based on the anatomical topography

Classification	Deformity (anatomical topography)	Characteristic presentation
Type-1	Sternum	Cleft sternum (+/– *Ectopia Cordis*) Currarino Silvermann syndrome
Type-2	Costal cartilage	Pectus excavatum Pectus carinatum
Type-3	Rib	Simple Complex (+/– syndrome)
Type-4	Combined costal cartilage and rib	Poland's syndrome VACTER syndrome
Type-5	Costo-vertebral junction	Osteogenesis imperfecta Syndromic

access correction in the late 1990s which was further followed by the development of conservative approaches in management of pliable CWDs from 2000's using vacuum bells for pectus excavtums and compressive braces for pectus carinatum [2, 3]. Based on these advances in management, a "Guideline for management of Pectus deformities" was proposed to offer an algorithm for patients with combinations of pectus deformities (Fig. 2.1) [1]. According to the guidelines: (A) Mild forms of pectus deformities in young children warrant a "wait and watch" approach, (B) Pliable pectus deformities in school age children and adolescent are suitable for conservative treatment (Vacuum bell-Pectus excavtaum or compression brace- Pectus carinatum), (C) Symmetric (or mild asymmetric) pectus deformities could be best managed by minimal access surgical procedures, and (D) Complex pectus deformities especially those with severe forms of asymmetry, combined formed (Pectus excavtum combined with Pectus carinatum), platythorax or cranial pectus carinatum require an open surgical correction. This guideline also offer flexibility to opt for a minimal access or open surgical procedure based on the severity of the deformity as well as the expertise of the surgeon.

Type 1- Sternum Deformities

Sternal clefts have an estimated incidence of 0.15 % of all CWDs and are formed when there is a defect in the fusion of the sternum [4]. It is presumed to be an embryonic developmental disorder; with the Hoxb gene implicated in the development of this deformity [5]. A classification proposed by Shamberger and Welch in 1990 divided sternal clefts into 4 types [6]:

- ***Thoracic ectopia cordis***: In this form of sternal cleft the heart is ectopic and devoid of a skin covering. The heart is generally located in an anterior position and has intrinsic anomalies. The sternal defect itself can be located cranial, caudal, central or may be total. Thoracic ectopia cordis can also be associated with omphaloceles. Since the thoracic cavity is hypoplasic, surgical correction is generally unsuccessful.
- ***Cervical ectopia cordis***: In these rare defects the heart is located more cranially and this condition is frequently associated with craniofacial anomalies.
- ***Thoracoabdominal ectopia cordis***: In this condition the heart is covered by a thin membranous or cutaneous layer within a caudal located defect. The heart which may be located either in the thorax or in the abdomen, is not rotated as in the previously types described, but is accompanied by intrinsic anomalies. This anomaly is a part of the Pentalogy of Cantrell.
- **Sternal cleft**: This is the most common among the types of sternal deformities and arises from a deficiency in the midline embryonic fusion of the sternal plates. Sternal clefts can be partial or complete. The partial deformities could have a cranial or caudal location. The caudal forms are often associated with a thoraco-abdominal ectopia cordis, whereas the cranial partial clefts exist as isolated malformations. Although the sternoclavicular joints are displaced laterally in sternal clefts, the clavicles retain a normal length. Sternal clefts can frequently be associated with other defects (maxillofacial hemangiomas, cleft lip or cleft palate, pectus excavatum, connectival nevi, supraumbilical raphe, or gastroschisis)

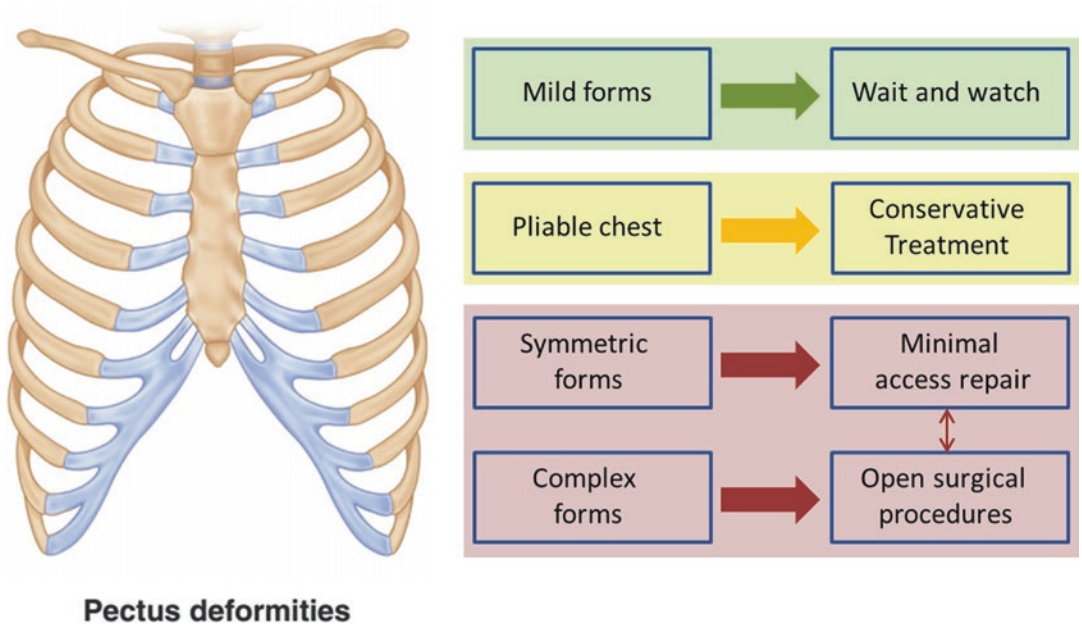


Fig. 2.1 "Guideline for management of Pectus deformities": (**a**) Mild forms in young children warrant a "wait and watch" approach, (**b**) Pliable pectus deformities in school age children and adolescent warrant conservative treatment (Vacuum bell-Pectus excavtaum or compression brace- Pectus carinatum), (**c**) Symmetric (*or mild asymmetric) pectus deformities should be offered minimal access repair options, and (**d**) Complex pectus (severe forms of asymmetry, combined forms, platythorax or cranial pectus carinatum) require an open surgical correction. Minimal access or open surgical procedure options offered are based on the expertise of the surgeon as well as the severity of the deformity

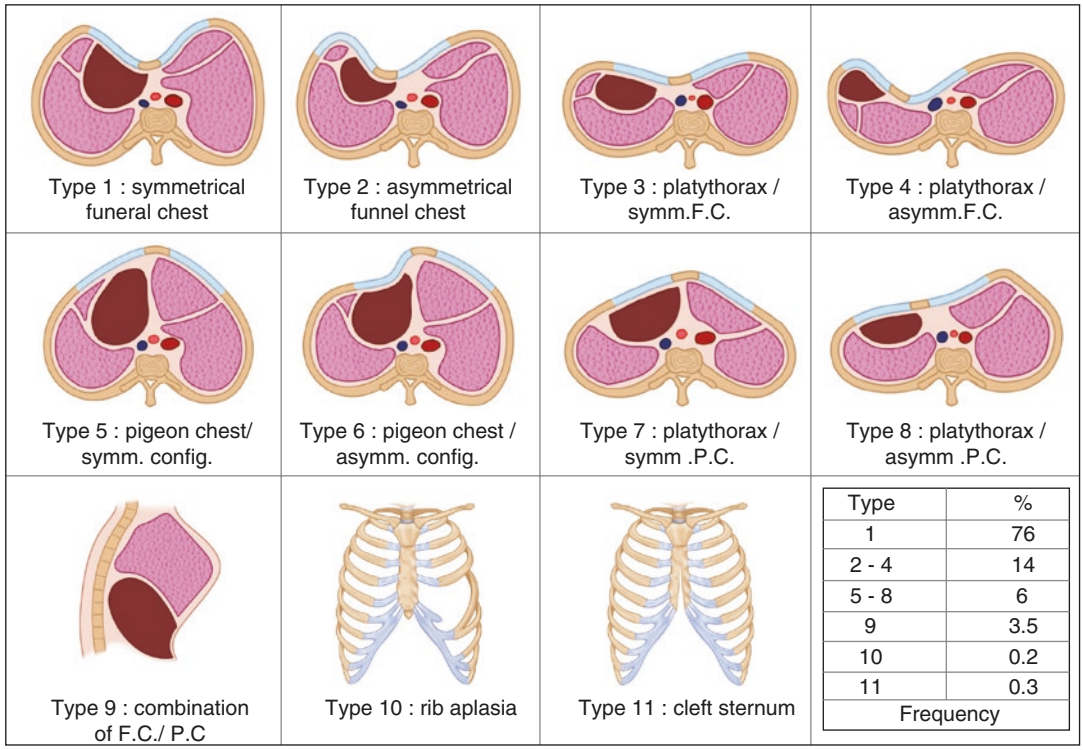
which must be carefully evaluated in the planning of any surgical procedure [7]. Other defects, such as cardiac defects, aortic coarctation, occular abnormalities, posterior fossa anomalies and obscured hemangiomas must be ruled out during evaluation of the patient.

Type 2- Costal Cartilage Deformities

Pectus deformities belong to the type of costal cartilage deformities which present a wide range of forms from pectus excavatum to pectus carinatum and their combinations. The patients are graded according to the Willital's classification, which is based on morphologic findings of the thorax and divides congenital chest wall deformities into 11 types – funnel chest (4 types), pigeon chest (4 types), combination of funnel and pigeon chest, chest wall aplasia and cleft sternum (Fig. 2.2) [8, 9] This classification allows a more accurate description and documentation of the deformities and takes into consideration the asymmetric presentation of pectus deformities along with inclusion of the rare forms.

Willital's classification of congenital chest wall deformities:

Type 1 Symmetric pectus excavatum within a normal configured thorax
Type-2 Asymmetric pectus excavatum within a normal configured thorax
Type-3 Symmetric pectus excavatum associated with platythorax
Type-4 Asymmetric pectus excavatum associated with platythorax
Type-5 Symmetric pectus carinatum within a normal configured thorax
Type-6 Asymmetric pectus carinatum within a normal congigured thorax
Type-7 Symmetric pectus carinatum associated with platythorax
Type-8 Asymmetric pectus carinatum associated with platythorax



Type	%
1	76
2 - 4	14
5 - 8	6
9	3.5
10	0.2
11	0.3
Frequency	

Fig. 2.2 Willital's classification of anterior congenital chest wall deformities [9]

Type-9 Combination of pectus excavatum and pectus carinatum
Type-10 Thoracic wall aplasia/hypoplasia
Type-11 Cleft sternum defects

- ***Pectus excavatum* (*PE*)**
  PE has an incidence of 1:100 to 1:1000 live births and accounts for 90% of the CWDs [10, 11]. It is characterized by the presence of a variably deep sternal depression due to uncontrolled growth of the lowest costal cartilages. PE is commonly congenital, but can appear during childhood or adolescence (Fig. 2.3). The etiology of PE is not clear with regards to the exact nature of the associated connective tissue disorders. Genetics studies on PE have shown the presence of an autosomal dominant transmission pattern, but autosomal recessive and X-linked patterns have also been associated with its development [12]. The overgrowth of costal cartilages has been presumed as the pathogenic mechanism leading to the development of PE [10, 13]. Collagen type II disorders have been demonstrated in the costal cartilages in PE [14], as well as overexpression or down regulation of some genes playing a role in the metabolism of cartilage and connective tissues, as collagen genes, matrix metalloproteinases, tumor necrosis factor-alpha, and filamin have also been implicated [10].

  PE presents with various degrees of severity which is important in assessment of the condition (Figs. 2.4 and 2.5). Simple patient evaluation can be performed by measuring the depth of the deformity using a caliper (Fig. 2.6). Three areas of reference are measured between the sternum and the vertebral column: A-Manibrium-Vertebral distance, B-Corpus-Vertebral distance and C-Xiphoid-Vertebral distance (Fig. 2.7). These distances provide information with regards to the level of the length of the sternum in relation to the

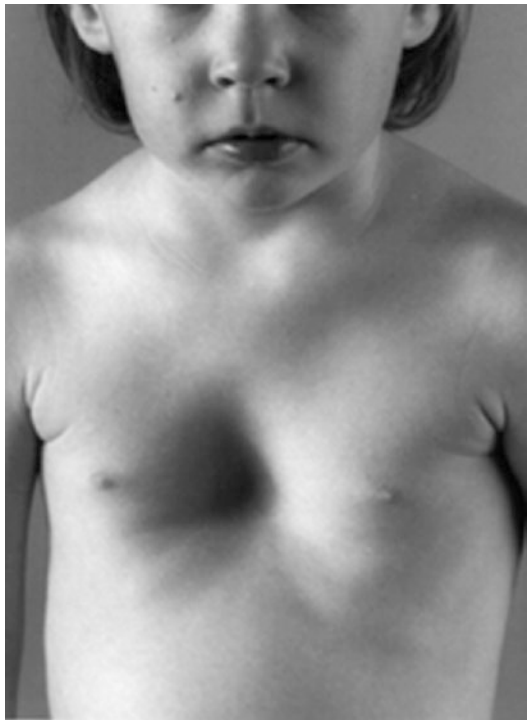


Fig. 2.3 Pectus excavatum in a 2-year old girl presenting with a symmetric deformity

Fig. 2.4 Asymmetric pectus excavtum in a 40-year old male patient with sternal rotation

vertebral column. Using a flexible ruler the contours of the thorax can be estimated (Fig. 2.8). These are then plotted at the point of maximum deformity to determine the variations in depth of the chest wall with regards to geometric proportions (Fig. 2.9).

However, Computerized Tomography (CT) scan are necessary to access the severity of the deformity and to calculate the Haller index to estimate the severity of chest depression in patients with PE (Fig. 2.10) (Haller index is the ratio of the transverse diameter and the anteroposterior diameter of the rib-cage. Haller index in a normal chest is around 2.5, but in pectus excavatum the index can reach 3.25 or even as high as 5.5) [15]. Another important feature in assessment is the symmetry. Asymmetric PE present with variable degrees of sternal rotation which are important to recognize and correct during repairs. Three-dimensional reconstruction images from CT can offer excellent images of the bony thorax with regards to the estimation of rib cage asymmetry and sternal rotation (Fig. 2.11). Another method to map the surface contours of the chest wall is the videostereoraster examination (Fig. 2.12). This examination can be performed preoperative and during follow-ups to compare the results of surgical repair.

- ***Pectus carinatum*** **(*PC*)**
  PC is less frequent than PE and has incidence of approximately 10–15 % [10, 16, 17] with a male predominance (Fig. 2.13). However, in some geographical areas of the world, PC is almost equal or more prevalent than PE [3, 18, 19]. The etiology of PC as in PE is also unknown, but the pathogenesis could be similar to PE which involves in an overgrowth of the costal cartilage [13]. Familial cases are not uncommon and cases of PC and PE within the same family have been reported [3, 10].

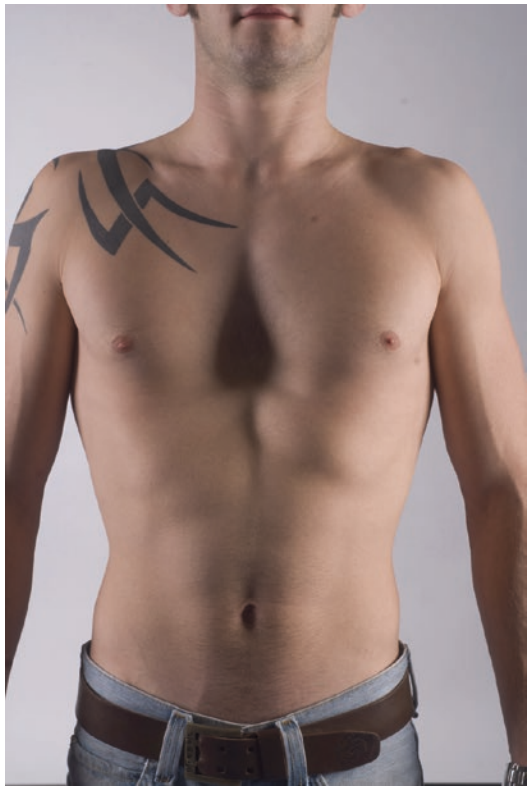


Fig. 2.5 Symmetric pectus excavatum in a 24-year old male patient

Connective tissue disorders and cardiac anomalies are rarely associated with PC [20]. Although PC usually appears later in life than PE, mainly during puberty, this deformity has also been observed in infants and children. With regards to estimation of the contours of the deformity, investigation that were performed in patients for PE- flexible rule and caliper measurement as well as CT can be performed for preoperative analysis (Fig. 2.14).

PC is been classified into 2 types according to the localization into the following types [17, 21]:

– ***Type 1- Chondrogladiolar***: The symmetric sternal protrusion is located in the inferior or mid sternum region. The lower ribs may be slightly or severely depressed on lateral side.
– ***Type 2-Chondromanubrial***: These have been frequently referred to as Currarino-Silverman syndrome or Pouter Pigeon Breast. Currarino-Silverman syndrome is a rare disorder characterized by premature fusion of manubrio-sternal joint and the sternal segments, resulting in a high carinate chest deformity; it is frequently associated with congenital heart disease [22]. However, chondromanubrial forms of PC are frequently isolated findings in patients (without cardiac association).

Reactive Pectus Carinatum This is observed in some patients who have undergone PE repairs. The occurrence of reactive PC which is evident after the first few months or years of minimal access as well as open repair of PE, and presents as a progressive ventral displacement of the sternum [23]. It is more frequent in patients with connective tissue disorders.

Type 3- Rib Deformities

Based on the radiological overview on pediatric rib deformities presented in 2002, the deformities in ribs are based on their origin- congenital or acquired [24]. Since the overview presents all radiological conditions that affect ribs, the following section refers to those rib deformities that are associated with CWD's. Table 2.2 summarizes the list of rib deformities that form the spectrum of CWDs (Table 2.2).

Congenital Deformities

1. ***Cervical ribs***: Cervical ribs arise from the seventh cervical vertebra. They are most commonly an incidental finding. Cervical ribs are rarely symptomatic in early childhood; but in older children and adults, compression of the brachial plexus or subclavian artery can give rise to the formation of a thoracic outlet syndrome [25].
2. **Altered rib count**: An alteration in the number of ribs has been found in trisomy 21 syndrome and in patients with the VATER association (Fig. 2.15) [26, 27]. It is not uncommon to see 11 pairs of ribs in the

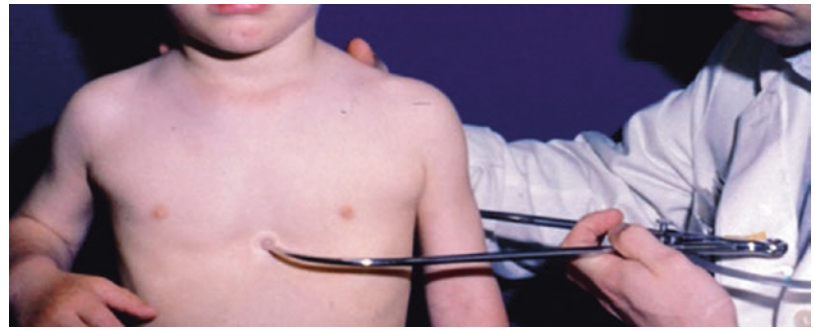


Fig. 2.6 Measurement of sterno-vertebral distances using a caliper

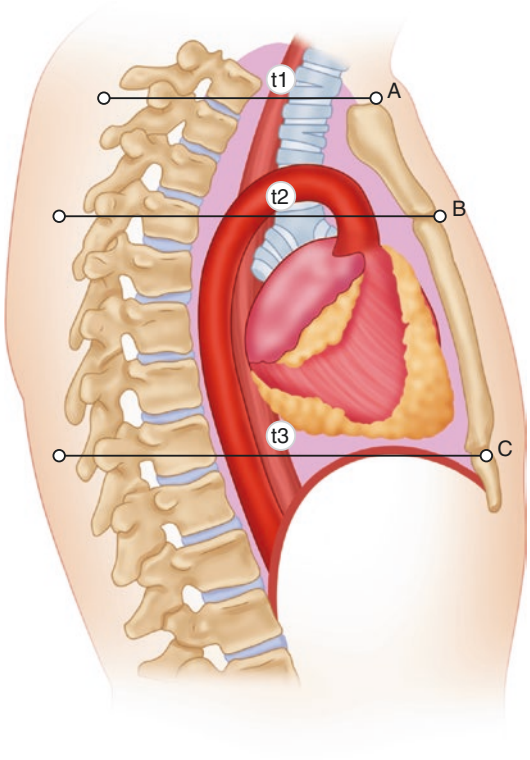


Fig. 2.7 Three areas of reference are measured between the sternum and the vertebral column using the caliper: A-Manibrium-Vertebral distance, B-Corpus-Vertebral distance and C-Xiphoid-Vertebral distance

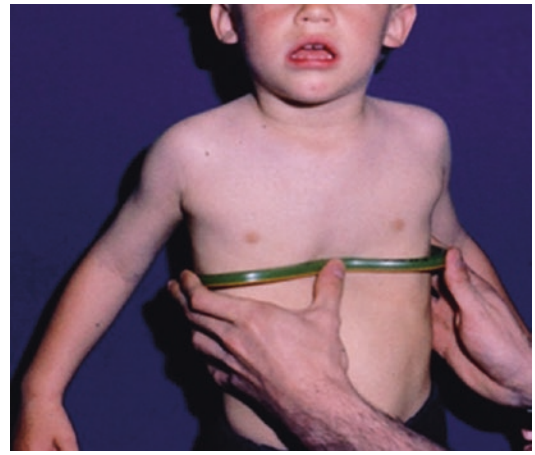


Fig. 2.8 The flexible ruler is placed anteriorly and then posteriorly at the point of maximum deformity to obtain the contours of the thorax

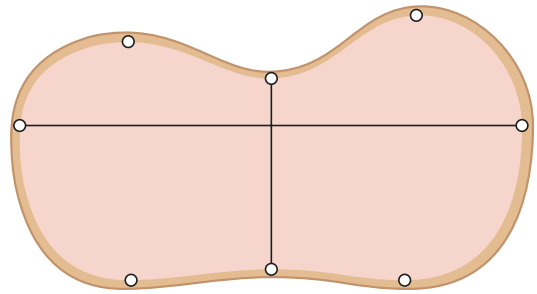


Fig. 2.9 The contours of the thorax obtained flexible ruler are plotted at the point of maximum deformity to determine the variations in depth of the chest wall

absence of associated anomalies; this situation occurs in 5–8 % of normal individuals. Eleven pairs of ribs occur in one-third of patients with trisomy 21 syndrome [27], as well as in association with cleidocranial dysplasia and campomelic dysplasia.

3. **Rib size variations**: Short ribs constitute an integral part of several syndromes. Due to their size, short ribs do not extend as far anteriorly to reach and connect with the sternum. This results in diminished volume of the chest

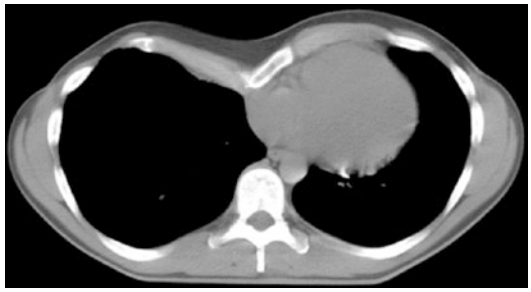


Fig. 2.10 Computed tomography scan of the thorax in a 14-year old patient showing the contours of a symmetric pectus excavtum with a rotation of the sternum

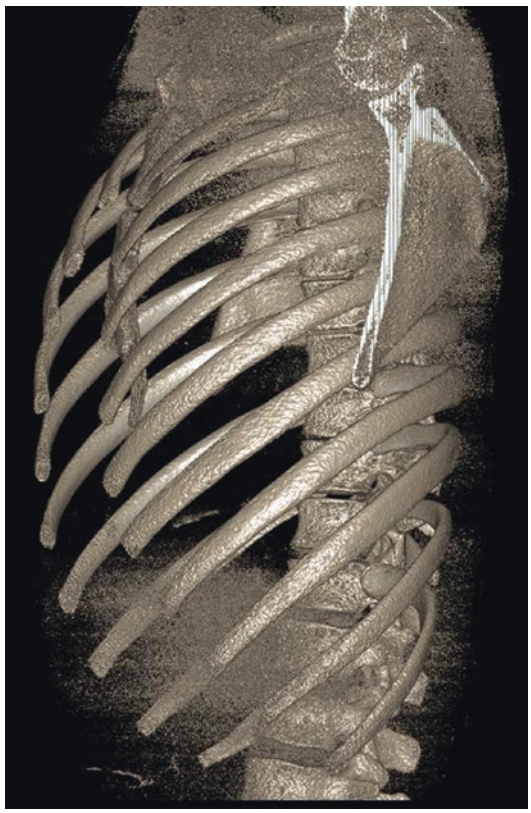


Fig. 2.11 Three-dimensional reconstructed CT images demonstrating sternal depression in relation to the rib cage in a 15-year old pectus excavtum patient

and restricts respiratory motion that further leads to respiratory insufficiency. Definitive diagnosis should be performed early in the neonatal period to exclude a lethal dysplasia so that a resuscitation policy can be planned.

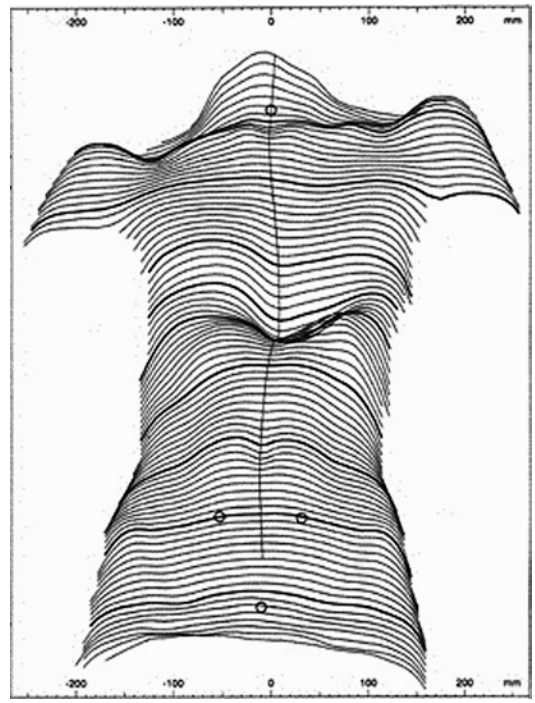


Fig. 2.12 Videoraster examination of a 16-year old male patient with slight asymmetric pectus excavtum deformity

Other conditions with rib size variations are the short rib-polydactyly syndromes (Saldino-Noonan, Majewski, and Verma-Naumoff); chest diameter is critically small in these syndromes.

(a) ***Thanatophoric Dysplasia*** —Thanatophoric dysplasia is a lethal dysplasia transmitted by a dominant gene mutation and is the most common lethal neonatal skeletal dysplasia after osteogenesis imperfecta type II. The ribs in these patients are very short and do not extend beyond the anterior axillary line. Associated findings are cloverleaf skull deformity, polydactyly, and hypoplastic iliac bones. Other conditions with these deformities include thanatophoric variants, homozygous achondroplasia, achondrogenesis and asphyxiating thoracic dysplasia.

(b) ***Jeune Asphyxiating Thoracic Dysplasia*** —Jeune asphyxiating thoracic dysplasia

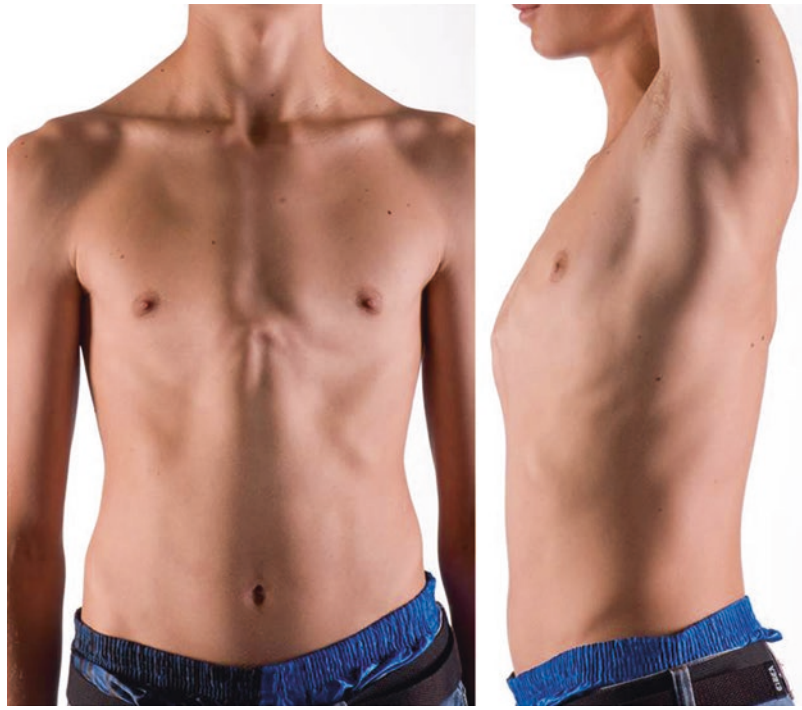


Fig. 2.13 Pectus carinatum deformity in a 16-year old male patient showing the protrusion of the sternum- frontal view (*left*) and lateral view (*right*)

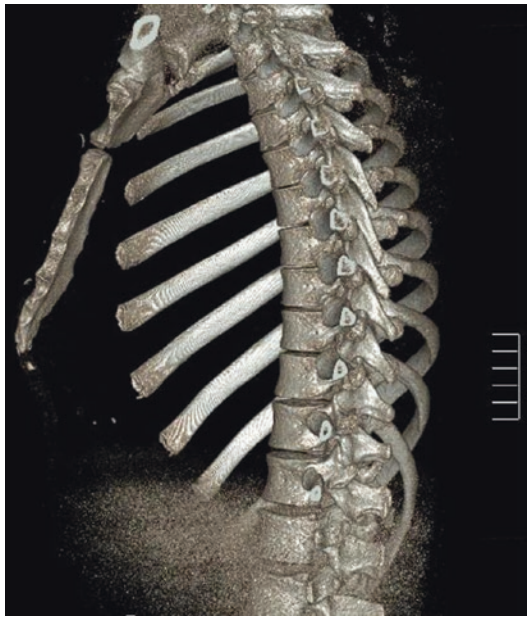


Fig. 2.14 Three-dimensional reconstructed CT images demonstrating sternal protrusion in relation to the rib cage in a pectus carinatum patient

is an autosomal recessive dysplasia [28]. The ribs are short, with a horizontal course, and the chest diameter is significantly decreased compared with that of the abdomen. Although the findings of the ribs on the chest radiograph may be suggestive of Jeune dysplasia, their appearance also resembles that of Ellis-van Creveld dysplasia. Children who do not have a severe affection and reach adulthood, are reported to have an increased incidence of medullary cystic renal disease [29].

(c) ***Ellis-van Creveld Chondroectodermal Dysplasia*** —Ellis-van Creveld chondroectodermal dysplasia is an autosomal recessive dysplasia in which the ribs are short, and the chest cavity is narrow. The narrow thorax amplifies the size of the heart; cardiomegaly is often present because of associated anomalies. Acromelic limb shortening, polydactyly,

Table 2.2 Rib deformities that cause chest wall deformitied are classified as congenital or acquired

Congenital deformities	Acquired deformities
Cervical ribs	**Metabolic disorders** Rickets Hyperparathyroidism
Altered rib count	**Iatrogenic causes**
Rib size variations Thanatophoric dysplasia Jeune Asphyxiating Thoracic Dysplasia Ellis-van Creveld Chondroectodermal dysplasia Achondroplasia	**Trauma** Child abuse Surgical trauma
Abnormal rib morphology Cerebrocostomandibular syndrome Rib notching Bifid ribs Slender ribs Trisomy 18 syndrome Neurofibromatosis	**Neoplastic disorders** Enchondromatosis Osteochondroma Langerhans cell histiocytosis Xanthogranuloma Ewing Sarcoma Primitive Neuroectodermal Tumor
	Infections

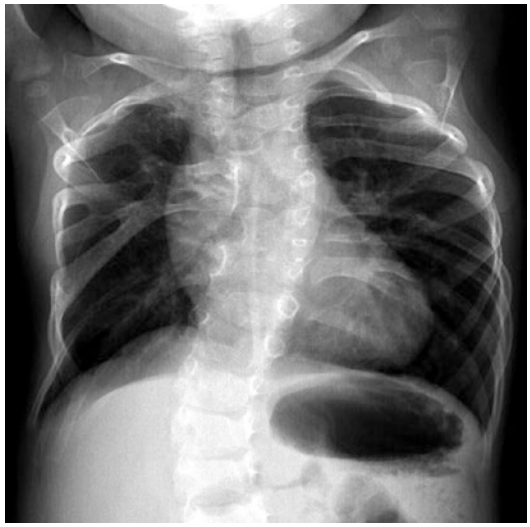


Fig. 2.15 Chest film showing alteration in the number of ribs and dyplasia of the right thoracic cage in a 2-year old with VATER association

and cone epiphyses are characteristic. The proximal humeral and femoral epiphyses may be prematurely ossified at birth. Diagnostic work up is used to rule out Jeune dysplasia and the short rib-polydactyly syndromes.

(d) ***Achondroplasia*** — In achondroplasia, the ribs are short and wide with concavity at the end of the ribs. The inheritance is autosomal dominant, with spontaneous mutations in 80 % of cases. Associated diagnostic features include macrocephaly, depression of the nasion, craniocaudal narrowing of the interpediculate spaces, square iliac bones, and a narrow pelvic inlet.

4. **Abnormal rib morphology**

(a) ***Cerebrocostomandibular Syndrome***.—Cerebrocostomandibular syndrome is very rare and the genetic transmission is not yet established [26]. These patients have eleven pairs of ribs which are characterized by abnormal costovertebral articulations and posterior ossification gaps that bear similarities to fractures. The posterior ossification gaps ossify in the adult age. Respiratory distress is common in neonates due to flail chest and airway abnormalities. Other features of this syndrome are microcephaly, micrognathia, and congenital heart disease.

(b) ***Rib notching***.—Concave notches are found on the inferior rib surface and are generally considered normal. Pathologic notching is vascular or neural in origin. Notching associated with aortic coarctation usually affects ribs 4–8 and is rare in affected children before the age of 8 years [30]. Rib notching is also encountered in neurofibromatosis and thalassemia.

Notching is unilateral, and on the right sided, with coarctation proximal to the origin of the left subclavian artery and a postoperative Blalock-Taussig shunt.

(c) **Bifid ribs**: Gorlin basal cell nevus syndrome is a rare, autosomal dominant syndrome characterized by multiple nevoid basal cell carcinomas that arise in childhood jaw cysts and bifid ribs. The fourth rib is the most commonly bifid. A single bifid rib is most commonly a normal incidental finding and may be detected as a palpable chest wall mass. Other rib anomalies include agenesis, supernumerary ribs, distorted shape, and fusion of adjacent ribs [31]. The lateral clavicle may be deficient. This condition is associated with features such as mandibular hypoplasia, macrocephaly, and mental retardation.

(d) **Slender Ribs**

- ***Trisomy 18 Syndrome*** — In this syndrome, eleven rib pairs are present, and they are classically hypoplastic and thin. The short sternum in these patients presents as a typical shield deformity of the chest. These children are spastic and neurologically delayed. The fingers are maintained in ulnar deviation with flexion deformities. Associated findings are dolichocephaly, micrognathia, and various multisystem anomalies. Other conditions that can cause such changes include Cockayne syndrome, trisomy 13 syndrome, Werdnig-Hoffmann disease and osteogenesis imperfecta.
- ***Neurofibromatosis***.—Peripheral neural tumors are the characteristics of neurofibromatosis, an autosomal dominant neurocutaneous syndrome. The ribs are slender; twisted, deformed, and separated by neurofibromas that arise from the intercostal nerves [32]. The mesenchymal abnormality in neurofibromatosis results in modeling deformity and even stranger appearance of the ribs. Intercostal neuromas can cause notching of the inferior surface of the ribs. Sphenoid dysplasia, modeling deformity of the long bones, and posterior vertebral body scalloping are associated findings. Cleidocranial dysplasia, myotonic dystrophy, Melnick-Needles osteodysplasty, and Werdnig-Hoffmann disease should also be considered when diagnosing these patienst.

Acquired Deformities

Acquired deformities can be grouped under those resulting from metabolic disorders, infections, neoplasms, trauma and iatrogenic causes.

1. **Metabolic Disorders**
   - ***Rickets***— Rickets in present times is more common with very low-birth-weight prematurity and secondary to renal or hepatic dysfunction and anticonvulsant administration. Delayed ossification of the osteoid matrix in immature bones is marked in the metaphyses around the knees, wrists and the rib ends. The hyperplastic rib osteoid may be palpable on the chest wall. Other differential diagnostic considerations for rib cupping and flaring include achondroplasia, hypophosphatasia, and metaphyseal chondrodysplasia.
   - ***Hyperparathyroidism***.—Hyperparathyroidism occurs most commonly secondary to chronic renal failure. Hyperparathyroidism results in subperiosteal and endosteal bone resorption. Brown tumors are a rare manifestation of hyperparathyroidism, in which well-defined lucent areas of lysis occur without adjacent reactive bone formation. The other differential diagnostic considerations are Langerhans cell histiocytosis, enchondromas and fibrous dysplasia.
2. **Iatrogenic Conditions**
   Generalized new bone formation occurs after prostaglandin administration in children with cyanotic heart disease. The formation has been reported to be as early as 6 days but is more evident after 30–40 days therapy. The changes are usually more symmetrical than those of infantile cortical hyperostosis (Caffey

disease or syndrome). After treatment, periosteal new bone becomes incorporated and remodeled [33]. Differential diagnostic possibilities include child abuse, extracorporeal membrane oxygenation, and infantile cortical hyperostosis.

3. **Trauma**
   - ***Child Abuse***.—Fractures of the ribs with child abuse are considered highly specific for child abuse [34]. Although abuse fractures can occur in any part of the ribs, they are more common in the posterior part because of the significant posterior levering force inflicted in the abuse situation. Healing fractures are more easily detectable than in the acute phase. The most important differential diagnoses to consider are osteogenesis imperfecta, long-term ventilator therapy in prematurity, birth trauma, and Menkes syndrome.
   - ***Surgical Changes***—Rib resections and deformities that result from thoracotomy are asymptomatic. However, fusion of the ribs that can lead to scoliosis and restrict the chest wall expansion may require surgical intervention. Fused ribs are also seen in patients with Gorlin syndrome.
4. **Infections**

   Rib osteomyelitis is most commonly acquired from an infectious source with the thoracic cavity, such as empyema or pneumonia. Although hematogenous spread is rare, but when present it manifests anteriorly near the costochondral junction or posteriorly near the rib angle [35]. The transformation resembles that of typical chronic long bone infection, in which there is formation of a sequestrum in the presence of periosteal fresh bone formation.
5. **Neoplasms**

   The incidence of primary neoplasms originating in the thoracic wall is 5–10 % of all bone tumors [36]. Large tumors extend both towards the inside and outside of the thorax; with the soft-tissue mass that extends externally being clinically obvious. As a result, primary rib tumors are significantly larger than bone tumors in other locations [37].
   - ***Enchondromatosis*** — These are characterized by the presence of medullary cartilaginous bone tumors, which are further subdivided by the presence of hemangiomas (Maffucci syndrome) or their absence (Ollier disease). Rib lesions are common in enchondromatosis. Enchondromas occur in association with a variety of systemic malignancies and are themselves associated with a 25 % prevalence of malignant degeneration [38, 39]. Differential diagnostic considerations are Langerhans cell histiocytosis and osteomyelitis.
   - ***Osteochondroma***.—Exostoses are benign bony masses with a cartilage cover. These can be solitary or associated with multiple hereditary exostoses. It has been estimated that rib osteochondromas arise in almost 50 % of patients with multiple hereditary exostoses [40]. Those rib exostoses that project externally are palpable on the chest wall. Internal exostoses can be asymptomatic [41]. Osteochondromas can mimic pulmonary nodules; calcified osteochondromas resemble granulomas.
   - ***Langerhans Cell Histiocytosis***.—Langerhans cell histiocytosis is presumed to be associated with immune dysfunction. Although most bone lesions are asymptomatic, some may present as a painful soft-tissue mass [42]. The ribs are commonly involved, and multiple bone involvement is not unusual. Differential diagnostic considerations include metastases, hyperparathyroidism (brown tumors), Ewing sarcoma, Askin tumor and lymphoma.
   - ***Xanthogranuloma***.—Xanthogranulomas are rare benign tumors that occur in the ribs and other flat bones. They are invariably solitary lesions and are almost twice as common in male patients [43]. The tumor has an excellent prognosis, and complete or partial removal is curative. The differential diagnosis includes aneurysmal bone cyst, hyperparathyroidism (brown tumor), and Langerhans cell histiocytosis.
   - ***Ewing Sarcoma***.—Ewing sarcoma is an extremely aggressive malignant tumor that occurs in adolescents and young adults. It is the most common malignant tumor that affects the ribs of children and adolescents [37]. Most

Ewing sarcomas have an associated soft-tissue mass that is significantly larger than the intraosseous tumor [44]. The differential diagnosis includes osteomyelitis; Langerhans cell histiocytosis, osteosarcoma, primitive neuroectodermal tumor, metastatic neuroblastoma, and lymphoma.

- ***Primitive Neuroectodermal Tumor***.— Primitive neuroectodermal tumor or Askin tumor is a rare malignant tumor that arises from the chest wall (Fig. 2.16). This tumor very closely resembles Ewing sarcoma, and differentiation depends on detection of neurosecretory granules at electron microscopy [45]. Rib destruction occurs in over 1/4th of the patients. Differential diagnostic considerations include osteomyelitis, Langerhans cell histocytosis, osteosarcoma, Ewing sarcoma, metastasis, and lymphoma.

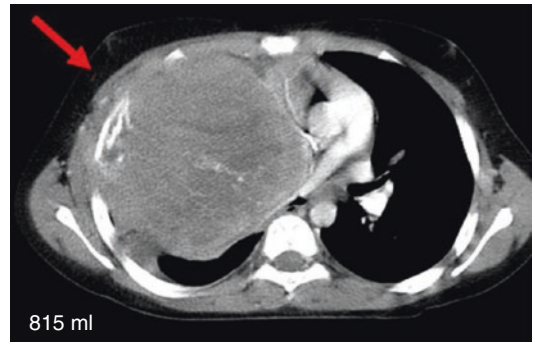


Fig. 2.16 Computed tomography image of the chest demonstrating the size and location (*red arrow*) of the Askin's tumor in a13-year old female patient

Type 4- Combined Costal Cartilage and Rib Deformities

Poland Syndrome (PS)

PS has an incidence of approximately 1:30,000 live births and is characterized by the absence or hypoplasia of the pectoralis major muscle, frequently combined with other ipsilateral abnormalities of the chest wall, breast and upper limb [46]. The defect is generally unilateral and in two thirds of cases right-sided. Very rare bilateral cases have been described [47, 48]. There is a male preponderance with a ratio of about 2:1 (Figs. 2.17 and 2.18). The etiology is unknown, but the most plausible hypothesis is the interruption of the vascular supply in subclavian and vertebral artery during embryonic life, leading to different malformations in the corresponding segments affected [49]. Beside this hypothesis, paradominant inheritance or the presence of a lethal gene survival by mosaicism have also been proposed to explain the genetic etiology of this anomaly [50, 51]. PS is usually sporadic, but the occurrence of familial cases has raised the theory of a possible transmission with an autosomal dominant pattern. Association of PS with other anomalies, as Moebius, Klippel Feil syndromes and Sprengel anomaly, has been reported [49, 52–54].

The thoracic defect is usually evident at birth in PS, but it can remain undiagnosed until the child gets older. The pectoral muscle deficiency causes an asymmetric aspect; but if there are costal anomalies associated the defect, these are more evident. In case of rib agenesis, particularly if multiple ribs are involved (generally the third and the forth rib), lung herniation and paradoxical respiratory movements are always present. Ribs can also be smaller or anomalous in patients with PS.

Anomalies like a PE or PC or both can occur concomitantly with PS, but in less than 10% of cases they require surgery. The breast region and nipple are frequently involved in these patients, which can range from a mild degree of breast hypoplasia to a complete absence of mammary gland. Associated cardiac and renal anomalies, as well as scoliosis, have been reported, but they are uncommon [53]. Dextroposition is reported frequently and is almost always associated with left PS. Patients with PS are asymptomatic, and there are have usually no limitations from the muscle defects.

Type 5- Costoverteberal Junction Deformities

- **Osteogenesis imperfecta**
  Osteogenesis imperfecta (OI) is a group of genetic disorders that mainly affect the bones [55]. There are at least eight recognized forms

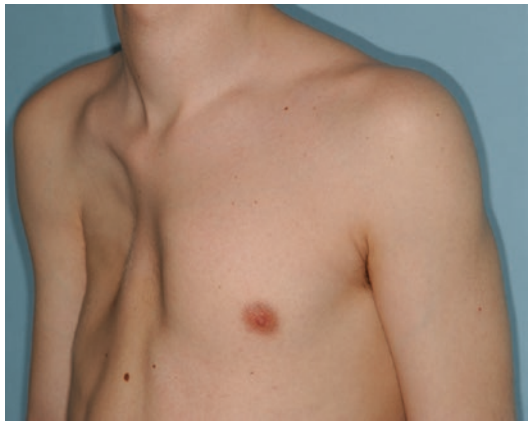


Fig. 2.17 Poland's syndrome with hypoplastic left chest wall in a 14-year old male patient

of osteogenesis imperfecta, designated type I through type VIII. The types can be distinguished by their signs and symptoms, although their characteristic features overlap. Type I is the mildest form of osteogenesis imperfecta and type II is the most severe; other types of this condition have signs and symptoms that fall somewhere between these two extremes. Increasingly, genetic factors are used to define the different forms of osteogenesis imperfecta. OI type II, can include an abnormally small, fragile rib cage and underdeveloped lungs. Infants with these abnormalities have life-threatening problems with breathing and often die shortly after birth.

- **Jarcho-Levin syndrome**
  Jarcho-Levin syndrome is a rare genetic disorder characterized by malformations of bones of the vertebrae and the ribs, respiratory insufficiency, and/or other abnormalities. Jarcho-Levin syndrome is a type of segmental costovertebral malformation [56]. Infants born with this syndrome have short necks, restricted neck motion due to abnormalities of the cervical vertebrae and short stature. In most cases, infants with Jarcho-Levin syndrome experience respiratory insufficiency and are prone to repeated respiratory infections that result in life-threatening complications. The vertebrae are fused and the ribs fail to develop properly, therefore, the chest cavity is too small to accommodate the growing lungs. There are two forms of Jarcho-Levin Syndrome that are inherited as autosomal recessive genetic traits and termed spondylocostal dysostosis type 1 (SCDO1) and spondylocostal dyostosis type 2 (SCDO2).

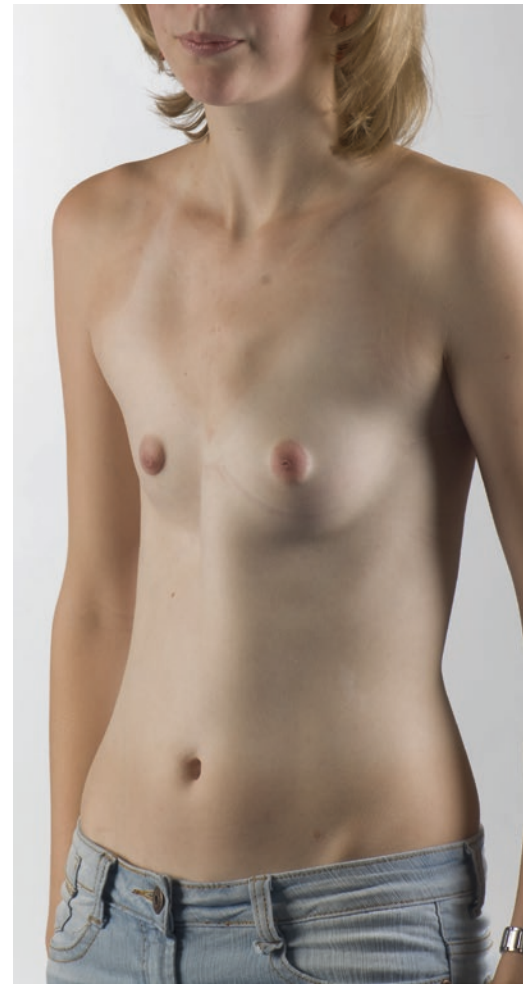


Fig. 2.18 Poland's syndrome with hypoplastic left chest wall with breast involvement in a 15-year old female patient

- **Aicardi syndrome**
  Aicardi syndrome is a rare genetic disorder first described by a French Neurologist Jean Aicardi in 1965. Aicardi syndrome is characterized by the following associations: (a) absence of the corpus callosum, either partial or complete, (b) infantile spasms, (c) lesions or "lacunae" of the retina of the eye

(lacunar chorioretinopathy), (d) other types of defects of the brain such as microcephaly, or porencephalic cysts and (e) costovertebral anomalies [57]. Aicardi syndrome only affects females, and in very rare cases, males with Klinefelter syndrome (XXY). Children are most commonly identified with Aicardi Syndrome around the age of 5 months. A significant number of these patients are girls who seem to be developing normally until around the age of 3 months, when infantile spasms commence. The known age range of affected children is from birth to the late forties.

- **Klippel-Feil Syndrome**
  Kippel-Feil Syndrome is a rare disease, initially reported in 1912 by Maurice Klippel and André Feil [58] characterized by the congenital fusion of any 2 of the 7 cervical vertebrae. The syndrome occurs in a heterogeneous group of patients unified only by the presence of a congenital defect in the formation or segmentation of the cervical spine. Klippel-Feil syndrome has following characteristics (a) scoliosis (b) spina bifida, (c) anomalies of the kidneys and ribs, (d) cleft palate (e) heart malformations (f) short stature. The disorder also may be associated with abnormalities of the head and face, skeleton, sex organs, muscles, brain and spinal cord, arms, legs, fingers and heart defects. These heart defects often lead to a shortened life expectancy, the average being 35–45 years of age among males and 40–50 among women. This condition is similar to the heart failure seen in gigantism.

Summary

CWDs encompass a wide range of anomalies involving the musculosketal system of the thorax. Enormous syndromes are associated with these deformities, detailed information on which is beyond the scope of this chapter. The classification of CWDs based on anatomic topography encompasses the spectrum of CWDs in a methodical way with attention given to each structure comprising the chest wall. Correct identification of the type of deformity according to its classification, an accurate diagnostic assessment and selection of the proper management option among those present, are the key features in the modern day approach to the treatment of CWDs.

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Surgical Anatomy of the Chest Wall

3

Atulya K. Saxena and Yasen Fayez Alalayet

The Chest Wall

The chest wall or thoracic wall is part of the thorax, which is the upper part of the torso extending from the neck to the abdomen. It is shaped like a truncated ovoid dome on anterior, posterior or lateral views and has a reniform shape on viewing its transverse section (Fig. 3.1). However, for the first 2 years from birth, when breathing is abdominal (diaphragmatic), the transverse section is more circular in shape. The thoracic wall consists of the osseo-cartilaginous throacic cage, the interconnecting muscles, the muscles on top, the fascia, the nerves and vasculature, the subcutaneous tissue, the skin, and the mammary glands that lie within the subcutaneous tissue. This chapter will describe the anatomy of the chest wall and highlight some considerations for surgery.

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Sternum

The sternum or breast bone is the central point of the osseo-cartilaginous throacic cage (Fig. 3.2). It is a long flat bone located in the middle of the anterior of the thoracic cage and consists of three parts: the manubrium, the body and the xiphoid process.

The manubrium, a trapezoid shaped bone with slightly concave surfaces, is located roughly at the level of the 3rd and 4th thoracic vertebrae. The middle of the superior surface, also known as the jugular notch or suprasternal notch, is easily palpable and flanked on either side by clavicular notches for the sternoclavicular joints. The sternoclavicular joint is an arthroidal, saddle type of synovial joint. The lateral surfaces provide depressions, above for the 1st costal cartilage, providing a synarthrosis (synchondrosis) and below a small articular facet for the upper end of the 2nd costal cartilage for an arthroidal joint, which is strong but flexible. The inferior surface is covered with cartilage for a Synarthrosis articulation with the body of the sternum, which lies in a different plane, projecting the sternal angle (of Louis), with the manubriosternal joint.

The body of the sternum is long and narrow. It lies roughly at the level of the 5th to the 9th vertebrae. The body of the sternum is composed of four primordial segments or the sternebrae which articulate with each other at primary cartilaginous joints (sternal synchondroses) and

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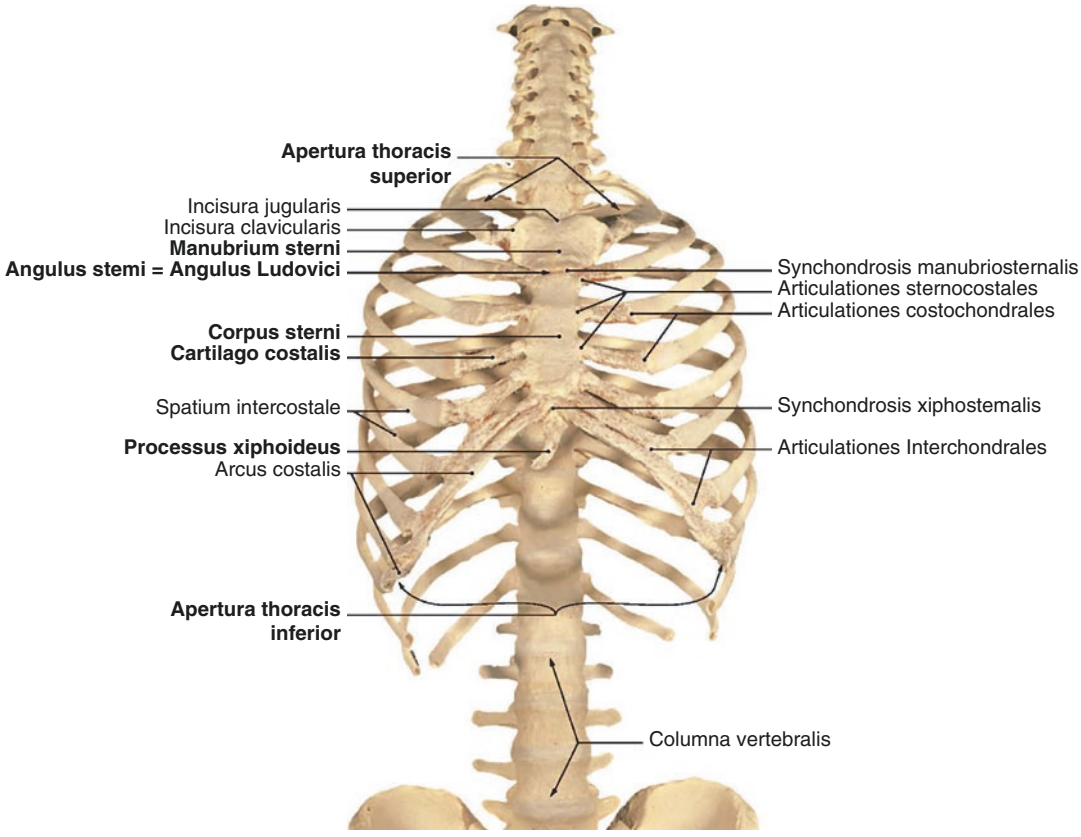

Fig. 3.1 The thoracic skeleton (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

fuse starting from the inferior joint moving upwards between puberty and the 25th year; leaving behind transverse ridges representing lines of fusion (synostosis). Its superior surface articulates with the manubrium and the superior angle with the 2nd costal cartilage. Along the lateral surfaces 4 costal notches are found from the 3rd to the 6th rib and the inferior angle articulates with the 7th rib cartilage. The inferior surface is narrow and provides for a synarthrosis (synchondrosis) articulation with the xiphoid.

The xiphiod is small, thin and elongated. It lies at the level of the 10th vertebrae. It may be bifid, curved or deflected to a side. It is cartilaginous and starts ossifying by the 40th year. The superior angle, along with the body of the sternum articulates with the 7th costal cartilage. The superior surface articulates with the body of the sternum at the xiphisternal joint and produces the infrasternal or subcostal angle of the inferior thoracic aperture. It also demarcates the inferior limit of the heart, the central tendon of the diaphragm and the superior limit of the liver,

Surgical Considerations

Pigeon Chest (pectus carinatum) occurs when the sternum projects forward and downward.

Funnel Chest (pectus excavatum) occurs when the sternum is pushed back into the thoracic cage.

Congenital anomalies are the most common forms of deformities found and are usually associated with overgrowth of the ribs. Slight deformities along with asymmetries of the thorax are common, however these do not restrain normal functioning in most cases.

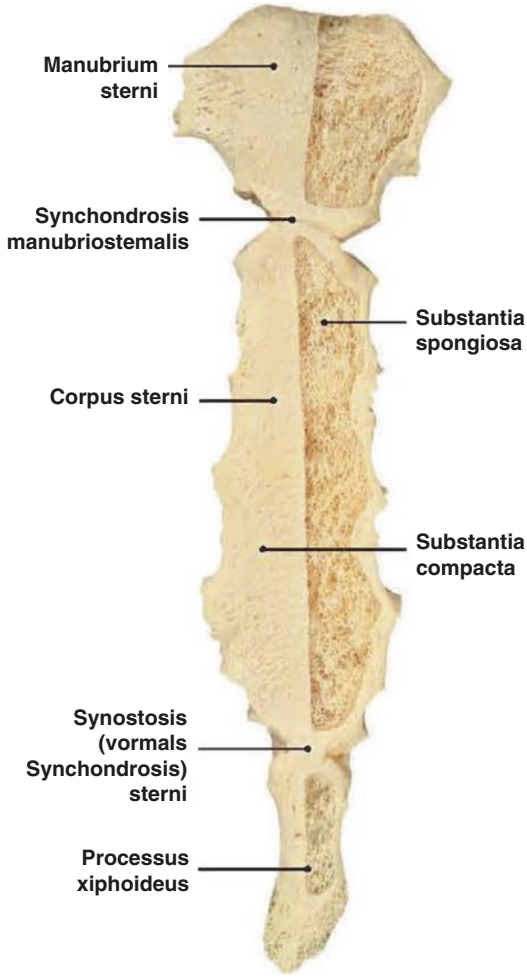

Fig. 3.2 The sternum (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

Dislocation of the sternoclavicular joint is much less frequent than the acromioclavicular dislocation. Sternoclavicular dislocations occur in violent falls or blow to the shoulder. Usually in these dislocations the ligaments are ruptured but the intra-articular fibrocartilage remains attached.

Ribs

The thoracic cage is mostly made up of flat curved bones, with a spongy interior containing hematopoietic tissue (bone marrow), called ribs that connect the vertebrae to the sternum (Fig. 3.3a, b). Depending on their way of connecting to the sternum these may be classified as true, false or floating ribs.

Those that connect to the sternum directly are called true (vertebrocostal) ribs (1st to 7th ribs), while those that connect indirectly are called false (vertebrochondral) ribs (8th, 9th and usually the 10th rib) and those that do not connect are known as floating (free vertebral) ribs (11th, 12th and sometimes the 10th ribs).

The first rib is the most curved, broad and shortest. Its upper surface has 2 shallow grooves for the subclavian vessels separated by the scalene tubercle, where the anterior scalene muscle attaches and on its head is a surface for articulation with the T1 vertebrae.

The 2nd rib is longer than the first and closer in form to typical ribs. It has the same curvature as the first but is not as flat. It has two surfaces for articulation with the T1 and T2 vertebrae and the upper surface has a tuberosity for the serratus anterior.

The 3rd to the 9th are typical ribs and have a dorsal (vertebral) and a ventral (sternal) extremity with an intervening body (shaft). The dorsal extremity includes the head, the neck and the tubercle of the rib. The head has an articular surface divided into two parts by a ridge. The neck is about an inch long and connects the head to the body at the tubercle. The tubercle is located at the junction of the neck and the body and has a smooth surface to articulate with the transverse process of the vertebrae and a rough surface that provides for the attachment of the costotransverse ligament. The body is thin, flat and arched, mainly at the angle of the rib, up to where the erector spinae muscles attach to the rib. The body has a costal groove along the inferior border on the internal surface which provides protection for the intercoastal nerves and vessels.

The 10th rib is similar to a typical rib except that it has a single articular surface on its head.

The 11th and 12th ribs also have single articular surfaces like the 10th, but they have neither neck nor tubercle and the 12th has no costal groove.

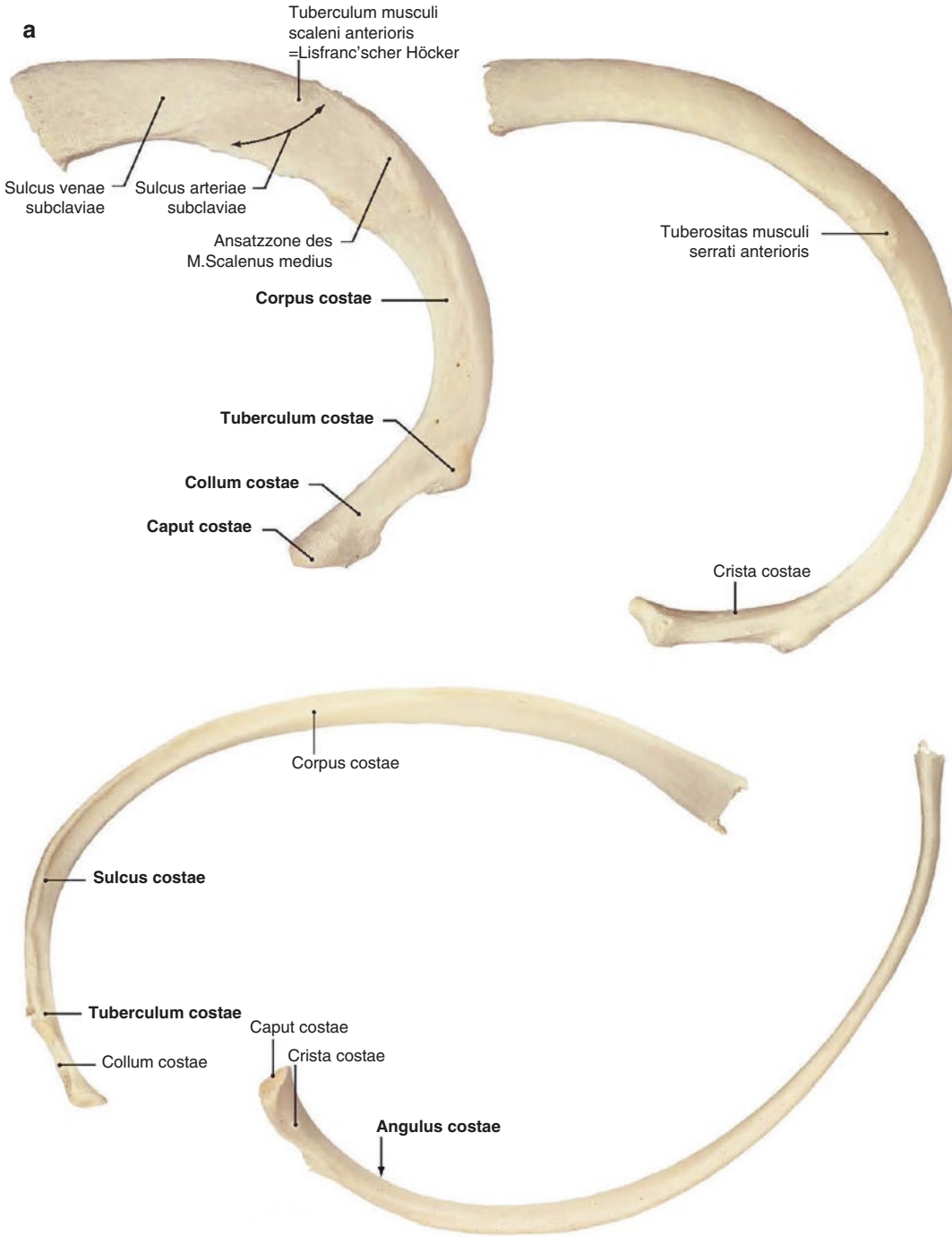

Fig. 3.3 (**a**) Typical and atypical ribs (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) Rib and Vertebrae articulation (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

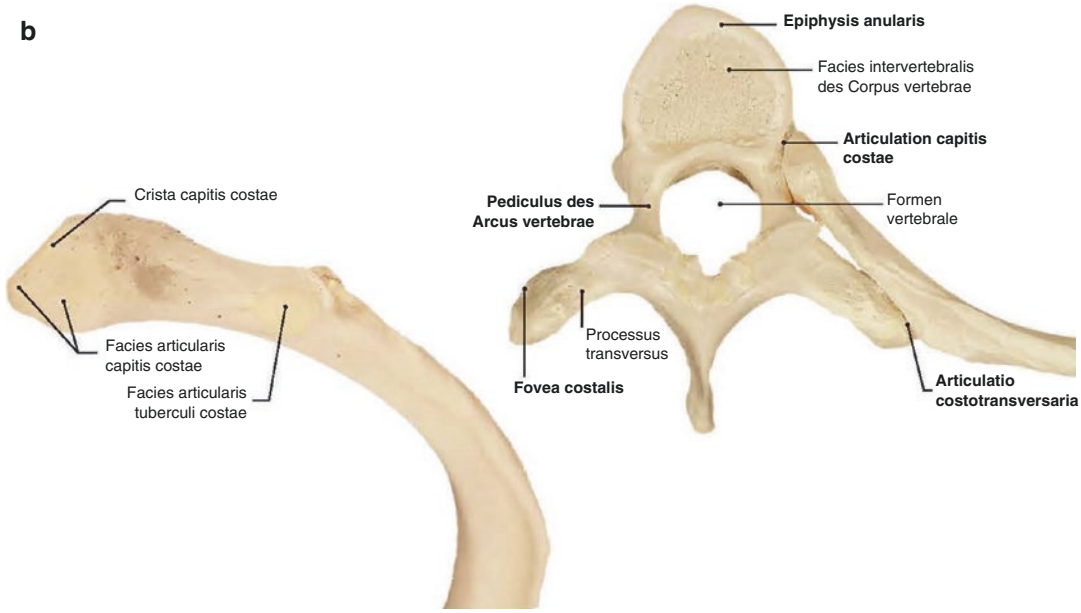

Fig. 3.3 (continued)

Coastal Cartilage

The costal cartilages provide a flexible attachment for the heads and the distal ends of the ribs (Fig. 3.1). The cartilage increases in length for the first 7 ribs and then decreases through the rest. The first 7 cartilages (and sometimes even 8th) attach directly and independently to the sternum. The 8th to 10th connect with the costal cartilages articulating a continuous, cartilaginous costal margin. The 11th and 12th cartilages form caps on the anterior ends of these floating ribs.

Intercostal Space

The intercostal spaces are the 11 spaces between the ribs and their costal cartilages. These spaces are occupied by intercostal muscles, membranes, and intercostal blood vessels and nerves (Fig. 3.4). The space below the 12th rib does not lie between ribs and is therefore referred to as the subcostal space.

Surgical Considerations

Supernumerary ribs, like cervical ribs, which are more common than lumbar ribs, are important as they may interfere with nerves and blood vessels and also may confuse the identification of vertebral levels in diagnostic imaging.

Rib fractures are painful because of the movement of the ribs in breathing and especially in sneezing, coughing, and laughing. The fracture of the first rib is rare but when occurs it may injure the brachial plexus and subclavian vessels, while lower rib fractures may tear the diaphragm and result in a diaphragmatic hernia.

Flail chest occurs when a segment of the thoracic cage fractures and detaches from the rest of it. The loose segment is extremely painful and moves paradoxically (inward during inspiration and outward during expiration), impairing ventilation.

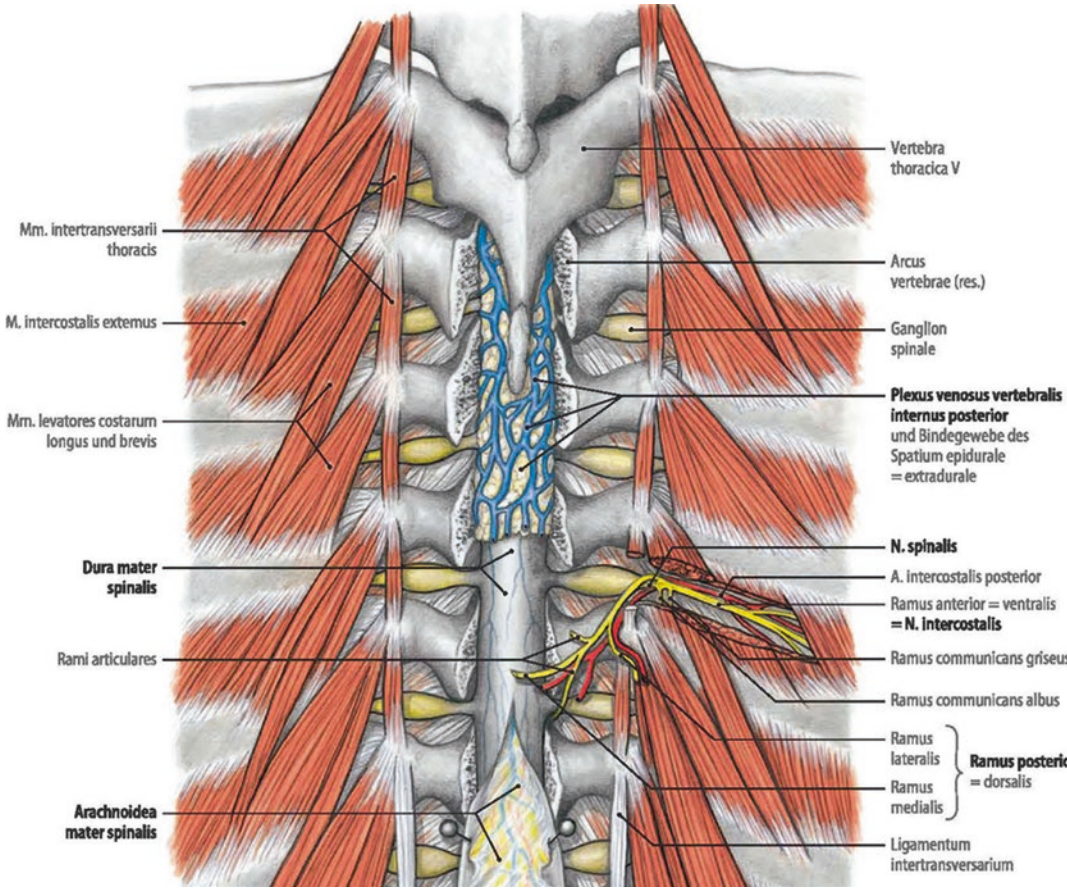

Fig. 3.4 Intercostal spaces (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

Vertebrae

Thoracic vertebrae are typical, independent vertebrae with bodies, arches, and the seven processes for muscular and articular connections (Figs. 3.3b and 3.5). The bodies have pairs of superior and inferior bilateral costal facets or demifacets that articulate with the heads of the ribs, a pair of costal facets on the transverse processes that articulate with the tubercles of the ribs (this is absent in the inferior two or three vertebrae), and long inferiorly slanting spinous processes. The facets are found in pairs along adjacent vertebrae encasing an interposed intervertebral disc. The two demifacets and the intervertebral disc thus create a socket to articulate with the head of the rib.

The first rib articulates only with vertebrae T1, making it an atypical vertebrae. Consequently, the superior costal facet of the vertebrae T1 is a complete facet while the inferior costal facet exhibits as a typical demifacet. The T10, T11 and T12 displays a single pair of costal facets positioned on their pedicles (with that of the T10 partly on the body as well).

The spinous processes of the vertebral arches are long and slope inferiorly and overlap the vertebra below. The superior articular facets face posteriorly and slightly laterally, while the inferior articular facets face anteriorly and slightly medially. The bilateral joint planes between the articular facets of the adjacent vertebrae create an arc, centering on an axis of rotation within the vertebral body, permitting small rotatory movements

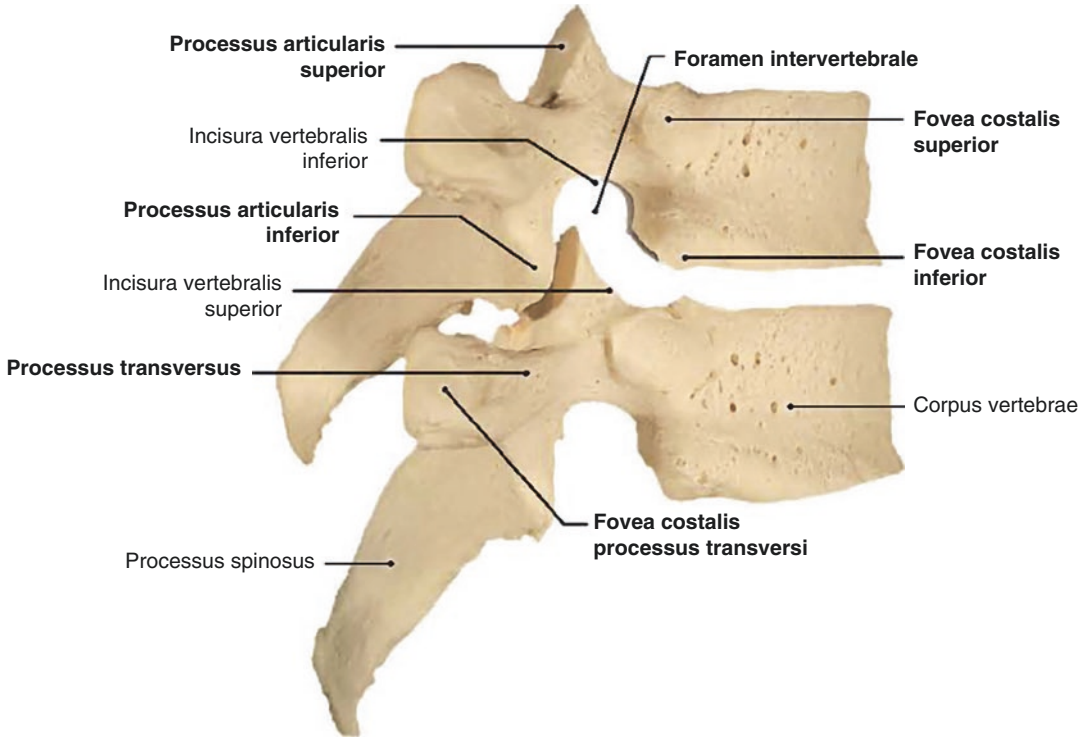

Fig. 3.5 Articular surfaces of the vertebrae (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

between adjacent vertebra, limited only by the attachment to the rib cage.

Muscles of the Thoracic wall

Muscles from the upper limb including the pectoralis major, pectoralis minor, subclavius, serratus anterior muscles and latissimus dorsi muscles attach to the thoracic cage (Figs. 3.6 and 3.7). Other muscles that attach to the thoracic wall include the abdominal muscles and some back and neck muscles.

The muscles of the thoracic wall include the external and internal intercostal. The external intercostals originate from the lower border of the rib and insert at the upper border of the rib below. They extend all the way from the tubercle of the rib to the costal cartilage. The external intercostal membrane replaces the muscle from costal cartilage to the sternum.

The internal intercostal muscles originate from the inner surface of the rib and insert at the upper border of the rib below. They extend from the sternum to the angles of the ribs. Internal intercostal membrane replaces muscle between angle of rib and vertebrae.

The subcostal muscles are found in the lower thorax and they lower the ribs when the last rib is fixed by the quadratus lumborum muscle. These originate the inner surface of the near angle of the ribs and insert 2 or 3 ribs below.

The transverse thoracic muscles are found in the inner chest wall. These originate from the posterior body xiphoid process of the sternum and sternal ends of the costal cartilages of ribs 4–6 and insert at the lower border and costal cartilages of ribs 2–6.

The levatores costarum is found in the posterior thorax. It originates from the ends and transverse processes of vertebrae C7 to T11 and inserts into the outer surface of the ribs immediately below the origin.

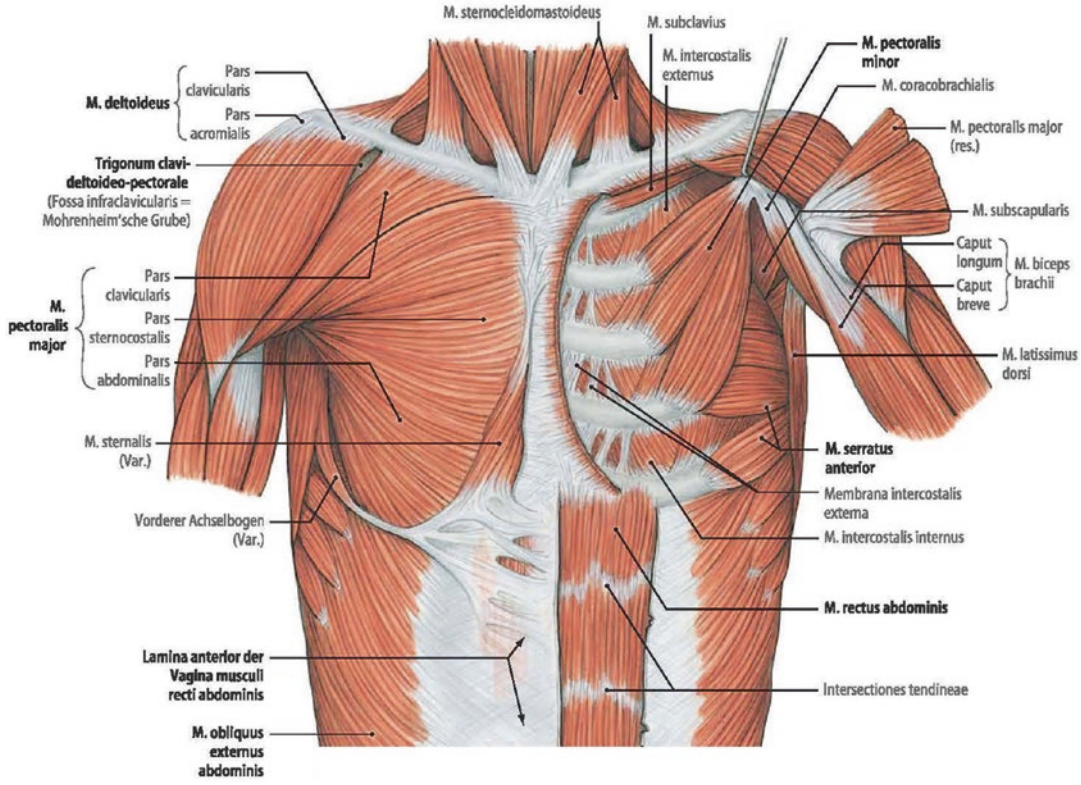

Fig. 3.6 Muscles of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

Serratus posterior superior is found on the posterior of upper thorax. It originates from the ligamentum nuchae, supraspinal ligament, and spines of vertebrae C7 to T3 and inserts on the upper borders of the ribs

Serratus posterior inferior is found on the posterior of the lower thorax. It originates from the supraspinous ligaments and spines of vertebrae T11 to T13 and inserts on the lower border of the ribs 9–12, lateral to the angles of the ribs.

The Diaphragm

The diaphragm is a dome shaped skeletal muscle with a dense collagenous connective tissue that separates the thoracic cavity from the abdominal cavity (Fig. 3.8).

The diaphragm originates form three parts; the sternal, the costal and the lumbar. The sternal part of the diaphragm contributes two muscular bands from the dorsal side of the xiphoid process, the costal contributes muscle origins from the costal cartilage and bones of ribs 7–12 and the lumbar provides the lumbocostal arches and crura. The lumbocostal arches are produced by the medial and the lateral arcuate ligament. The medial arcuate ligament attaches medially to the body of the 1st and 2nd lumbar vertebrae and crosses the psoas muscle to attach laterally to the anterior transverse processes of the same (1st and 2nd lumbar) vertebrae. The lateral arcuate ligament attaches medially to the anterior transverse process of the first lumbar vertebrae and crosses the quadratus lumborum muscle to attach laterally to the tip of rib 12. The crura has a right part that is larger and originates from the bodies and discs of the lumbar vertebrae 1–3 with the medial most fibers crossing in front of the aorta to the left and a left part that originates from the bodies and discs of the 1st and 2nd lumbar vertebrae with some fibers crossing to the other side. The

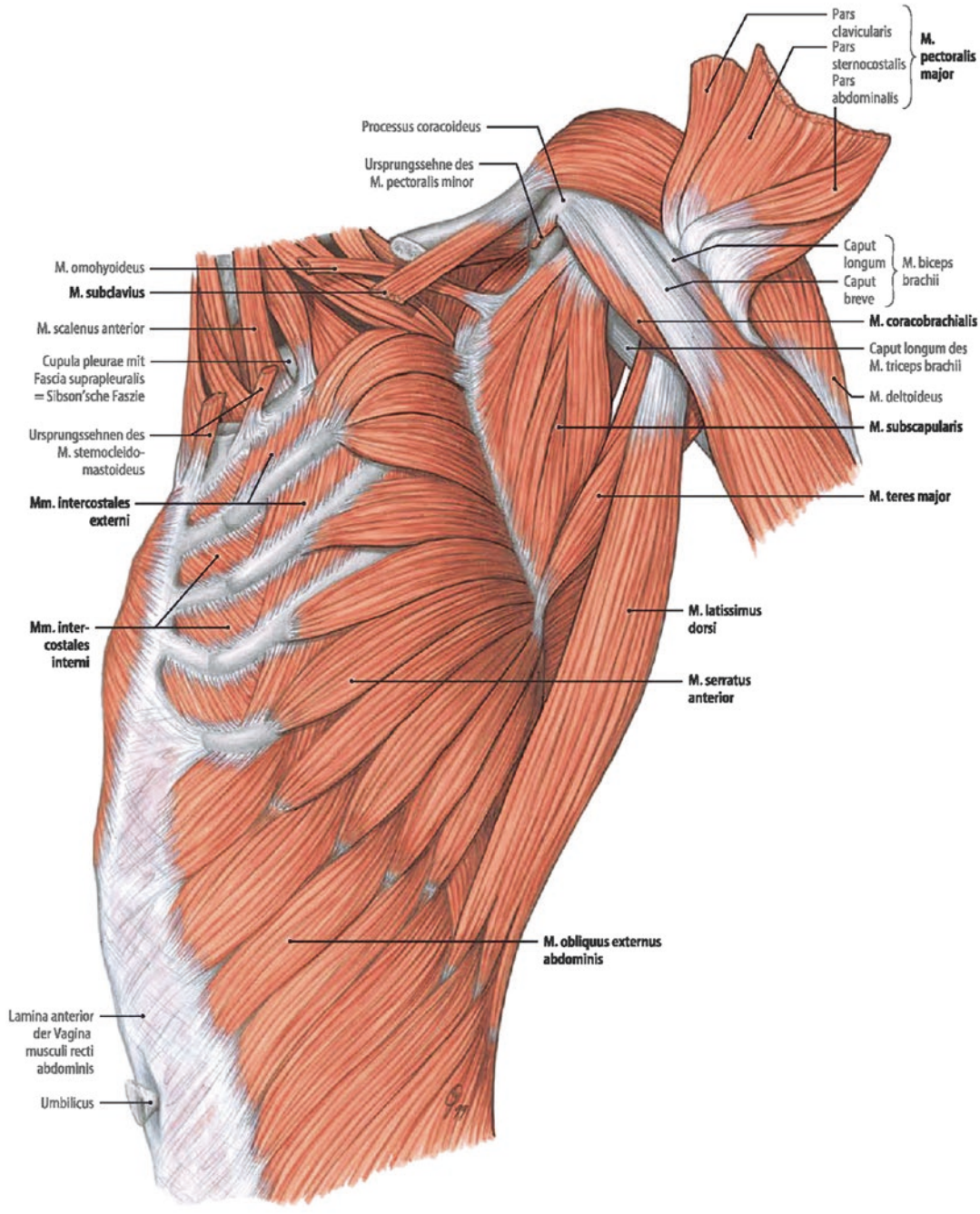

Fig. 3.7 Lateral view of the muscles of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

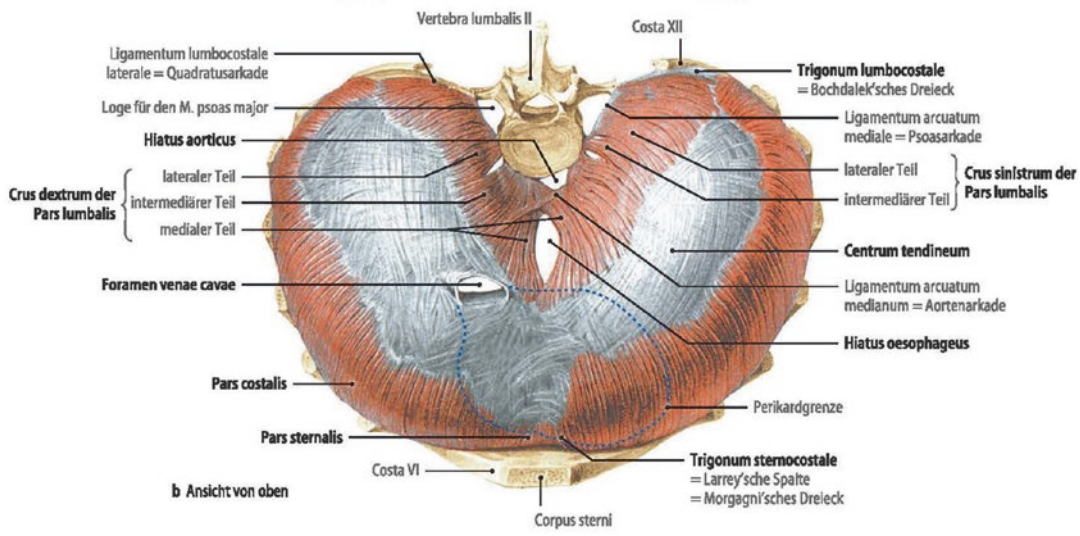

Fig. 3.8 Diaphragm (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

sternal, costal and lumbar parts of the diaphragm insert at the central tendon near the center of the diaphragm and are supplied by the phrenic nerve of the cervical plexus.

The diaphragm has three orifices; one each for the esophagus, the aorta and the inferior vena cava. The esophageal hiatus, at the level of the 10th thoracic vertebrae provides passage for the esophagus and the right and left vagus nerves. The aortic hiatus, to the left of the midline at the level of the 12th vertebrae provides passage for the aorta, azygos vein and thoracic duct. The vena cava formen, to the right of the midline, at the level of the disc between the 8th and 9th thoracic vertebrae, transmits the vena cava and small branches of the right phrenic nerve. A few minor openings are also found in the diaphragm, at the right crus for the right greater and lesser splanchnic nerves, the left crus for the greater and lesser splanchnic nerves and the hemiazygos vein, and anteriorly between sternal and costal parts of the diaphragm for the passage of the superior epigastric artery.

Fascia of the Thoracic Wall

The names of the fascia of the thoracic wall correspond to the muscles they are associated with. The pectoral or pectoralis fascia thus is the deep fascia over the anterior thoracic wall associated with the pectoralis major. Deeper to that is found the clavipectoral fascia, suspended from the clavicle and enclosing the pectoralis minor muscle. Internally, the thoracic cage is lined with a thick fibroareolar layer called the endothoracic fascia.

The endothoracic fascia attaches the lining of the lung cavity to the thoracic wall and becomes fibrous over the apices of the lungs as the supra-pleural membrane.

Nerves of the Thoracic Wall

The thoracic wall is supplied by 12 pairs of thoracic spinal nerves that divide into anterior and posterior primary rami (Fig. 3.9). The anterior rami of nerves T1 to T11 form the intercostal nerves that run along the inferior margin in costal grooves, while 1st and 2nd run along the internal surfaces of the ribs. That of the T1 divides further into a large superior part that joins the brachial plexus and a small inferior part that presents as the 1st intercostal nerve. The intercostobrachial nerve emerges from the 2nd intercostal nerve (sometimes also the 3rd) and penetrates the serratus anterior to enter the axilla and arm. The lateral cutaneous branch of the 3rd intercostal nerve may give rise to a second intercostobrachial nerve. The anterior ramus of nerve T12, coursing

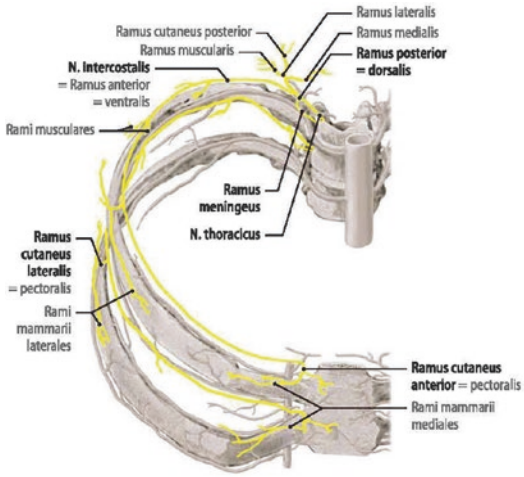

Fig. 3.9 Intercoastal nerves of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

inferior to the 12th rib, is the subcostal nerve. The posterior rami of thoracic spinal nerves pass posteriorly, immediately lateral to the articular processes of the vertebrae, to supply the joints, muscles, and skin of the back in the thoracic region.

The group of muscles supplied by the posterior ramus and anterior ramus (intercostal nerve) of each pair of thoracic spinal nerves are the intercostal, subcostal, transverse thoracic, levatores costarum, and serratus posterior muscles associated with the intercostal space that includes the nerve.

The branches of a typical intercostal nerve include the rami communicantes, the collateral branch, pleural branches, the lateral cutaneous branch, the anterior cutaneous branch, the muscular branches, and the peritoneal sensory branches. The rami communicantes or communicating branches connect the intercostal nerves to a ganglion of sympathetic trunk, adjacent to the vertebral column. The collateral branches arise close to the angle of the ribs and run parallel to the main nerve along the upper border of the rib below supplying the intercostal muscles and giving rise to the pleural branches supplying the parietal pleura. The lateral cutaneous branch pierce the internal and external intercostal muscles splitting into anterior and posterior branches to supply the skin of the lateral thoracic and abdominal walls. The anterior cutaneous branches penetrate the muscles and membranes of the intercostal space to provide the terminating ends of the intercostal nerves by dividing into medial and lateral branches to supply the skin on the anterior of the thorax and abdomen. The muscular branches supply the intercostal, subcostal, transverse thoracic, levatores costarum, and serratus posterior muscles. Finally the peritoneal sensory branches are similar to the pleural sensory branches but arise from the lower intercostal nerves because the lower intercostal spaces are more related to the peritoneum that to parietal pleura.

Arteries of the Thoracic Wall

The thoracic wall receives blood supply from the subclavian artery, the axillary artery and the thoracic aorta (Fig. 3.10a, b).

The subclavian artery supplies blood through the internal thoracic and supreme intercostal arteries. The internal thoracic arteries (previously known as the internal mammary arteries) arise close to the root of the subclavian artery and descend into the thorax posterior to the 1st costal cartilage and then run lateral to the sternum and posterior to the six upper costal cartilages and intervening internal intercostal muscles. After the 2nd costal cartilage, the internal thoracic artery runs anterior through slips of the transverse thoracic muscle to communicate with the parietal pleura. The internal thoracic arteries give rise to the anterior intercostal arteries supplying the superior six intercostal spaces and terminate in the 6th intercostal space by dividing into the superior epigastric and the musculophrenic arteries. The superior epigastric artery crosses the sternocostal triangle of the diaphragm, descends along the posterior of the rectus abdominis and anastomoses with the inferior epigastric artery. The musculophrenic artery supplies several intercostal spaces, pierces the diaphragm, and anastomoses with the deep circumflex iliac artery.

The axillary artery supplies blood through the superior and lateral thoracic arteries. The superior thoracic artery passes between the pectoralis minor and the pectoralis major to the lateral side of the chest and supplies the first and second intercostal spaces as well as to the superior portion of

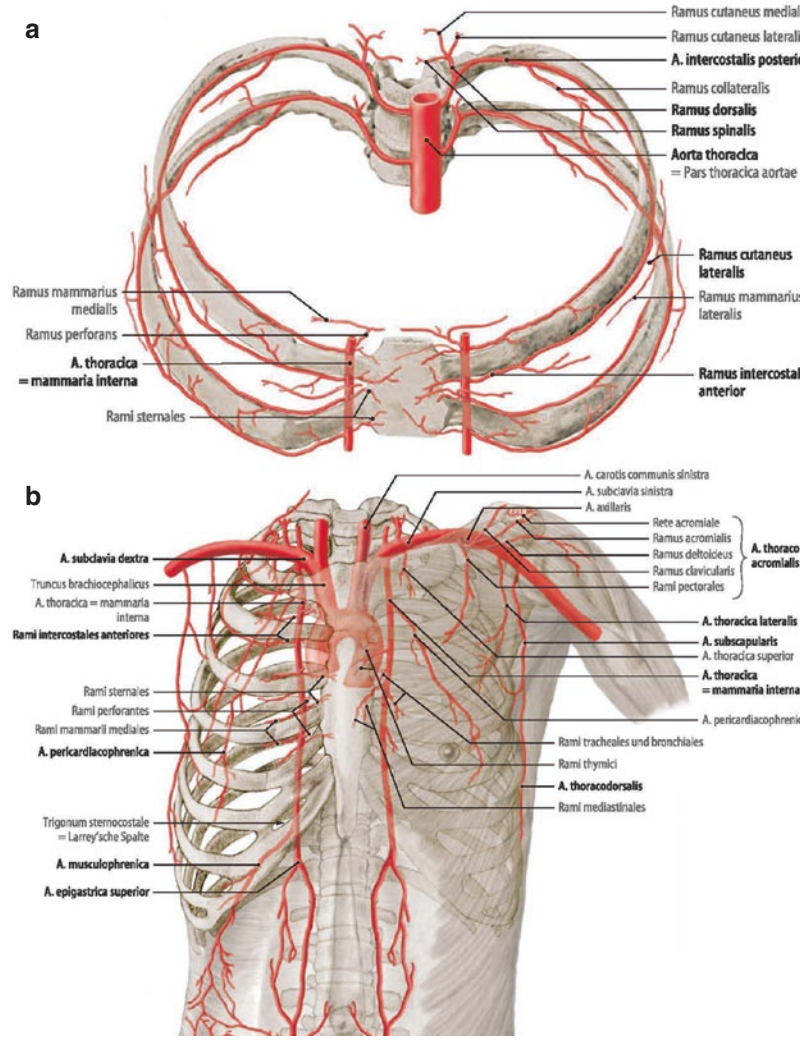

Fig. 3.10 (**a**) Intercoastal arteries of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) Arteries of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

serratus anterior. The lateral thoracic artery trails the lateral lower border of the pectoralis minor.

The thoracic aorta gives rise to the posterior intercostal and subcostal arteries. The posterior intercostal arteries arise from the superior intercostal artery for the first two intercostal spaces and from the thoracic aorta for the 3rd to the 11th intercostal spaces. The longer right arteries cross the vertebrae and pass posterior to the oesophagus, thoracic duct, azygos vein, and the right lung and pleura to reach their destinations. The arteries produce posterior branches that accompany the posterior rami of the spinal nerve to supply the spinal cord, vertebral column, and back muscles. Small collateral branches cross the intercostal spaces to run along the superior borders of the rib and along with the terminal branches anastomose with anterior intercostal arteries. The anterior intercostal arteries supply the anterior of the upper six intercostal spaces and pass laterally in the intercostal space dividing to two branches, one near the inferior margin of the superior rib and the other near the superior margin of the inferior rib.

Veins of the Thoracic Wall

The thoracic wall is drained by the intercostals veins, which are made up of the anterior and the posterior intercostals veins (Fig. 3.11). The

anterior intercostals veins of the upper six intercostals spaces drain into the internal thoracic vein while those of the 7th to 9th spaces end in the musculophernic veins. The anterior intercostals veins anastomose with the posterior intercostals veins. The posterior intercostals veins are the supreme intercostal in the 1st intercostals space draining directly into the corresponding and nearby brachiocephalic veins, the superior intercostals vein in the 2nd to 4th intercostal spaces uniting to form the superior intercostals vein and the subcostal below the bottom rib, with the veins in spaces 5–11 without specific names. Most posterior intercostal veins drain to the azygos/hemiazygos venous system going further into the superior vena cava.

The right superior intercostal vein drains into the superior vena cava, while the left superior intercostal vein empties into the left brachiocephalic vein. This requires the vein to cross anteriorly the arch of the aorta or the root of the great vessels arising from it and between the vagus and phrenic nerves. The left bronchial veins and often the left pericardiacophrenic vein drain to the left superior intercostals vein, which also communicates with the accessory hemiazygos vein inferiorly.

The internal thoracic veins are also found in the thoracic wall and are associated with the internal thoracic arteries.

Thoracic Apertures

The thoracic cage is open on either end providing the superior and inferior thoracic apertures. The smaller superior aperture connects with the neck and upper limb. The larger inferior aperture provides the origin for the diaphragm, which completely occludes the opening.

Superior Thoracic Aperture

The superior thoracic aperture or the anatomical thoracic inlet slopes anteroinferiorly and has a circumference demarcated by the manubrium, the 1st pair of ribs, the cartilages of the 1st pair of ribs and the vertebra T1 (Fig. 3.12a, b). The structures that cut through the superior thoracic aperture to enter the thoracic cavity include the trachea, esophagus, nerves, and vessels that supply and drain the head, neck, and upper limbs.

Inferior Thoracic Aperture

The inferior thoracic aperture or the anatomical thoracic outlet has its circumference demarcated by the Xiphisternal joint, the 11th and 12th pair of ribs and the 12th thoracic vertebra (Fig. 3.13a, b). The diaphragm covers the circumference of the inferior thoracic aperture and separates the

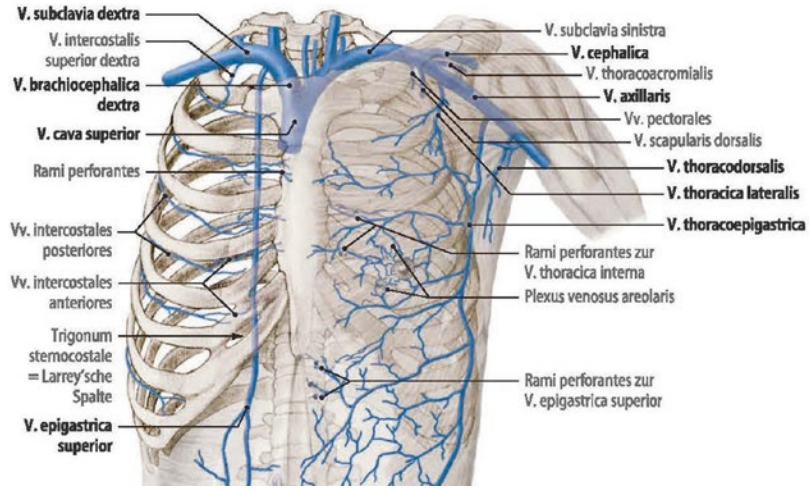

Fig. 3.11 Veins of the thoracic wall (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

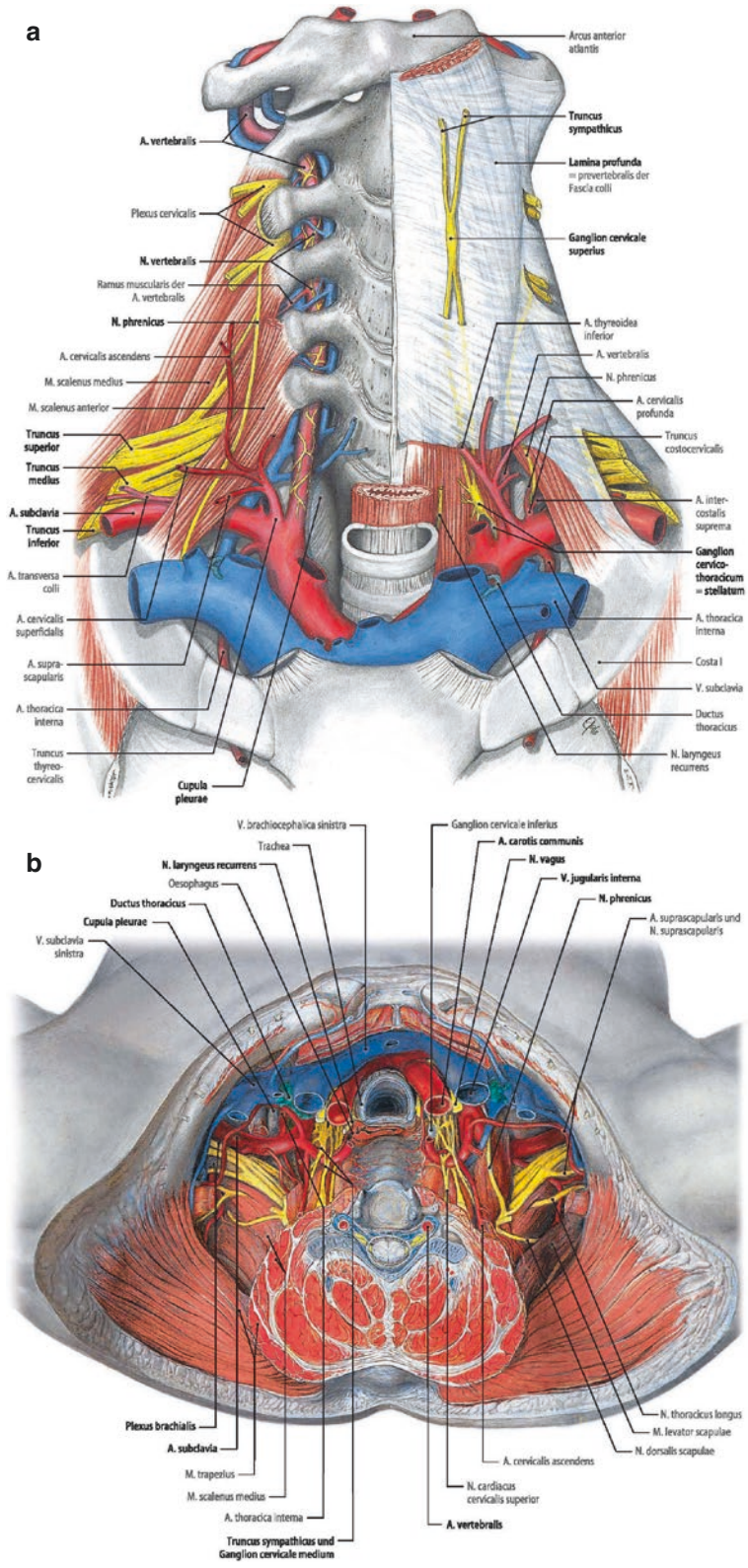

Fig. 3.12 (**a**) The superior thoracic aperture (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) The superior thoracic aperture (Cross sectional view) (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

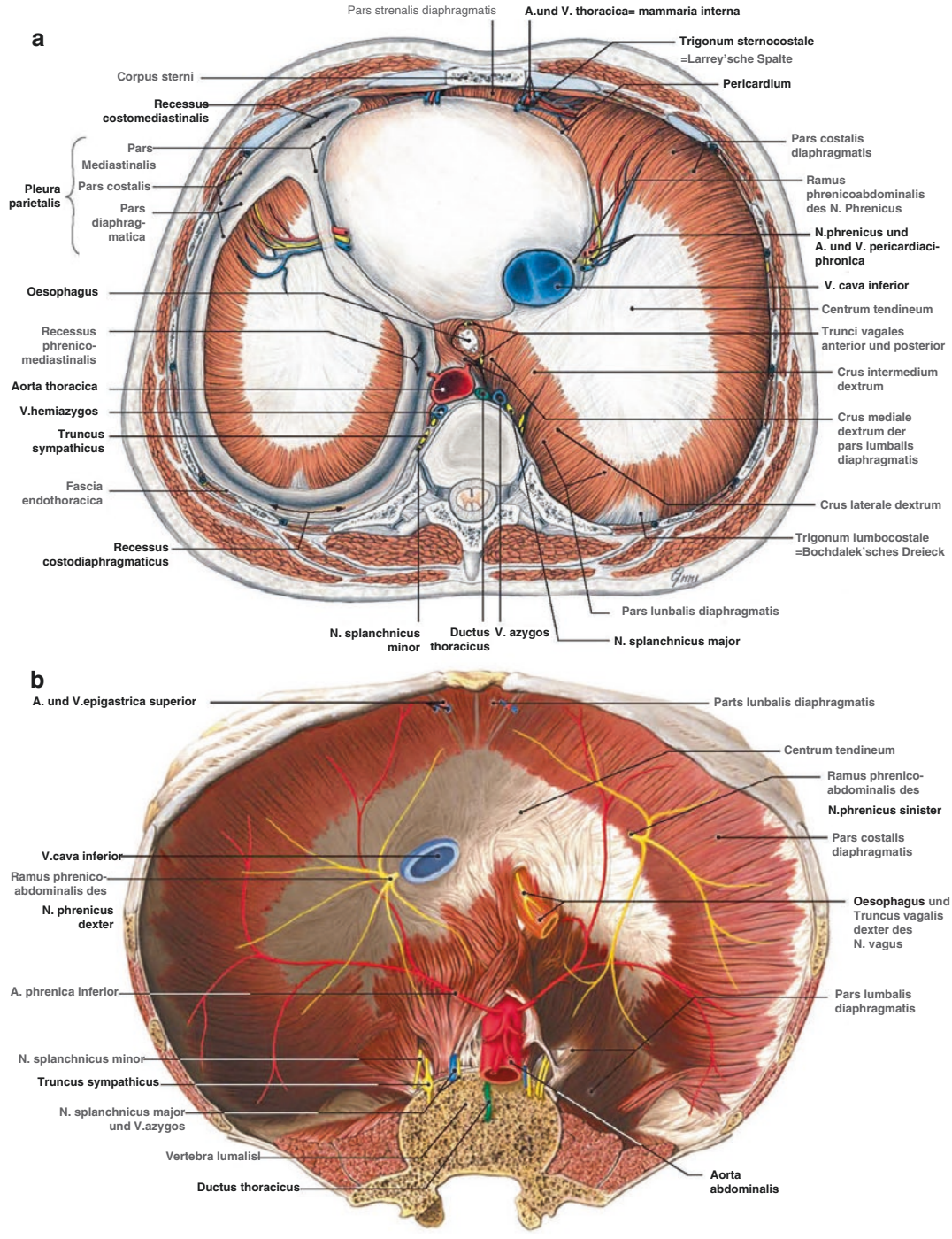

Fig. 3.13 (**a**) The inferior thoracic aperture (Thoracic view) (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) The inferior thoracic aperture (Abdominal view) (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

thoracic cavity from the abdominal cavity, allowing only the passage of the esophagus and the inferior vena cava, through it and the aorta, posterior to the diaphragm. The liver, spleen and stomach lie superior to the plane of the inferior thoracic aperture, pushing the dome of the diaphragm to the level of the 4th intercostal space.

The Mammary Glands

The mammary glands can be seen vertically from the 2nd to the 6th or 7th rib and horizontally from the border of the sternum to the midaxillary line. The center of the mammary gland is located just above the mammary papilla or nipple, which can be seen at the 4th interspace.

Each mammary gland is made up of 15–20 separate lobes set together like a citrus fruit and is more prominent in females. The mammary gland lies in the superficial fascia with the glands embedded in adipose tissue and separated from the muscle fascia by connective tissue (Fig. 3.14a, b). The suspensory ligaments of Cooper run from the dermis through the glands to the superficial fascia.

The mammary glands are supplied by the anterior and lateral cutaneous branches of the 2nd to the 6th intercostal nerves. From the axillary artery, the mammary glands are supplied by the supreme thoracic artery, the pectoral branch of thoracoacromial, and the external mammary artery, which emerges from the lateral thoracic arteries. It is also supplied by the cutaneous branches of the intercostal arteries arising from the 3rd to the 5th instercostal spaces and the perforating branches of the internal thoracic artery from the 2nd to the 4th intercostal spaces. Veins initiating from the anastomotic circle around the papilla pass peripherally to the axillary, internal thoracic, lateral thoracic, and the upper intercostal veins. The lymphatic drainage originates from the extensive perilobular plexus which journey along the lactiferous ducts towards the areola forming the subareolar plexus and draining into the pectoral group of axillary

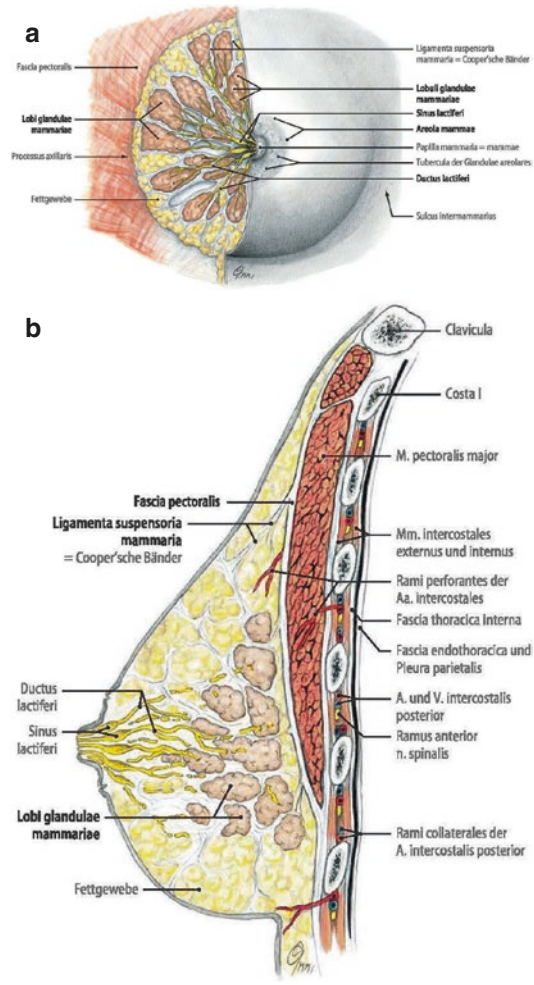

Fig. 3.14 (**a**) The mammary gland (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) The mammary gland (Cross sectional view) (From Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

nodes. The medial side of the gland drains to the parasternal nodes, the upper gland to the apical axillary nodes or supraclavicular nodes and the caudal part of the gland to the abdominal lymph nodes.

This chapter described the basic anatomy of the chest wall and some surgical considerations, essential for surgery. More detailed anatomy and surgical considerations will be explained in further chapters in the context of the surgical conditions, procedures and instruments used.

Suggested Further Reading

1. Gray H. Gray's anatomy: descriptive and surgical. New York: Cosimo Classics; 2010. 750 p. p.
2. McMinn RMH, Abrahams P, Boon J, Spratt J. McMinn's clinical atlas of human anatomy. 6th ed. Edinburgh: Mosby Elsevier; 2008. 386 p. p.
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5. Tortora GJ, Derrickson B. Principles of anatomy and physiology. 11th ed. New York;; Chichester: Wiley; 2006. xxxii, 1146 p. p.

Sternalis Muscle

4

Athanasios Raikos and Panagiota Kordali

Introduction

Sternalis muscle is an aberrant muscle found on the anterior thoracic wall. The muscle is well known to anatomists but not very familiar to clinicians such as surgeons and radiologists. Furthermore, medical students and doctors in training are also unfamiliar with the entity as it is not included in standard medical textbooks [1–4]. The muscle is of interest to researchers for more than 400 years and attracted lots of controversy on its embryological origin, terminology, and function. The first description on the muscle was from Cabrol in 1604 in his book "*Alphabeton anatomikon*". Much later, in 1726, Du Puy as stated by Turner, provided a detailed description of a bilateral example of sternalis muscle and supported that the muscle is a phylogenetic remnant carried over from other primates [5].

Sternalis muscle is one of the most variably termed anatomic variants with at least 21 different synonyms found in the literature as can be seen in Table 4.1. The common term for the muscle nowadays is sternalis and comes from its non-standard attachment and parallel arrangement with the sternum. In many cases the muscle's origin and insertion in not related to the sternum at all but it is still called sternalis for ease of communication. Christian supported that the term sternalis is very general and should be used only for parasternal muscles related to pectoralis major [7].

Characteristic Features of Sternalis Muscle

Sternalis has a variety of appearances, although it is generally considered as a flat, strip-like, or oblique oriented muscle lying always anteriorly to pectoralis major and covered by the superficial pectoral fascia (Table 4.2). On its posterior plane it is covered by the anterior pectoral fascia [2]. It might appear in one or in both sides of the anterior thoracic wall maintaining a parasternal position or crossing the sternum. It can be found in various forms, single-slip, multi-slip, Y-shaped, X-shaped, and cross type (Fig. 4.1).

It is believed that sternalis is homologous to other muscles of the anterior chest, specifically the main four in the literature is the sternocleidomastoid [9], rectus abdominis [10], pectoralis major [11], and panniculus carnosus, a subcutaneous muscle correspondent to platysma in humans [12]. The sternalis can take rise from various locations such as the upper sternum, infraclavicular region, pectoralis major, sternocleidomastoid

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A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_4

Table 4.1 Synonyms for sternalis muscle according to the literature

Terminology
1. Abdominoguttural
2. Abdomino-cutaneous
3. Accesorius ad rectum
4. Anomalus sterni
5. Cutaneous pectoris
6. Episternalis
7. Parasternal
8. Pectoris rectus
9. Praesternalis
10. Presternalis
11. Rectus abdominalis superficialis
12. Rectus sternalis
13. Rectus sterni
14. Sternalis
15. Sternalis brutorum
16. Sternalis Japonicus
17. Superficial rectus sterni
18. Supracostalis
19. Supracostalis anticus
20. Thoracicus
21. Triangular muscle anterior or external to sternum

Table adopted and modified from Arraez-Aybar et al. [6]

tendon, and sternal angle [11, 13–15]. The insertion of the muscle is also variable, some examples for it is the costal cartilages, anterior pectoral fascia, lower ribs, lower sternum, sheath of rectus abdominis muscle, and aponeurosis of abdominal external oblique muscle [2, 8].

The size of sternalis varies from a few short muscle fascicles to well-developed muscle. In a study utilizing 948 multidetector computed tomography (MDCT) datasets, a sternalis was found in 10.5 % of cases with the average height of 77.9 ± 25.1 mm, width 19.4 ± 12.2 mm, and thickness 2.8 ± 1.3 mm [16]. In a second more extensive study on 6,000 MDCT datasets a sternalis was found in 5.8 % of cases and the mean length, width, and thickness were 111 ± 33 mm, 17.7 ± 9.9 mm, and 4.1 ± 1.7 mm respectively [17]. Nevertheless, muscles up to 26 cm long and 4.6 cm wide have been reported in the literature [18].

Sternalis' innervation in variable and might depend on the topographic location of the muscle. According to a review on 35 studies, 55 % of cases were innervated by branches of external and internal thoracic nerves, 43 % from intercostal nerves, and 2 % had double innervation from intercostal and thoracic nerves [18].

The vascular supply of sternalis has not been emphasized in the literature. However, the muscle is expected to receive supply and drainage from adjacent vessels according to its topographic location in the chest. The supply should be similar to pectoralis major, thus from anterior perforating (anterior intercostal branches) of internal thoracic artery and pectoral branches of lateral pectoral vessels. In a report of two cases with sternalis, perforating branches of the internal thoracic artery supplied the aberrant muscles [19].

Incidence

The incidence of sternalis varies among different populations and ethnic groups as well as between the two genders. The reported global incidence in autopsy material and living population is up to 23.5 % [2, 4, 20]. It is more common is females than males, thus 13 % and 8.4 % respectively [16], while others support that the incidence between the two genders is the almost equal [12]. The bilateral sternalis is more common in females than males, with an incidence of 8.7 % and 6.4 %, respectively [3].

The reported incidence in European population is 1.9–9.9 % [2]. Specifically, the incidence in a study on English population was 4 % [21], at least 3 % in Greeks [22], 1.5 % in Germans [23], 2.9 % in Bulgarians [2], 4.6 % in French [24], and 7.4 % in Portuguese [25]. In Asian population the frequency varies between 0.5 and 23.5 % [2, 20]. Specifically, as reported in a review it was found with a frequency of 18.2–23.5 % in North-Chinese [2], 4.1–15.6 % in Japanese [2, 26]. In Taiwanese the incidence is the lowest of all populations at 0.5 % [20]. In Malaysians is 2 % [27] and 6.2 % in Koreans [28]. Moreover, it was found with a frequency of 4 % in Saudi Arabians [29], 9.3 % in Turks [30], and 4–8 % in Indians [2]. Additionally, in Americans the reported incidence ranges is from 2.9 to 6.4 %

Table 4.2 Characteristic features of sternalis muscle

Etiology	Congenital anatomic variation.
Types	Unilateral, bilateral, cross-type, Y-type, X-type. Single or multiple muscle slips.
Shape	Flat, oblique, fan-shaped muscle.
Muscle origin	Upper sternum and the infraclavicular region such as pectoralis major, sternal angle, sternocleidomastoid muscle, ventral and lateral body wall muscles of thorax and abdomen or the rectus abdominis sheath, rectus abdominis muscle, remains of the panniculus carnosus muscle sheet, abdominal external oblique muscle or from the ventrolateral part of the diaphragm.
Muscle insertion	Costal cartilages, anterior pectoral fascia, lower ribs, sheath of the rectus abdominis muscle, aponeurosis of the abdominal external oblique muscle, sternal angle.
Innervation	Branches of the external or internal thoracic (pectoral) nerves, intercostal nerves or from both nerves.
Incidence	Cadavers: 0.5–23.5 %. Mammograms: 0.01 %. Multidetector CTs: 5.8–10.5 %.
Appearance risk factors	Unknown.
Symptoms	Usually asymptomatic. May cause changes in the ECG and breast or chest asymmetry.
Gender ratio	Slightly more frequent in females.
Imaging	Plain X-ray: not visible. Mammogram: anomalous vertical shadow in cranio-caudal view. CT/MRI: parasternal structure. Multidetector CT: longitudinal muscle lying onto pectoralis major muscle.
Differential diagnosis	Benign and malignant breast lesions, extra-abdominal desmoid tumors, granular cell tumors and other pathology such as diabetic mastopathy, abscesses, hematomas, sclerosing adenositis, lymphadenitis, fat necrosis and surgical scars.
Function	Unknown. Might facilitate in shoulder joint and chest movement.
Surgical interest	May cause diagnostic dilemma. Can be used as a flap in reconstructive surgery of the head and neck, anterior chest wall, and breast.
Clinical significance	ECG alterations, chest and breast asymmetry.

Table adopted and modified from Raikos et al. [8]

in white-Americans and 4.1–15 % in African-Americans [7, 12]. Finally, as reviewed, the incidence in Africans ranges from 4.2 to 14.3 % [2].

Radiologic MDCT datasets on 948 Japanese patients revealed an overall incidence for sternalis of 10.5 %. It was found more frequently in females than in males, thus 13 % and 8.4 %, respectively. Twenty-five percent were bilateral and 75 % unilateral type. From the unilateral cases 45 were on the right side, 28 on the left, and 2 were cross type. In comparison, the muscle was wider in males than females [16]. In a similar study on a much larger population comprising 6,000 Chinese adults the frequency of sternalis was 5.8 % but this time it was slightly more often in males, 6 % comparing to 5.5 % in females [17].

The incidence of sternalis in mammographies does not agree with findings in other imaging modalities and tissue examining methods. In a study on 1580 datasets no cases where found at all, whereas in a very large study on 32,000 mammographies only four cases where identified giving a mere frequency of 0.01 % [31]. The very low incidence in mammography images is grounded to pitfalls in mammographic interpretation, insufficient exposure of tissues in the mediolateral projection, and unfamiliarity of the radiologist with sternalis' muscle entity [4, 31].

Embryology

The embryological origin of sternalis muscle is on debate. Synoptically, the suggested way to study the morphological significance and embryonic origin of a muscle is by examining its innervation [6]. Another area of investigation relates to the origin, insertion, and direction of its muscle fibers. Molecular embryology studies aid to confirm the actual origin of a muscle.

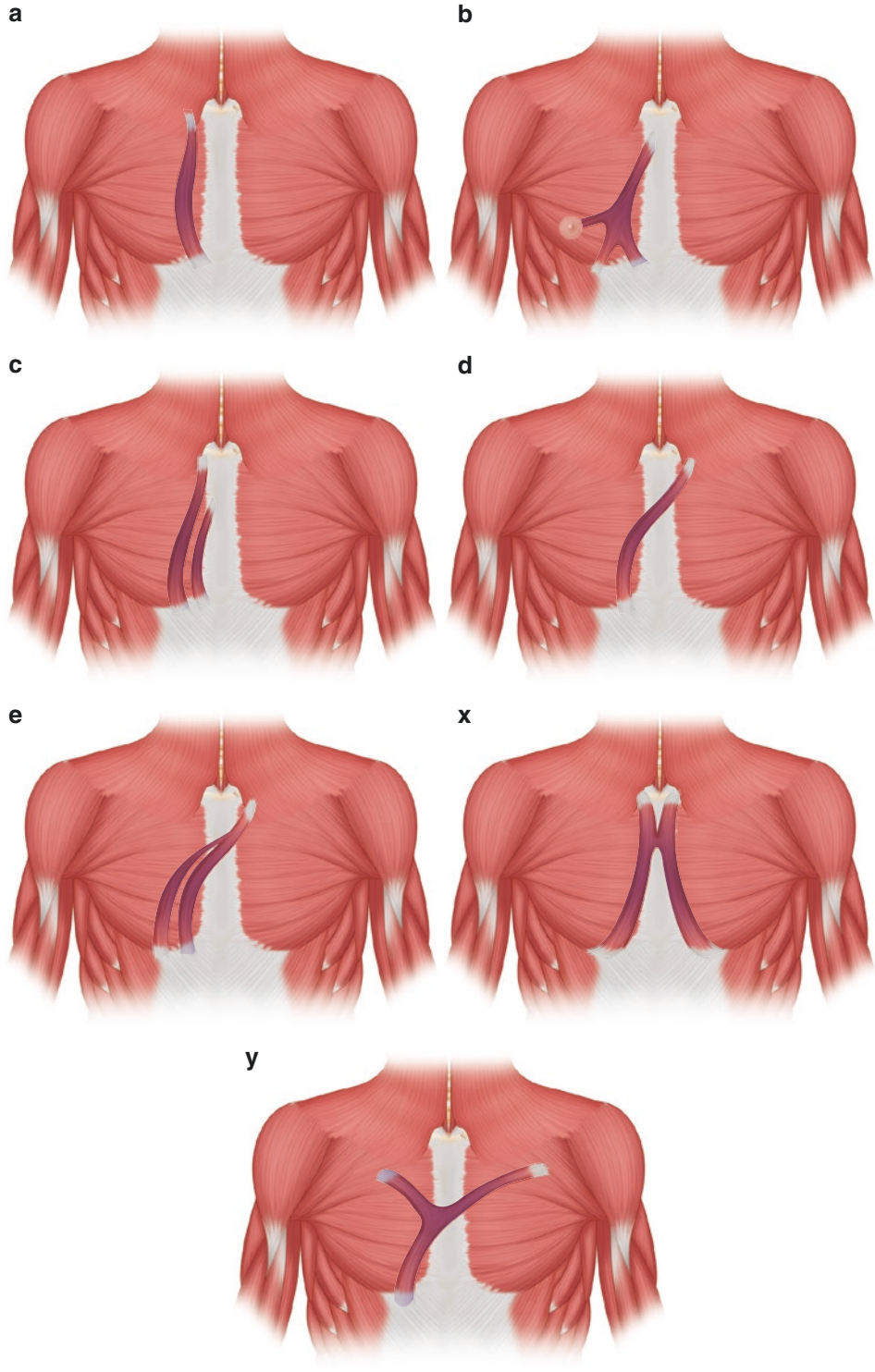

Fig. 4.1 Schematic representation of sternalis muscle classification. Type A: Single muscle belly. Type B: Single belly with attachment to the ipsilateral nipple. Type C: Multiple bellies (two or more). Type D: Single belly crossing over sternum. Type E: Multiple bellies crossing over sternum. Type X: X-shaped sternalis with tendinous connection to the sternocleidomastoid. Type Y: Y-shaped sternalis with muscle bellies passing onto other muscles or to well-defined fascicules of the sternalis (Classification adopted and modified from Raikos et al. [4])

The most popular theories support that the embryological origin of sternalis is either from pectoralis major, sternocleidomastoid, ventral and lateral body wall muscles of thorax, the rectus abdominis sheath, the rectus abdominis muscle, the abdominal external oblique muscle or from the ventrolateral part of the diaphragm, or it might be a remnant of the panniculus carnosus muscle sheet [4, 12, 32–34]. Among researchers there are some who believe that sternalis is a unique muscle and does not take its origin from other muscles [5].

It is hypothesized that sternalis derives from hypaxial myotomes/dermotomes from which ventral and lateral body wall muscles of the thorax and abdomen develops [8]. Similarly, it is been supported that sternalis is part of the ventral longitudinal column of muscles in the pectoral region [35]. Sternalis' appearance might be the effect of atavism of muscle fibers of the pectoral cutaneous muscle of lower animals and it has been co-related with fetal anencephaly [5, 34, 36].

Function

The function of sternalis muscle is still unknown. It may participate in the shoulder joint and chest movement or have an accessory role in the lower chest wall elevation [3, 28]. In case of pectoralis major absence the sternalis, if present, might enlarge to compensate pectoralis function [3]. The muscle can be used in the surgical field if big enough and identified promptly. Specifically, it can be used as a muscle flap in reconstructive surgery of the head and neck, anterior chest wall, and breast [37, 38].

On the other side, the presence of the muscle can affect negatively the proper positioning of breast implants in augmentation mammoplasty during submuscular pocket dissection as sternalis might interfere with the medial part of the prostheses [39]. If prominent in size, sternalis can impose aesthetic implications in the form of chest or breast asymmetry [39], or even medial deviation of the ipsilateral nipple-areola complex [40].

Clinical Correlations

Clinically, sternalis is not related to any symptoms. It can be found accidentally during surgery of the anterior chest wall without any significant alterations in surgical times [37], while other support that surgical times might be affected significantly [39]. Although, it is reported that may cause changes in the electrocardiogram [6] and may cause breast or chest asymmetry [39]. Also, it may cause diagnostic dilemma during breast surgery, anterior chest wall plastic reconstruction, and interpretation of mammograms and CT scans of the anterior chest wall [6, 8]. Its appearance in the anterior chest wall can be confused with tumors such as benign and malignant breast lesions, extra-abdominal desmoid tumors, granular cell tumors and other pathology like diabetic mastopathy, abscesses, hematomas, sclerosing adenositis, lymphadenitis, fat necrosis, and surgical scars [31, 41]. In cases of breast carcinoma, the presence of sternalis muscle may interfere with the pathology in the region as the depth, at which the internal mammary nodes are irradiated, especially those lesions infiltrating the medial quadrants is changed.

Identifying sternalis in plain mammographies can be challenging. In a large study on 32,000 women examined it was identified successfully in only four cases [31]. On further assessment, ultrasonography of the breast could confirm the initial diagnosis and provide the dimensions of the aberrant tissues [42]. Row MDCT or MRI scans are useful imaging modalities for precise identification of aberrant anterior chest wall musculature [16, 17, 43]. Any sternalis presence should be recognized promptly during mastectomy, whereas blood supply to the aberrant muscle must be spared. During positioning of tissue expanders for breast reconstruction should take into account the configuration of sternalis, if present. Failure to adapt surgical technique may lead to improper placement of the prostheses, as well as insufficient muscle coverage [44].

Sternalis can co-exist with other anomalies like vascular variants of the kidneys, testis, adrenal glands, aberrant anatomy of the hepatobiliary system [44], or other chest muscles deformities

[18, 45]. Furthermore, a positive correlation of sternalis with anencephaly has been reported in fetuses [34].

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Cartilage Histology in Chest Wall Deformities

5

Felix Muller, Christoph Brochhausen, Volker H. Schmitt, and Salmai Turial

Introduction

Chest wall deformities represent a heterogeneous group of malformations of the thoracic skeleton. From the pathophysiological point of view chest wall deformities associated with syndromes such as Jeune-Syndrome or Poland-Syndrome should be distinguished from malformations without such a syndromal background. The latter group is based on pectus excavatum and pectus carinatum. Pectus excavatum represents the most frequent chest wall deformity. There are a number of surveys regarding potential clinical symptoms, several diagnostic methods and multiple variations of surgical or even non-surgical treatment options. However, the etiology of pectus excavatum and pectus carinatum is still not well understood. Until today different etiological hypotheses exist, but none of these can fully explain the appearance of pectus excavatum and pectus carinatum. The association of chest wall deformities with several connective tissue diseases led to the supposition that the cause of pectus excavatum and pectus carinatum might be an intrinsic costal cartilage abnormality. One potential reason for the weakness of etiological knowledge could be that the morphological changes in different chest wall deformities have not been systematically analysed and compared in detail. To facilitate this, our centre began to systematically analyse histomorphological cartilage specimens after open chest wall corrections. The long-term aim is to set-up a network of clinicians, scientists and patients associations to further optimize our knowledge on the pathophysiology of chest wall deformations and to give a scientific base for innovative conservative or surgical treatment options. Furthermore, the results of the morphological analysis could give a basis for future developments of potential tissue engineered treatment options for such pathological conditions.

In the present chapter the first morphological and immunohistological data from chest wall deformities are presented. Based on the presentation of the different etiological models the morphological, immunohistological and ultrastructural changes of pectus excavatum have been described. Furthermore, results of other morphological studies will be also reviewed in this chapter.

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Aetiological Theories on Pectus Excavatum and Pectus Carinatum

Even if there are several hypotheses, since its first description by Bauhinus in 16th century the formal pathogenesis of pectus excavatum and pectus carinatum is not clear until today. However, in his first description Bauhinus already gave his idea of the reason for chest wall deformities, given as a hypertonus of the diaphragm during embryological development. In contrast, during the end of the 19th and the beginning of the 20th century one of the leading hypotheses for the cause of pectus excavatum was a high intrauterine pressure to the sternum due to an abnormal position of the embryo. The idea was that either the chin [1] or the heel [2] exert pressure on the sternum. An alternative hypothesis at that time did not accept a cause of this malformation during embryological development and favoured an acquired defect due to a permanent strain on the sternum arising during the resoling of shoes, leading to the term "cobbler chest". In the early 20th century Brown made a thickened ligamentum substernale responsible for a traction force leading to pectus excavatum [3]. In this context, Sutherland hypothesised a congenital dysbalance between the anterior and the posterior muscle strands of the diaphragm leading to traction on the xiphoid process of the sternum [4].

So far, all these hypotheses on the cause of chest wall deformities focused on parameters located outside the cartilage tissue itself. There was a unique hypothesis taking into account the sternal tissue or the rip cartilage respectively and this discussed a luetic or rachitic involvement of these tissues [5]. The general view that the origin of chest wall deformities is to be found outside of the sternal tissue or that of the sterno-costal-cartilage completely changed during the second half of the 20th century. During that time the focus in pathophysiological research on chest wall deformities changed from anatomical aspects to biochemical aspects of the sterno-costal-cartilage. With that change, the sterno-costal cartilage itself and potential changes in this tissue became the focus of pathophysiological considerations of pectus excavatum and pectus carinatum. Actually, two pathophysiological hypotheses of pectus excavatum and pectus carinatum exist. One view emphasizes a altered metabolism within the sterno-costal cartilage resulting in a biomechanical weakness of the cartilage. The alternative hypothesis favoured an excessive growth of the sterno-costal cartilage. The latter hypothesis was postulated as early as 1944 [6] gathering that idea from a very early description by Flesch in 1873. According to that hypothesis an overgrowth of the sterno-costal cartilage results either in pushing the sternum in a ventral direction, leading to pectus carinatum, or pushing it dorsally, leading to pectus excavatum.

The hypotheses of metabolic disorder or cartilage overgrowth respectively opened innovative perspectives for detailed morphological analysis. However, comprehensive systematic studies of the changes of pectus excavatum and pectus carinatum at histopathological, immunohistological or ultrastructural level have failed until today. Nevertheless, there are a number of valuable case reports and smaller studies analysing different aspects of cartilage metabolism in chest wall deformities. The data from these observations give important insights into sterno-costal cartilage in chest wall deformities and thus offer innovative perspectives for further studies with a greater number of specimens in different chest wall deformities. Such comprehensive and comparative studies could give valuable evidence for new patho-physiological mechanisms at cellular and subcellular level in different chest wall deformities. Thus, the important question to be answered is whether pectus excavatum and pectus carinatum are different presentations of the same basic pathophysiological mechanism.

Cartilage Metabolism in Pectus Excavatum and Pectus Carinatum

Disturbances of cartilage metabolism could be a potential cause of chest wall deformities. This brings into focus the main elements of cartilage tissue, namely the chondrocytes and the extracellular matrix components. In this context, changes in the number of chondrocytes, in their metabolic

activity or in their cellular and subcellular integrity are of special interest. The cellular pathology of the chondrocytes in the sterno-costal cartilage could play a key role in elucidating the pathophysiological mechanisms of chest wall deformities since these cells synthesize the fibrillar and non-fibrillar extracellular matrix components. Furthermore, these cells are responsible for the balance between synthesis and degradation of extracellular matrix components and thus for the remodelling of cartilage. In particular, changes in the composition of extracellular matrix components are crucial for the mechanical stability of the cartilage, and this forms the basis of the hypothesis of mechanical weakness as a potential cause of pectus excavatum and pectus carinatum. Studies by Fokin et al. gave evidence for a variable cell quality and a disturbance of the organisation of the extracellular matrix of the cartilage in patients with Pectus excavatum. Based on molecular biological analyses the authors found an up-regulation of different collagens, such as collagen 2A (Col2A) and collagen 9A (Col9A), the up-regulation of filamin-genes as well as a down-regulation of metalloproteinases, which they interpreted as a sign of increased growth activity of the cartilage [7, 8]. However, an increased proliferative activity of chondrocytes, which would be a crucial parameter of increase cartilage growth, has not been detected until today. An experimental approach to the hypothesis of cartilage overgrowth came from Nakaoka et al. who measured the lengths of rips and sterno-costal cartilage in symmetric and asymmetric chest-wall deformities. They could not find any differences in the length of cartilage between the rips of the deepest point of sternal sinking compared to that of the other ribs. They concluded that the depression of the sternum could not be sufficiently explained by cartilage-overgrowth [9]. However, the authors clearly described the limitation of their results, namely the lack of normal control specimens. Furthermore, they highlighted the need for trials with normal sterno-costal cartilage as base-line control to test the hypothesis of cartilage overgrowth. In fact, the lack of normal sterno-costal cartilage as control material is one of the major weaknesses of hypothesis testing in histopathological chest wall deformity research. In the light of this an early theory of a combination of growth disturbance called “sterno-costal dysostosis” with a dysfunction of the respiratory musculature by Maneke remained hypothetical [10]. Even if the proposed pathophysiological model could not be proved two interesting elements remained: Maneke postulated a developmental defect in the mesenchyme, which referred to a defect during an early stage of sterno-costal development, when the condensation of mesenchymal cells directs the shape of the prospective rib and the sterno-costal cartilage. This view of an early mesenchymal disturbance would support the suggestion that pectus excavatum and pectus carinatum represent different occurrences of the same pathology. Until today this important question of whether pectus excavatum and pectus carinatum are different representations of the same pathology, or should be viewed as the result of two different pathophysiological mechanisms, is still unclear. Nevertheless, there is increasing evidence that the underlying pathological changes could be induced by metabolic changes in the cellular compartment of the sterno-costal region.

An interesting observation was made by Geisbe and co-workers in the late sixties of the last century. They described premature signs of aging in the sterno-costal cartilage of pectus excavatum patients, and this lends support to the hypothesis of metabolic disturbances as a crucial pathophysiological parameter for pectus excavatum [11]. They found degradation of the extracellular matrix, especially a demasking of parallel-aligned fibre components, which represents a sign of premature aging of the cartilage. Interestingly, they also observed a correlation between the degree of matrix degradation and the severity of the pectus excavatum. In general, according to their results the authors stated that in the sterno-costal cartilage of pectus excavatum specimens a premature aging of one decade became obvious. These morphological changes could be correlated with an increase in the activity of catabolic enzymes for mucopolysacharides such as hexoaminidase, glucuronidase and carboxypeptidase, all enzymes involved in the

process of demasking of fibres within cartilage. Furthermore, the authors were able to demonstrate in a rat animal model that a mobilisation of the sternum from the ribs induced a funnel like deformation within 80 days [11]. These findings indicate that weakness or instability of the thorax as a result of attraction forces and pressure differences between the visceral and parietal serosa could play a role in the pathogenesis of pectus excavatum. However, in their experiments they were not able to clarify whether the metabolic defect within the cartilage is a result of inadequate mechanical forces. The concept that mechanical forces could play a role in the pathomechanisms of chest wall deformities is further supported by observations that both clinical and experimentally induced scoliosis lead to deformation of the thorax [12–14]. Nevertheless, how these conditions are transformed into metabolic disturbances of the chondrocytes remains a matter of debate.

A potential explanation for metabolic disturbances was presented by Rupprecht and co-workers who demonstrated abnormalities in the content of trace elements in cartilage from patients with pectus excavatum, namely a decrease of zinc and an increase of magnesium and calcium [15]. Since zinc represents an important trace element in the metabolism of cartilage the authors hypothesized that in these specimens a replacement of zinc by magnesium and calcium appears to occur. The authors also stated that this trace element dysbalance could be the cause of the metabolic lesion seen ultrastructurally in cartilage from pectus excavatum patients as dilatation of the endoplasmic reticulum, Golgi vesicles, and cristae of the mitochondria as well as abnormal collagen fibres [15]. On the topic of collagen abnormalities Hecker and co-workers were unable to find any differences in the molecular weight or the amino-acid sequence between collagen II in normal cartilage and in cartilage of pectus excavatum patients [16]. The conclusion that trace element metabolism could play a role in the pathophysiology of pectus excavatum is in line with experimental data from O'Dell and co-workers, who demonstrated that a depletion of zinc in the nutrition leads to a decreased activity of chondrocytes [17]. It still has to be shown that mechanical stress can influence trace element metabolism in cartilage and what the underlying mechanisms for these influences are. Furthermore, detailed study is required to see if mechanical stress and trace-element disturbances in congenital pectus excavatum and pectus carinatum represent two elements of the same pathophysiological cascade or if they are part of independent pathomechanisms which could both lead to these chest wall deformities. This argument raises the question of whether there are more than two independent pathomechanisms which could lead, for example at different stages of thorax development, to pectus excavatum and pectus carinatum. In this context, the light microscopic results from the study of Rupprecht and Freiberger are of special interest. They described blood vessels within the sterno-costal cartilage of pectus excavatum specimens and interpreted this as a developmental vestige as a result of a disorder of the sterno-costal cartilage [18]. In contrast to these findings Craatz and co-worker described small blood vessels not only in the specimens from pectus excavatum patients but also from normal sterno-costal cartilage of adults [19].

Feng and co-workers proposed a reduced biomechanical stability in cartilage in pectus excavatum patients as a result of disturbances in collagen distribution and composition [20]. In their experiments they showed a markedly reduced biomechanical quality of cartilage from pectus excavatum specimens in response to tensile force, pressure and flexure in comparison to normal sterno-costal cartilage. These results could represent the missing link in the theory that defective cartilage is an important requirement for the subsidence of the cartilage in pectus excavatum. In a recent morphological study by David and co-workers the changes in the extracellular matrix of pectus excavatum specimens was described as an immature pattern with a normal cell/matrix ratio. It becomes evident in view of these observations that future studies need to clarify whether the changes are indeed comparable with immature sterno-costal cartilage or if they only resemble immature cartilage, but are actually a result of pathophysiological degradation [21]. However, all existing studies on the pathomechanisms of pectus excavatum and

pectus carinatum have been queried by conclusions from a recent analysis by Tocchioni and coworkers [22]. From their morphological analysis of cartilage from pectus carinatum specimens, these authors stated that matrix composition as well as cell number and size of costal cartilage are all dependent on the subject and not on the disease. In addition, they argued that the microscopic organization of cartilage correlates with the stabilization of the defective shape but not with the onset of the deformity [22].

In summary, a review of the various studies in the literature indicates that even after half a century of research into the pathophysiology of chest wall deformities evidence for a definitive formal pathogenesis with elucidation of the cellular pathomechanisms is minimal. In the following contribution histological, immunohistological and ultrastructural findings will be presented to illustrate research trends in that field and to present innovative aspects of current research on cellular pathology in pectus excavatum and pectus carinatum.

Morphological Findings in Pectus Excavatum and Pectus Carinatum

Since pectus excavatum is much more frequent than pectus carinatum histological and immunohistological findings in sterno-costal cartilage from pectus excavatum patients will be demonstrated in the present description.

Histological Characterization of Pectus Excavatum Specimens

In our own studies sterno-costal cartilage histomorphologically showed a typical pattern with few layers of elongated chondrocytes in the periphery (Fig. 5.1a) and more or less cell-rich central areas with round-shaped chondrocytes arranged in typical cell cluster (Fig. 5.1b). The elongated chondrocytes in the periphery were arranged parallel to the surface of the cartilage and had uniform small, round nuclei. In the central portion of the sterno-costal cartilage the chondrocytes were arranged in numerous cell clusters in diffuse distribution, but also single, typically round-shaped chondrocytes were evident. The single cells as well as the cell clusters showed uniform round nuclei without any signs of atypia. The extracellular matrix showed a heterogenous pattern. Accentuated in the central portion of the specimens hyaline extracellular matrix alternates with areas of mucoid matrix degeneration (Fig. 5.1c) and fibrillar demasking of the matrix (Fig. 5.1d). The distribution of these signs of degradation was heterogeneous and did not show a relationship with the distribution of single cells and cell clusters. Furthermore, in our cases of pectus excavatum blood vessels could be identified within the periphery and central areas of the sterno-costal cartilage (Fig. 5.1d), which confirmed former findings by Rupprecht and Freiberger [18] as well as these from Craatz and co-workers [19]. The distribution of vessels within the cartilage was variable without any correlation with the peripheral or central zone of the cartilage. In addition, no relationship was seen between the numbers of blood vessels and the content of single cells or cell clusters. Interestingly, in our experiments the vessels were accompanied by variable amounts of a cell- and fiber-rich soft tissue. To clarify whether such blood vessels represent developmental relicts, as suggested by Rupprecht and Freiberger, or if these are normal variants, as interpreted by Craatz, there should be a prospective study with a suitable number of specimens and the relevant clinical data. In our own experiments it seems that the quantity of vessels in pectus excavatum is higher than that found in fetal sterno-costal cartilage. Furthermore, vessels in fetal sterno-costal cartilage showed an accompanying fibrous tissue to a significantly lesser extent. We also found interesting differences in the arrangement of cells between cartilage from pectus excavatum patients and fetal sterno-costal cartilage. Thus, in the latter a homogeneous distribution of partly round-shaped, partly elongated chondroid cells could be found. The inhomogeneous pattern of cell distribution with numerous clusters of chondrocytes was not evident in the fetal tissue. In our experiments we used fetal sterno-costal cartilage to compare its parameters with that in pectus excavatum and pectus carinatum specimens, since the pathophysiological hypothesis of a developmental disorder in the sterno-costal

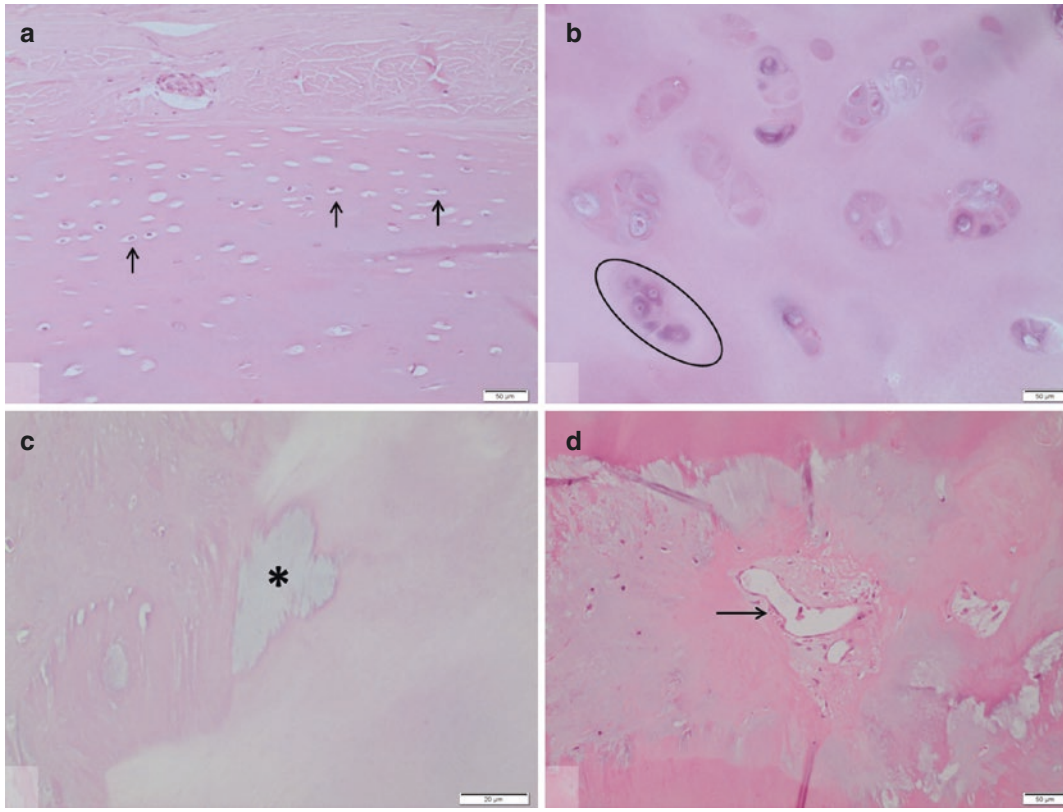

Fig. 5.1 Histological characterization of sterno-costal cartilage from pectus excavatum specimens: The peripheral zone of the cartilage is characterized by flattened elongated chondrocytes (**a**) whereas in the central portion round shaped chondrocytes arranged in typical clusters were evident (**b**). Accentuated on the central portion of the specimens mucoid matrix degeneration of the extracellular matrix (**c**) and small blood vessels with surrounded by fibrous tissue (**d**) were seen

cartilage remains unsolved to the present day. However, as described above our findings indicate relevant differences between the sterno-costal cartilage of pectus excavatum patients and fetal sterno-costal cartilage. In this context it is also important to take into account that in our experience there are no differences in the histomorphological changes in pectus excavatum and pectus carinatum.

Immunohistological Findings in Pectus Excavatum Specimens

Some interesting pathophysiological hypotheses could be corroborated by immunohistological techniques. One of the most interesting hypotheses is that of cartilage overgrowth. Even if the cellularity in cartilage is not as high as in other types of tissue, it contributes to growth in a double sense, as chondrocytes are responsible for the extracellular matrix components and as they are the unique cellular element of this tissue. That means that overgrowth should be represented not only by activated chondrocytes synthesizing large amounts of extracellular matrix components but also by an appropriate level of proliferation of these cells. The proliferative activity of cells can be detected immunohistochemically by use of antibodies against Ki-67, which is an antigen corresponding to a nuclear nonhistone protein expressed by cells in all phases of proliferation, namely the G1, G2, M and the S phase. In a fully automated, standardized staining protocol using monoclonal antibodies (Dako, Germany) we were not able to detect proliferative cells, neither in the pectus excavatum specimens (Fig. 5.2a), nor in

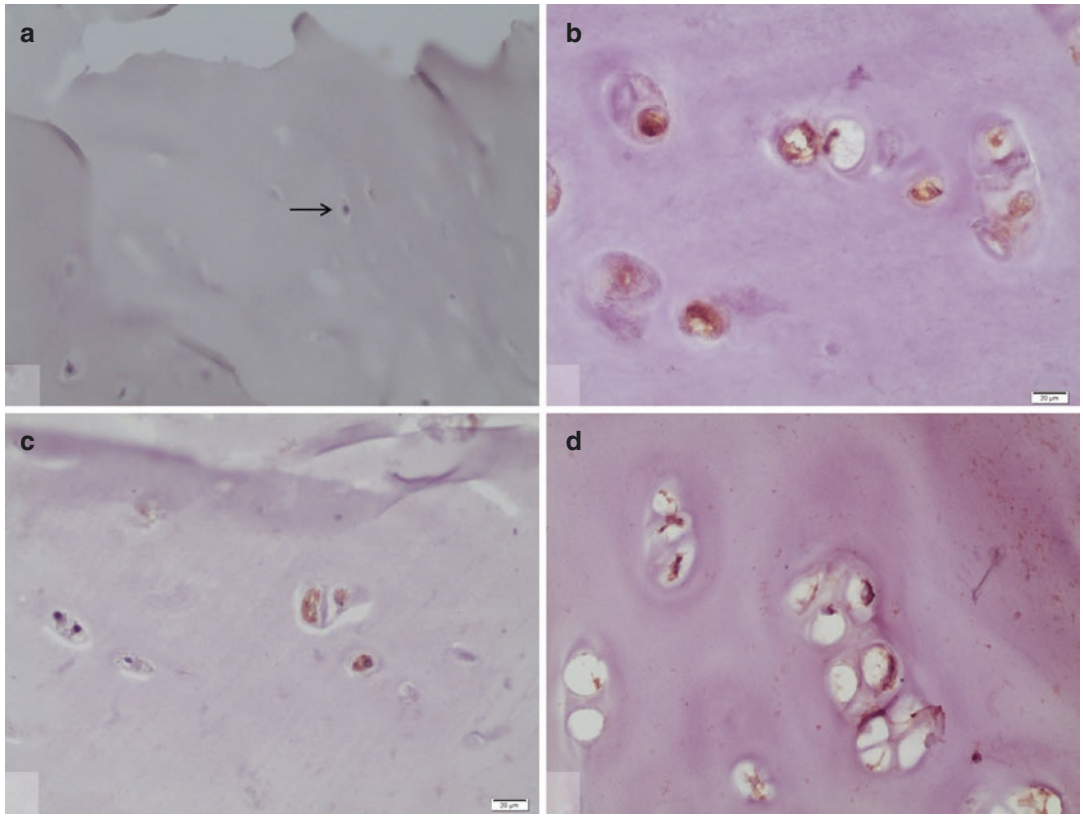

Fig. 5.2 Immunohistological findings: No positivity for Ki-67 could be detected in our specimens (**a**). The chondrocytes reacted positive for COX-2 as a sign of synthetic activity (**b**). The reaction with antibodies against EP-2 showed an inhomogeneous pattern with negative and positive cells within the specimen (**c**). In contrast, the chondrocytes reacted homogenously positive with antibodies against EP-4 (**d**)

fetal cartilage. These findings correspond with the well-known fact that mature chondrocytes and chondroid cells showed a highly reduced proliferative activity.

With respect to the synthetic activity of chondrocytes arachidonic metabolism is worthy of consideration, since several experimental studies clearly demonstrated that not only matrix synthesis [23] but also phenotypic differentiation [24] and proliferation [25] of chondrocytes are influenced by that pathway. The active molecule in cartilage metabolism is prostaglandin E_2, an arachidonic acid metabolite which is synthesized by enzymatic action of cyclooxygenase-1 (Cox-1) and cyclooxygenase-2 (Cox-2). The biological effects of cyclooxygenases are mediated by transmembrane G-protein-coupled receptors, of which 4 isoforms are described, namely prostaglandin E_2 receptor- 1 (EP-1), prostaglandin E_2 receptor- 2 (EP-2), prostaglandin E_2 receptor- 3 (EP-3), prostaglandin E_2 receptor- 4 (EP-4). Following receptor binding, inositol-triphosphate and cyclic-AMP as second messenger regulate matrix synthesis and proliferation of chondrocytes. Pectus excavatum specimens showed in our investigations no expression of cyclooxygenase-1 and a moderate expression of Cox-2 (Fig. 5.2b), accentuated in the central areas, indicating metabolic activity in this area. However, the expression pattern of cyclooxygenases showed no differences between the pectus excavatum cases and fetal sterno-costal cartilage. Regarding the expression of prostaglandin receptors pectus excavatum specimens and fetal sterno-costal cartilage were negative for EP-1, whereas both reacted positively with

antibodies against EP-2 (Fig. 5.2c) and EP-3. This was more evident in the central portions of the specimens. A marked difference in the expression pattern of prostaglandin receptors was found with EP-4 expression (Fig. 5.3d), which was positive in the center of the pectus excavatum specimen but negative in fetal sterno-costal cartilage. Since experimental data from cultured articular cartilage chondrocytes demonstrated the stimulation of catabolic effects via EP-4, our findings indicate a potential role of EP-4-mediated mechanisms in the pathogenesis of pectus excavatum and pectus carinatum as that would be a mechanism to explain matrix degradation. The stimuli for this EP-4 expression and its functional role in matrix degradation in pectus excavatum and pectus carinatum should be addressed in future *in vitro* studies.

Ultrastructural Changes in Pectus Excavatum and Pectus Carinatum

First ultrastructural changes of cartilage in pectus excavatum were demonstrated by Rupprecht and co-workers [15], who described ultrastructural alterations both in chondrocytes and in extracellular matrix components. On the cellular level the authors found mainly abnormal chondrocytes with intracytoplasmic lipid droplets, dilated cisterns of the endoplasmic reticulum and dilated vesicles of the Golgi apparatus. Furthermore, swollen mitochondria were detected as well as spots of glycogen and diffuse osmophilic deposits, interpreted as calcium-deposits. The extracellular matrix was characterized by abnormal matrix components with signs of disintegration, such as diffuse osmophilic spots of proteoglycans and irregularly arranged, partially thickened collagen fibres.

In our own studies we could confirm signs of disintegration of the extracellular matrix with a disruption of the fibrillar architecture and areas of dehiscent, partially thickened collagen fibers. In a portion of the thickened fibers there was a loss of the characteristic fiber striation. Furthermore, a loss of proteoglycan granules in the extracellular matrix was detectable. On the cellular level we also observed intracellular lipid droplets in chondrocytes of the periphery and the central portion of specimens, partially abnormal mitochondria with rarefication of cristae and dilated endoplasmic reticulum.

A highly interesting finding was the detection of centrioles in chondrocytes of the sterno-costal cartilage from pectus excavatum specimens (Fig 5.3c). Physiologically, centrioles are the core of primary cilia, which in the case of chondrocytes have important functions in mechanotransduction [26, 27]. In this context, experimental data from immortalized chondrocytes clearly indicates the role of primary cilia as essential organelles in mechanotransduction, since it could be demonstrated that the presence of primary cilia in chondrocytes is a prerequisite for an increased synthesis of glucosaminoglycans and collagen II in response to mechanical stress [28]. Taking these findings into account, the lack of the cilium in chondrocytes could be the reason for the disturbance of the extracellular matrix composition. As a consequence, this could explain the biomechanical weakness of sterno-costal cartilage from pectus excavatum patients, which is clearly evident [20]. In contrast to the lack of cilia we have observed complete cilia in chondrocytes from fetal sterno-costal cartilage. However, in our fetal and pectus excavatum specimens not all chondrocytes showed primary cilia or centrioles. A potential explanation for this finding could be the presence of so-called pacemaker cells, which coordinate the matrix synthesis in response to mechanical stress. Actually, we have evidence for this hypothesis from exhaustive morphological analysis of our samples obtained from pectus excavatum patients.

The ultrastructural basis of cartilage changes in pectus excavatum with conclusive interpretations and the potential transformation as a general pathophysiological model for further chest wall deformities are complicated by our finding of crystalline inclusions in single chondrocytes in a case of pectus carinatum [29]. In this we could not detect any primary cilia or centrioles within the chondrocytes. However, the finding of crystalline inclusions (Fig. 5.3e, f) especially in mitochondria is interpreted as a morphological

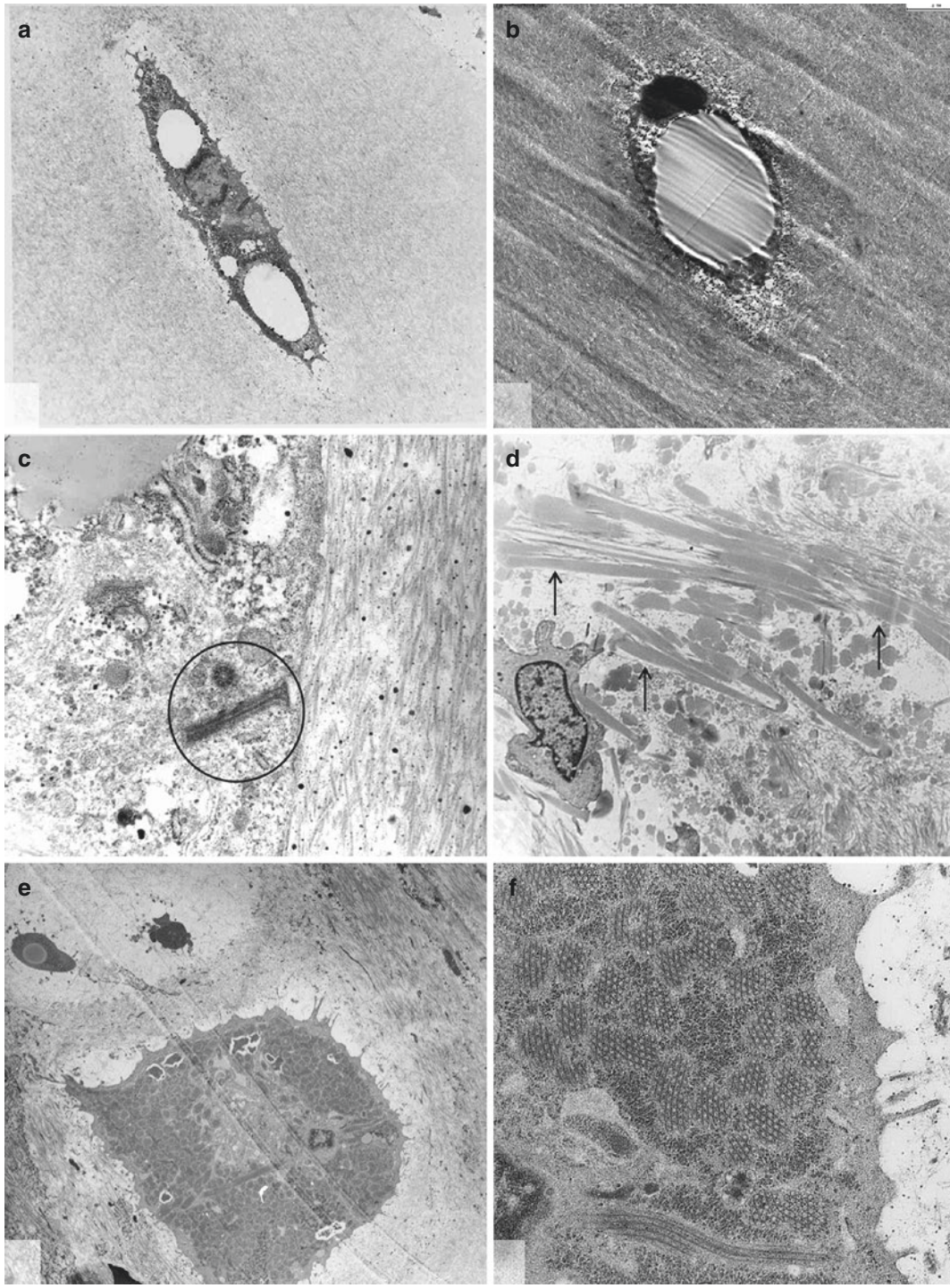

Fig. 5.3 Summary of the ultrastructural findings: Detection of lipid-droplets in the chondrocytes of the periphery (**a**) and in the center of the cartilage (**b**). In a case of pectus excavatum centriols (O) without adjacent primary ciliar were seen in a chondrocyte in the periphery (**c**). The extracellular matrix showed irregular, partial thickened collagen fibres (**d**). In a case of pectus carinatum chondrocytes with crystalline inclusions were evident (**e**, **f**)

correlate for disturbances in cellular metabolism, in particular enzymatic activity. As described above, biochemical studies have demonstrated decreased levels of zinc and increased levels of calcium and magnesium in the tissue of chest wall deformities. Since zinc-dependent enzymes and zinc finger proteins are involved in cartilage formation [30], the dysbalance of zinc, calcium and magnesium could play an important role in the pathomechanisms of pectus carinatum, confirming the hypothesis of a metabolic lesion as a relevant pathogenetic factor.

Taking together the ultrastructural features in pectus excavatum, namely the presence of centrioles and the absence of the cilium, and those in pectus carinatum as seen by the presence of crystalline inclusions, it is evident that detailed studies are still required to clarify the issue of whether the cellular pathology in both entities and thus the formal pathogenesis is different.

Future Perspectives in Pathophysiological Research of Pectus Excavatum and Pectus Carinatum

The present contribution clearly demonstrates that after reviewing the different histological, immunohistochemical and ultrastructural changes the cellular pathology of pectus excavatum and pectus carinatum remains incompletely understood. Furthermore, it becomes obvious that despite various experimental and clinicopathological studies the formal pathogenesis remains hypothetical. So far, no applied methodological setting has been able to clarify the pathophysiology and the pathomechanisms of pectus excavatum and pectus carinatum. Moreover, one could conclude that all the different methods and experiments to date only serve to confirm the diversity of the findings. In view of this the question arises whether or not there is really one pathophysiological mechanism leading to pectus excavatum and pectus carinatum, or if the deformation of the thorax is in fact a result of independent pathological mechanisms, thus making these two deformities separate clinical entities. In this context, the different ultrastructural findings, with lack of a primary cilium and the presence of centrioles in pectus excavatum, and the presence of paracrystalline inclusions in chondrocytes of pectus carinatum are of special interest. More exhaustive analysis with a greater number of specimens is necessary to answer these questions in a conclusive manner.

One potential explanation for this diversity of findings without clear evidence for a causal cascade of cellular mechanisms is the lack of knowledge of the physiological changes of normal sterno-costal cartilage with age and within the normal fetal and postpartal development. Thus, it would be of great interest to have more information on normal histomorphology of the cartilage in this localization, the immunohistological and ultrastructural pattern of the chondrocytes in this tissue and its biochemical composition. Of special interest for a better understanding of the pathogenesis of pectus excavatum and pectus carinatum are the questions of which changes represent normal variants of the normal situation and which are pathological, but unspecific changes. For this purpose, it is important to establish the normal ratio of cells and matrix components, the normal distribution and composition of cells in detail in different age groups and developmental stages. Since this will not be possible in human sterno-costal cartilage appropriate animal models are required for such studies. An interesting finding in the literature is that in an animal model of experimental scoliosis chest wall deformities arise [12]. On the other hand, a chest wall deformity in animals can also lead to scoliosis [31]. These findings would support the hypothesis that there might not be one pathophysiology in pectus excavatum and pectus carinatus in the sense of special clinical entities, but that these chest wall deformities represent a result of complex disturbances of mechanical forces in the thoracic cage. Nevertheless, these animal models, especially the scoliosis model, could serve as a valuable source of costo-sternal cartilage for systematic structural, immunohistological and molecular biological analysis as proposed above. Furthermore, a systematic collection of cartilage from different chest wall deformities could help

establish greater cohorts for such studies in human patient material. Therefore, the set-up of an interdisciplinary network with the establishment of a tissue bank for resected specimens enriched with clinical data would be an innovative approach to accelerate comprehensive research projects, which hopefully will lead to a better understanding of the formal pathogenesis and cellular pathology of pectus excavatum and pectus carinatum.

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Bone Mineral Density as a Marker for Pectus Correction

6

Manabu Okawada, Geoffrey J. Lane, and Atsuyuki Yamataka

Introduction

Pectus excavatum (PE) is the most common congenital chest wall anomaly (90 %) encountered, followed by pectus carinatum (5–7 %), and other less common anomalies (5 %). PE occurs in an estimated 1 in 300–1000 births, with male predominance (male-to-female ratio of 3:1). The condition is typically noticed at birth, and more than 90 % of cases are diagnosed within the first year of life. Worsening appearance and onset of symptoms are usually reported with the onset of the teenage growth spurt [1]. Patients present most commonly with a combination of esthetic concerns and restricted pulmonary function causing exercise-induced dyspnea [2].

For the past half century, PE has been corrected by subperichondrial removal of the offending costal cartilage, mobilization of the sternum, and stabilization; the technique was first described by Ravitch in 1949 [3, 4]. In the late 1980s, Donald Nuss began to use a minimally access approach by temporarily placing a convex metal bar substernally through small, bilateral incisions. It is associated with less morbidity than the traditional Ravitch repair, and because it is effective in the long term, the Nuss procedure has become widely accepted as the standard technique for the surgical treatment of PE [5–12].

However, the Nuss procedure is not free from complications. Since the thorax is forcibly distorted by the placement of correction bars, patients often develop serious postoperative pain with ineffective sputum evacuation that can cause pneumonia. In fact, if postoperative pain is severe enough the bars may need to be removed. Many authors advise that a pectus bar (PB) should be removed 2–3 years after insertion, to prevent abnormal rib growth although there is no objective basis for this. Thus, there are no established indications for PB insertion and removal available [8, 13].

Bone Mineral Density (BMD)

We hypothesized that bone mineral density (BMD) might be a feasible marker for determining the timing of PB insertion and removal and conducted a study of 23 PE patients (17 male, 6 female) who had PB removal. Age at PB insertion/removal, duration of insertion, clinical outcome, and BMD were assessed. Clinical outcome was determined subjectively by a single surgeon; excellent = no depression, good = mild depression, fair = moderate depression, and poor = recurrence requiring repeat intervention. BMD was measured using dual-energy X-ray absorptiometry

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(QDR-2000 Hologic Inc, Bedford, MA, USA) because of low radiation exposure, greater precision, and accuracy [14–18] according to a technique that is described elsewhere [17]. In this study, we measured BMD supine using L2 through L4 (Fig. 6.1).

In all 23 patients, BMD was examined just prior to insertion of the PB (in-BMD) and just prior to removal of the PB (out-BMD). We calculated %BMD by dividing subject BMD by standard mean BMD for age and sex matched healthy Japanese children [19]. i.e., %BMD = 1.0 is normal (Fig. 6.2). In our series, age at insertion ranged from 4.3 to 12.7 years and age at removal ranged from 6.3 to 14.1 years. Duration of insertion ranged from 1.4 to 3.9 years. Clinical outcome was described as excellent in 7, good in 11, fair in 3, and poor (recurrence) in 2. Mean ages at insertion/removal according to outcome are shown in Table 6.1. An interesting point is that mean age of insertion/removal in the poor (recurrence) group was younger than for the other 3 groups (excellent, good, and fair) (p = NS). In the 2 cases with recurrence, PB was inserted at 5 and 4 years of age and removed at 6 and 8 years of age, respectively. In these 2 cases NP was performed earlier than usual because of clinical indications (obvious chest pain and respiratory distress).

The dotted lines in Fig. 6.2 show standard mean BMD for age and sex matched healthy Japanese children. As shown in Fig. 6.2, the majority of the 21 subjects in the excellent, good, and fair outcome groups had BMD at the time of PB removal close to standard mean values. However, out-BMD in the 2 subjects in the poor (recurrence) outcome group (0.57 and 0.58 g/cm^2 respectively) was much lower than standard, and also lower than the subjects in the other 3 groups.

Figure 6.3 shows %BMD at the time of PB removal according to subject group. Again, both BMD and %BMD were below normal in the 2 subjects in the poor (recurrence) outcome group. In the 9 cases with no recurrence, PB removal was between 6 and 8 years old and BMD was normal and %BMD was higher than in the 2 cases with recurrence (0.88 and 0.90). However, this difference was not statistically significant

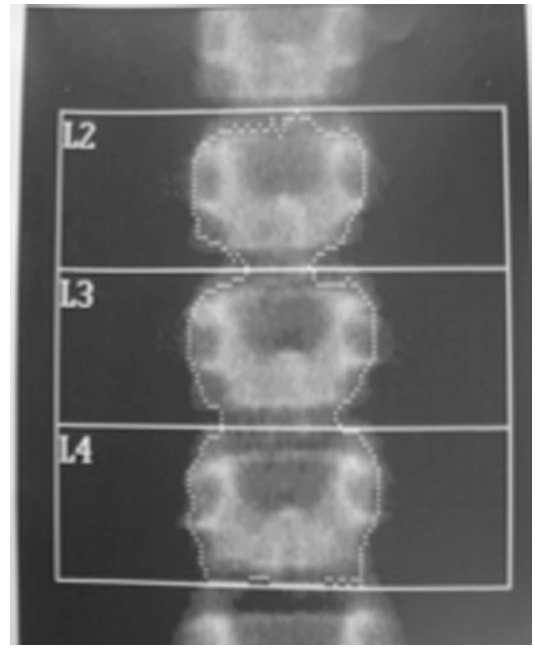

Fig. 6.1 BMD measured at L2 – L4 is regarded to reflect bone status throughout the body using dual-energy X-ray absorptiometry (QDR-2000 Hologic Inc, Bedford, MA, USA)

(p = 0.08). Both poor outcome cases had re-do NP (PB reinsertion) at the ages of 9 and 8 years respectively, and in both, PB was removed 2 year later. Outcome was excellent in both cases, and interestingly, BMD at PB removal after re-do NP were normal (0.72 and 0.70, respectively), meaning their %BMD were closer to 1.0 (0.95 and 0.97) than previously (0.88 and 0.90).

Determinants of BMD in healthy persons are genetic ethnic factors, hormonal status, calcium intake, physical activity, and weight. In fact 46–62% of BMD variance has been attributed to genetic factors. Generally, BMD is higher in blacks than whites and lower in Chinese and Japanese. Total body BMD is lower in girls of Asian background compared with Caucasian girls that may be related to calcium intake. A widely held theory is that blacks have greater skeletal muscle mass than whites genetically, and this greater mass causes added stress on bone, thereby resulting in higher BMD. African population are also believed to have higher circulating estradiol concentrations which may also increase

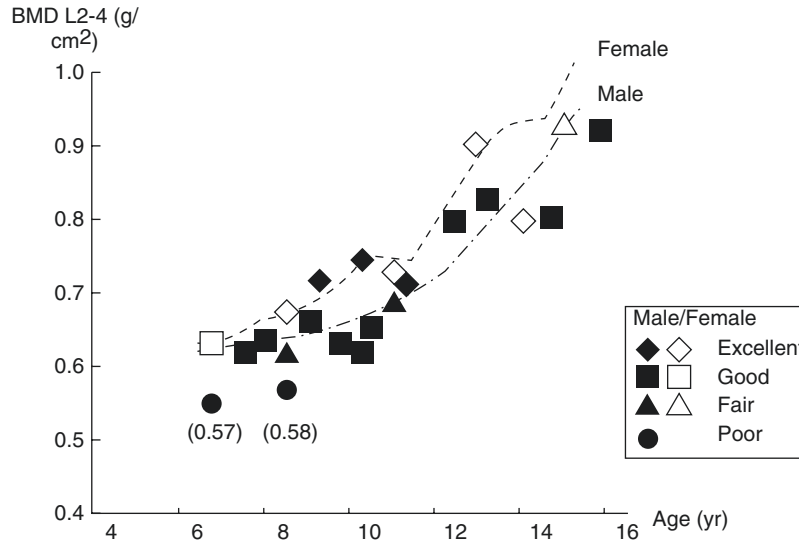

Fig. 6.2 Standard mean BMD for age and sex matched healthy Japanese children. BMD of patients in the excellent, good, and fair outcome groups were close to standard BMD

Table 6.1 Mean age at PB insertion/removal according to outcome

	Mean age (yr)	
Clinical outcome	PB insertion	PB removal
Excellent (n = 7)	8.4 ± 2.5	10.7 ± 2.1
Good (n = 11)	7.5 ± 2.7	9.5 ± 2.7
Fair (n = 3)	10.1 ± 2.2	12.6 ± 3.2
Poor (n = 2)	***5.1 ± 0.6***	***7.7 ± 0.5***

secretion of growth hormone, which in turn can lead to an increase in bone mass [20].

During childhood and adolescence, BMD increases until peak bone mass is reached. Peak bone mass and subsequent bone loss are important determinants of osteoporosis later in life. It is essential to know which factors influence BMD in childhood, with the goal of achieving optimal peak bone density. A few studies have shown that persons who consume greater quantities of calcium early in life achieve greater bone mass later.

During puberty, there is a large increase in BMD. Lumbar spine BMD and total body BMD increased significantly with higher Tanner stages. At the same time, growth hormones as well as sex steroid levels increase, and both have a positive influence on BMD. The influence of puberty on BMD is higher in girls than in boys. In multiple regression analysis, Tanner stage did not correlate significantly with BMD in boys, whereas in girls it was the major determinant. Estrogen is an important determinant of BMD in girls during puberty and girls with early menarche and regular periods had higher BMD. Later puberty and amenorrhea are risk factors for low BMD in girls [14].

Correlation with Pectus Deformities

Severity of deformity in PE is commonly evaluated objectively using the Haller index (ratio of transverse to antero-posterior diameters of the chest) obtained using chest CT [21]. At our institution, surgical repair is indicated if the Haller index is over 3.2, and the patient is clinically symptomatic, with chest pain and/or respiratory distress and manifestations of psychological distress based on cosmetic issues.

Although NP has gained wide acceptance for the minimally invasive repair of PE, it is associated with a recurrence rate of some 4.3% [6] and this has been generally attributed to the timing of PB insertion/removal [8]. Our data indicate that the majority of subjects with good outcome had normal BMD at the time of PB removal; whereas those in the poor outcome (i.e., recurrence) group had BMD markedly below normal. Thus, bone status appears to be related to clinical outcome in MARPE patients.

According to the standard mean BMD curve for age and sex matched healthy Japanese children, normal BMD begins to increase markedly around 8–9 years old and plateaus around 14–15 years old [19]. Figure 6.4 shows standard mean BMD results measured quarterly as dotted lines, with a plateau for males at 13 years and a plateau for females at 11 years 3 months. Thus, we postulated that the optimum age for PB insertion should be around 8–9 years old and that the optimum age for PB removal should be around 13–14 years old, because ossification appears to be too well established after the age of 15 years for NP to be clinically effective (Fig. 6.4).

The MARPE procedure is currently indicated for patients aged 5–20 years. However, from clinical experience, the ideal age is 8–12 years because, the chest wall is still flexible, stabilization of the bar can be achieved easily, thoracic epidural can be used safely, and the child is mature enough to understand the operation and postoperative instructions. In addition, when performed in younger children, the risk for recurrence later in life (particularly during puberty) appears to be increased [1]. Thus, both BMD and %BMD could be used routinely as objective markers for planning the timing of PB insertion/removal.

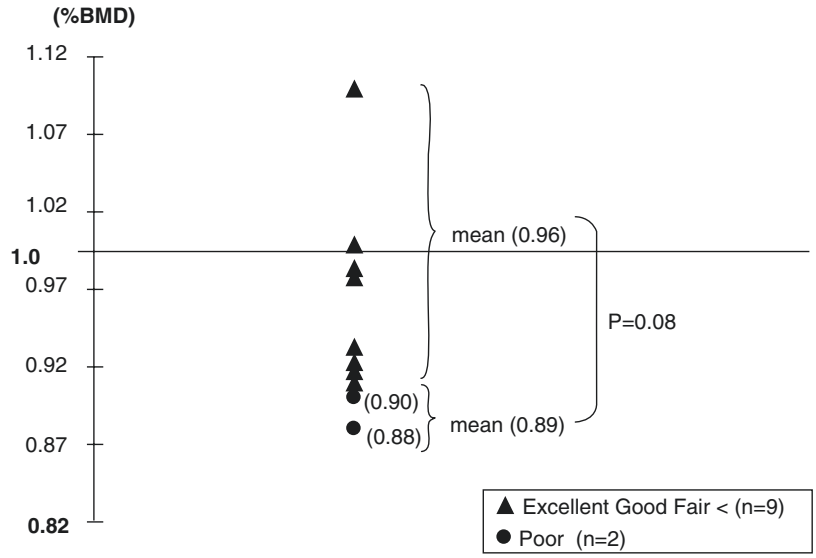

Fig. 6.3 %BMD in 11 patients who had PB removal between 6 and 8 years of age

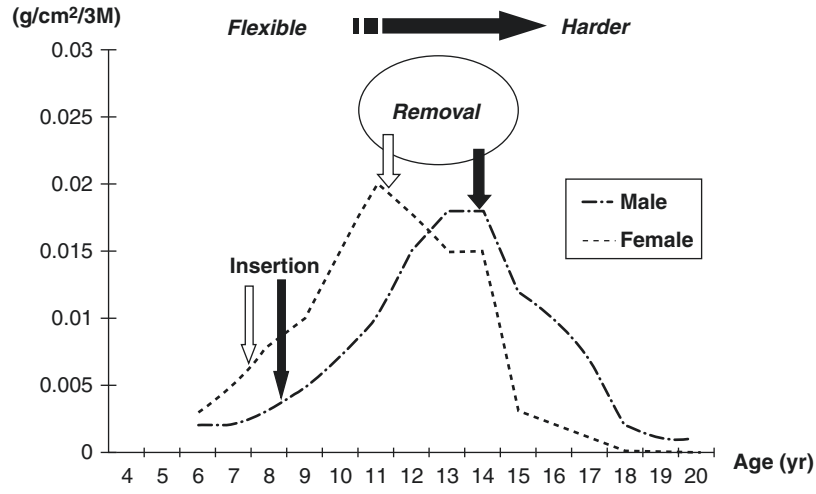

Fig. 6.4 Quarterly changes in BMD. There is one plateau for males around the age of 13 years, while females have a single plateau at 11 years and 3 months of age

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Biochemical and Histological Differences Between Costal and Articular Cartilages

7

Michael W. Stacey

Abbreviations

AFM	Atomic Force Microscope
cDNA	Complimentary Deoxyribonucleic Acid
DNA	Deoxyribonucleic Acid
ECM	Extra Cellular matrix
FCD	Fixed Charge Density
GAG	Glycosaminoglycan
MMP	Matrix Metalloproteinase
PBS	Phosphate Buffered Saline
PC	Pectus Carinatum
PCR	Polymerase Chain Reaction
RNA	Ribonucleic Acid
RT-PCR	Reverse Transcriptase Polymerase Chain Reaction
SEM	Scanning Electron Microscope
SLRP	Small Leucine Rich Proteoglycan
SNP	Single Nucleotide Polymorphism
VNTR	Variable Number of Tandem Repeats

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Introduction

Costal cartilages, a type of hyaline cartilage, are bar like structures connecting the ribs to the sternum and allows for rib cage flexibility. Unlike articular cartilage of the joints, which is only a few mm thick, costal cartilage can approach approximately 1 cm in diameter. Like other cartilage types, the body of the tissue does not have nerve or blood supplies. Costal cartilage does have a surrounding perichondrium that is vascular and provides nutrients. Nutrients diffuse into the cartilage, but it is estimated that diffusion coefficients are in the order of 200 mm, a fraction of the diameter of costal cartilage. This results in a dilemma regarding costal cartilage structure and function. Centrally located cells are deprived of oxygen and cell metabolism creates an acidic environment. Indeed, chondrocyte gene expression is up regulated in these conditions, and needs to be accounted for in future experiments; however, cells still require some minimal nutrient and gas exchange to function. Our group examined costal cartilage immunohistochemically and by scanning electron microscopy to determine the structure and protein content of costal cartilage, and genetically by examination of gene expression of key cartilage related genes.

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Cellular Distribution

Histologically, the proteins of cartilage are produced by chondrocytes that are sparsely distributed throughout the secreted extra cellular matrix (ECM), occupying only approximately 2 % of cartilage. Few studies have been undertaken on chondrocyte distribution within cartilage, yet cell density and arrangements are considered to be critical to function. Cellular clusters [1], pairs [2], and rows [3], have been reported. A more extensive study [4] in the superficial zone of articular cartilage identified complex patterns that appear to be location specific. For example, chondrocytes of the articular surface of ankle joints were as pairs, whereas strings of chondrocytes were found in half of the superficial chondrocytes of the femoral condyles of the knee joint, and circular clusters were found in the patella-femoral groove of the femur [4]. It has been suggested that chondrocyte groups form after cell division with incomplete separation within the cellular microenvironment allowing direct communication between cells [5]. In cross sections of costal cartilage removed from a patient with pectus carinatum we observed 98 % of cells were single or paired towards the periphery whereas 8 % appeared as clusters of 3–4+ cells in central regions, consistent with the notion that younger cells that have undergone one or no divisions are peripheral, with older cells having undergone more divisions being embedded more centrally. In a control sample, ~13 % of cells were in clusters of 3–4+ in central regions. Although the control sample showed cell clusters nearly twice that of a PC sample, verification of rib number and site was not possible, and thus in this instance site specific variation cannot be confirmed. No strings were observed in PC or control sample. A spatial relationship between collagen fiber alignment and cellular organization has been suggested [3], with chondrocytes running parallel to adjacent fibers. When we examined longitudinal sections of costal cartilage, we also note the presence of lacunae between the large fibrous structures [6]. The predominance of single and doublets in costal cartilage suggests cells undergo relatively few divisions. The absence of extensive strings and clusters is likely due to the different biomechanical forces experienced by costal cartilage compared to articular cartilage of joints.

The forces experienced by costal cartilages are very different to those of load bearing cartilage and a comprehensive investigation of the cellular distribution along the length of individual costal cartilages may reveal important insights into apparent weaknesses observed in patients with chest wall deformities.

Costal Cartilage Structure

Collagen Type IIAI (COL2A1) Presence in Costal Cartilage

Recent work [7] described a decrease in the biomechanical stability of costal cartilage in pectus excavatum patients and suggested a disorderly arrangement and distribution of collagen fibers. Other authors have suggested that atypical collagen fibers may be implicated in chest wall deformities [8, 9]. The arrangement of collagen fibers in costal cartilage has not been described in detail; however, highly ordered fiber formation was described in the surrounding perichondrium [10]. A lack of understanding of molecular and ultrastructural properties hampers understanding events leading to these disorders.

Collagen type II is a major constituent of hyaline cartilage, consisting of ~70 % of cartilage dry weight. It is a fibrous protein that is cross-linked to other fibers and adds structure and strength. Mutations in collagen genes have been described in many skeletal dysplasias, but no abnormalities have been described in patients with chest wall deformities. The presence of COL2A1 in costal cartilage is shown in Fig. 7.1. Staining appears to be intense surrounding cellular lacunae, with a more uniform staining pattern within the matrix. This staining is consistent with COL2A1 staining in hyaline cartilage. Negative controls showed no staining (data not shown).

Aggrecan Presence in Costal Cartilage

Aggrecan, a large proteoglycan present at 3–10 % cartilage dry weight, serves as an attachment for chondroitin and keratin sulphates,

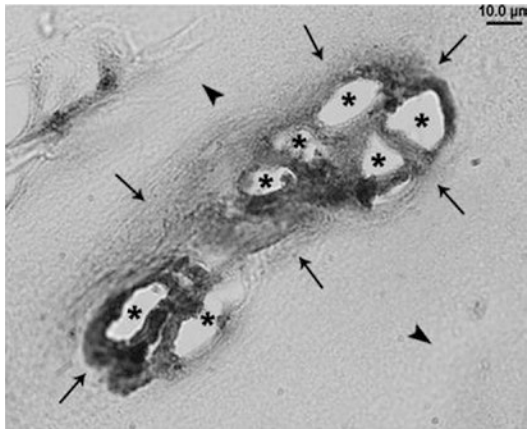

Fig. 7.1 Immunohistochemistry using a COL2A1 specific human monoclonal antibody showing intense COL2A1 staining (*arrows*) around cellular lacunae (*), with a lighter, more uniform COL2A1 stain over the matrix (*arrow heads*) [44]

highly negatively charged proteins that are responsible for the fixed charge density of cartilage. This fixed charge density (FCD) is the source of all electrochemical events in cartilage [11] and is responsible for cation movement into the tissue along with water to produce osmotic swelling that gives cartilage its unique physical properties. Abnormalities in aggrecan can result in weakened cartilage, as exemplified in osteoarthritis where breakdown of aggrecan, reduced FCD, and associated reduction in osmotic pressure, result in weakened cartilage susceptible to wear and tear.

Hypoxia or low pH has been shown to act as a trigger for aggrecan production through induction of hypoxia inducible factor 1-α and *SOX9*, as well as inhibit *COL1A1* expression [12, 13]. Similarities with inter vertebral discs are noteworthy. Cells embedded within the centrally located nucleus pulposus experience hypoxia and express aggrecan under the regulation of the hypoxia induced P13K/AKT signaling pathways via modulation of *SOX9* [14]. It appears that as cells become centrally located they, due to lack of blood supply, experience hypoxia and lower pH. Consistent with this hypothesis, we observed aggrecan deposition by chondrocytes in centrally located regions of costal cartilage compared to those cells at the periphery (Fig. 7.2). The induced negatively charged environment draws in water and cations, e.g., Na^+ that was confirmed by Electron Probe Micro Analysis in a section of costal cartilage (Fig. 7.3).

Although collagens and aggrecan are major protein constituents of cartilage, major differences between articular cartilage and costal cartilage in form and function are very apparent and thus major differences in protein content and deposition are expected. The presence and driving force of aggrecan and associated negatively charged proteins are common to both; however the arrangement of collagen fibers in response to their unique sites primarily reflect function. This draws the question as to how fibers are arranged in costal cartilage, and what proteins may play a role in the process of fibrillogenesis.

Small Leucine Rich Proteoglycans (SLRPs)

Small leucine rich proteoglycans (SLRPs) are extra cellular matrix molecules that bind strongly to collagen and other matrix molecules. They are associated with collagen fibril formation and therefore important in the proper formation of ECMs. The co-operation, sequential, timely, orchestrated action of SLRPs that shape architecture and mechanical properties of the collagen matrix and overall importance in disease is reviewed by Ameye and Young [15]. Indeed, SLRP knockout mice exhibit disorganized collagen fibers and loss of connective tissue function [16, 17]. Fragmentation of SLRPs is associated with degeneration of matrix in meniscus, knee and hip cartilage [18]. The name arises from their relative small size of approximately 40 kDa compared to ~200 kDa for the large aggregating proteins aggrecan and versican, the presence of numerous adjacent leucine rich repeats and one or very few glycosaminoglycan (GAG) side chains. SLRPs are generally expressed in a very tissue specific manner. Mechanistically, it is accepted that horseshoe shaped SLRPs interact with individual collagen fibrils by their concave surfaces, and the space inside the horseshoe accommodates a single

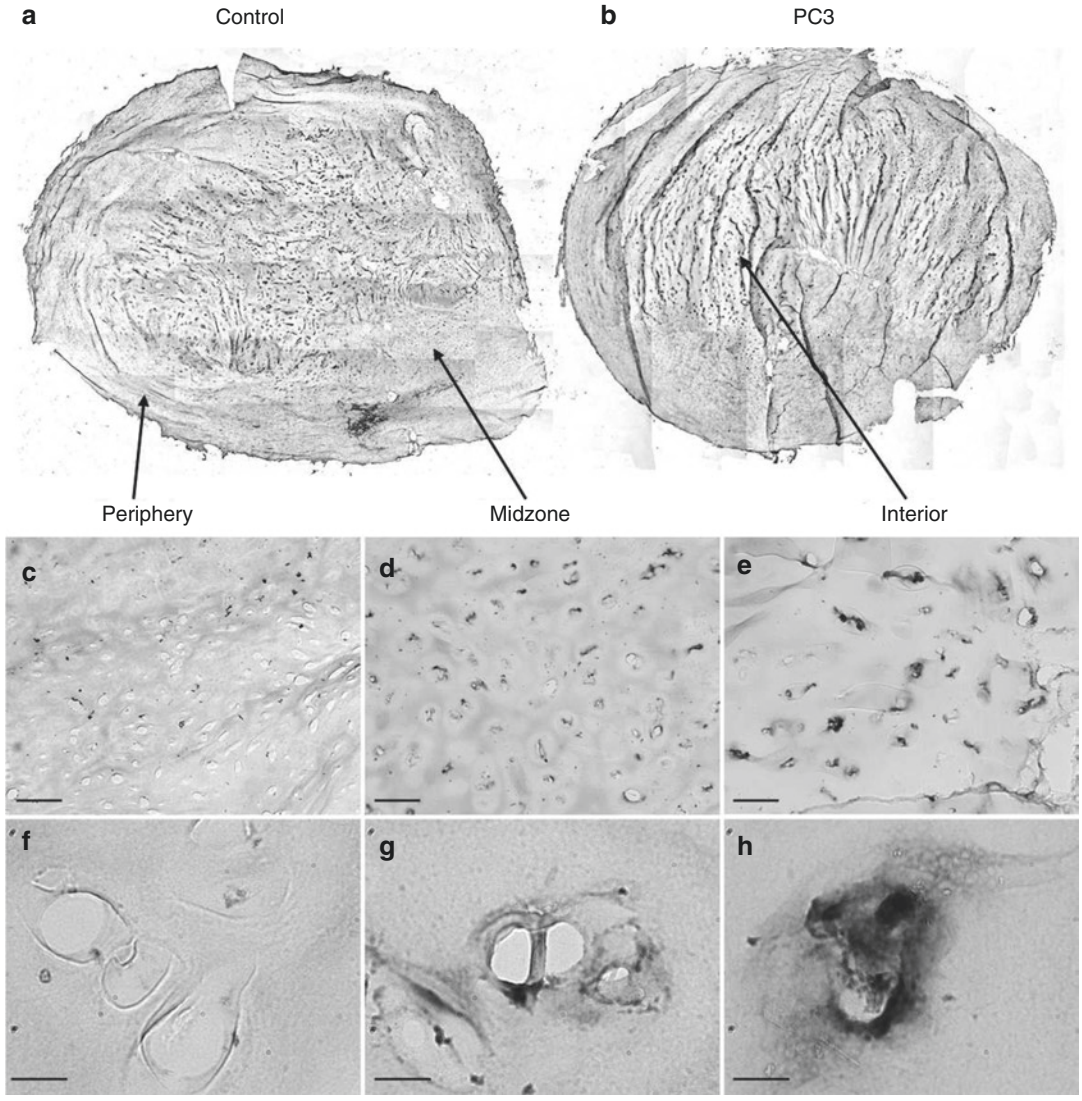

Fig. 7.2 Localization of aggrecan by immunohistochemistry in transverse cross-sections of costal cartilage. (**a**) Distribution of aggrecan in whole control section. (**b**) Distribution of aggrecan in whole PC3 section. (**c**–**e**) Distribution of aggrecan in control at 10× magnification from (**c**) periphery, (**d**) midzone, and (**e**) interior regions. Scale bars, 100 μm. (**f**–**h**) Distribution of aggrecan of aggrecan in control at 100× magnification from (**f**) periphery, (**g**) midzone, and (**h**) interior regions. Scale bars, 10 μm. Notice the localization of aggrecan becomes more intense around each lacuna in the interior compared to the periphery

triple helix of collagen [15]. Mutations in SLRPs may be important as predisposing genetic factors for diseases of the ECM.

Reports of decreased biomechanical stability of costal cartilage in pectus excavatum and suggested disorderly arrangement, distribution, and atypical collagen fibers in chest wall deformities [7–9], led us to examine the size and distribution of two SLRPs expressed in cartilage, decorin and biglycan [19]. Both regulate ECM growth and fibrillogenesis. Interestingly, both sequester TGFβ, controlling availability of this growth factor and thus growth of cartilage. The important roles of decorin and biglycan in fibrillogenesis and sequestration of TGFβ makes them interesting molecules to investigate in disorders related

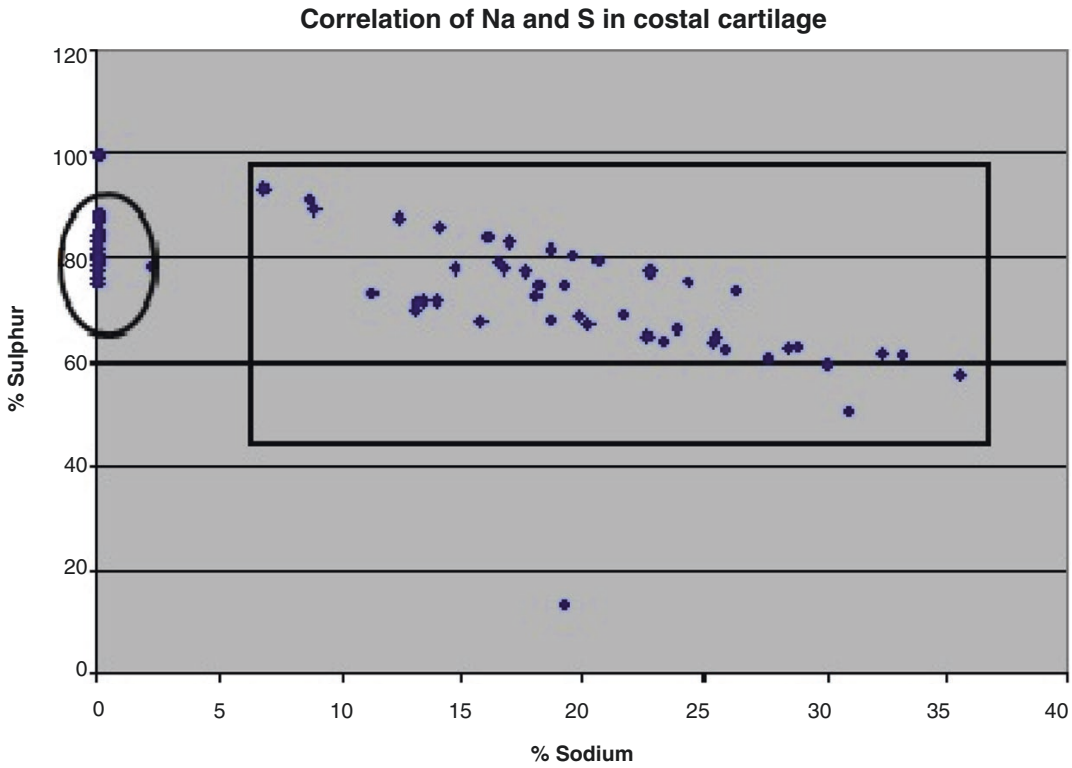

Fig. 7.3 Electron probe microanalysis of 100 equally spaced points over a transverse section of costal cartilage. Sodium is present only in the central 48 points (*boxed*), and not at the peripheral points, which contain Sulfur only (*circled*) showing that positively charged ions were drawn to central regions to achieve electroneutrality [6]

to abnormal cartilage growth and strength. There may be a mechanistic role for these two SLRPs in chest wall deformity. Decorin and biglycan are prominent class I members of the SLRP family and are homologous (57 % identity at the amino acid level) but with divergent patterns of expression. Despite their similarities, biglycan and decorin have distinct functions which may partially result from differences in GAG chains; biglycan has two chondroitin sulphate chains and decorin has one, respectively (Fig. 7.4a, b).

Biglycan deficiency has been shown to cause spontaneous aortic dissection and rupture in mice [20], a characteristic of Marfan syndrome, a syndrome known to exhibit chest wall deformities. Decorin function is consistent with functions related to fibrillogenesis [17, 21]. Null mutations lead to abnormal collagen architecture in mice suggesting a mechanistic role for these proteins in skeletal disorders.

Decorin and biglycan have different isoforms, the pro and mature forms that show differential expression [22–24]. Proforms have a 14 amino acid N-terminal pro-peptide that is cleaved in the mature forms. The abundance of pro-forms of both SLRPs is tissue and age dependent, with the mature form being expressed more highly in juvenile tissue, and the pro form in adult [25, 26]. Structurally, costal cartilage has been shown to consist of straw-like structures running parallel to the length of the tissue. Gene expression in this tissue shows high levels of decorin compared to biglycan [6]. The complex arrangement of fibers observed in costal cartilage and the role of decorin and biglycan in these structures has not been explored. We investigated the presence and distribution of the different isoforms of decorin and biglycan. This was achieved by immunohistochemistry of costal cartilage from teenage patients with pectus carinatum and an age-matched control

using antibodies to the different isoforms of decorin and biglycan [19, 22]. Our results (Fig. 7.5) show the presence of mature form of decorin and pro-and mature forms of biglycan in the interterritorial matrix in patient and control samples. Prodecorin was localized only to the cells [19].

No apparent differences were observed between an age-matched control and patient samples; however, proforms of decorin and biglycan are maintained evolutionarily, suggesting they play an important, undetermined, functional role.

Overall immunohistochemistry shows intense perilacunae staining for collagen type II, mature biglycan, and mature decorin. This is indicative of orchestration of the ECM production occurring soon after proteins are secreted from the cell. Biglycan likely initiates collagen fibril organization that is further assembled by decorin to form the large nanostraw-like structures characteristic of costal cartilage. Many other proteins will play a crucial role in the final network [19, 27]. Decorin and biglycan sequester TGFβ, controlling availability of this growth factor to chondrocytes. This needs to be proven in the environment of costal cartilage as it assumes the presence of this growth factor within the matrix, and thus a means of transport to arrive there. The components of decorin and biglycan, (leucine rich repeats, absence or presence of propeptide, and variable O-and N-linked glycosylated side chains), allow for multiple interactions with collagen fibrils and may be an important factor in cartilage stability in relation to formation of chest wall deformities. Careful analysis of the composition of GAG sidechains may be warranted as the sugar content of such chains has been suggested to play a role in binding of collagen fibers and pathology [28].

Glycosylation

Increasing attention is being given to the glycosylated side chains of proteins, particularly with relevance to disease [29]. Differences in side chains of decorin and biglycan have been described from different cartilage sites, including non-glycosylated decorin and biglycan in nucleus pulposus of intervertebral discs [30]. Decorin has a single O- and three N-linked glycosylation sites. Biglycan has two O-linked and two N-linked sites. O-linked sites are typically covalently bound by chondroitin/dermatan sulphate (Fig. 7.4). Dermatan sulphate is linked to these molecules in skin where defects of glycosylation have been described in Ehlers Danlos syndrome underlying collagen fibrillogenesis [28]. In cartilage chondroitin sulphate is covalently bound in the O-position. Glycanated side chains show length variation, with shorted chains being associated with tighter collagen fiber configuration [31]. More recent work [32] has shown that decorin may bind to one collagen fibril by its core protein and to another by its side chain. Additionally, they demonstrate that closely related side chain molecules (chondroitin-4-sulphate and chondroitin-6-sulphate) have very different effects affecting fusion and layout of collagen fibers. This exemplifies the importance of recognizing subtle variations in these molecules and their biological consequences in addition to enzyme systems that are responsible for synthesis and assembly of these molecules. Sulphate anion transporter abnormalities have been described in chondrodysplasias [29]. Interestingly, a recent report [33] describes a mutation in the GAL3ST4 gene in a single Chinese family showing dominant inheritance of pectus excavatum. GAL3ST4 is a member of the sulfotransferase family that catalyzes the C-3 sulfation of galactoses in O-linked glycoproteins. Sulfation of proteoglycans is crucial for normal development of bone and cartilage [34], and defects in genes encoding catalytic machinery responsible for sulphate biosynthesis have been reported [35, 36]. The inheritance of chest wall deformity is extremely complex [19, 37, 38], however the importance of the role of enzymes responsible for glycosylation cannot be overlooked and this report [33] may be corroborated with other genes on these pathways.

Scanning Electron Microscopy

Suggestions that atypical collagen fibers may be implicated in chest wall deformities led us to investigate ultrastructural aspects of costal cartilage. Scanning electron microscopy (SEM) was

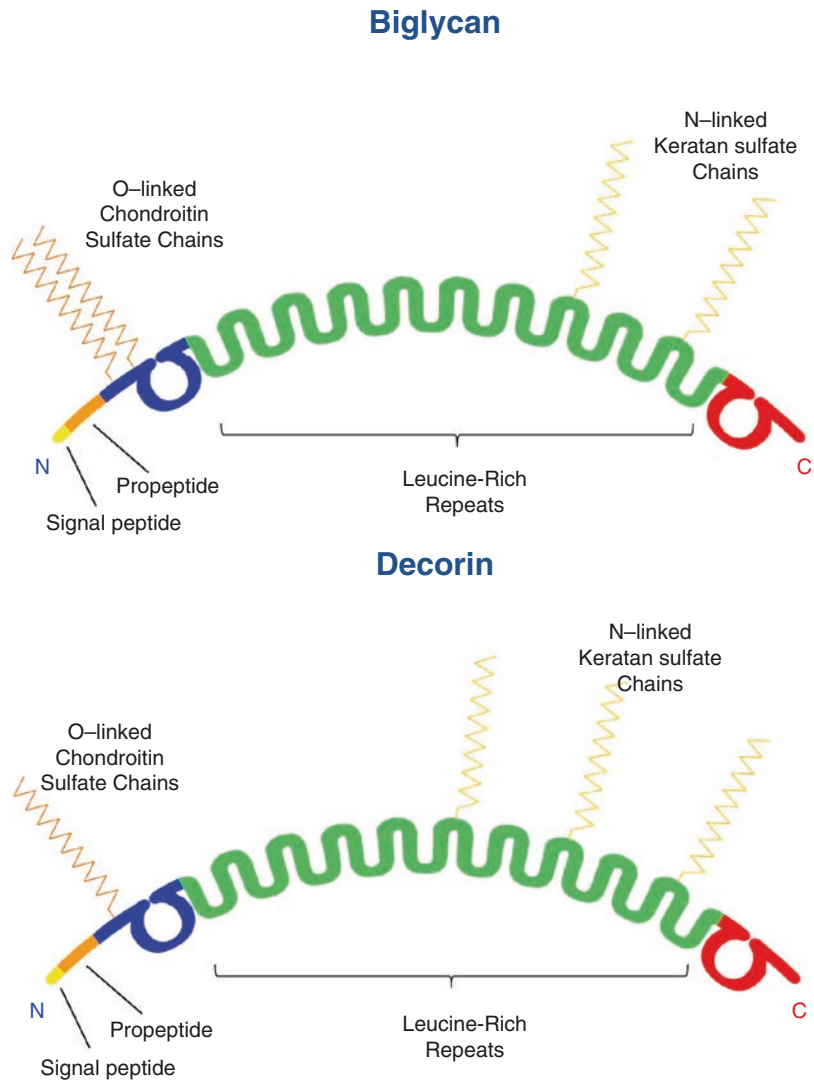

Fig. 7.4 A schematic showing salient features of biglycan and decorin

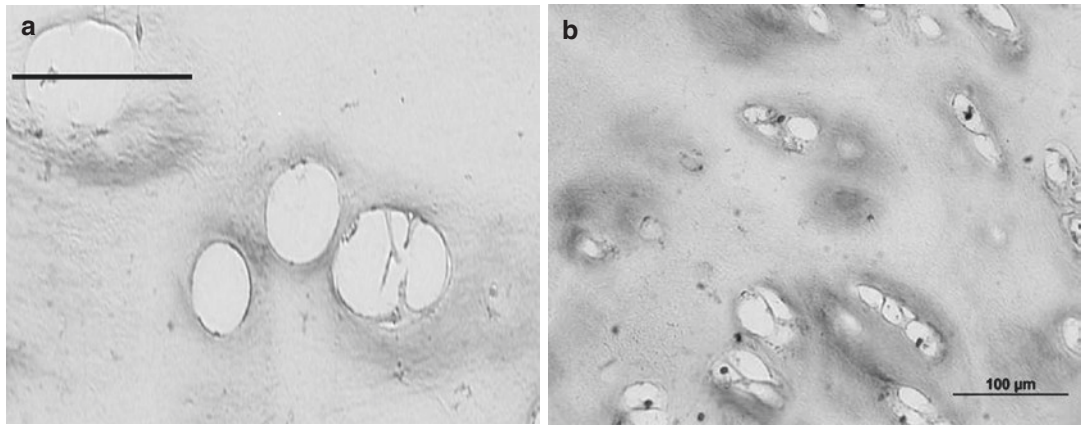

Fig. 7.5 Detection of biglycan (LF-112) (Bar 50 um) and decorin (LF-136) (Bar 100 um). Both show dark staining around lacunae, however decorin appears to be slightly more intense and more widely distributed

undertaken on a transverse section of costal cartilage to investigate distribution of collagen fibers in this tissue. Figures 7.6a, b are representative SEM images of a transverse cross-section of costal cartilage. Figure 7.6a shows a fracture in the cartilage exposing collagen fibers of approximately 600 nm diameter. Fibers come together to form an extremely large complex of many μm (arrowed) that run parallel to the length of the cartilage. Figure 7.6b is a higher magnification of the boxed area and shows that each fiber forms a nanostraw of approximately 650 nm external diameter and 250 nm internal lumen diameter. Images of longitudinal sections show a well-defined organization with bundles of collagen fibers of approximately 20 μm diameter and cellular lacunae, arrowed in Fig. 7.6c.

We measured the diameters from 150 clearly defined fibers from SEM images and found that most (51.1 %) were in the range from 1 to 100 μm. The smallest (<0.1 μm) would most-likely represent collagen fibrils, the midsize (~1 μm) would represent the microtubes and the largest (~100 μm) would be large fascicle-like structures [6].

This work shows unique ultra-structural properties of costal cartilage. The presence of straw-like structures shows that a large degree of complex extracellular matrix formation occurs. Form and function are inextricably linked in biology and the role of these structures remains to be verified. Bundles of fluid filled straws would certainly add strength while allowing flexibility during movement. Indeed, movement may be a driving force for fluid transport within costal cartilage, allowing some degree of nutrient and gas exchange for internally located cells. Assuming that the strength of costal cartilage is related to the sum of individual nanostraws, or conversely that weakness may be reflected in a more deformable nanostraw, we set out to determine mechanical properties of individual nanostraws. Young's modulus is a means to measure the elastic properties of materials that are stretched and compressed and can be described as

$$\text{Stress}\,(\text{N/m}^2)\,\text{expressed as force}\,(\text{F})/\text{area}\,(\text{m}^2)$$

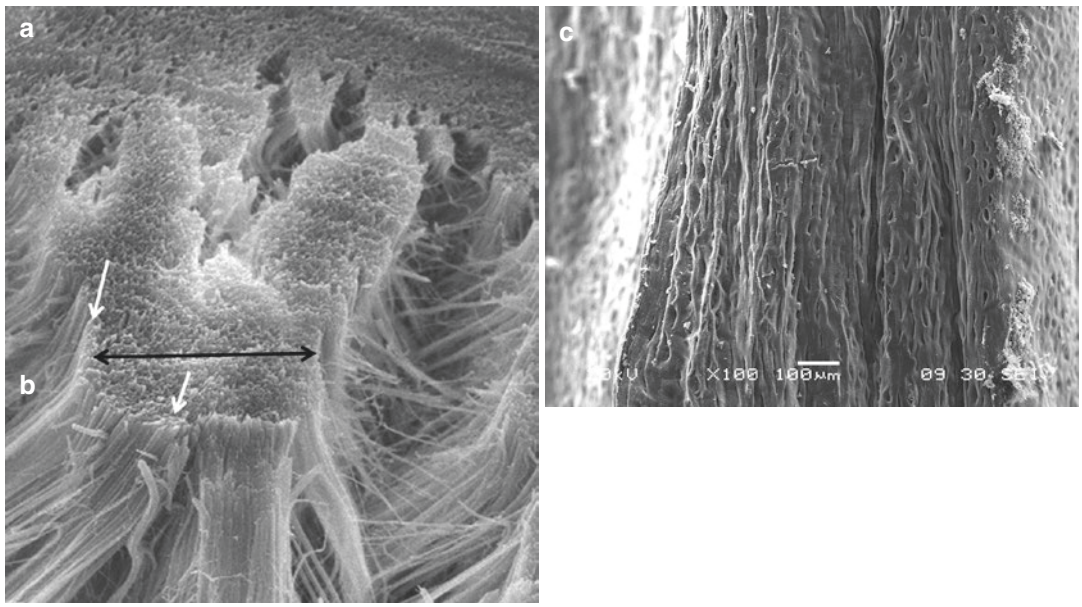

Fig. 7.6 SEM images of normal costal cartilage. (**a**) Transverse section (×2500) showing large numbers of dense fibrils running longitudinally (*arrowed*). (**b**) Magnification (×10,000) of the *boxed area* in **a** and shows the presence of collagen nanostraws (*arrowed*). Each straw is approximately 650 nm in diameter, with a lumen diameter of approximately 250 nm. (**c**) Longitudinal section (×500) showing large bundles of collagen fibers, formed from multiple collagen nanostraws, of approximately 20 μm diameter (*white arrow*)

Table 7.1 Example of a range of Young's moduli from compliant rubber to hardened steel [6]

Material	Approximate Young's modulus (10^9 N/m^2; GPa)
Bone	9
Cartilage	2.4
Tendon	5.5
Rubber	0.01–0.1
Pine wood	9
Stainless steel	180

Strain (m/m) expressed as elongation or compression of object (dL)/length of object (L).

$$\text{Young's modulus} = \text{stress/strain} = (F/A)/(\Delta L/L)$$

Stress and strain are resisted by collagen fibers and changes in the properties of collagen fibers would influence Young's modulus. Many pathological processes change tissue elasticity and is the basis of palpitation as a diagnostic tool. Some interesting values for Young's modulus are given in Table 7.1, where low values are derived from compliant materials, and high values from resilient material.

Cartilage values in Table 7.1 are for articular cartilage, however, reported values depend very much on biological sample preparation and measurement technique [39]. Our rationale for atomic force microscopy was that individual nanostraws would have characteristic Young's modulus of elasticity, and that these may be reduced in samples from patients with chest wall deformities due to abnormalities in the assembly of nanostraws. Costal cartilage from patients with chest wall deformities have often been described as weak, particularly in those who do not do well in surgery.

Atomic Force Microscopy

The atomic force microscope (AFM) is a very high resolution scanning probe microscope that has found applications from the biological to the material sciences and has several advantages over transmission and scanning electron microscopy, including the absence of electron-induced specimen damage, ambient operation, preservation of biological morphology, and the ability to be utilized on live or fixed tissues. Analysis of biological samples frequently necessitates their fixation and protein cross-linking by chemical fixation, although fixation itself can cause tissue distortion. The AFM probes the surface topography of a sample to a very high resolution irrespective of whether the tissue is live or fixed. Probing of live tissues opens the possibility of investigating biomechanical measurements, for example, Young's modulus of elasticity [40–42]. Because AFM probing can be undertaken when the sample is submerged, it is possible to maintain live samples under physiological conditions.

It was proposed that the straw-like structures observed in costal cartilage act as a means of nutrient and gas transport; additionally they provide biomechanical support [6]. This is analogous to the pressure induced fluid flow in the canaliculi-lacunae network described in bone [43]. Here stress induced microcirculation in canaliculi of approximately 200 μm in diameter was investigated to show that flow can nourish 4–5 layers of concentric osteocytes and also suggest that stress induced flow may be important in bone remodeling where lack of flow may have pathological consequences, e.g., osteoporosis. Further characterization of collagen nanostraws is warranted if such a model is to be applied to fluid flow in costal cartilage.

In order to further characterize collagen nanostraws, brief homogenization and enzymatic digestion of cartilage with trypsin and hyaluronidase was used to isolate individual samples [44]. Individual nanostraws were examined for D-zone spacing and Young's modulus of elasticity. D-Zone bands are characteristic of collagen fibers, reflect the underlying regular arrangement of fibrils, and are estimated to be approximately 67 nm in hydrated and 64 nm in dehydrated samples [45]. The D-Zone patterns were measured from an SEM image compared to a digested and homogenized AFM image in air (Figs. 7.7a, b, respectively) and found mean D-Zone values of 63 nm and 65 nm from 10 zones each. These results are consistent with shorter D-Zones in dehydrated collagen forms suggesting that the underlying arrangement of fibers in costal cartilage derived from a patient with pectus carinatum is comparable to normal values under these conditions.

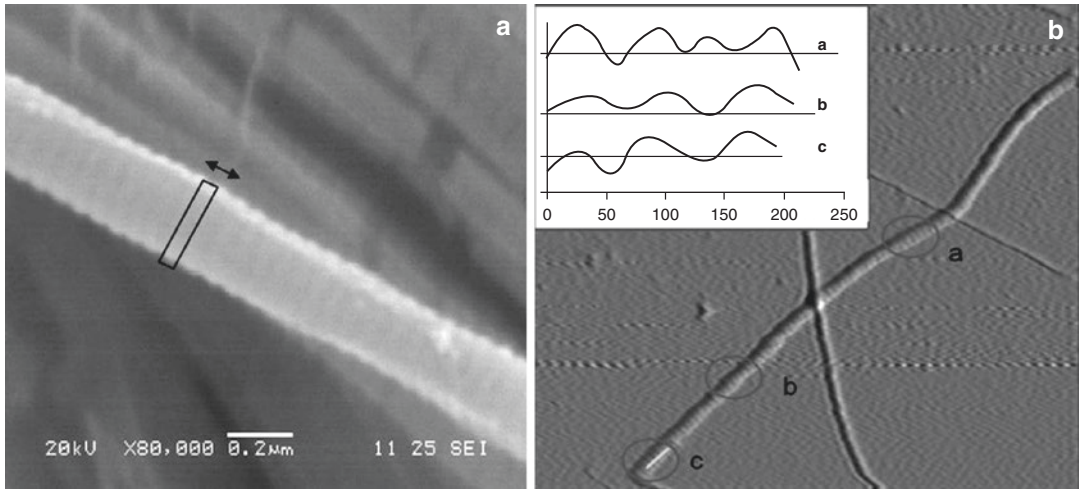

Fig. 7.7 SEM (**a**) and AFM (**b**) of characteristic D-zones of collagen fibers. A single D-zone is blocked and *arrowed* in **a**. The *insert* in **b** shows variation in D-zones over a single fiber [44]

To determine Young's modulus of elasticity, individual isolated nanostraws were attached onto poly-L-lysine cover glass. Force measurements were performed using frequency modulation force spectroscopy [46], and the resulting force data was modeled using the Derjagin, Muller, Toropov (DMT) model [47–49] via an in-house data analysis program written in MATLAB® (version 2009, Mathworks). Figure 7.8 shows typical force measurement on a nanostraw for digested, homogenized and fixed specimen in air. Utilizing the DMT model the modulus of elasticity from six separate measurements is found to be 2.06 ± 0.35 GPa.

Collagen nanostraws are structures significantly larger than individual collagen fibers and may be cross-linked by many structural proteins. Force measurements published in the literature, conducted for dehydrated collagen fibrils obtained from the common sea cucumber and analyzed at ambient conditions, resulted in values ranging between 1 and 11.5 GPa [50–52]. These values are high compared to reported hydrated, unfixed samples, where values of 2–5 MPa are reported [53]. These values strongly depend upon ionic concentration, hydrogen bonding, and hydration forces, all of which can influence interactions of tropocollagen molecules and, therefore, the elastic modulus. High values observed in Fig. 7.8 from costal cartilage (23 MPa) are derived from fixed samples, and due to cross-linking of proteins, create a more ridged structure and thus a higher Young's modulus.

Overall, these results show the unusual tubular network of costal cartilage that is hypothesized to act as a means of fluid and gas transport. To study nano-fluidic transport, such structures necessitate the accurate measurements of their dimensions. Interestingly, previous reports suggest that in rabbit tibia these structures correspond to the known biomechanical properties of the tissue, and would act as a dampening system during compression by resisting lateral fluid flow in the tissue and directing it against the compressive force [54]. Our study demonstrates that the protocols adopted for these measurements have significant influence on size measurement. Clearly, costal cartilage has large fiber dimensions with complex structures that are formed through finely tuned fibrillogenesis that ultimately reflect the biology of this understudied tissue type. The complex inheritance of chest wall deformities suggests that these processes are under the control of many genes.

Analysis of Candidate Genes

Aggrecan is an integral part of cartilage and mutations in the *ACAN* gene are associated with skeletal dysplasias [55, 56]. Patients with pectus excavatum commonly exhibit scoliosis, and *ACAN*

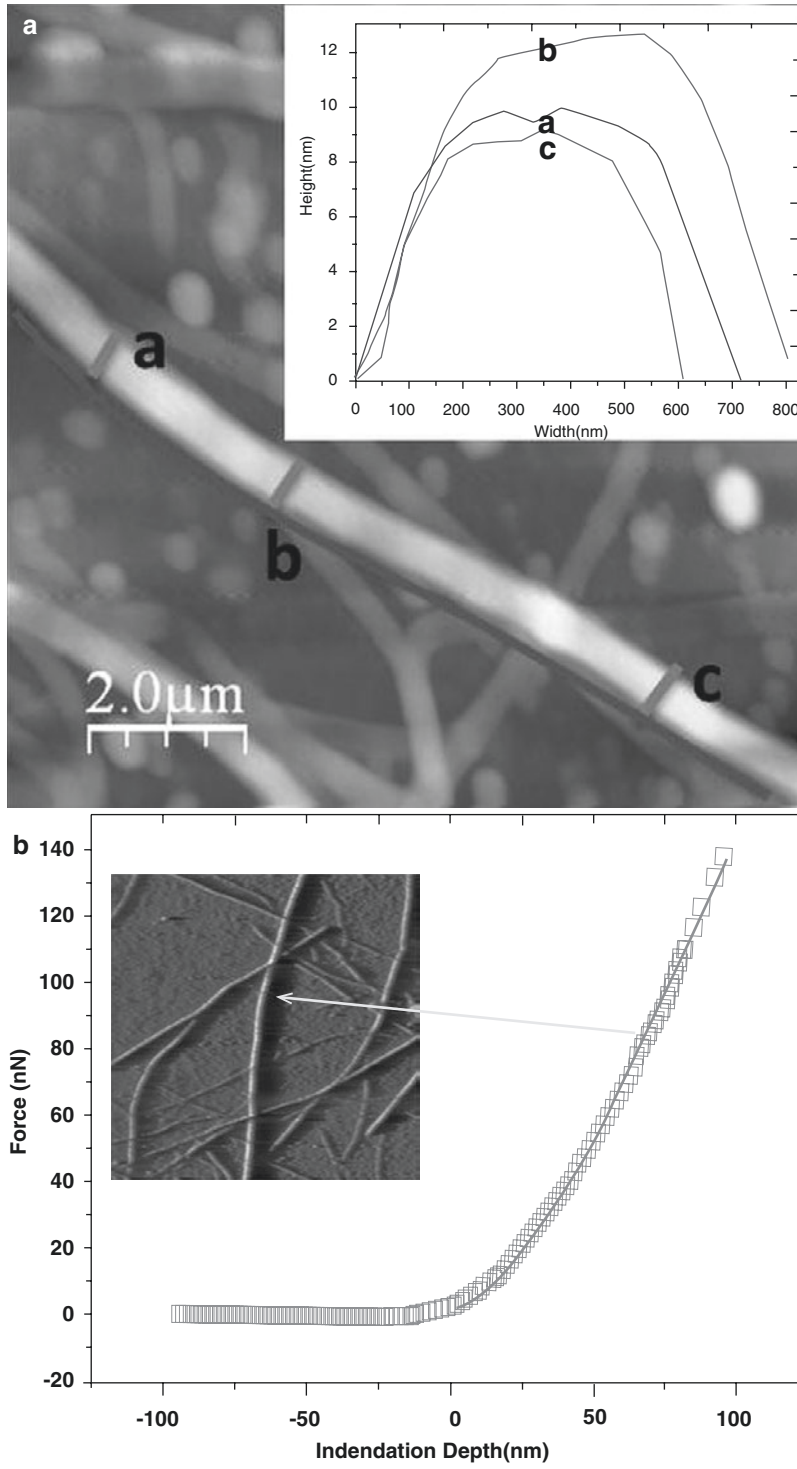

Fig. 7.8 (**a**) Representative topography of costal cartilage digested and homogenized. Contrast covers height variation of 390 nm. *Insets* in the figures show height distribution of nanostraws at various locations. (**b**) Force versus indentation depth data on a nanostraw for homogenized digested fixed sample in PBS buffer. Analysis based on the DMT model gives the modulus of elasticity of $E = 23 \pm 3$ MPa in PBS buffer. Experimental data is shown by symbols, while the curve-fit of data to the DMT model is shown by *solid lines* [44]

has been investigated as a candidate gene in familial idiopathic scoliosis [57, 58]. The CS1 domain of the *ACAN* gene exhibits length polymorphisms due to a variable number of tandem repeats (VNTR), 19 amino acids in length. Each repeat acts as an attachment site for chondroitin sulphate [59]. The number of *ACAN* VNTRs determines the number of GAG side-chains. The presence on aggrecan of a large number of highly charged chondroitin sulphate chains generates an osmotic swelling pressure and is important in maintaining structural integrity of the tissue. Smaller repeat sequences may result in mechanical shearing and tearing [60], and are associated with rheumatoid arthritis and spinal disc degeneration [56, 61]. It was hypothesized that abnormalities of costal cartilage in patients with pectus excavatum may be due to variation in number of repeat sequences outside of the normal reported range of 26–28 [59, 62] that would result in a concomitant change in chondroitin sulphate anchorage sites and compromised structural characteristics.

For this investigations were performed on the size and frequency distribution of *ACAN* VNTRs in patients with pectus excavatum and correlated overall allele sizes (genotype) to Haller index (Fig. 7.9) [63]. This was achieved by isolating DNA from venous blood of patients or by isolation from chondrocytes derived from patient costal cartilage and amplifying VNTR regions by polymerase chain reaction (PCR) [59, 62].

Genotyping identified 15 alleles ranging from 19 to 34 repeats, with alleles 25–28 accounting for 94 % and 84.7 % respectively in patients and controls. Allele distribution differed between patient and control groups ($\chi^2 = 48.58$, $p < .009$) such that patients had 0.43 fold fewer 25 repeats ($\chi^2 = 7.41$, $p < .025$) and 1.5 fold more 27 repeat alleles ($\chi^2 = 145.32$, $p < .001$) compared to controls. Overall, however, we observe an allele frequency of 0.120, 0.866, and 0.014 in patients for <26, 26–28, and >28 alleles respectively, consistent with the normal observed range [59, 62]. There is no apparent bias of allele genotype with increased Haller index, therefore a specific combination of VNTRs does not predispose to increased severity in pectus excavatum.

Patients showed phenotypic variation, and subgroups were identified where a genetic component may be influential. Females (16 % of patients), showed a significant increase in severity compared to males ($t(250) = 2.36$, $p < .019$; Mean + SD: 5.7 + 2.1 vs. 4.8 + 2.2), and tended to have a decreased number of VNTRs, consistent with a hypothesis of reduced attachment sites for chondroitin sulphate and weakened cartilage.

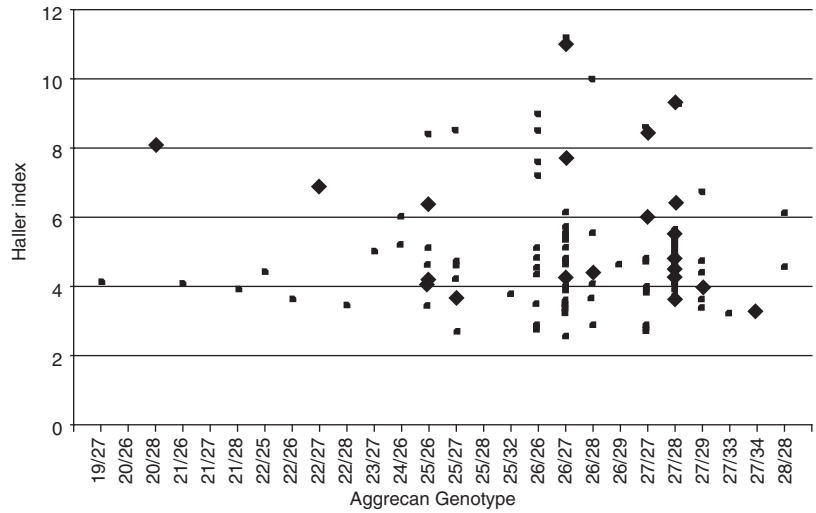

Fig. 7.9 Correlation of aggrecan genotype (VNTR repeats/allele) with Haller index in male (squares) and female (diamonds) patients [63]

Marfan phenotype patients (10.4% of patients), exhibited phenotypic findings consistent with Marfan appearance such as long limbs, arachnodactyly and high-arched palate without absolute diagnostic criteria for Marfan syndrome [64]. There was no apparent correlation in the number of VNTRs compared to the non-Marfan patients, suggesting that VNTRs do not have a differential role in this subgroup.

Repeat surgeries are the smallest subgroup (3.2% of patients) and showed no correlation between Haller index and surgical outcome, suggesting initial presentation is not an indicator of outcome.

Furthermore variation in a functional VNTR were investigated and identified, a useful marker for first-pass analysis. Investigation of SNPs will allow a more refined description in the inheritance of this, and other candidate genes, in the role of inherited chest wall deformities.

Analysis of Gene Expression in Costal Cartilage

Cartilage formation is a complex process with many interacting components. To determine gene expression in costal cartilage investigations were performed on twelve candidate genes based upon structural and functional importance in cartilage formation. Table 7.2 lists each gene and chromosomal location. Chest wall deformities show a sex bias, being more prevalent in males compared to females (4:1) [37, 38], suggesting that genes located on chromosome X may be of importance. Males have a single X chromosome (XY) and therefore defects in genes on this chromosome cannot be compensated for by genes at a second allele as in females (XX). Four genes were identified on chromosome X with relevance to cartilage formation (Table 7.2).

Aggrecan: A large aggregating proteoglycan that serves to anchor highly negatively charged keratin and chondroitin sulphate molecules ultimately responsible for generating osmotic pressure within cartilage.

Biglycan: *BGN*, a SLRP on the X-chromosome that encodes for the protein involved in assembly of collagen fibrils within the extracellular matrix of cartilage. It is closely related to decorin, possibly through gene duplication, and carries two glycosaminoglycan side chains. It strongly binds the growth factor TGF-β, controlling bioavailability.

Tissue inhibitor of metalloproteinase-1: *TIMP1* is located on the X chromosome and plays a role in the maintenance and turnover of the extracellular matrix within cartilage. It functions as an inhibitor of matrix metalloproteinases (MMPs), specifically MMP-8 and MMP-13, which are both collagenases.

Voltage-gated calcium channel-α1F: *CACNA1F* is a gene that encodes for a voltage-gated calcium channel and is found on the X chromosome. It functions to control the amount of

Table 7.2 Candidate genes investigated in this study [6]

Gene	Name	Chromosome location
ACTB	β-Actin	7p22
ACAN	Aggrecan	15q26.1
BGN	Biglycan	Xq28
CACNA1F	Voltage-gated calcium channel-α1F	Xp11.23
COL1A1	Collagen α-1 chain	17q21.33
COL2A1	Collagen type II α-1	12q13.11
DCN	Decorin	12q21.33
FBN1	Fibrillin 1	15q21.1
NYX	Nyctalopin	Xp11.4
SOX9	SRY (Sex determining region Y)-box 9	17q24.3
TGFβ1	Transforming Growth Factor-β1	19q13.2
TIMP1	Tissue inhibitor of metalloproteinase 1	Xp11.23

calcium that enters the cell upon membrane polarization and may be linked to bioelectric components of cartilage.

Nyctalopin: The *NYX* gene was investigated because of its location on the X chromosome and its function as a SLRP. It is associated more with eye function, and defects in the gene result in a number of eye related anomalies including night blindness

Collagen α-1 chain: *COL1A1* encodes for Type I α collagen fiber found in most connective tissues. Although not expressed highly in articular cartilage, it acts as a marker of cartilage differentiation.

Collagen type II α-1: *COL2A1* encodes for collagen Type II-α fibers found in cartilage where mutations in this gene have been associated with chondrodysplasias. It is highly expressed in articular cartilage and is essential for cartilage to resist compressive forces.

Decorin: *DCN* is a SLRP that plays a role in matrix assembly. It has an important role in binding collagen fibrils and strongly influences fiber size and shape. It has a single glycosaminoglycan side chain. It binds to COL1A1, COL2A1 and the growth factor TGF-β, controlling bioavailability.

Fibrillin 1: *FBN1* encodes a large matrix proteoglycan that serves as a structural component in force bearing microfibrils and binds to TGF-beta. Mutations in this gene are associated with Marfan syndrome where chest wall defects are common. One of these mutations creates an N-glycosylation site that disrupts multimeric assembly [28].

Sex determining region SRY box-9: *SOX9* is a homeobox class of DNA binding proteins. It is a potent activator of *COL2A1* and may also regulate the expression of other genes involved in cartilage formation by acting as a transcription factor for these genes.

Transforming Growth Factor-β1: *TGFβ1* is a multifunctional protein that controls proliferation and differentiation in many cell types. It regulates many other growth factors, and stimulates chondrocyte cell growth through the MAPK3 signaling pathway.

β-Actin: *ACTB*, was used throughout as a reference housekeeping gene and from which relative levels of gene expression were calculated.

For this, costal cartilages were immediately placed into a solution of RNAlater after surgery to preserve the integrity of expressed genes. RNA was extracted as described previously [6]. RNA was reverse transcribed to produce cDNA and amplified by RT-PCR on a BioRad CFX96 real time system (Fig. 7.10). Gene expression was measured by incorporation of SYBR green into amplified products. All primers were designed

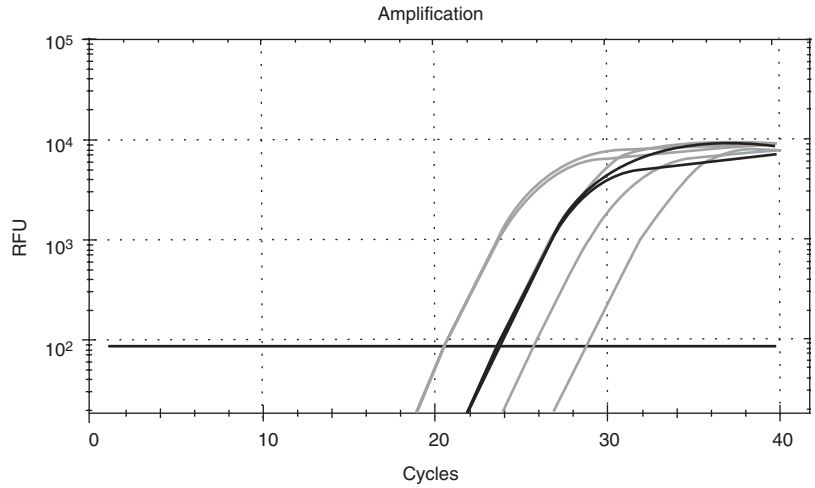

Fig. 7.10 Gene expression curves showing left to right, expression of *DCN*, *TIMP-1* (overlapping actin) *ACTB*, *BGN*, and *TGFB*. High expression is displayed as a curve farthest to the left and lower expression moving to the right

specifically for gene amplification (Qiagen CA, USA). Relative fold differences in gene expression were calculated as $2^{-(CtGOI-CtHKG)}$, where CtGOI is the Ct value of the gene of interest compared to the CtHKG, which is the Ct value for the house keeping gene [65].

Costal cartilage from individuals with chest wall deformities is described as abnormally grown and weak. Typically, surgical repair takes place during teenage years to early 20s. Phenotypically, there is considerable variation of the clinical condition of PC, reflecting the complex nature and inheritance observed in these families. Variation in gene expression between samples is, therefore, expected; however, it is unknown whether the expression of matrix genes will be affected by surgical procedures. We compared gene expression of 4 patients with pectus carinatum to an age-matched-control. *COL2A1*, *DCN*, *ACAN*, and *TIMP1* are all highly expressed compared to *ACTB*, however, when normalized to control (=100%) significant reductions in expression are observed with sample variation (Table 7.3).

Compared to control, PC1 showed significant reduction in expression of *DCN* ($p<0.001$) and *TIMP1* ($p<0.001$). PC3 showed significantly lower expression of *COL2A1* ($p<0.001$) and like PC4, both showed decreased expression of *ACAN* ($p<0.03$ and $p<0.024$, respectively). PC4 also showed significantly higher expression of *TIMP1* ($p<0.001$) and decreased expression of *BGN* ($p<0.04$). PC2 showed significant reduction in expression of *COL2A1* ($p<0.01$), *DCN* ($p<0.0002$), *TIMP1* ($p<0.001$), *BGN* ($p<0.03$) and *FBN1* ($p<0.01$). This sample, like all PC samples, was immediately processed from the operating room, although results suggest possible degradation of this sample.

Table 7.3 Percent fold difference in gene expression of four patients with pectus carinatum compared to β-actin and normalized to an age-matched control

	PC1	PC2	PC3	PC4
COL2A1	100	*63	*35	89
ACAN	122	93	*42	*205
DCN	*25	*25	92	117
TIMP1	*21	*29	107	*174
ACTB	100	100	100	100
BGN	43	*24	60	*22
COL1A1	125	100	92	100
FBN1	NA	*39	107	65
SOX9	91	61	46	191
TGF-β1	83	72	50	22

Significant differences in expression between control and patients are marked with * [6]

Many patients with chest wall deformities are considered Marfanoid-like [64] without fulfilling all criteria for diagnosis of Marfan syndrome, including mutations of the fibrillin-1 gene. The expression of this gene was not significantly different between control and patients, with the exception of PC2 ($p<0.01$). Expression of the X-linked genes *NYX* and *CACNA1F* was not detected in any samples. Overall, deregulation of *TIMP1* expression was evident in 3/4 PC samples, and expression of *DCN* was significantly lower in 2/4, suggestive of roles for fibrillogenesis and matrix turnover.

The differentiation status of cartilage can be equated to the ratio of *COL2A1*, present in differentiated cartilage, to *COL1A1*, present at higher levels in more undifferentiated cartilage. We compared ratios of gene expression from our samples to published data.

Ratios of the differentiation markers *COL2A1*:*ACAN* and *COL2A1*:*COL1A1* are low in PC patients and control (Table 7.4) compared to rabbit articular cartilage (1090 and 1790, respectively) but both are highly comparable to the nucleus pulposus region of lumbar discs (23 and 930 respectively), [66]. The ratios of *ACAN*:*COL1A1* fall between those reported for fully differentiated rat chondrosarcoma cells (78.4) and dedifferentiated chondrocytes cultured from costal cartilage (4.6) [67]. A high expression ratio of *COL2A1*:*COL1A1* (294.6) in human articular cartilage has been reported [65], but here results are referenced to *GAPDH* rather than *ACTB*. Overall, these results suggest costal cartilage is at an intermediate stage of differentiation and likely represents the different functional requirements of this tissue compared to articular cartilage. The differentiation similarities between lumbar discs and costal cartilage are of interest. The high incidence of scoliosis in patients with chest wall deformities indicates that defects of cartilage of a specific

Table 7.4 Gene expression ratios in costal cartilage from pectus carinatum and age-matched control

	COL2A1/ACAN	*COL2A1/COL1A1*	*ACAN/COL1A1*	*DCN/BGN*
PC1	35	878	25	8
PC2	29	701	24	13
PC3	36	427	12	19
PC4	19	990	53	69
Control	43	1117	26	13

differentiation status may be very important. Small differences exist however between patients and between patients and control (Table 7.4), suggesting that gene ratios measured here are not major contributors to chest wall abnormalities in these samples. Interestingly, *DCN* is expressed at high levels compared to *BGN*. As well as binding growth factors, both SLRPs have a role in fibrillogenesis and were hypothesized to play a role in the etiology of chest wall deformities. The high *DCN*/*BGN* ratio strongly suggests the importance of decorin expression in costal cartilage morphology. Decorin is present at high levels during tendon (fibrocartilage) development and persists until thick fibers are formed [17], thus parallels with costal cartilage (hyaline cartilage) are apparent.

Conclusions

Biological properties of human costal cartilage are a much understudied field. In this chapter preliminary data that investigates these properties were described. Sample characterization is of upmost importance and future studies should attempt to utilize samples from different but identified ribs, and the site of control samples should be verified for comparative purposes. Acquisitions of healthy age match controls are not easy because the age of patients tend to be teens to twenties. It has been suggested that rib abnormalities may be secondary to events of the thorax, with costal cartilage responding to micro-environmental factors, changing their biological characteristics as a result. The 'chicken and egg' paradox needs to be resolved, and identification of biological causes identified. This is particularly relevant to patients who do not do well in surgery, where a biological basis may underlie their prognosis and outcomes.

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Syndromes Associated with Pectus Deformities

8

Eva E. Amerstorfer and Amulya K. Saxena

Introduction

Pectus excavatum (**PE**) represents the most common deformity of congenital malformations of the chest wall. The prevalence of PE is about 1 in 300–1000 live births and affects predominantly males with a ratio of 4:1 and is evident at birth and progresses with growth [1]. Although its etiology has still not been identified yet, about 40 % of patients have relatives with chest wall deformities [2, 3]. A recent family analysis for PE on 34 families [4], it was postulated to have an autosomal dominant inheritance in 14, an X-chromosomal inheritance in 6 and an autosomal recessive inheritance in 4 for isolated PE. However, as few family members of this descriptive family study presented other connective tissue traits, an underlying connective tissue pathology cannot be ruled out.

PE is associated with structural abnormalities and disturbed synthesis of type II collagen in the costal cartilage, but also with abnormal ion distribution such as calcium, magnesium and zinc [3, 4]. Affecting the connective tissue, it may be associated with hereditary connective tissue disorders such as the classical connectivopathy "Marfan's syndrome".

Recently, in an investigation [3] on PE patients stated that 2/3rd of PE patients had further symptoms that are consistent with the MASS phenotype which is characterized by the combination of a mitral valve prolapse, non progressive aortic root enlargement, skeletal and skin alterations [5] which is as another tissue connectivopathy more frequent than the Marfan's, Ehler-Danlos or Poland's syndrome. The MASS phenotype is an autosomal dominant disorder that results from a mutation in the FBN1 gene that differs from Marfan's syndrome by a slight, generalized laxity of connective tissues and a non progressive aortic root enlargement without a risk to progress to aneurysm or predisposition to dissection [5–7]. Its criteria have been defined in the revised Ghent nosology [6] (Tables 8.1a and 8.1b). However, practically it is difficult to differentiate patients with Marfan-like features from MASS patients, wherefore it is important to follow these patients clinically and echocardiographically to identify an increased aortic risk and worsening of the mitral valve prolapse in individuals [3, 7].

Pectus carinatum (**PC**) describes the protrusion of the sternum and adjacent ribs due to overgrowth of the costal cartilages. It may be unilateral or bilateral, then causing a complete

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Table 8.1a Revised Ghent criteria for diagnosis of Marfan syndrome and related conditions

In the absence of family history:	**Marfan syndrome**
1. Aortic root diameter (Z ≥ 2) or aortic root dissection	
AND ectopia lentis	
2. Aortic root diameter (Z ≥ 2) or aortic root dissection	
AND FBN1 mutation	
3. Aortic root diameter (Z ≥ 2) or aortic root dissection	
AND systemic scores* (≥7points)	
4. Aortic root diameter (Z ≥ 2) or aortic root dissection	
AND ectopia lentis AND FBN1 mutation	
In the presence of family history:	
5. Ectopia lentis AND family history of Marfan syndrome	
6. Systemic scores* (≥7points) AND family history of Marfan syndrome	
7. Aortic root diameter (Z ≥ 2 and > 20 years old or Z ≥ 3 and < 20 years old) or aortic root dissection AND family history of Marfan syndrome	
Ectopia lentis with or without systemic score features AND with an FBN1 mutation not known with Aortic root diameter or aortic root dissection OR no FBN1 mutation	**Ectopia lentis syndrome**
Aortic root diameter (Z <2) AND systemic scores* (≥5 with at least 1 skeletal feature) without ectopia lentis	**MASS** (**myopia**, **mitral valve prolapse**, **borderline** (**Z < 2**) **aortic root dilatation**, **striae**, **skeletal findings phenotype**)
Mitral valve prolaps AND Aortic root diameter (Z <2) AND systemic scores* (<5) without ectopia lentis	**Mitral valve prolaps syndrome** (**MVPS**)

Compiled from information found in "The revised Ghent nosology for the Marfan syndrome." by Loeys et al. [6]

Table 8.1b Scoring of systemic features according to the revised Ghent nosology for diagnosis of Marfan syndrome and related conditions

*Scoring of systemic features	
Wrist AND thumb sign	3 (wrist OR thumb sign e 1)
Pectus carinatum	2 (PE or chest asymmetry =1)
Hindfoot deformity	2 (plain pes planus =1)
Pneumothorax	2
Dural ectasia	2
Protrusio acetabuli	2
Reduced upper segment/lower segment ration AND increased arm/height AND no severe scoliosis	1
Scoliosis or thoracolumbar kyphosis	1
Reduced elbow extension	1
Facial features (3/5) (dolichocephaly, enophthalmos, downslanting palpebral fissures, malar hypoplasia, retrognathia)	1
Skin striae	1
Myopia >3 diopters	1
Mitral valve prolapse (all types)	1
Total: (max. 20 points; score ≥7 indicates systemic involvement)	

Compiled from information found in "The revised Ghent nosology for the Marfan syndrome." by Loeys et al. [6]

elevation of the anterior chest wall. **PC** is less common then **PE** but represents the second most common pectus deformity accounting for about 5–20 % of anterior chest wall deformities [8]. It has a more pronounced male predominance compared to **PE** and occurs in most cases sporadic, although familial cases have been observed in about 1/3rd of PC patients [8]. Like **PE**, **PC** may also be a syndromic feature of an underlying genetic syndrome or connectivopathy, wherefore investigations for other organ or musculoskeletal malformations are recommended [9–11].

PC can be divided into 3 types: the classic keel chest, a lateral pectus carinatum and the "Pouter Pigeon chest", which represents a marked elevation of the second costal cartilages and of the sterno-manubrial joint. The classic keel chest is characterized by a protrusion of the caudal third of the elongated sternum with usually maximum prominence at the sterno-xiphoidal junction also described by "pyramidal chest" [8]. **PC** may also often cause significant physiological and body image issues to the adolescent or adult patients, especially when a pointed pyramidal chest is accentuated..

Cleft sternum describes a rare chest deformity due to a failure of ventral cell migration or fusion of the sternal bands which takes place between the 6th–9th week of gestation [12]. Its clinical manifestation ranges from an indentation in the manubrium to a complete non-fused cleft sternum. An association with a supraumbilical raphe, presenting as a postoperative scar or keloid, is frequently observed. Furthermore, it may be associated with cavernous hemangiomata [13]. A non-syndromic cleft sternum is mostly sporadic with a low recurrence risk, although familial presentation has been reported [14]. As a syndromic feature it may occur as a cleft of the inferior sternum in the Cantrell pentalogy which is characterized by a midline supraumbilical abdominal wall defects such as an omphalocele, rectus muscle diastasis or absent umbilicus, a deficient diaphragmatic pericardium and anterior diaphragm and cardiac malformations involving atrial or ventricular septum defect, double outlet right ventricle, truncus arteriosus, ectopic heart or pulmonary venous drainage anomaly [15]. The Cantrell pentalogy is generally a sporadic disease, although mutations of the gene on Xq25-26.1 with X-linked inheritance have been assumed [13]. A cleft sternum may also be a syndromic feature of the uncommon PHACES (**P**osterior fossa abnormalities and other structural brain abnormalities, **H**emangioma(s) of the cervical facial region, **A**rterial cerebrovascular anomalies, **C**ardiac defects, aortic coarctation and other aortic abnormalities, **E**ye anomalies, and **S**ternal defects and/or Supraumbilical raphe) syndrome, which describes a neurocutaneous association characterized by ventral developmental defects and haemangiomas with a female predominance and sporadic incidence [16].

The most frequent syndromes associated with PE or PC represents the Noonan and Marfan syndrome. They and other syndromes or chromosomal aberrations which may display pectus deformities as one of their syndromic features are listed in Tables 8.2a and 8.2b and the most frequent Noonan and Marfan syndromes as some extraordinary ones are described in brief below. In this chapter the concept of using figures has been employed for schematic representation of the syndromes in which colour based presentation of the organ systems was done to enable a better overview of the pathologies associated with individual syndromes.

Noonan Syndrome

The Noonan syndrome describes a common autosomal dominant inherited dysmorphology which is characterized by congenital heart defects (such as cardiomyopathy, atrial septal defect and pulmonary valve stenosis), a broad or webbed neck, a pectus deformity with cranial **PC** and caudal **PE**, short stature later in life with normal measurements at birth, facial dysmorphism involving ptosis, hypertelorism, low-set and posteriorly rotated ears, cryptorchidism, coagulation defects and variable mental development [17] (Fig. 8.1). In 1963 Noonan and Ehmke [18] first described this syndrome. It is caused by a mutation in various genes involved in the RasMAPK (*RAt* Sarcoma viral oncogene homolog – mitogen

Table 8.2a Proven or suspected monogenic syndromes and chromosomal aberrations with PE and/or PC listed in POSSUM (Pictures Of Standard Syndromes and Undiagnosed Malformations, version released 2013) and/or WBDD (Winter-Baraitser Dysmorphology Database, version 1.0.14)

Syndromes	Inheritance	Gene location	Gene
Aarskog syndrome – Facio-digito-genital dysplasia		**Xp11.21**	FGD1
Aarskog-like syndrome (Xu 2010) without the characteristic hyperextension of the proximal interphalangeal joints of Aarskog syndrome	AD/AR?		
Acrocapitofemoral dysplasia	AR	**2q33-q35**	IHH?
Acro-cephalo-polysyndactyly, type 4 – Goodman syndrome	AR		
Acro-cranio-facial dystostosis, Kaplan type	AR		
Acrofacial dysostosis, Catania type	AD/X-linked		
Acropectoral syndrome	AD	**7q36**	
Acro-pectoro-vertebral dysplasia – severely malformed hands and feet	AD	**2q36**	
Acrorenal syndromes – renal abnormalities, missing fingers or other limb reduction defects	AD/AR		
Adducted thumb syndrome – Craniosynostosis, arthrogryposis, cleft palate	AR		
Al-Awadi-Raas-Rothschild syndrome	AR	**3p25.1**	WNT7A
Al-Gazali (1996) – webbed neck-facial dysmorphism-congenital heart disease	AR		
Allan-Herndon-Dudley syndrome	X-linked	**Xq21**	MTC8
Alpha-mannosidosis	AR	**19p13.2**	MAN2B1
Anterolateral bowing of tibia, contralateral polydactyly of great toe			
Antley-Bixler syndrome	AR	**10q26, 7q11.2**	FGFR2
Antley-Bixler syndrome, genital anomalies, disordered steroidogenesis	AR	**7q11.2**	POR
Apert syndrome – Acro-cephalo-syndactyly type 1	AD	**10q26**	FGFR1 FGFR2
Aplasia pectoralis major, renal anomalies			
Arterial tortuosity syndrome	AR	**20q13.1**	GLUT10
Arthropryposis, X-linked tapes 1–3	X-linked	**Xp11.3-q11.2**	
Aspartylglucosaminuria	AR	**4q32-q33**	AGA
Autosomal dominant mental retardation, craniofacial and genital anomalies	AD	**11q13.1-q13.2**	PACS1
Bannayan-Riley-Ruvalcaba syndrome	AD	**10q22-23**	PTEN
Baraitser-Patton syndrome – Noonan-like with slow hair growth			
Baratela-Scott syndrome – Abnormal facies, skeletal dysplasia, dislocated patella, developmental delay	X-linked?		
Barth syndrome – Cardioskeletal mitochondrial myopathy, neutropenia	X-linked	**Xq28**	TAZ1 G4.5
Basal cell naevus syndrome – Gorlin-Goltz syndrome	AD	**9q22.3, 1p32**	PTCH1 PTCH2
Beals contractural arachnodactyly	AD	**5q23-5q31**	FBN2
Becker naevus syndrome			
Blepharophimosis, deafness, mental retardation, short stature, camptodactyly			
Blepharophimosis, radioulnar synostosis			

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Bowed tibiae, radial ray defects, osteopenia	AR		
Brachydactyly, syndactyly, short stature, microcephaly, mental retardation			
Brachycephaly-deafness-cataracts-mental retardation			
Braddock-Carey syndrome – Thrombocytopenia, Robin sequence, agenesis of corpus callosum			
Branchio-oto-renal syndrome	AD	**8q13.3**; **19q13.3**	EYA1 SIX5
Branchio-skeleto-genital syndrome – Elsahy-Waters syndrome	AR		
Campomelic dysplasia/dwarfism	AD	**17q24.3-q25.1**	SOX9
Camptodactyly, tall stature, hearing loss	AD	4p16.3	FGFR3
Camptodactyly-type Guadalajara	AR		
Carbohydrate-deficient glycoproteins syndrome IIA	ÂR	**14q21**	CDGS2
Cardiofaciocutaneous syndrome	AD	**12p12.1** **7q34**	KRAS BRAF MEK1 MEK2
Cardio-skeletal syndrome, Boehme-Shotar type	AD		
Cartilage-hair hypoplasia – Metaphyseal chondrodysplasia, McKusick type	AR	**9p13**	RMRP
Cataract, hypertrichosis, mental retardation syndrome	AR		
Catel-Manzke – cleft palate, accessory metacarpal			
Cerebro-costo-mandibular syndrome	AD/AR		COG1
Charcot-Marie-Tooth disease with other defects: hereditary motor or sensory neuropathy has been reported with dysmorphism (Ruiz 1987) and with neurological defects		**1p36-p35** **8p21** **17p11.2** **1q22** **3q13-q22** **5q23-q33** **8q13-21.1** **8q24** **11q23** **19q13** **Xq13.1**	
Choanal atresia, lymphoedema	AR	**1q32**	PTPN14
Chondrodyslasia, Megarbane-Dagher-Melki type	AR		
Chondrodysplasia, joint dislocations, Megarbane-Ghanem type	AR		
Coffin-Lowry syndrome – moderate to severe mental retardation, characteristic facial features, short stature, pectus deformity, mitral valve dysfunction, and tapering fingers in males.	X-linked	**Xp22**	RSK2
Congenital disorder of glycosylation, type IIa (Alazami 2012, Alkuraya 2010)	AR	**14q21**	MGAT2
Congenital hypothyroidism, cerebellar hypoplasia, mental retardation			
Connective tissue dysplasia, Spellacy type	AR		
Cranioectodermal dysplasia – Levin syndrome: short limb dwarfism, narrow chest, dolicocephaly, sparse thin hair, distinctive face, dental abnormalities and brachydactyly	AR	**3q21** **14q24.3** **2p24.1**	WDR10 WDR35

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Craniosynostosis, Passos-Bueno type	AD		
Cryptorchidism, arachnodactyly, mental retardation, other defects Benthem-Driessen-Hanveld syndrome			
Cutis laxa, developmental delay, epiphyseal dysplasia, deafness (Armstrong 2003)			
Dandy-Walker syndrome – Cutis laxa, immunodeficiency: mental retardation, macrocephaly, coarse facies with hypertelorism, down-slanting palpebral fissures, long philtrum and large low set ears; sparse scalp and body hair, thin dystrophic nails, pectus excavatum and marked lumbar lordosis			
D-bifunctional protein deficiency – Pseudo-Zellweger syndrome	AR	**5q23.1**	HSD17B4
Desbuquois syndrome	AR	**17q25.3**	CANT1 DTDST
Distal arthrogryposis, mental retardation, Chitayat-Hall type	AR		
Distal arthrogryposis, Rivera type	AR?		
Dominant brachyolmia Brachyolmia, Maroteaux type	AR	**12q24.1-12q24.2**	TRPV4
Dundar (2001) – acropectoral syndrome	AD	**7q36**	
Dursun syndrome – Familial pulmonary arterial hypertension, leukopenia, ASD	AR	**17q21**	G6PC3
Dyggve-Melchior-Clausen syndrome, Smith-McCort syndrome	AR	**18q12** **18q21.1**	DYM RAB33B
Dysequilibrium syndrome, Cerebellar hypoplasia, VLDLR-associated CAMRQ1 (Cerebellar ataxia, mental retardation, with/without quadrupedal locomotion-1)	AR	**9q24**	VLDLR
Dysosteosclerosis	AR		
Dysspondyloenchondromatosis: Short stature from birth, flat midface, short and broad thorax, unequal limb length, multiple enchondromata of flat and tubular bones with vertebral segmentation defects resulting in severe kyphoscoliosis.	AD?		
Edwards-Gale syndrome – Camptobrachydactyly	AD		
Ehlers-Danlos syndrome, Friedman type	AD		
Ehlers-Danlos syndrome, musculocontractural type – Adducted thumb-clubfoot syndrome	AR	**15q14**	CHST14
Facio-auriculo-thoracic syndrome	AR		
Facio-thoracic-genital syndrome	AR		
Faciothoracoskeletal syndrome, Gul type	AR		
Femoral epiphyseal dysplasia, myopia, deafness	AR		
Fetal alcohol syndrome		**12q24.2**	
Fibrochondrogenesis 1: Short-limbed skeletal dysplasia with characteristic bone histology, flat face, small nose with anteverted nares, long philtrum, small mouth, cleft palate, narrow thorax.	AR	**1p21**	COL11A1 COL11A2
Fibrodysplasia ossificans progressiva, Myositis ossificans progressiva – Munchmeyer syndrome	AD	**2q23-q24**	ACVR1 NOG

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Fitzsimmons-Guilbert syndrome: **Spastic paraplegia**, **brachydactyly**, **cone shaped epiphyses**			
Floating –Harbor syndrome – Pelletier-Leisti syndrome	AD	**16p11.2**	SRCAP
Fronto-metaphyseal dysplasia: **Twisted ribs**, **increased interpedicular distance of lumbar spine**, **flared ilia**, **mildly flared and bowed long bones**, **arachnodactyly**, **undermodelled short tubular bones**, **advanced ossification of femoral and tibial epiphyses**.	X-linked	**Xq28**	FLNA
Giuffre-Tsukahara syndrome: **Radio-ulnar synostosis**, **microcephaly**, **short stature**, **mental retardation**	AD/AR	**Xp22.13-q21.33**	
Gurrieri syndrome	AR		
Hirschsprung disease – microcephaly-mental retardation	AD	**2q22**	SIP1
Hirsutism, **hypoplastic nails**, **developmental delay**	AR		
Holt-Oram syndrome	AD	**12q21-24** **14q23-24**?	TBX5
Homocystinuria	AR	**21q22.3**	CBS
Humero-spinal sysostosis, **congenital heart disease**	AR	**10q22.1**	CHST3
Hurler syndrome	AR	**4p16.3**	IDUA
Hypertelorism, **distinctive facies**, **brachydactyly**, **genital anomalies**, **mental retardation**	AR/X-linked?		
Hypodontia, **choroids calcification**, **unusual facies**, **Pallotta-Fusilli type**	AD		
Hypomelanosis of Ito	X-linked?		
Hypospadias, **hypertelorism**, **lid colomba**, **deafness**	AR		
Ichthyosis-cheek-eyebrow-syndrome	AD		
Idaho 2 syndrome, **Craniosynostosis**			
Isolated thoracic dysostosis (**Rabushka 1973**)	AD		
Joubert syndrome, **Partial or complete agenesis of the cerebellar vermis**, **oculomotor anomalies with jerky eye movements**, **hypotonia**, **ataxia and impaired equilibrium**, **severe mental retardation**, **and episodic tachypnoea/apnoea in neonatal period**, **variable phenotype has included hemifacial spasm**, **retinal dystrophy**, **chorioretinal colobomata**, **post-axial polydactyly**, **and tongue tumours**.	AR	**9q34.3** **7q32** **6q23** **2q13** **3q11.2** **1q12.2** **4p15.3** **2q33** **10q24.1** **Xp22.3-p22.2** **5p13**, **2q24** **16q12** **16q23**	INPP5E AHI1 NPHP1 CEP290 ARL13B MKS3 RPGRIP1L TTC21B CC2D2A OFD1 KIF7 TCTN1 TMEM237 CEP41 TMEM138 C5ORF42 TCTN3 ZNF423 TMEM231
Juvenile hyaline fibromatosis	AR	**4q21**	CMG2
Juvenile idiopathic osteoporosis	AR		
King-Denborough – dysmorphic features, **myopathy**, **malignant hyperthermia**	AD	**19q13.1**	RYR1

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Khaldi, F., et al. 'An autosomal recessive disorder with retardation of growth, mental deficiency, ptosis, pectus excavatum and camptodactyly' (1988)	AD		
Kniest dysplasia, Metatropic dwarfism, type II	AD	**12q13.11-q13.2**	COL2A1
Kyphomelic dysplasia	AR		
Lack of subcutaneous tissue, arthritis, skeletal dysplasia			
Lateral meningocele syndrome – Osteosclerosis, meningoceles, "empty sella" – Lehman syndrome	AD		
Lehman (1977) – osteosclerosis, abnormalities of nervous system/meninges	AD		
Lenz microphtalmia syndrome	X-linked	**Xq27-Xq28** **Xp21.2-p11.4**	BCOR
LEOPARD syndrome (multiple Lentigines, Electrocardiographic conduction abnormalities, Ocular hypertelorism, Pulmonary stenosis, Abnormalities of genitalia, Retardatio of growth, sensoneural Deafness)	AD	**6q** **12q24**	PTN11 RAF1
Lethal chondrodysplasia, Blomstrand type	AD	**3p22-p21.1**	PTHR1
Loeys-Dietz syndrome – aortic aneurysm, hypertelorism, arterial tortuosity, CP	AD	**9q22** **3p22** **15q22.3** **1q41**	TGFBR2 TGFBR1 SMAD3 TGFB2
Lowry-Wood syndrome - epiphyseal dysplasia, microcephaly, nystagmus	AR		
Lujan – Fryns syndrome	X-linked	**Xq25-q26**	MED12 UPF3
Mandibulofacial dysostosis, Toriello type	X-linked		
Marden-Walker syndrome, Blepharophimosis, arthrogryposis, myopathic facies, microcephaly, kyphoscoliosis, arachnodactyly, renal anomalies and mental retardation. CNS anomalies such as Dandy-Walker anomaly and hypoplastic corpus callosum reported.	AR		
Marfanoid hypermobility syndrome		**Xq22.3**	
Marfanoid mental retardation syndrome, Fragoso-Cantu type	AR		
Marfan syndrome	AD	**15q21**	FBN1
Maroteaux-Lamy syndrome – Mucopolysaccharidosis 6	AR	**5q11-q13**	ARSB
MASS phenotype	AD		
Megarbane, et al. 'Spondyloepimetaphyseal dysplasia of Maroteaux (pseudo-Morquio type II syndrome): report of a new patient and review of the literature' (2004)			
Mental retardation, hypotonia, skin hyperpigmentation			
Mental retardation, microcephaly, Tolmie type			
Mental retardation, polydactyly, uncombable hair – Kozlowski-Krajewska type	AR/X-linked?		
Mental retardation, short stature, DeLozier-Blanchet type			
Mesomelia-synostosis syndrome	AD	**8q13**	
Mucopolysaccharidosis type VII	AR	**7q21.11**	MPS7

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Metaphyseal chondrodysplasia, tracheomalacia, Kaitila type	AR		
Metaphyseal dysplasia, cataract, mental retardation			
Metaphyseal acroscyphodysplasia: Mental retardation, short stature., wedge-shaped epiphyses of knees Bellini disease	AR		
Metatropic dysplasia	AD/AR		
Microcephalic primordial dwarfism, type 3	AR		
Midface asymmetry, brachydactyly			
Morquio syndrome – mucoploysaccharidosis 4: coarse facies, fine corneal opacities, enamel hypoplasia, mandibular prognathism, short neck, broad chest, pectus carinatum and aortic incompetence	AR	**16q24.3**	GALNS GLB1
Mowat-Wilson syndrome: Microcephaly, mental retardation., Hirschsprung's disease, dysmorphic facies		**2q22**	ZEB2
Mucolipidosis 1	AR	**6p21.3,** **10p14-q2**	
Mucopolysaccharidosis 7 – Sly syndrome	AR	**7q21.11**	GUSB
Mucosal neuroma syndrome	AD	**10q11.2**	RET
Multiple anomalies, mental retardation, athetoid cerebral palsy			
Multiple congenital anomalies, Hirschsprung disease, Toriello type (1988)			
Multiple congenital anomalies, mental retardation, Buenos Aires type -Mutchinick syndrome	AR		
Multiple congenital anomalies, optic nerve colobomas, renal anomalies, arthrogryposis multiplex	AD		
Multiple epiphyseal dysplasia with Robin sequence			
Multiple epiphyseal dysplasia, macrocephaly, distinctive facies, Al Gazali-Bakalinova type	AR	**15q26**	
Multiple epiphyseal dysplasia-microcephaly-nystagmus	AR		
Multiple joint dislocations, cataracts	Mosaicism?		
Mutchinick syndrome			
Myotonia, mental retardation, spinal abnormalities – Richieri-Costa-Garcia-Da Silva syndrome	AR		
Nail-patella syndrome – Onycho-osteo dysplasia – HOOD syndrome – Osteo-onychodysplasia – Turner-Kieser syndrome – Fong disease – Osterreicher-Turner syndrome	AD	**9q34.1**	LMX1B
Neuraminidase deficiency with beta-galactosidase def. – Goldberg syndrome	AR	**20q13.12**	CTSA
Neurofibromatosis-Noonan syndrome	AD	**17q11.2**	NF1 PTPN11
Nevoid basal cell carcinoma syndrome (Gorlin)	AD	**9q22-q31**	PTCH1
Nevo syndrome	AR	**1p36.3-p36.2**	PLOD
Noonan syndrome	AD	**12q24.1** **12p12** **2p22** **3p25**	PTPN11 KRAS SOS1 RAF1

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Noonan-like/multiple giant cell lesion syndrome	AD	**12q24.1**	PTPN11
Noonan-like syndrome, loose anagen hair		**10q25**	SHOC2
Occipital horn syndrome	X-linked	**Xq13**	MNK
Ocular colobomas, cleft palate, genital, skeletal, and craniofacial abnormalities, mental retardation MCA/MR, Khalifa type	AR/X-linked?		
Opitz-G syndrome	X-linked	**Xp22**	MID1
Opsismodysplasia	AR		
Orofaciodigital syndrome, type 3	AR		
Osteogenesis imperfecta type I, III, IV	AD, AR	**17q22, 7q22.1**	COL1A1 COL1A2
Osteoporosis, macrocephaly, mental retardation, blindness, joint laxity	AR		
Osteoporosis-pseudoglioma syndrome	AR	**11q12-13**	LRP5
Oto-facio-osseous-gonadal syndrome	AR		
Paes-Alves syndrome	AR		
Parana hard skin syndrome	AR		
Parastremmatic dysplasia	AD	**12q24.1**	TRPV4
Partial familial lipoatrophy, Dunnigan type	AD	**1q21-q23**	FPLD3 FPLD3 PLIN1
Patterson syndrome – Pseudoleprechaunism			
Pectus excavatum, macrocephaly, short stature, dysplastic nails Zori-Stalker-Williams syndrome	AD		
Platyspondylic chondrodysplasia, San Diego type	AR	**4p16.3**	FGFR3
Polyosteolysis with hyperostosis syndrome	AR		
Postaxial acrofacial dysostosis syndrome – Miller syndrome	AR	**16q22.2**	DHODH
Premature ageing, hypogonadism with Marfan syndrome			
Progressive macrocephaly and hamartoma, Halal type	AD?		
Pseudoachondroplasia	AD	**19p13.1**	COMP
Pseudodiastrophic dysplasia	AR		
Pseudomarfanism – Dinno-Shearer-Weisskopf syndrome	AD		
Ptosis, pectus excavatum, camptodactyly, mental retardation, short stature – Khaldi-Gharbi syndrome	AR		
Rigid spine syndrome, Severe classic multiminicore myopathy		**1p35-1p35**	SEPN1
Renal cysts, ear anomalies, abnormal hair, dysmorphic facies			
Robinow syndrome, autosomal recessive form – Covesdem syndrome	AR	**9q22**	ROR2
Robin sequence, pectus excavatum, rib and scapular anomalies, Stalker-Zori type	AD		
Rubinstein-Taybi syndrome – Broad thumbs/great toes, characteristic facies, mental retardation	AD	**16p13.3, 22q13**	CREBBP EP300
Ruvalcaba syndrome	AD		

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Saldino-Mainzer syndrome, Skeletal, retinal, cerebellar defects, nephropathy – Conorenal syndrome	AR	**16p13.3**	IFT140
Saul-Wilson syndrome			
SCARF syndrome	X-linked		
Schilbach-Rott syndrome, Ocular hypotelorism, blepharophimosis, submucous cleft palate, hypospadias	AD		
Schiel-Stengel-Rutkowski syndrome			
Schwartz-Jampel syndrome – Myopathy, dwarfism, chondrodystrophy, ocular, facies Osteochondromuscular dystrophy, Chondrodystrophic myotonia	AR	**1p34-p36.1**	HSPG2
Scurvey – congenital malformations in man related to Vitamin C deficiency, rare before age 2 years: irritability, bleeding disorder, the sternum appears depressed, with elevated rib margins ("scurvy rosary").		**8p21.1**	GULO
Seckel syndrome – primordial dwarfism, proportionate short stature, severe microcephaly, mental retardation, and a typical 'bird-head' facial appearance	AR	**3q22.1-q24** **18p11.31-q11.2** **14q23** **15q21**	SCKL1 SCKL2 SCKL3 SCKL5 SCKL6 SCKL7 PCNT2
Secundum ASD, various cardiac and noncardiac defects MCA, Megarbane-Stephan type	AD		
Sedaghatian lethal metaphyseal chondrodysplasia	AR		
Shprintzen-Goldberg – arachnodactyly, craniosynostosis, hernia		**3p22** **15q21**	TGFBR2 FBN1
Simpson-Golabi-Behmel syndrome	X-linked	**Xq26** **Xp22**	GPC3 GPC4
Skeletal dysplasia, mental retardation – Katsantoni-Cote type			
Somlo (1993) – marfanoid syndrome with polycystic renal disease	AD	**16p13**	
Sotos syndrome 2		**19p13.3**	NFIX
Spondyloepimetaphyseal dysplasia, joint laxity	AR		
Spondyloepimetaphyseal dysplasia, Leonard-Hughes type			
Spondyloepimetaphyseal dysplasia, matrilin 3 type	AR	**2p25-24**	MATN3
Spondyloepiphyseal dysplasia-brachydactyly and distinctive speech Fantasy Island syndrome	AD/X-linked?		
Spondyloepiphyseal dysplasia tarda, Leroy-Spranger type	AR		
Spondyloepiphyseal dysplasia, Sensenbrenner type			
Spondylometaepiphyseal dysplasia, Brazilian type	AD		COL2A1
Spondylometaepiphyseal dysplasia, Kozlowski type	AD	**12q24.1**	TRPV4
Spondylometaepiphyseal dysplasia, short limb-hand type	AR	**1q12-qter**	DDR2
Spondylometaepiphyseal dysplasia, Strudwick type	AD	**12q13.11-q13.2**	COL2A1

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Spondyloperipheral dysplasia	AD	**12 q13**	COL2A1
Spondylothoracic dysplasia – Jarcho-Levin syndrome	AR	**19q13.1-q13.3** **15q26.1**, **17p13.1**	DLL3 MESP2 HES7 LFNG
Spruijt, et al. 'Identification of a novel EYA1 mutation presenting in a newborn with laryngomalacia, glossoptosis, retrognathia, and pectus excavatum' (2006)			EYA1
Stickler syndrome – Hereditary progressive arthro-ophthalmopathy – Cervenka syndrome	AD/AR	**12q13.11-q13.2** **6q12-14** **6p21.3** **1p21** **1p33-p32.2**	COL2A1 COL9A1 COL11A2 COL11A1 COL9A2
Stratton-Parker syndrome, Growth hormone deficiency, Wormian bones, cardiac defect			
Tibial bowing, pectus excavatum	AD		
Thoraco-laryngo-pelvic dysplasia, Barnes syndrome	AD		
Three M syndrome	AR	**6p21.1**	CUL7
Toriello-Carey syndrome, Agenesis of corpus callosum, Robin sequence, facial anomalies	AR		
Torticollis, keloids, cryptorchism, renal dysplasia – Goeminne syndrome	X-linked	**Xq28**	
Tricho-rhino-phalangeal syndrome, type 1 and 3	AD	**8q24.12**	TRPS1
Unusual facies, pectus carinatum, joint laxity (Guizar-Vazquez)	AR		
Urethral obstruction sequence – Prune belly syndrome – Triad syndrome – Eagle-Barret syndrome			
Weissenbacher-Zweymuller syndrome	AR	**6q21.3**	COL11A2
Wiedemann syndrome, Microcephaly, micropenis, hand/foot anomalies			
Williams syndrome – Williams-Beuren syndrome – Elfin facies syndrome	AD	**7q11.23**	ELN RFC2 GTF2I GTF2IRD1 MAGI2 CYLN2 LIMkinase1
X-linked mental retardation, alpha-thalassaemia	X-linked	**Xq13**	ATRX
X-linked mental retardation, marfanoid habitus	X-linked	**Xq26.1**	ZDHHC9
X-linked mental retardation, agenesis of the corpus callosum, coloboma, micrognathia	X-linked	**Xq13.1**	IGBP1
X-linked mental retardation, short stature, muscle wasting, facial dysmorphism	X-linked	**inv(X)p21.1**; **q22.1**	NXF5
X-linked mental retardation, Snyder-Robinson type	X-linked	**Xq21**	
X-linked, mental retardation, spastic paraplegia, Claes-Fryns type	X-linked	**Xp11.22-p11.21**	JARID1C
Zellweger syndrome	AR	**7q11.23**	PEX1,2,3,5,6, 12,14,16,19

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Chromosome aberrations			
Diaphragmatic hernia, lung hypoplasia, microcephaly, coarse face with 8mostly) full lips, bulbous nasal tip, prominent forehead and deep-set eyes		**Del 1q41 ->42**	**Chromosome 1**
Pre- and postnatal growth retardation, cardiac defects, mental retardation, macrocephaly or micro-/brachycephaly, facial dysmorphisms, wrinkled skin, Pierre-Robin sequence, pulmonary hypoplasia and flexion contractures		**Dup 1q**	
Defective central nervous system, heart and lung, overlapping flexed fingers, facial dysmorphism		**Mosaic trisomy 1**	
Mental and postnatal growth retardation, prominent broad nasal bridge and bulbous nose, high-arched palate, micrognathia and anomalies of fingers/toes		**Del 2p**	**Chromosome 2**
Beaked nose with abnormal bridge, deep-set eyes, myopia, high narrow palate, low-set abnormal ears, scoliosis, pectus carinatum, long slender fingers with flexion contractures, spasticity and hypertonia (Wenger & McPherson 1997)		**Del 2p12-p11.2**	
Long face with high forehead, fine hair, telecanthus, down slanting palpebral fissures, large ears, broad and high nasal root with prominent tip, high palate, everted lower lip, arachnodactyly, pectus excavatum, kyphoscoliosis, mitral and aortic valva anomalies (Chabchoub 2008)		**Microdeletion 2p15**	
Developmental delay, microcephaly, corpus callosum defects, cardiac anomalies, prominent forehead, low-set and malformed ears, tendency to recurrent severe infections		**Del 2q13->q21**	
Severe mental retardation and behavioural problems, facial asymmetry, scaphocephaly, receding forehead, thick coarse hair, prominent supraorbital ridges, thick eyebrows with synophrys, proinent eyes, long nose, thin nasal bridge, anteverted nares, high narrow palate with bifid uvula, small teeth, low-set posteriorly rotated long ears, short neck, pectus excavatum, scoliosis, tapering fingers, sandal gap of the toes, hypoplastic metatarsals and macroorchidism (Prontera 2009)		**Del 2q31.22->q32.3**	
Moderate-severe mental retardation, short stature, rounded face with short nose and flattened nasal bridge, short metacarpals and metatarsals		**Del 2q37**	
Short stature, microcephaly, severe mental retardation, aortic stenosis, high prominent forehead, hypertelorism, depressed nasal bridge, long/narrow trunk and scoliosis, soft skin and hyperextensible fingers with arachnodactyly		**Dup2p**	
Growth and mental retardation, cardiac, renal and gastrointestinal anomalies, facial dysmorphism		**Dup2q**	
Severe mental retardation, congenital heart disease, kidney defects, placid personality, pre-/postnatal growth retardation, asymmetric skull and face, telecanthus, ptosis, micrognathia, low hair line, synophrys		**Del 3p25->pter**	**Chromosome 3**
Profound growth and mental retardation, cardiac abnormalities, annular pancreas, renal anomalies, facial dysmorphism		**Del 3p->p14**	
Hypertelorism, epicanthus, epicanthus inversus and ptosis, broad and high forehead, broad flattened nasal bridge, profound growth and mental retardation, frequent cardiac abnormalities, with annular pancreas and renal anomalies in some cases.		**Del 3p(p12p14)**	
Moderate mental retardation, growth delay, hypotonia, horseshoe kidney, hypospadias, facial dysmorphism		**Del 3q29**	
Variable clinical features; developmental delay, autistic features, skeletal abnormalities, dysmorphic facies (long face, long nose, high nasal bridge and broad nasal tip, short philtrum, large ears. Orofacial clefting, cardiac defects, ocular anomalies and genitourinary malformations		**Microdeletion 3q29**	
Cutis marmorata, frontal bossing, hirsutism and short neck		**Mosaic trismoy 4**	**Chromosome 4**

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Severe growth and mental retardation, hyperextensible joints, triangular face and multilobulated ear tags		**Del 5p15**	**Chromosome 5**
Bilateral epicanthal folds, low-set dysplastic ears, short nose with anteverted nostrils, conically shaped fingers, generalised increase of subcutaneous fat, multiple fine venous telangiectasia on back, mild pectus carinatum, and a general muscular hypotonia		**Del 5q21.1->q23.1**	
Prenatal lymphoedema, marked muscular hypotonia and delayed motor milestones, but speech development within normal limits, wide fontanelles, failure to thrive with postnatal short stature, and multiple minor anomalies including bell-shaped chest, minor congenital heart disease, and distinct facies including upslanting palpebral fissures, short nose and prominent cheeks (Rauch 2003, 2007)		**Del 5q35.3ter**	
Mild developmental delay, macrocephaly, bell-shaped chest, brachydactyly, dysmorphic facies with telecanthus and anteverted ears		**Del 5qter**	
Cardiac defects, developmental delay, anterior chamber anomalies, hypertelorism, downward slanting palpebral fissures, smooth philtrum and deafness		**Del 6p25**	**Chromosome 6**
Brain, heart and kidney abnormalities, short neck, facial dysmorphism, clino- or syndactyly		**Del 6p22->24**	
Micro-/Brachycephaly, absent pulmonary valve, flat face, hypertelorism, bulbous nose and malformed ears		**Del 6q**	
Developmental delay, autism, seizures, hypotonia, mild microcephaly, enlarged ventricles, absent corpus callosum, ear anomalies, epicanthal folds and flat philtrum		**Del 6q27**	
Severe mental retardation, low birth weight, microcephaly, hypertelorism, ptosis, prominent nasal bridge, small mouth, pointed chin and low-set ears		**Dup 6p**	
Craniosynostosis, urogenital and cardiac defects, facial dysmorphism, mental retardation, anomalies of hands and feet		**Del 7p21->pter**	**Chromosome 7**
Craniosynostosis, mental and growth retardation, craniofacial abnormalities		**Del 7p15.3->21.2**	
Supravalvular aortic and/or peripheral pulmonary artery stenoses, elfinlike hypotonic face with thick lower lip, large mouth, long smooth philtrum, periorbital fullness, stellate iris pattern, full cheeks, dental anomalies, constipation and feeding difficulties, hypercalcemia in infancy, moderate growth and developmental delay, outgoing personality		**Del 7q11.21**	
Mental retardation, cardiac and genitourinary abnormalities, dysmorphic facies		**Recomb 8**	**Chromosome 8**
Congenital heart malformations, microcephaly, IUGR, mental retardation, characteristic hyperactive impulsive behaviour		**Del 8pter**	
Pre-/Postnatal growth retardation, microcephaly, narrow forehead, epicanthus, moderate-severe mental retardation, congenital heart defects		**Del 8p**	
Severe growth and mental retardation, congenital heart disease, absent gallbladder, renal and skeletal anomalies, facial dysmorphism		**Dup 8q**	
Cardiac, renal and skeletal defects, large babies with deep palmar and plantar creases, coarse expressionless face with thick lips, prominent ears, absent/dysplastic patellae, velopharyngeal insufficiency, mild-moderate mental retardation		**Mosaic trisomy 8**	
Dysmorphic and mentally handicapped		**Del 9q22.1->22.32**	**Chromosome 9**
High birth weight and macrocephaly, mental retardation, triangular face, frontal bossing, epicanthal folds, small mouths and thin upper lips		**Del 9q22.3**	
Kleefstra syndrome: Hypotonia, developmental delay, natal teeth, single umbilical artery, microcephaly, facial dysmorphism		**Del 9q34**	
Intrauterin growth retardation, ventriculomegaly, contractures, renal anomalies, mental retardation, hypertelorism, beaked/bulbous nose, cleft li/palate		**Tetrasomy 9p**	

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Mental disability, pre- and postnatal growth retardation, ano/genital defects, cardiac and renal anomalies, microcephaly, triangular face, hypertelorism, strabismus, prominent nasal bridge, low-set ears, micrognathia, short neck		**Del 10q26**	**Chromosome 10**
Mental and growth retardation, skeletal, heart and renal anomalies, microcephaly, round and flat face, high-arched eyebrows, downslanting short palpebral fissures, tender upper lip		**Dup 10q**	
Heart and urogenital defects, thrombocytopenia, trigonocephaly, hypertelorism/telecanthus, short nose, microretrognathia, carp-shaped mouth		**Del 11q**	**Chromosome 11**
Pyloric stenosis, growth and developmental delay, hypertelorism, ptosis, low-set and rotated ears, sparse hair		**Del 12q**	**Chromosme 12**
Moderate mental retardation, bouts of aggressive behaviour, normal growth, microcephaly, prominent forehead, hypoplastic supraorbital ridges, long eyelashes, deep-set eyes, strabismus, paranasal broadening, unilateral cleft lip and palate, large mouth with cubid-bow-shaped upper lip, small ears		**Del 12q24**	
Ulnar deviation, developmental delay, cardiac anomalies, shawl scrotum, facial dysmorphism		**Dup 12p**	
Brain malformations (holonprosencephaly), cleft lip/palate, polydactyly, variable organ defects		**Trisomy 13**	**Chromosome 13**
Relatively mild phenotype including mild developmental delay, craniofacial dysmorphism, pectus excavatum, narrow shoulders, malformed toes, and cafe-au-lait spots (der Kaloustian 2011)		**Del 13q12.11**	
Growth and mental retardation, congenital heart disease, micropenis/undescended testes, body asymmetry, streaky/linear hyperpigmentation, facial dysmorphism, short neck		**Mosaic trisomy 14**	**Chromosome 14**
Craniosynostosis, facial dysmorphism, severe mental retardation, tetralogy of Fallot, limb anomalies, late-onset obesity, scalp defect		**Del 15q15->q22.1**	**Chromosome 15**
Postnatal growth and mental retardation, bulbous nose, arachno-/camptodactyly, cardiac and genital defects		**Dup 15q**	
Left microphthalmia, persistent hyperplastic primary vitreous and posterior coloboma, right posterior pole coloboma, pectus excavatum, mild hypotonia, mild delays in speech and motor development, and anxiety disorder with social difficulties and speech delay(Bardakjian 2010).		**Microdeletion 16p11.2**	**Chromosome 16**
Microcephaly, hyperactivity-attention deficit disorder or autism and dysmorphic, no definitive facial phenotype (Shinawi 2010).		**Microduplication 16p11.2**	
Severe mental retardation, postnatal growth deficiency, hypotonia, seizures, microcephaly, cortical atrophy, partial agenesis of the corpus callosum, facial anomalies, long fingers and bilateral talipes equinovarus		**Del 17p13**	**Chromosome 17**
Pre-/Postnatal growth and mental retardation, heart defect, microcephaly, hypertelorism, downslanting palpebral fissures, thin upper lip, microganthia		**Dup 17p**	
Growth and mental retardation, microcephaly, frontal bossing, widow's peak, downturned mouth, short neck, hirsutism with sparse sclap hair		**Dup 17q**	
Growth and mental retardation, seizures, skeletal and genital defects, facial dysmorphisms		**Del 18p**	**Chromosome 18**
Mental retardation, abnormal behaviour, obesity, dysmorphic features		**Del 18q12.1->q21.1**	
Pre-/Postnatal growth deficiency, developmental delay, microcephaly, round face, hypertelorism, small nose, low-set and dysplastic ears, preauricular pits, sensorineural hearing loss, small chin, swallowing difficulties, ventricular septal defect, III/IV cutaneous syndactyly		**Dup 18p11**	

(continued)

Table 8.2a (continued)

Syndromes	Inheritance	Gene location	Gene
Developmental delay, abnormal craniofacial features and ventricular septal defect, occipital plagiocephaly, large forehead, bilateral epicanthic folds, small bulbous nose with slightly anteverted nares, thin upper lip, dimpled chin, low set ears, oblique crease of the ear lobules, pectus excavatum, small umbilical hernia and single palmar crease (Blanc 2008).		**Dup 20q13.2**	**Chromosome 20**
Cardiac and intestinal defects, peculiar facial grimacing with tongue thrusting, hypotonia and delayed motor milestones, increased leukaemia risk		**Trisomy 21**	**Chromosome 21**
Congenital lymphedema of hand and/or feet, short stature, short/webbed neck, low posterior hairline, cubitus valgus, nail hypoplasia, broad chest, infertility, left-sided cardiac anomalies		**Monosomy X**	**X Chromosome**
Multiple congenital anomalies with mental retardation, short stature, facial dysmorphisms		**Dup Xq**	
Mental retardation, cleft palate or bifid uvula, coarse facies, radioulnar synostosis, hypogenitalism and cardiovascular defects, verbal skills extremely poor – Klinefelter syndrome		**49, XXXXY**	
Small baby with joint contractures, dysmorphic facies and multiple congenital anomalies		**Tetraploidy**	

Adapted and updated from information found in "Etiology of chest wall deformities – a genetic review for the treating physician." By Kotzot and Schwabegger [13]; *AD* indicates autosomal dominant and *AR* autosomal recessive

activated protein kinase) pathway which is involved in cell proliferation, – differentiation, – survival and – death [17] and which is also associated with other syndromes such as the LEOPARD syndrome, the cardiofaciocutaneous syndrome, the Costello syndrome and other RASopathies [19, 20]. In 50 % the mutated gene is the protein tyrosine phosphatase nonreceptor type 11 (PTPN11). The incidence of Noonan syndrome is reported to be 1:1000–2500 births with a weak genotype-phenotype correlation [17]. Due to autosomal dominant inheritance, the recurrence risk for the patient's offspring is 50 %. However, as the phenotype is variable, familiar genetic counselling is recommended for evaluating the recurrence risk for siblings of non-syndromic parents [13].

Noonan-like/Multiple Giant Cell Lesion Syndrome

Noonan-like/Multiple Giant Cell Lesion Syndrome (NS/MGCL) was until recently considered to be a distinct syndrome differing from Noonan by the development of multiple giant cell lesions of the jaw, resembling a keratocytic odontogenic tumour (Fig. 8.2) [21]. Nowadays it is considered as a phenotypic variation within the Noonan syndrome and other syndromes associated with the Ras/MAPK pathway [22]. According to the genes of the syndromology following the Ras/MAPK pathway, gene mutations of PTPN11, SOS1, RAF1, BRAF or MAP2K1 have been documented in patients developing MGCL [23]. Usually, all patients with NS/MGCL present with the typical dysmorphic features of NS. The MGCLs of the mandible and to a lesser extent of the maxilla are usually noted at premature age and generally present with enlargement of the jaws [23]. Histologic examinations show cystic masses consisting of multinuclear giant cells of the osteoclastic type and collagen fibres [23]. Recurrence rates after resection or curettage of the lesions is reported with 11–49 % [24]. Therefore, regarding a MGCLs a wait-and see attitude until physical maturity was recommended [23].

Marfan's Syndrome

Marfan's syndrome is a well-known connectivopathy that manifests due to several identified mutations in the gene which encodes for the glycoprotein fibrillin-1 (FBN1), localized on chromosome 15q21. It affects 2–3 cases in 10.000

Table 8.2b Proven or suspected monogenic syndromes with cleft sternum listed in POSSUM (Pictures Of Standard Syndromes and Undiagnosed Malformations, version released 2013) and/or WBDD (Winter-Baraitser Dysmorphology Database, version 1.0.14)

Syndromes	Inheritance	Gene location	Gene
Achondrogenesis 2 Langer-Saldino type achondrogenesis	AD	**12q13.11-q13.2**	COL2A1
Achondrogenesis, type 1B	AR	**5q32.1-q33.1**	DTDST
Acro-pectoro-vertebral dysplasia F syndrome	AD	**2q36**	
Al-Awadi-Raas-Rothschild syndrome Limb/pelvis-hypoplasia/aplasia syndrome	AR	**3p25.1**	WNT7A
Amniotic band syndrome Early amnion rupture spectrum	sporadic		
Arthrogryposis, craniofacial and ophthalmological anomalies – Arthrogryposis multiplex congenita, Al-Ghamdi type	AD?		
Atelosteogenesis, type 3	AD	**3p14.3**	FLNB
Asternia			
Bohring (1996) midline body wall defects – facial anomalies			
Campomelic dysplasia Camptomelic dwarfism	AD	**17q24.3-q25.1**	SOX9
Cantrell pentalogy	X-linked	**Xq25-26.1**	
Cleidocranial dysplasia – Marie-Sainton disease	AD	**6p21**	RUNX2
Coffin-Lowry syndrome	X-linked	**Xp22.2-p22.1**	RSK2
Coffin-Siris syndrome – Brachymorphism-onychodysplasia-dysphalangism syndrome	AD/AR?	**7q32-34**	ARID1A ARID1B
Congenital hypothyroidism	AD/AR		
Craniofacial dysostosis, omphalocele, limb defects			
Craniosynostosis, fibular aplasia – Lowry syndrome	AR		
C syndrome	AR	**3q13**	
Dandy-Walker malformation – facial hemangiomas	AR		
Desbuquois syndrome, Spondylometaphyseal dysplasia, advanced bone age Chondrodystrophy, joint dislocation, glaucoma, mental retardation Spondylometaepiphyseal dysplasia, Piussan-Maroteaux type	AR	**17q25.3**	CANT1
Dysosteosclerosis	AR/X-linked?		
Ectopia cordis – cleft lip/palate			
Fetal varicella syndrome			
Goltz – focal dermal hypoplasia	X-linked	**Xp22, 9q32-34**	PORCN
Herrmann-Opitz arthrogryposis syndrome VSR syndrome	AD		
Kaplan, et al. 'Ectopia cordis and cleft sternum: evidence for mechanical teratogenesis following rupture of the chorion or yolk sac', (1985)			
Mental retardation, hypopituitarism, distal arthrogryposis	AR?		
Mental retardation, polydactyly, uncombable hair, multiple congenital anomalies., Kozlowski-Krajewska type	AR/X-linked		
Mesomelia-synostoses syndrome, Acromesomelia, synostoses, soft palate agenesis with facial and renal abnormalities, mesomelic short stature, acral synostosis – Verloes-David-Pfeiffer type	AD	**8q13**	

(continued)

Table 8.2b (continued)

Syndromes	Inheritance	Gene location	Gene
Midline raphe, sternal cleft, dental anomalies	AD		
Miles-Carpenter – x-linked mental retardation, fingertip arches, contractures	X-linked	**Xq21**	
Ozlem (2008) – anophthalmia-anal atresia-rhizomelia	AR		
MOMO syndrome, Macrosomia, Obesity, Macrocephaly, Ocular abnormalities			
Multiple anomalies, mental retardation – Teruel type	AR		
Multiple anomalies, mental retardation, ventral midline anomalies, Bohring type			
Multiple congenital anomalies – Van Allen-Myhre type			
Ocular colobomas, cleft palate, genital, skeletal, and craniofacial abnormalities, mental retardation – Khalifa type	AR/X-linked?		
Perlman syndrome – Renal dysplasia, Wilms tumour, foetal gigantism, hamartomas	AR	**2q37.2**	DIS3L2
PHACE syndrome, Haemangioma, cardiac/eye/brain anomalies	X-linked?		
Pterygium colli medianum – midline cercical cleft			
Pulmonary agenesis – unilateral	AR		
Raas-Rothschild, et al. 'Giant congenital aortic aneurysm with cleft sternum, supraumbilical raphe, and hemangiomatosis: report and review' (2000)			
SCARF syndrome	X-linked		
Split hand/foot – tibial defects	AR, AD		
Sternal clefts – teleangiectasia/hemangiomas			
Sternal malformation-vascular dysplasia association – Sternal cleft, facial haemangioma	AD?		
Teruel (1987) – absent abdominal musculature, microphthalmia, joint laxity	AR		
Thoracoabdominal schisis, limb defects			
Uygur (2004) – omphalocele, ectopia cordis, absent tibia, oligodactyly			
Thoracoabdominal syndrome Midline defects	X-linked	**Xq25-q26.1**	
Van Allen-Myhre – ectopia cordis, split hand/foot, skin defects	AR		
Weyers syndrome – Weyers' ulnar ray/oligodactyly syndrome, Cleft lip/palate, ulnar hypoplasia	AD?		
X-linked mental retardation, alpha-thalassaemia	X-linked	**Xq13**	ATRX
Yunis-Varon – cleidocranial dysostosis plus amniotic band syndrome	AR		
Chromosome aberrations			
Partial agenesis of corpus callosum, ASD, dysmorphic features include frontal bossing, prominent occiput, low set ears, micrognathia, short sternum, short and broad hands and feet (Fagan 1989)		**Del 4q21.1-q22.1**	**Chromosome 4**
		Submicroscop. Del 9q22	**Chromosome 9**

Adapted and updatedfrom information found in "Etiology of chest wall deformities – a genetic review for the treating physician." By Kotzot and Schwabegger [13]; *AD* indicates autosomal dominant and *AR*, autosomal recessive

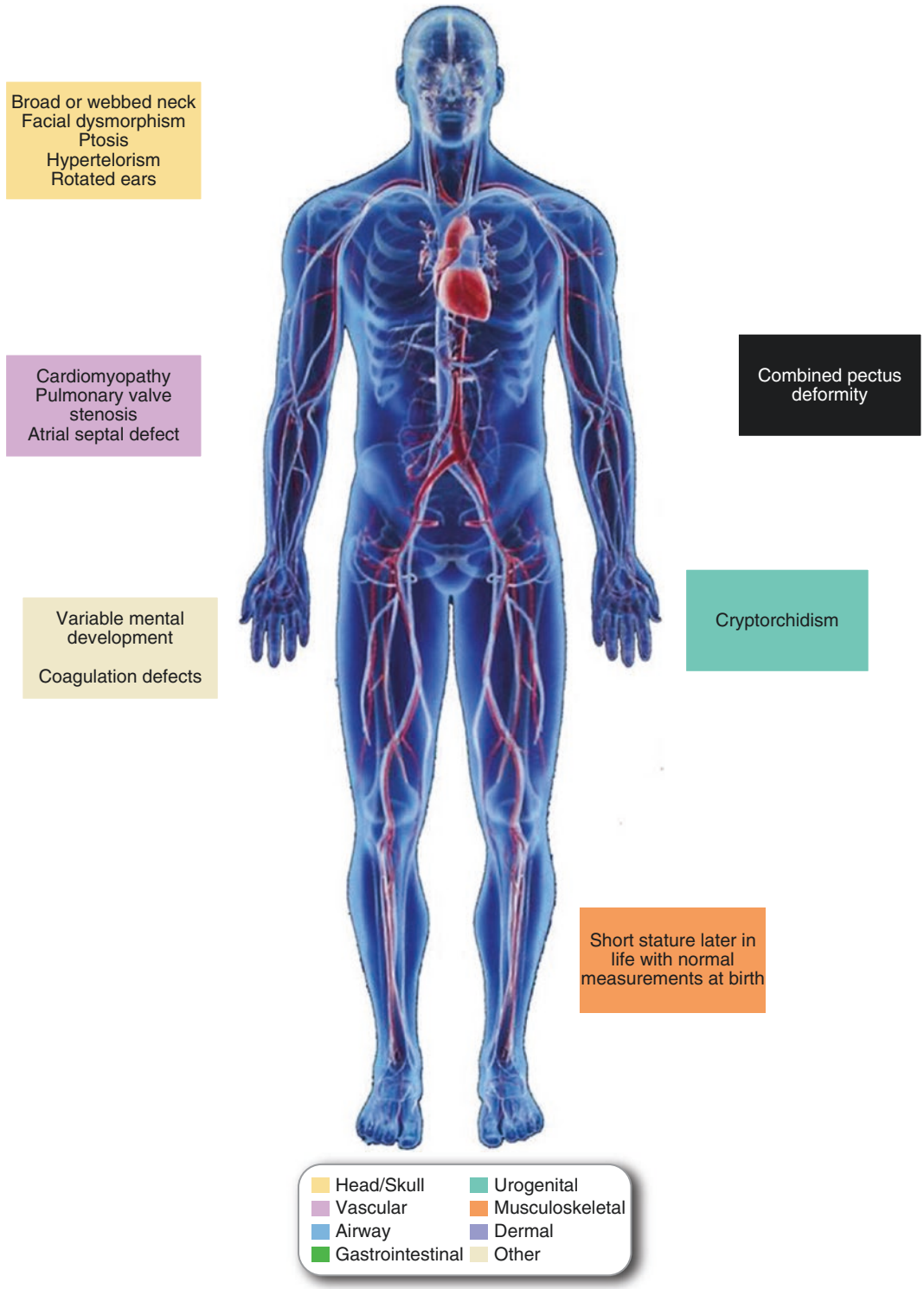

Fig. 8.1 Schematic characteristics of Noonan's syndrome

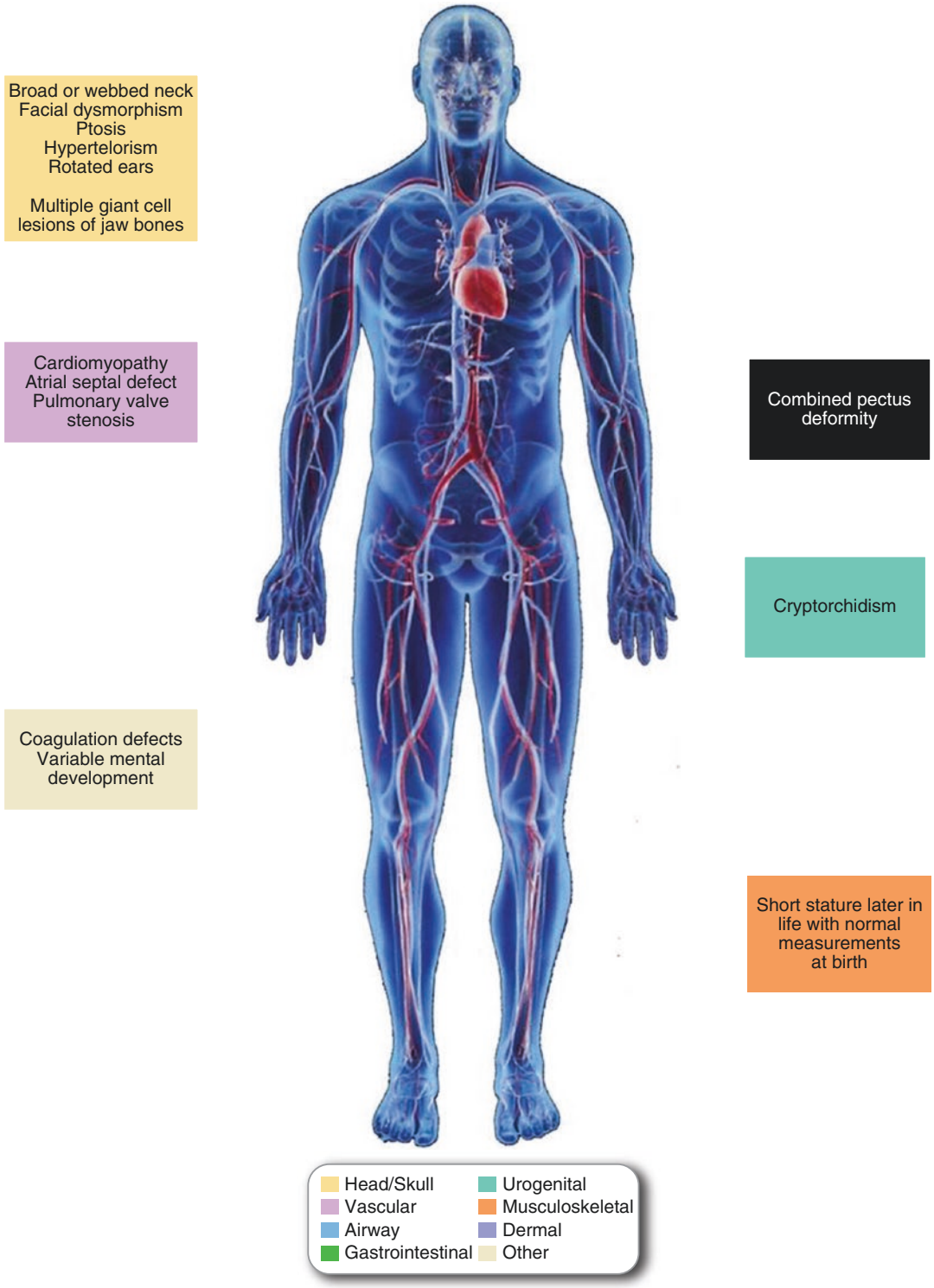

Fig. 8.2 Schematic characteristics of Noonan-like/Multiple Giant Cell Lesion Syndrome

births and about 2 thirds of the patients present with a pectus excavatum [25, 26]. The penetrance is described as almost 100 % with interfamiliar and intrafamiliar variability. Although most mutations are de novo with a low recurrence risk for sibs, the risk for recurrence in a patient's child is 50 % [13]. Multiple organ systems are involved in Marfan's syndrome varying in range and severity from isolated features to a severe presentation with a poor genotype-phenotype correlation, with the cardiovascular, respiratory, ocular, and skeletal system mostly being affected [27] (Fig. 8.3). Apart from family history,, diagnosis of Marfan's syndrome is established based on presence of the clinical hallmarks ectopia lentis and aortic root dilatation/aneurysm, detection of the genetic mutation and other clinical manifestations as defined in the revised Ghent criteria [6] (Tables 8.1a and 8.1b). Cardiovascular diseases such as aortic dilatation or dissection predominantly cause morbidity and mortality of these patients [28, 29] and have to be carefully assessed when a **PE** correction is planned. Regarding the optimal time-point of surgical PE correction, it has been proposed that, if possible, the surgical repair should be performed until the skeletal maturity is nearly completed [30]. A substernal strut such as used in the minimal invasive repair of PE according to Nuss seems to be essential [26] and is associated with a good outcome [25, 26].

Loeys-Dietz Syndrome Type IB

The Loeys-Dietz syndrome (LDS) represents a rare autosomal dominant inherited syndrome that is characterized by the triad of arterial tortuosity and aneurysm, hypertelorism and bifid uvula or cleft palate [31, 32] (Fig. 8.4). This genetic syndrome is caused by an increased transforming growth factor ß (TGF-ß) -signalling due to a heterozygous mutation of the TGF-ß receptor type I or II genes (TGFBR1 or −2), resulting in a predisposition to aggressive and widespread vascular disease, characterized by aggressive arterial dilatation and aneurysms that have a greater risk of rupture or dissection which describes the major difference to Marfan related disorders [31–35]. Depending on clinical manifestations the syndrome is classified as LDS type I or II and according to the underlying gene mutation TGFBR1 or −2 subclassified into LDS Type I or II A/B respectively [31, 32]. LDS Type I patients have vascular and craniofacial symptoms such as classic hypertelorism, bifid uvula or cleft palate, but also craniosynostosis, dolichostenomelia, blues sclerae, malar hypoplasia and retrognathia, while LDS Type II patients do not have any craniofacial dysmorphic features besides an isolated bifid uvula but represent at least 2 signs of vascular Ehler-Danlos syndrome characterized by translucent or velvet skin, joint laxity, wide atrophic scars, bruisability or visceral rupture) [31, 32]. Further manifestations may be mild developmental delay, neurocognitive anomalies, congenital heart defects, as well as skeletal anomalies such as **pectus deformity**, scoliosis, talipes equinovarus, arachnodactyly or camptodactyly [31, 32]. In 25 % of patients LDS is inherited from an affected parent in an autosomal dominant manner, while in 75 % LDS is caused by a de novo mutation of the TGFBR1 or −2 genes [36]. In order to prevent catastrophic vascular events, cardiovascular evaluation and genetic screening for TGFBR1 and −2 mutations are recommended in every patient suspicious of LDS [36].

Poland's Syndrome

Poland Alfred first reported on a patient with absence of the pectoralis major muscle and syndactyly on the ipsilateral side in 1841 [37] which was defined as Poland's syndrome/anomaly 100 years later [38]. Poland's syndrome is a mostly sporadic occurring disruption sequence and characterized by a deformity of the thoracic wall, of the upper extremity and vertebral anomalies and affects the right side in about 75 % (Fig. 8.5). It usually includes a Sprengle deformity (high scapula or congenital high scapula), scoliosis, **PE**, hypoplastic or fused ribs and possible hypoplastic or absence of the pectoralis major muscle, the nipple or even of the entire breast, but also brachy -, oligo- or syndactyly. Its incidence is about 1 in 25,000–32,000 births and usually

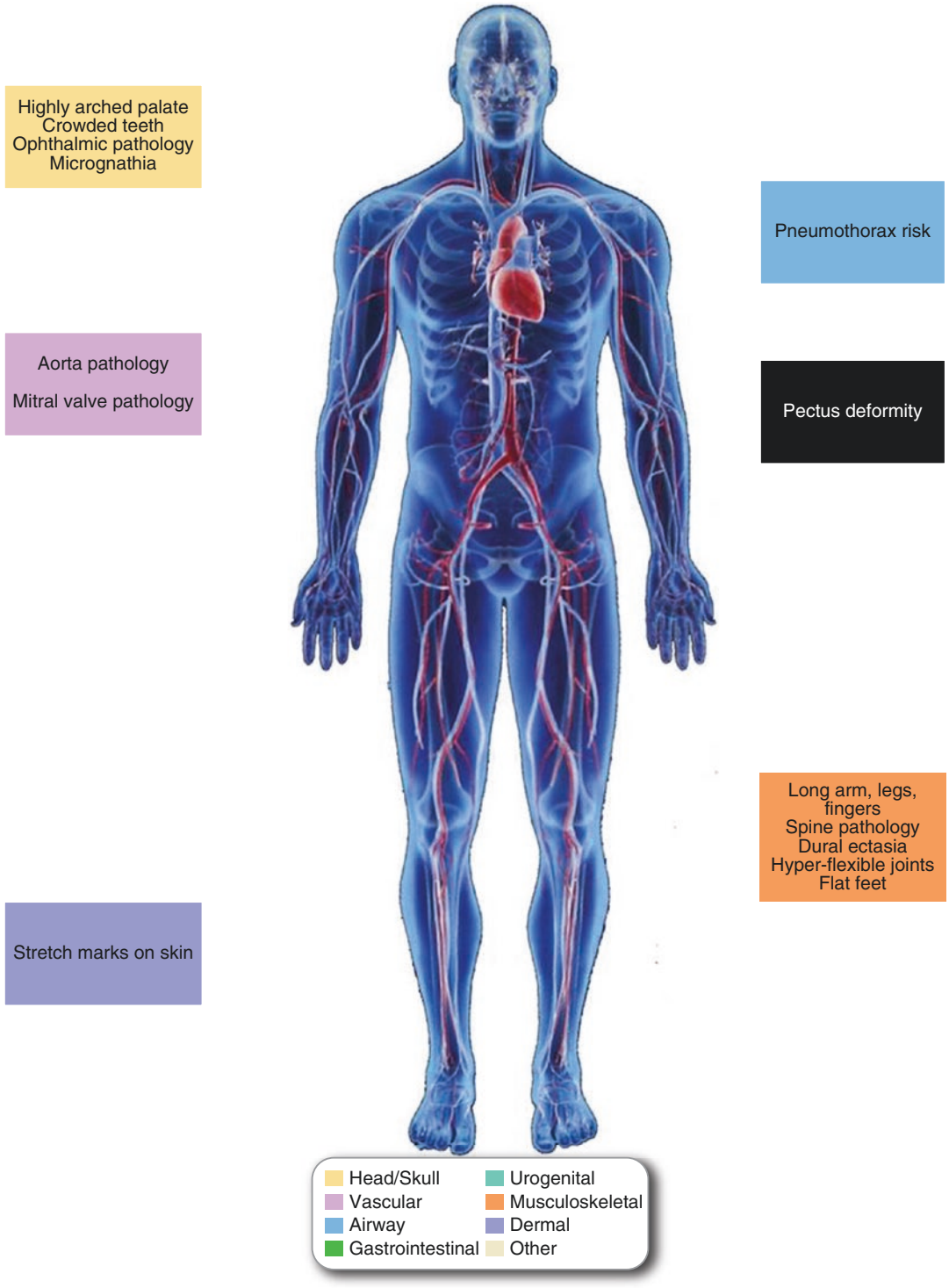

Fig. 8.3 Schematic characteristics of Marfan's syndrome

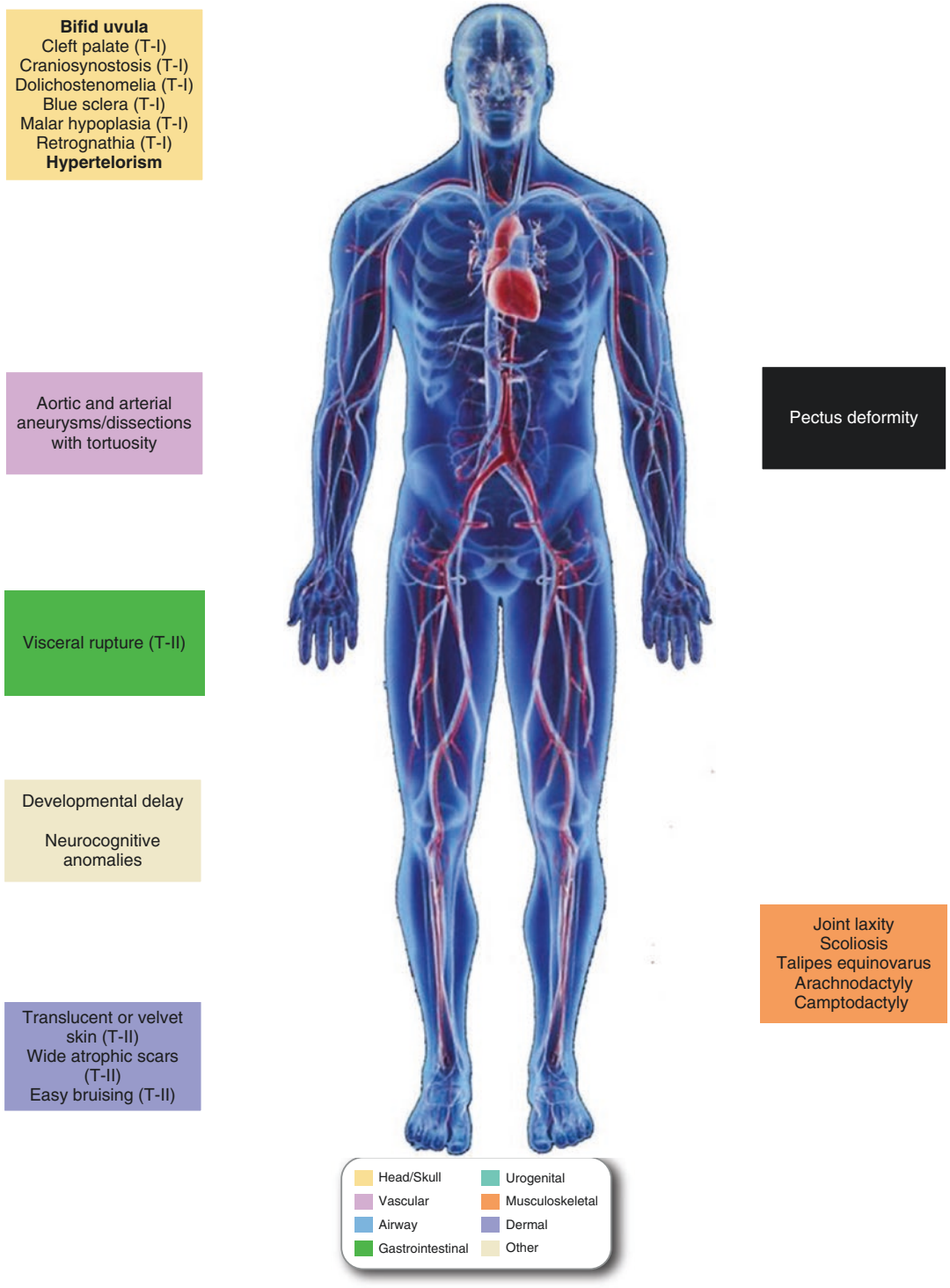

Fig. 8.4 Schematic characteristics of Loey-Dietz's syndrome

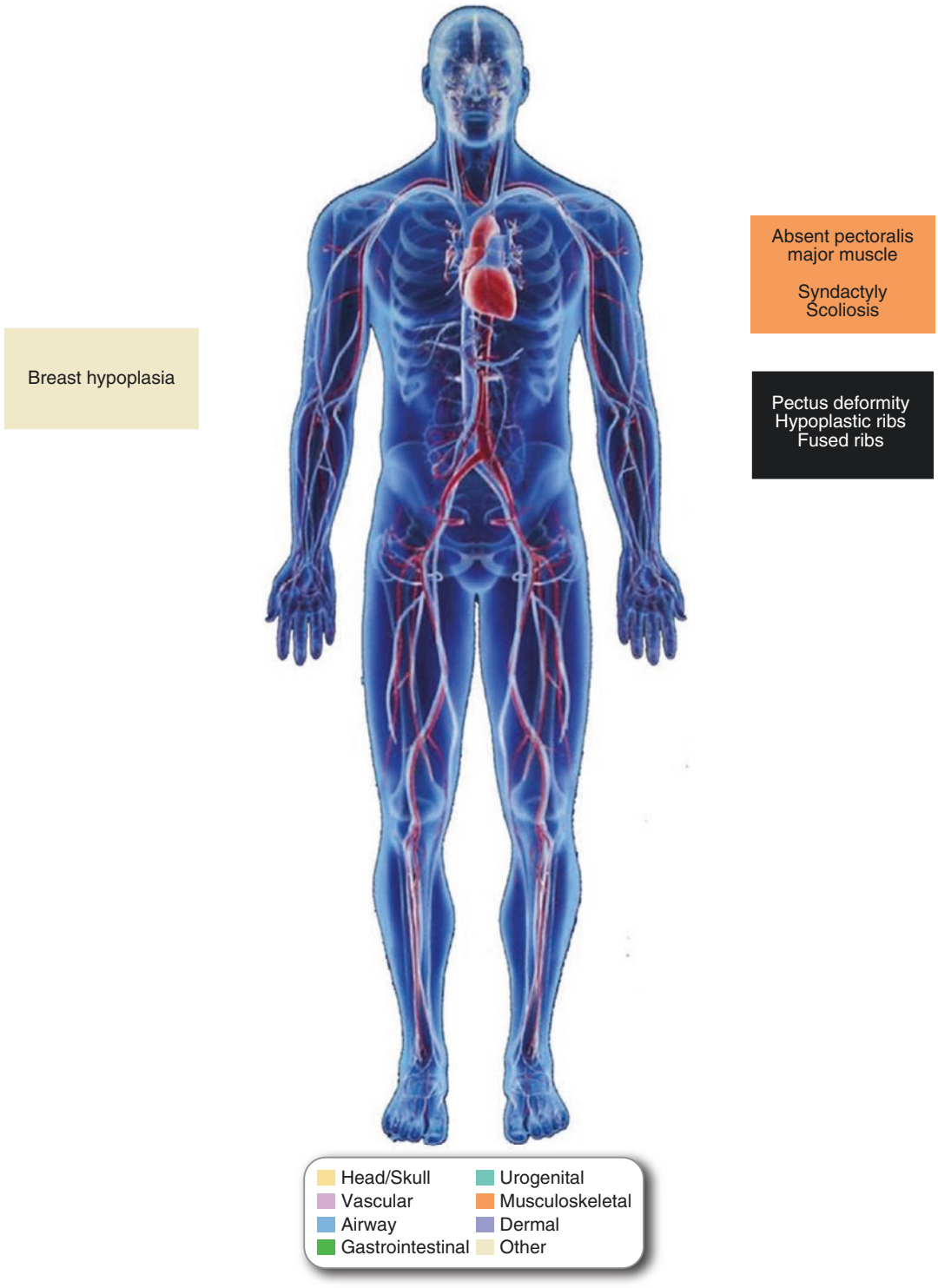

Fig. 8.5 Schematic characteristics of Poland's syndrome

affects men with a male to female ratio of 2–3:1 [39]. A disruption of the lateral plate mesoderm, or in the subclavian artery supply have been discussed as the underlying cause, which has not been clarified yet [13]. Rare bilateral symptoms have been observed in a few familial cases and then indicate an autosomal dominant inheritance with a recurrence risk of 50 % for the patient's offsprings, whereas a unilateral Poland's anomaly is assumed to be nongenetic with a low recurrence risk for siblings and offsprings [13].

Moebius anomaly

This anomaly represents another disruption sequence which differs from the Poland's syndrome by additional uni- or bilateral facial and abducens palsies and a facial dysmorphology, such as epicanthic folds, micrognathia and/or dysplastic ears [40] (Fig. 8.6). Verzijl et al. [40] described it as a syndrome of rhombencephalic maldevelopment manifesting as general motor disability with poor coordination and respiratory abnormalities. It is as the Poland' anomaly usually a sporadic anomaly with a low recurrence risk for siblings and offsprings, although a rare familial inheritance with autosomal dominant, autosomal recessive and X-linked traits have been suggested [40].

Jeune's Syndrome

The Jeune's syndrome or also termed as "asphyxiating thoracic dystrophy" is a rare autosomal recessive skeletal dysplasia which is characterized by skeletal malformations that involve a small, narrow, chest and variable limb shortness, and is associated with a considerable neonatal mortality due to respiratory distress [41] (Fig. 8.7). Postaxial brachy- or polydactyly of both hands and/or feet are observed in 20 % [42]. In addition to the severely constricted thoracic cage, as the major phenotypic feature of Jeune's syndrome, later in life, variable renal, hepatic, pancreatic, and retinal complications may occur [41]. The gene mutation locus has been mapped to chromosomes 15q13 [43]. Recently, mutations were found in the IFT80 gene, localized on chromosome 3q and encoding for an intraflagellar protein, IFT, which is essential for the development and maintenance of motile and sensory cilia [44]. Prenatal indications for Jeune syndrome are an abnormal small thorax, short limbs, polyhydramnios, and unidentifiable fetal respiratory movements at the prenatal ultrasound examinations [45, 46]. It has been observed that when the patients survive the neonatal period, the thoracic malformation becomes less pronounced and the respiratory problems improve, while the short stature and distal shortening of the limbs become more obvious with age, wherefore intensive care is recommended for the first years of life [41].

De Novo 2.1.: Mb Deletion 13q12.11

Recently, a male patient with a mild developmental delay and minor dysmorphic features due to a new de novo 2.1-Mb heterozygous deletion within the 13q12.11 region was reported on [47] (Fig. 8.8). This deletion contains 16 genes, involving GJA3, GJB2, GJB6, FGF9, IFT88 and LATS2, which have previously not been associated with any of the phenotypic characteristics that were found in this patient. Apart from a developmental motoric and speech delay, the patient displayed dysmorphic facial features which were dolichocephaly with a prominent occiput and bitemporal narrowing, attached ear lobes with underdeveloped helices, droopy eyelids and narrowing of the plapebral fissures, telecanthus and bilateral epicanthic folds, depressed nasal bridge with anteverted nares, a high narrow arched palate, small teeth, and a long smooth philtrum; skeletal dysmorphic features represented by narrow dropping shoulders, mild **PE**, 11 rib pairs, slightly short clavicles, square shaped toes with increased interdigital space between the 2nd and 3rd toes; and skin features involving bilateral distal axial triradii and 4 small café-au-lait spots. A unilateral cryptorchidism was corrected at 1 year of age. There was no family history. It was speculated that the apparently

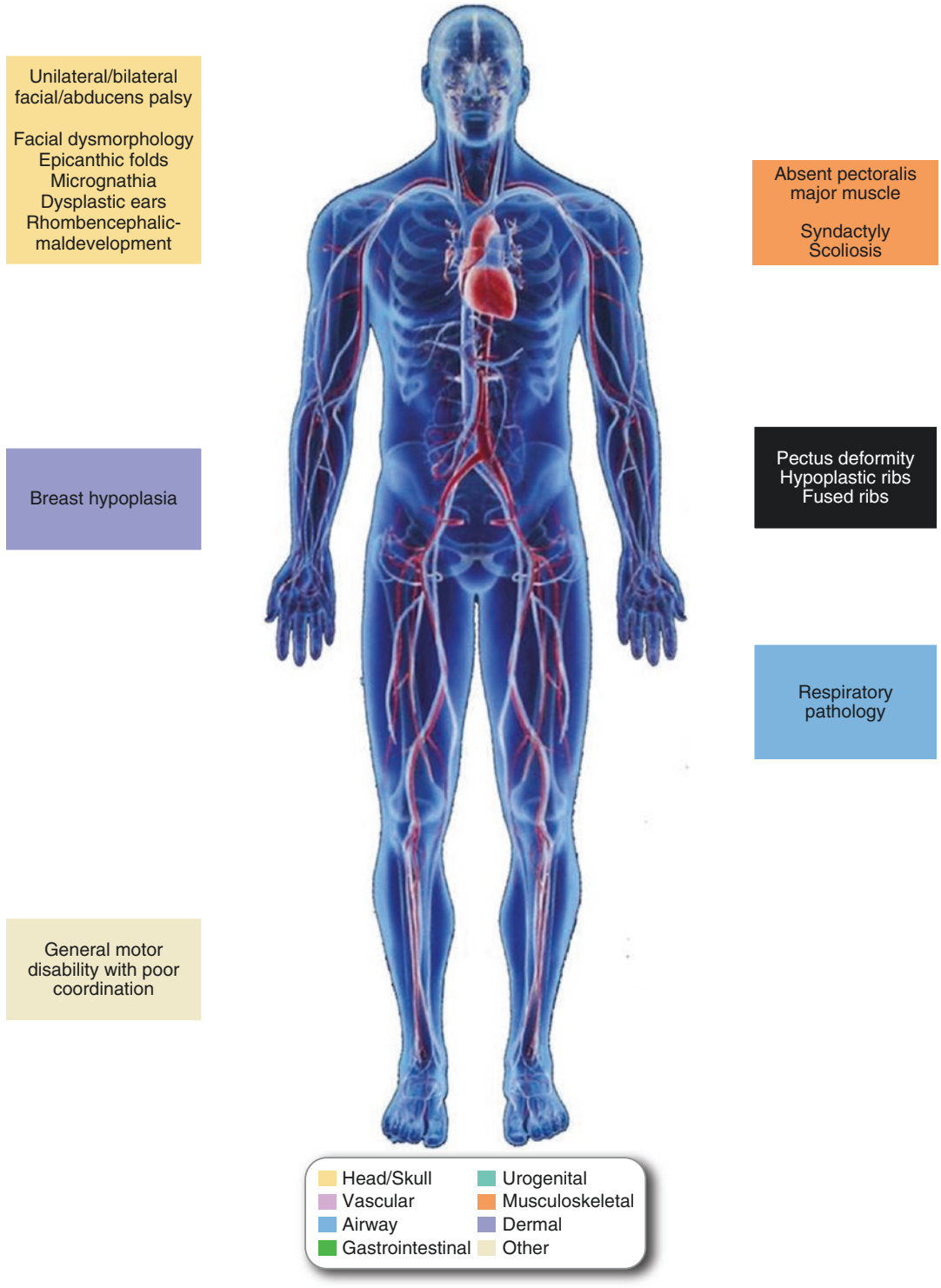

Fig. 8.6 Schematic characteristics of Moebius anomaly

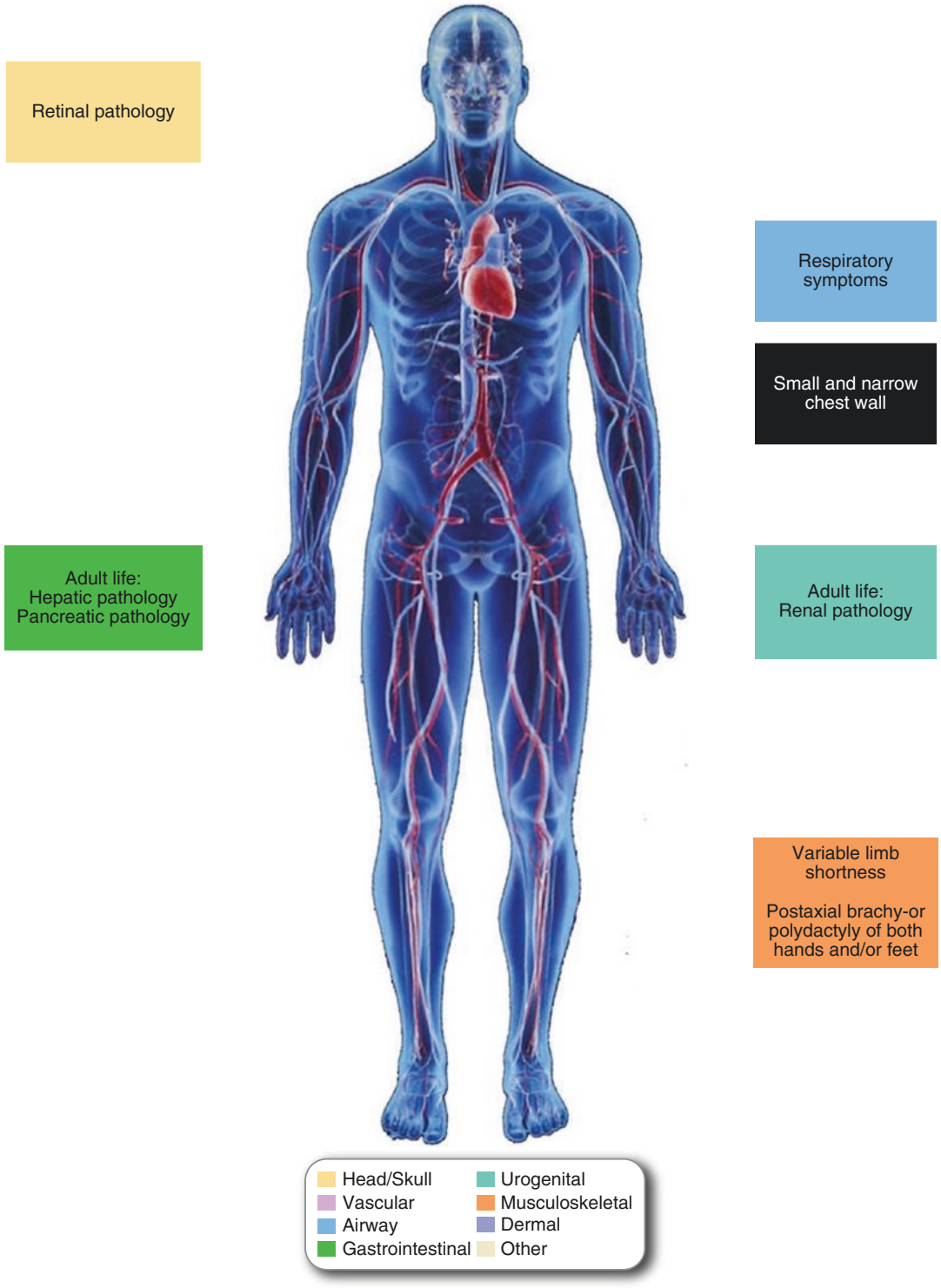

Fig. 8.7 Schematic characteristics of Jeune's syndrome

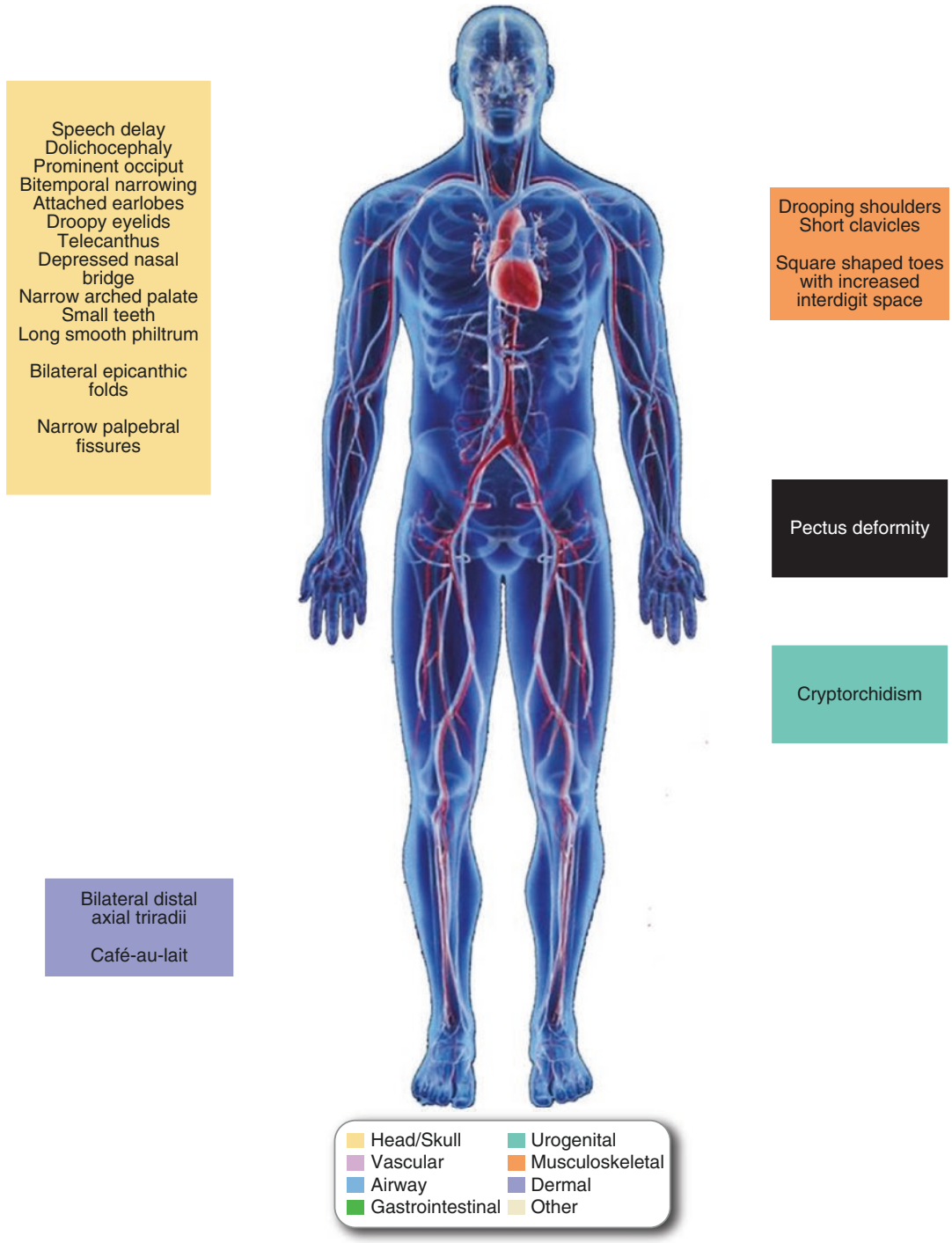

Fig. 8.8 Schematic characteristics of De novo 2.1. – Mb deletion 13q12.11

normal chromosome carries mutations in one of the genes that may cause this contiguous gene deletion syndrome [47].

De Barsy Syndrome

De Barsy syndrome represents a rare syndrome which is characterized by cutis laxa, a progeria-like appearance and ophthalmological malformations (Fig. 8.9). Most patients also present with neurologic and orthopaedic abnormalities. This syndrome was first described by de Barsy in 1967 [48]. A female patient with de Barsy syndrome developing a **PE** during the first year of life was recently described by Kivuva et al. [49]. That patient was born to double-first cousins from a consanguineous Pakistan family. She had the characteristic progeria-like appearance with distinctive facial features such as prominent forehead, brachycephaly, large fontanelles with spaced sutures, hypotelorism, sparse hair, everted lower eyelids and deficient upper eyelids, blue sclera, cloudy corneas, pinched nose with hypoplastic nares and large ears. Furthermore, she had thin, overlapping fingers and thumbs and failed to thrive. There was a marked developmental delay and abnormal athetoid movements [49].

The underlying pathogenesis of de Barsy syndrome has not been elucidated yet. An elastin deficiency associated with functional impairment of other genes was suggested by Guerra et al. [50]. De Barsy syndrome differs from other cutis laxa syndromes by a progeria-like appearance and associated ophthalmological, orthopaedic or neurological abnormalities.

Gorlin-Goltz Syndrome

Gorlin-Goltz, Basal Cell Nevus or Nevoid Basal Cell Carcinoma Syndrome (NBCCS) represents a rare autosomal dominant inherited disorder that may involve all organ systems with complete penetrance and variable expressivity (Fig. 8.10). The hallmarks are odontogenic keratocysts, nevoid basal cell carcinomas and bifid ribs. Its overall prevalence is 1 in 60.000 and it is 3-fold higher in males than females. About 30 % of the patients do not document a family history, and in 40 % a de novo mutation is found. Genetically, the syndrome develops due to a haploinsufficiency of the PTCH1 gene which is a tumor suppressor gene that encodes for the receptor *patched-1* which is involved in the Sonic hedgehog pathway, being responsible for cell proliferation and growth. A chromosomal 9q22.3 microdeletion is also associated with Gorlin-Syndrom, as it leads to the loss of one PTCH1 gene copy [51]. These genetic features result in signs that manifest in the skin, skull and ribs (e.g., pits in the skin of the palms and feet soles, skeletal abnormalities of the spine, bifid, fused or markedly splayed ribs, **pectus deformity** (**e.g.**, **PE**), Sprengel deformity, syndactyly of the digits, cleft lip or palate, macrocephaly, frontal bossing, hypertelorism) and are typically apparent from birth or become evident in early childhood. In case of a high index of suspicion due to clinical major and minor diagnostic criteria [52] and once the diagnosis is established all organ systems should be screened for benign or malignant manifestations. Besides genetic counselling of all family members, a close follow-up is mandatory as malignant tumors such as the nevoid basal cell carcinoma or medulloblastoma may occur and eventually may provoke a fatal outcome [53].

King-Denborough Syndrome

The King and Denborough first reported in 1972 on male patients who developed malignant hyperthermia (MH) (Fig. 8.11) [54]. In a total of 3 studies they described the phenomenon of hyperpyrexia which is thought to be anaesthetic induced and further documented that a few of the MH patients had congenital progressive myopathy, cryptorchidism, **PC**, thoracic kyphosis, short stature and lumbar lordosis with just a few patients also displaying facial features such as ptosis, downslating palpebral fissures, low set ears, micrognathia, crowded teeth and a short webbed neck [54–56]. Nowadays, this syndrome is characterized by the triad of susceptibility to malignant hyperthermia (MH), myopathy and

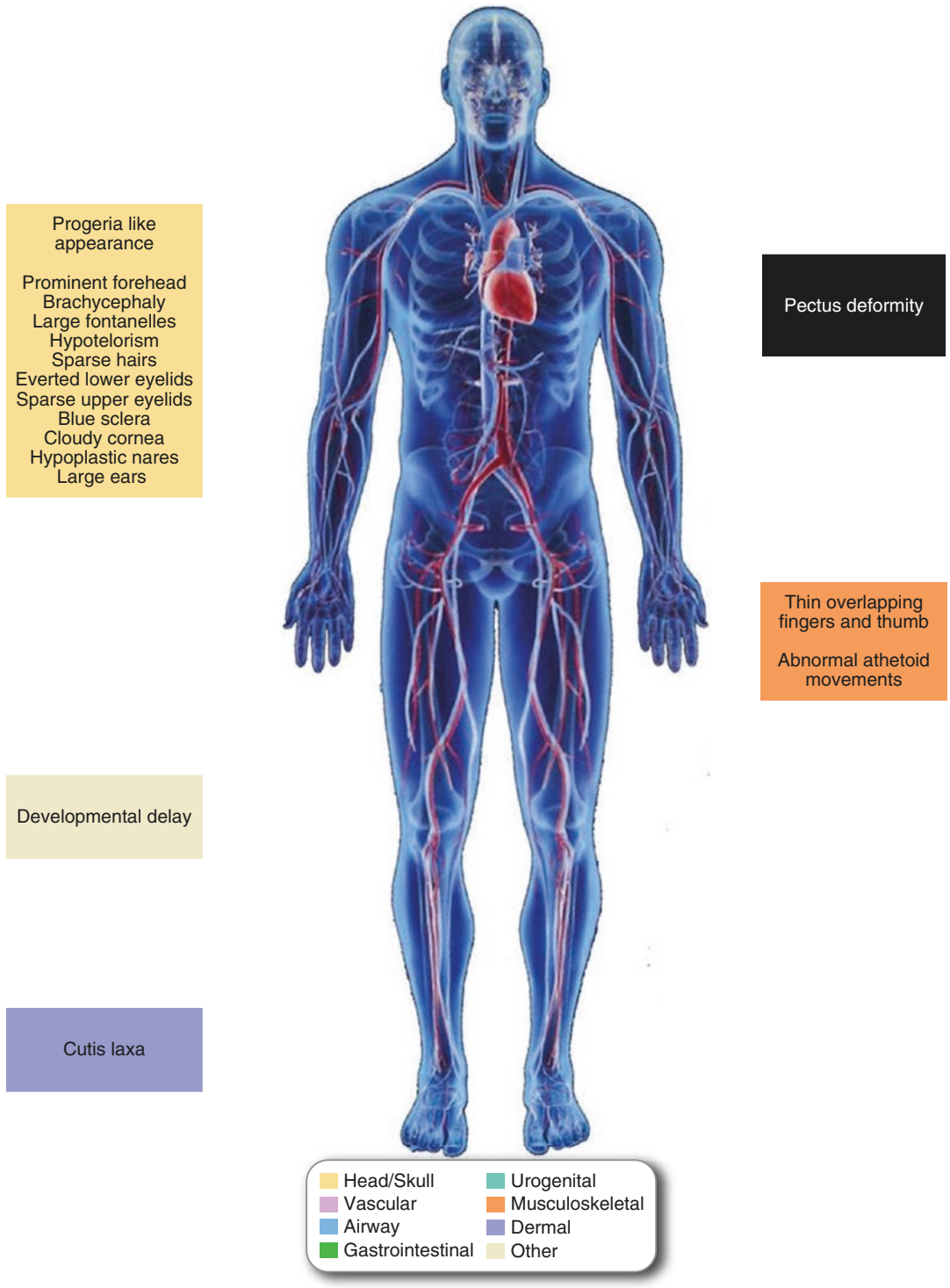

Fig. 8.9 Schematic characteristics of De Barsy Syndrome

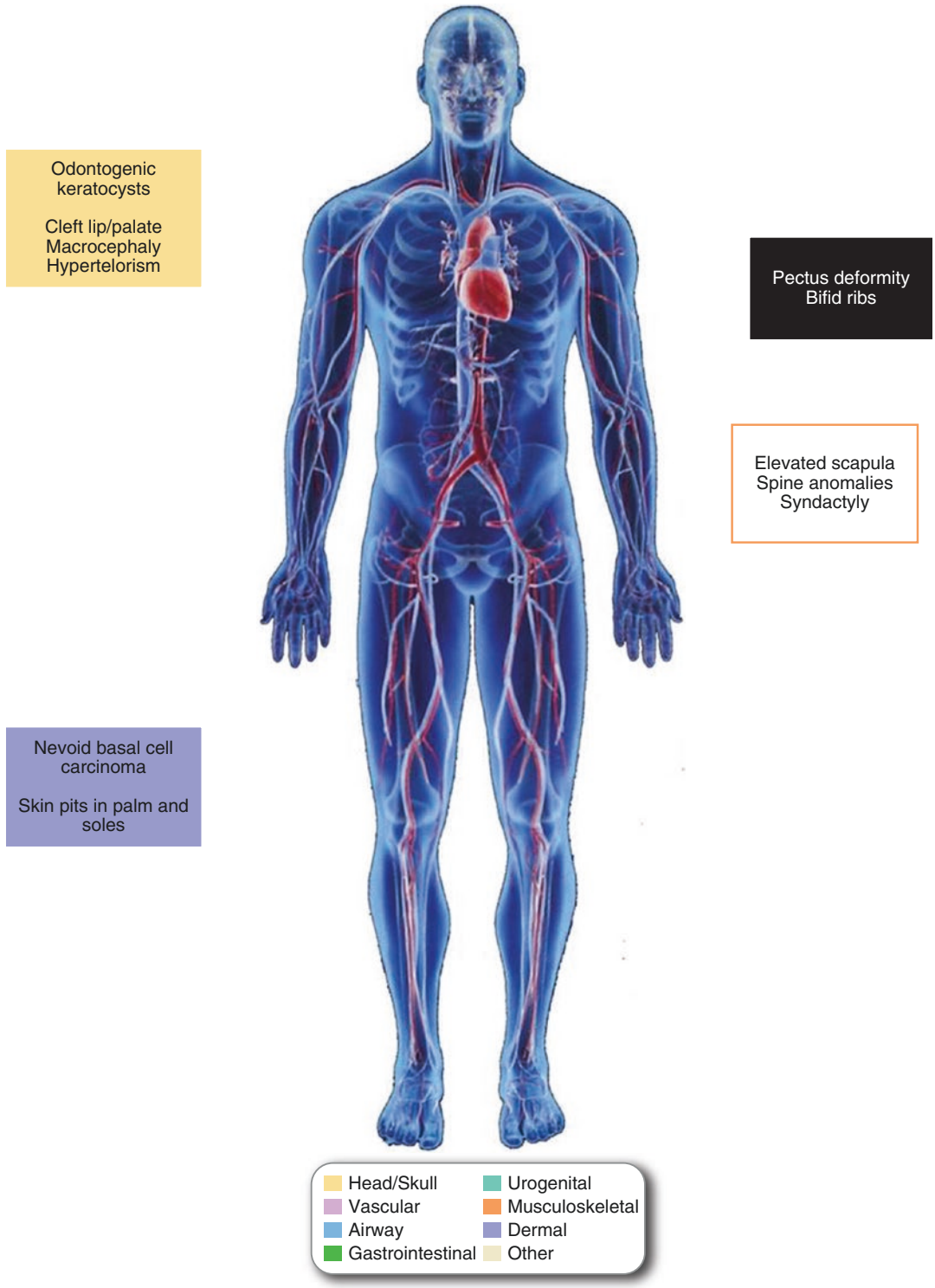

Fig. 8.10 Schematic characteristics of Gorlin Goltz Syndrome

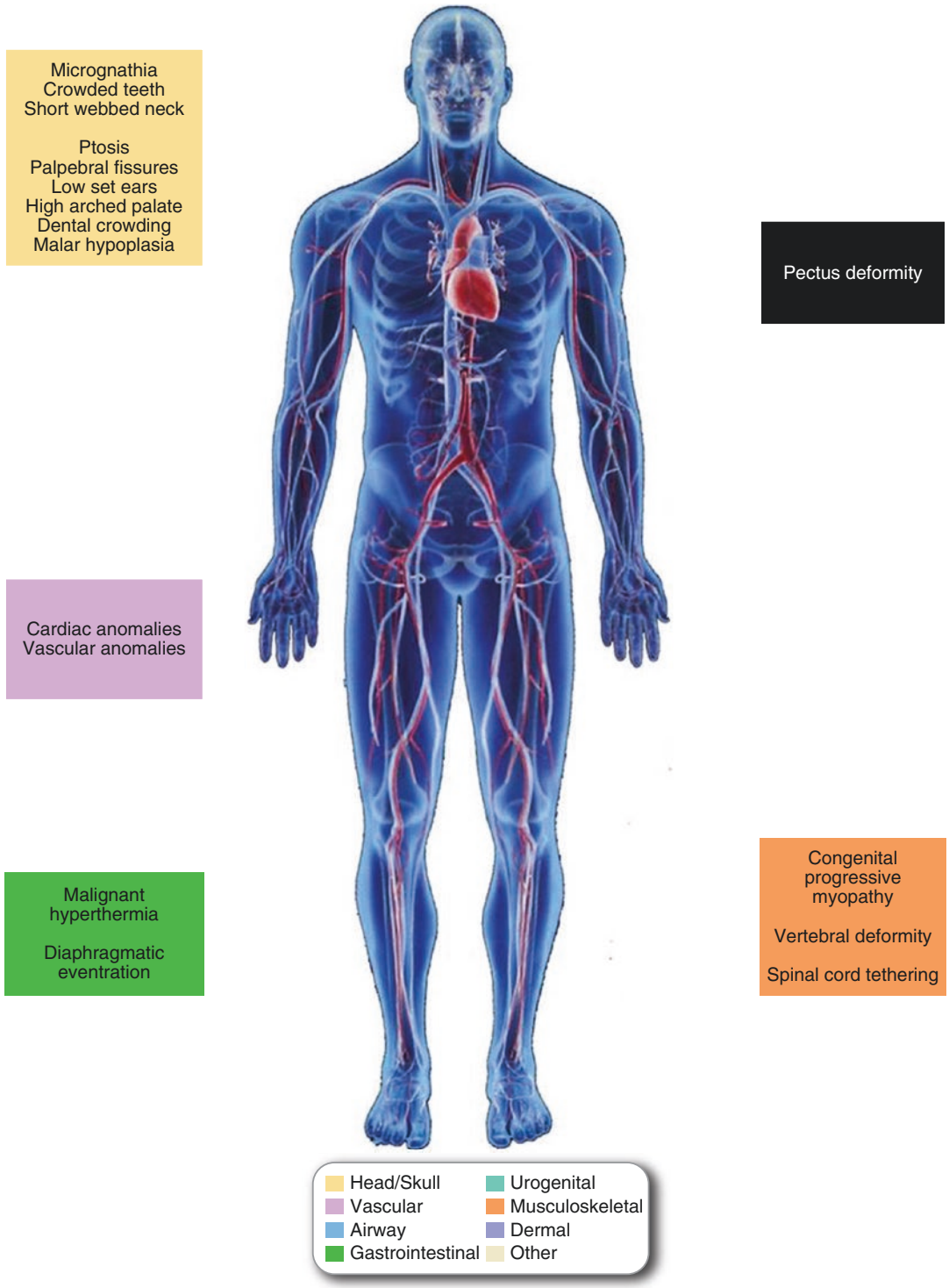

Fig. 8.11 Schematic characteristics of King Denborough syndrome

variable dysmorphic features that include skeletal abnormalities such as scoliosis, kyphosis, lumbar lordosis, and **PC/PE**, vertebral fusion, eventration of the diaphragm, spinal cord tethering and facial anomalies such as ptosis, downslanting palpebral fissures, low-set ears, high-arched palate, dental crowding/malocclusion, malar hypoplasia, micrognathia, and webbing of the neck [57]. The cause of this syndrome, also short termed as the "King Syndrome" or KDS has not been fully elucidated yet. MH may be a primary disease that is autosomal dominant inherited [58] without the underlying myopathy, however, it is frequently observed in patients with muscle disorders such as Duchenne muscular dystrophy [59], myotonic dystrophy [60] or central core disease [61]. It has been documented that resting Creatin-phosphate-kinase (CPK) levels are elevated in some patients [55, 62]. Muscle biopsies have shown variable myopathic features such as variation in fibre size and few, small, or atrophic type I muscle fibres [57, 63–65]. A recent study by Dowling [65] stated an autosomal dominant inheritance with variable expressivity mutations of the skeletal muscle ryanodine receptor (RYR1) gene, which was first documented as a novel heterozygous missense mutation in a KDS patient [66], as the possible underlying cause for KDS but also pointed to a possible genetic heterogeneity. Chitayat has reported on a patient with the features of myopathy, kyphoscoliosis, joint contractures and a facial dysmorphy that was associated with King syndrome but who also displayed unrelated features such as normal stature, hyperextensible joints, **PE**, hypoplastic thenar and hypothenar eminences with clinodactyly of the 5th fingers and 2–5th toes [63]. Further the patient presented cardiac anomalies that combined dilated ventricles, aorta and pulmonary artery with a mild pulmonary insufficiency and palpitations and had normal genitalia. In contrast to other patients this patient underwent 3 anaesthesias without developing MH [63]. Although, it has been described that MH may develop during later anaesthesias [67], a clinical malignant hyperthermia reaction may not be experienced due to a lack of exposure to potentially triggering agents during general anaesthesia or a due to a low penetrance of the malignant hyperthermia susceptibility trait [65]. This observation alerts precaution against MH in every patient who has a myopathic disease or a family history of MH, as MH displays the major cause of death in these patients [63]. Due to the small number of reported cases, diagnostic criteria have not been established yet, especially, as biopsies of the skin and muscle may be inconclusive [63].

Interstitial Deletion 2(p11.2 p13)

Deletion 2p11.2p13 represents a very rare chromosomal aberration which has been reported in 4 cases in literature (Fig. 8.12) [68–71]. As the phenotype has not always been described the phenotype is just beginning to emerge. Wenger et al. [71] documented on a male patient who presented at term birth with eventration of the diaphragm, scoliosis, pectus carinatum, cryptorchidism, hypertonia, long slender fingers with flexion contractures, clinodactyly and overlapping toes, camptodactyly and facial dysmorphism which involved a beaked nose with a transverse crease across the nasal bridge, deep-set eyes with myopia, low-set abnormal ears and high narrow palate. The boy died on an unknown cause at the age of 2 months. The karyogram of the patient revealed the cytogenetic 46,X,del (2) (p11.2p13) karyotype, while the karyotype of the parents was normal [71].

Prune Belly Syndrome

The prune-belly syndrome is characterized by a triad of 3 symptoms which are abdominal muscle deficiency, bilateral cryptorchidism (or undescended testes) and genitourinary abnormalities (Fig. 8.13). It is also termed as the Eagle-Barrett syndrome [72] or the "triad syndrome" [73] and was first documented in 1901 by Osler [74]. The incidence is described as 1 in 29,000–40,000 live births [75]. This syndrome predominantly occurs in male patients with a few either incomplete or questionable cases reported in females [76]. The full spectrum of prune-belly syndrome abnormal-

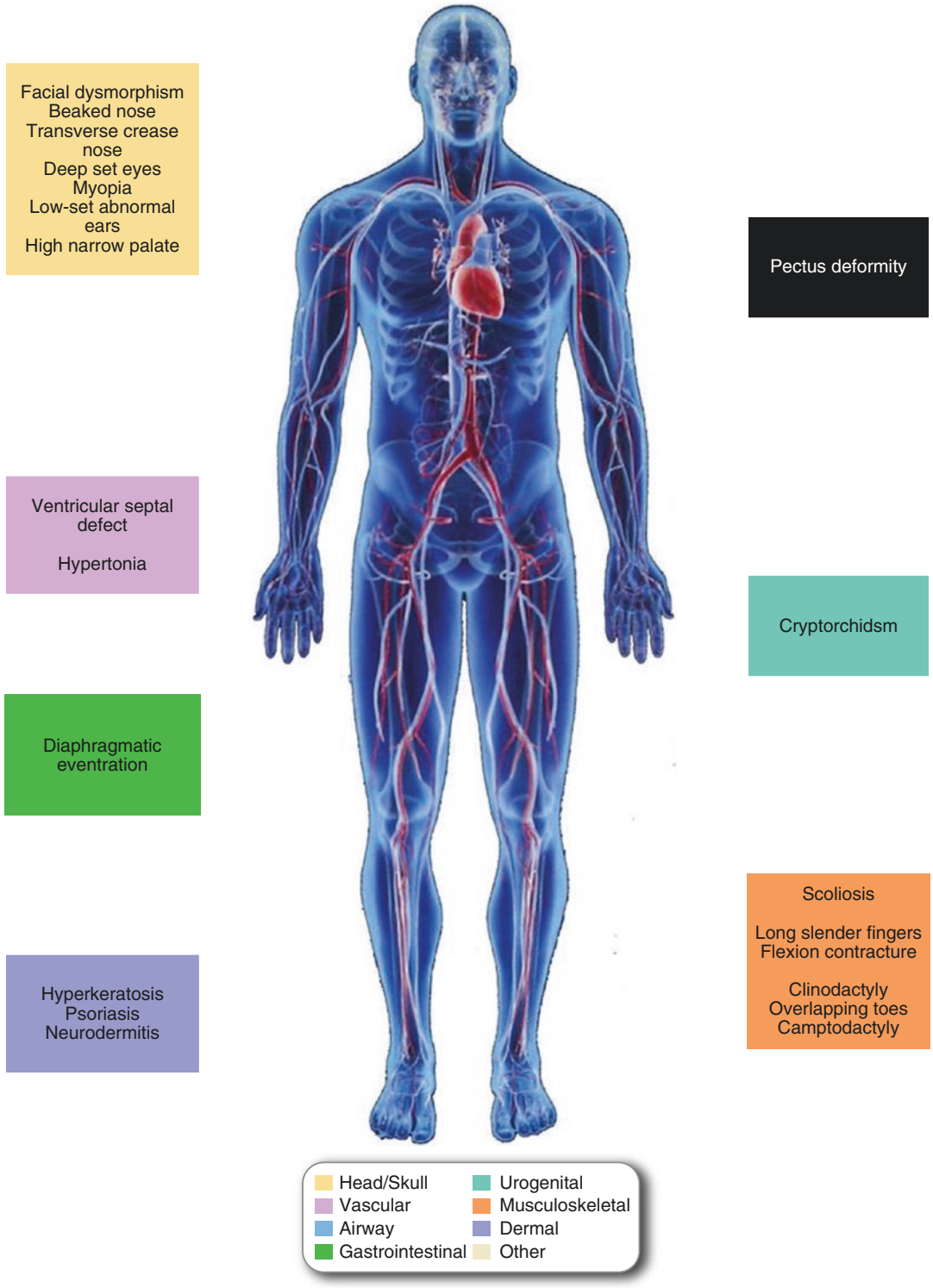

Fig. 8.12 Schematic characteristics of Interstitial Deletion 2(p11.2 p13)

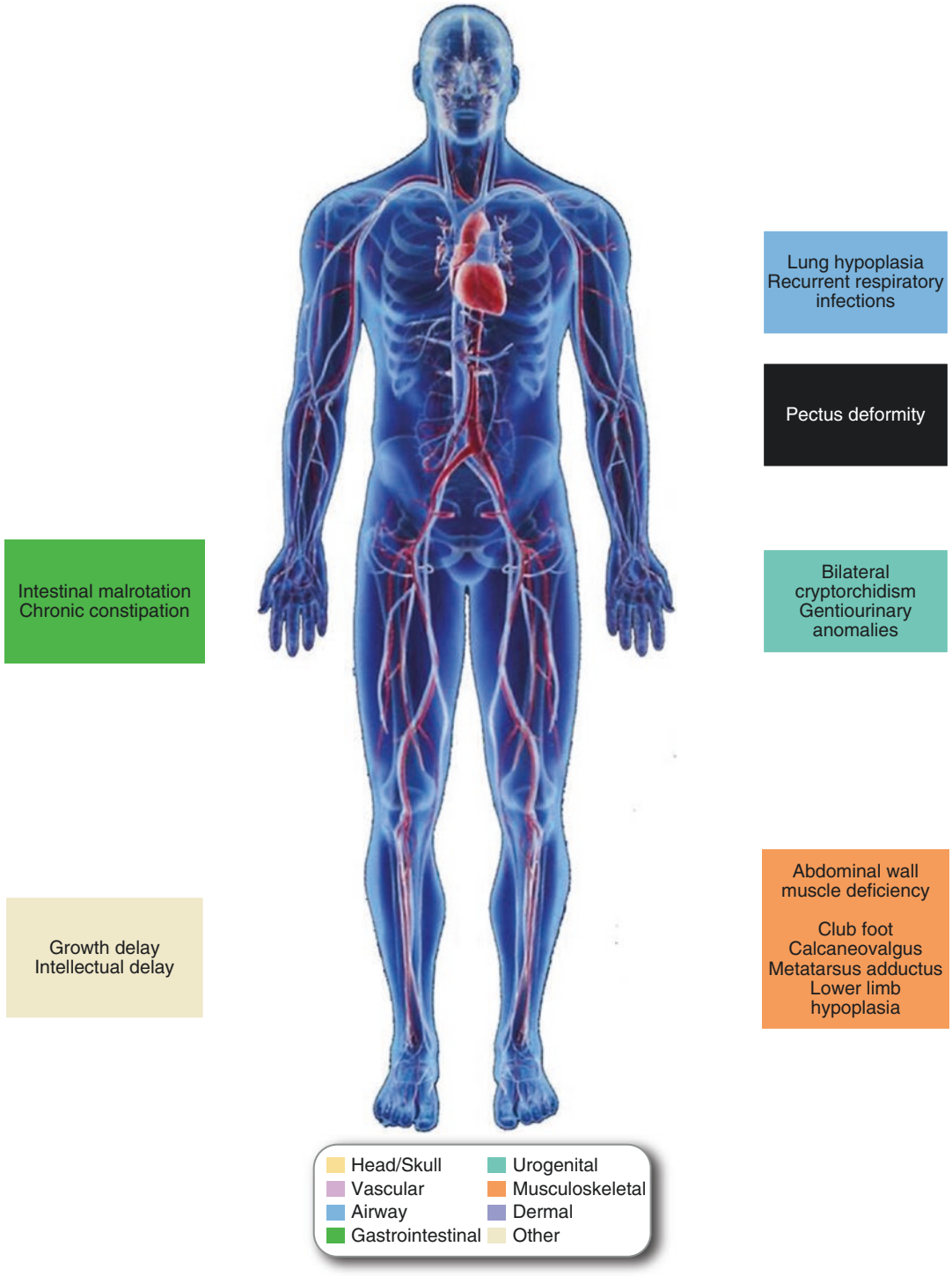

Fig. 8.13 Schematic characteristics of Prune belly syndrome

ities was defined in 1986 by the Toronto study and documented an incidence of non-urologic problems of 75 % [77]. With advanced medical and surgical management of these patients, morbidity and mortality because of genitourinary malformations decreased and other features of this syndrome become apparent [78]. Non-urologic problems that are described in prune-belly patients are pulmonary (lung hypoplasia secondary to oligohydramnion [75] and recurrent respiratory tract infections [77]), gastrointestinal (intestinal malrotation, imperforate anus, small bowel stenosis and chronic constipation [77, 79]; but may also be concomitant with gastroschisis [80, 81]), developmental (growth and intellectual delay [77]) and orthopaedic (clubfeet, calcaneovalgus, metatarsus adductus, lower limb hypoplasias ranging from complete amelias to hypoplastic or shrivelled legs or feet with absent rays, hip dysplasia, thoracic cage deformities including scoliosis and PE and pectus carinatum) [77]. Loder et al. [78] have assessed the musculoskeletal aspects of this syndrome in a retrospective review and defined a prevalence of musculoskeletal diseases associated with this syndrome with 45 %. Reported prevalence of musculoskeletal diseases associated with the prune-belly syndrome are 26 % for clubfeet, 5 % for hip dysplasia, 4 % for spinal deformities and 8 % for chest wall deformities (5 % for **PE** and 3 % for **pectus carinatum**) [78]. With increasing age and since most prune-belly syndrome patients survive into adulthood nowadays, musculoskeletal abnormalities such as pectus deformities causing distinctive morbidity and therefore may require corrective surgery.

Ruvalcaba Syndrome

Ruvalcaba et al. [82] reported in 1971 on a rare familial multiple congenital anomalies syndrome that combined skeletal dysplasia, facial anomalies and mental retardation (Fig. 8.14). In 50 % of the reported patients skeletal anomalies involving short stature, small hands and feet with short fingers and toes, broad hips, joint limitation and vertebral anomalies occur, while facial anomalies are characterized by microcephalus with an abnormal usually beaked nose with hypoplastic nasal alae, microstomia with a narrow maxilla and a thin upper lip vermilion [82–87]. A **pectus deformity** has been observed in 6 of 16 documented patients [82, 83, 85, 87]. In 1989 another female patient who presented additional anomalies was reported on by Bialer et al. [83]. This 22-year old female patient additionally presented with features such as upslanting palpebral fissures, torus palatinus, **pectus excavatum** different to **pectus carinatum** observed in the other reported patients, equinovarus deformity, hypotonia, hiatal hernia with gastrointestinal reflux and recurrent pulmonary infections and further presented genitourinary anomalies that involved unilateral renal hypoplasia, an accessory ovary and an atretic fallopian tube. There was no familial history and the karyogram showed a 46,XX karyotype [83]. The underlying cause has not been identified yet. As familial history with interfamilial differences has been documented [82, 87, 88], an autosomal dominant inheritance with incomplete penetrance has been suspected [87, 88]. However, due to the variability of the manifestations, an unbalanced cytogenetically inapparent chromosome rearrangement was also suggested [83]. As a few patients represented renal anomalies, such as renal hypoplasia [83], abnormal positioning of the kidney [82, 85], megaureter and hydronephrosis [87], renal ultrasound and contrast studies were implicated in evaluating these patients [83].

New Syndrome with Apparent AR-Inheritance

In 2012, the case of 2 brothers from consanguineous parents with multiple congenital abnormalities and mental retardation was published [89]. The 2 brothers displayed the features of developmental and mental retardation, hypotonia, inability to sit and to hold the head straight, short stature, pylorus stenosis that required pyloromyotomy, **pectus excavatum**, brachycephaly due to craniosynostosis, frontal bossing, depressed nasal bridge, high arched-wide palate, downslant

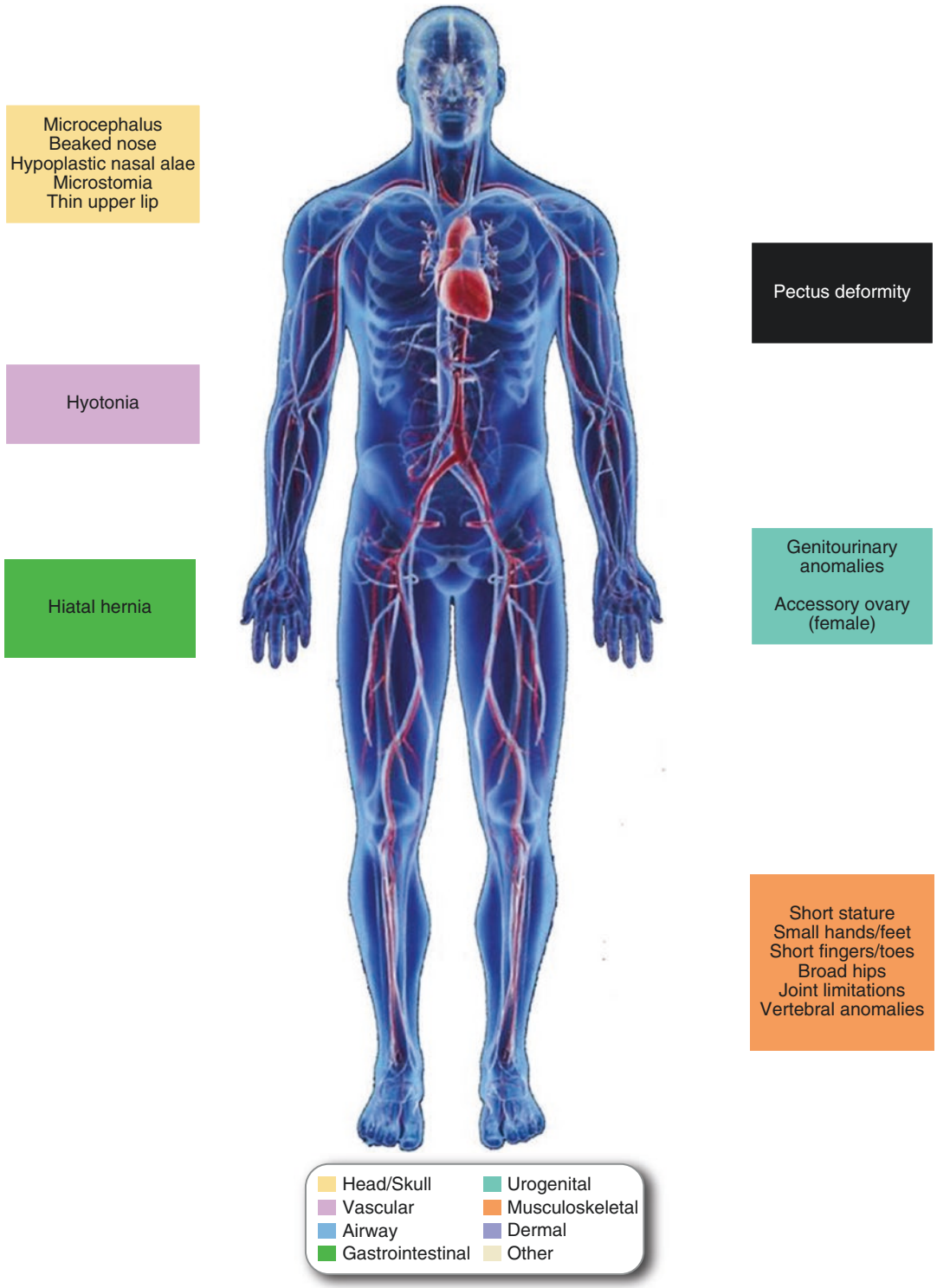

Fig. 8.14 Schematic characteristics of Ruvalcaba syndrome

palpebral fissures, low-set large ears, thin upper lip, bilateral cryptorchidism and thalassemia intermedia. The karyotyping and mutation analyses were normal which were also found to be normal in the healthy sister and parents and did not match clinically, neurologically, or by molecular study with any other known syndromes and were therefore characterized as a new apparent autosomal recessive disorder [89].

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9 Chest Wall Deformities and Musculoskeletal Defects in Congenital Diaphragmatic Hernia

Elke Zani-Ruttenstock and Amulya K. Saxena

Introduction

Congenital diaphragmatic hernia (CDH) has an incidence between 1 in 2600 and 1 in 3700 live births [1, 2]. The commonest type of CDH occurs in the postero-lateral aspect of the diaphragm and it is called Bochdalek hernia (Fig. 9.1a) Less common forms comprise the Morgagni hernia (the defect is in the anterior aspect of the diaphragm on either side of the xiphisternum) (Fig. 9.1b) and a congenital absence of the hemidiaphragm (Fig. 9.1c) Malformation of the diaphragm allows the abdominal viscera to migrate into the chest during fetal life, inhibiting lung development.

More recent studies suggest that the lungs have a primary intrinsic defect before being further impaired by secondary viscera herniation ("dual-hit hypothesis") [3]. The resultant abnormality leads to disordered lung growth, leading to pulmonary hypoplasia and persistent pulmonary hypertension. Both lungs are affected, the ipsilateral lung more than the contralateral one [4]. Approximately 80 % are left sided, less than 5 % are bilateral, and about 15 % are right sided [1, 2]. The size of the defect varies from small to very large, involving most of the hemidiaphragm.

In the past, CDH was considered a surgical emergency, where prompt reduction of the abdominal viscera was needed to expand the lung. The increased knowledge of the pathophysiology of CDH has led to a different approach, where prolonged preoperative stabilization has proven useful. The operative repair of small size defects is usually by primary closure in the presence of sufficient diaphragmatic tissue. Large defects, on the other hand, might require the application of a prosthetic patch. Data from the Congenital Diaphragmatic Hernia Study Group report prosthetic patch repair rates of up to 51 % [5]. Although patches have offered a good short-term solution in repairing large diaphragmatic defects, they could be complicated with patch disruption and hernia recurrences in up to half of the patients by the age of 3 years with many patients having multiple recurrences [6]. Moreover, scarce information exist on the diaphragmatic integration of patches in CDH survivors during childhood.

Since the widespread implementation of delayed operative repair with extra-corporal membrane oxygenation and gentle ventilation

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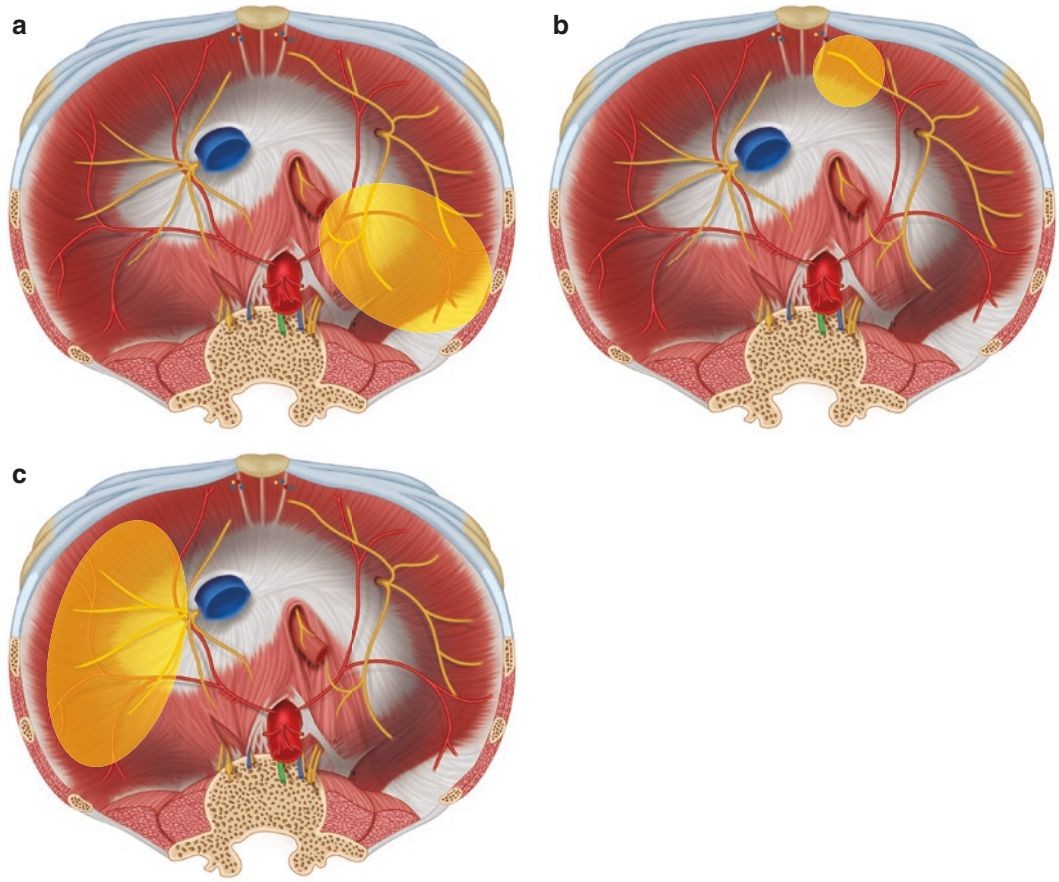

Fig. 9.1 (**a**) Postero-lateral Bochdalek hernia (Modified from Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**b**) Antero-medial Morgagni hernia (Modified from Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010). (**c**) Congenital absence of the right hemidiaphragm (Modified from Tillman B. *Atlas der Anatomie des Menschen*. Springer Berlin Heidelberg 2010)

strategies starting in the mid-1990s, overall survival of CDH has dramatically increased [7, 8]. With the improvements in survival, increased postoperative and long-term morbidity has been reported, together with a greater need for monitoring and supportive interventions in infancy and early childhood [9, 10]. It is well recognized that CDH survivors suffer from morbidities related to their underlying pathology (pulmonary hypoplasia), as well as from other significant morbidities that result from the intensive and aggressive maneuvers used to resuscitate the critically ill neonate. Among the long-term sequelae of CDH musculoskeletal defects have been increasingly reported.

Incidence

Chest wall deformities are present in approximately 16–55 % of the CDH survivors [10–12], whereas the incidence of scoliosis has been reported in approximately 5–30 % of CDH survivors, although large studies are lacking [10, 13–19].

Types of Musculoskeletal Defects

- Chest wall deformities (16–48 %)
  (a) Pectus excavatum (PE) (10–47 %) (Fig. 9.2)
  (b) Pectus carinatum (PC) (up to 12 %) (Fig. 9.3)
- Scoliosis (5–30 %)

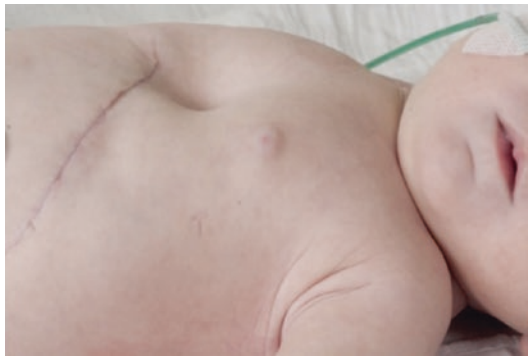

Fig. 9.2 Three-week old infant post left-sided Bochdalek CDH patch repair. A significant pectus excavatum deformity was already present at time of birth, which points to the fact that these deformities exist despite the type of repair offered for CDH repair

Pathophysiology

Congenital diaphragmatic hernia–associated chest deformities are particularly troublesome, as they tend to be asymmetric and progressive. Little is known about etiology and natural course of these conditions. The etiology can be related to the embryology of CDH, early postnatal ventilation strategy, or even the surgical treatment. The close relationship between development of the lung, diaphragm, and thoracic cavity is the reason for which deformities of the chest wall are more common in patients with CDH [13].

Several factors contribute to thoracic deformities. Firstly, the thoracic cavity on the affected side may be reduced in size due to a smaller lung volume. Secondly, the increased work of breathing in these children may contribute to the development of a pectus abnormality because more negative intrapleural pressure is required to inflate the lungs [12–14]. Thirdly, more negative intrathoracic pressure promotes retraction of the chest wall in its most compliant section, i.e., the cartilaginous anterior wall [12, 20].

Since chest wall deformities are more common among patients with anatomically large diaphragmatic defects, it has been suggested that repair of a large defect causes a degree of tension that would interfere with the normal development of the thoracic cage and would promote asymmetry and/or flat chest [11, 12]. It has been

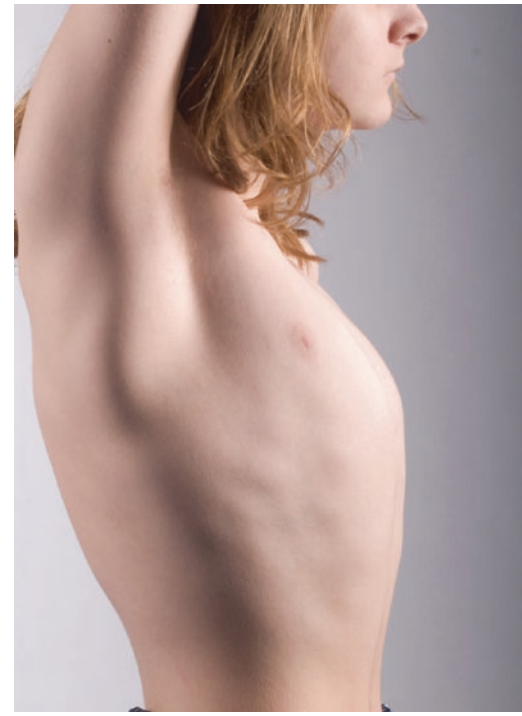

Fig. 9.3 Pectus carinatum with protrusion of the sternum and ribs

reported that CDH patients with a clinically asymmetric chest and scoliosis are prone for pulmonary function impairment [19, 20]. In other CDH patients with pectus excavatum, a reduced lung function compared to normal CDH patients has been demonstrated [21, 22]. However, it is controversial whether surgical correction results in improvement of pulmonary function [22–24].

Furthermore, musculoskeletal defects are reported to occur more frequently following CDH patch repair [11, 15, 16] resulting in a search for new patch materials that will not increase hernia recurrence while "growing" with the child to prevent spine and chest wall changes [9]. The last decades have seen a wide range of materials reported for use in CDH patch repairs, including autologous muscles, although the perfect patch material has yet to be identified.

Finally, the technique of patch placement also plays an important role in CDH recurrence and/or musculoskeletal changes. Patches placed under tension are more likely to pull away from the chest wall during growth to cause a recurrence or conversely

pull the chest wall inward to cause force line traction to the costal structure and consequently chest contour changes [16, 25]. Early results of tension-free, cone-shaped patch repairs are promising [26]. Their use will have to be followed prospectively to determine whether this technique truly makes a difference in preventing CDH recurrence and musculoskeletal changes. However, a significant proportion of patients with an initial primary repair still developed this problem (21 %) [11].

Chest asymmetry is more common among those who had a thoracotomy. Division of the serratus anterior during thoracotomy, rib resection and repeated thoracotomies, or postoperative empyema have been shown to increase the likelihood of postoperative thoracic deformities [12].

The impact of innovative surgical approaches to CDH repair has also been observed in the follow-up of CDH patients. Proponents of minimal access surgery have supported its benefits in minimizing pain, maximizing cosmesis, and limiting future chest wall deformities. Multiple reports have detailed the early and frequent CDH recurrence rates following thoracoscopic repair; [27–30] however, there are mid-term and long-term studies in progress. Larger series and surgical conquest of the steep learning curve will have to be achieved with minimal access repairs.

Scoliosis in CDH Patients

In comparison to idiopathic scoliosis, scoliosis in CDH patients is considered an early onset with significant tendency to progression. These patients need to be referred to pediatric spinal services early in order to enable prompt intervention with braces and/or growing rods. Patients with scoliosis most often present with unleveled shoulders, waistline asymmetry (one hip "sticking out" more than the other), or a rib prominence. Back pain is sometimes the presenting complaint.

Clinical Assessment of Scoliosis

- Detailed history
- Examination
- Radiological investigation
- Early referral to pediatric orthopedics for vertebral column evaluation

In the vast majority of affected patients, a diagnosis of chest deformity occurs during the first 2 years post-repair [11]. For further details on diagnosis and management of scoliosis please refer to the next chapter.

Clinical Evaluation of Pectus Excavatum (PE)

- Complete history and physical examination, which includes questions regarding onset, duration, and progression of chest wall deformity in context of the CDH.
- The morphology of the PE should be described in detail with relation to the CDH surgery and the location of the scar. The degree of depression of the body of the sternum and the lower costal cartilages should be documented. The deformity may be described as symmetric or asymmetric with a deeper depression on one side of the sternum. Asymmetry is often associated with sternal rotation, and this should be identified in the initial assessment. The deepest point of the PE should be noted, which is generally close to the sterno-xiphoid junction in CDH patients. The deformity may be defined as localized (cup-shaped), diffuse (saucer-shaped), or long and unilateral (grand-canyon type).
- Photographic documentation is very helpful in following these patients before and after repair (Fig. 9.4). If possible videosteroraster examinations should be performed.
- Chest films with both anterior-posterior and lateral view to estimate the level of the diaphragm, which may be altered in CDH patients with or without patch repairs, especially if a minimal access repair is planned to minimize the risk of trocar associated injury (Fig. 9.5).
- Symptom assessment should be thorough, with focused questioning regarding the pulmonary and cardiac systems. Most young children with PE are asymptomatic, common

complaints are exercise intolerance, lack of endurance, and shortness of breath on exertion. Body image is also an important part of the history because these patients may be at risk of negative body image perception, social withdrawal, and even suicidal tendencies. PE has been determined to have lower Health-Related Quality of Life (HRQL) scores than age-matched peers. The symptom correlation of pectus patients with CDH has not be evaluated.

- In case of clinical or radiological detection of scoliosis – early referral to pediatric orthopedics for vertebral column evaluation and early commencement of management [31].

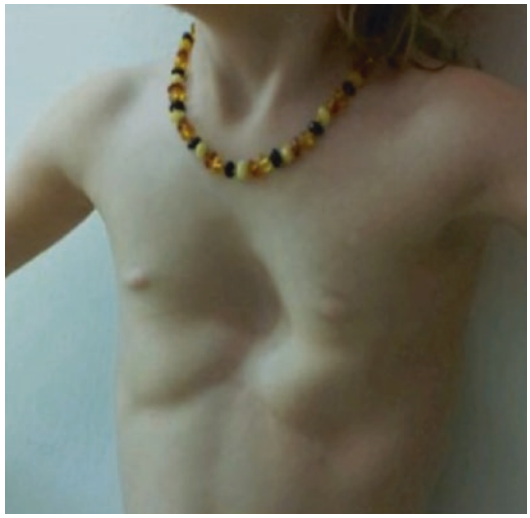

Fig. 9.4 Infant after left sided Bochdalek hernia recurrence repair at 6 months of age with significant Pectus excavatum

Objective Evaluation of PE

Pulmonary function testing, CT or MRI of the chest, and a cardiac evaluation to include electrocardiogram and echocardiogram are a routine aspects of the evaluation for a patient with PE. The pulmonary function tests need to be evaluated in the context of the hypoplastic lung which is present in patients with CDH. More recently, MRI of the chest with cardiac function assessment has been used to avoid radiation exposure and replace the echocardiogram. Radiographic evaluation with a thoracic CT scan

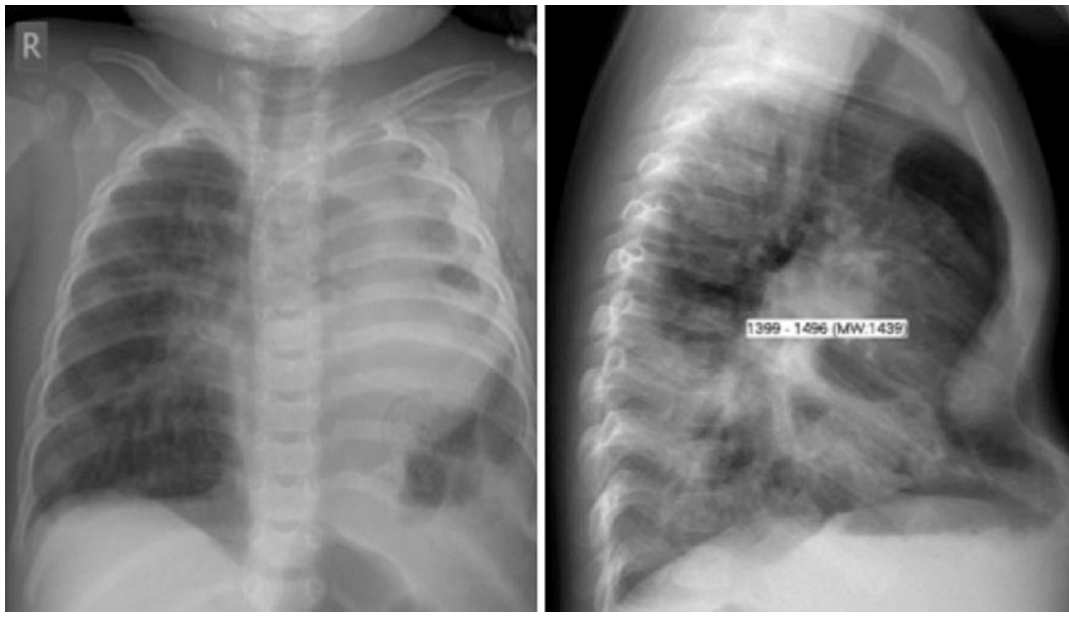

Fig. 9.5 Anterior/posterior and lateral chest x-ray of same infant at 6 month of age with left sided Bochdalek hernia recurrence after patch repair and significant progression of pectus excavatum

or MRI allows the physician to grade the severity of the PE by calculating the Haller index that uses the internal measurements of the transverse and anterior-posterior dimension at the deepest point of the deformity.

Surgery for PE

Surgical correction options for pectus excavtum patients with CDH need to be carefully evaluated. In case the pectus deformity is identified at an early age, conservative treatment options may be initiated when the child is around the first decade of life. If there is failure of conservative treatment, surgical options should be offered. Many points should be considered when planning surgery, especially if large size patches were utilized for CDH repair. If the minimal access approach is favored, preoperative estimation of the diaphragm level is important to avoid injury to the diaphragm or liver on the right and spleen on the left while inserting the optic port in the thorax. Care should also be taken not to place the pectus bar close to the sterno-xiphoid area especially if a large patch was used. If the option for open repair is taken, care should be taken during dissection of the retrosternal space while freeing the caudal part of the sternum from the scar remnants. Procedural details and steps as outlined in the chapters with details on surgical techniques should be carefully evaluated with regards to the altered morphology in patients with CDH repairs.

Clinical Evaluation of Pectus Carinatum (PC)

- A complete history and physical examination is warranted. There is always a prominent protrusion of the sternum, with several patterns that may be present (Fig. 9.3). Although rare in association with CDH, pectus carinatum is often a secondary deformity which is possibly due to attached and rigid "repaired" diaphragm that results in disproportionate chest wall groeth.
- Most children will have not significant symptoms with PC. Tenderness when lying prone is a common complaint and musculoskeletal pain of the chest and the epigastrium may also be present. Psychological distress and negative body image perception can be identified, and in severe cases patients may complain of physical deficits such as shortness of breath.
- Posteroanterior and lateral chest x-ray, followed by either CT or MRI [31].

Management

For patients who are identified with pectus carinatum in the first decade of life, Orthotic bracing is recommended as the chest wall is malleable chest wall. Please refer to the chapter on Orthotic bracing for more details. However, if the chest wall is rigid, surgical options as outlined in the chapters for surgical techniques should be offered.

Conclusion

Chest wall and spinal deformities are common among patients with repaired diaphragmatic defects. In the majority of patients, these deformities are mild and do not need treatment. Conservative treatment options can be offered when the patients are in their first decade of life and the chest wall is not rigid. Surveillance of patients with CDH repairs should be followed up with until adolescence with regards to chest wall development, so that the suitable window for conservative treatment, if necessary, is not missed.

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Idiopathic Scoliosis

10

Christine Wibmer and Vinay Saraph

Definition and Etiology

The term scoliosis was supposedly used by Galen, for the first time. It is derived from the Greek word σκολιός [skol-ee-os'] meaning crooked. One of the most common deformities of the spine, scoliosis has been recognized since ancient times [1]. In 1741, Nicolas Andry first used the crooked spine as a symbol for orthopedics [2].

Scoliosis is defined as a lateral curve of the spine measuring more than 10° deviation from the normal vertical line on plain radiograph [3]. Although the primary deformity is in the frontal plane, the deformity of the spine is three-dimensional: lordosis in the sagittal plane, scoliosis in the frontal plane and vertebral rotation in the axial plane.

Milder forms of scoliosis cause minimal trunk asymmetry and the implications are mostly cosmetic. With worsening of the deformity, structural changes are seen including asymmetric growth of the vertebral bodies, pedicles, lamina, facet joints, transverse process and spinous process. Severe deformities are markedly disfiguring, with disturbed relationships between intrathoracic and abdominal organs. Although severe spinal deformities generally cause few problems in childhood long-term follow-up studies have documented significant morbidity in early and late adulthood: chronic back pain and early degenerative changes in the spine, decreased vital capacity, higher likelihood of cor pulmonale, as well as psychological disorders [4].

Scoliosis can have several underlying etiologies: idiopathic, neuromuscular, syndrome related, and congenital. The etiology of idiopathic scoliosis remains unknown, thus the diagnosis is one of exclusion of other diseases. Idiopathic scoliosis represents the largest subset, accounting for nearly 80 % of patients with structural scoliosis.

Although the etiology of idiopathic scoliosis remains unknown, substantial research has been performed. The etiological theories range from disorders of bone, muscles, intervertebral discs, growth abnormalities, central nervous system causes and genetic factors [5]. Whether these abnormalities are the cause of the scoliosis, or occur secondary to the development of curves is unknown.

Several studies strengthen the proposed genetic etiology of this disease [6–11]. Both location of the curve, curve severity and curve pattern appear to be inherited separately in adolescent idiopathic scoliosis (AIS). It seems a polygenic, multifactorial condition with a variable expression and a low penetrance of its cumulative alleles [12].

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Epidemiology

Approximately 0.5 % of all children develop idiopathic scoliosis. Girls are affected in a higher percentage than boys (male to female ratio 1:4). The number of patients who need treatment, conservative or surgery, is lower at around 0.25 % of all cases [13]. The incidence numbers are derived from school screening programs, introduced in the 1970ies in several countries, or from data collection of scoliosis centers. After a short increase in incidence at the beginning of school screening programs the incidence levels decreased to a lower level again. Furthermore, as diagnostic possibilities increase, the incidence of true idiopathic scoliosis, particularly the infantile form, has decreased [14, 15].

Clinical Presentation

Most of the cases are diagnosed incidentally as this condition is not painful. At stages with more severe curves the scoliosis is apparent as a trunk deviation and rib cage deformity even in upright position, otherwise it becomes visible only when the patient is asked to perform the forward-bend test. Deformity in the thoracic spine is seen as a prominent scapula, or a 'rib-hump'. This 'rib hump' presents on the convex side of the curve as the axial rotation is directed to the convex side, too (e.g. clockwise rotation in a right-convex curve) followed by rotation of the whole thoracic cage. In the thoracolumbar and lumbar spine an asymmetrical lumbar prominence is seen on the convex side. An uneven shape of the paravertebral musculature may also be visible (Fig. 10.1a, b).

Pulmonary symptoms, e.g. diminished vital capacity, occur in severe deformities and are observed in curves exceeding 60°.

Pain, leg length discrepancy, altered muscle tonus are usually signs of other underlying causes for scoliosis. Back pain in adults on the other hand may be the result of an undiagnosed idiopathic scoliosis during childhood or adolescence.

Classification

Several classification systems for idiopathic scoliosis have been proposed, to help indicate a treatment course or allow a prognosis of curve progression.

Age at Onset

The Scoliosis Research Society recommends classifying idiopathic scoliosis correspondent to age at first diagnosis [16]. Idiopathic scoliosis is accordingly classified as follows:

Infantile scoliosis: age 0–3 years
Juvenile scoliosis: age 4–10 years
Adolescent idiopathic scoliosis: age 10–18 years
Adult idiopathic scoliosis: Onset of deformity after reaching skeletal maturity.

This traditional chronological classification of scoliosis is clinically relevant, as the natural progress of the disease varies within the subtypes. During childhood, the lungs grow in size, the alveoli and arteries multiply and the pattern of vascularity changes. Severe scoliotic deformity before the age of 5 years will inadvertently restrict cardiopulmonary growth. Several reports have advocated a distinction between early-onset and late-onset scoliosis because of the effects of the deformity on cardiopulmonary development [17–19].

Localisation of Curve

The scoliotic curves can also be classified according to the technical description of the curve locations [20].

- Cervical – apex between C2 and C6
- Cervicothoracic – apex between C7 and T1
- Thoracic – apex between T2 and T11
- Thoracolumbar – apex between T12 and L1
- Lumbar – apex between L2 and L4
- Lumbosacral – apex below L5

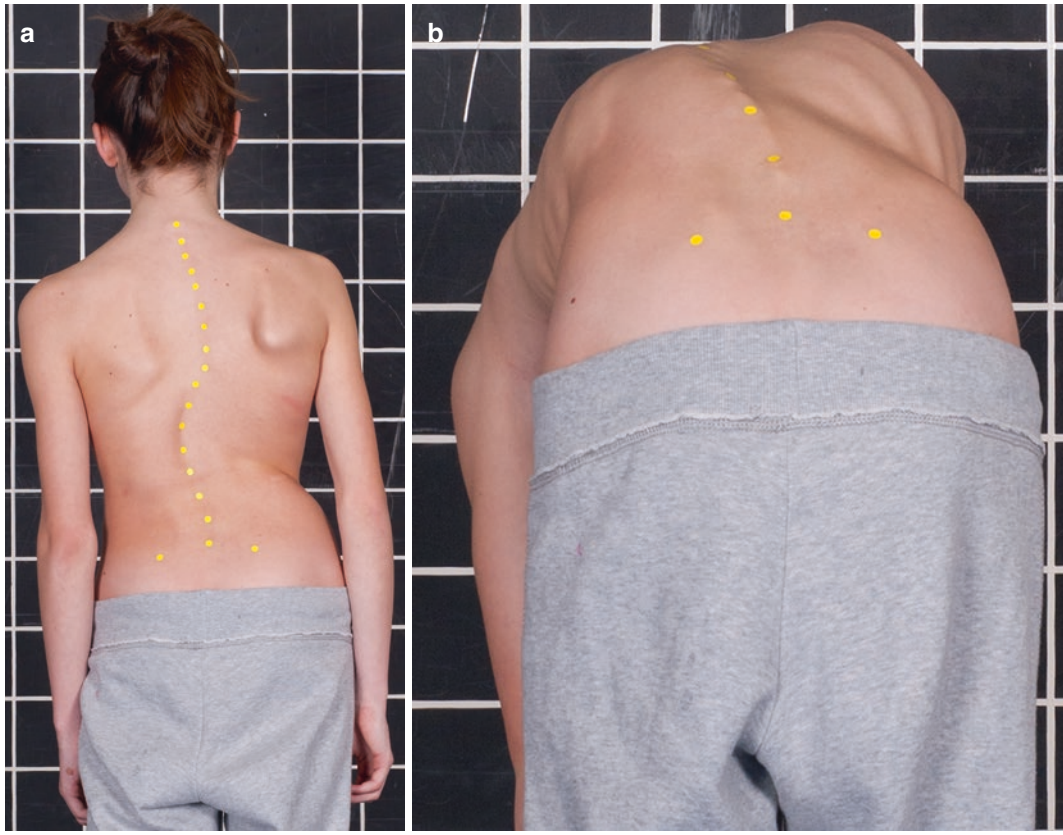


Fig. 10.1 Patient with Idiopathic thoracolumbar scoliosis. Markers have been placed over the spinous processes to delineate the spinal column. (**a**) In Standing, note the deviation of the trunk, prominence to the scapula on the right side and asymmetry of the flanks. (**b**) On forward-bending (Adams Test), the fixed rotational component presents as a rib hump on the right, and lumbar bulge on the left

Classifications Pertaining Surgical Decision Making

Patterns of the scoliotic curves must be well understood for surgical correction. Decisions need to be made regarding the extent of fusion, surgical approach, and the type of the surgical instrumentation.

The most common classification was presented by King in 1983, where thoracic scoliosis curve patterns were described based on radiological and clinical criteria [21]. Five curve types have been described in this classification based on anterior-posterior plain radiograph and bending radiographs. According to the curve type, a determination of when to instrument the thoracic curve alone, and when to instrument both the thoracic and lumbar curves is possible. The King-Moe Classification has certain limitations. The classification addresses scoliosis with thoracic curves only. Primary lumbar and thoracolumbar curves, as well as triple curves were not included. The sagittal plane balance and its role in deciding the fusion levels are not included.

Lenke et al. proposed a comprehensive classification system, which considers both frontal and sagittal plane deformity and is designed to guide surgical decision making for all curve patterns [22]. The utility of the classification system in surgical decision making has been well documented [23–31]. A detailed description of the surgical classifications of idiopathic scoliosis is beyond the scope of this chapter.

Clinical and Radiological Features

In the clinical examination the forward-bend test (Adams-test) [32] is performed to assess lower and upper trunk rotation. The patient bends forward, with knees straight and palms together. The asymmetry of upper and lower trunk can be quantified using a scoliometer which measures the rotation at the apex of the curve, or by documenting the height of the rib-hump or the lumbar bulge in centimeters. Trunk deviation can be measured clinically using a plumb line starting at C7, the deviation quantified in centimeters from the gluteal cleft.

The lateral deviation of the spine is described using the Cobb angle on plain radiograph of the total spine with the patient standing [33]. The end vertebra is the most cephaled or caudad vertebra of a curve, and is also the most tilted from the horizontal. The apical and caudal end vertebrae of a curve define the upper and lower end of the scoliotic curve, and are the most tilted from the horizontal. These vertebral endplates are used to measure the Cobb angle (Fig. 10.2). The apical vertebra marks the most rotated vertebra and the apex of the curve. It also is the most deviated vertebra from the central sacral vertebral line. A Cobb angle between 10° and 24° marks a mild curve, between 25° and 39° moderate and over 40° a severe deviation.

The vertebral rotation can be measured using the Nash-Moe method [34]. This method is applicable on plain anterior-posterior radiograph. The landmarks for measurement are the apical vertebra and here the shadow of the pedicle on the convex side of the curve. The percentage of displacement of the convex pedicle with respect to the vertebral body width enables to approximate the angle of vertebral rotation. Other methods like Perdriolle or Stokes method have been developed to measure vertebral rotation [35].

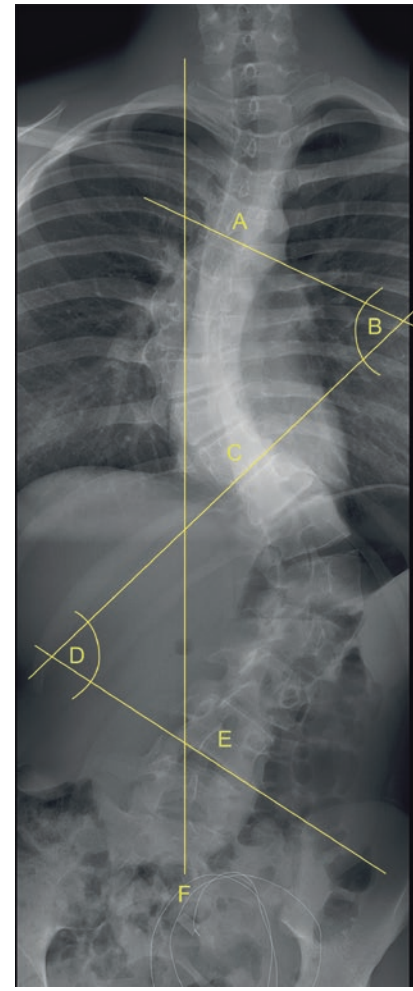


Fig. 10.2 Basic radiological measurements in idiopathic scoliosis. In the figure, *A* and *C* mark the upper and lower end vertebrae for the thoracic curve, the angle between them (*B*) is the Cobb angle for the thoracic curve. *C* and *E* represent the end vertebra for the lumbar curve, angle *D* is the lumbar Cobb angle. *F* represents the central vertical sacral line (CVSL) which demonstrates the shift of the spinal axis in the coronal plane

Course of Disease

A progression of the deformity is the dreaded course of disease, with rigid spinal and rib cage deformity and pulmonary complications as the final state. But not every case leads to progression. The majority of all diagnosed cases (approximately 80 %) will never progress [36, 37]. This leads to the problem of adequate prognostication of curve progression to prevent over- or under-treatment.

Several clinical parameters are associated with curve progression:

Female gender: Girls are generally more at risk of curve progression.

Remaining growth: The probability of progression of the scoliotic curve increases with higher remaining growth potential at the time of first diagnosis. The following growth parameters are associated with progression:

- First diagnosis of scoliosis before the onset of menarche,
- Risser grade 0 or 1. This is a method of measuring skeletal maturity according to ossification of the iliac apophysis [38].
- Low skeletal maturity (skeletal age under 11 years in girls, under 13 years in boys) according to Greulich and Pyle, where skeletal maturity is determined according to degree of ossification seen on the x-ray of the hand [39–42].
- Tanner index of maturity 1 or 2. Here the evaluation of maturity is performed according to appearance of secondary sexual characteristics on clinical examination [43, 44].

Severe Curve at Diagnosis

- Cobb >30°,
- Thoracal location of deformity
- Double curve

The so called Hueter-Volkmann effect can partly explain the possible progression of deformity during growth period. It describes the suppression of growth on the concave side of the curve and leads to asymmetric growth and remodeling of the vertebral bodies, pedicles, lamina, and facet joints [45].

All of these factors are associated with curve progression, but no reliable hazard ratio could be developed out of these observations alone for juvenile and adolescent idiopathic scoliosis. The rib-vertebral angle difference (RVAD) has been documented to be a reliable radiological criterion for predicting progression of the curve in infantile idiopathic scoliosis. A convex RVAD above 20° is associated with a progression of the scoliotic curve; a convex rib-vertebral angle below 68° is an equally accurate indicator for curve progression [46, 47].

Recent genetic research found 53 genetic markers with prognostic impact regarding curve progression. A DNA-based prognostic test has been developed to predict spinal curve progression in white patients with AIS. [48–51].

After reaching skeletal maturity the risk of progression is much lower and depending on the grade of deformity. Thoracic curves measuring less than 50° and lumbar/thoracolumbar curves below 30° are unlikely to progress. Curves with Cobb angles above 50° may progress slowly with 0.5–1° per year.

Treatment

The severity of the curve, the location of the curve, and remaining trunk growth are the main factors determining whether conservative treatment or surgery are appropriate. Selection of treatment modality in idiopathic scoliosis in skeletally immature patients is suggested by the European Spine Society as follows:

Scoliotic Curve less than 20° Cobb: Observation, Clinical examination every 3–6 months depending on the age and growth rate of the patient.

Scoliotic Curve between 20° and 40° Cobb: Brace Treatment and Physiotherapy if more than 1 year of pubertal growth is remaining.

Scoliotic Curve above 40° with documented progression: Surgery.

Physiotherapy and Bracing

Spinal Bracing is usually combined with physiotherapeutic scoliosis specific exercises (PSSE) and is generally recommended in curves between 20° and 40° in skeletally immature children with the aim to prevent progression of the deformity. Conservative treatment is recommended in these cases until skeletal maturity.

Exercise based therapy used alone or in conjunction with Bracing is controversially mentioned in the literature. Several reports claim the inefficiency of physiotherapy and exercise based

programs in preventing progression of curves in idiopathic scoliosis [52–55].

Exercise based therapies however are a logical approach to improve and maintain flexibility and function of the spine, particularly in combination with Bracing. Several studies document decreased progression in patient populations who were treated with exercises [56]. Reported modes of physical therapy are the Lyon method, Side-Shift method, Dobosiewicz method, SEAS method, asymmetric trunk mobilization and Schroth therapy; the latter popular in central European countries [56, 57].

It has to be emphasised that the success of physical therapy depends on meticulous application of these methods by specifically trained therapists as well as compliance of the patient to perform the exercises regularly.

Most surgeons recommend Brace treatment for cases in which the progression of the curve exceeds 5° when the initial Cobb angle has lied between 20° and 40° (Figs. 10.3 and 10.4). Early bracing may be considered when a strong positive family history for progressive scoliosis is present. The Milwaukee brace, developed in the 1940s, was one of the first braces for scoliosis management and has become the standard for comparison to other designs of braces. The design which originally provided longitudinal traction between the skull and the trunk had potential to maintain upper thoracic curves. The patient acceptance for this brace, however, was poor [58]. Underarm braces (Wilmington, Boston, Chenau, Providance, SpineCor) are more popular due to increased compliance by the patients.

The aim of a spinal brace is to provide corrective molding of the trunk and spine during growth. The recommended wearing schedule for braces is 18–23 h a day. Several braces (Charleston, Providence) are effective when worn part-time or as nighttime braces. However, in curves greater than 35°, generally full-time bracing is recommended. Bracing should be continued until skeletal maturity, indicated by unchanged height measured consecutively 6 months apart, Risser sign 4 (females) or 5 (males), postmenarchal 18–24 months, or skeletal maturity on bone age determination [59]. The indications and efficacy of brace treatment has been extensively reviewed by Schiller JR et al. [60].

Brace treatment for progressive curves provide the standard of care in most of the developed world. The time for initiation of bracing is difficult – very mild curves are easier to control with bracing; however, using this policy, one tends to unnecessarily brace many children. Waiting till the curve progresses from 20° to 30° is precarious, as the margin between 30° (conservative therapy) and 40° (operative management in case of further progression) is very small. It needs to be mentioned that managing a teenager with brace can be very demanding. The resistance of adolescents who might have to wear a brace for 3–5 years is understandable [61, 62]. Regular clinical follow-ups, motivation of the patient, communication with the physiotherapist and the family are essential for a successful brace treatment.

Surgical Treatment

Surgical treatment in idiopathic scoliosis has to be individualized for every patient. A detailed description of the indications, available surgical implants and selection of type of surgery for the patient is beyond the scope of this chapter. The guidelines for surgical treatment shall be discussed. The interested reader is advised to refer to literature on pediatric spinal surgery.

General recommendations propose surgery in skeletally immature patients if the scoliotic curves are above 40° with documented progression. However, if the location of the scoliotic deformation is thoracolumbar, even curves with lower angles might need surgery, when presenting a higher deviation of the trunk. Long-term results of the first dorsal instrumentation system, the Harrington rods, have documented lasting effects of stabilization of the spine [63]. Spinal instrumentation systems for correction of scoliosis are being improved and the newer systems permit a better three-dimensional correction, more stability, and the possibility to instrument the

spine without fusion in younger patients with the possibility of growth modulation using periodic lengthenings.

Certain aspects need to be considered in the surgical patient:

1. Evaluation of growth potential of the segments planned to be fused is essential. This can be calculated using percentile growth charts, or using a formula for potential shortening caused by a spinal fusion. The formula is: centimeters of shortening = 0.07 × number of segments to be fused × number of years of growth remaining [64]
2. If substantial growth potential in the segments to be fused is remaining, one should consider continuing the conservative treatment with bracing and PSSE unless curve progression

Fig. 10.3 Illustrative example of a modified Stagnara Brace in juvenile idiopathic scoliosis. This type of brace is applicable in young patients with flexible spines, where correction can be achieved with minimal translational supports. (**a**, **b**) Clinical pictures. (**c**, **d**) Radiograph without and with brace, showing the correction of the scoliosis

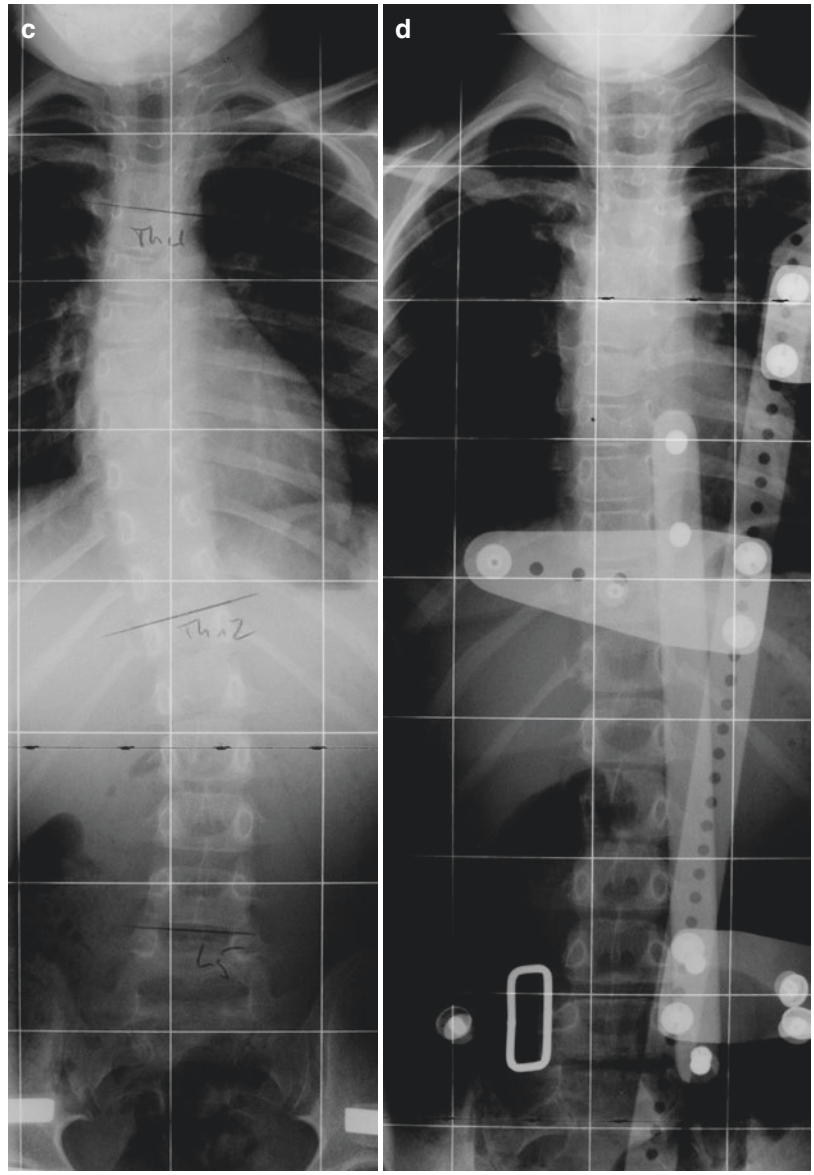


Fig. 10.3 (continued)

cannot be prevented. In that way it is possible to modulate the remaining growth and not perform spinal fusion at an early age. Frequent follow-up to detect curve progression is then necessary. When surgery is mandatory in infantile and juvenile scoliosis with significant remaining growth potential, spinal implants which permit sequential lengthening can be used to allow growth of the spine and correction of the spinal deformity at the same time. The management of patients with this type of instrumentation, however, is very tedious and good cooperation of the family is a prerequisite for successful treatment [65]. At the end of spinal growth these patients need definitive fusion.

3. Preoperative evaluation should include complete evaluation of blood, urine, cardiorespiratory system, evaluation of bladder function as well as special radiographs to differentiate structural from nonstructural curves (Side-bending radiographs).

4. Basically, structural curves are fused. Non-structural curves – curves showing good correction on side bending radiographs need not be fused (Fig. 10.5a–d). Selection of the fusion levels for any given patient is individual and depends on several factors and remains a topic of controversy. The selection of fusion levels in idiopathic scoliosis aims to preserve as many motion segments as possible.

In the majority of the patients with idiopathic scoliosis who need surgery according to established recommendations, the curve progression can be controlled with conservative treatment until the patient reaches the end of the adolescent growth spurt. The aim of spinal instrumentation is to stabilize the spine until a stable arthrodesis is achieved. This can be achieved posteriorly, anteriorly, or combination of anterior and posterior approaches.

Fig. 10.4 Illustrative example of a Cheneau Brace in adolescent idiopathic scoliosis. This type of brace is applicable adolescents with relatively rigid curves, correction is achieved by translational and rotational forces. (**a**, **b**) Clinical pictures. (**c**, **d**) Radiograph without and with brace, showing the correction of the scoliosis

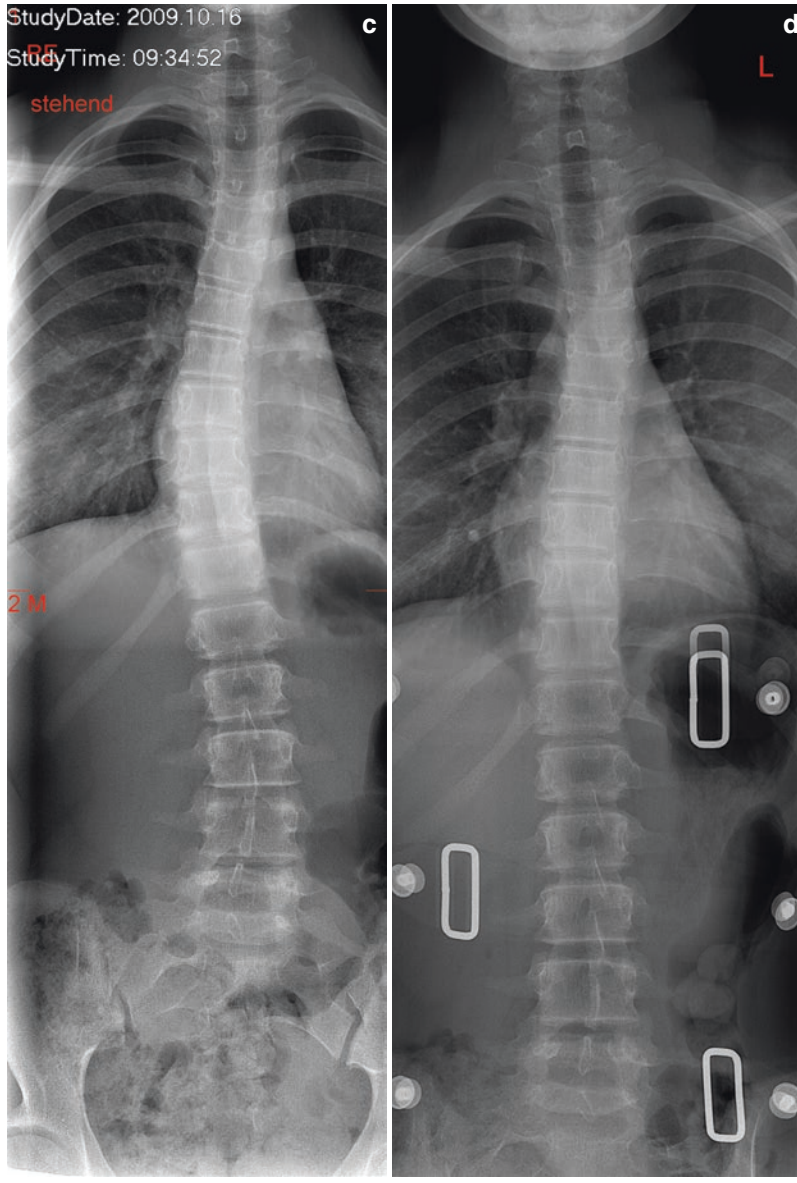
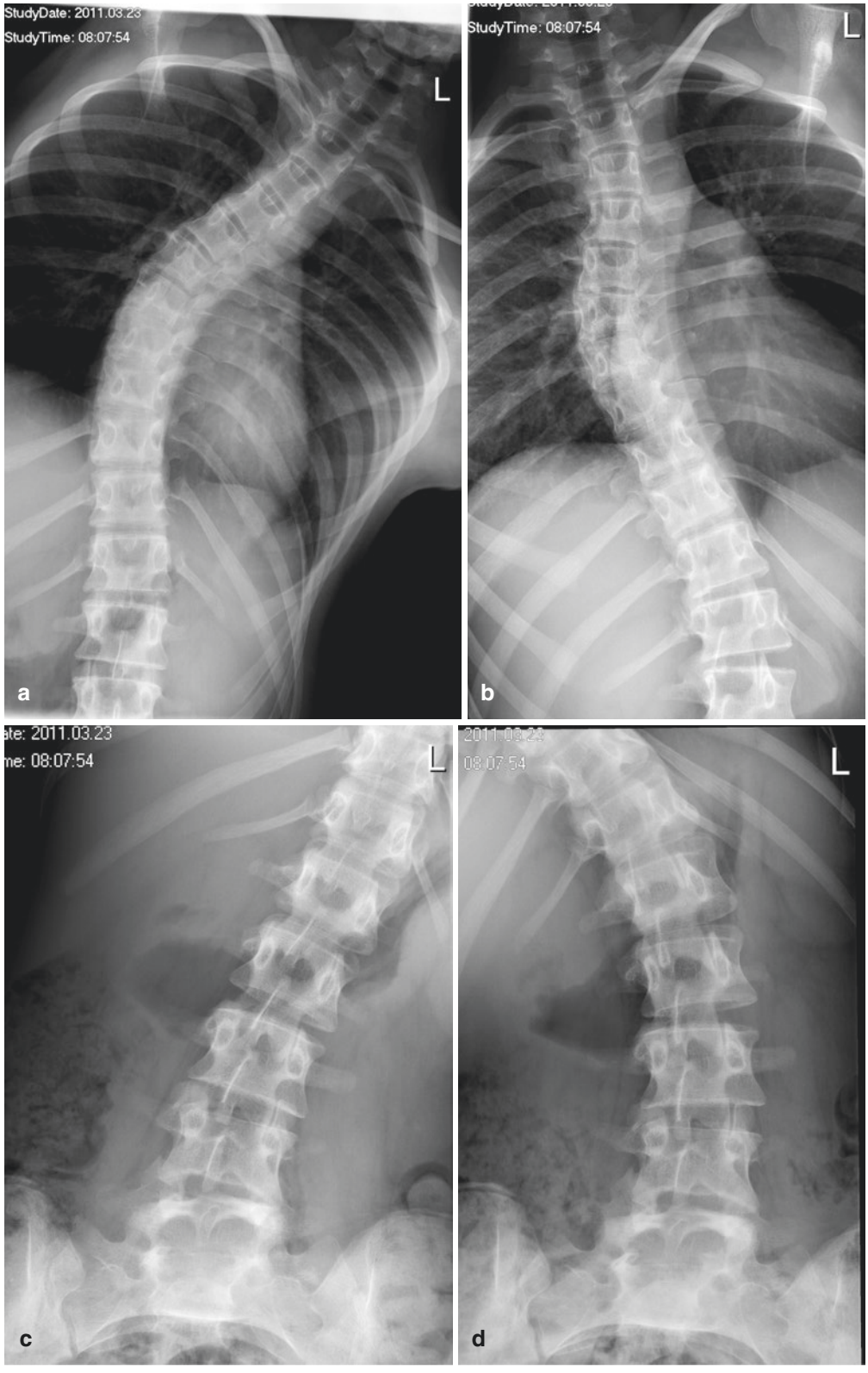


Fig. 10.4 (continued)

Fig. 10.5 (**a**–**d**) Side-bending radiographs of the patient shown in Fig. 10.6. Bending radiographs show that the thoracic curve is structural, the lumbar curve is non-structural and shows almost complete correction in the coronal plane. (**a**) Left thoracic bending view. (**b**) Right thoracic bending view. (**c**) Left lumbar bending view. (**d**) Right lumbar bending view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The selection of approach depends on the type of the scoliotic curve, sagittal deformity, as well as experience and expertise of the surgeon. Anterior approaches are mostly used to correct lumbar and thoracolumbar curves. The advantages of anterior approach are direct access to the vertebral bodies and intervertebral discs and the ability to achieve enhanced correction in the frontal and axial plane, but anterior spinal instrumentation tends to decrease the sagittal plane lordosis. This can be corrected by using interbody support and rigid instrumentation systems. Proponents of anterior surgery claim to be achieving similar or greater correction of the scoliosis with fewer levels of instrumentation [66, 67]. Disadvantages of anterior approach include the invasive approach through abdomen, or thorax and abdomen; chronic pain, disturbing scar and bulge at the incision [68].

In severe and rigid curves (curves greater than 70–75°) a combined anterior and posterior approach may be indicated. Anterior release increases the curve flexibility, permitting greater correction with posterior instrumentation. In addition, fusion can be performed anteriorly as well. Minimally-invasive techniques for anterior release using thoracoscopy and small abdominal approaches are gaining popularity. Thoracoscopic scoliosis surgery with instrumentation has been reported and is applicable for well selected isolated thoracic scoliotic curves [69, 70].

Posterior spinal instrumentation and fusion is the most preferred surgical option for fusion in adolescent idiopathic scoliosis. Improvements in the spinal implants, the flexibility to use pedicle screws, hooks or sublaminar wires permit adequate correction and fusion of the majority of curves. Using the posterior approach one can avoid breaching the thoracic or abdominal cavity. Instrumentation and fusion can be performed posteriorly. Additional surgery like rib-hump resection can be performed in the same session. The posterior central scar is generally more acceptable to the patient than the oblique abdominal or thoracoabdominal scars (Figs. 10.6 and 10.7).

Complications of Surgical Treatment

Complications of scoliosis surgery in idiopathic scoliosis have been decreasing with advances in anaesthesia, intraoperative and postoperative patient monitoring, blood loss management neurological monitoring, and improvement of instrumentation systems.

The incidence of neurological complications after spinal surgery in all types of scoliosis is reported by the Scoliosis Research Society to be 1 %. Patients with congenital malformations and hyperkyphosis are at greater risk for neurological complications.

Complications in the early postoperative period include respiratory compromise, early wound infections, and neurological injury. Neurological deficit may present in the postoperative period due to vascular hypotension, mechanical irritation from the spinal implants, or compressive hematoma.

Hardware related complications may present in early or late postoperative period. In cases of pseudoarthrosis, implant failure is to be anticipated. Delayed infection, loosening, or metal reaction may cause postoperative pain.

Improper selection of the fusion levels or changes in sagittal or coronal alignment may result in junctional problems. Levels adjacent to the fused segments are subjected to more mechanical stress and may present with early degenerative changes.

The mainstay of treatment for idiopathic scoliosis in children remains periodic clinical examinations. The majority of all curves do not need definitive treatment. If progression of the scoliosis above 20° is documented, consequently undertaken conservative treatment with a corrective brace and intensive and regular exercise programs or physiotherapy prevents further progression. When a progression of curves above 40° is documented, surgery should be advised before further deterioration occurs. Optimal timing of scoliosis surgery can avoid long instrumentations and fusions. The goal of surgery in children and adolescents with idiopathic scoliosis is to stop the progression of a curve and provide the patient with a balanced spine.

Idiopathic Scoliosis and Chest Wall Deformities

The interrelation of growth of the spine, chest wall, and lungs is well recognized. Following pathologies will be enlightened in this chapter:

Disturbed lung development
Pectus excavatum
Pectus carinatum
Deformity in association with rare disorders

Growth of the thorax is exponential in the first 5 years of life, with a fivefold increase in the thoracic volume. Growth oft he lungs occurs by both volume expansion and tissue hypertrophy. The increase in the number of alveoli occurs predominantly before the age of 8, the most rapid phase is seen in the first 5 years of life. In late childhood the increase in lung volume is predominantly due to increase in size of the alveoli [71–73].

The height of the spine represents 60 % of the sitting height. After the first period of growth acceleration in early childhood, the second occurs during the pubertal growth spurt. In patients with spinal deformities, corrected standing height can be estimated using arm span measurements, the standing height being approximately 97 % of the arm span, with minimal gender differences [74, 75].

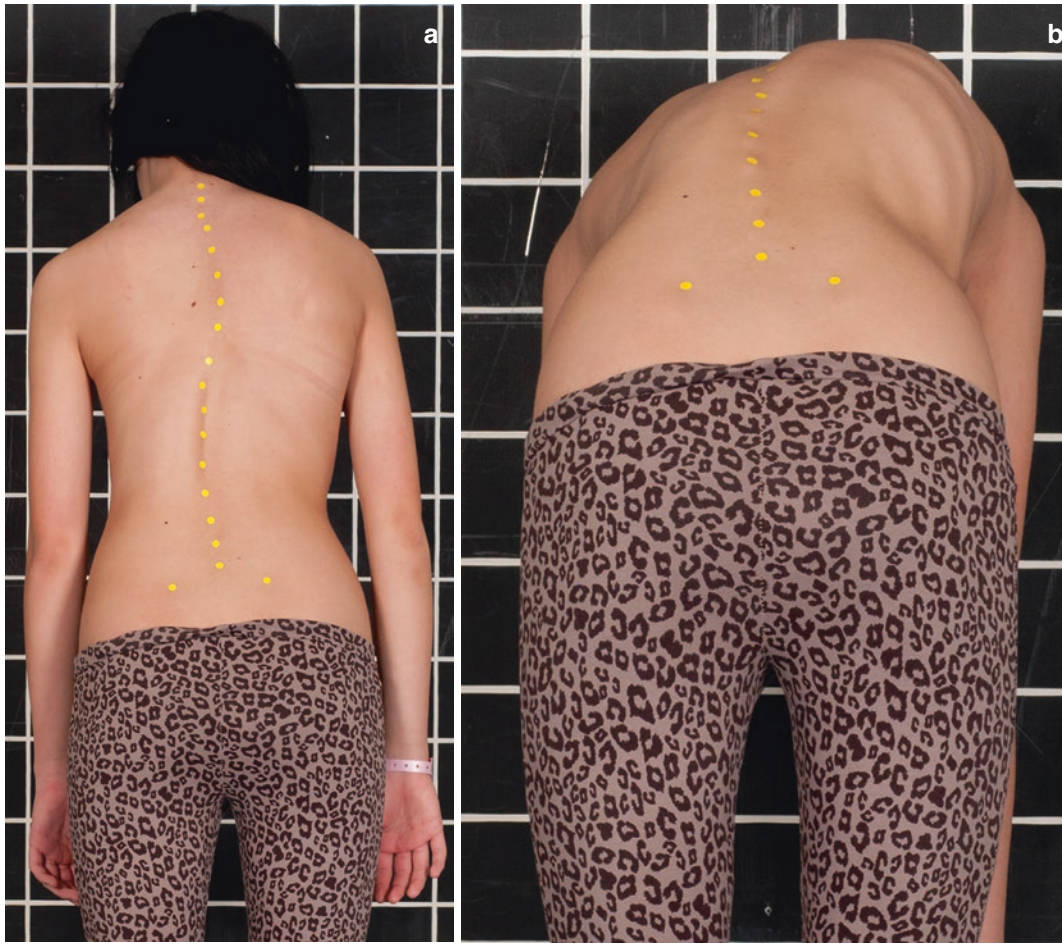

Fig. 10.6 (**a**–**d**) Clinical appearance in a case of adolescent idiopathic scoliosis before surgery and postoperative result. Note improvement in rib-hump and trunk balance due to three-dimensional correction of the spine. (**a**) Preoperative standing. (**b**) Preoperative forward bending. (**c**) Postoperative standing. (**d**) Postoperative bending

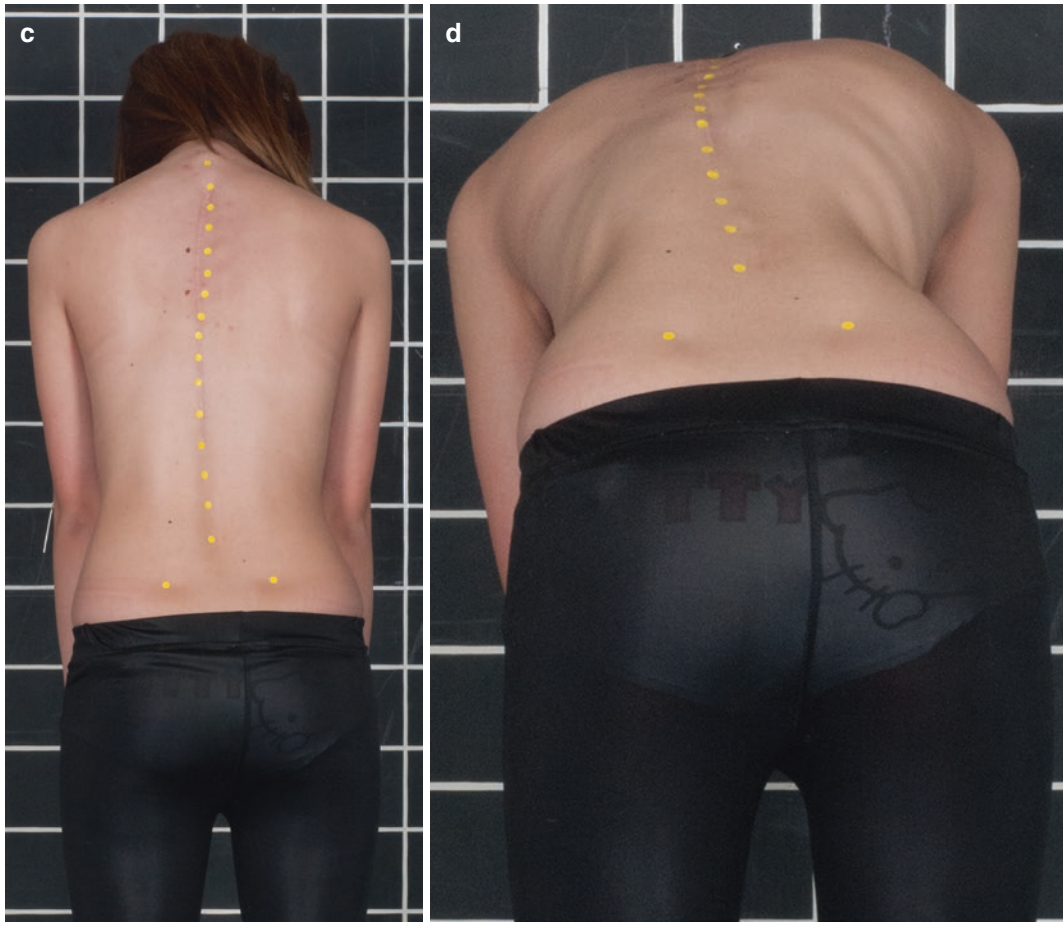

Fig. 10.6 (continued)

In view of the rapid development of the spine and the thorax, early onset deformities, either isolated or in combination, demand treatment in order to prevent disturbance of lung development. Although conservative treatment is sufficient in the majority of chest wall deformities associated with idiopathic scoliosis, a progression of the deformity particularly in the first 5 years of life might necessitate surgical intervention, either from the pediatric surgeon or the pediatric orthopedic surgeon.

An association between anterior chest wall deformities and idiopathic scoliosis is described in the literature but is poorly defined. Reported prevalence of combined severe anterior chest wall deformities and spinal deformities range between 5 and 22 % [76, 77]. The presence of chest wall deformities does not influence the course of disease in idiopathic scoliosis. Thus, the management of scoliosis is, as mentioned above, conservative in the majority of cases.

The associated deformities of the anterior chest wall may need more invasive treatment. For a pectus excavatum, when coexisting with idiopathic scoliosis, there are relatively few options for conservative management. In cases showing progression of the deformity, a surgical option for correction of the deformity should be discussed with the patient and his/her family. Pectus carinatum on the other hand can be managed conservatively with special bracing and shows satisfactory results, particularly if the treatment is initiated in the early growth years. The success of bracing depends on the remaining growth years

which permit remodelling of the thoracic cage, and the compliance of the patient. A recent survey by the Canadian Association of Pediatric surgeons showed that over 80 % of the surgeons agreed that bracing was generally preferable to surgical repair; and that bracing should be the first line of treatment [78]. Surgical correction is generally necessary in cases showing progression during brace treatment, or noncompliance with brace treatment with eventual progression of the chest wall deformity [79]. A progression of the chest wall deformity is not associated with a progression of the spinal deformity in idiopathic scoliosis.

Although not the topic of this chapter, chest wall deformities and scoliosis associated with rare

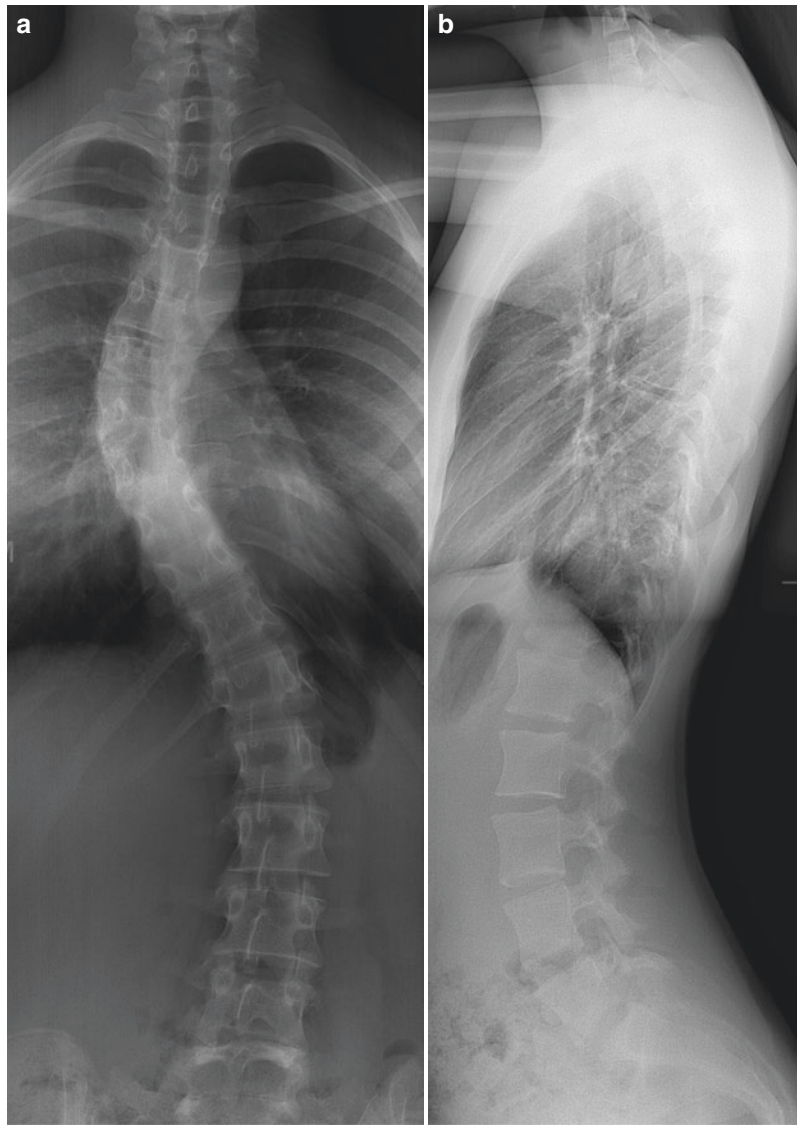

Fig. 10.7 (**a**–**d**) Preoperative and postoperative radiology in the patient illustrated in Fig. 10.6. Note the impressive but non-structural lumbar curve which was not fused. Postoperative radiographs showing correction of the thoracic hypokyphosis; the non-structural lumbar curve shows spontaneous regression. (**a**) Preoperative PA radiograph. (**b**) Preoperative lateral radiograph. (**c**) Postoperative Frontal radiograph. (**d**) Postoperative lateral radiograph

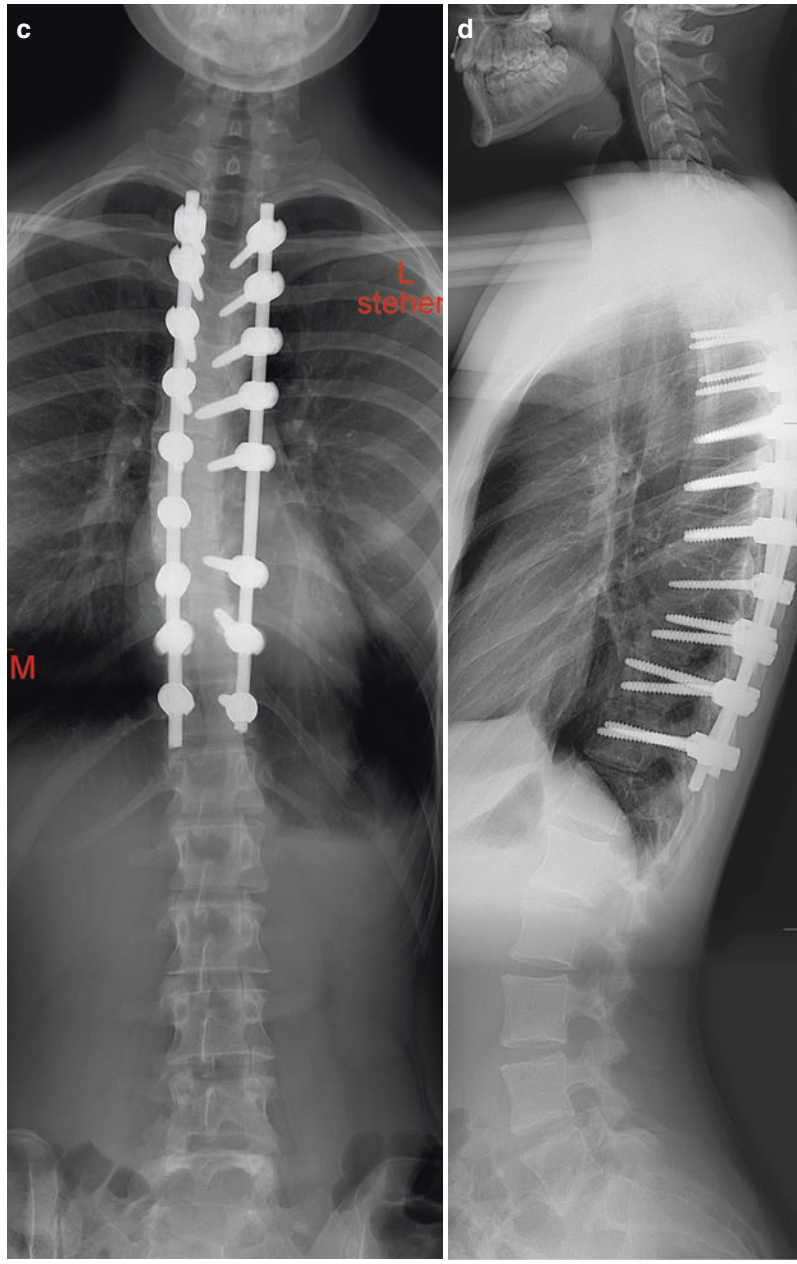

Fig. 10.6 (continued)

pediatric disorders and exotic congenital syndromes pose special challenges for the pediatric and the spinal surgeon. Typical disorders associated with severe chest wall and spinal deformities are Marfan Syndrome, Jeune Syndrome, Jarcho-Levin Syndrome and the Cerebrocostomandibular Syndrome. The management of these conditions is very demanding, and need familiarity with the disease, a multi-speciality involvement in accurate diagnosis of the condition. The co-morbidities and mortality of the condition need to be evaluated prior to defining the treatment options [80].

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11 Psychologic Effects, Body Image, and Pectus Excavatum and Carinatum

Robert E. Kelly Jr. and Michele L. Lombardo

Introduction

Young people with pectus excavatum or carinatum seek treatment for physical symptoms, concerns about chest appearance, or both. It has long been recognized that the appearance of the chest is a major concern to patients with pectus excavatum. Although pectus excavatum was described in 1596, the first surgical correction was not even attempted until 1911. Surgical progress was rapid, however, and already by 1949, Charles W. Lester of New York wrote, "The psychological aspect of the situation is of great significance". The girls do not pay much attention to the deformity until their breasts begin to develop and they start to wear a brassiere. Then they become conscious of it. The boys notice their deformity earlier probably because they run around unclad with other boys who take delight in calling attention to any deformity with derisive language. Many children take this teasing without trouble, but others cannot and they are entitled to a corrective operation [1]. In 1958, Adkins and Gwathmey of Washington, D.C. stated, "The other and equally valid indication for surgical correction is for cosmetic and psychologic reasons. A depression or deformity of the chest wall may be a source of considerable embarrassment, especially during adolescence and young adulthood. Consequently, operative correction of such a deformity is well justified even in the absence of symptoms" [2]. Mark M. Ravitch, in the influential 1962 textbook, *Pediatric Surgery*, offered that, "Subject to more argument, perhaps, is the psychologic importance of the essentially cosmetic deformity. It…is very rare for parents to present the psychologic effects as reason for operation, or children either…they want the operation only if it is indicated for "reasons of health." But after operation, attention is centered almost entirely on the visible correction of the deformity and the relief this gives, usually to three generations of the family. Many children, both males and females, come for operation in early adolescence, when they become reluctant to undress before their fellows" [3]. By the 1970's, operations designed to restore the contour of the chest wall with implants illustrated the need to improve the appearance [4]. Lacquet and colleagues reported from the Netherlands in 1998 that surgical treatment of pectus excavatum affected patients' self-perception: "The psychologic results were usually striking. In many cases it was only after the operation that patients admitted to having been distressed by their deformity. They lost all traces of an inferiority complex, and the boys rather proudly displayed

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their surgical scar. Generally most patients were very thankful and enjoyed the result of their operation" [5].

Body Image Concept

E. Einsiedel, a clinical psychologist from Mainz, Germany, wrote on the dual nature of patients concerns in 56 patients with funnel chest. He referred to this as "interlocking-systemic diagnosis". He reported that children over 11 years old "display as a whole more psychological disorders. Along with specific embarrassment reactions, social anxiety, feelings of stigma, limited capacity for work, orientation toward failure, reduced tolerance of frustration and temptation, limited capacity for communication, and even markedly depressive reactions are observed." The frequency of occurrence of each was reported, and varied from 58 to 94 % of the children. Children less than 11 years old were much less frequently affected (4.5–40 % depending on category) [6].

Physical attractiveness as a separate field of study within psychology was advanced by seminal work by Dr. Thomas F. Cash of Old Dominion University. He researched "the influences of various aspects of physical appearance on human behavior" [7]. He uncovered evidence that physical appearance had a major effect on physical and mental health and identified links between self-concept, social competence, and personality functioning. This work validated the concept that persons who believe themselves to be less attractive are less social, self-accepting, and independent [8]. Behavioral changes range from benign to maladaptive. Cash indicates that women who perceive themselves as obese use more facial makeup, for example [7]. Anorexia nervosa, on the other hand, is an example of a psychopathology induced by the same concerns. Depression is another psychopathology linked to adverse opinion of one's attractiveness. There is an inverse relationship between body image and depression [7].

Successful treatment of congenital craniofacial deformities was developed in the 1960s and 70s by Paul Tessier in France, Milton Edgerton in Virginia, and others. These treatments were shown to have positive effects on body image [9–12]. The concerns of burn patients regarding body image have received increased attention as well [13].

Body Image in Pectus Excavatum

Given the many clinicians who noted psychological improvements in pectus patients after surgery, scientific investigation of these effects followed, at first on a small scale. In Vancouver, Canada, Roberts and colleagues conducted structured interviews of five children who had undergone the Nuss procedure, and their parents, studying then for a total of ten subjects. They used Keith and Schalock's quality of life model. They concluded that the patients interviewed had a significant improvement in quality of life, which they attributed to the surgery [14].

In considering development of psychological evaluation, discussion of the clinical experience is a starting point. Surgeons find that adolescents and young adults who present for evaluation of pectus excavatum or carinatum often must be directly asked to remove their shirts for physical exam. For females, this may be due to modesty. Males do not typically exhibit such modesty issues; however, there remains a real reluctance to be examined. In contrast, after operation, boys often lie in bed recovering from operation with no shirt on, and no sheet over their chest. At clinic follow up 2 weeks postop, they are like Superman in a phone booth, rapidly removing their shirt, eager to show off their new chest.

Evaluation of Body Image in Pectus Excavatum

When approached about this phenomenon, Cash and associates developed a psychological tool, the Pectus Excavatum Evaluation Questionnaire, or PEEQ. This 24 question inventory includes questions concerning both physical symptoms and body image. It was designed to evaluate the

quality of life. To validate the new instrument, we administered a child and parent version of the questionnaire to 22 parents and 19 children (ages 8–18) before surgery and 6–12 months after repair by Nuss procedure. The instrument had high test-retest reliability. Children reported significant improvements in exercise intolerance, shortness of breath, and tiredness. Of nine questions asking the children how they feel or act about their bodies, all but one question showed significant improvement after surgery. Parents also reported significant improvements in their child's exercise tolerance, chest pain, shortness of breath, and tiredness, and decreases in the frequency of the child being frustrated, sad, self-conscious, and isolated. These data were reported by our group in 2003 [15].

Subsequently, as part of a multicenter study of pectus excavatum, the PEEQ was administered by the research coordinator, by telephone, to parents and patients (8–21 years of age) before and 1 year after Nuss or open surgery. From 2001 to 2006, 264 patients and 274 parents completed the postoperative questionnaire. Preoperative psychosocial functioning was unrelated to objective pectus excavatum severity (computed tomographic index). Patients and their parents reported significant postoperative changes. Improvements occurred in both physical and psychosocial functioning, including less social self-consciousness and a more-favorable body image. Ninety-seven percent of patients thought that surgery improved how their chest looked. We concluded that surgical repair of pectus excavatum can significantly improve both the body image difficulties and limitations on physical activity experienced by patients [16].

By request, we subsequently provided the PEEQ without cost to other investigators, who have confirmed these results.

The Vancouver group followed up their 2003 report in 2008 with an analysis of 43 patients who underwent surgery. They were studied both with the PEEQ and with the Child Health Questionnaire (CHQ-CF87). Results were compared to age-matched Australian norms. The majority of patients underwent the open operation, and 44 % the Nuss procedure. The study concluded that patients undergoing surgery for pectus excavatum by either Nuss or Ravitch procedure have similar clinical and health-related-quality of life (HRQL) outcomes. As a group the PE patients have poorer HRQL scores than age-matched population norms [17].

In London, England, the Royal Brompton Hospital Group used a modification of the PEEQ questionnaire (which they called NQ-mA for Nuss questionnaire modified for adults) and a new Single Step Questionnaire (SSQ) to assess 20 male patients with median age 18 years (range 14–37 years). The questionnaires were adequate to measure disease-specific quality of life changes after surgery and were able to confirm that the Nuss procedure improves the quality of life in young male adults with pectus excavatum [18]. Separately, they showed that the SSQ correlated with the PEEQ modified for an adult population.

Recently, there has been great deal of publication related to health-related quality of life with pectus excavatum. From Denmark, Jacobsen, Pilegaard and colleagues studied 172 children and adolescents ages 8–20 years, 86 % of whom were males. They used the Child Health Questionnaire CHQ-CF87 for children and the CHQ-PF50 for adults. They also used the PEEQ modified for adults (NQ-mA) and the SSQ using an instrument other than the PEEQ have shown that in five subscales of self-esteem, behavior, emotional role, mental health and family activities, the PE group had a better HRQoL [19].

Ohno and colleagues from Osaka, Japan, evaluated 36 patients with pectus excavatum aged 1–22 years old. Respondents were asked whether they suffered psychological distress and if they wanted surgery. The severity of the deformity was measured radiographically. The severity of the deformity was worse in the patients who suffered psychological distress [20].

The group from Graz, Austria performed a comprehensive psychological investigation of 17 patients in 2003, and repeated the battery of tests in 2007 after Nuss operation to correct pectus excavatum at a mean age of 19.6 years. Nearly all patients' preoperative expectations were confirmed postop. Data from SCL-90-R, measuring mental exposure, had normal range. The authors

conclude that "the long-time follow-up can make us sure that the Nuss procedure as a physical treatment has positive effects on physical as well as psychologic aspects of young adults." Patients were pleased with the cosmetic effects of operation [21].

In Erlangen, Germany, 90 patients and 82 controls were assessed with the PEEQ modified for adults, also the FKB-20 Body Image Questionnaire, the Dysmorphic Concern Questionnaire, and the Diagnostic Interview for Mental Disorders-Short Version (Mini-DIPS), the General Depression Scale, and a self-rating of self-esteem were used to evaluate general psychological impairment. The study reported that, compared with control group results, the physical quality of life was reduced in patients with pectus excavatum, while the mental quality of life was decreased in patients with pectus carinatum. Body image was highly disturbed in all the patients. Patients with pectus carinatum appeared to be less satisfied with their appearance than those with pectus excavatum. Body image did not influence physical quality of life. Patients displayed no elevated rates of mental disorders according to Diagnostic and Statistical Manual of Mental Disorder, fourth edition (DSM-IV) criteria. Based on mean scores, patients rated their appearance significantly more negatively than adult raters but only slightly different than other adolescent raters. Adolescent raters' judgment was related to chest wall deformity characteristics. Self-rating seems to be related to psychosocial factors. The group suggests that effective interventions focusing on social interactions are needed, since adolescents' evaluation of appearance might affect patients' psychological functioning [22].

In Hanover, Germany, a study was conducted to demonstrate that the improvement of quality of life demonstrated following Nuss procedure persisted after removal of the bar. Forty patients were assessed a mean of 54 months following Nuss procedure. Patients were interviewed 6 months after the bar was put in, and then 23 months after bar removal on average. There was a high level of persistent satisfaction after bar removal, and the positive impact of MIRPE (Nuss procedure) on psychosocial and physical well-being was documented, which persisted up to 4 years after bar removal [23].

A follow up from the group from Hanover evaluated correlation between general practitioner (GP) and patient evaluation of outcome. This was done because of concern that patient ratings may be biased by high preoperative patient expectations. Thirty-nine patients and their GPs were asked to complete a questionnaire to evaluate their opinion on psychosocial and physical well-being of the patients. Only 56 % of the GP's completed the questionnaire, but there was a highly significant correlation between GP and patient ratings. Detailed analysis showed that GPs scored several items differently to their patients. GPs scored social activity and preoperative self-esteem lower, but pain and impairment of daily activities during the period with the implanted bar significantly higher than the patients [24].

In Sichuan, China, a study of 337 children aged 6–16 years over a 3 year period was performed. Psychosocial functioning was assessed by a Child Behavior Checklist. A Chinese Mandarin version of the 1991 CBCL, previously validated for reliability and discriminant validity in Chinese children, was used [25, 26]. Four hundred and fifteen patients were initially evaluated, but 38 patients did not complete the study, and 40 were later found to be ineligible. The deformity was first noted at 4, 5, or 6 years of age in 75 % of children. Only three patients first noted it after the age of 10 years. Dissatisfaction and being teased about the deformity were major factors in seeing treatment. Thirty-seven percent of patients asked their parents to be taken for treatment. Avoidance of exposing the chest in public was reported by 44 % of children. Teasing, always by non-family members, was reported by 23 % of children; notably, 97 % of the time teasing was perpetrated by other children. Age correlated strongly with psychosocial problems: compared with the group under 9 years, the group from 12 to 16 years was at higher risk for such problems. Multivariate analysis showed that age, severity of the malformation, and being teased correlated with psychosocial

problems. As the authors discussed, the correlation of psychosocial distress with severity of the depression stands in marked contrast to the CHKD/EVMS report [16]. The authors use a different tool to study the psychosocial distress. Mother's education was not a significant predictor or psychosocial problems. This work suggested that children with PE have more psychosocial problems than children from the general population. Multiple medical and psychosocial factors were associated with patients' impairment of psychosocial functioning [27].

Body Image in Pectus Carinatum

There has been little publication concerning these effects in pectus carinatum. However, in 2013, a group from Istanbul, Turkey, studied 30 patients who had undergone minimally invasive repair of pectus carinatum using the PEEQ. There was found to be significant improvement following surgery, confirming for the first time that the findings in pectus excavatum were also seen in pectus carinatum [28].

Conclusion

In summary, concerns about physical appearance are present in the vast majority of children and adults with pectus deformity; operation to improve chest wall contour improves body image.

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12 Anesthesia and Pain Therapy for Surgery of Chest Wall Deformities

Maria Vittinghoff and Anton Gutmann

Abbreviations

NSAIDs	Non-Steroidal Anti Inflammatory Drugs
PACU	Post Anesthesia Care Unit
PCA	Patient-Controlled Analgesia

In most institutions minimal access repair of pectus excavatum (MARPE) is preferred. Therefore in this chapter the focus will be predominantly on pectus excavatum which is the most common deformity and on the management of patients undergoing MARPE. Although MARPE is performed through minimal skin incisions, this procedure is associated with significant injury of intercostal muscles and nerves irritation/damage and therefore offers no reduction in postoperative pain when compared with open procedures [1, 2]. From the patient's point of view this surgical approach in which minimal skin incisions are involved is thought and expected to be associated with minimal pain. In order to avoid respiratory suppression and delayed mobilization due to systemic opioid administration, adequate postoperative pain management by regional anesthesia using catheter techniques is now the management of choice. Several analgesic methods for MARPE including thoracic and lumbar epidural analgesia have been described [1–7]. Senturk et al demonstrated a superior prevention of acute and long-term thoracotomy pain when thoracic epidural analgesia was offered preoperatively when compared with sole intravenous opioid administration [8]. Thoracic epidural analgesia bears a risk for severe complications such as dural puncture, epidural abscess, and spinal hematoma. Potential damage of the spinal cord should be considered when choosing a safe method for postoperative analgesia in an esthetic procedure such as MARPE.

For this, a paravertebral block is administered outside of the spinal canal is preferred as it is associated with minimal risks for spinal damage. Figure 12.1 illustrates the difference between epidural and paravertebral access for regional anesthesia to clarify this point. In the case of chest wall deformities two paravertebral catheters are required for bilateral analgesia (Fig. 12.2). The continuous paravertebral block which has originally been described by Lonnqvist for pain management after thoracotomy in children was found to be superior to epidural analgesia using bupivacaine [9, 10]. While this technique carries a potential risk of infection in paravertebral spaces and pneumothorax, minimal risk of spinal cord injury and epidural abscess still exist. While

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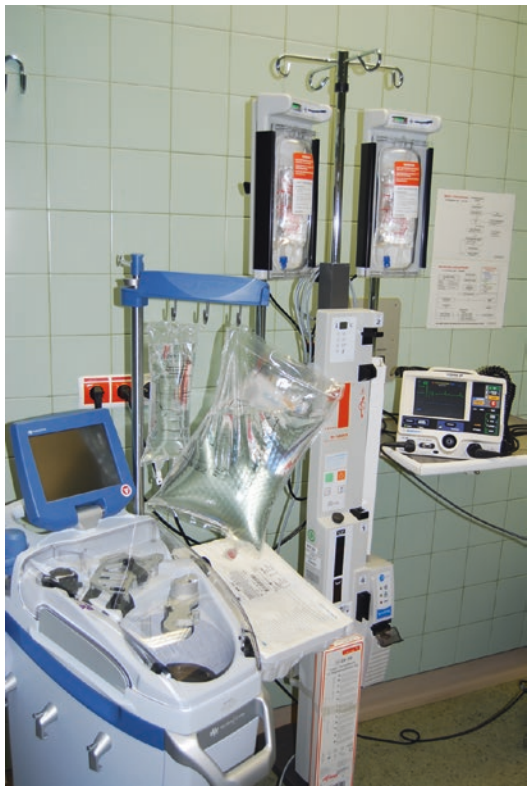

Fig. 12.1 Emergency equipment

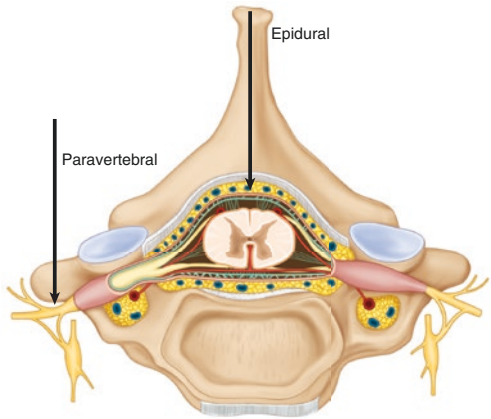

Fig. 12.2 Epidural/paravertebral space: The two arrows illustrate the epidural (*middle black arrow*) and paravertebral access (*left black arrow*) for regional analgesia. The epidural access includes the risk for spinal cord damage, whereas the risk with paravertebral access is limited to single segmental spinal nerve damage

trauma in open procedures is evident, the amount of intercostal trauma and tension to the elevate structures, that is caused by the inserted pectus bar, may vary among patients and therefore cause different intensities of pain. Even the fact whether single or double-bar placement for MARPE is used, influences the intensity of pain. Application of two pectus bars in suitable patients has been determined to be associated with decreased postoperative pain after MARPE [11].

Age is also known to influence pain intensity after MARPE [12]. Regional anesthesia has repeatedly been demonstrated to be superior in pain management compared to intravenous opioid analgesia in chest wall excursion after trauma and after pectus excavatum repair [13–16]. However, some other groups do not recommend epidural anesthesia over intravenous Patient-Controlled Analgesia (PCA) [17]. When PCA is used, a small dose of ketamine can be given to reduce the total administered dose of intravenous opioids and hence decreases the incidence of pruritus [18].

Preoperative Evaluation and Preparation

The most effective preoperative medication is a friendly, reassuring visit from the anesthesiologist. Additional to preoperative history and physical examination, exercise intolerance and respiratory tract infections must be evaluated carefully. **Laboratory tests** should include: red and white blood cell count, coagulation test and tests depending on the presence of any underlying conditions. Whether **blood products** must be made available or not depends on the surgical method. MARPE generally does not need substitution of blood products. But in case of unexpected major bleeding, depending on the weight of the patient, one to two units of packed red cells should be cross matched and be available prior to commencement of the surgical procedure. **Cardiovascular and pulmonary exercise performance** tests are rarely pathologic in common chest wall deformities like funnel chest. Mild decreases in forced expiratory flow and lung volumes as well as in

cardiac output resulting from limitation of diastolic filling have been observed in these patients [19]. Irrespective of that, the insertion of a pectus bar through the chest wall has been found to result in significant changes in respiratory mechanics and gas exchange [20] Therefore, **teaching and training of deep diaphragmatic breathing** during the preoperative preparation is crucial as thoracic excursions will be altered when the patient wakes up postoperatively. In case of a patient with concomitant asthmatic disease, treatment must be optimized and continued perioperatively. Computed tomographic scans if available can offer additional information in the preoperative planning. Normal eating until the evening before surgery and intake of clear fluids until 2 h before the induction of anesthesia is encouraged. Oral premedication in most institutions is done with midazolam. **Prophylactic antibiotic treatment** is provided according to the hospital protocols. During the preoperative visit the attending anesthetist should explain the details of general anesthesia to the patient and present the concept of patient controlled analgesia for the postoperative pain management. In order to avoid respiratory depression and delayed mobilization due to systemic opioid administration, adequate postoperative pain management by **regional anesthesia using catheter techniques** is preferable. Several analgesic methods including thoracic and lumbar epidural analgesia as well as bilateral paravertebral block may be employed. The lumbar epidural approach with the intention to thread the catheter further up into the thoracic region may result in inadequate pain therapy. On the other hand, thoracic epidural analgesia is also at risk for severe complications such as dural puncture, epidural abscess, and spinal hematoma. Potential damage of the spinal cord should be considered when choosing a safe method for postoperative analgesia. Bearing these aspects in mind, it is preferred to perform a paravertebral block outside of the spinal canal and minimize the risk for spinal damage. Although this technique carries a potential risk of infection in paravertebral spaces and pneumothorax, it poses minimal risk of spinal cord injury and epidural abscess formation. This risk-benefit analysis has to be kept in mind when offering and explaining the different possibilities of regional anesthesia to the patient. Furthermore absolute (such as coagulation disorders and patient's rejection) and relative (e.g. pre-existing neurological deficits) contraindications for performing a central regional anesthesia have to be considered. Although patient controlled regional anesthesia would be the preferable method for postoperative pain management, the alternative of intravenous patient controlled analgesia has to be offered as well. The advantage of this method is to avoid neurological side effects at the cost of possible opioid induced over sedation, respiratory depression and nausea and vomiting. After careful decision-making the chosen method has to be explained in detail to the patient and informed consent has to be taken. At the same time the patient has to be advised in the use of the PCA-infusion pump.

Induction, Maintenance and Monitoring of Anesthesia

As a matter of principle, the anesthetist should use those drugs and methods he is most experienced with for induction and maintenance of anesthesia. Deep anesthesia and muscle relaxation during intrathoracic manipulation is crucial to avoid injury due to spontaneous movement or coughing of the patient. In our institution **total intravenous anesthesia** is favored over balanced anesthesia using an inhalational agent for maintenance. However, Nitrous Oxide should not be used for endoscopic procedures, as the pleura may be punctured during the procedure and pneumothorax cannot be excluded. General anesthesia is induced with fentanyl (2 μg/kg), propofol (2–5 mg/kg) and rocuronium (0.6 mg/kg) or cisatracurium (0.1–0.15 mg/kg). After orotracheal intubation, pressure controlled ventilation is performed; after which remifentanil and propofol are used for maintenance of anesthesia. The placement of an epidural catheter allows intraoperative supplementation with local anesthetics and postoperative pain management

with epidural anesthetics. Intraoperative **invasive arterial blood pressure monitoring** is preferred and always installed for the MARPE in our institution. A central venous line is not mandatory but may be helpful in patients with poor vascular access. Additional devices such as urinary catheter, nasogastric tube and a temperature probe are used for intraoperative monitoring. In MARPE severe complications are rare but the most feared and life threatening complication is cardiac perforation. To be prepared for **sudden major blood loss** the cross matched red blood cell units, an autologous blood collecting systems such as a cell saver and a rapid infusion system (Fig. 12.1) should be on hand in the operating room. Prophylactic antibiotics are administered according to the hospital guidelines.

Regional Anesthesia

Catheter Placement

An agreement exists, whereby in adult patients the regional catheter is placed in the awake or only slightly sedated patient. On the contrary, in children the regional catheter is placed under absolute sterile conditions after induction of general anesthesia. Using either the lumbar or thoracic approach the epidural catheter needs to be placed optimally [21]. In children the median approach to the epidural space is preferred (Fig. 12.2). Especially in small children the pediatric anesthetist has to be aware of the different anatomical and pharmacological conditions specific for this age group. The skin-epidural distance in which the epidural space has to be expected can be calculated using Busoni's formula: (Age in years × 2) + 10 mm = distance (mm) [22]. Also, for the puncture of the epidural space Touhy-needles of different age adjusted sizes and the fitting indwelling catheters are used (Table 12.1). After identification of the epidural space, using the loss of resistance technique with sterile saline, the epidural catheter is advanced a few centimeters over the tip of the epidural needle (Figs. 12.3 and 12.4). In the literature, an increasing number of publications are now advocating the use of ultrasound to guide placement of epidural

Table 12.1 Size of needle for epidural catheters

Weight in kg	Size of needle
10–30 kg	19 gauge
>30 kg	18 gauge

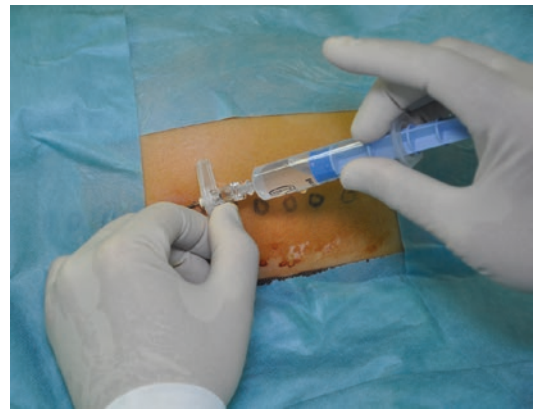

Fig. 12.3 Identification of epidural Space with loss of resistance technique

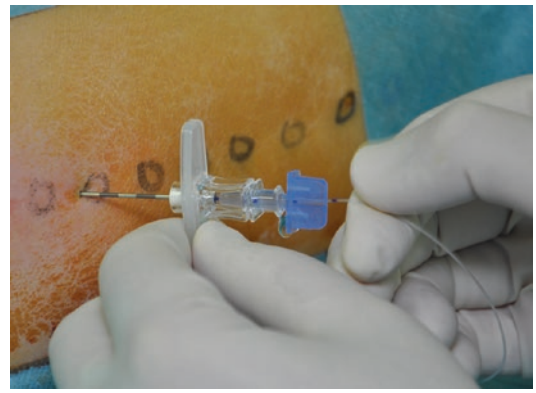

Fig. 12.4 Insertion of epidural catheter

catheters. Once the catheter is positioned and a test dose is administered, the catheter is secured to the skin and an anti-bacterial filter is attached to the catheter before the full dose of local analgesia is administered (Fig. 12.5). A bilateral paravertebral block may be an alternative regional technique when thoracic epidural block is contraindicated (Fig. 12.2). This method which was first published by Lönnquist provides adequate intra- and postoperative analgesia. The disadvantages of this method are that two regional catheters have

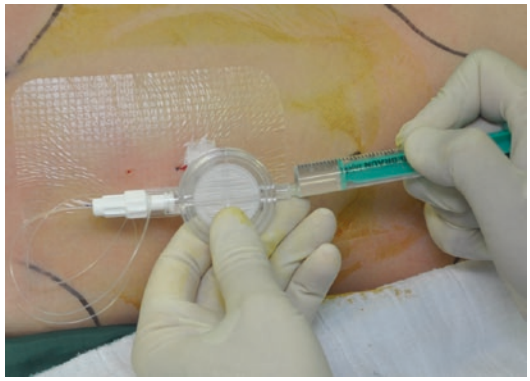

Fig. 12.5 Epidural catheter with bacteria filter

to be placed and therefore also two PCA infusion pumps have to be provided during the postoperative period.

Test Dose

When an 18G needle is used for the initial puncture, in case a blood vessel is inadvertently punctured, there is a high probability that blood would flow passively backwards through the needle bore and could be aspirated through the catheter. Hence using a smaller gauge needle is not advisable, as there is a smaller chance to aspirate blood through smaller needles if the catheter is mal positioned. To eliminate the danger of an intravascular position of the catheter, as far as possible, a test dose with Lidocaine 1 % plus Epinephrine 1:200,000, 0.1 ml/kg with a maximum of 5 ml is administered after positioning of the epidural catheter [23, 24]. The effect of intravascular epinephrine would be an increase in heart rate. But it is well known, that is still not a 100 % reliable test. Bearing this in mind, it is advisable to administer local anesthetic drugs slowly and in small increments. During the injection phase heart rate (rhythm, S -T wave) and blood pressure have to be monitored carefully [25].

Local Anesthetics for the Intraoperative Management

The local anesthetic drugs (Tables 12.2 and 12.3) which are most frequently used for epidural anesthesia are bupivacaine, levobupivacaine and ropivacaine. Lidocaine is only used for the test dose [26–28]. In general, it is advantageous to have a motor block during the operation. For this reason higher concentrations of the local anesthetic drugs are used intraoperatively even when the patient is under general anesthesia (Table 12.4). The amount (in ml) of the local anesthetic drug, which is used, depends on following factors:

- Concentration of local anesthetic drug
- Age of the patient
- Position of the epidural catheter (lumbar or thoracic epidural)

Considering these facts and the maximum amount of local anesthetic drug that is allowed pro kg of bodyweight for the single dose the following volumes of local anesthetic drugs are administered as follows:

- Lumbar: 1 ml/kg (max. 30 ml)
- Thoracic: 0.5–0.75 ml/kg (max. 20 ml)

Table 12.2 Local anesthetic drugs: maximal single dose

Local anesthetic drug	Dosage (mg/kg)
Lidocaine	4–5 (test dose)
Bupivacaine	2–2.5
Levobupivacaine	2–2.5
Ropivacaine	3–4

Table 12.3 Characteristics of different local anesthetics

	Bupivacaine	Levobupivacaine	Ropivacaine
Protein binding	96 %	97 %	93–96%
Onset	10–15 min	8–12 min	8–10 min
Analgesia duration	3–6 h	3–7 h	2.5–9 h
Motor block	Pronounced/longer	Medium/longer	Less/shorter

Table 12.4 Concentration of intraoperative local anesthetics

Local anesthetic drug	Concentration (%)	Maximal dosage (mg/kg)
Bupivacaine	0.25–0.5	2–2.5
Levobupivacaine	0.25–0.5	2–2.5
Ropivacaine	0.2–0.375	4 (in children)
Ropivacaine	0.375–0.75	4 (in adults)

Adjuvants

In order to potentiate and to prolong the effect of the local analgesic drugs adjuvants are added. The most frequently used adjuvants are **adrenaline, clonidine, opioids (morphine, fentanyl sufentanil) and s-ketamine** [29, 30].

Adrenaline

In the history of regional anesthesia adrenaline (1: 200 000) has been the most commonly used adjuvant [31]. Adrenalin causes vasoconstriction of the epidural blood vessels. This reduces the vascular uptake of the local anesthetic drug, decreases the maximum plasma levels and prolongs its effect. But since adrenaline is most effective with short acting analgesic drugs, its use is contraindicated with ropivacaine which has its own vasoconstrictive properties.

Clonidine

Clonidine is an alpha2 – agonist which is well known as antihypertensive drug, but was used over the years in a variety of routes mainly for its sedative effects [32]. The epidural application of clonidine causes a direct analgesic effect by acting on the dorsal horn of the spinal cord, thus potentiating both effect and the duration of the local analgesic drug [33–36]. A very welcome side effect is a certain amount of sedation which is due to its action over the locus ceruleus. However, sedation does not depend on the amount of clonidine in the cerebrospinal fluid, but on its plasma levels and administered dose. Only when 2 μg/kg or more are administered through the epidural route, this sedative effect can be observed. Since the plasma levels are very low, changes in blood pressure are rare and not observed. The dosage of Clonidine is as follows:

- Single dose: 1–2 μg/kg
- Continuous infusion: 0.2–0.4 μg/kg/h

Opioids

Opioids are frequently used as adjuvants for epidural anesthesia. They act through their receptors which have been identified at the dorsal horn level of the spinal cord. Due to vascular uptake, epidural opioids can cause the same adverse effects as administered by any other parenteral route, however with the one difference, that the plasmatic levels are quite low. The usual adverse effects are:

- Nausea and vomiting
- Itching
- Urinary retention
- Respiratory depression

In general, respiratory depression is a very rare complication in older children and adults when opioids are epiduraly used in the recommended doses, but this may occur hours after administration. Hence, it is recommended that all patients with epidural opioids are accurately monitored during the postoperative period, at least with pulsoxymetry.

Morphine when compared to other opioids is a much more hydro soluble substance. This is the reason for its slow uptake and the slow onset of analgesia which is delayed by at least 30 min. But epiduraly administered morphine generates a centripetal diffusion. This means that caudally administered morphine causes analgesia at the thoracic level after 30–60 min. The mean duration of its analgesic effect is approximately 8–10 h, with a maximum up to 24 h.

Dosage of Morphine (epidural):

- Single dose: 30–50 μg/kg
- Continuous infusion: 2–4 μg/kg/h

Sufentanil is a liposoluble opioid and has a rapid onset within some minutes. Sufentanil does not show any centripetal diffusion and its analgesic effect is limited to only a few segments. Therefore it is frequently used for segmental thoracic epidurals in adult patients. Sufentanil generates a high vascular uptake and reaches a plasma level which causes a certain amount of

central analgesic effects. The duration of the epidural effect is 4–6 h.

Dosage of Sufentanil (epidural):

- Single dose: 0.75 µg/kg (max 20 µg in adults)
- Continuous infusion: 0.03–0.075 µg/kg/h (children – adults)

Fentanyl is also a liposoluble opioid with a rapid onset and an effect limited to the segmental level at the point of administration. Fentanyl most frequently causes itching, which is very inconvenient for the patient. The analgesic effect of epiduraly administered fentanyl lasts only 1–2 h [29].

Dosage of fentanyl (epidural):

- Single dose: 1–2 µg/kg
- Continuous infusion: 0.25–0.5 µg/kg/h

Ketamine

Recently Ketamine and S-Ketamine have also been used as adjuvants in epidural anesthesia. For many years ketamine has been stabilized with preserving substances which are supposed to be neurotoxic. Therefore, only S-Ketamine which is free of preservatives should be used for epidural application.

Dosage S-Ketamine (epidural):

- Single dose: 0.5–1 mg/kg
- Continuous infusion: no references available

At our Centre, clonidine and morphine have proven to be most effective and therefore only these two adjuvants are used in our practice.

Additional Drugs for a Multimodal Pain Concept

Since intra- and postoperative pain therapy should be considered as a **multimodal concept**, the first dose of a non-opioid analgesic drug is administered intraoperatively. The aim of Non-Steroidal Anti Inflammatory Drugs (NSAIDs) is an anti-inflammatory effect that cannot be provided by local anesthetics and opioids alone and these have been found helpful in reducing their needed amount. It is very important, that the patient is free of pain, when he/she leaves the operating theatre. If the patient complains of pain when waking up after surgery, an intravenous analgesic drug should be administered as rescue medication. After this another top up of the local analgesic drug has to be administered before the patient is transferred to the recovery room. The intravenous rescue medication bridges the gap until the onset of the effect of local analgesic drug commences (Table 12.5).

Table 12.5 Characteristics of possible drugs for a multimodal pain concept

	Dose	Intervall	Remarks
Opioid drug			
Piritramid	0.1–0.2 mg/kg	4–6 hourly	First dose 30 min before end of surgery
Tramadol	1–2 mg/kg	3–4 hourly	
Nalbuphin	0.2 mg/kg	3–4 hourly	First dose at end of surgery
Non opioid drug			
Diclofenac	1 mg/kg	2–3 times daily	
Ibuprofen	10 mg/kg	2–3 times daily	

Since Nalbuphin is a partial antagonist, it must not be given, when morphine or any µ-receptor agonist opioid is used as an adjuvant in epidural anesthesia. Special problems in patients with repair of pectus excavatum are psychosocial rather than somatic handicaps [37]. Therefore in our experience, in the perioperative setting additional **anxiolysis** is often required. This experience has also been confirmed by another institutions that have experience in managing patients undergoing MARPE [38].

Emergence of Anesthesia

The intraoperative anesthetic management aims for extubation at the end of surgery and patients should be admitted for close postoperative monitoring to the Post Anesthesia Care Unit (PACU). In the case of endoscopic surgery gas must be completely evacuated from thorax by repeated recruitment maneuvers. During positive pressure

recruitment up to 30 mmHg for 10–20 s, the endoscopic port's valve must be opened and maintained in an underwater seal. Connecting the port to underwater can help to determine whether the entire insufflated gas is evacuated or not (Fig. 12.6). Before extubation a chest radiograph should be performed to rule out residual pneumothorax or lobar atelectasis.

Postoperative Pain Management

Regional Analgesia Catheters

The postoperative management of epidural catheters depends on the facilities of the Center where the patient is operated and nursed

Intermittent Administration of Boluses on Demand of the Patient

This is the least effective method of using an epidural catheter and should only be chosen if there is absolute no infusion device available to administer a continuous infusion of the local analgesic drug. In this case the anesthetist on duty has to be called when the patient starts to complain of pain. Due to time delays until the bolus is administered and the deferred onset time of the analgesic drug, a high number of patients are unsatisfied with this method. On the other hand it also increases the recurrent manipulation at the anti-bacterial filter rendering the danger of contamination and increasing the risks of catheter infection.

Continuous Administration of the Local Analgesic Drug

The continuous administration of the local analgesic drug is a very effective method, provided that the rate of infusion is chosen adequately. A disadvantage might be that the patient cannot adjust the pain therapy by himself to certain stress situations, such as during physiotherapy.

Patient-Controlled Analgesia (PCA) Using a Special Infusion Pump

Patient-controlled regional analgesia is the most effective and elegant way of postoperative pain management using an epidural catheter (Fig. 12.7). The experiences over many years have shown that a fairly high continuous rate together with additional boluses on demand of the patient is the most effective method. The continuous rate should be chosen high enough to provide good analgesia at rest, thus allowing the patient a fairly undisturbed sleep at night. The bolus doses allow the patient to adjust the pain therapy to stress situations as mobilization, change of dressings, physiotherapy and so on. For postoperative epidural analgesia bupivacaine 0.125 % and ropivacaine 0.2 % or levobupivacaine

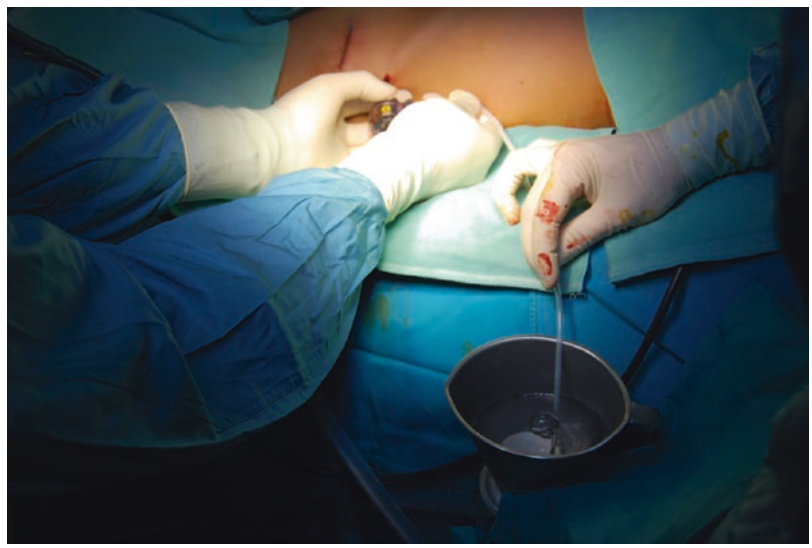

Fig. 12.6 Recruitment maneuver

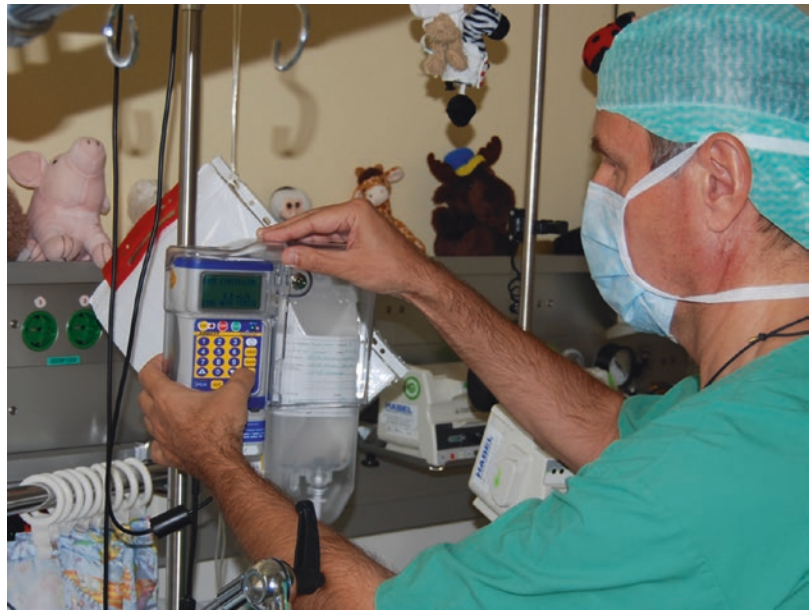

Fig. 12.7 Infusion device for patient controlled regional analgesia

0.125 % are most frequently used. At the author's institution ropivacaine 0.2 % is preferred for several reasons [39]:

- Ropivacaine 0.2 % provides excellent analgesia
- At this concentration it rarely generates a motor blockade which is very inconvenient to the awake patient
- Compared to other long acting local analgesic drugs, it has a better profile when considering its side effects, when relating to cardio- and neurotoxicity

Adjuvants are also used in the postoperative epidural analgesia. If **clonidine** was used intraoperatively, this is added to the analgesic drug reservoir in a concentration if 1 μg clonidine/ml local analgesic drug (e.g. 200 μg clonidine added to one bag of 200 ml ropivacaine 0.2 %). In the case of intraoperative use of **morphine,** this is added in a concentration of 20 μg/ml ropivacaine 0.2 %. For the postoperative continuous use of regional analgesia, very divergent dosage regimes have been recommended. Most of all in children, the suggestions are to use very low doses which have been found to provide inadequate analgesia. The following dosage regime are recommended for the patient controlled epidural analgesia delivered by a special infusion pump; a protocol that has been used over many years successfully at our Centre (Table 12.6). Above a continuous rate, boluses are allowed with limits of 4 h along with which a bolus lock out time of 15 min is applied. The regional analgesia should always be combined with a regular dose of a non-opioid analgesic drug and an opioid for rescue medication has to be available on demand of the patient. Minor side effects of regional analgesia include vomiting, pruritus, development of Horner's signs and urinary retention in the postoperative period. Vomiting and pruritus are well known side effects of epidural morphine but are reversible with reduction of the administered analgesics and addition of supplemental drugs, e.g. ondansetron for treatment of vomiting and naloxone for treatment of pruritus. The occurrence of Horner' sign is a

Table 12.6 Dosage regime for patient controlled epidural analgesia

Continuous rate	0.1 ml/kg/h (max. 8 ml/h)
Bolus	0.1 ml/kg (max. 6 ml)
Lock out time	15 min
4 h maximum	1.2–1.5 ml/kg (max. 52 ml)

well-known side effect of thoracic epidural or paravertebral infusions of local anesthetics but is in most patients well tolerated and completely revers after reduction of the local anesthetic volume.

Patient-Controlled Intravenous Analgesia (iv-PCA)

Postoperative pain relief may also be provided by patient-controlled intravenous analgesia [40]. In the literature morphine is the most commonly used opioid for iv-PCA, but also fentanyl, piritramid and tramadol have also been reported. In our Centre iv-PCA with piritramid is preferred for postoperative pain management after chest wall surgery [41, 42]. For the purpose of multimodal pain control, the first dose of a non-opioid drug (ibuprofen 10 mg/kg or diclofenac 1 mg/kg) is applied after induction of anesthesia. In case of intraoperative use of short acting opioids, such as fentanyl and remifentanil a loading dose of the long acting opioid (piritramid 0.1–0.15 mg/kg) has to be administered at least 30 min before the end of the procedure. For the postoperative pain management in children and adolescents piritramid is provided with a PCA-infusion pump using a background infusion with bolus administration and a 4 h limit (Table 12.7). In the literature the addition of a background infusion is discussed controversially. In adult patients it could be demonstrated, that the use of a background infusion clearly increases the risk of respiratory depression, most of all in the elderly population. But a meta-analysis by George concluded that the addition of a continuous background infusion is not associated with a higher incidence of respiratory events in pediatric patients [43].

Table 12.7 Dosage regime for patient controlled intravenous analgesia with piritramid

Continuous rate	20 µg/kg/h
Bolus	30 µg/kg
Lock out time	5 min
4 h maximum	0.2 mg/kg

Management and Monitoring of Patient Controlled Analgesia on the Ward

The aim of the intraoperative analgesic regime is that the patient leaves theatre absolute free of pain. In the recovery room the special infusion pump (pain manager) for postoperative regional analgesia or iv-PCA is started. After the recovery time, the patient is transferred to the ward together with his pain manager. In the authors institution special instructed and trained nurses in the ward care for these patients. All patients with a PCA pain manager have to be monitored with pulse-oxymetry for at least 24 h; whereas patients with epidural opioids or patients who receive additional sedative drugs have to be monitored for the whole period of PCA-use [44]. The nurses check in regular time intervals the vital parameters, grade of sedation, adverse effects and most of all the actual analgesia state of the patient. In the literature a number of different pain scales are recommended. Up to an age of 4 years the "Children's and Infants' Postoperative Pain Scale" (CHIPPS) is the most common used tool [45]. For older children the Smiley Scale may be used. In our Centre, for children older than 4 years of age and adolescents the revised Faces Pain Scale is used [46] whereas in adult patients a visual analogue scale (0–10) is used. All the previously mentioned pain scores rank a maximum of ten points, and are therefore easily comparable. Zero means no pain at all; ten points to the worst imaginable pain. The nurses also have to be provided with detailed written orders concerning the management of patients with regional catheters and iv-PCA. Most of all they have to know when exactly an attending and responsible anesthetist have to be informed. In our Centre, recommendations to inform the doctor on-call include:

- SaO2 below 94 (first administer oxygen)
- Pain score above 4 despite several boluses and administration of a rescue medication
- Severe PONV (Patient has vomited more than two times)
- Increasing or newly developed motor blockade
- Increasing paresthesias
- Other side effects

The responsible team of anesthetists has to provide a 24 h pain service [47]. The anesthetist on duty has to perform a ward rounds at least once a day and is if possible twice a day where all patients with patient controlled analgesia are visited. During these ward rounds, in patients with regional catheters the puncture sites are inspected and the dressings are changed if necessary. In all patients the pain scores over the last hours are checked and the pain therapy with the PCA – pain manager is adapted if necessary. In general, the regional catheter is left in place until after a stress test (continuous rate is stopped) when the patient is free of pain without demanding further bolus doses. In certain cases the regional catheter has to be removed ahead of time if:

- Local inflammation or swelling at puncture site
- Local pain at puncture site
- Massive leaking of the catheter
- High temperature after all other causes have been excluded

In these cases the tip of the catheter should be examined for bacterial colonization [48, 49]. In patients with iv-PCA the background infusion needs to be reduced as soon as possible, aiming for sole bolus administration at the second or third postoperative day. Using patient controlled analgesia for intra- and postoperative pain management leads to a high patient satisfaction, but it requires close cooperation between surgeons, anesthetists, physiotherapists and the nursing staff in the wards.

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Part II

Clinical Investigations

Preoperative Assessment of Chest Wall Deformities

13

Seth D. Goldstein and Paul M. Colombani

Preoperative Assessment of Chest Wall Deformities

Congenital anterior chest wall deformities are defects characterized by either over- or undergrowth of the cartilaginous structures of the thoracic cage. The most common of these are pectus excavatum, pectus carinatum, and Poland syndrome, all of which can present as initial disease or as recurrence after prior surgical repair. Rarer anomalies that may present in infants include sternal cleft, pentalogy of Cantrell, and Jeune's constricting thoracic dystrophy. Precise identification of the type and severity of the defect is important when evaluating patients for surgical management. Guiding principles include routine assessment of pain, body image issues, and adverse effects on cardiopulmonary function in the context of the natural development of the chest wall during childhood and puberty (Table 13.1).

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Pectus Excavatum

Pectus excavatum is the most frequently occurring chest wall anomaly and can present with a wide range of severity of sternal depression and rotation at various ages. The male to female ratio is approximately 4:1 [1]. Deformities may be narrow and deep or wide and shallow, with especially pronounced progression during puberty. Due to its characteristic external appearance, it is easily recognized and patients will often present for evaluation in childhood or even infancy, though the most common age for surgical evaluation is around the time of the adolescent growth spurt in the early teenage years [2].

Most prepubescent patients with pectus excavatum are asymptomatic, as otherwise healthy young children have significant pulmonary reserve. During adolescent growth and maturation, the thoracic cage develops increased rigidity, at which point the defect can attain physiologic significance. Most patients report at least some difficulty with shortness of breath, dyspnea on exertion, or exercise intolerance [3]. Though many patients carry a diagnosis of mild (or exercise-induced) asthma at the time of surgical evaluation, the deficit is typically manifested on pulmonary function tests as restrictive pulmonary disease [4]. Chest pain and cardiac palpitations may also contribute to exercise intolerance. Compression of the heart resulting in a decrement in cardiac function is less common but may occur in severe disease. Interestingly, mitral valve pro-

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Table 13.1 Recommendations for assessment of chest wall deformities

	Preoperative workup	Indications for surgical repair
Pectus excavatum	History and physical examination Caliper measurements Static and exercise pulmonary function tests Chest CT Echocardiography as needed	Severe exercise intolerance Body image issues Intractable pain Caliper measurement greater than 2.5 cm Haller index > 3.0 Pulmonary function test results less than 80 % of predicted for age, height, and gender Echocardiographic evidence of right heart compression
Pectus carinatum	History and physical examination Static and exercise pulmonary function tests if symptomatic Chest CT in severe instances	Failure or inability to tolerate constrictive orthotic bracing Abnormal pulmonary function tests Presentation after puberty
Poland anomaly	History and physical examination Chest CT with 3D reconstruction	Exposure of visceral organs to possible injury Patient dissatisfaction with cosmesis of breast aplasia

lapse may occur in as many as 25 % of patients. Self-conscious embarrassment regarding body image may develop during puberty, exacerbating avoidance of sports or other group activities. The psychosocial issues surrounding body image in teenagers can be significant and life-altering [5].

On physical examination, the depressed deformity is easily visualized and palpated. Even in the absence of connective tissue disorders, patients tend to have an asthenic build. The depth of the defect and presence of sternal rotation should be assessed. Thoracic kyphosis and scoliosis are commonly noted, which can lead to the rounded shoulders and "pot belly" secondary to lax rectus abdominis musculature. Most defects involve the lower end of the sternum with the inward turning of costal cartilages around ribs 4–7 at the level of the nipples. Pain is usually on the left sternal border, likely related to tension on the chondrosternal joints (Fig. 13.1).

There are a number of various caliper and CT-based anthropometric measurements that can be obtained to assess severity and guide treatment options. The simplest is with obstetrical calipers. The calipers are used to measure the distance between the posterior midline overlying the vertebral spinous process and the deepest aspect of the depressed sternum, which is compared to the distance between the same posterior point and the chest wall at the midclavicular line at that cranial-caudal level. A difference in measurements of more than 2.5 cm is indicative of a moderate to severe excavatum defect [6].

Patients with significant symptoms or deep caliper measurements should be further evaluated for surgical repair with CT imaging. A CT scan without contrast will identify the precise levels of the defect, the amount of sternal rotation, and the presence of any compressive effects on intrathoracic

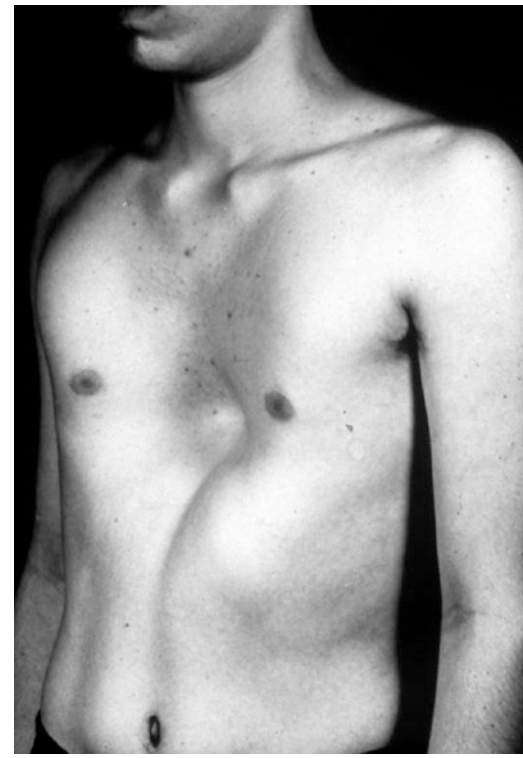

Fig. 13.1 A typical preoperative photo of pectus excavatum. Classic features noted include rounded shoulders, a sunken chest, sloped ribs, and a pot belly

structures. Additionally, this will allow calculation of the Haller index [7], which is the most commonly used measure of pectus excavatum severity. To do this, the transverse width of the chest between the ribs at the deepest level of the sternal defect is measured and divided by the anterior-posterior dimension at the same level from the anterior surface of the vertebral body to the posterior sternum, as demonstrated in Fig. 13.2. The Haller index is the quotient of this division; an index greater than 3 is considered significant, and warrants consideration for repair. Other CT-based pectus severity indices have been recently proposed and are currently being evaluated [8, 9] (Fig. 13.2).

Both static and exercise pulmonary function tests should be obtained if there is any known or suspected respiratory component of a patient's presentation. Forced vital capacity (FVC) and forced expiratory volume in 1 s (FEV1) should be compared to normal values for age, height, and gender. Exercise pulmonary testing can elicit the actual physiological effects of the disease by measuring the oxygen delivery to tissues (VO_2, L/min), which is often below normal in patients with moderate to severe defects [10].

Echocardiography can serve as an adjunct to standard diagnostic testing if a decrement in cardiac function is suspected. As previously mentioned, mitral valve prolapse is common finding in patients with pectus excavatum. A sufficiently depressed sternum can compress the right-sided chambers of the heart and interfere with diastolic filling. Any effect on filling can decrease cardiac output and is another indication for surgical correction.

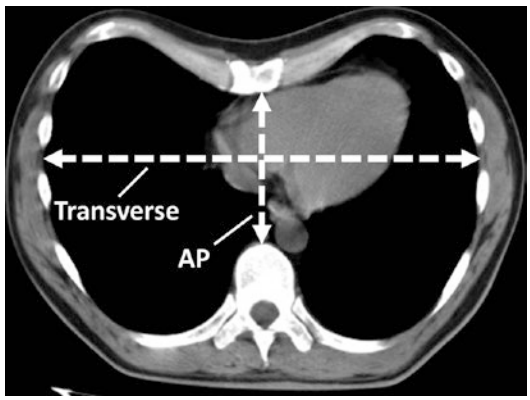

Fig. 13.2 Depiction of the Haller index calculation. The anterior-posterior (AP) distance between vertebra and sternum is divided by the transverse width of the chest at that level

Clinicians should be aware that though rare, the association with connective tissue disorders is higher than in the average population. Approximately 1 % of patients with pectus excavatum have an underlying condition such as Marfan syndrome, Ehlers-Danlos syndrome, or Loeys-Dietz syndrome. Any family history or stigmata on physical examination that indicate a connective tissue disorder should be referred for genetic, cardiac, and ophthalmologic evaluation.

Overall, subjective indications for surgical correction of pectus excavatum include physiologic compromise, psychological body image issues, and severe pain. Objective correlates are caliper measurements greater than 2.5 cm, a Haller index greater than 3.0, below-normal pulmonary function tests, or echocardiographic evidence of right heart compression.

The timing of surgery remains controversial [11]. Recent reports have suggested that pectus excavatum can be safely repaired in early childhood as young as 5 years old [12]. Nevertheless, in our opinion pectus repair before the pubertal growth spurt is likely a risk factor for recurrence and can also inhibit full development of the thorax, leading to an acquired restrictive dystrophy [13]. Our practice is to wait for patients to reach the early to middle teenage years before offering surgical repair. Typically patients undergo minimally-invasive repair with bar removal after 3 years, which helps to prevent recurrence during adolescent growth. Fortunately, true cardiopulmonary impairment appears rare before puberty in our experience. Though pectus excavatum presents along a spectrum of severity, the symptoms of exercise limitations and self-image-issues are not typically present in the vast majority of the preadolescent patients. However, younger patients with significant compromise may be candidates for earlier repair. The safety of repair at an earlier age has been established with acceptable short-term results, but long-term follow up of these children has not yet accumulated to demonstrate that the thoracic remodeling is permanent.

Pectus Carinatum

Pectus carinatum is an overgrowth of costal cartilages causing the sternum to protrude forward, either unilaterally or bilaterally. Approximately 80 % of patients are male and 25 % have a family history of a chest wall defect [14]. It tends to present during puberty rather than earlier childhood. Although this tends to be of less physiologic consequence than pectus excavatum, symptoms of dyspnea on exertion may exist and should be investigated with pulmonary function tests. The most common chief complaints are pain and body image issues. Frequent trauma and athletic injuries of the chest are also often reported, related to the sternal protrusion. Associated mitral valve prolapse has been reported even in the absence of connective tissue disorders [3].

Physical examination will reveal the overall severity of the deformity. The condition is characterized by the elevation or overgrowth of the costal cartilages causing protrusion at either the sternomanubrial joint (Pouter chest) or more inferiorly along the costochondral junctions on the gladiolus, or body of the sternum.

Patients with significant symptoms and severe defects warrant correction. Even body image issues in the absence of pain are a legitimate indication for active management. During the late school age to early teenage years, comtemporary first line therapy is constrictive orthotic bracing, which can be applied during puberty and is successful in eliminating the defect in over 80 % of cases [15, 16]. Surgery is indicated for failure of bracing or presentation during adulthood and is typically performed after the completion of the adolescent growth spurt. Open Ravitch-type repair is most commonly needed in these cases. In some instances, pectus carinatum may occur following surgical repair of pectus excavatum; these cases are typically treated successfully with bracing and observation [17].

Poland Anomaly

Patients with Poland anomaly have a unilateral underdevelopment of ipsilateral upper extremity and chest wall musculature. Absence of the pectoralis minor and the sternal portion of the pectoralis major with spared clavicular portion is characteristic, and syndactyly is commonly associated. The right side is more often affected than the left. Aplasia or hypoplasia of the breast, nipple, and costal cartilages may occur.

On physical examination, most patients have relatively normal use of their arm and shoulder. Chest wall asymmetry is usually obvious in terms of the features described above. The absence or presence of an ipsilateral latissimus dorsi muscle is of importance. The stage of breast development in females should be additionally noted.

If additional diagnostic testing is needed, it is often CT to precisely identify the number and level of ribs involved as well as the orientation of underlying structures. This assists surgical planning and may be facilitated by three-dimensional reconstruction.

The most important consideration for surgical management of Poland anomaly is protection of the lung and potentially heart from injury when there is significant loss of bony protection of the thoracic cage. Surgical repair can be undertaken to protect these organs as well as to provide a cosmetic repair, particularly in females. Surgery should typically be delayed until patients have completed their teenage growth. Autologous rib grafts and polypropylene mesh can be used to bridge the gaps where ribs are absent. A latissimus dorsi flap can be used to cover the chest wall repair and serve as a pectoralis muscle. This provides an excellent cosmetic result and protects underlying visceral organs, but should be delayed in females until breast development is complete.

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Index for Measurement of Pectus Excavatum

14

Shawn D. St. Peter

Pectus excavatum, as a defect wherein the central chest is displaced posteriorly relative to the remainder of the anterior chest wall, has been subjected to quantification to place objective value to the degree of the defect. The initial measure proposed, which became the standard for the past few decades was the Haller index (HI). The HI was initially described in 1987 [1]. This metric was defined as the ratio of the distance between the anterior spine and posterior sternum to the widest transverse diameter of the chest. The measurement is typically derived from computed tomography (CT). In general, the deep point at the bottom of the sternum is chosen as the location to measure the denominator (Fig. 14.1). In other words, the number means the number times the chest is wider than it is deep at the deepest point of the sternal depression.

In the initial description, the authors documented a width to depth ratio of 3.25 as a discriminator to define the patient as having a significant enough pectus excavatum defect to be a potential candidate for repair. The number was established by utilizing a group of normal controls compared to pectus excavatum patients who underwent repair. The authors found that the control patients had an index of <3.25 as opposed to the pectus patients. Of course physical limitations secondary to the defect, psychosocial disposition and other variables are utilized in making this decision, it has been generally held that surgical candidacy revolves around an HI greater than or equal to 3.25 [1–3]. However, we remain cognizant of the fact that this number is supposed to simply assess the degree of severity anatomically. There may not be a substantial correlation between physical limitation and the HI. While it would be intuitive that patients with a deeper defect and a higher HI should be more difficult repair or elevate with a bar. This should result in longer operative time and more post-operative pain. However, in a series of 271 pectus excavatum repairs with bar placement, there was no correlation with operative time, duration of hospitalization, or bar infection [4].

More importantly than whether the HI carries clinically meaningful prognostic value, it may not adequately define the presence or absence of disease. There have been little data subsequently generated to validate the accuracy of the HI in separating those patients with a pectus deformity from patients with normal chest contour. This is important since the HI has been used as a diagnostic tool differentiating severe enough disease to warrant repair from those who may be observed without intervention.

The initial report utilized 19 controls, of which four were under 6 years of age, and 33 patients who underwent correction [1]. Interestingly, the HI was clinically incorporated with the

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aforementioned threshold value without further studies to validate the diagnostic accuracy of the HI. Our group recently investigated the accuracy of the HI in separating pectus patients from normal controls [5]. For this thoracic CT scans were used from patients who had undergone pectus excavatum repair and a cohort of normal controls. Pectus patients underwent repair upon the agreement of the operating surgeon, patient and family that the defect was severe enough to mandate repair and there would be a substantial improvement from bar placement. Every patient who had undergone bar placement at our institution with a retrievable chest CT scan was used. The aim was to establish a comprehensive cohort of controls representative of all children in the age range of those evaluated for correction. For this ten males and ten females were recruited from each year of age between 8 and 18 who had undergone a thoracic CT scan for an indication other than pectus excavatum. This gave a 20 8-year olds, 20 9-years olds and so on to create a population of 220 patients with a normal chest. Patients who had been evaluated for pectus excavatum or had this as a diagnosis within their medical record could not be considered for the control group. Each CT scan was reviewed and both correction and Haller indices were calculated using the Synapse® Radiographic Information System (Fujifilm Medical Systems, Stamford, CT). The ruler tool was used by drawing lines in the appropriate positions which generates the length of the line in millimeters. The most inferior image of the sternum was used to calculate the indices. The Haller index was calculated in a standard fashion (Fig. 14.1).

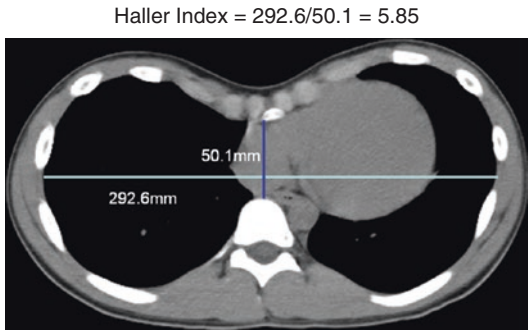


Fig. 14.1 Demonstration of the calculation for the HI

The results were a bit disturbing in that there were several pectus patients whose HI was low enough well within the controls and control patients who had a high enough HI to be will

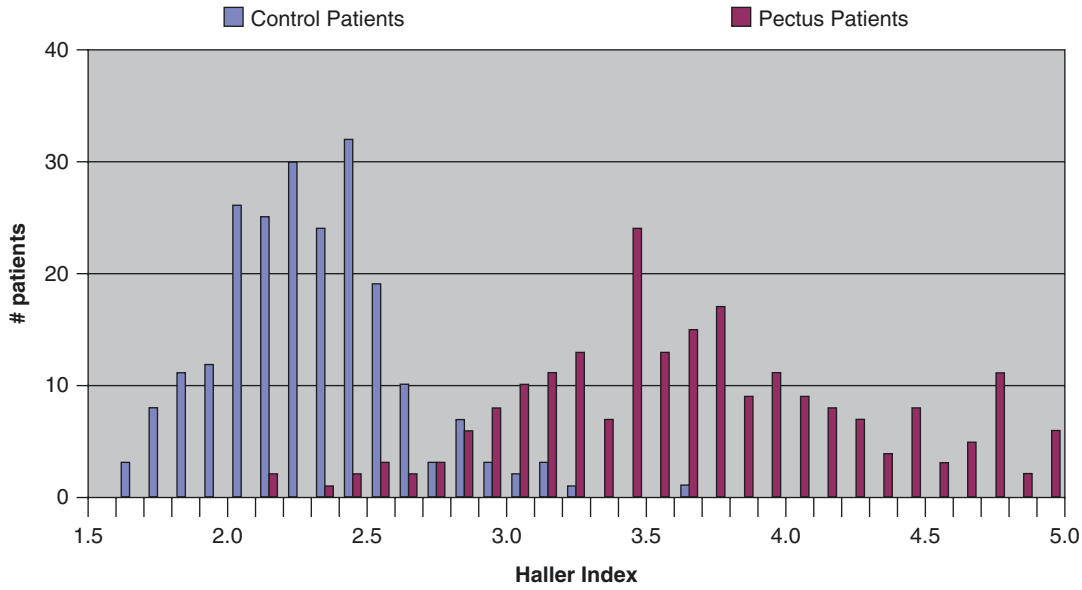


Fig. 14.2 Histogram for the distribution of HI plotting pectus patients with normal controls. In order to demonstrate both curves for pectus and control patients for HI, those with an HI over 5.5 are not displayed as this end of the curve is long tail of sporadic outliers

within the pectus patients (Fig. 14.2). In total 47.8 % of the total population of both groups fell between the highest control HI and the lowest pectus HI. This is obviously an unacceptable amount of overlap for a measurement that we are to be using to separate these two groups – those who would benefit from a bar repair from those who would not. Consider that abnormal physiologic parameters for any disease process are normally determined by establishing a normal distribution and using two standard deviations above or below the mean which leaves 2.5 % of the assumed normal population who may meet abnormal criteria by chance in either direction. The overlap between disease and control groups is large, but in addition, the HI of 3.25 as a surgical threshold also failed as we found 19 % of the pectus patients who had undergone repair possessed an HI under this value. These are patients who clearly have the defect as well as the typical clinical manifestations of the disease but the HI does not represent the severity of the defect. An example of such a patient is demonstrated in Fig. 14.3. On the flip side, we measured a control patient with a HI of 3.75. This case is depicted in Fig. 14.4 where there is clearly no central thoracic depression. In fact, the sternum is the most anterior point on this patient's chest. These cases demonstrate the limitation of the HI and the reason it should be abandoned as a clinical tool. The measure depends heavily on the width of the chest as the numerator of a simple fraction. Therefore, the patient with a deep chest that is narrow side-to-side can have an impressively severe pectus excavatum defect, which when assessed by the HI, generates a number that is no where near the historically chosen value of 3.25.

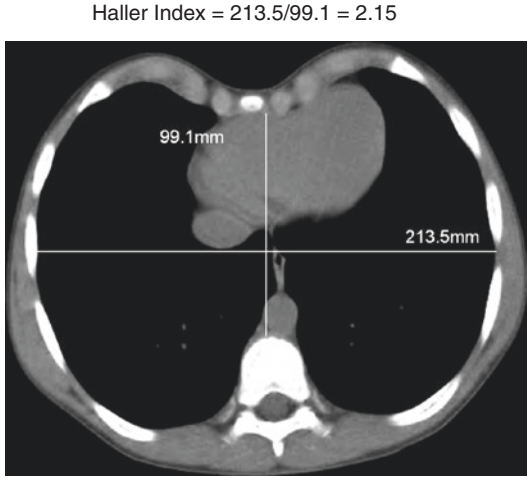


Fig. 14.3 CT image of the pectus patient with the lowest HI

In order to overcome the inadequacy of the Haller index and provide a reliable tool that will precisely quantify the severity of the defect, we developed the pectus correction index [5]. This equation was developed to remove width from the process. Instead the deepest point of the sternum is compared to the most anterior protrusion of the chest wall. The pectus correction index (PCI) requires drawing a horizontal line across the anterior spine. Then two distances are measured, the minimum distance between the posterior sternum and the anterior spine as is utilized for the HI, and the maximum distance between the line placed on the anterior spine and the inner margin of the most anterior portion of the chest on the same CT slice chosen for the minimum distance. The difference between the two lines is simply the amount of defect the patient has in their chest. If this difference between the measurements is then divided by the maximum prominence of the chest (the longer measurement) and multiplied by 100, it generates the percentage of chest depth the patient is missing centrally (Fig. 14.5).

Since current standard for pectus excavatum repair is to place a bar under the sternum which otherwise rest outside the ribcage, the repair is simply a bridge created across the two anteriorly protruding areas of ribcage on each side of the sternal portion where the bar crosses beneath. As a result the deep portion is brought to the level of the anterior chest regardless of the overall depth or width of the chest. Therefore the PCI also provides the number which represents the percentage of chest depth to be corrected by bar placement. This is the percentage of chest to be regained by bar placement.

As opposed to the HI, the PCI cannot possibly provide a number that deviates much from zero in patients who do not have a pectus deformity. This is because the sternum will be near

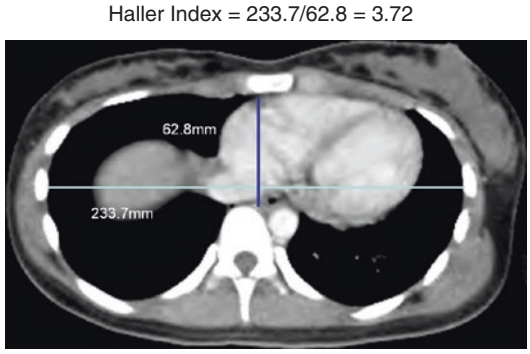


Fig. 14.4 CT image of the normal patient with the highest HI

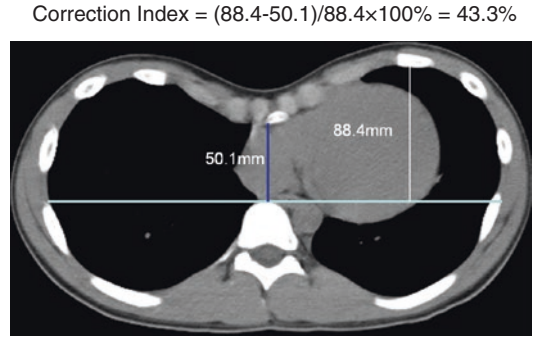
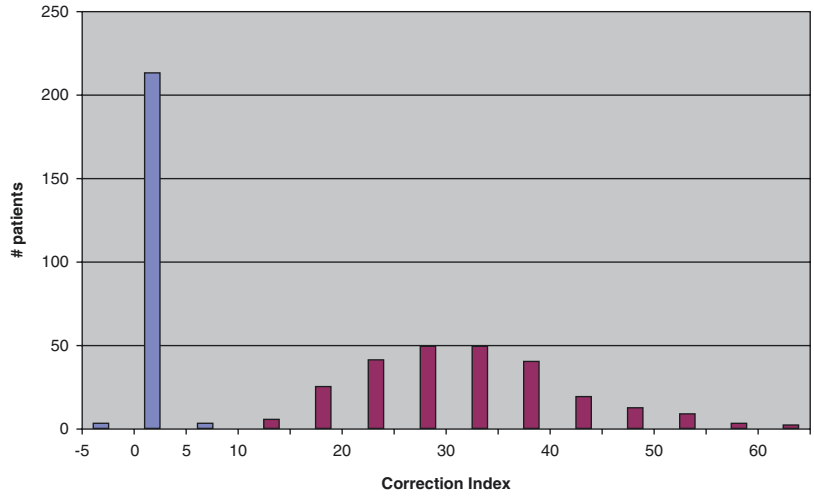


Fig. 14.5 Demonstration of the calculation for the PCI

Fig. 14.6 Histogram for the distribution of PCI plotting pectus patients with normal controls



the anterior margin of the chest. Once a patient has a PCI of 10 or 10 % of the chest depth is lost at the inferior margin of the sternum, then by definition, they have a pectus excavatum. In order to test this theory, measured the PCI on the aforementioned study population of 252 patients who had undergone bar placement and 220 normal controls was also measured. It was found that all the controls were measured around zero and the smallest PCI found on any of the pectus patients was 16. When the pectus patients were plotted on the same horizontal axis as the controls, there was absolutely no overlap found in the populations (Fig. 14.6). Therefore the PCI can be expected to provide no false positive or negative due to the nature of the measurement. The only way a patient can generate a number over 15 is if the distance from spine to sternum is 15 % shorter than the surrounding anterior chest wall, which will be a clinically obvious pectus excavatum. Since completing this validating study of the PCI we now utilize a PCI of 15 as a threshold for consideration of operative correction. The PCI has been remarkably useful as over 100 bar repairs have been performed at our Centre since the completion of that study, with no bar being placed in patients with a PCI under 15.

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Anthrometric Index for *Pectus Excavatum*

15

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Abbreviations

AI	Anthropometric Index for *pectus excavatum*
HI	The Haller Index
LVI	Lower Vertebral Index
PEX	Pectus excavatum

Introduction

Congenital deformities of the thoracic wall have been the object of medical scrutiny since the 15th century [11]. Among such deformities *pectus excavatum* (PEX) is the most common and is characterized by a strongly depressed sternum along the spinal cord [33]. Scoliosis may occur in 21.5 % of PEX patients [35], a combination that adversely affects the quality of life of these patients.

The literature contains several theories for the etiology of PEX: [shortening of the diaphragm tendon (Brodkin 1953); sternum suffering abnormal growth (Ebstein 1882); flaccidity in the presternum region (Kleinschmidt and Kirschner 1944) cited by Trench and Saad [33], [congenial deviation of the heart leftwards [37], and [abnormal growth of costa cartilage [13]. PEX deformity can be classified as symmetric and asymmetric [3]. Usually, when PEX is asymmetric, it is the right side that is more greatly affected by the deformity.

Indication to PEX treatment may be grounded on the following aspects: functional [21, 22], aesthetic [4], psychological [6] and quality of life [20]. As for its treatment PEX may either be conservative or surgical.

Among the authors who advocate conservative treatment is Haje et al. [9] who proposed the use of orthoses to compress the rib-cage in an attempt to remodel its architecture into a more normal configuration. Haecker [8], made use of a suction device that when applied over the anterior chest wall intermittently for a period of up to 36 months, corrects the depressed sternum.

The surgical proposals in turn follow two main venues:

I. surgeries without any bone placement or use of filling materials like silicone, used to restore the external configuration of the chest closest to normal as possible [7, 15, 23];
II. surgeries with bone replacement that can be divided into:
    1. Surgical treatment with bone placement without the use of prosthetics. In this technique anatomical structures are used to anchor the sternum into its new position

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once it has been detached from costal cartilage [14, 27, 34, 37];
2. Surgical treatment with bone replacement and prosthetics, when the support of the sternum is achieved resorting to prosthetics, which can be made of:
   (a) Metal [31, 1, 26];
   (b) Polypropylene [31];
   (c) Bio-absorbable material [24].

Pectus excavatum assessments have been made along the years either in a subjective manner resorting to criteria like surgical scars, persistence, and relapse of the deformity [18] or objectively, which include:

(a) Clinical evaluations in which the focus is to characterize the external configuration of the chest, as can be verified in many studies: [12, 16, 17, 19, 29, 30].
(b) Evaluations of the defect by means of imagery that show the internal configuration (bone configuration) of the chest. Didactically, these may be divided into:
   1. Methods that resort to chest X-rays, like in Welch [36], Backer et al. [2], and Derveaux et al. [5].
   2. Methods that make use of computerized tomography, like in Haller et al. [10], Nakahara et al. [25], and Matsui et al. [24].

We shall now describe some such measuring methods adopted to establish thoracic architecture so that we can comprehend the context in which the AI for PEX was created.

Knutson, in 1967, as well as Haller in 1978, purported to create a method to objectively evaluate PEX. For such they set out to develop devices that could draw the configuration of the anterior thoracic wall in a axial section so that the dimension of the deformity could be characterized graphically. In both techniques the patient remained in a horizontal supine position on a flat table while a probe ran along the defective chest. The topography of the chest was mechanically transmitted to a screen where the defect was recorded in an axial profile. In this manner it was possible to collect objective quantification of the deformity as well as assess the results after the treatment.

Hümmer and Willital in 1984 (Fig. 15.1) developed a ratio index between the external sagittal diameter at the level of xiphoid process and the external sagital diameter at the level of sternum furculum. For these authors normality was between 115 and 145 as long as the measurements were taken at exhaling. This one-dimensional index characterized the approach or distancing of the sternum to the spinal cord, and the measurements were obtained by means of a pelvimeter.

In 1985 Horst et al. worked on measurements obtained from Moiré's topography based on three-dimensional optics [16]. The images of

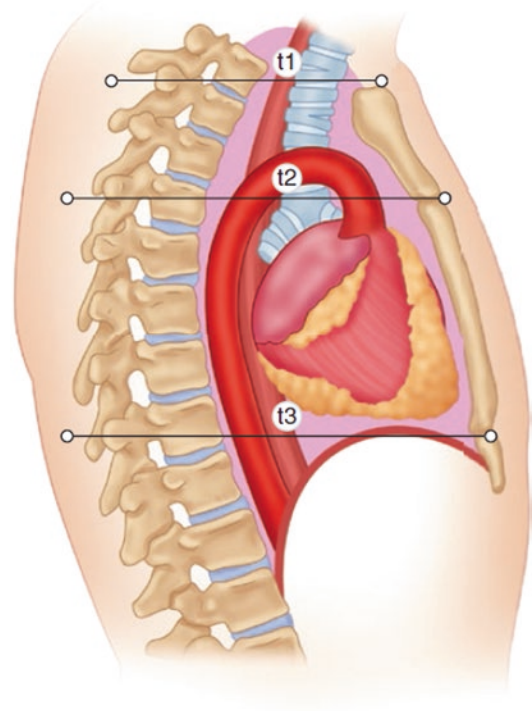

Fig. 15.1 Representation of the measures used by Hümmer and Willital in 1984. ($t_{1=}$ sagittal measurement of the sternum furculum from the vertebral column at the same level). ($t_{2\,=}$ sagittal measurement from the manubrium to the vertebral column at the same level). ($t_{3\,=}$ sagittal measurement form the xiphoid appendix to the vertebral column at the same level). ($t_3/t_{1\,=}$ index of the chest in funnel)

deformed patients made with the application of Moiré stripes produced patterns that were converted mathematically in profiles, and these allowed for the creation of two funneling indexes, I and II. The former was related to the sagittal profile (sternum depression in relation to the manubrium and navel) while the latter related to the axial profile (sternum depression in relation to the right and left anterior chest wall).

Concerned with an objective measurement Welch, in 1958, compiled an index obtained from the X-ray of the chest profile, which was determined as being the quotient between the sagittal diameter between the posterior plate of the sternum and the anterior portion of the vertebral body, at the level of thoracic vertebra 9, and the sagittal diameter between the anterior plate of the sternum and the vertebral spinous process at the level of thoracic vertebra 3. By means of this index, he classified PEX as light (0.75–0.5), moderate (0.5–0.3) and severe when the value was under 0.3 (Fig. 15.2).

Backer et al. [2] and Derveaux et al. [5] (Fig. 15.3), also investigated the indexes obtained from X-rays of the chest in profile. Those studies produced the Lower Vertebral Index (LVI), which is the ratio between the sagittal diameter of the vertebral body and the sagittal thoracic diameter in the posterior portion of the vertebral body in relation to the posterior plate of the sternum at the distal third of the sternum or at the deepest point of the deformity. According to the authors, this index also allows for objective evaluation which enables the identification of PEX and analysis of the results after treatment.

Backer et al. [2] also studied the frontal-sagittal index obtained from X-ray measurements taken of the posterior-anterior profile of the thorax and defined as the ratio between: (B1), the lowest sagittal diameter of the posterior plate of the sternum in relation to the anterior portion of the thoracic column (X-ray of the thoracic profile), and (B2), the largest transversal diameter (latero-lateral) obtained from the posterior-anterior thorax (Fig. 15.4). It is important to point out that index is the inverse of that of Haller which will be mentioned subsequently, and which is one of the main forms of objectively evaluating PEX.

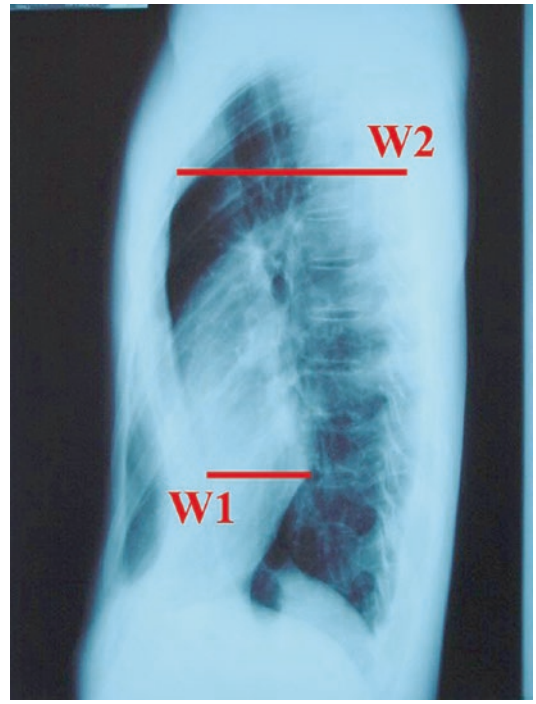

Fig. 15.2 Measurements for the calculation of the Welch index = W1/W2 (*W1* = Distance between the posterior plate of the sternum and the anterior portion of the vertebral body at the level of thoracic vertebra 9. *W2* = the distance between the anterior plate of the sternum and the vertebral spinous process at the level thoracic vertebra 3 [36] (**Source:** [28])

Haller et al. [10] (Fig. 15.5) developed the index that carries his name, the Haller Index (HI) which is arguably the objective evaluation that is most widely used by authors who investigate PEX. The index is dependent on a chest computer tomography which allows for the collection of the necessary measures for its calculation. HI is the ratio between the maximum latero-lateral distance from pleura to pleura and the minimum anterior-posterior distance of the posterior plate of the sternum in relation to the anterior portion of the vertebral body, at either the highest level of the deformity or the lower third of the sternum. According to the authors the index enables screening of patients for surgery or not, or determines their normality. The cut point for such screening is 3.25. In 2005 we had the opportunity to work with HI comparing it to the LVI and the AI for PEX. We reached a cut point value

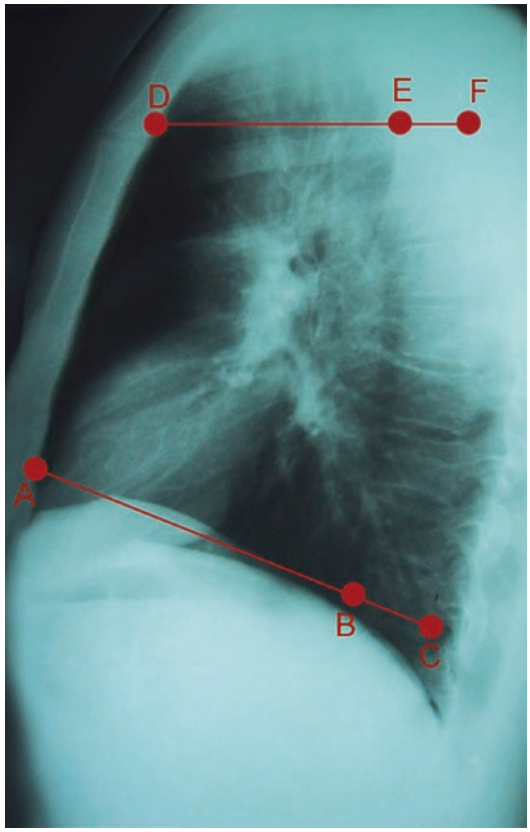

Fig. 15.3 Measurements for the calculation of the Lower Vertebral Index = BC/AC. *BC* = Sagittal diameter of the vertebral body at the distal third of the sternum or at the deepest point of the deformity. *AC* = thoracic sagittal diameter of the posterior portion of the vertebral body in relation to the posterior plate of the sternum [5] (**Source:** [28])

of 3.1 among patients who underwent surgery to correct PEX and normal individuals, which was very close to that reported by Haller in the original publication.

Nakahara et al. [25] examined measurements of tomographic sections that allow for the assessment of PEX patients prior to and after surgical treatment, comparing them to subjective impressions. The measures (Fig. 15.6) that were

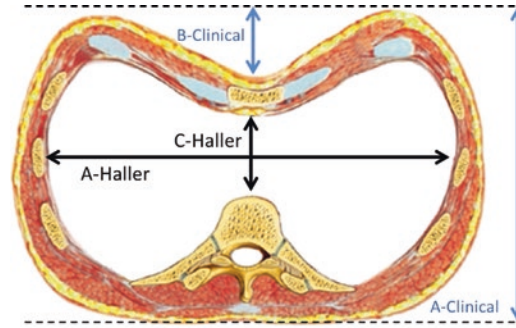

Fig. 15.5 Illustration of an axial section of the thorax related to the Anthropometric Index measurements for *pectus excavatum* (B-clinical/A-clinical) and those of the Haller Index (A-Haller/C-Haller). *A-clinical* = maximum anterior-posterior external distance; *B-clinical* = highest depth of the defect; *A-Haller* = maximum latero-lateral distance; *C-Haller* = minimum anteroposterior distance of the posterior plate of the sternum relative to the anterior portion of the vertebral body. *Both indexes calculated from the level of either where the deformity was the deepest or the distal third of the sternum* (**Source**: [29, 30])

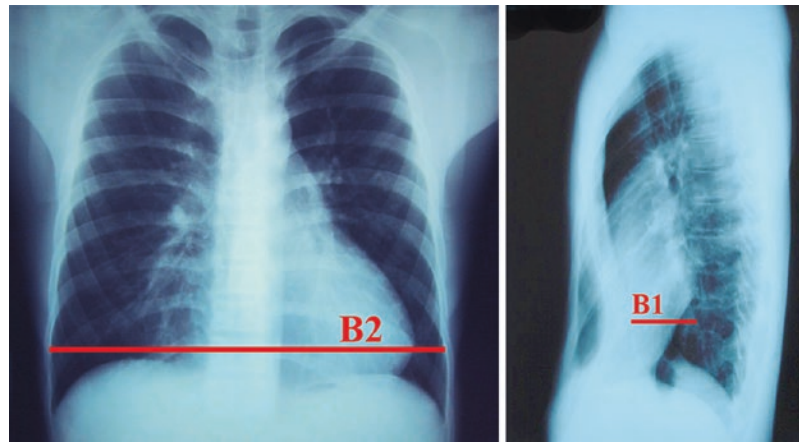

Fig.15.4 Representation of the measures obtained from chest X-rays intended for the calculation of the frontal-sagittal index (B1/B2) [2] (**Source**: [28])

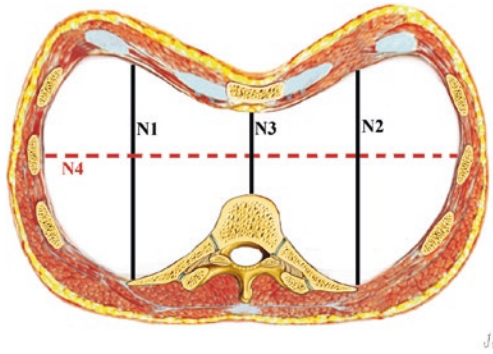

Fig. 15.6 Measurements collected from a tomographic section at the deepest point of deformity, Nakahara et al. [25]. Degree of depression (N2/N3); degree of asymmetry (N1/N2); degree of flattening (N4/N2) [25] (**Source:** [28])

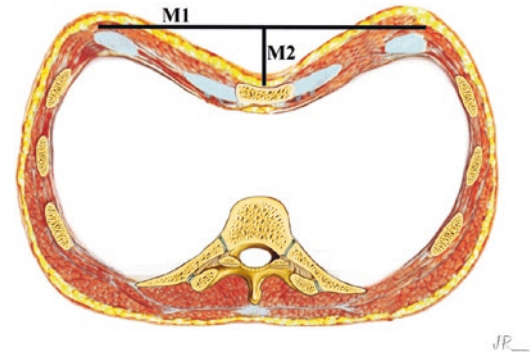

Fig. 15.7 Illustration demonstrating the obtained measures in the tomographic section at the level of deepest depression made by Matsui et al. [24]. Retraction Index = M2/M1 (**Source:** [28])

obtained show the highest deformity and with that the authors were able to determine:

(a) Degree of depression: N2/N3 [ratio between the maximum internal anterior-posterior distance of the left hemithorax (N2) and the least anteroposterior distance of the anterior portion of the vertebral body relative to the posterior portion of the sternum plate (N3)].
(b) Degree of asymmetry: N1/N2 [ratio between the maximum anterior – posterior internal distance of the right hemithorax (N1) and the maximum anterior-posterior internal distance to the left hemithorax (N2)]
(c) Degree of flattening: N4/N2 [ratio between the highest latero-lateral internal distance (N4) and the maximum internal anteroposterior distance to the left hemithorax (N2)].

Matsui et al. [24] quantified the results of treatment in PEX patients by means of measurements collected from tomographic sections at the highest level of deformity, and compared them to those collected after surgery. By doing so they created yet another index dubbed the Retraction Index, the ratio between M2 and M1 (Fig. 15.7), where M1 is the measure between the two points at the beginning of the depression in the right and left hemitorax not considering the soft tissue, and M2 is the maximum depth of the deformity.

Anthropometric Index (AI) for *Pectus Excavatum*

The team of thoracic surgery of the Hospital das Clínicas of the Faculdade de Medicina da Universidade de São Paulo, following the line of thought of the authors above in that an objective evaluation is the best manner in which to assess results, have used the Anthropometric Index (AI) for *Pectus Excavatum*. Thus, a methodology was conceived, one that would allow for the diagnosis and follow-up treatment, that would be easy to carry out (even in the ambulatory), and achieved with the use of simple instrumentation. Also, it had to be accessible and free of the need to resort to imagery (which would eliminate the need of irradiation), not be affected by the environment, and finally, be of low cost [29, 30].

The calculation of AI is based on the ratio between measures B and A. Both are obtained with the patient in supine position, on a level table parallel to the floor and during deep inhaling. The index was established based on an observational study including 20 PEX patients and 30 individuals with normal thoracic morphology.

Measure A, or maximum anteroposterior distance (Fig. 15.8) is the highest anteroposterior distance from the tangential coronal plane relative to the dorsal region and from the tangential coronal plan to the highest point of the anterior thoracic wall. When the plane of the ruler over

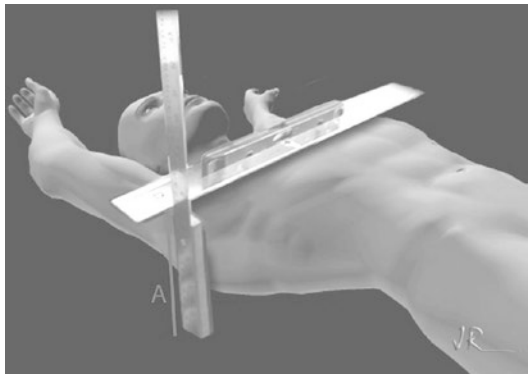

Fig. 15.8 Measure A: maximum anteroposterior measurement at either the level of deepest deformity or distal third of the sternum (**Source**: [29, 30])

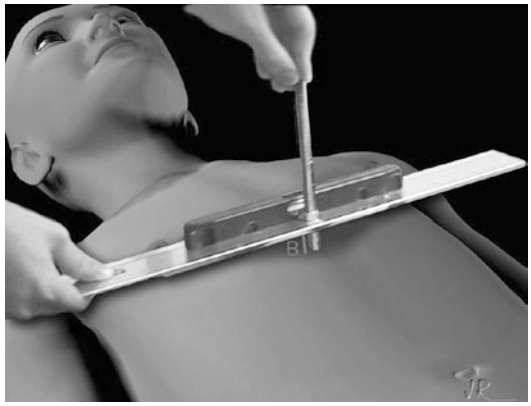

Fig. 15.9 Measure of the maximum deformity. Distance between the highest point of the anterior chest wall and the deepest point of the pre-sternum region at the deepest point of the deformity or the distal third of the sternum (**Source**: [29, 30])

the chest is parallel to the dorsal plane the distances are recorded at the level of deepest deformity or at the distal third of the sternum in the case of individuals without anterior thoracic deformity. The horizontal ruler is fitted with a level so that with the aid of a vertical measuring the highest point of the anterior thoracic wall can be gauged, be it to the right or left, determining the maximum sagittal distance.

Measure B, or the maximum depth of the deformity (Fig. 15.9), is the distance between the tangential plane to the highest point of the anterior chest wall and the plane of the deepest point of the pre-sternal region at the level of the greater deformity or distal third of the sternum. Both planes must be parallel to each other. The horizontal ruler is fitted with a level and an intersecting vertical measuring shaft that runs through an orifice at its center so that the highest point of the anterior thoracic wall can be gauged, are it to the right or left. With the use of the measuring shaft the maximum depth of the deformity can then be determined.

At the time AI was developed it was compared to the IH and LVI and (Figs. 15.10 and 15.11), which are largely accepted indexes by those who investigate PEX. The comparative analysis that followed showed that there is significant correlation between the three [28].

In regards to sensibility and specificity AI proved to have similarities observed in the two consecrated indexes (Figs. 15.12, 15.13, and 15.14) considering the following cut point between PEX patients and normal individuals; for AI the cut point was 0.12, for HI it was 3.1, and for LVI it was 0.25. There was accuracy of 80 %, 85 %, and 77.28 % respectively.

The team of thoracic surgery of the Hospital das Clínicas of the Faculdade de Medicina of the Universidade de São Paulo has given preference for the adoption of the AI grounded on the following arguments: (a) the methodology contains sensitivity and specificity that are comparable to those of consecrated indexes in medical literature; (b) AI considers soft parts and therefore leads the investigator to the real configuration of the chest; (c) there is no risk to the patient; (d) it is of low cost; (e) AI measurements can be collected readily in the ambulatory; (f) easiest way to measure the surgical results, (g) there is no environmental interference. We are well aware that this method may suffer some alteration, even in those individuals whose thorax is morphologically normal. This group of investigators thus continues to examine such variations and these shall be the object of future studies following the same line of research.

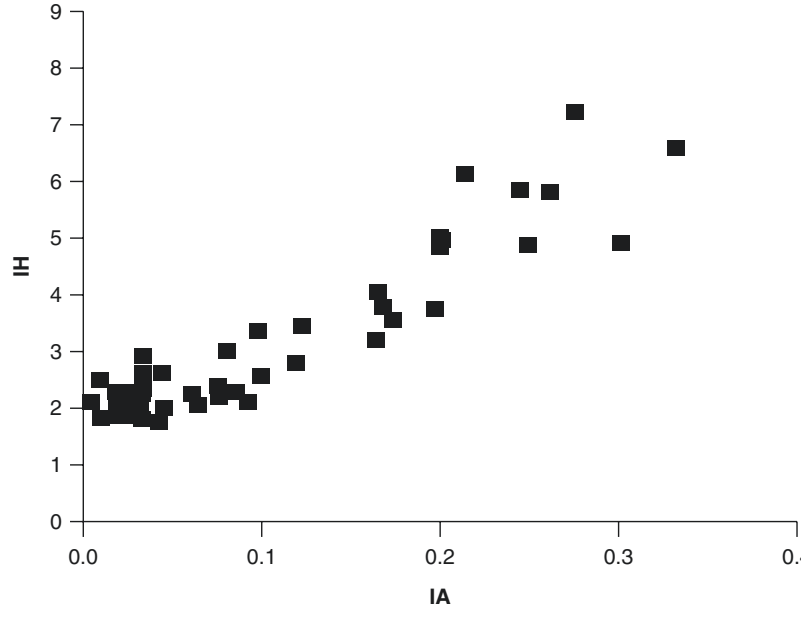

Fig. 15.10 Correlation of the AI (Anthropometric index for *pectus excavatum*) and the HI (Haller Index). Spearman correlation test: p < 0,001. IA = AI and HI = IH (**Source:** [28])

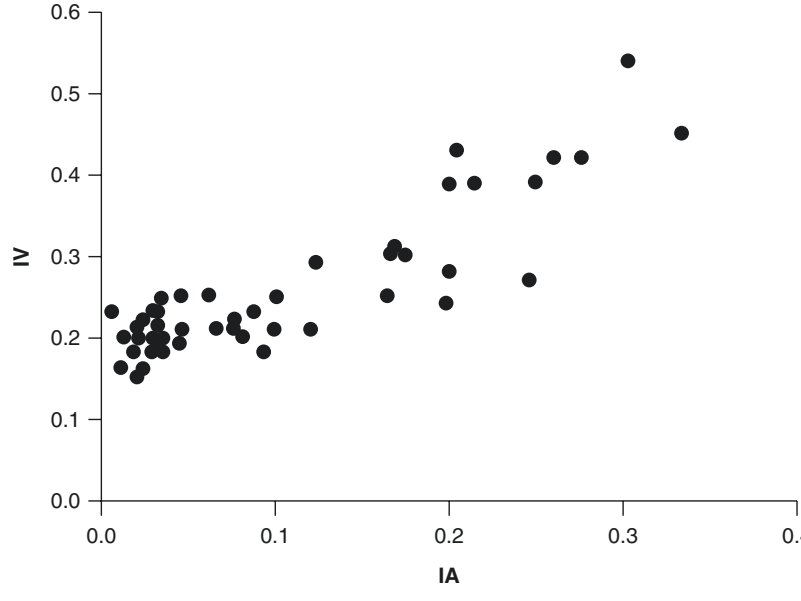

Fig. 15.11 Correlation of the AI (Anthropometric Index for *pectus excavatum*) and the LVI (Lower Vertebral Index). Spearman correlation test p < 0,001. IA = AI and IV = LVI (**Source:** [28])

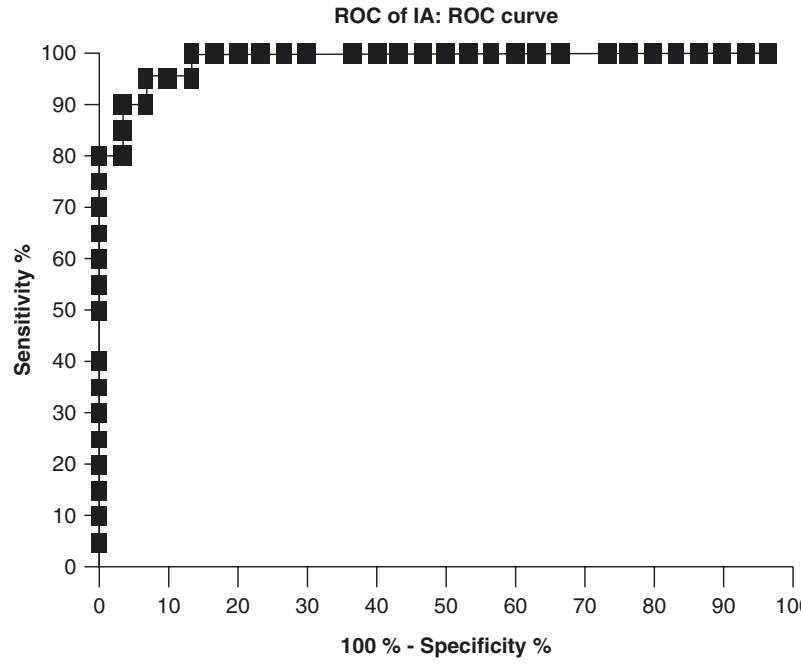

Fig. 15.12 Characteristics of the operational curve of AI (Anthropometric Index). Area under the curve = 0.99, standard error = 0.01 $p < 0.001$. IA = AI (**Source:** [28])

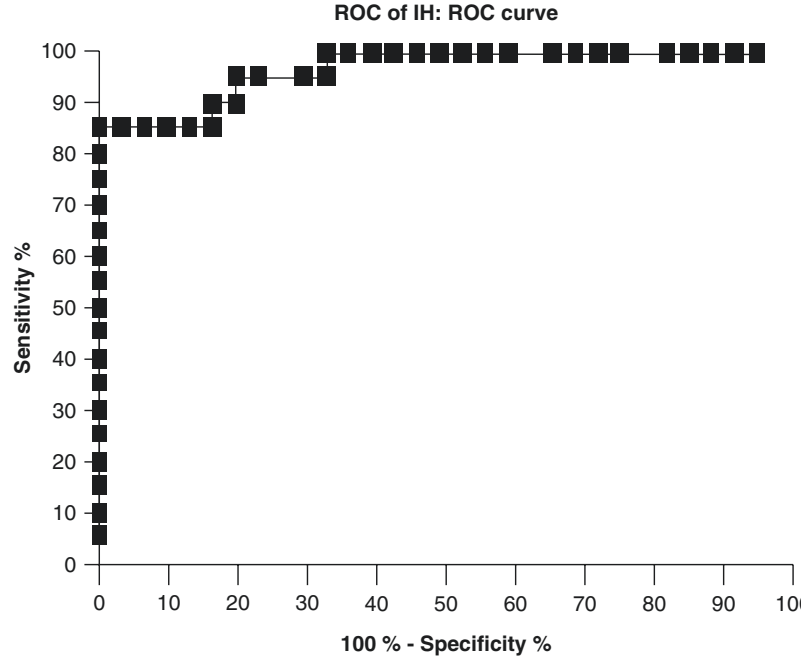

Fig. 15.13 Characteristics of the operational curve of HI (The Haller Index). Area under the curve = 0.96, standard error = 0.02 $p < 0.001$. IH = HI (**Source:** [28])

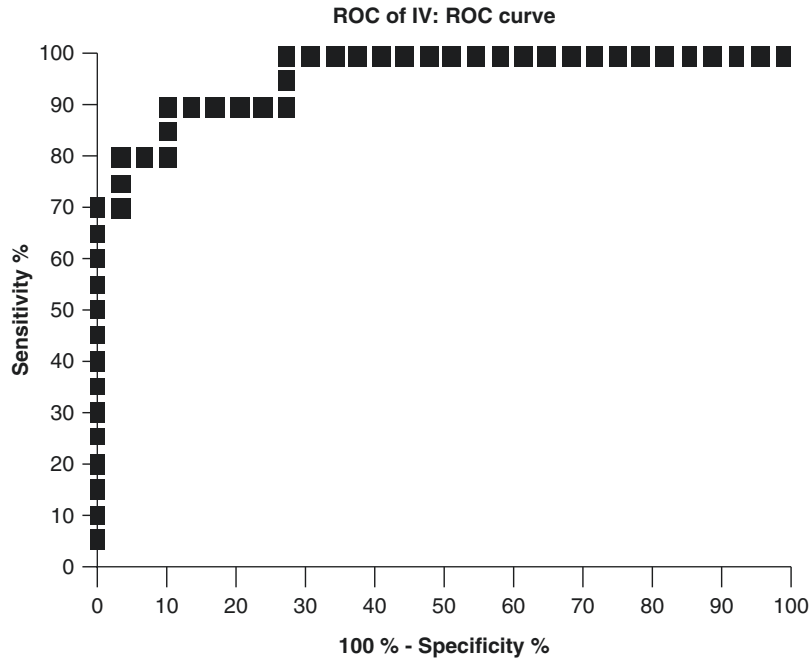

Fig. 15.14 Characteristics of the operational curve of LVI (Lower Vertebral Index). Area under the curve = 0.96, standard error = 0.02 $p < 0.001$. IV = LVI (**Source:** [28])

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Spiral Computed Tomography

16

Erich Sorantin and Sabine Weissensteiner

Principle of Computed Tomography

X-ray computed tomography (CT) is a medical imaging method, where the patient is placed on the CT table and an x-ray tube rotates around the body and sends x-rays through the body. Those x-rays experience a varying attenuation along their way depending on the body part they are transversing. Based on the mathematical algorithm of "Backprojection" a cross sectional image is computed – hence the name Computed Tomography (Fig. 16.1). The unit of the attenuation coefficients is called "Hounsfield Units" (HU) which range from −1024 to 3071. There are two fixed points of that scale: water equals to 0 HU and air to −1024 HU. Lower values usually represent tissues with lower densities e.g. fat is less dense that water and therefore has a range from −80HU to −120HU.

Historically images were generated in the axial (transverse) plane, perpendicular to the long axis of the body but modern scanners allow to compute reformations in various planes or even as volumetric (3D) representations of structures.

In the beginning of the CT era one row of detectors acquired a single row of data, after which the table had to moved and the next rotation was done ("Step-Rotate-Step"). In the mid 1990's CT couch could be moved continuously during tube rotation – thus helical or spiral scanning came into existance. This revolutionary technique utilizes two slip rings on the CT equipment: one for power supply and other for data reading – consecutively no cables were necessary and the tube could rotate continuously. Therefore another synonym for helical CT is slip ring CT.

At the end of the 1990's more detector rows were combined for larger detectors thus a new generation of CT machines was launched: the Multi Detector CT (MDCT).

In the last years there was a dramatic increase in the numbers of detector rows and therefore today CT machines with 4, 8, 16, 32, 40, 64, 128 up to 320 slices are used [1]. These modern CT scanners employ multiple rows of detector arrays allowing rapid scanning and wider scan coverage. Multiple detector rows along the Z axis (longitudinal axis of the patient, i.e., head to toe) permit simultaneous scanning of more than one slice.

All new CT systems have multiple detectors with a single or dual x-ray source, and a number of new dose reduction tools have become available commercially [2, 3]. Moreover the "Backprojection-Algorithm" for image computation was replaced by statistical reconstruction, which enables dose savings up to 50 %.

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Automated Exposure Control (AEC)

For dose saving, CT scanners are equipped with automatic exposure control, which dynamically controls the required tube current according to the user settings. Similar to AEC in conventional x-rays, CT AEC automatically selects scan mA on the basis of patient attenuation estimated from scout views (Fig. 16.2). In clinical practice, the mA automatic selection process is usually based on a user-specified acceptable image noise level, since noise is one of the major markers for image quality.

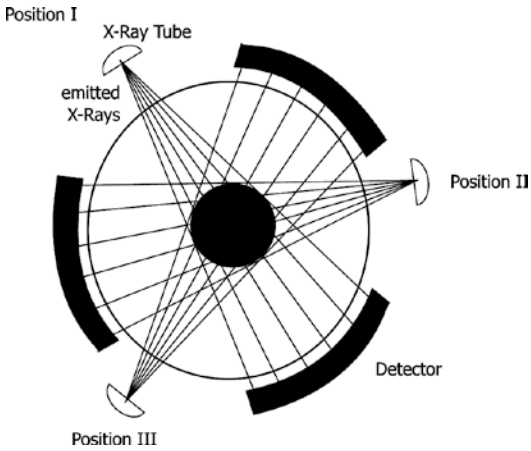

Fig. 16.1 Scheme of CT – an x-ray tube rotates around the body; emitted x-rays are attenuated in the body. To the sake of simplicity only three positions are displayed. Based on "Back Projection" (similar to GPS triangulation) actually up to 4000 projections are used for image generation (Modified from Sorantin et al. [25])

Therefore with AEC it should be possible to keep the image noise constant through the whole scan area. The software adapts the dose according attenuation e.g. in chest CT increasing the dose at the shoulder girdle, reducing it in the mediastinum and increasing it again in the lower chest aperture. This can be achieved either according to the absorption profile of the scout-view or by synchronous image noise computation during scanning with dynamic real time dose adaption [1].

That kind of dose reduction tool does not totally free the operator from selection of scan parameters, and awareness of individual systems is important. Scanning protocols cannot simply be transferred between scanners from different manufacturers and should be determined for each MDCT [2, 4].

Multiple scanning parameters affect CT radiation dose – e.g. slice thickness, image exposure (tube current and voltage), scanning modes, pitch and scanner geometry [2, 3].

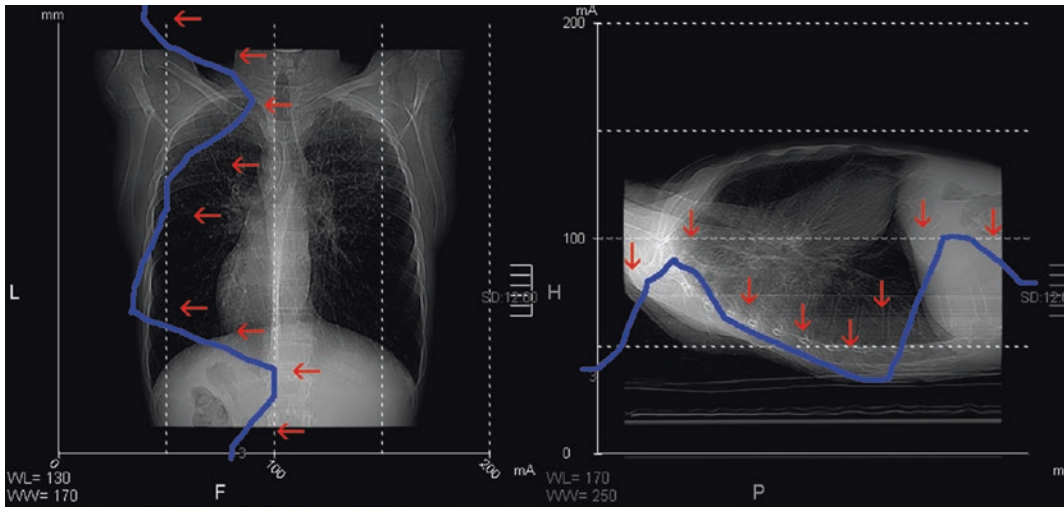

Fig. 16.2 Dose profile from a chest CT – *blue line* (marked with *red arrows*) depicts the dose profile, which was calculated by the CT machine based on the chosen parameter settings and patients anatomy (Modified after [5])

Slice Thickness, Reconstruction Parameters and Image Exposure

Half slice thickness needs double dose for the same noise level, which corresponds to image quality. Controversy to single slice scanners, where slice thickness could not be changed after data acquisition this is not any more true for MDCT – almost any slice thickness can reconstructed afterwards. One rule in MDCT is: **Scan thin - read thick** [1].

Tube current influences linearly dose – double tube current means double dose. Vice versa noise is inversely related, higher tube currents means less noise if all other factors are kept constant. Voltage (kV) has a square influence on dose - for example going down from 120 to 100 kV reduces the dose by one third, from 120 to 80 kV to less than 50 % [5].

Scanning Mode

The type of scanning is important too – either single slice (also called "sequential scanning"), helical mode, or volume acquisition are available, most commonly using multi-detector technique. The delivered dose varies considerably (more than double) between the different scan modes [2].

Pitch

Helical scanning with MDCT scanners is identical to that with SDCT scanners – rotation and table movement occur simultaneously with continuous data acquisition. Helical pitch was calculated as table movement per rotation divided by slice thickness. Due to sophisticated reconstruction parameters it is possible to move the CT table faster than the slice thickness thus saving dose, however with the disadvantage of lower image quality. In case of high contrast examinations like CT-Angiography this effect is neglectable [6].

Pitches of less than 1 implied x-ray beam overlap (and thus double irradiation of some tissue) and are not used, since it is a hallmark of helical CT, that overlapping slices can be reconstructed after data acquisition without adding radiation to the patient. Heart CT with ECG triggering is the only exception to this rule.

In MDCT the term pitch is not only dependent on table movement and slice thickness. More parameters like table speed, beam collimation have to be considered [1, 4]. The International Electrotechnical Commission CT Safety Standard specifically addressed the definition of pitch, re-establishing the original definition (distance of table travel normalized to the total beam width) as the only acceptable definition of pitch [7].

Scanner Geometry

Cone beam effects in CT are associated with the divergent nature of the X-ray beam emitted from the x-ray tube. This divergence means that the Z-axis of the X-ray beam is somewhat wider when it exits the patient than when it entered the patient. Those artifacts have to be corrected mathematically.

Additionally all helical MDCT's suffer from overranging and overbeaming when used in helical mode. Overranging refers to the effect, that in order to have data for first and last slice reconstruction the scanners needs half a rotation at the beginning and end of the scan range. Obviously this effect is more prominent with wider detectors (=more rows) and small scan ranges e.g. children. Nowadays some vendors have provided an adaptive collimation technique. These filters open and close x-ray collimation at the beginning and end of the scan [1, 8]. Overbeaming is caused by the imperfect slice sensitivity profile, meaning that is not rectangular but trapezoid causing a penumbra of radiation which cannot be used for image reconstruction. Due to the overlap of those penumbras some areas get an enormous amount of radiation. This effect is more prominent in CT with smaller detectors (=less rows).

Generally children should be investigated on at least 64 slice scanners. Volume scanners do not suffer from the "Overbeaming Effect" and only to a less degree the "Overranging Effect".

Furthermore, there are a number of aspects specific to MDCT, that systematically increase or decrease patient dose compared SDCT. One important point should be listed here: if the identical mA settings are used for MDCT that were used in SDCT, even for a scanner from the same manufacturer, there can be an unnecessary

increase in patient dose. This is primarily due to differences in the distance from the x-ray tube to the patient and detector array, although differences in tube and detector design between the scanner models also play a role [1, 2].

Users need to understand the relationship between patient dose and image quality and be aware that image quality in CT is often higher than that necessary for diagnostic confidence. Therefore a close cooperation between the referring physician and radiologist as well as teamwork between radiologists and radiographers are indispensable for accomplishing a targeted, optimized examination.

Optimized CT Scan Technique

Computed tomography calls for advanced expertise when planning the exam and interpreting the images. Deep knowledge of the technical aspects in order to be able to minimize radiation exposure is expected from all users.

Best Practice of a CT Chest Examination

Cooperative and well informed patients are helpful for a meaningful CT examination. Good results were achieved when explaining the investigation to the patient, also with some training involving breath exercises.

When lying on the table, patients should be correct positioned in the center of the scanner (Fig. 16.3). Arms up in chest CT decreases attenuation (Fig. 16.4), improves images quality and saves dose when using AEC [5].

The length of the scout view should be as short as possible, tightly adapted to the region of interest. Including the neck and the abdomen for a chest CT makes no sense. Exact positioning with the available laser finder is mandatory (Fig. 16.5). Nearly all modern CT scanners can put the tube below the CT table - thus saving in chest CT at least 50 % dose for breast and thyroid.

Furthermore pediatric adapted exposure settings for the scout view are mandatory, since all vendors deliver their machines with fixed values [1, 5] – thus in a small patient the non-optimized scoutview can deliver the same dose as the whole examination.

Planning scan length planes on the scout view should only cover the clinical question. Common practice, e.g. in chest CT, is to scan from above the pulmonary apex through the lung base, which is, for example, not necessary in heart CT. There is a linear correlation between the tube current-time product (mAs) and radiation exposure. A simple way to reduce radiation exposure is to lower the tube current (mA) – consecutively image noise will be increased, the effect being more prominent in low contrast areas (e.g. soft tissue) than in high contrast areas (e.g. bones, lungs) [9].

A comparison of CT scanners from different manufacturers based on their mAs is of limited value since the actual radiation exposure is affected by the CTDI which may differ by a factor of up to 1.7 between different vendors. Due to differences in pre-filtering and geometry, a setting of 10 mAs in one machine may correspond to 17 mAs in another one [1, 2, 9].

Kilovolt settings correlate with square of dose. For adults keep tube voltage at 120 kV. For younger patients up to the age of 14, a tube voltage of 100 kV is recommended [1, 5].

In chest deformities, like funnel chest, the bone skeletal deformity is of particular interest, thus representing a high contrast situation in which more noise can be accepted. Assuming, that there are optimized settings for standard chest CT available, then in machines equipped with AEC a noise level of about one third over the standard CT represents a good starting point [5]. In older machines without AEC reducing mAs settings by 50 %, as compared to the standard settings, would be the selection of choice. It has to be considered, that if voltage is lowered the dose is reduced by the square multiple e.g. dropping voltage from 120 to 100 kV means a reduction of dose about 30 % and therefore mAs can only be reduced about 20 %.

The need for protocol optimization has been seen in Europe already since the 90s and has led

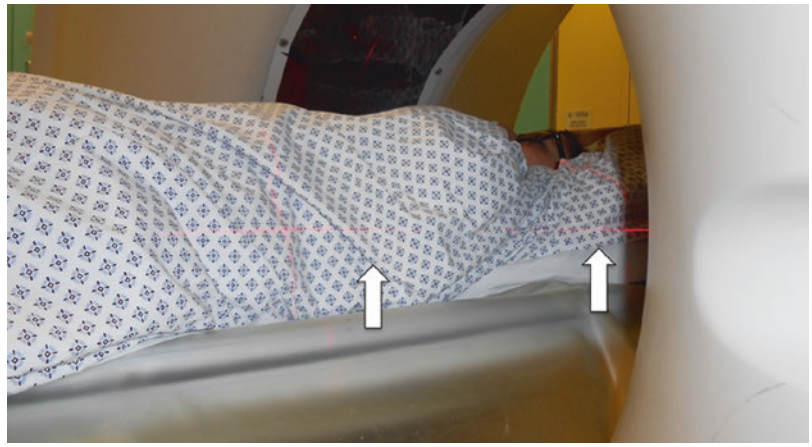

Fig. 16.3 Exact positioning of the patient in the center of the scanner is mandatory. *White arrows* demonstrate the laser finder

Fig. 16.4 For chest CT positioning of the patient with arms up improves image quality and reduces in CT machines equipped with AEC

to the EU Quality Criteria for Computed Tomography [6]. An update has led to the formation of European Guidelines for Multislice Computed Tomography [10].

Imaging in Chest Deformities

Several possibilities for imaging of chest deformities are on offer e.g. funnel chest exist – plain chest X- Rays and CT being the most frequent choices for investigation. Chest films demonstrate a 2D attenuation image of the 3D space and time, usually during a breath hold. Those images can be enhanced by fixing a wire in the depressed area of the chest and obtaining an image in lateral position so that the severity of funnel chest can be demonstrated (Fig. 16.6). Using standard chest radiographs the severity of funnel chest can be characterized and quantified by the "Vertebral Index (VI)" and the "Fronto-Sagittal Index (FSI)" [11].

CT and its variants allow contiguous data acquisition, thus forming an ideal basis for 3D-reconstructions, multiplanar reconstructions (MPR) being one of the most frequent ones. In MPR, image data is not reconstructed in the standard axial plane, but e.g. in coronal or sagittal axial ones (Fig. 16.7). Therefore, indices like the VI and FSI can also be calculated. Due to the 3D data acquisition and processing capabilities like selection of the optimal plane, it can be assumed

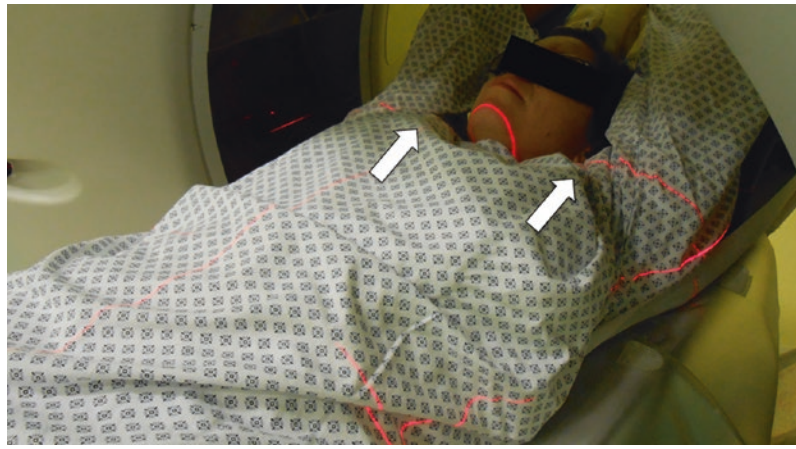

Fig. 16.5 The starting line (laser finder) for the scoutview is depicted by *white arrows*

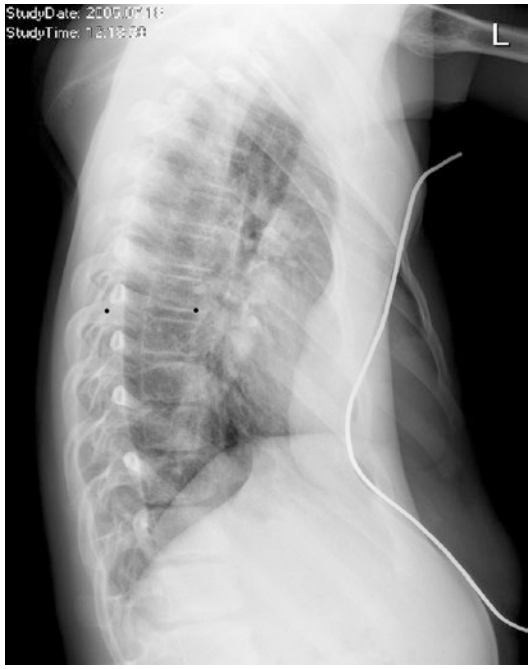

Fig. 16.6 Lateral image in a patient with funnel chest. The wire demonstrates the depression of the sternum, but cannot show its rotation

that those parameters exhibit less variances as compared to those obtained by plain chest films.

In CT, bones are characterized by high attenuation values (Hounsfield Units), thresholding techniques can be used to select those pixels. This is a technique called "Shaded Surface Display" and by rotating the 3D model different properties can be studied (Fig. 16.8). A complete different technique is represented by "Volume Rendering". Contradictory to "Shaded Surface Displays", where a certain threshold (or threshold range) is used to select those pixels belonging to bones, in "Volume Rendering" every pixel gets an particular opacity, ranging from 0 % (= completely transparent) to 100 % (=completely opaque). The pixel's transparency are set using a so called "opacity curve" superimposed on the histogram of all data volume's pixel (Fig. 16.9). An example of "Volume Rendering" in funnel chest is shown in Fig. 16.10.

Besides the deformity display, an automated classification of funnel chest seems be an advantage - especially for follow-up investigations. In our computer laboratory a novel approach based on CT has been developed [12]. In this, the first the slice with the greatest deformity is selected following which digital image processing is used in which the outer and inner contour of the chest is segmented, followed by the calculation of bounding hull (= convex hull) and automated computation of six new indices (Fig. 16.11), which form the basis for separation of three groups: normal, borderline and funnel chest cases (Fig. 16.12). The same algorithm could be

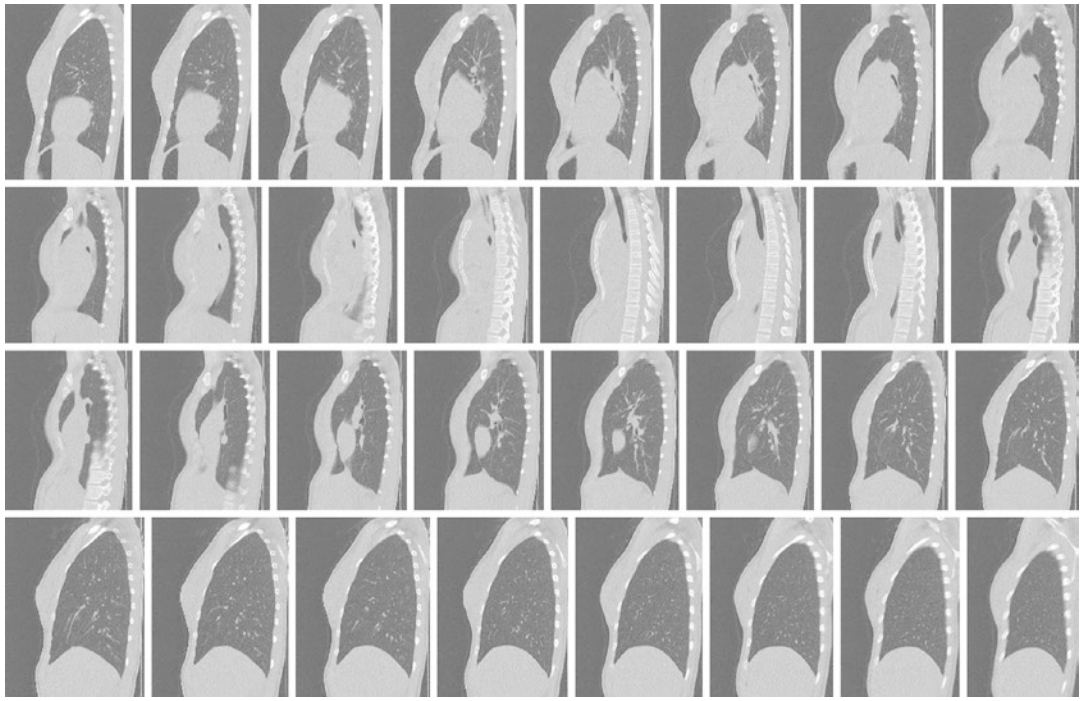

Fig. 16.7 MPR reconstruction in sagittal orientation in the same patient as in Fig. 16.6. An image series was computed, thus allowing selecting the plane with biggest deformities. Such MPR's can be generated in any arbitrary direction

applied with some modifications to Magnetic Resonance Images.

Ideally an imaging method for funnel chest would be completely non-invasive. Saxena et al. reported raster stereograph [13] for that purpose. Another possibility is based on the projection of different stripe patterns on the patient's chest – from those patterns a 3D geometry can be computed (Fig. 16.13).

Why Children Are Different

Since the invention of Computed Tomography (CT) in the early 70's, CT has experienced an evolving role in daily clinical praxis. The invention of spiral (helical) CT in the middle of 90's enabled for the first time contiguous data acquisitions with the latest progress to volume CT using 320 rows. Due all these technical achievements there is a steady progress in CT examinations with the consequence of delivering a considerable amount of radiation burden to patients. For example, only 10 % of all radiation based imaging modalities is CT, but it is responsible for delivery of about 50 % of the total collective dose [14].

Children differ in many ways from adults – not only in size, weight and shape. First of all the psychological situation during an imaging study has to be considered. If the natural environment of a child gets compared with that of a hospital, then is obvious that fears are created: everybody looks strange (white coats), rooms are white (some even with glazed tiles), you have to be quite, get undressed, are put in strange devices, commands like "breath hold" are given etc. It can imagined what a child feels, seeing an intensive care patient with a lot of tubes, plasters and much more passing by.

Therefore all efforts have to be done, to create a friendly and patient environment – this starts at

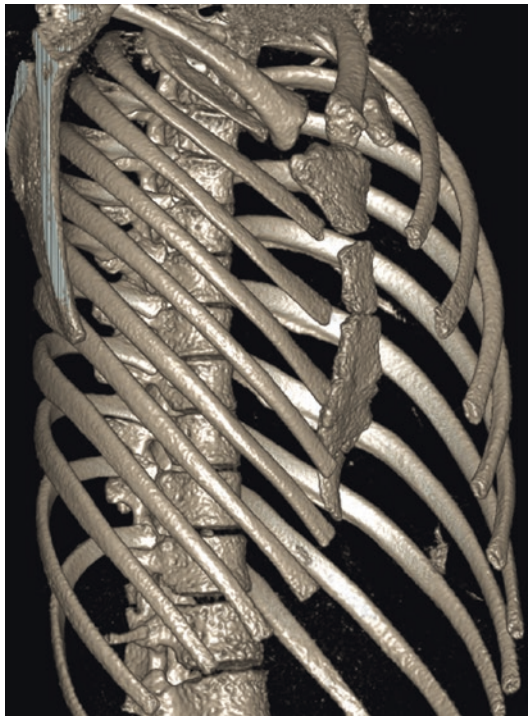

Fig. 16.8 3D funnel chest reconstruction using "Shaded Surface Display" technique – bones have high attenuation values and therefore those pixels can be easily selected in native (non-intranvenous contrast enhanced) CT. Afterwards all of those pixels are converted to a surface and highlighted by an artificial lamp

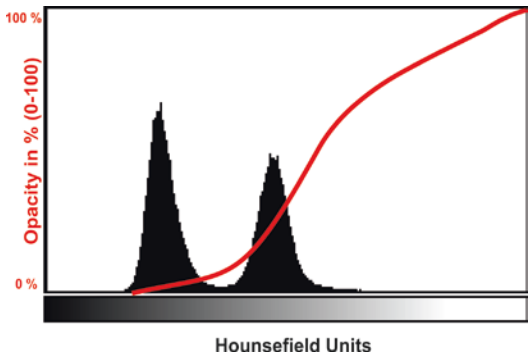

Fig. 16.9 Principle of opacity curve – histogram of scan volume is displayed; Hounsefield Units are on the x-axes. Superimposed on the histogram there is the opacity curve in red – ranging from 0 % opacity (equals complete transparent pixel) to 100 % opacity (equals complete opaque pixels). By changing the opacity curve the different reconstructions of Fig. 16.10 can be generated

the counter and ends after the discharge of the child from the imaging unit. Every involved person has to keep the psyche of the small patient and the accompanying persons in mind. If a child starts crying almost all is lost. Moreover, all necessary handling must kept to an absolute time minimum, since even in cooperative children this phase could end earlier than expected.

Radiation sensitivity is considerable higher than in adults – at least by a factor 10. This fact can be explained by the higher cell turn over as it is necessary for growth as well as the higher absorption rate per tissue [15]. Pediatric Radiology faces the multidimensional problem of child development representing a 4D issue. There are three dimensions in space and the fourth dimension is time, representing maturation. Everything is changing, weight, height, bone density, number of alveoli, body proportions and radiation sensitivity. On Fig. 16.14 the change in body proportion is shown. In addition, the numbers in grey circles describes the relative radiation sensitivity. It is easy to recognize, that in e.g. newborns the head and extremities exhibit greater radiation sensitivity than later in life. In contradiction, the gonads are less susceptible in early life. Both can be explained easily by maturation – a newborn has only a few reflexes and the usual position is with flexed extremities. At the age of 1 year baby starts walking, therefore a lot of maturation takes place in the brain and makes it more susceptible to radiation. On the other hand, babies are obviously not sexual active and therefore gonads do not need a high cell turn-over.

There is a broad range of dose reported for pediatric CT so for example a range from 1.0 mSv up to 24.0 mSv in children's head CT [17] – higher doses inherently associated with cancer development [15, 18]. If the dose for head CT is not adopted for children it is estimated that 1 in 1000 of those patients will die of cancer after a single, not optimized, cranial CT [15, 19]. It can be assumed, that those principles are valid for chest CT too.

The ALARA (**A**s **L**ow **A**s **R**easonable **A**chievable) principle, taught us, to use an

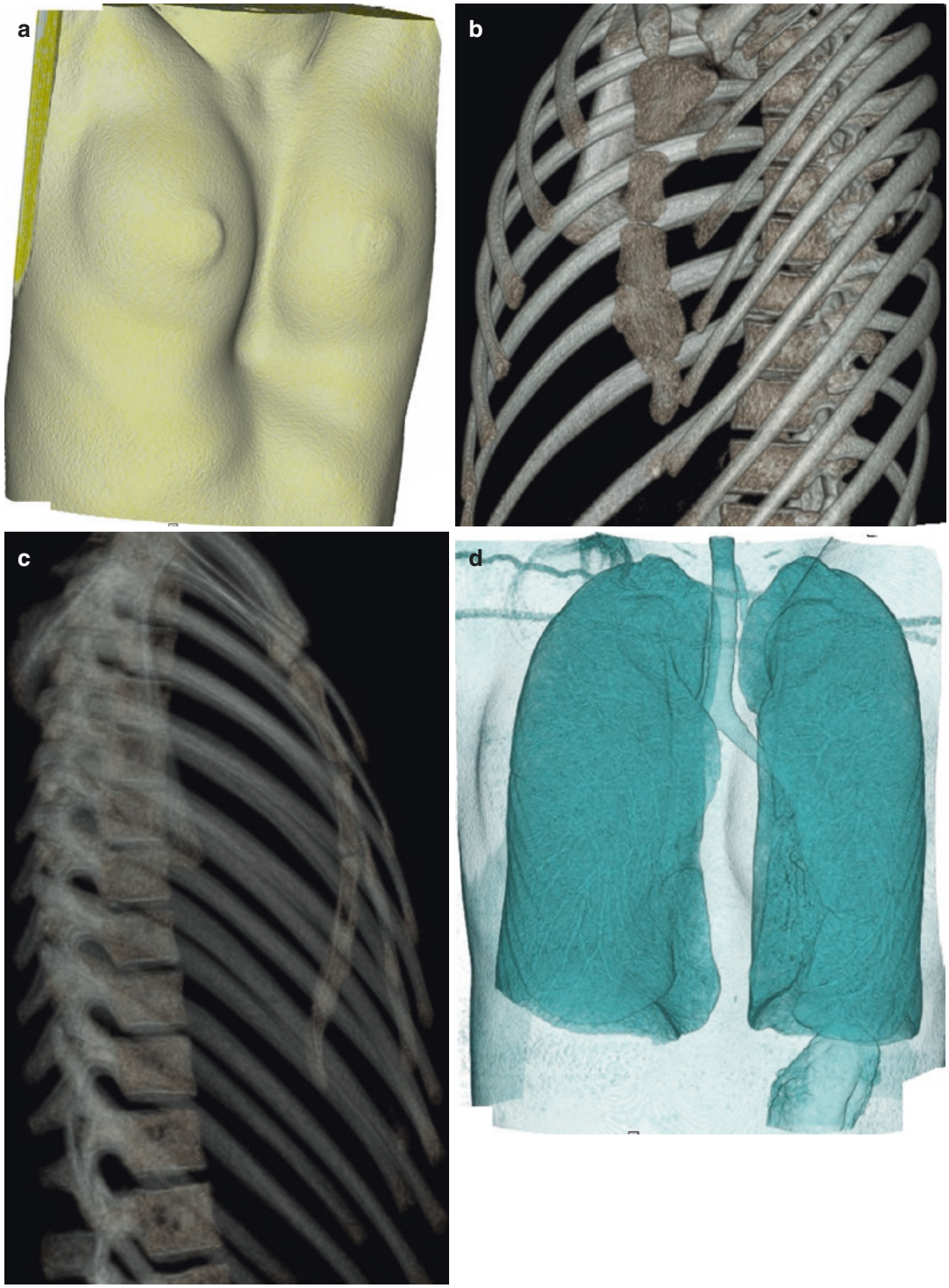

Fig. 16.10 Four different examples of volume rendering, the different images were computed from the same dataset, only the opacity curve was changed and point of view. (**a**) Opacity curve was set in order to demonstrate soft tissue settings, in (**b**) to demonstrate the skeleton, in (**c**) same opacity settings as in (**b**) but the view point was changed to lateral in order to demonstrate better the funnel chest deformity and in (**d**) opacity curve was manipulated to depict the airways and air filled lung

imaging modality with lowest possible exposure settings, which answer the clinical question [20]. Therefore, in Pediatric Radiology a certain CT protocol has to consider the above mentioned facts and in combination with the clinical question as well as impact on patient management. Figure 16.15 presents a summary of all parameters, which have to be adjusted for a CT protocol. The complexity of that chart makes clear, that pediatric CT is a highly sophisticated investigation and persons involved should undergo a specialized training.

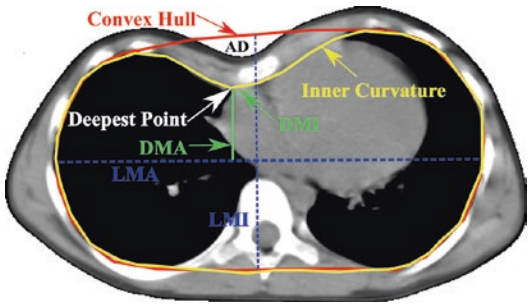

Fig. 16.11 Scheme of new indices for automated funnel chest classification. Using a novel approach the inner and outer contour of the chest is segmented after selection of the slice with biggest deformity [12], computation of the bounding hull (= convex hull) and the following six new indices: *MA* length of major axis, *LMI* length of minor axis, *DMA* distance of the deepest point from major axis, *DMI* distance of the deepest point from minor axis, *ACH* area of convex hull of the inner curvature, *AD* area of deformation

Dosemanagement

As mentioned in the previous sections, one of the biggest challenges of today's radiology is, to reduce the radiation dose during CT examinations without compromising the image quality. In general, higher radiation doses result in higher image quality with lower noise, while lower doses lead to increased image noise and thus affecting sharpness too.

CT is used increasingly to replace conventional x-ray studies. It is important that patient dose is given careful consideration, particularly with repeated or multiple examinations. Even when CT is the optimal investigation for the clinical problem and recommended by guidelines, many parameters have to be set optimally in order to guarantee lowest dose for the answer of the clinical questions. Dose reduction strategies were given in the previous sections.

Referring physicians, surgeons, radiologists and radiographers have to understand the relationship between patient dose and image quality. They are aware that, often, image quality in CT is higher than that needed for diagnostic confidence. Images of the highest quality are not essential for all diagnostic tasks, but rather the level of quality (e.g., low noise, medium or low dose) is dependent on the diagnostic task.

There is increasing awareness of how adapting exposure factors can contribute to the management of patient dose. However, the rate at

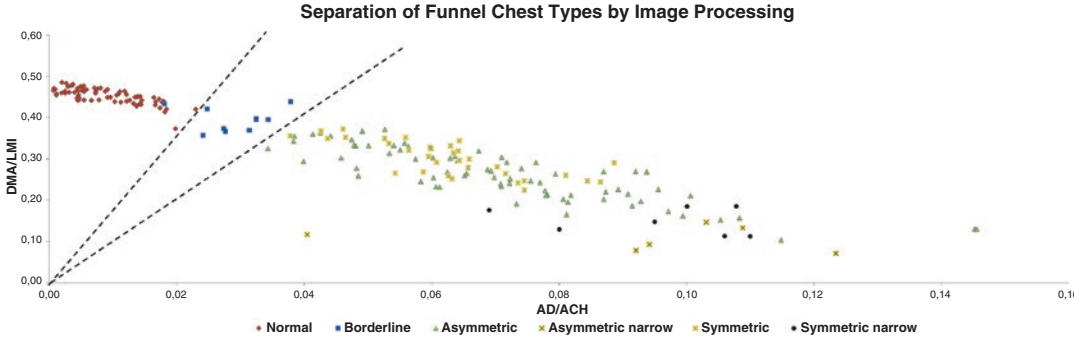

Fig. 16.12 Chart showing classification results on the indices in Fig. 16.11 – three groups can be separated: normal, borderline and funnel chest cases

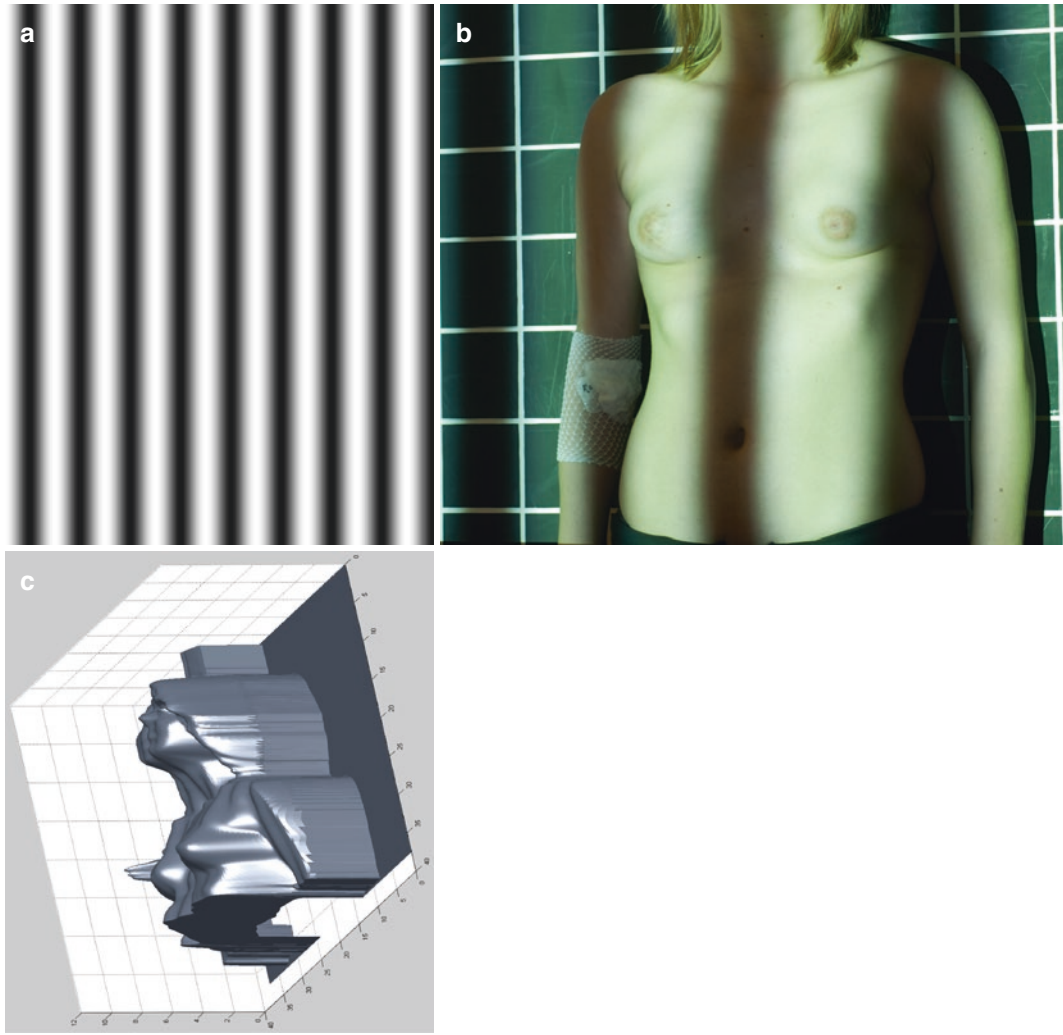

Fig. 16.13 (**a**) Light stripe patterns are demonstrated; (**b**) image shows projection of those patterns on a patient's chest and (**c**) exhibits the result after 3D reconstruction of the chest

which technology is changing requires permanent attention to management of patient dose.

Dosemanagement in Clinical Practice

Publication 102 from ICRP [2] suggests following important care that should be taken with regards to dose in clinical practice.

Justification of CT use is a shared responsibility between requesting clinicians and radiologists. It includes justification of the CT study for a given indication and classification of the clinical indications into those requiring standard dose CT and those requiring only low dose CT. Scanning parameters should be based on study indication, patient size, and body region being scanned so that patient dose can be managed based on these parameters.

Guidelines (selection criteria for CT examinations) are necessary in order to avoid inappropriate studies. In addition, alternative non-radiation imaging techniques should be considered.

Training of requesting physicians and CT staff can help in the management of scan indications, protocols, and patient dose.

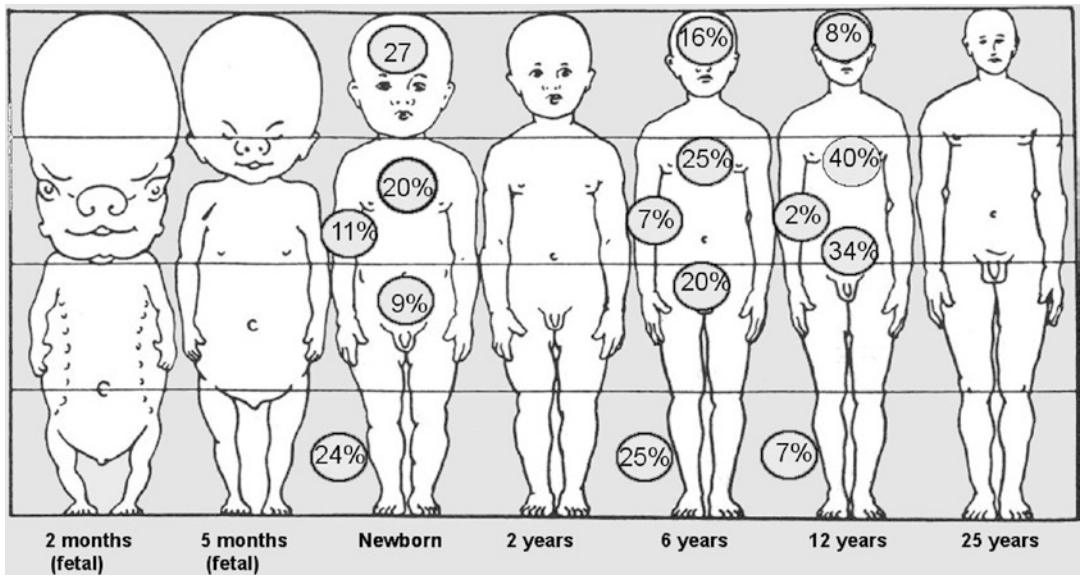

Fig. 16.14 Development scheme in children showing the hanging proportions with the head prominence within the first years. Numbers in circles refer to the relative radiation sensitivity of a specific organ system (modified after [16])

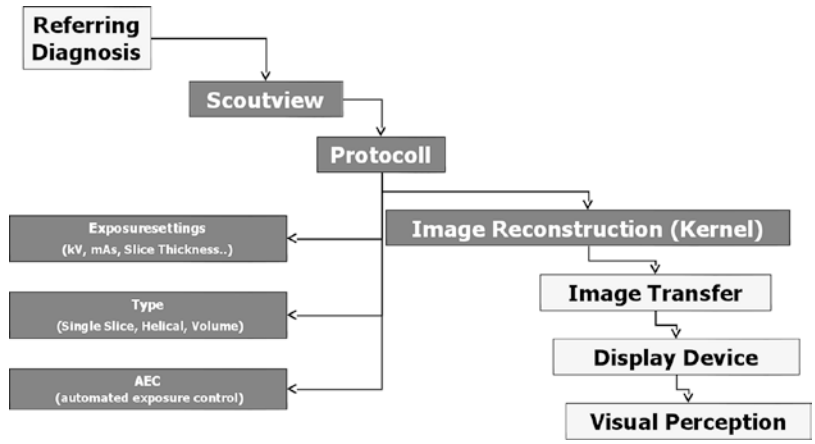

Fig. 16.15 Scheme of parameters, which have to be considered for a CT protocol (Adopted from [5, 21])

Dose Parameters in CT

Radiation-induced health effects are correlated with the mean absorbed dose to organs and tissues (i.e., the organ dose). Since organ dose generally cannot be measured directly, a number of indirect approaches are used to evaluate the impact of CT practices on organ doses. The Computerized Tomography Dose Index (CTDI) is measured with the use of a 16 or 32 cm diameter CT dosimetry phantom. The Dose Length Product (DLP) represents the product of the CTDI and the scan length. Using these directly measured quantities, or calculation methods, medical physicists can estimate organ doses and the quantity effective dose [2].

Effective doses are expressed in milli Sievert (mSv) and are useful for comparison among different diagnostic medical procedures and also background radiation (e.g. terrestrial and cosmic radiation). Depending of the location, the annual value of effective dose for background radiation

Table 16.1 Diagnostic reference levels from the federal office for radiation protection (www.BFS.de) for chest CT [21]

Study	Age/weight	$CTDI_{vol}$ –16 [mGy]	$CTDL_{vol}$ –32 [mGy]	DLP –16 [mGy*cm]	DLP –32 [mGy*cm]
	≤5 kg (newborn)	3.0	1.5	40.0	20.0
	6–10 kg (≤1 year)	4.0	2.0	60.0	30.0
Chest	11–20 kg (2–5 years)	7.0	3.5	130.0	65.0
	21–30 kg (6–10 years)	10.0	5.0	230.0	115.0
	31–50 kg (11–15 years)	/	8.0	/	230.0
	51–80 kg (>15 years)/adults	/	12.0	/	400.0

is in the range of 2–4 mSv/year. Typical effective doses in a chest CT are in the range of 5.5–8 mSv. Chest CT involves larger radiation doses than a conventional x-ray procedures for the chest (range 0.01–0.02 mSv) [2, 18]. When a CT scan is justified by medical need, the associated risk is small relative to the diagnostic information obtained [18].

Since children are smaller than the 16 or 32 cm dosimetry phantom, CTDI values are heavily underestimated – up to 200–300 %. This problem was addressed by the AAPM Report 204 where a better parameter for CT investigations of the body trunk was defined – the "Size Specific Dose Estimates (SSDE)" [22].

Diagnostic Reference Levels (DRL)

The radiological protection principle of dose limits used for exposure of workers and the general public does not apply to medical exposures for patients. Assisting in the optimization process of medical exposure to patients, the concept of diagnostic reference level (DRL) has been introduced. CTDI and/or DLP are the quantities proposed as DRL for CT. Determination of local DRLs should be done using a sample of ten standard sized patients (e.g. common chest CT). Mean values of these findings should be compared with the DRL of other organizations (e.g. EMAN, ICRP, National Institute of Radiation Protection). If the own levels are higher than nationally or internationally set DRL, appropriate corrective actions should be applied to reducing the dose [2, 3].

In a several nationwide German surveys, DRLs for adult and pediatric patients have been evaluated by the Federal Office for Radiation Protection (Bundesamt für Strahlenschutz – BfS) -the latest version of which being published in 2010. The following table (Table 16.1) is extracted and translated from the original document and presents $CTDI_{vol}$ and DLP for different ages in Chest CT [23].

Surveys of reference dose quantities (CTDI, DLP) and effective doses and comparisons of dose quantities with diagnostic reference levels have been demonstrated to be a useful strategy for promoting dose management [24].

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Structured Light Technique for Measurement of Pectus Excavatum

17

Marcin Witkowski and Wojciech Glinkowski

Introduction

The recent development of electro-optical instrumentation allowed the development of three-dimensional (3-D) structured light scanners which may be used to measure the surface of human body. Such scanners offer high measurement resolution and accuracy at an affordable price. Moreover, the time of measurement is very short (less than one second) and the measurement method does not require to expose the patient to any kind of possibly harmful radiation. This chapter describes an application of a 3-D measurement system for assessment of pectus excavatum (funnel chest) deformity.

Being the most common congenital deformity of the chest wall, pectus excavatum (PE) is reported to affect from 0.23 to 1% of population [1–3]. An inward depression of the sternum that is characteristic to PE may in moderate-to-severe cases be the cause of exercise intolerance and, rarely, chest pain. Limited physical fitness accompanied by an altered body image may be reasons for low self-esteem and lack of self-acceptance by adolescent and young adult patients. Psychological and cosmetic indications for surgical treatment of PE gained more importance after introduction of minimally invasive and cosmetic surgical methods.

Most widely used methods for identification of PE cases rely on physical examination, charts, preoperative photographs, and computed tomography (CT) scans. Physical examination should pay attention to asymmetry index, relative length of depression to the sternum, fraction of sternum affected, degree of sternal torsion, and localized vs. diffuse morphology [4]. Preoperative photographs are used to document the deepest point of the pectus, the length of the pectus relative to the sternum, the relative amount of the sternum affected, symmetry of the pectus, localized or diffuse (cup or saucer) morphology, and the presence of flaring of the lower costal margin. Although standard (two-dimensional, 2-D) photography is not sufficient to document three-dimensional characteristic of the deformity, preoperative photographs can be used to determine chest lines, the area affected by the depression, the vertical length of the deformity, and the length of the sternum affected. Methods such as physical examination, measurement of the chest circumference with a metric tape and anthropometric measurement called chest cyrtometry are performed for basic documentation [5, 6]. Also an anthropometric index defined as a maximum anteroposterior measurement in the region of the greatest deformity divided by the measurement of the greatest depth of the defect is a simple method to assess the deformity.

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Radiological examination sometimes may additionally define the orientation and relationships of the ribs, costal cartilages and the sternum for preoperative planning. However, plain photographs cannot become concurrent to CT assessment and Haller Index (HI) calculation [7, 8], which remains a gold standard for preoperative planning and decision making. The Haller Index calculated from CT scans was developed to provide an objective measure of the depth of the deformity for determining surgical eligibility [9]. Kim et al. [10] developed additionally indices that can automatically be extracted by computerized image to describe the depression, the asymmetry, of the chest-wall, eccentricity, flatness and circularity index, to present the deformation of the chest-wall in details. Despite of several reported pectus excavatum severity classifications the Haller Index remains the most commonly used. Haller Index (Fig. 17.1) greater than 3.2 is used as one of the criteria for surgical correction. Proposed value was accepted as the cutoff between mild and moderate pectus excavatum.

There are cases where a single level HI may not supply sufficient information about chest-wall deformity. Then a multiple level slice examination may supplement an assessment for more accurate diagnosis and preoperative planning [11]. The main disadvantage of using HI for estimation of PE severity is the exposure to ionizing radiation. Due to current trends to reduce overall yearly radiation exposure new methods of PE assessment are developed, particularly for repeated monitoring.

Background Work and Motivation

Promising results of the authors' experiments on 3-D body measurement [12–14] as well as described by other researchers were the starting point for developing an optical 3-D system for torso measurement and an automatic data processing algorithms to assess the shape and severity of deformity of pectus excavatum.

Our first trunk measurement system was only able to measure from a single direction at a time and required the patient to sit in a specially constructed frame (Fig. 17.2). Rotating the frame with the patient around a vertical axis allowed capturing the whole surface of the trunk. The measurement process was time-consuming and was uncomfortable for both the patient and the operator.

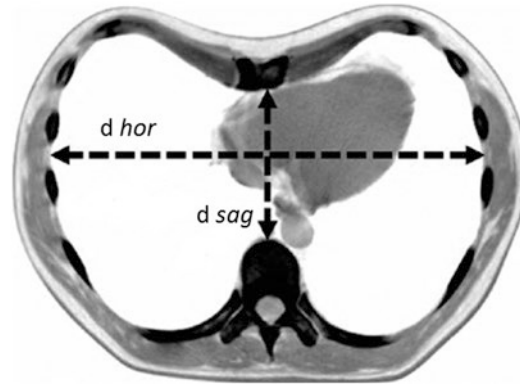

Fig. 17.1 Distances measured on a CT-scan for Haller Index calculation

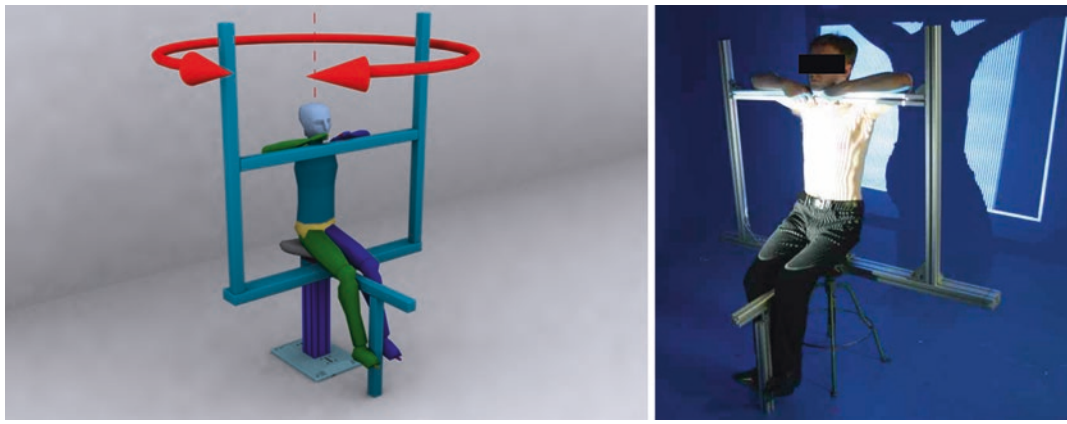

Fig. 17.2 Taking a 3-D measurement with an early-stage version of the measurement system required the patient to sit in a special frame

Several versions of the measurement system were developed and installed in the Baby Jesus Clinical Hospital, Medical University of Warsaw (MUW) since then. The system used at present is a high-accuracy optical markerless 3-D measurement system and may be used for non-invasive, quick and relatively inexpensive assessment of pectus excavatum deformity but is also used for posture screening [15] and other applications. The system facilitates an automatic measurement and data processing software for analysis of the PE deformity. The process is fully automatic and does not require the any operator input apart from preparing the patient at the beginning of the measurement. As a result the deformity is evaluated in an objective way. The custom developed 3-D scan index (I_{3ds}) is automatically calculated from measured torso surface outline and shows correlation with CT Haller Index.

The Measurement System

The measurement system is composed of four modules which measure the patient from four directions simultaneously. Each directional module is an optical full-field 3-D scanner based on structured light projection method [16]. The main components of each module are (Fig. 17.3):

Fig. 17.3 The components of the measurement system as installed in a laboratory at Medical University of Warsaw: a modified DLP projector at the bottom, a CCD camera at the top and a PC at the side

– a projection unit, which is a Digital Light Processing (DLP) projector (XJ-A250, Casio, Japan),
– a detection unit which is a Charge Coupled Device (CCD) 1 MPix FireWire camera (Flea FL3-FW-14S3M-C, Point Grey Research, Canada) and
– a PC-class computer.

In order to attain the highest accuracy and repeatability of the measurement the projectors had to be customized. Standard lenses had to be replaced by custom designed ones in order to allow preserving stable parameters of the optical system. The factory-installed high-speed fans introduced vibrations in the image and had to be replaced by larger low-speed fans.

The directional modules of the measurement system installed in the laboratory at Medical University of Warsaw (MUW) are located in the corners of the room 5.2 × 5.2 m at height 1.9 m from the floor level (Fig. 17.4). The modules are fixed to the walls in order to keep their position unchanged. Any modifications of hardware configuration or optics parameters must be followed by a re-calibration of the measurement system. The calibration process requires a dedicated calibration board with circular markers. Thanks to an automated calibration procedure there is no need of user interaction and the whole process takes 60 min.

For the actual measurement the patient is asked to stand still inside the calibrated measurement volume and hold breath for a few seconds. Then a series of patterns is projected onto his or her body surface. These patterns include sinusoidal fringes and modified binary Grey codes. The shape of body surface is calculated based on the raster deformation according to the Temporal Phase Shifting (TPS) method [16].

The measurement takes less than a second. All directional modules perform the measurement simultaneously. In order to avoid detecting patterns projected by another unit the adjacent modules are separated spectrally (Fig. 17.5). Data set produced by the measurement system is in the form of set of (x, y, z) points that represent the surface of patient's body (Fig. 17.6). The common

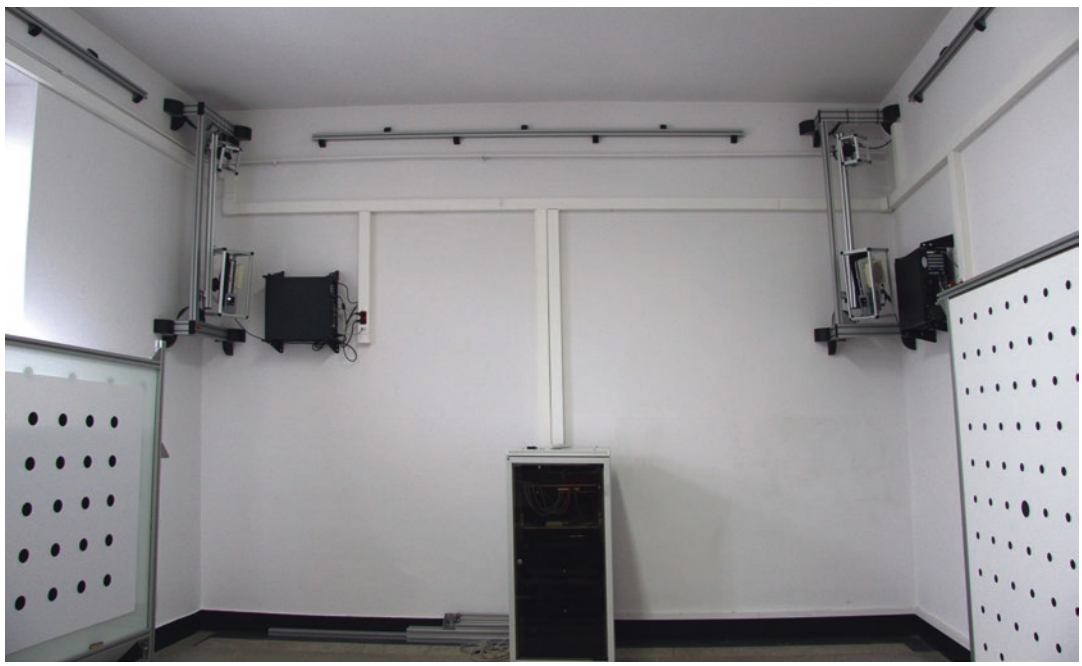

Fig. 17.4 A perspective view on two measurement modules as installed in the laboratory at Medical University of Warsaw

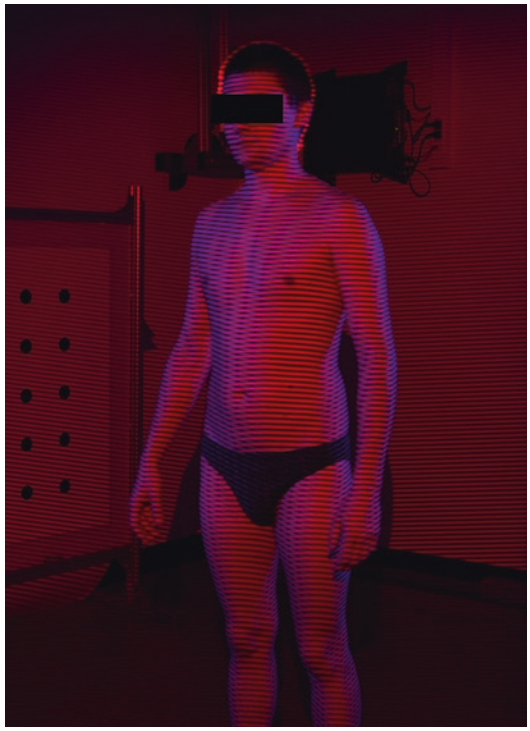

Fig. 17.5 Healthy subject's body covered with red and blue sinusoidal fringes projected during the measurement

coordinate system used by all modules is set based on global calibration process during which spatial relationships between directional modules are found. As a result directional point clouds are merged automatically without additional computations (Fig. 17.7).

The metrological values of the measurement system are as follows:

- measurement volume size: 1.5 m × 1.5 m × 2.0 m,
- accuracy: 0.4 mm,
- data acquisition time: 0.7 s (all modules simultaneously),
- maximum number of points: 4 million.

Automatic Analysis of Chest Surface

The method for PE assessment developed and used by the authors takes advantage of a dedicated 3-D scan index (I_{3ds}) describing the shape outline of the trunk at the area where the deformity represents the highest chest depression. The index is

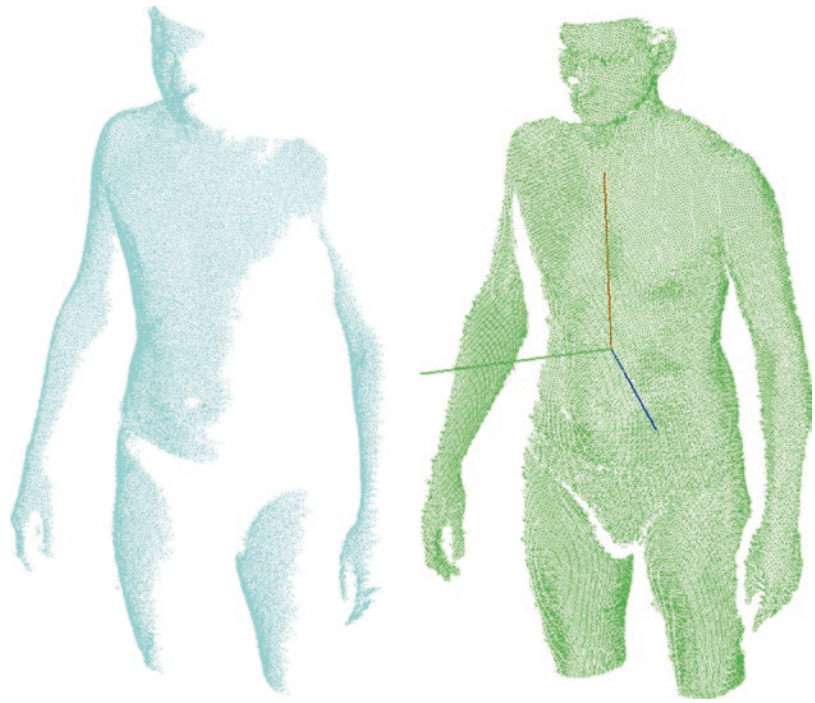

Fig. 17.6 Exemplary point clouds captured by the front left and front right directional measurement modules

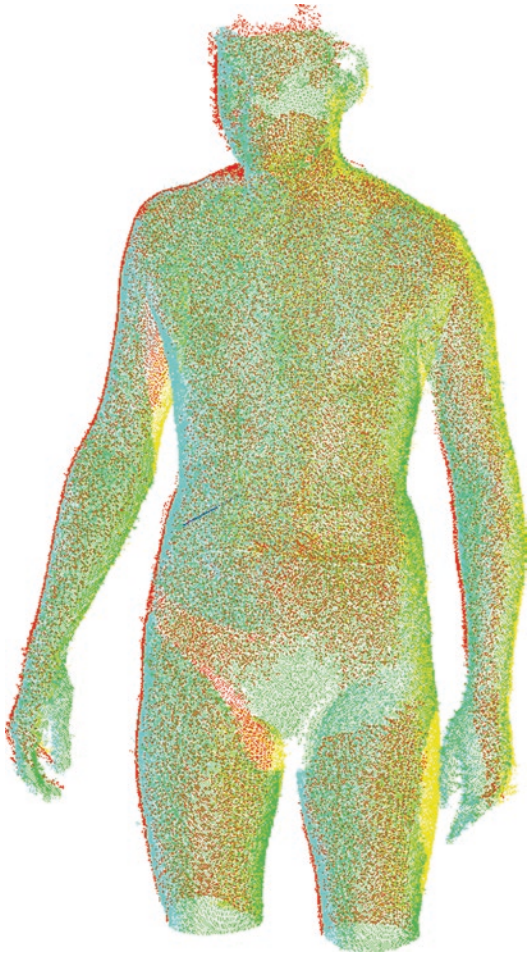

Fig. 17.7 Merged directional point clouds representing the whole body surface measured. Different colors represent points originating from different directional modules

calculated based on a transverse slice of the trunk in a similar way to Haller Index in order to allow for a comparison with the latter. The main and most meaningful difference is that (I_{3ds}) is calculated based on external surfaces while HI is calculated based on the dimensions of internal structures. In a mathematical sense (I_{3ds}) has been defined as the distance from the most depressed point of the front chest wall to the surface of the back thoracic wall in the midline divided by the external surface width of the chest at the same level (in the same slice). The (I_{3ds}) formula is given below (1):

$$I_{3ds} = \frac{D_{LR}}{D_{FB}} \tag{17.1}$$

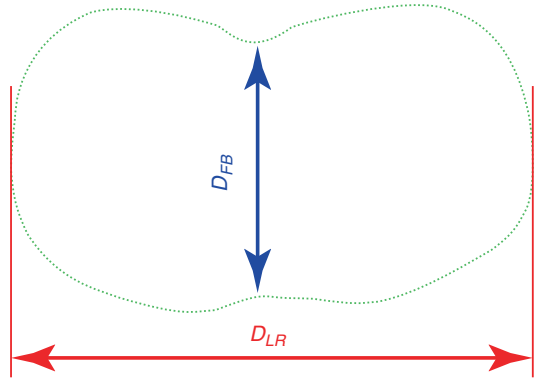

Fig. 17.8 The distances used to calculate the (I_{3ds}) index

where: I_{3ds} – the calculated index, D_{LR} – the length of the slice along the transversal axis, D_{FB} – the distance from the most depressed point of the front chest wall to the back chest wall (Fig. 17.8).

The algorithm used for analysis of the 3-D point cloud is as follows. First, the point cloud needs to be segmented into body parts in order to extract the fragment that represents the torso. All point cloud fragments other than representing the chest are ignored in further analysis. Then the remaining point cloud representing patient's trunk is cut into thin horizontal slices. In the next step for each slice a value of (I_{3ds}) index are calculated. The slice for which (I_{3ds}) reaches the lowest value is regarded as the slice of the most significant deformation and this particular value of the (I_{3ds}) index is regarded as the (I_{3ds}) value representative for the patient under examination. The following paragraphs will present a more detailed description of the developed algorithm (Fig. 17.9).

Segmentation of a Point Cloud Representing the Whole Body Surface

In order to extract the trunk part from the point cloud representing the whole body of the patient an algorithm for body part segmentation is used. The segmentation algorithm uses an assumption that the measured person is standing in an upright position on his or her both legs and the arms do not touch the torso.

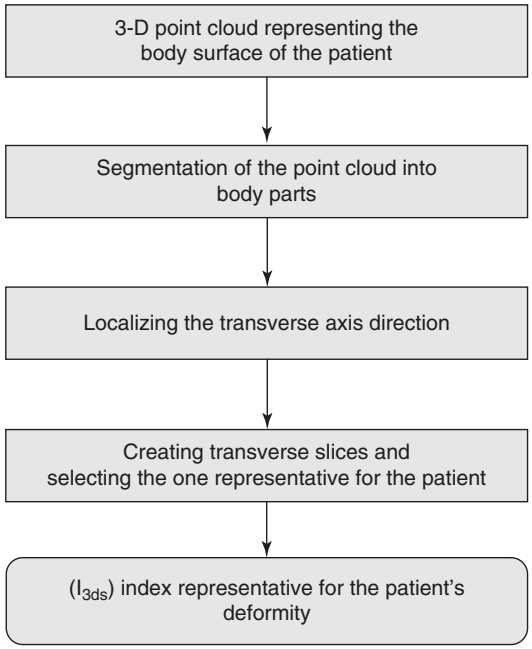

Fig. 17.9 The main stages of the developed algorithm

The point cloud is divided into horizontal slices from which separate point groups are determined. Body fragments are created from adjacent point groups that have similar properties (e.g. center of mass (CoM), circumference, number of points). This rough segmentation allows to differentiate main body parts: upper and lower limbs, head, trunk (two parts divided at an armpit height) and pelvis area (Fig. 17.10). The trunk fragment is extracted for (I_{3ds}) calculation in the next steps of the analysis algorithm.

Localizing the Transverse Axis Direction

The measurement system vertical axis OY is used as the examined patient's longitudinal axis along which the cross-sections are performed. However, the direction of patient's transverse axis is unknown as it depends on the position taken by the person during the measurement and may differ significantly from ne measurement to another. For the analysis purposes the patient's transverse axis is defined as a horizontal line that passes

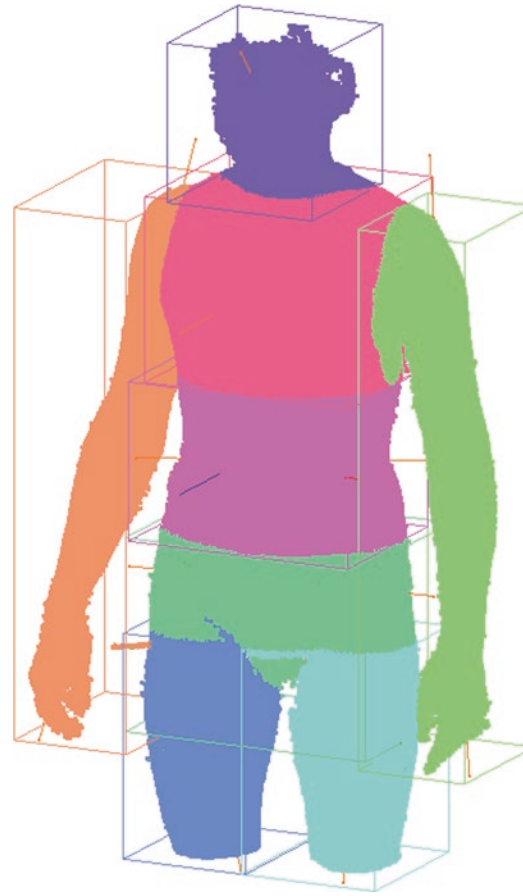

Fig. 17.10 A point cloud representing body surface segmented into body parts

through the left and right dimples related to the posterior superior iliac spines (Fig. 17.11). The 3-D positions of the dimples are automatically localized by means of back surface analysis as described in [15]. For further analysis the point cloud is rotated around the vertical axis in such a way that the patient's transverse axis matches the OX axis of the measurement system.

Preparing Transverse Slices

When the orientation of the model has been determined, the thoraco-abdominal fragment of the point cloud is sliced with horizontal planes transverse to the patient's torso (Fig. 17.12). The 5 mm slice height has been chosen as optimal.

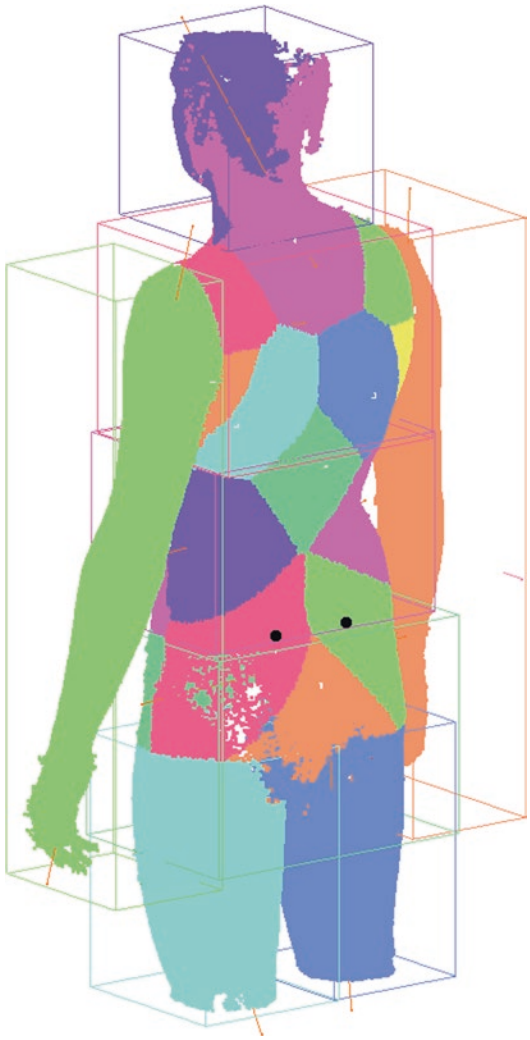

Fig. 17.11 Areas related to anatomical landmarks on the back surface. Characteristic points selected with black dots

Gain in accuracy due to lower slice height is negligibly small. Each slice is checked whether it is a closed or open curve. Slices that form open curves located above the level of axillae are neglected.

Calculation of the (I_{3ds}) Index

For each closed slice the value of the (I_{3ds}) index is calculated. The index is a quotient of two distances. The first (nominator) is the distance from the most depressed point of the front chest wall to the back chest wall. The denominator is the width of the slice, which is defined as the size of the slice along the transversal axis.

Fig. 17.12 Transverse slices among which the most deformed slice is searched (open slices have been neglected). Different colors apply to different slices

The slice for which its (I_{3ds}) index reaches the minimum is regarded as the most representative slice for the patient and its value is regarded as the (I_{3ds}) index value representative for the patient (Fig. 17.13).

The presented method is fully automatic in both the measurement and data analysis stages. In particular, there are no markers attached to patient's skin and there is no requirement to point the deformation extreme with mouse by the operator.

Clinical Examinations

Studies conducted by the authors have presented a relationship between the Haller Index and (I_{3ds}) index calculated based on the 3-D optical scan of the patient [17]. The study was conducted on 12 patients with pectus excavatum. The

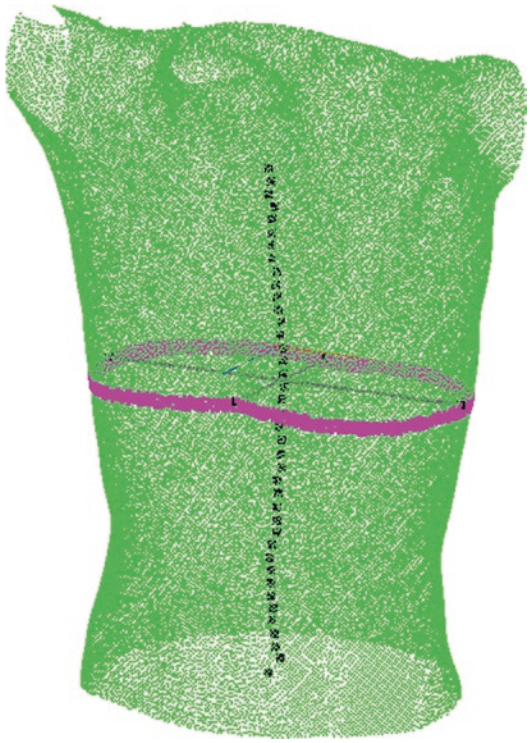

Fig. 17.13 The most representative slice for the examined patient. The characteristic distances used to calculate the (I_{3ds}) value are also presented

preliminary series consisted of white adolescent and young adult males at average age 16 years and 5 months (12–19 y.a.). The cases were documented through a paired examination of patient 3-D images and CT scans. CT Haller Index was calculated for all cases. Thorax shapes were 3-D scanned and analyzed according to the presented method. Statistical analysis confirms relationship between Haller Index and (I_{3ds}) index for the studied cases (significance level $\alpha = 0.05$ and Pearson's correlation coefficient $R = 0.93$).

A comparison of slices acquired by means of CT and 3-D optical scan for two exemplary patients is presented in the following figures. The trunk contour measured with the 3-D scanner is presented in red color. (Fig 17.14)

There are some noticeable differences between body contours in slices captured with CT and 3-D optical scan. The slices acquired with use of the 3-D scanning system are up to 10 mm narrower in the transverse direction of the patient at the same time being up to 10 mm bigger in the anteroposterior direction. The reason for this is a difference in body position taken during CT procedure (reclined) and 3-D optical scanning (suppine). The mentioned shape differences result from gravity acting on patient's body.

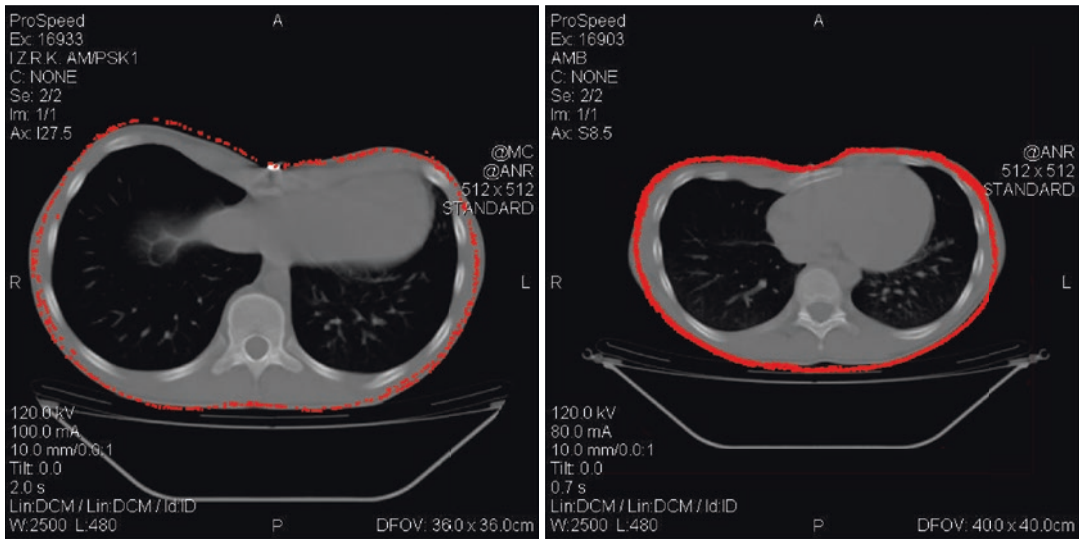

Fig. 17.14 The comparison of CT images (grayscale images) and corresponding slices captured with the 3-D optical scanner (red contours) for Patient 1 (a) and Patient 2 (b)

Conclusion

The chapter presents an automatic method for an optical three-dimensional markerless shape measurement and assessment of pectus excavatum severity. The measurement system utilizes structured light projection method (Temporal Phase Shifting and modified Gray codes) to acquire a highly accurate 3-D model of patient's body surface. The measurement process has been completely automated and does not require the user input in any form.

The 3-D shape of patient's torso is analyzed in order to localize the most pronounced depression. The characteristic slice is described by a 3-D scan index (I_{3ds}) that is a quantitative measure of the deformation. The studies confirm a positive correlation between (I_{3ds}) and Haller Index.

Localizing the characteristic slice as well as calculating the index value is performed automatically without user intervention. In effect the PE assessment method presented in this chapter is an objective one.

The measurement method based on light projection is completely safe for the patient and may be performed repeatedly (pre- and post-operatively) not posing a threat to the patient's health.

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Breast and Chest Wall Asymmetries

18

Allen Gabriel, Craig N. Creasman, and G. Patrick Maxwell

Introduction

Following the introduction of the silicone gel prosthesis in 1962, breast augmentation has become one of the most frequently performed operations in plastic surgery [1, 2]. It is estimated that more than 3 % of the adult female population in the United States (between 2 and 4 million) has undergone breast augmentation [3]. Recent data shows that breast augmentation still remained as the most popular aesthetic surgical procedure in 2012. As we strive to minimize reoperation rates in our breast surgery patients, identifying asymmetries preoperatively will aid in having a more concise operative plan. This can also aid in more appropriate and improved pre-operative consultations with the patient.

With any type of breast surgery, whether performing an augmentation, reduction, reconstruction, or revisionary surgery, our goal is to create a symmetrical and aesthetically pleasing breast form. Clearly achieving these goals entail developing a breast mound that is proportional to the body and importantly a nipple that is located at the most anteriorly projecting portion of the breast which has minimal ptosis and is teardrop in shape [4].

It is also important to keep in mind that each patient will have her own perspective of what an aesthetically pleasing breast may be and will have different end-result expectations as the surgeon, some of which may not be achievable. Therefore we work on using different primitive techniques to help the patient visualize what her breast form may look like under clothing and therefore quantify the volume necessary to achieve the desired goal. In 1981, Brody had patients choose brassieres with well-formed cups and pad them appropriately until they arrived at a size that suited the image they were trying to obtain and volume was then assessed with water filled bags [5]. This did account for identifying some of the gross asymmetries present but subtle asymmetries were difficult to obtain.

Other plastic surgeons have tried using cookbook calculations to determine the volume required to achieve aesthetic breasts [6–8]. Each surgeon uses their own technique of assessment for identifying pre-operative asymmetries and it

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has become clear that selecting an implant for a patient can be very complicated [9, 10]. Preoperative planning worksheets with questions and measurements to achieve the appropriate volume for a specific type of implant have also had their limitations [11, 12].

Identifying chest wall asymmetries has also been difficult unless it's a major deformity that can easily be identified by the naked eye. It is worth noting that in a study performed to evaluate breast asymmetries, the soft tissue evaluations between the examiners was consistent statistically. However, there was inconsistent identification of chest wall asymmetries between the evaluators making it a statistically insignificant finding [4]. This confirms that chest wall asymmetries can be difficult to visualize and many believe that the incident rate of chest wall asymmetries is low and therefore not be noticed. In a recent study, Gabriel et al evaluated the incidence of breast and chest wall asymmetries in healthy females with advanced 4D photography [13]. Overall, 100% of the women had some degree of asymmetry (soft tissue and/or chest wall) confirmed by 4D photography [13]. These findings underscore the importance of developing a systematic preoperative breast and chest wall analysis that can be individualized for each patient. The resulting asymmetries should then be discussed with the patient, along with the potential for continued or even more pronounced asymmetry postoperatively [14].

Anatomy

An interesting finding of the asymmetry study showed that left chest wall had increased convexity in majority of the cases [13]. An example of this is shown in Fig. 18.1c. This anatomical deformity is majority times only seen if the patient is supine and is being evaluated from a worm's eye view. One may wonder why the left side is more convex and why the right side of the chest has more of a downward slope. Even though this has not been described to date, one simple explanation is that embroyolgially the heart is developed and beating strongly by 6 weeks which will have an effect on the shape of the ribs as they develop on the left.

The 12 paired ribs develop from cartilaginous costal processes of the maturing thoracic vertebrae with rib development beginning at 9 weeks. This is followed by secondary ossification centers appear at 15 years [15]. The first seven "true" ribs connect to the sternum via the costal cartilages by day 45. It is possible that during this time the ribs gain convexity to make room for the developing heart as the ribs are still developing and are malleable to curvature changes.

Rib abnormalities can be divided into congenital and acquired deformities. Not all of the deformities are symmetric and bilateral and therefore extensive asymmetries in the chest wall can be observed in some patients. Some of the more significant deformities that lead to asymmetries include the family of dysplasias that can also lead to respiratory insufficiency, since the abnormal size and length of ribs can diminish the volume of the chest. Clinically abnormal rib shapes that are frequently noticed in patients include Cerebrocostomandibular Syndrome; rib notching, Pectus Excavatum and Pectus Carinatum. Among the acquired conditions that can lead to abnormal rib development include Rickets, hyperparathyroidism. Other unfortunate acquired conditions also include trauma, infection and neoplasms at an early age.

Discussion

Chest wall asymmetries are a common finding if careful evaluation is performed. There are many instances during development and growth that can lead to unilateral changes of the rib cage. This is a significant finding for plastic surgeons that perform elective breast surgeries on women. Elective breast surgeries are on the rise and all

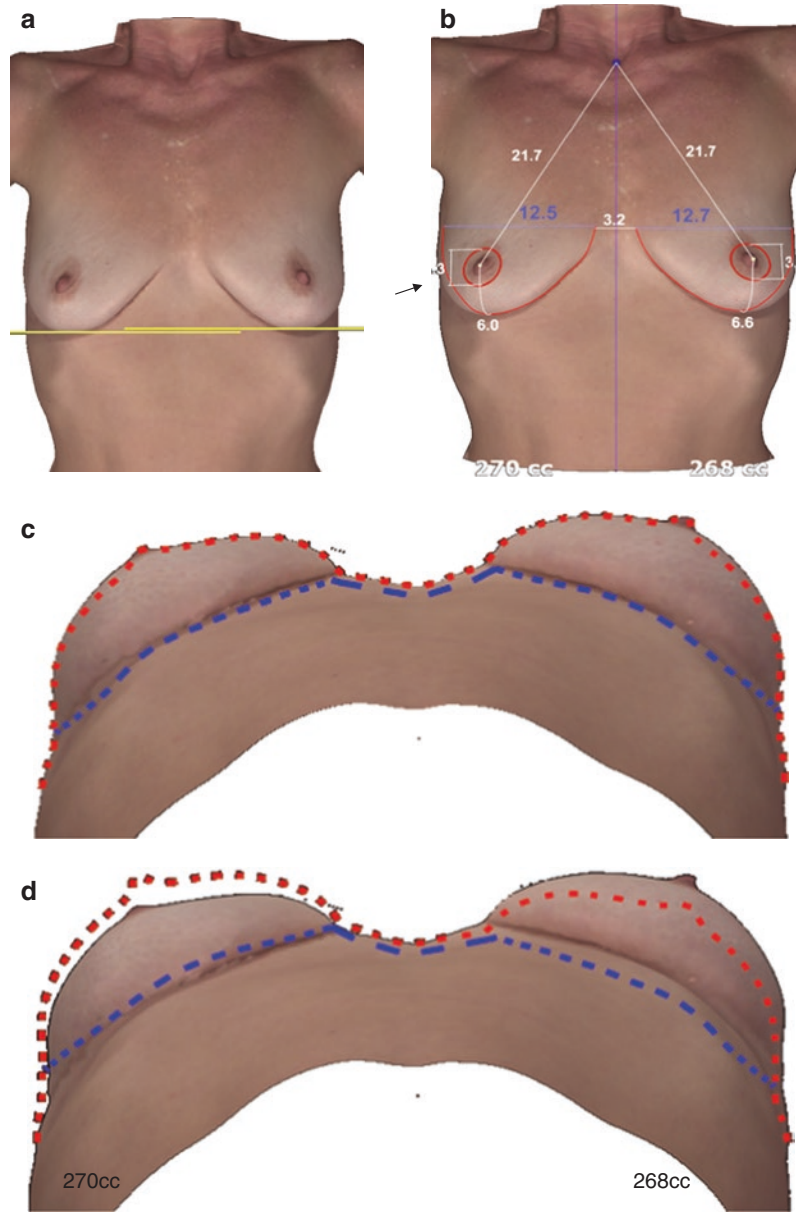

Fig. 18.1 Example of chest wall asymmetry: (**a**) Patient's AP view; (**b**) Patients AP view following automated biodimensional measurements and volume characterization. (**c**) Worm's eye view of patient's chest. Red line delineating the soft tissue envelope, and blue line the chest wall. (**d**) Worm's eye view of patient's chest with superimposed soft tissue and chest wall outlines as mirror images with identification of chest wall asymmetry. Even though the volume is identical the presenting anatomy is very different

efforts are being made to decrease the re-operation rates and increase patient satisfaction with long term aesthetic results. In addition to following sound surgical principles, the importance of identifying breast/chest asymmetries in the preoperative consultation can add to our goal. First, appropriate measurements and identifying both soft tissue and chest wall asymmetries prepares the surgeon to perform an efficient operation with anticipating the outcome. Second, obtaining a clear patient consent and explaining the underlying asymmetries that are present by getting the patient involved in decision making process and understanding their goals and desires,

achieves a happy patient and good outcome as the end result.

There is evidence now that all breast forms have soft tissue, chest wall asymmetries or both. Knowing preoperatively, the extend and cause of the asymmetry (chest wall or soft tissue), it will guide the plastic surgeons in choosing appropriate techniques and implants [13]. Biodimensional principles remain the gold standard for selecting the appropriate implant. A recent survey showed that limited number of plastic surgeons used the base width as one of their major determinants to choose an implant [16].

Breast surgery can be as one of the most demanding operations. For many years, plastic surgeons have had tools to assess the soft tissue envelope of the breast but as always it has been very subjective in terms of assessing the chest wall. We all rely on our artistic eye and judgment in the operating room and utilize multiple seizers to achieve symmetrically breasts. It is clear that most likely our operative time may also decrease if all aspects of the asymmetries have been identified and documented preoperatively, since the surgeon will have a better understanding of what type and/or volume of implant is needed to achieve an aesthetically symmetrical breast.

Appropriate and thorough preoperative evaluation will allow the surgeon to select and plan a suitable operation. Choosing the proper technique begins with designing incisions based on scar placement and length. Minimizing scar appearance is fundamental to any operation in plastic surgery. However, scars should not be avoided if they are necessary to provide adequate and durable results. A balance must be accomplished between scar placement and efficacy, as the final result will depend on the harmony of the breast shape and scar appearance.

Knowing that all patients have some degree of asymmetry, looking for these asymmetries and identifying them in the preoperative evaluation is very important to have a happy patient postoperatively. Implants magnify asymmetries and patients may notice them more postoperatively, even though they were present all their adult life.

Using an advanced photography tool in breast surgery consultation, eliminates the subjective factor of evaluating the chest wall asymmetry and identifying the existing volume of each breast (Figs. 18.1, 18.2 and 18.3). The figures show three different scenarios of asymmetries encountered in our clinical practice with the most common being the combination of both (Figs. 18.1, 18.2 and 18.3).

Appropriate preoperative counseling with each patient can eliminate the majority of postoperative complaints. By explaining that breast asymmetries are the rule rather than the exception and that subtle differences preoperatively may be more obvious after breast augmentation, patients will have a more realistic expectation for their final result.

Knowing that 100% of patients have asymmetries present in one or more of the dimensions, involving chest wall and soft tissue, more diligence should be applied to the pre-operative evaluation to identify these asymmetries and communicating them to the patient.

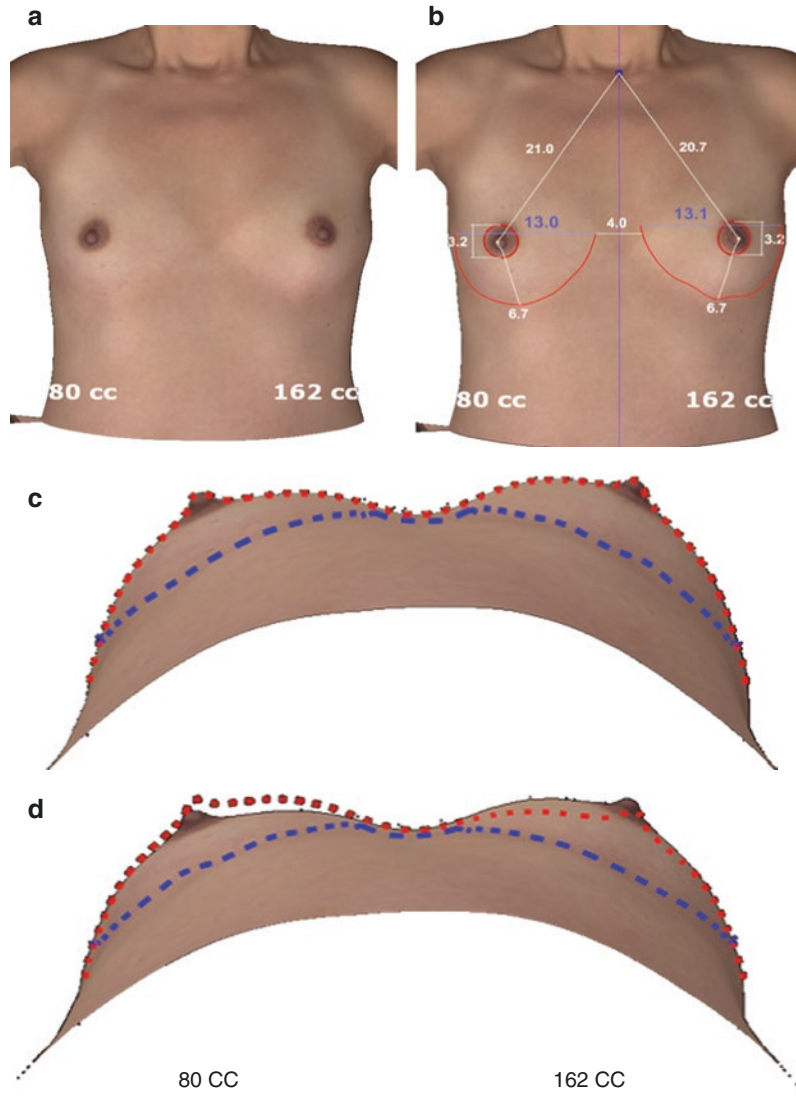

Fig. 18.2 Example of soft tissue asymmetry: (**a**) Patient's AP view; (**b**) Patients AP view following automated biodimensional measurements and volume characterization. (**c**) Worm's eye view of patient's chest. Red line delineating the soft tissue envelope, and blue line the chest wall. (**d**) Worm's eye view of patient's chest with superimposed soft tissue and chest wall outlines as mirror images with identification of soft tissue asymmetry

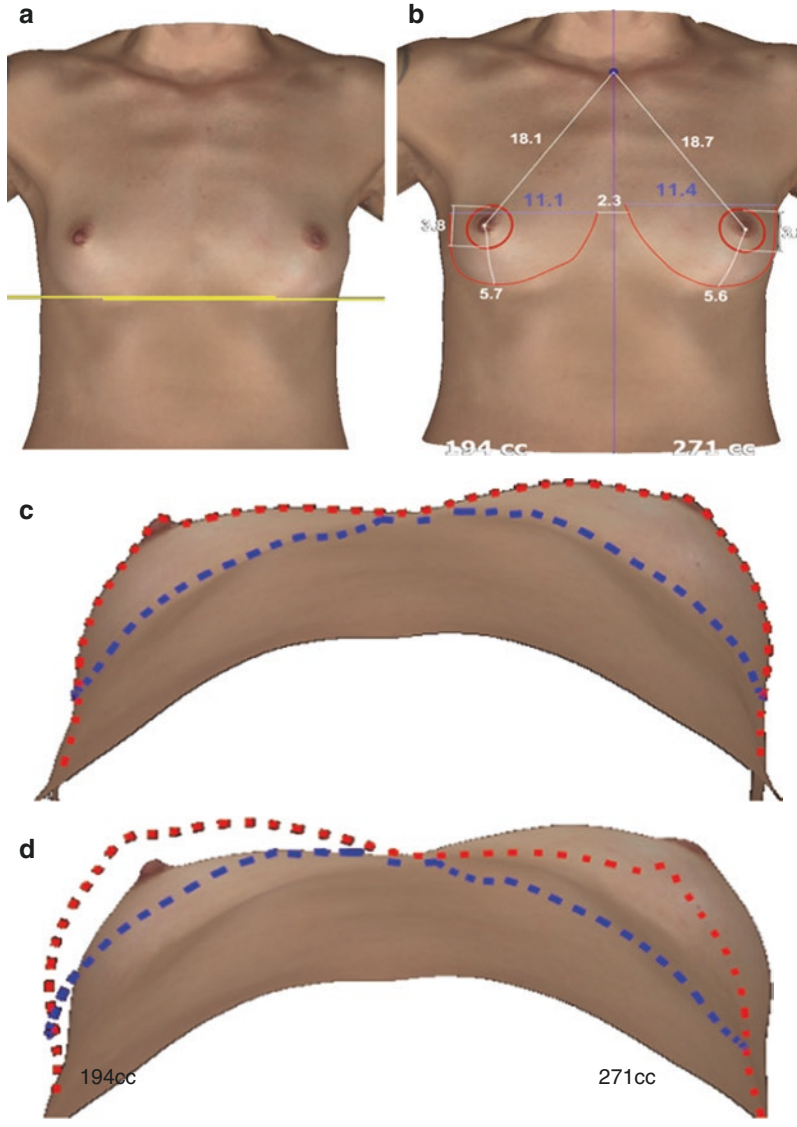

Fig. 18.3 Example of soft tissue and chest wall asymmetries: (**a**) Patient's AP view; (**b**) Patients AP view following automated biodimensional measurements and volume characterization. (**c**) Worm's eye view of patient's chest. Red line delineating the soft tissue envelope, and blue line the chest wall. (**d**) Worm's eye view of patient's chest with superimposed soft tissue and chest wall outlines as mirror images with identification of soft tissue and chest wall asymmetries

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Exercise Performance Testing in Pectus Excavatum Patients

19

Christoph Castellani, Jana Windhaber, and Peter H. Schober

Introduction

Psychological problems with disturbed body perception render a large proportion of patients with pectus excavatum to complain about reduced exercise capacity [1–3]. A series of 152 patients that were investigated in a report demonstrated 51 % to be associated with reduced exercise tolerance, 43 % with fatigue and 32 % complained about shortness of breath on exertion [4]. The impression of the mediastinum by the sternal grove possibly in combination with a plathythorax may be pathologies leading to symptoms described above. Compression of the mediastinum may lead to reduced venous backflow to the heart and thus reduced preload. Doppler examinations of pectus patients have shown increased supine to upright stroke volumes [3] and a lower cardiac index [5] when compared to healthy controls. In contrast, it has also been reported that no significant differences of ejection fraction and fractional shortening are observed when pectus patients are compared to healthy controls [6].

Restriction of space and thorax excursions by the pectus excavatum may lead to a restrictive pulmonary disease with reduced vital capacity (VC) and total lung capacity (TLC). Regarding lung function tests in pectus patients, there is a general agreement that vital capacity (VC) and forced expiratory volume in 1 s (FEV_1) is reduced [1, 2, 4, 7] although interpretation of this data is debateable. As possible result of these data, pectus patients were found to have significantly reduced oxygen uptake (VO_2) and physical work capacity in exercise performance testing [3, 5, 8, 9].

Although reports demonstrate pathologies in lung function, cardiac ultrasound and performance testing, there seem to be discrepancies between the patients' subjective description and the results of investigations [5, 10]. When treating pectus patients lung function and exercise performance tests help to detect possible pathologies behind the symptoms described by the patient and additionally allow for postoperative monitoring.

Test Modalities

At our centre, lung function tests by spirometry and exercise performance tests such as spiroergometry are routinely performed preoperatively, 3–6 months after the operation (with the Pectus bar *in-situ*) and > 3 months after Pectus Bar removal. Additionally, body fat is calculated by Calliper method and body mass index is recorded. The patients are questioned about sports activities, training frequencies and for occurrence of symptoms during exercise (chest pain, shortness of breath, reduced capacity).

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Spirometry

Spirometry is the most common test in assessing lung function. A flow sensor is used to measure the inspiratory and expiratory flow of the patient optimally at rest in a sitting position (Fig. 19.1). These parameters are used to calculate lung volumes such as vital capacity (VC), forced expiratory volume in one second (FEV_1), the Tiffenau index (FEV_1/VC), the peak expiratory flow (PEF) and the mean expiratory flow at 25 and 50 % of vital capacity (MEF 25 and MEF 50). Especially in children and adolescents, the test results have to be scaled to obtain comparable results. Usually predicted normal values (scaled for age, gender, weight and height) are programmed in the software and used to express test results as percentages of the predicted value.

$$Result_{(\%predicted)} = \frac{Result_{measured}}{Result_{expected}} \cdot 100$$

The following paragraphs give a short overview of the most common lung volumes:

- **Vital Capacity [Inspiratory Vital Capacity (IVC)]:** This is the volume which can be expired after maximal inspiration (Fig. 19.2). The expected normal value (in litres) is calculated by the equation given below:

$$VC_{expected} = \frac{h^3}{f} \cdot \left(1.03 - \frac{age_{years} - 25}{100} \cdot 0.75\right)$$

Where h is the height in metres and f is a correction factor ($f_{male} = 1.0$; $f_{female} = 1.1$).

- **The Forced Expiratory Volume in one second (FEV_1)** is the amount of air a patient can exhale in one second of maximal expiration. FEV_1 is a measure for the diameter of the intrathoracic (small) airways. It is most commonly expressed as Tiffenau index (TI).

$$TI = \frac{FEV_1}{VC} \cdot 100$$

Generally TI is > 75 %; a reduction of TI is considered as a sign of obstruction of the smaller airways (Asthma, Chronic Obstructive Pulmonary Disease-COPD).

- **The Peak Expiratory Flow (PEF)** and the **Mean Expiratory Flow at 25 and 50 % of VC (MEF25 and MEF50)** give information about the transport capacity of the large airways.

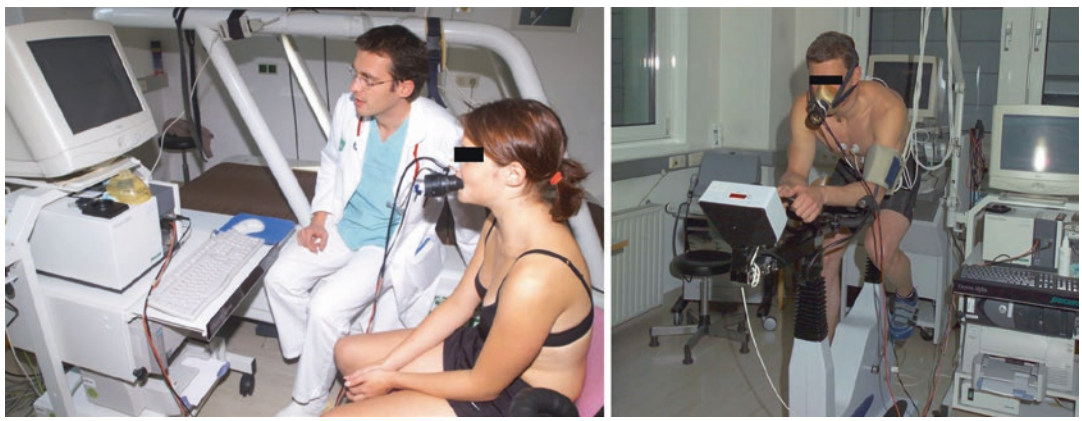

Fig. 19.1 Clinical setting for spirometry investigation in a patient with pectus deformity (*left*) and exercise performance (*right*)

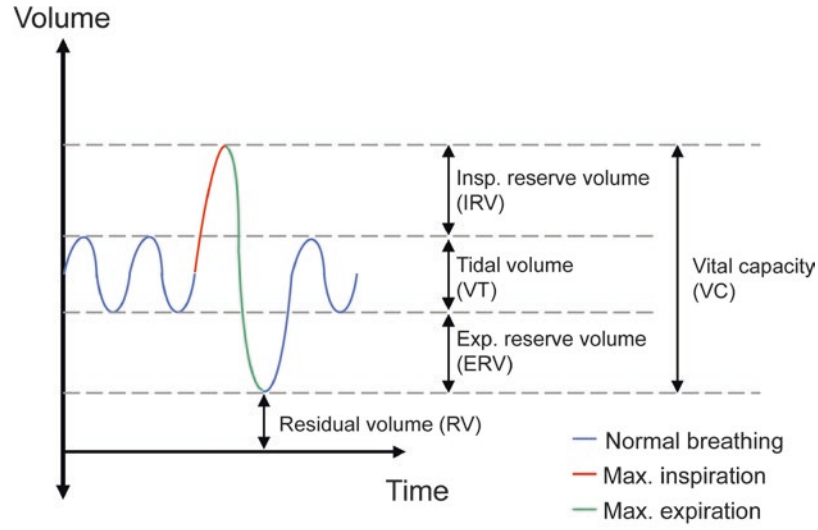

Fig. 19.2 Lung volumes at spirometry testing

The biggest advantages of spirometry testing are its availability, ease in performing the tests, low costs and the possibility to combine spirometry with ergometry. Disadvantages are that cooperation of the patient is essential to obtain reliable test results. Additionally, certain lung volumes such as total lung capacity (TLC) and residual volume (RV) cannot be detected by spirometry and require investigations through body plethysmography (see below).

Body Plethysmography

By measuring changes in volume and pressure in a closed system, this test allows to determine the total lung capacity and the residual volume additionally to the parameters described in spirometry. Although, more precise than spirometry, technical difficulties and costs for this method are higher than for conventional spirometry; and the use of the additional information gained is questionable. Hence, conventional spirometry for lung function testing is the preferred modality of investigation.

Overview of Results of Lung Function Tests

Pre-operative examinations performed in a series of 307 patients reported, demonstrated a reduced FEV1, VC, MEF25 and MEF50 in pectus patients compared to normal expected values. A body plethysmography in the same collective also proved reduced TLC and elevated RV [11]. In another large series reported on 408 patients, almost similar results were found, however with significantly lower VC and FEV_1 compared to expected values [7] (Table. 19.1).

In a smaller series of 21 patients, significantly reduced VC and FEV_1 in pectus excavatum patients compared to expected values were demonstrated; with TLC and RV however demonstrating no significant changes [8]. Normally one would expect reduced VC as a sign for restriction in a pectus excavatum patient to go proportionately along with reduced TLC and elevated RV. A possible reason for this finding may be a lack of patient motivation in spirometry testing which underlines the necessity of good patient cooperation during spirometry.

In contrast to these results, another study showed significantly reduced VC but no

Table 19.1 Spiroergometry in pectus excavatum patients according to the data from the literature

Author	Patients (N)	$VC_{\%predicted}$	$FEV_{1\ \%predicted}$	$TLC_{\%predicted}$	$RV_{\%predicted}$	$PEF_{\%predicted}$
Kelly RE et al. [11]*	307	90 (<0.0001)	89 (<0.0001)	94 (<0.0001)	111 (<0.0001)	–
Kubiak R et al. [12]**	15	64 (0.035)	63 (0.035)	83 (0.302)	–	–
Tang M et al. [6]*	49	96 (ns)	87 (<0.001)	–	–	81 (ns)
Lawson ML et al. [7]*	407	87 (<0.0001)	86 (<0.0001)	–	–	–
Malek MH et al. [8]*	21	86 (<0.0001)	87 (<0.01)	92 (<0.0001)	108 (ns)	–
Jiang X et al. [13]*	27	92 (0.01)	82 (0.0007)	111 (ns)	166 (0.0008)	–

Data displayed as mean (*) or median (**); p-values for the comparison to expected values are displayed in round brackets; *ns* = not significant

differences for PEF, FEV1 and TI in pre-operative examinations of pectus excavatum patients compared to healthy controls [6]. Data from selected studies is displayed in Table. 19.2.

Based on the assumption that reduced VC and TLC is a result of chest size restriction in pectus excavatum patients; leads to the hypothesis that these parameters might be related to pectus severity indices. However, several smaller series could not demonstrate correlations between parameters of lung function and depth of the pectus excavatum [10, 12]. In an investigation of 327 pectus patients in which lung function tests were performed, the patients were divided between those with a Haller Index above and below the median. In these patients, a correlation of FEV1, VC, TI and TLC could be demonstrated, but this correlation could not be shown for residual volume and the functional residual capacity when compared to the severity of pectus excavatum [14].

Exercise Performance Tests

In pectus excavatum patients, our preference is to perform a bicycle spiroergometry with a gender and body-weight adapted setting (Table. 19.2).

In order to obtain comparable results in spiroergometry certain criteria have to be observed:

- Consumption of a normal diet by the patient
- No physical training within the last 48 h
- No competitive sport participation/hard (exhaustive) training within the last 2 weeks
- Good health and general condition
- Optimal ventilation of the room to grant inspiratory oxygen content of 21 %
- Optimal room temperature (20–22° Celsius)

The spiroergometry testing is performed after taking the medical history, collection of anthropometric data and taking a spirometry at rest. For the spiroergometry test, the patients are required to breathe through a mask allowing flow measurements and determination of the expired CO_2. Additionally, an electrocardiogram (ECG) is continuously recorded. These parameters allow determining the performance capacity, maximum oxygen uptake per kilogram bodyweight ($VO_{2max/kg}$) and oxygen pulse

- **Oxygen uptake (VO_2):** is a measure for the capacity of an individual to transport and use oxygen during exercise and thus for the aerobic capacity of the patient. Typically the absolute oxygen uptake (VO_{2max} in l/min) increases with growth. To relativize the test results in adolescents the oxygen uptake is usually related to body weight ($VO_{2max/kg}$ in ml/kg/min).
- **Oxygen pulse:** describes the oxygen uptake per heartbeat and gives information about cardiac output and cardiac reserve.

It is important to ascertain the cooperation of the patient during all these tests. Capillary lactate

Table 19.2 Body weight adapted ergometry settings used in our centre

Body weight (kg)	Start at (W)	Steps (W)	Step duration (min)
25–30	15	15	2
31–35	20	20	2
36–40	25	25	3
41–45	30	30	3
46–50	35	35	3
51–55	40	35	3
56–60	45	40	3
>60 male	50	50	3
>60 female	50	40	3

tests allow objectifying symptoms such as breathlessness or exhaustion as described by the patients during the test. Lactate levels >5 mmol/l are signs for a cooperative patient and reliable test results.

Overview of Results of Exercise Performance Tests

Overall there is scarce data which is limited to small series of patients with regards to the comparison of exercise performance test results in pectus excavatum patients with expected the values or values in control patients. Furthermore, the parameters examined are heterogenic which making comparisons of different reported results extremely complex. As all test parameters strongly depend on the training condition of the patients and the state of training can hardly be quantified between pectus excavatum patients and healthy controls; comparisons are difficult to interpret. A further reported investigation performed on 20 patients demonstrated significantly reduced maximum oxygen uptake (VO_{2max}) when compared to expected values, but no differences for $VO_{2max/kg}$ [8]. Similarly another reported study showed no significant differences in $VO_{2max/kg}$ for pectus excavatum patients (n = 49) when compared to healthy controls (n = 26) [6]. Another study demonstrated significantly lower work rate, VO_2 and O_2-pulse in pectus excavatum patients (n = 13) also compared to healthy controls (n = 20) [3]. Overall it remains unclear if reduced exercise tolerance depends upon the physical condition itself or a lack of training. Regarding the pathologic self-perception and self-esteem of pectus excavtum patients with many adolescents reporting problems with undressing in group sports or at the swimming pool; a lack of training may well be attributed to the socio-psychological side effect of pectus excavatum.

Lung Function and Exercise Performance After Surgical Correction of Pectus Excavatum

Lung Function

Regarding lung function tests the reports in the literature are contradictive. For the Minimal Access Repair of Pectus Excavatum (MARPE) method, most Centres agree that a temporary significant decrease of VC is observed in these patients as long as the Pectus bar is *in-situ* [10, 15, 16]. Restriction of thorax movements is the most likely explanation for this finding (Table. 19.3).

After Pectus bar removal, the data is however inconsistent with some Centres reporting improvements (compared to pre-operative levels) [7, 12] where others have found no significant differences [9, 15, 16]. Similarly no significant improvements could be demonstrated for the FRC [15]. Available data for open pectus repairs is limited to small series with none of the Centers demonstrating significant improvements of VC [2, 17].

Similarly to VC results for FEV1 are controversial with some Centres reporting significant improvements [7, 9, 12] and others who could not show any significant differences [15, 17]. The reason for the discrepancies between different studies remains unclear at present.

Exercise Performance Tests

In these tests, the most commonly examined parameter is the oxygen uptake per kilogram body weight and minute ($VO_{2/kg}$). Similar to

Table 19.3 Selected data for changes of lung function by surgical correction (MARPE; RA = Ravitch method) of pectus excavatum in the literature

	$FRC_{\%predicted}$			$VC_{\%predicted}$			$FEV_{1\ \%predicted}$		
Author (method)	Pre-OP	With bar	Post-OP[a]	Pre-OP	With bar	Post-OP[a]	Pre-OP	With bar	Post-OP[a]
Aronson DC et al. [15] (MARPE)	93 (111)	96[b] (111)	96[d] (53)	90 (111)	86[b] (111)	89[d] (53)	95 (111)	93[b] (111)	95[d] (53)
Bawazir OA et al. [16] (MARPE)	–	–	–	93 (47)	76[b] (32)	87[d] (10)	79 (47)	70[b] (37)	80[c] (10)
Kubiak R et al. [12] (MARPE)	–	–	–	64 (15)	–	81[b] (15)	63 (15)	–	85[b] (15)
Lawson ML et al. [7] (MARPE)	–	–	–	85 (45)	–	90[b] (45)	84 (45)	–	89[b] (45)
Sigalet DL et al. [9] (MARPE)	–	–	–	91 (26)	–	93[d] (26)	78 (26)	–	84[b] (26)
Quigley PM et al. [2] (RA)	–	–	–	81 (15)	80[d] (15)	–	–	–	–
Kowalewski J et al. [17] (RA)	–	–	–	85 (22)	–	88[d] (22)	78 (22)	–	82[d] (22)

[a]Post-OP refers to results (iv available) after removal of metal struts. Data displayed as mean, numbers of patients evaluated are given in round brackets
[b]Difference to Pre-OP significant
[c]Difference to measurement with bar significant
[d]Not significant

respiratory parameters data reported is not unanimous and studies have focused on MARPE with little data available for open repair procedures. In the first report on Exercise performance tests, a significant postoperative (Pectus bar *in situ*) decrease of the mean VO_2 from 36 to 29 ml/kg/min was reported [18]. Contrary to this a report from our Centre demonstrated no differences [10] while another report demonstrated improvements [6] (Table. 19.4).

In another investigation comparing preoperative measurements to tests after Pectus bar removal no significant differences were reported [9], however this could not be confirmed in our patients where a significant decrease was observed [10]. Significant improvements could however be demonstrated with regards to oxygen pulse and VO_{2max} in litres [9, 16]. Thus it seems unlikely that discrepancies are direct causes of the operation and it is more likely that changes in training frequencies and/or intensities are the underlying cause.

Results from Our Single Centre Cohort

An investigation conducted at our Center involved 46 patients with pectus excavatum [10]; in which all patients underwent the MARPE. Exercise performance testing and lung function were performed pre-operatively, 6–12 months after operation with the Pectus bar *in situ* and within 3 years after Pectus bar removal. In exercise performance tests, a significant decrease of the maximum oxygen uptake (ml/kg/min) and the performance capacity was evident (Table. 19.5). As described above, the operative procedure was put into question as reason for these changes. Most of our patients reported changes in their training habits

Table 19.4 Selected data for changes of exercise performance by surgical correction (MARPE; RA = Ravitch method) of pectus excavatum in the literature

	$VO_{2max(l)}$			O_2-Pulse$_{\%predicted}$			$VO_{2/kg}$		
Author (method)	Pre-OP	With bar	Post-OP[a]	Pre-OP	With bar	Post-OP[a]	Pre-OP	With bar	Post-OP[a]
Bawazir OA et al. [16] (MARPE)	1.63 (40)	1.39[b] (30)	2.06[b, c] [10]	–	–	–	–	–	–
Sigalet DL et al. [9] (MARPE)	1.70 (26)	–	2.08[b] (23)	77 (26)	–	83[b] (23)	34 (26)	–	36[d] (23)
Castellani C et al. [10] (MARPE)	–	–	–	–	–	–	44 (43)	43[d] (42)	42[b] (42)
Tang M et al. [6] (MARPE)	–	–	–	–	–	–	26 (38)	28[b] (38)	–
Quigley PM et al. [2] (RA)	–	–	–	–	–	–	40 (5)	40[d] (15)	–

[a]Post-OP refers to results (iv available) after removal of metal struts. Numbers of patients evaluated are given in round brackets
[b]Difference to Pre-OP significant
[c]Difference to measurement with bar significant
[d]Not significant

Table 19.5 Results of the authors' investigation [10]

Time	N	Pmax/kg (Watt)	VO_{2max} (ml/kgBW/min)	VO_{2max} (ml/kgLBW/min)	$PC_{\%expected}$
Pre-OP	43	3.3 ± 0.5	43.8 ± 6.5	49.8 ± 5.5	105 ± 28
With strut	42	3.3 ± 0.6	42.6 ± 6.9	49.3 ± 6.9	106 ± 25
After removal	42	3.3 ± 0.7	42.2 ± 7.2[a]	49.8 ± 7.3	97 ± 33[a]

[a]Significant difference to pre-operative test results
PC performance capacity, *Pmax* maximum performance, *BW* body weight, *LBW* lean body weight

suggesting changes in physical conditioning. Regarding the antropometric data of our patients, a significant increase in body mass index and body fat mass could be demonstrated. When relating VO_{2max} to the lean body weight (body weight minus body fat) there were no significant differences throughout the investigation, pointing towards a change of life style and training habits as main cause for the changed findings in exercise performance tests.

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Correlating Haller Index and Cardiopulmonary Disease in Pectus Excavatum

20

Meera Kotagal, Jordan W. Swanson, and Jeffrey R. Avansino

Abbreviations

CT	Computed tomography
FEF	Forced expiratory flow
FEV1	Forced expiratory volume in one second
FVC	Forced vital capacity
HI	Haller Index
LVEF	Left ventricular ejection fraction
PFTs	Pulmonary function tests
PGI	Pectus gracilis index
RV	Right ventricle
TLC	Total lung capacity

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Introduction

Patients with pectus excavatum frequently attribute symptoms such as exercise intolerance, early fatigability, and dyspnea on exertion to their chest wall deformity [1]. While multiple studies have associated pectus excavatum with poor quality of life measures and subjective complaints [2, 3], direct links between the chest wall deformity, patient symptoms and concrete evidence of cardiorespiratory dysfunction have been more difficult to elucidate. It is unclear to what extent a chest wall deformity impacts exercise tolerance.

Understanding the physiologic ramifications of pectus excavatum is essential for identifying appropriate indications and candidates for surgical repair. A conventional belief was that while severe pectus deformities may cause physiologic impairment, the majority of deformities did not, at least to the degree that such impairment could be objectively measured. This resulted in the critique that undertaking repair of pectus excavatum deformities amounted to a cosmetic procedure. However, many experienced practitioners report anecdotal evidence of exercise limitation and subsequent improvement in exercise tolerance after repair across the pectus deformity spectrum [4–6]. As a result, there has been increasing emphasis on standardizing measurement of both deformity severity and physiologic impairment.

The Haller Index (HI), or pectus index, was proposed by Haller et al. in 1987 as an objective

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measure of pectus excavatum severity [7]. The HI is the ratio of transverse to anteroposterior chest wall diameters, determined from a single axial computed tomography (CT) scan at the point of maximal pectus deformity. In their study of 33 patients selected for surgical pectus repair and 19 age-matched control patients, Haller et al. found that all of the surgical cases but none of the controls had a HI exceeding 3.25, suggesting HI as an objective criterion for pectus excavatum repair. This chapter aims to review the existing literature on correlating cardiopulmonary disease with the Haller Index in patients with pectus excavatum.

Pulmonary Disease

There is relatively strong evidence in the literature associating pulmonary dysfunction with pectus excavatum. The majority of studies demonstrate that pulmonary dysfunction in these patients is restrictive in nature [8]. Both Lawson and Kelly found that patients with pectus excavatum have reduced forced vital capacity (FVC), forced expiratory volume in one second (FEV1) and forced expiratory flow (FEF) [9, 10].

Multiple studies have demonstrated a correlation between HI and pulmonary dysfunction. Quigley et al. found that patients with pectus excavatum had a lower FVC than controls, and that abnormalities in the total lung capacity (TLC) correlated best with the HI [3]. Lawson found that the percentage of patients with abnormal pulmonary function tests (PFTs) – FVC, FEF 25–75 %, TLC – increased as the HI increased [11] (Fig. 20.1). Our group found that patients with pulmonary (or combined cardiopulmonary dysfunction) had a significantly higher HI than patients without cardiopulmonary limitation [12] (Fig. 20.2). Not all studies, however, have demonstrated correlation between pulmonary disease and severity of pectus deformity, as determined by the HI. Morshuis et al. found that pulmonary function follows a restrictive pattern in pectus excavatum patients, but that it is not related to age, severity of deformity or pulmonary symptoms [6].

While the evidence correlating pulmonary dysfunction with pectus excavatum severity is relatively strong, analysis of the physiologic impact of surgical repair has not been as positive. Multiple studies have demonstrated that pulmonary function is similar, or worse, post-operatively, depending both on timing of repeat evaluation and the type of surgical repair. Kaguroaka et al. demonstrated similar pulmonary function to baseline levels at 42 months post-operatively [13]. Borowitz found no significant differences after Nuss repair, although mean baseline pulmonary function was normal in the patient population preoperatively [14]. Quigley

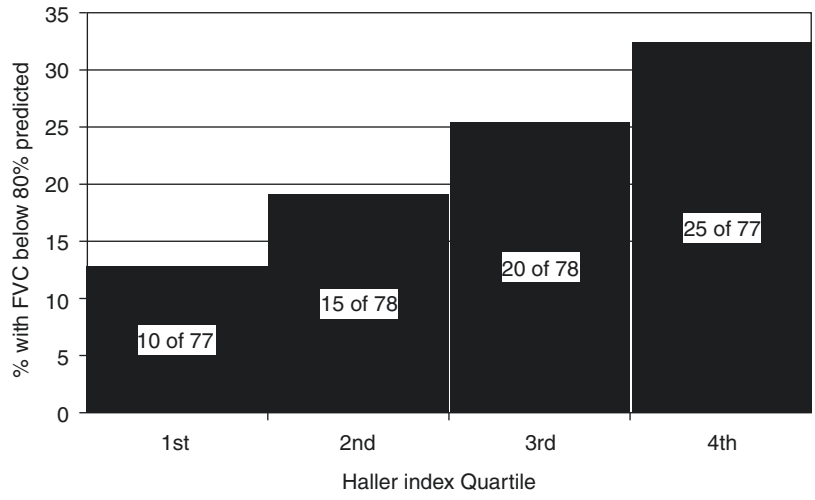

Fig. 20.1 Relative frequency of reduced FVC by Haller Index among 310 North American pectus excavatum patients (Lawson et al. [11]. With permission from Elsevier)

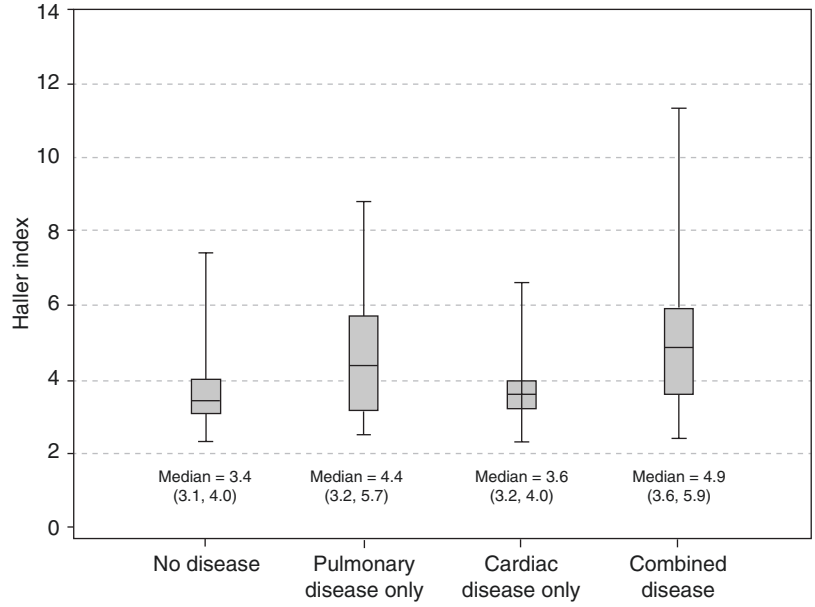

Fig. 20.2 Haller Index differs based on pulmonary function: box plot of HI by disease group (Swanson et al. [12]. With permission from Elsevier)

et al. found no change in FVC post-operatively. A meta-analysis of existing data on the impact of surgical repair on pulmonary function found no change between pre- and post-operative pulmonary function [15] (Fig. 20.3). Some studies have reported that patients actually have worse pulmonary function post-operatively, a fact that has been attributed to pain and decreased chest wall elasticity associated with open repair employing thoracotomy or sternotomy [6, 16]. Despite lack of consensus showing pulmonary benefit from surgical repair with either the Nuss or Ravitch repair, studies have intermittently identified isolated improvements in TLC, FEV1, and other PFTs post-operatively, particularly after Nuss repair and bar removal [9, 17, 18].

From this data we can conclude that while pectus excavatum is often associated with restrictive pulmonary dysfunction, repair of this deformity has not resulted in improved pulmonary function post-operatively.

Cardiac Disease

Efforts to determine whether patients with pectus excavatum have associated cardiac dysfunction have also yielded mixed results. Based on anatomical and physiologic principles, a long-standing hypothesis holds that an excavatum deformity could impact cardiac function by compressing the right ventricle (RV), particularly inhibiting ventricular dilation to increase cardiac output during exercise. However, the data have not universally supported this hypothesis. The majority of studies evaluating cardiac physiology (using exercise testing) have shown no difference between patients with pectus excavatum and normal controls. Quigley et al. found that the maximal heart rate and oxygen pulse (a proxy for stroke volume) did not vary significantly between pectus excavatum patients and controls [3]. Using exercise testing, Ghory et al. found no difference in maximal workload, oxygen consumption, cardiac output and stroke volume between patients with pectus excavatum and healthy controls [19]. Rowland et al. found that pectus excavatum patients had lower resting and submaximal exercise stroke volumes than controls, but there was considerable overlap between the two groups, leaving the clinical significance of the finding unclear [20]. In an effort to correlate cardiac dysfunction with HI, our group examined cardiac function over the spectrum of Haller indices in patients with pectus excavatum. We found no difference in oxygen

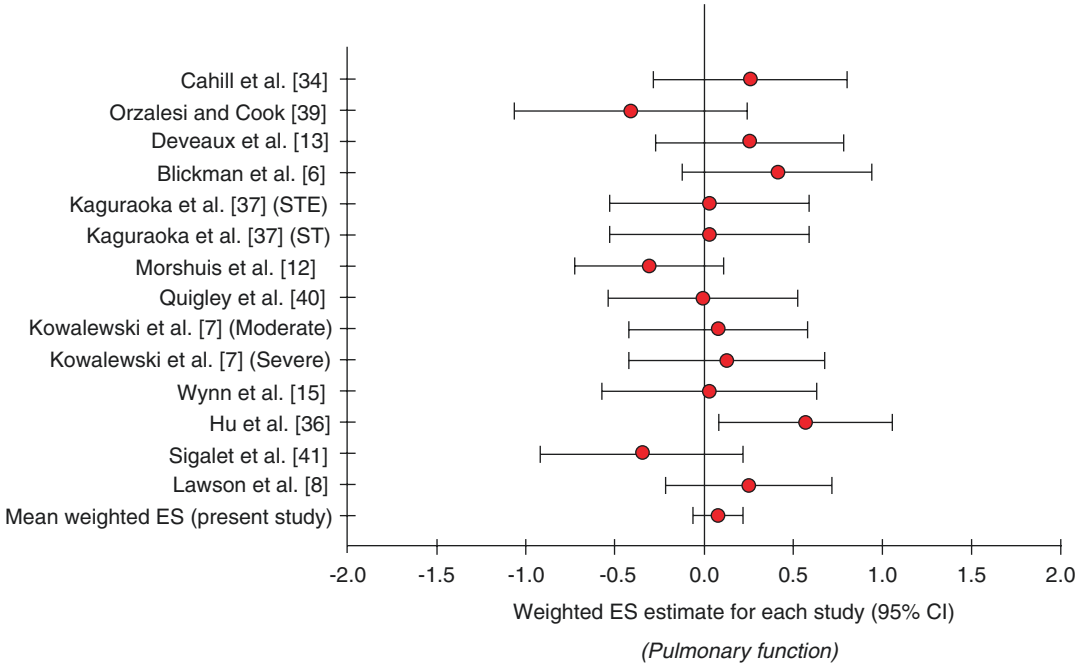

Fig. 20.3 Weighted effect size and 95 % confidence intervals of each study analyzed in the meta-analysis by Malek et al. [15] (With permission from Oxford University Press)

pulse or oxygen consumption between patients with normal or abnormal HI [12] (Fig. 20.4). Of note, oxygen pulse is calculated value based on other variables, and as such does not have its own norms and may be an insensitive measure for mild pathology.

However, when studies have evaluated cardiac mechanics (using echocardiography) pre-operatively, they have found that patients with pectus excavatum have decreased cardiac filling [21], stroke volume [22], and work performance [23]. These differences in cardiac volumes suggest that pectus deformity may result in cardiac compression, even if it does not manifest in decreased cardiac function.

The data on the impact of surgical repair on cardiac function has proven even more controversial. Multiple studies have demonstrated an improvement in cardiac function post-operatively. Quigley et al. found that patients with pectus excavatum had a slight increase in oxygen pulse post-operatively [3]. Krueger et al. found that end-diastolic right ventricular diameter, area, and volume as well as left ventricular ejection fraction (LVEF) increased significantly after surgical repair of pectus excavatum [24]. Kowalewski et al. using echocardiography, found an increase in right ventricular volume post-operatively but no correlation between the change in RV volume and the change in HI between pre and post-operative evaluation. There was, however, a correlation in this study between pre-operative right ventricular volume and the change in HI [25] (Fig. 20.5).

Two meta-analyses have examined this question. The first, by Malek et al. used existing data from eight studies to create a single index for cardiac function. They found that there was a significant improvement in this cardiac function index post-operatively [26]. However, this meta-analysis was critiqued by Guntheroth et al. who subsequently performed their own meta-analysis [27]. In their meta-analysis, they found no increase in left ventricular size, stroke volume or cardiac output post-operatively. An additional study found that stroke volume increased

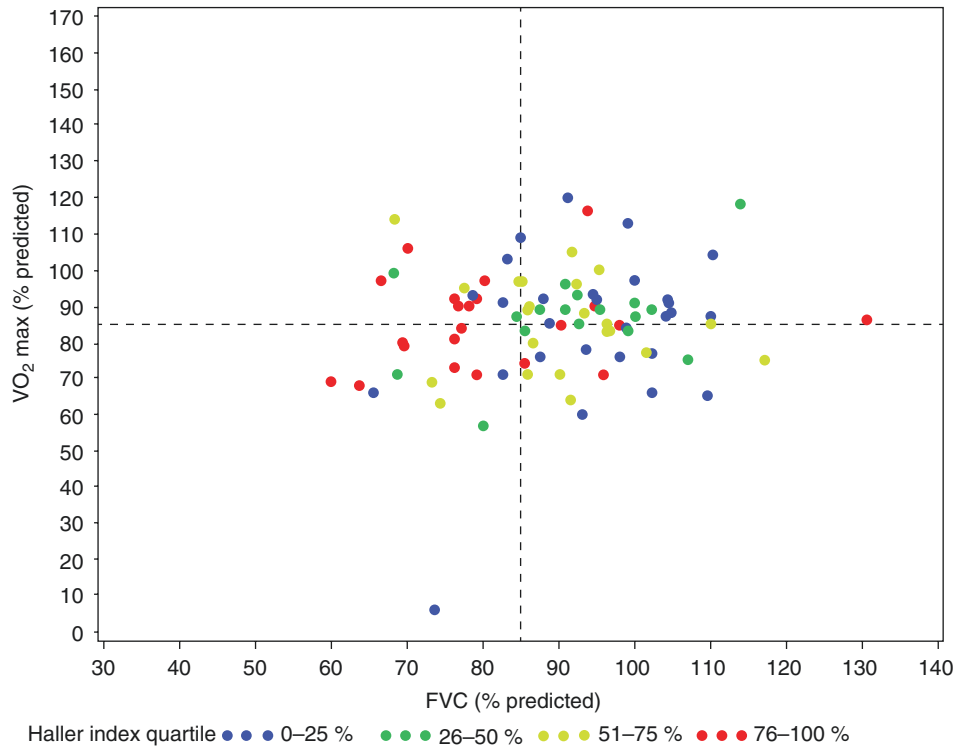

Fig. 20.4 Distribution of Haller Index by cardiac and pulmonary dysfunction. *Dotted reference lines* at 85% predicted denote cut-off values for cardiac and pulmonary dysfunction, such that observations in the *upper right* quadrant show no cardiac or pulmonary dysfunction and observations in the *lower left* quadrant show combined (cardiac and pulmonary) dysfunction (Swanson et al. [12]. With permission from Elsevier)

post-operatively, but when normalized for the growth of (often adolescent) pectus excavatum patients, there was no change in the cardiac index [18]. The lack of normalization of stroke volume in multiple studies has been a critique of this literature [16].

Subjective Symptoms and Exercise Tolerance

In contrast to a paucity of consistent, objective findings of cardiopulmonary dysfunction in pectus patients either at baseline or after surgical repair, there is greater consensus on subjective evidence and reports of exercise intolerance in this patient group. Separate studies consistently found exercise limitation reported by 57–58% of patients with pectus excavatum [3, 25]. Patients frequently report fatigability, shortness of breath with exercise and decreased stamina compared to healthy controls [1]. In addition to describing the presence of symptoms pre-operatively, the literature has widely reported that even in the absence of concrete data demonstrating improvement in cardiorespiratory function, patients report subjective improvement in symptoms and exercise tolerance [5, 6, 18, 25, 28]. Studies have also found that (alongside improvements in subjective exercise ability) patients also report improvements in psychosocial functioning – including body image satisfaction and feelings of frustration and isolation – after repair of pectus deformities [2]. The correlation of subjective symptoms and exercise tolerance with Haller indices has not been evaluated.

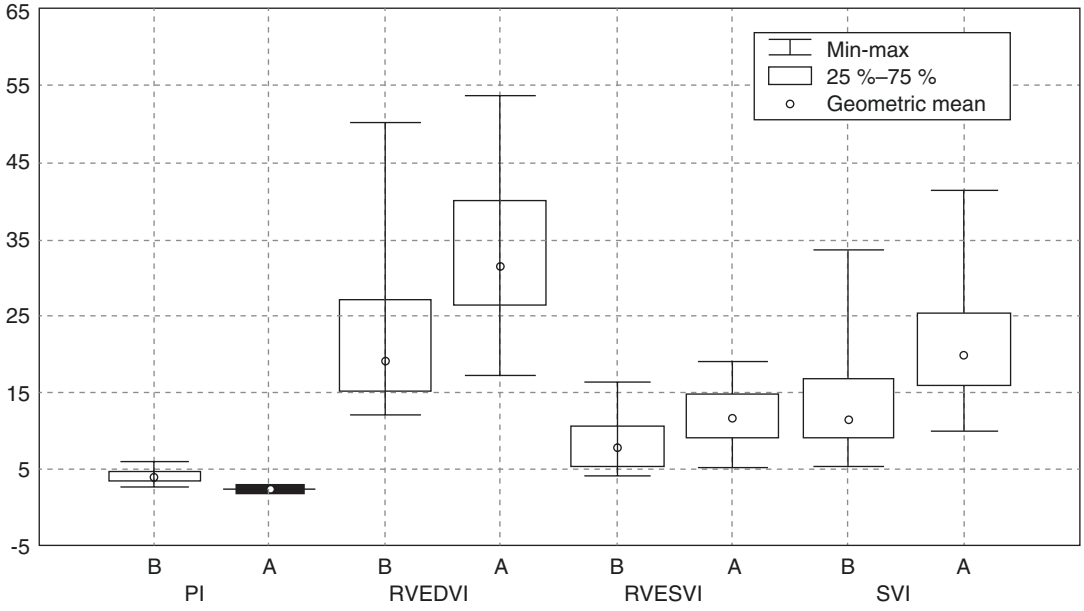

Fig. 20.5 Box and whiskers plot of Haller Index (HI), right ventricular end-diastolic volume index (*RVEDVI*), right ventricular end-systolic volume index (*RVESVI*), and stroke volume indices (*SVI*) before (*B*) and after (*A*) surgery (Kowalewski et al. [25]. With permission from Elsevier)

Summary

In conclusion, while many patients with pectus excavatum report cardiorespiratory symptoms pre-operatively and subsequent improvement post-operatively, concrete data in support of these subjective changes has been more difficult to identify. The literature does seem to suggest that patients with pectus excavatum, particularly of greater severity as demonstrated by a higher Haller Index, have pulmonary dysfunction that is restrictive in nature. The data on cardiac dysfunction remain less conclusive. Attempts to correlate the severity of HI with cardiorespiratory function have found that a higher HI is associated with measurable pulmonary limitation [3, 11, 12].

Recent research has elucidated other indices of chest wall deformity that may provide useful in correlations between severity of chest wall deformity and cardiopulmonary dysfunction. Redding et al. describe an upper chest wall deformity, known as pectus gracilis, which has an associated standardized measure of chest wall deformity called the pectus gracilis index (PGI) [8]. The PGI is a chest width-to-depth ratio measured at the gladiolar-manubrial-sternal junction, and it correlates highly with the HI (Fig. 20.6). Elevations in the PGI are associated with reductions in the vital capacity expressed as a percent of predicted (Fig. 20.7). More importantly, however, the PGI continues to correlate inversely with vital capacity, even after adjusting for the HI. The presence of an upper chest wall deformity such as pectus gracilis in patients with pectus excavatum may indicate that pectus is not just a deformity of the lower sternum, but instead a marker of an overall chest wall abnormality. Future research focused on a complete evaluation of the entire thorax in patients with pectus excavatum may provide further clues into the links between chest wall deformities and cardiopulmonary dysfunction.

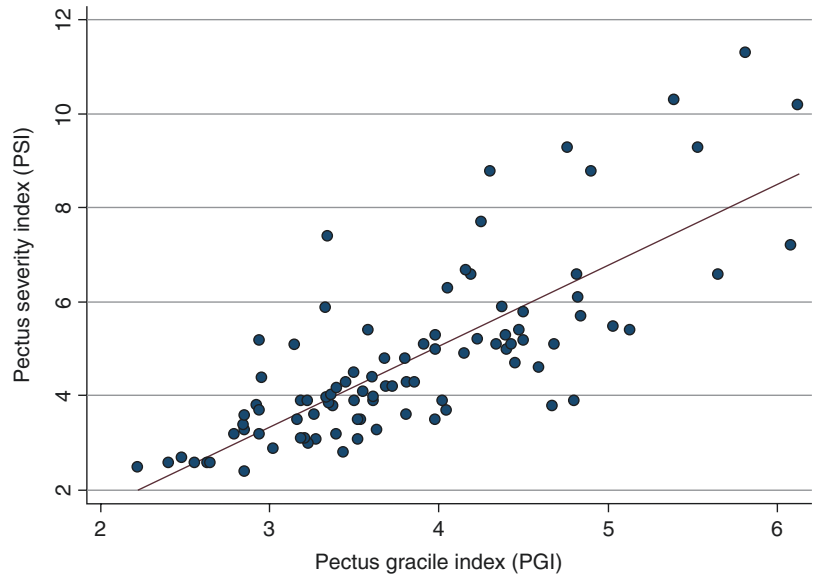

Fig. 20.6 Relationship between the pectus gracilis index (*PGI*) and the Haller Index (Redding et al. [8]. With permission from Wiley)

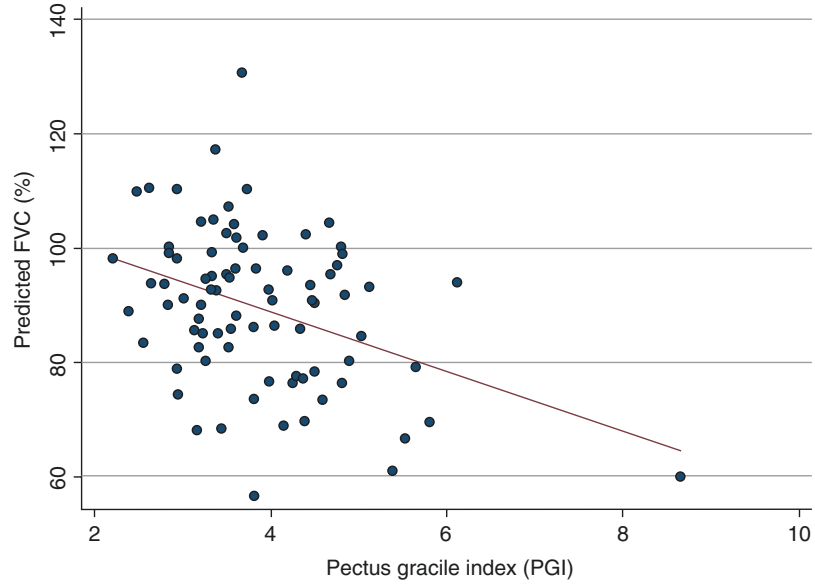

Fig. 20.7 Relationship between the PGI and vital capacity expressed as a percent of predicted based on published norms (Redding et al. [8]. With permission from Wiley)

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Cardiological Aspects of Symptomatic Pectus Excavatum in Adults

21

Paul F. Höppener, Hans A. Kragten, and Ron Winkens

Abbreviations

CT	Computed Tomography
GNT	Nitroglycerine
LVEF	Left Ventricular Ejection Fraction
MRI	Magnetic Resonance Imaging
PE	Pectus Excavatum
PFS	Pulmonary Function Studies
PS	Paediatric surgeon
PVCs	Premature Ventricular Contractions ("ventricuar extrasytoles")
RV	Right Ventriclar
SPES	Symptomatic Pectus Excavatum
TEE	Transesophageal Echocardiography
TS	Thoracic surgeon
TTE	Transthoracic Echography

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Introduction

Twenty years ago most GPs, internists and pediatricians regarded pectus excavatum (PE) principally as a cosmetic problem, and it was discussed as such in standard texts [1]. A relation between PE and clinical symptoms was traditionally not recognized by most surgeons. Even nowadays there is no consensus among doctors as to whether or not the deformity can produce symptoms, sometimes even severe enough to justify a surgical procedure [2].

It is the focus of this chapter to discuss the relation between PE and symptoms, with a overview of the trends over the past decades.

In the past 20 years a steadily increasing flow of reports has appeared from surgical groups in North America and continental Europe, describing a relationship between symptoms and this malformation, the possible benefits of surgical treatment, the complications of such operations, and the psychological burden associated with the condition [1–29]. Such literature reveals that pectus surgery is common, with series of many hundreds of cases being reported by thoracic surgeons with 20–40 years experience in pectus surgery (Table 21.1).

Robicsek and Watts give insight into the history of the past 80 years in their recent article "Surgical correction of pectus excavatum. How did we get here. Where are we going"[25].

An excerpt from this publication is cited:

> The clinical picture of pectus excavatum varies from no discomfort to total disability. Paradoxical

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Table 21.1 Reports from experienced pectus surgeons

Name of first author	Period	Nr of patients	Country	Year publication	Reference
Barauskas	1968–2002	853	Lithuania	2003	[6]
Fonkalsrud	1971–2009	912	USA	2009	[30]
Goretsky	1987–2003	1124	USA	2004	[14]
Haller	1949–1988	700	USA	2000	[31]
Kelly	1987–2000	100	USA	2008	[32]
Robicsek	1970–2000	±600	USA	2000	[25]
Saxena	1984–2004	1262	Germany	2007	[33]

breathing, a common symptom in patients with pectus excavatum, was first mentioned by Nageonette-Wilbourschewitz in 1925. Compression of the heart was not only recognized but also radiologically documented by Pohl in 1928. The most common symptoms mentioned were palpitation, exertional dyspnea, fatigue and dull precordial pain. In the following decades several studies demonstrated a significant improvement in exercise tolerance, dyspnea and chest discomfort after surgical correction.

Pathophysiology

Any structural or functional abnormality of the chest wall has the potential of affecting significantly the breathing but also the individual organs of the thoracic cavity [2, 34, 35].

Unfortunately, the expansion of the intrathoracic space does not seem to result in significant improvements in lung growth and function, suggesting that the restriction of the thorax is only part of the pathophysiology of these conditions [36].

Many studies performed during the past four decades have failed to document consistent improvement in pulmonary function at rest after surgical repair of pectus excavatum (PE), despite considerable symptomatic improvement. Most importantly, the pattern of lung function appeared to vary widely among affected patients [2, 30, 37]. Only anatomically severe PE is associated with abnormal pulmonary function [38]. Malik et al. conclude in their article "Pulmonary function following surgical repair of pectus excavatum. A metaanalysis", that the average effect for surgical repair of PE on pulmonary function was very small and not statistically significant [37].

Fonkalsrud et al. (2009) concluded, summarizing their experience with 912 symptomatic PE patients during the last 40 years, that physiologic impairment and reduced exercise capacity is primarily the result of impaired cardiovascular performance rather than ventilatory limitation [30] He refers to the long term cardiopulmonary effects after surgery, as described in 21 children by Sigalet et al. [39] (see also Chap. 23).

Huang (2009) evaluated prospectively 10 patients, aged 4–54 years. The function of the right ventricle was evaluated by intraoperative transesophageal echocardiography (TEE) and showed significantly increased values after retrosternal dissection and post-turning of the pectus bar [40].

Krueger et al. (2010) assessed in a prospective study 17 adult PE patients, with an age range of 17–54 (mean age of 28 years), expressing a variety of symptoms mainly related to impaired exercise tolerance and aesthetic concerns. All patients underwent intraoperative TEE with evaluation of RV dimensions and LVEF before and after surgical correction of their pectus excavatum deformity. Surgical correction of PE according to Ravitch-Shamberger technique resulted in a significant increase in end-diastolic RV dimensions and a significantly increased LVEF [41].

Comparable results were obtained by Neviere et al. in a similar study of 70 adult patients, with an age range of 18–64 (mean age of 28 years) [42]. The data of these three studies support the concept that closed repair directly contributes to sustained hemodynamic improvement. There is scarce evidence regarding haemodynamic effects of heart compression like enlarged

venous pressure and pulmonal hypertension [7, 23, 43–45].

In severe forms of pectus excavatum the inward deformation of the sternum creates an impression on the right atrium or ventricle. This compression of the heart was not only recognized but also radiologically documented by Pohl in 1928 [25]. Nowadays we can document the presence of cardiac compression in many more convincing ways by using modern imaging techniques like computed tomography (CT) of the thorax, magnetic resonance imaging (MRI) of the heart, TEE and angiography (see also Chaps. 16 and 20). The sternal depression, that characterizes the deformity, decreases significantly the anteroposterior dimensions of the rib cage. This decrease impedes the ability of the heart to expand adequately and inhibits proper filling of the right-ventricle.

This in turn limits the ability to increase the stroke volume in order to meet increased metabolic demands (e.g. during exercise). The cardiovascular function may be further complicated by the displacement of the heart and possible rotation of the great vessels [2, 4, 11, 14, 18, 20, 26–28, 30, 33, 36, 44, 46, 47].

In children and young adults, with a still flexible chest, compression of the right atrium and right ventricle can be avoided by deviation of the heart to the left. This flexibility decreases as age advances and the chest wall gets stiffer. By this time, side deviation is no longer possible and cardio compression increases. Thus, senior patients (>50 years of age), having an asymptomatic PE during their youth, can develop gradually and in slow progression a symptomatic PE. Senior patients develop exactly the same clinical picture as children and young adults, but at lower levels of exercise. Seniors get more complaints due to the slightly developing collapse of the spine with advancing age, which in itself is normal, the calcification of the costal cartilage, and the decreased mobility of the heart in the chest. The distance between sternum and heart will decline further, the inwards bent sternum will press as a wedge on the right atrium and the right ventricle [19, 23, 30, 44, 48].

Symptomatic Pectus Excavatum in Adults

The deformity of the chest in Pectus excavatum usually progresses slowly as the child grows. Most young children are asymptomatic, because young children have significant cardiac and pulmonary reserves. Additionally, a child's chest wall is very pliable. As children mature, however, the deformities become more severe, and the chest wall develops increased rigidity. They find that they have difficulty keeping up with their peers when playing aerobic sports [19]. They are young and in good health, the complaints occur only during high level exercises and are often are not mentioned or taken seriously for that reason [23].

Complaints and symptoms encountered in children and adults with a symptomatic pectus excavatum (PE) may consist of fatigue, reduced exercise tolerance, shortness of breath, palpitations, mitral valve prolapse and chest pain. Fonkalsrud et al. are the first to publish about PE in adults. Only 10 years afterwards the subject was picked up by a few more authors from several countries. Below an overview and cited excerpts from these publications.

Fonkalsrud EW (USA, 1999) "Repair of Pectus Excavatum and Carinatum in Adults" [12]. "During the past 11 years we have been consulted by an increasing number of patients over the age of 20 years who have severe untreated pectus deformities with worsening symptoms and limitations and who are very desirous of having surgical correction. There is sparse published information regarding the repair of pectus deformities in adults."

Jaroszewski (USA, 2010) "Current Management of Pectus Excavatum" [20]. "Many patients do not undergo repair during childhood and subsequently experience a progressive worsening of symptoms and cardiopulmonary function with increasing age. Patients with PE can present with many different symptoms. The symptoms often vary in severity and their effect on patients' daily activities. The severity of the defect does not necessarily correlate with the severity of symptoms. Many patients are asymptomatic at a younger age

but start experiencing symptoms as they enter their teens. This may be because of a worsening of the defect or an increase in exercise and physical activity".

Iida H. (Japan, 2010) "Surgical Repair of Pectus Excavatum" [28]. "Patients experience signs as they grow older, and more than half of the adult patients have some physical signs. Most patients report worsening of symptoms during adolescence, and patients even with mild deformities frequently report symptoms such as chest pain, chest discomfort, and palpitations after puberty, which may be caused by contact of the heart with the chest wall".

Frantz FW (USA, 2011) "Indications and guidelines for pectus excavatum repair" [3]. "Although infants and young children with PE rarely have physiologic symptoms associated with the defect, many older children and adolescents do develop cardiorespiratory symptoms, such as exercise intolerance".

Robicsek and Watts LT. (2011) "Surgical correction of pectus excavatum [25]. How did we get here_Where are we going".

"Also, it was shown that patients, who suffered but had no apparent ill effects from the deformity and live a seemingly unrestricted normal life at an early age, may later develop various degrees of cardiorespiratory embarrassment due to compression of the heart and blood vessels, loss of lung volume, paradoxical breathing and recurrent pulmonary infections."

Krueger T. (Switzerland, 2010) "Cardiac Function Assessed by Transesophageal Echocardiography During Pectus Excavatum Repair" [49] "PE is often asymptomatic during early childhood but symptoms like easy fatigability and decreased endurance usually appear when patients with PE reach adolescence and are involved in competitive sports".

Symptomatic Pectus Excavatum in Seniors (SPES)

Symptomatic Pectus Excavatum in Seniors, as such, was never mentioned in literature before we published about SPES. Quite often our surgical procedures for patients with SPES took place after they had been turned away by colleagues, who were not familiar with the clinical picture [19, 23] (Table 21.2).

"Symptomatic Pectus Excavatum in Seniors" (SPES) is usually not recognized by physicians because they are not familiar with the clinical picture. Sometimes it is even denied that a funnel chest possibly could cause of compression of the heart. In a period of 3 years (2006–2009) 138 patients with a PE were examined by our group, who consulted for cosmetic and somatic problems

Table 21.2 Publications on symptomatic pectus excavatum in seniors (SPES)

First author	Adult patients	Refers to	Year of publication	Reference
Majid et al.	1	48-year-old man	1979	[7]
Fonkalsrud	47	Patients with severe pectus deformities with worsening symptoms and limitations	1999	[50]
Saxena et al.	>1	Range 2–53 years Median age 14.9	2007	[33]
Guldemond et al.	1	68-year-old man	2008	[19]
Jaruszewski et al.	1	78-year-old man	2009	[48]
Krueger	>1	Range 17–54 years Median age 28	2010	[49]
Park et al.	1	68-year-old man	2010	[46]
Neviere et al.		Range 18–62 years Median age 28	2011	[42]
Kragten et al.	19	Patients with serious and invalidating symptoms	2011	[23]

at our regional Atrium medical Centre in the south of the Netherlands. The 96 patients younger than 49 years came mainly for esthetic reasons, but 27 % (n = 26) appeared to have mild to considerable somatic complaints. Two pairs of patients were brother and sister. Nearly all 42 senior patients (n = 38) were referred to our department after Dutch newspapers had drawn attention to a case history published by our group, which reported that symptoms such as shortness of breath and tiredness in elderly people could be related to an already long existing funnel chest [19, 23, 51]. The clinical picture of the 42 senior patients showed complaints of fatigue and low exercise tolerance, shortness of breath, palpitations, inspiratory obstruction and sometimes chest discomfort or pain. Nearly 45 % of the 42 patients (n = 19) were diagnosed as suffering from SPES. They had a rather deep funnel chest, considerable signs and symptoms and cardiocompression was visible on CT or MRI. There was a long history of a slow but steady progression of their complaints and a declining quality of life. It was remarkable that the serious, and sometimes invalidating, complaints had started only in their fourth or fifth decade of life. 63 % of our SPES patients (n = 12) were labeled as "patients with unexplained cardiovascular complaints". The first surgical procedure for a patient with SPES took place at our Centre after he had been turned away often by (thoracic) surgeons [19, 30, 48].

Clinical Picture of Symptomatic PE in Adults and Seniors (SPES)

Signs, symptoms and pathological findings most frequently encountered in children and adults with a symptomatic PE are fatigue, reduced exercise tolerance, shortness of breath, palpitations, mitral valve prolapse and chest pain.

Being young and in good health, these complaints occur during high level exercises and are often not mentioned or taken seriously for that reason [5, 14, 30, 38, 52].

Senior patients develop gradually and in slow progression exactly the same clinical picture, at lower levels of exercise [19, 30, 48]. Certainly, at advanced age, ischemic heart disease and COPD could fit into this clinical picture and therefore should be excluded.

See Tables 21.3, 21.4, and 21.5 for signs and symptoms in symptomatic pectus excavatum.

*[1]Palpitations

In PE the mechanical pressure on the heart can initiate Premature Ventriculart Contractions (PVCs), which patients describe as "palpations", as they are experienced as hard blows to the excavated part of the chest wall. Although this is not a serious complaint, these palpations are very frightening for the patient. The PVCs are sometimes quite frequent, unregulated and chaotic. Occasionally they initiate a nodal or supraventricular tachycardia (Fig. 21.1).

*[2]Other Complaints

Some patients in our research population observed that their edema of the legs, hemorrhoids , carpal tunnel complaints or tendovaginitis stenosans of some fingers had disappeared after corrective surgery. (see Part VII, Cases). No reference to this subject was found in literature.

*[3]Electrocardiography (ECG)

The radiological findings of heart compression and displacement are usually associated with pathological ECG findings. There are no specific ECG changes that can be pathognomic for the diagnosis PE with heart compression.

Barauskas found in a period of 35 years (1968–2002) pathological ECG changes in 382 (71.8 %) of his pediatric patients, postoperatively ECG findings were within normal limits in 330 (86.8 %) of the cases. In 52 (13.2 %) patients, all belonging to the age group of over 10 years, pathological ECG findings persisted postoperatively. The most common findings were incomplete right bundle branch block, repolarization disturbances, electrical axis deviation, tachycardia and bradycardia. He correlated pathological ECG findings with the data obtained by radiological examination and the correlation between them was statistically significant [6].

Table 21.3 Complaints and Synonym used in some publications [1–31]

Complaints	Synonym used in some publications	Specific references
Exercise intolerance	Exercise limitation; progressive loss of stamina and endurance with exercise; lack of endurance; limited work performance; decrease in stamina and endurance during exercise	[8, 11, 20, 23, 29]
Fatigue	General weakness; early fatigability; becoming easily fatigued	[11, 21, 25, 29]
Dyspnea at rest	Short of breath at rest; short of breath even under moderate exertion	[9, 11, 13–15, 25, 29]
Exertional dyspnea	Short of breath during strenuous exercise; difficulty climbing more than two flights of stairs without becoming short of breath	[9, 11, 13–15, 25, 29]
Tachypnea	Tachypnea with physical exertion	[20, 29, 30, 52]
Chest pain	Pain in the anterior chest; dull precordial pain; chest tightness	[5, 14, 24–26, 28, 30, 38, 53]
Palpitations[*1]	Heart pounding; irregular heartbeats	[13–15, 25]
Frequent respiratory infections, asthma	Asthma/asthma-like symptoms; mild restrictive lung disease	[12, 13, 22, 36, 38]
Lightheadedness	Dizziness	[27, 38]
Poorly eating	Appetite loss which may be the result of compression of the stomach by the deformed chest wall	
Other (infrequent)[*2]	Edema of the legs, hemorrhoids, carpal tunnel syndrome, snapping fingers	

Table 21.4 Incidence of Complaints, symptoms and pathological findings of 557 surgical patients in one clinic, median age 13.3 years, with a range from 21 months to 29 years.

Complaints, symptoms and pathological findings	%
Shortness of breath, lack of endurance, exercise intolerance	93
Frequent respiratory infections	34
Asthma/asthma-like symptoms	34
Chest pain, with or without exercise	71
Cardiac compression (by CT, echo)	85
Cardiac displacement (by CT, echo)	74
Murmur on examination	26
Mitral valve prolapse	15

From "Chest wall anomalies. Pectus excavatum and pectus carinatum" [14]

[*4]Transthoracic Echography (Ultrasound Examination of the Chest)

Mitral valve prolapse and/or mitral insufficiency have an incidence of about 1 % in a normal population. In several publications incidences about PE incidences varying from 8 to 58 % are mentioned [3, 21, 26].

Due to the abnormal anatomical shape of the chest in pectus excavatum, information obtained through the apical view of a transthoracal echogram (TTE) is unreliable and therefore disputable. The effect of mechanical pressure on the heart is often not clearly visible on the TTE. Transesophageal echocardiography (TEE) is superior to TTE, but not easily applicable in normal routine as it is a patient-unfriendly method of investigation. On the other hand preoperative and postoperative TEE is an important tool in monitoring the haemodyamic changes after relieving cardiac compression [42, 49, 59, 65].

[*2]Haller Index

Calculated on a CT scan by obtaining the ratio of the horizontal distance of the inside of the ribcage and the shortest distance between the vertebrae and sternum.

Table 21.5 Examination of PE patients; common findings

Examination	Common findings	Specific literature references
Inspection	Pectus excavatum (depth? shape?)	
Auscultation	Functional heart murmur	[9, 14, 20, 30, 54]
Medical Photography	Chest deformity, pectus excavatum	[55]
Electrocardiography (ECG)[*3]	Lateral displacement of the heart, right axis deviation, right bundle branch block, depressed ST-segments, arrhythmias like PVCs, SPVCs, sinus tachycardia, AV-nodal tachycardia, supraventricular tachycardia and Wolff Parkinson-White syndrome	[4, 7, 11, 20, 26, 30, 44]
Transthoracic Echography (TTE), standard supine[*4]	Mitral valve prolapse, mitral insufficiency, decreased left ventricular ejection fraction, elevated right atrial pressure, cardiac compression and changes in right ventricular geometry	[3, 14, 18, 21, 23, 26, 27, 29, 40, 42, 56, 57]
2-view chest radiography	Haller index[*5] (global estimation), inward deviation of sternum	[60–62]
CT MRI	Haller index, inward deviation of sternum, cardiac compression	[18, 25–27, 31, 58, 60–62]
Pulmonary function studies (PFS)	See Chaps. 19 and 22	[3, 6, 37, 40, 42, 63, 64]
Cardiopulmonary exercise testing	See Chaps. 19 and 22	[3, 6, 35, 40, 42, 63]

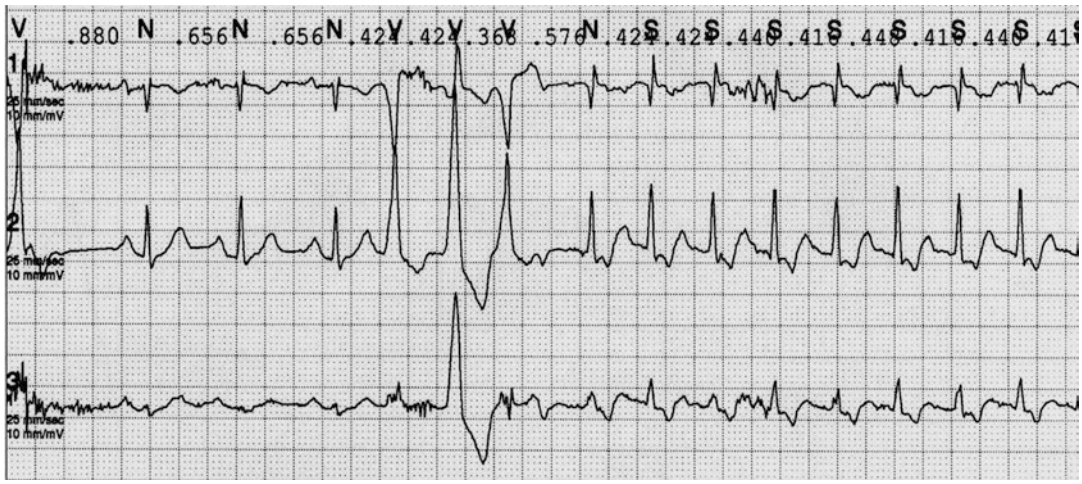

Fig. 21.1 ECG of a 65-year-old male SPES patient showing 3 normal beats (*N*), followed by 3 PVCs (*V*), initiating a supraventricular tachycardia (*S*)

Postural Components and Position of Diaphragm

Postural complaints and symptoms are quite usual in PE. In the upright or forward bending position the sternum is pressed inward even further. A full stomach will push up the diaphragm and reduce the space in the chest even more.

Ventricular catheterization studies have demonstrated diminished stroke volume and cardiac output in preoperative patients during upright exercise, which improved considerably after

repair. These findings are not demonstrable in supine patients evaluated at rest and may account for some of the reports of minimal physiologic impairment in PE patients [30, 39, 66–68].

Most physicians are not aware of this postural effect and therefore do not ask specific questions regarding this aspect of the complaints and symptoms. Patients complaining about dyspnea will tell their physicians, if asked for more information, that they experience the shortness of breath as an obstruction or "clogging" of the trachea. While breathing they have to inhale air rather forcefully against a resistance in their airways. Breathing through the mouth, abdominal breathing and lifting up the ribcage will facilitate breathing and reduce the complaints (Fig. 21.3). Most senior patients are good observers and have invented their own methods to decrease their complaints (Table 21.6; Figs. 21.2, and 21.3).

Table 21.6 Postural components and position of diaphragm

Components worsening symptoms	Measures taken to reduce symptoms
Local pressure	
On the sternum On the upper abdomen (belt) On the neck (tie)	Wider pants No belt, but braces to keep the pants up No tie
Leaning forward	
Work in stooping posture Reading book or paper Administrative work Working on the computer Fixing shoelaces Walking Cycling	Avoid leaning forward as much as possible Read standard Higher level of desk Placing the computer monitor at eye level Putting feet on a chair while fixing shoelaces Walk upright when admiring the tops of the trees Higher positioning of steering wheel
Rotating or bending the neck	
Looking down Maximal rotation of the neck	Avoid this attitude at daytime Proper position while sleeping
Normal lunch or dinner	Eat small portions, more often

Protocol for Preoperative Cardiological Screening

In our opinion, many published strategies and protocols for preoperative screening are too ambitious and only practicable in the setting of a university clinic. As their added value is minimal and the added workload is considerable, expiratory pulmonary function studies (PFS) and treadmill cardiac evaluation are only performed on indication [23]. A new item, not mentioned in literature and introduced by us, is the use of a Triflow Spirometer, a very simple tool to detect any obstruction of the airways during inhalation.

Taking the patient's habitual exercise history is a critical component of the evaluation process. Therefore, it is recommended that the mode (i.e. type of exercise performed), frequency (i.e. sessions per week), duration (i.e. hours per week), and length of time of the exercise regimen is documented systematically. A good tool for this is the Borg Rating of Perceived Exertion (RPE) scale [42].

In our Centre we gradually have developed a format for the "ideal protocol" for preoperative cardiological screening of all adult and senior patients with a clinical picture suspicious of symptomatic pectus excavatum. All results are

Fig. 21.2 Working on the computer, lower part of the screen at eye level

Fig. 21.3 Walking, wearing braces, forced abdominal breathing, head lifted as much as possible and pushing the ribs upwards with the hands

scored in the so called SPES-score and a Pectus Evaluation Index-score (PEI-score) and recorded in the electronic medical file of the patient [23].

Standard examinations:
History taking
Physical examination
Medical photography
Electrocardiography
Transthoracic echocardiography (TTE)
2-view chest radiography
Chest CT or MRI of the thorax
Triflow forced inspiration test[*1]

Special examinations only on indication:
Expiratory pulmonary function studies
Transesophageal echocardiography (TEE) before and after surgical correction
Treadmill cardiac evaluation
Measuring of the central venous pressure
Angiography

Registration and calculating scores
To assess to what extent our patients suffered from symptoms related to SPES the signs, symptoms and possible test results were arranged in a SPES-score from 1 to 12 (maximum) by giving a severity score to each finding. The SPES-score was 0 in case of absence of a PE, when there were no specific symptoms or when pre-existing disorders, like COPD and cardiac ischemia, could be responsible for the symptoms (Table 21.7).

The results of the visual inspection of the funnel chest by the surgeon and the radiological analysis were arranged in the PEI-score. An ordinal scale from 0 (flat or no pectus excavatum) to 5 (deep or wedge formed excavation) was applied [58].

Patients with a SPES-index >5, a PEI-score >3 and complaints of cardiac compression were eligible for surgery. After careful instruction they were asked by the surgeon to decide if they wanted a possibly curative correction of their PE [23].

[*1]Triflow forced inspiration test
The usual pulmonary function studies measure flow and volume at expiration. SPES patients have problems with their inspiration, which they describe as a "clogged intake of air."

The Triflow Incentive Breathing Spirometer is widely used by recently operated heart patients to promote slow sustained maximal inspiration and to achieve full expansion of the lungs (Fig. 21.4).

A simple and cheap diagnostic tool was used by our group to measure the degree of inspiratory obstruction. SPES patients with a decreased inspiratory flow were only able to achieve, and maintain, the inflow required to draw two balls to the upper position, and keep them more than 1 min in place. Immediately after their operation the third ball would remain in position for several minutes.

Table 21.7 SPES-score calculation

Signs and symptoms	SPES score		
	1	2	3
Dyspnoea	Minor	Moderate	Severe
Palpitations	Minor	Severe	
Fatigue, low exercise tolerance	Minor/moderate	Severe	
Chest pain	Yes		
Postural symptoms	Possible	Clearly present	
ECG/Holter recording:			
(Supra)ventricular arrhythmia; Erratic conduction; Partial AV-block	Clearly present	Severe	
Echocardiography:			
Mitral valve insufficiency and/or prolapse; Tricuspid valve insufficiency; Enlarged right atrium	Minor	Clearly present	
Spirometry/Triflow forced inspiration test:			
Inspiratory obstruction	Moderate	Clearly present	

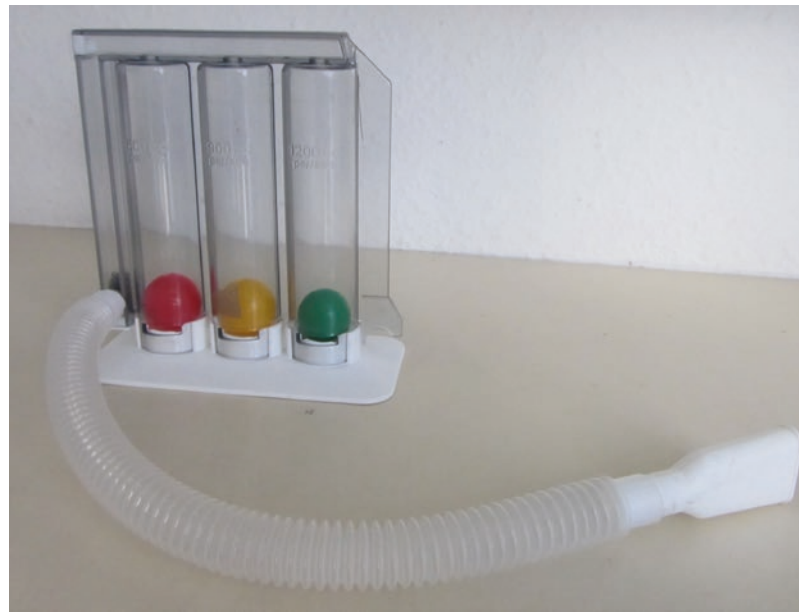

Fig. 21.4 The Triflow model is created with three chambers, each with a ball inside that will raise as the patient takes a breath, as deep as possible, through the mouthpiece. The balls indicate air pressure and air flow strength

Cases

Three patients from our Centre are presented, described in an earlier publications and on our website www.spesweb.nl. All patients have given their consent to publish this material, including the images.

Patient One

A 57-year-old man with an 8 year history of slowly increasing loss of stamina and shortness of breath. Also complaining about periods of fast palpitations, coming up suddenly and stopping after maximal 1 h. Previously healthy and feeling

well. Once a year he was running the local half Marathon and has been performing well. He owns a garage and does all the car maintenance work himself.

For 2 years there has been a progressive loss of stamina and endurance with exercise and he had to interrupt his work frequently because of increasing fatigue. His complaints increase when he works in a stooping position during car maintenance or leans forward doing administrative work. The last few months he has not been able to perform his work properly and he is tired after a short walk. His complaints were elsewhere diagnosed as unexplained cardiac complaints. After reading an article in the local paper about the diagnosis and treatment of SPES in our hospital, he could persuade his cardiologist to refer him to our hospital for further diagnosis and treatment.

A moderate degree of funnel chest with maximal central depression of 3.2 cm was evident (Figs. 21.5 and 21.6). The pulse was regular, blood pressure was 130/80 mmHg in both arms. An electrocardiogram at rest showed normal sinus rhythm and conduction. Echocardiography showed a minimal prolapse of the mitral valve. The MRI in the transverse and posteroanterior view showed a pectus excavatum with formation of osteophytes and compression of the heart. SPES-index: 8 and PEI-score: 4 (Figs. 21.7 and 21.8).

A modified Ravitch procedure was performed. The sternum, which clearly pressed firmly against the heart, was lifted 5 cm and fixed with 2 cerclage wires and 2 supporting Marlex strips. Postoperative recovery was fast, some pain on the breast was the only remaining complaint. After 3 months he could perform his mechanical work as he used to do many years ago. After 1 year he performed well in running the half marathon. Feeling of general well-being on a scale of 10: before the operation 3 points, after the operation 8 points.

Patient Two

A previously healthy 59-year-old male patient complained of progressive heart palpitations, fatigue and postural dyspnoea; bending over caused a clear increase of dyspnoea. At repeated examinations no overt abnormality or explanation was found, except a supraventricular arrhythmia and a nodal tachycardia. Drug treatment with sotalol and verapamil had only temporary effects. In the 8 years to follow the symptoms increased and led to considerable physical impairments and orthopnoe. He tried to cope with his impairments

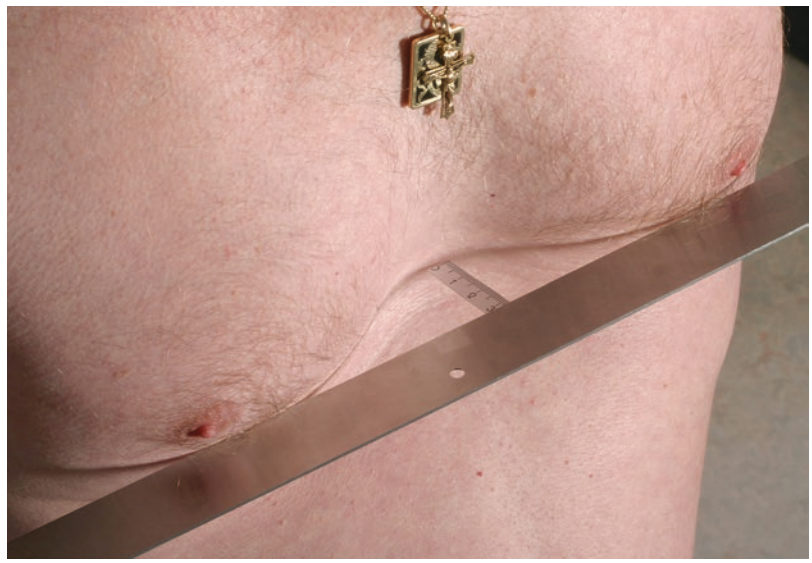

Fig. 21.5 Moderate pectus excavatum, measured depression 3.5 cm

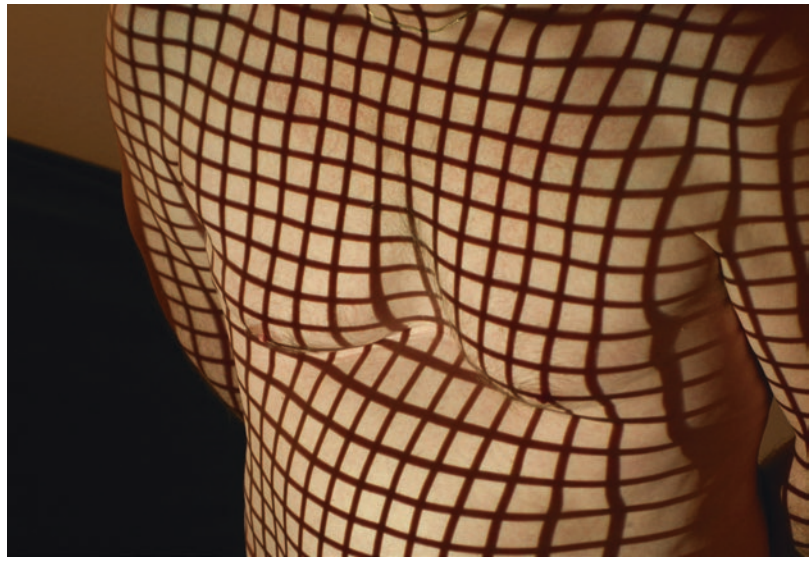

Fig. 21.6 3-D gridprojection photograph [55] (Photo courtesy of Hans van Dijk, Atrium Medical Center, Heerlen)

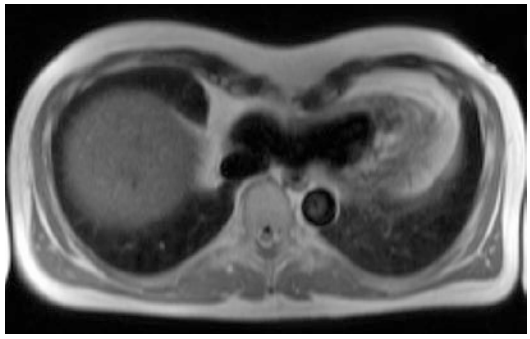

Fig. 21.7 Transverse MRI thorax; cardiocompression

by a series of measures such as eating smaller amounts of food, reducing exercise, etc. Climbing stairs without resting became impossible. The dissatisfactory diagnosis was: ventricular extrasystoles, nodal tachycardia, and congestive heart failure of unknown origin. Meanwhile, the patient himself noticed that dyspnoea not only occurred after exercise but also postprandially, and depended on posture (bending over, in particular). In a desperate attempt to seek relief of discomfort, the patient started using glyceryl trinitrate (GTN, nitroglycerine), resulting in a dramatic but unfortunately only temporary improvement.

Angiography was completely normal, the effect of the GTN could not be explained. So, 8 years after the first onset of symptoms, an explanation for the patient's condition was still lacking. Both the patient, his sister and his brother had a pectus excavatum, and this had usually not been noticed by the consulted doctors and if noticed labelled as "harmless". After a web search, the patient came up with an explanation: his complaints were caused by PE. Initially, reactions were skeptic but ultimately his cardiologist, impressed by the available information on internet, referred him to an experienced thoracic surgeon. A lateral chest x-ray with the patient bending over and a lateral computed CT of the thorax revealed an impression of the heart by the sternum, which at the time had not been noticed nor recorded by the examining doctor (Figs. 21.9, 21.10, 21.11 and 21.12). SPES-index: 9 and PEI-score: 5

Ten years after the patient's complaints and symptoms had first appeared, a modified Ravitch procedure was carried out, preoperatively measured central venous pressure was 5 cm in the supine position and raised to 10 cm when the patient was sitting upright. Soon thereafter, the physical condition of the patient improved rapidly and he was almost completely relieved from fatigue and shortness of breath. After 3 months, heavy physical exercise was possible again, with only minor arrhythmia. Feeling of general well-being on a scale of 10: before the operation 4 points, after the operation 8 points.

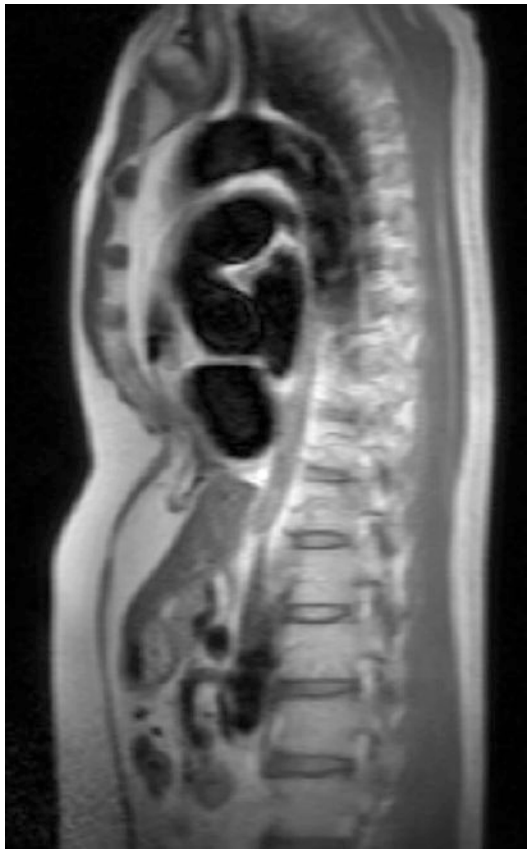

Fig. 21.8 Lateral MRI thorax, cardiocompression

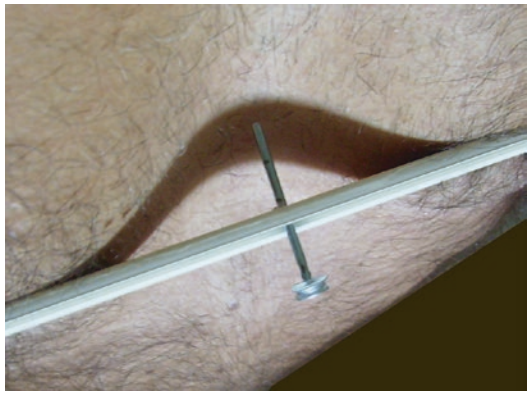

Fig. 21.9 Measuring the excavation

Patient Three

A previously healthy 67-year-old female patient had been familiar with asthmatic complaints since her childhood. Eight years ago she developed a slowly progressing shortness of breath after exercise. More and more she was feeling tired and dizzy. She had a dull pain in the anterior chest and there were frequent palpitations. For 6 months walking up more than one stair at a time had been nearly impossible, and even at rest she had been short of breath. For 2 years her legs had been swelling and she had been experiencing a decline of consciousness while bending forwards.

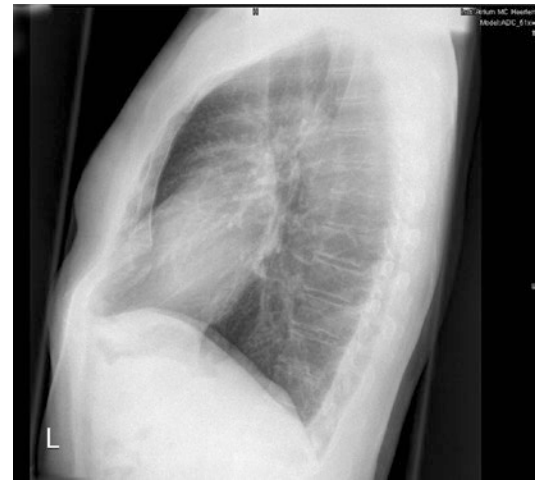

Fig. 21.10 Lateral Xray chest (2005)

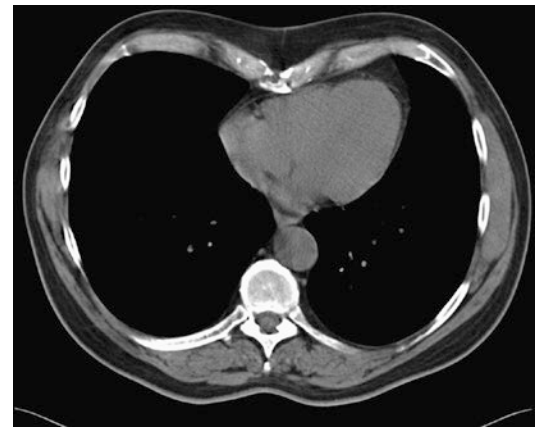

Fig. 21.11 Transverse CT thorax (2005); cardiocompression

The patient had a pectus excavatum. After reading an article in the local paper about the diagnosis and treatment of SPES, she consulted her GP who referred her to the local pulmonologist. After examination he referred her to our hospital for

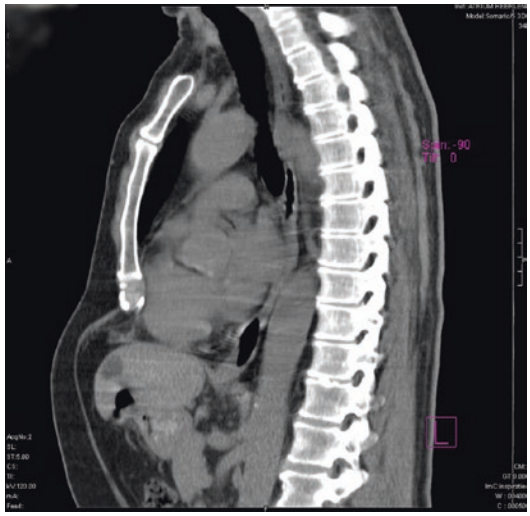

Fig. 21.12 Lateral CT thorax (2005); cardiocompression

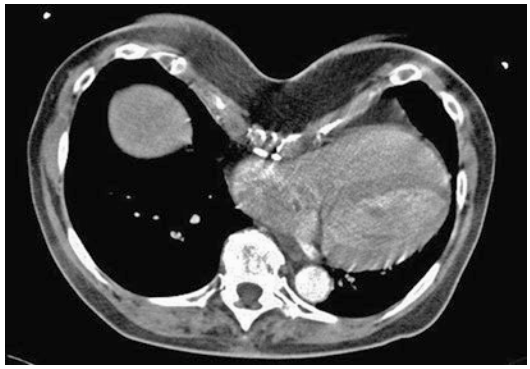

Fig. 21.13 Transverse CT thorax; severe compression of the right atrium and the right ventricle (Courtesy of Ms. Annet Piek)

treatment of her severe symptomatic pectus excavatum. There was an evident pectus excavatum with an external depth of 4 cm. Both legs were edematous, obvious cardiac abnormalities were not found. Right heart catheterization (requested by the pulmonologist) showed increased pressure in the A.pulmonalis with exertion, and the arterial pressure in the Rt atrium had increased upto an average of 13 mmHg (normal = 5–7 cm). Transverse and lateral computed tomograms of the thorax showed a severe compression of the right atrium and the right ventricle of the heart. SPES-index: 11 and PEI-score: 5 (Figs. 21.13 and 21.14).

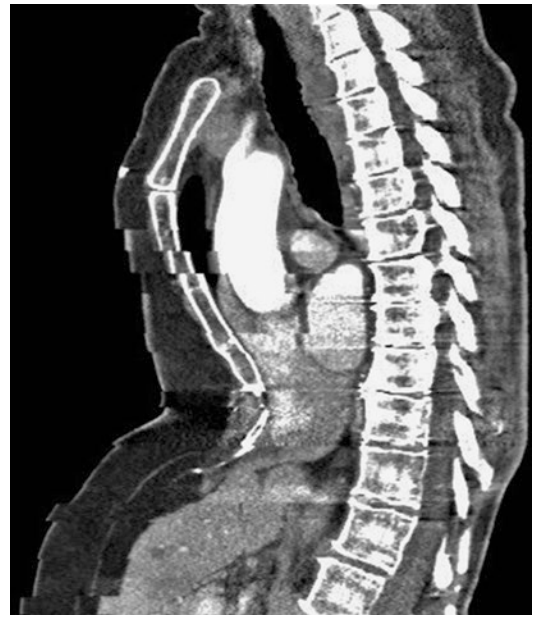

Fig. 21.14 Lateral CT thorax; severe compression of the right atrium and the right ventricle (Courtesy of Ms. Annet Piek)

The physical condition of the patient improved gradually after a modified Ravitch procedure was carried out. After 3 months the distance between the sternum and spine appeared to have increased 5 cm (from 3.8 cm to 9.3 cm). Her previous complaints and symptoms had disappeared nearly completely and she could resume her former rather healthy way of life. Feeling of general well-being on a scale of 10: before the operation 4 points, after the operation 7 points.

Discussion and Conclusions

There is no consensus among doctors as to whether or not pectus excavatum can produce symptoms sometimes even severe enough to justify a surgical procedure.

For many decades PE has been considered to be an incidental finding without clinical significance. However, there is growing evidence from literature, that PE may cause complaints and symptoms such as heart palpitations, fatigue and postural dyspnoea.

The former reluctance of clinicians to perform a surgical correction of a pectus excavatum can partly be explained by the fact that surgical repair of pectus excavatum does not significantly improve pulmonary function. Malek and colleagues suggest that these findings may be a result of testing pulmonary function under conditions in which pectus excavatum does not manifest itself [37]. In our opinion pulmonary function testing should only be done on indication, to exclude expiratory pulmonary obstruction. The clinical picture of the patient, as judged by the surgeon or cardiologist in follow up examinations, and the improvement of the quality of life, as experienced by the patient, were our indicators for the postoperative relief of symptoms [23].

Chest radiography should be used as a primary modality for preoperative imaging of pectus excavatum. However, in most elderly patients additional information from a CT scan or MRI will be required to measure the PA chest-diameter and to discover if cardiac compression is present.

Due to the abnormal anatomical shape of the chest in pectus excavatum, information obtained through the apical view of a transthoracal echogram is unreliable and therefore disputable [65]. More reliable results can be obtained by transesophageal echocardiography(TEE). Three recently published prospective studies showed clearly, that surgical correction of pectus excavatum resulted in a significant increase in end-diastolic RV dimensions and a significantly increased LV ejection fraction. End-diastolic right ventricular (RV) dimensions and left ventricular (LV) ejection fraction were assessed by use of intraoperative TEE before and after surgical correction of pectus excavatum [24, 40, 42, 49]. Our own data show a clear relation between a pectus excavatum in senior patients and complaints like shortness of breath, palpitations, chronic fatigue and symptoms like ventricula ectopic beats [23].

A postural component, aggravating the complaints and symptoms, was often present. Most doctors are not aware of this postural effect on complaints and symptoms and therefore do not ask specific questions regarding this aspect. Quite understandable, as there was up till now only scarce hard evidence available about the negatively reduced pumping capacity of the heart during upright exercise, and hemodynamic improvement after surgical correction [42, 49, 59, 65, 67].

ECG recordings showed moderate or serious conduction disturbances and arrhythmia [23]. Heart failure and ischemic heart disease can present a similar clinical picture and should be excluded. Cardiac compression by the inward deformation of the sternum and the presence of a postural component are indicative for diagnosing SPES. The clinical picture we encountered in our patients with a pectus excavatum suggests a cardiovascular problem. In fact 12 of the 19 patients with SPES were diagnosed previously as having unexplained cardiovascular complaints.

More research should be undertaken to collect more evidence. Up till now the postural component aggrevating complaints and symptoms has escaped attention. More specific tools for measuring the degree of inspiratory obstruction are wanted. The hemodynamic consequences of cardiac compression in pectus excavatum should be examined more systematically.

Medical schools and postgraduate training should include this subject in their curriculum or at least refrain from spreading the misapprehension that PE is merely a curiosity. Diagnosing SPES is relevant because surgical reconstruction of the chest can provide complete relief of symptoms.

Symptomatic Pectus excavatum is a cardiological problem, as has become amply evident from literature.

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Assessment of Right Ventricular Function in Pectus Excavatum

22

Haritha Reddy, Prashant Vaishnava, and Rajeev L. Narayan

Introduction

The right ventricle (RV) has been an elusive and misunderstood entity within the realm of cardiology. While previously thought to contribute little to overall cardiac function, recent advancements in cardiac imaging have demonstrated the pivotal role of the RV with respect to the etiology of multiple disease processes and the critical role the RV plays with respect to overall cardiac function. This chapter will discuss the principles of normal right ventricular anatomy, function and hemodynamics, and help to elucidate key differences in the behavior of the RV when compared to the left ventricle (LV). Furthermore, the impact of chest wall disorders, specifically pectus excavatum, on right ventricular function will be explored. In order to understand the impact of pectus excavatum on RV function, the principles of echocardiographic and cardiac magnetic imaging will be introduced.

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Principles of Normal Right Ventricular Anatomy, Function and Hemodynamics

As the "forgotten ventricle," the right ventricle (RV) has often been neglected [1]. Recently, however, the substantial role of RV function has been more significantly elucidated, and studies using all available cardiac imaging modalities have demonstrated the pivotal role of the RV with respect to the etiology of multiple disease processes [1, 2]. Nonetheless, most efforts in adult cardiac imaging have focused on left ventricular (LV) structure and function. Developments in cardiac magnetic resonance imaging (CMR) and echocardiography over the past two decades have provided new insights, however, into the critical role the RV plays with respect to overall cardiac function.

The right and left ventricles differ substantially in terms of their morphology, structure and function. This is likely due to their embryological origin. RV myocardial precursor cells have a different origin than that of LV precursor cells [3, 4]. LV precursor cells originate from the primary heart field in the anterior plate mesoderm, whereas RV precursor cells originate from the secondary heart field. This embryological difference may, in part, explain the differential response

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of right and left ventricular myocytes to abnormal hemodynamic parameters within the heart [5]. During the fetal period, the right ventricle pumps blood to the systemic circulation and placenta, contributing up to 65 % of cardiac output [6]. After birth, the right ventricle remodels to accommodate for low impedance pulmonary flow. Once this initial remodeling is complete, the right ventricle loses its ability to convert back to its fetal form and therefore its ability to respond to abnormal hemodynamic loading, especially increased pressure [6]. The genetic expression involved in adaptive remodeling is different in the right and left ventricles and studies have demonstrated that molecular changes in the RV myocardium induced by pressure loading are significantly different from those in the LV myocardium [5]. Due to its inability to adapt to different pressures the right ventricle is more prone to fail with chronic pressure loading [6].

The physiology of right and left ventricles differs greatly. The RV pressure-volume relationship, which is generally presented visually as a loop, appears as a more trapezoidal shape versus a more rectangular shape found in the left ventricle (Fig. 22.1) [7].

This difference is a reflection of the less defined isovolumetric phases in the right ventricle [7]. Furthermore, the right ventricular cardiac output is very sensitive to afterload, given its inability to effectively respond to increases in pressure. Even modest increases in afterload can lead to significant decreases in RV output [6]. In addition, the three-dimensional orientation of the myocytes in the RV differs from that in the LV resulting in different contraction patterns. The right ventricle contracts with the walls moving toward the septum and a longitudinal motion of the base towards the apex, whereas the left ventricle contracts circumferentially and is associated with a twisting motion similar to the wringing out of a towel [6, 7]. Torsional motion is less pronounced in the RV, and these differences affect RV functional assessment.

The mediastinal position and ventricular morphology are also a source of difference between the RV and LV. The anterior position of the RV in the chest limits echocardiographic visualization, and as a result, are better imaged using cross-sectional imaging techniques such as CT and MRI, which are less limited by near-field resolution, and are therefore generally considered superior to echocardiographic imaging. Two-dimensional imaging is further limited by the complex RV morphology. In contradistinction to the ellipsoid ventricular shape of the LV, the RV has a complex geometric shape with a triangular appearance in the sagittal plane, and a

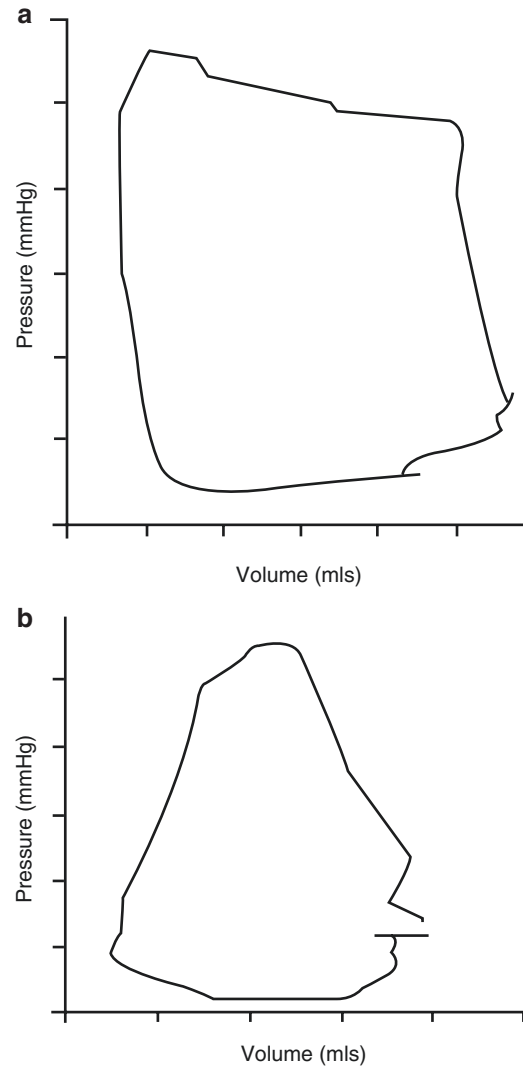

Fig. 22.1 (**a**) The rectangular left ventricular pressure-volume loop. (**b**) The trapezoidal right ventricular pressure-volume loop (With permission from *Cardiology Clin*. 2002;20:341–49. Elsevier)

crescent shape in the coronal plane [6, 7]. RV anatomy is divided into the inflow tract which contains the tricuspid valve, chordae tendinae, papillary muscles and highly trabeculated myocardium; and the outflow tract which contains smooth myocardium and the pulmonic valve. A third apical region is also noted, and opposes the left ventricle at its apex. The RV inflow and outflow tracts are positioned in different planes, and thus present a further obstacle to two-dimensional RV imaging.

The normal RV also features a thin wall that is normally from 3 to 5 mm in adults [8]. This can be difficult to differentiate from the surrounding structures in the mediastinum. Prominent RV trabeculations further complicate detection of endocardial surfaces required for echocardiographic assessment of RV volume, mass and function.

In summary, the RV is a complex cardiac chamber with varying embryological origin that causes it to have a unique anatomy, morphology and hemodynamic response that creates difficulty with respect to assessment with cardiac imaging. Despite these obstacles, advanced cardiac imaging techniques utilizing echocardiography and cardiac magnetic resonance (CMR) imaging have helped to improve our ability to elucidate and understand normal RV function.

Features of Right Ventricular Function on Echocardiography

As mentioned, the right ventricle is difficult to image by two-dimensional echocardiography due to its location in the anterior chest, its thin wall and coarse trabeculations. In addition, its retrosternal location creates an acoustic barrier for ultrasound waves [8, 9]. Cardiac magnetic resonance imaging (CMR) and CT are less limited by near-field resolution and therefore offer better visualization of the right ventricle [6–8].

Currently, CMR is considered the gold standard technique to identify right ventricular volume and RV ejection fraction [6]. However, it is only reliable when adequately standardized. Small differences in methodology, such as patient respiratory patterns during acquisition or inclusion/exclusion of RV trabeculations or the inflow/outflow tracts in calculations of RV volumes can potentially lead to dramatic variability in results, leading to misinterpretation of RV function. Other disadvantages to CMR include its accessibility and costs [6, 7]. Add to this, the general exclusion of patients with claustrophobia and those with implanted metal, and the utility of CMR in RV functional assessment becomes limited.

Clinically, echocardiography is the most frequently used modality to assess RV function and especially to follow RV function over time. Echocardiography is relatively inexpensive and much more readily available, even in resource-limited settings. Recent advances in imaging technique and transducer quality have allowed for the inclusion of both two and three-dimensional echocardiographic imaging. RV volumes obtained via three-dimensional echocardiography have been shown to correlate well to CMR [6]. In clinical practice, serial two-dimensional echocardiography is used to follow RV dilatation; however volume measurements obtained do not correlate well to three-dimensional volumes. Therefore, for those patients who need accurate measurements to guide therapeutic treatment, CMR images are generally recommended [6, 7].

Assessing right ventricular systolic function is more complicated than left ventricular systolic function, and there is no single ideal parameter used in clinical practice. Unlike LV systolic function, for which left ventricular ejection fraction (LVEF) has emerged as a universally accepted parameter to assess LV systolic performance; RV systolic function is less well defined. RV ejection fraction is more difficult to measure than LVEF given the aforementioned complex geometry, causing gross miscalculations with respect to RV volumetric analysis. The most clinically used technique to assess RV function in most echocardiographic laboratories is subjective visual assessment of the RV function, in which an experienced echocardiographer utilizes "eyeball assessment" to provide qualitative assessment of RV function as normal, mildly reduced, moderately reduced, or severely reduced. This method,

however, has poor sensitivity, and further has poor intra-observer and inter-observer variability. As a result, numerous quantitative parameters have emerged in the literature to assess RV function. These include: percent fractional area change, Doppler assessed myocardial performance index, tricuspid annular plane systolic excursion (TAPSE), tissue Doppler derived tissue velocity, and myocardial deformation studies such as strain and strain rate [6–8].

Percent fractional area change (FAC) is perhaps the simplest of quantitative parameters that are calculated using two-dimensional imaging techniques. It is calculated using the four-chamber view on an echocardiogram, and is equal to the difference between end-systolic area and end-diastolic area divided by the end-diastolic area (Fig. 22.2a). A fractional area change of <40 % is considered abnormal. While simple to use, and easy to obtain, with excellent inter-observer variability, RV FAC does not take into account the RV outflow tract's effect on systolic performance [8].

As RV geometry has been difficult to assess using two-dimensional echocardiography, Doppler ultrasound techniques have been long proposed as an alternative method of objective RV functional assessment. Blood-pool Doppler ultrasound techniques to measure RV inflow or outflow velocities can be used to noninvasively assess RV hemodynamics, including RV systolic pressures, pulmonary artery systolic pressures and pulmonary artery diastolic pressures. Additionally, blood-pool Doppler ultrasound data have been used in the assessment of RV function by measuring systolic time intervals. The ratio of pre-ejection to ejection times has long since been suggested as an indicator of LV and RV function [9]. This concept was expanded by Tei and colleagues, who proposed the use of the sum of isovolumetric relaxation and isovolumic contraction times divided by ejection time as an indicator of global systolic and diastolic performance. This ratio was named the myocardial performance index (MPI) (Fig. 22.2b) [10]. The longer the isovolumic phases, the higher the MPI and the worse the ventricular performance (normal value 0.28 ± 0.04) [8–10]. The MPI concept to assess RV performance is surprising given the short or absent isovolumic periods in normal right ventricular contraction. However, with increasing loading parameters, the isovolumic periods become longer and more easily measurable. These intervals are also altered by RV function, and with progressive deterioration in RV function, there is progressive prolongation of isovolumic contraction and diastolic dysfunction with prolonged isovolumic relaxation. The RV MPI has been shown to be a strong prognostic factor for disease progression in patients with pulmonary hypertension, and has correlated well with right ventricular ejection fraction (RVEF) measured by CMR in patients with congenital heart disease. However, despite these excellent data, the MPI varies considerably with both function as well as loading parameters, and can be difficult to reproduce as simultaneous measurement of both RV inflow and RV outflow is not possible using Doppler technique. As a result, imaging must be separated temporally, and changes in heart rate or loading between captured cardiac cycles can lead to misrepresentations of the MPI. Currently, the MPI has not been used clinically with widespread acceptance.

Normal RV function is dependent on longitudinal shortening. As a result tricuspid annular plane systolic excursion (TAPSE) was proposed as a measure of systolic function based on the principle that longitudinal shortening is a good indicator of overall ventricular performance. It is easily calculated using an M-Mode cursor placed over the lateral tricuspid annulus in the two-dimensional four-chamber echocardiographic view, with a direct measurement of longitudinal displacement of the annulus in centimeters (Fig. 22.2c). Normal values should be greater than 1.5 cm in adults [8]. TAPSE has been found to correlate well with both RVEF as well as RV FAC percent change, and is associated with survival in patients with pulmonary arterial hypertension. TAPSE, however, is a regional parameter that can only be used to measure longitudinal shortening in the lateral free wall of the right ventricle, and in patients with regional dysfunction in the outflow portion of the ventricle, TAPSE fails to correlate with global RVEF. Since TAPSE

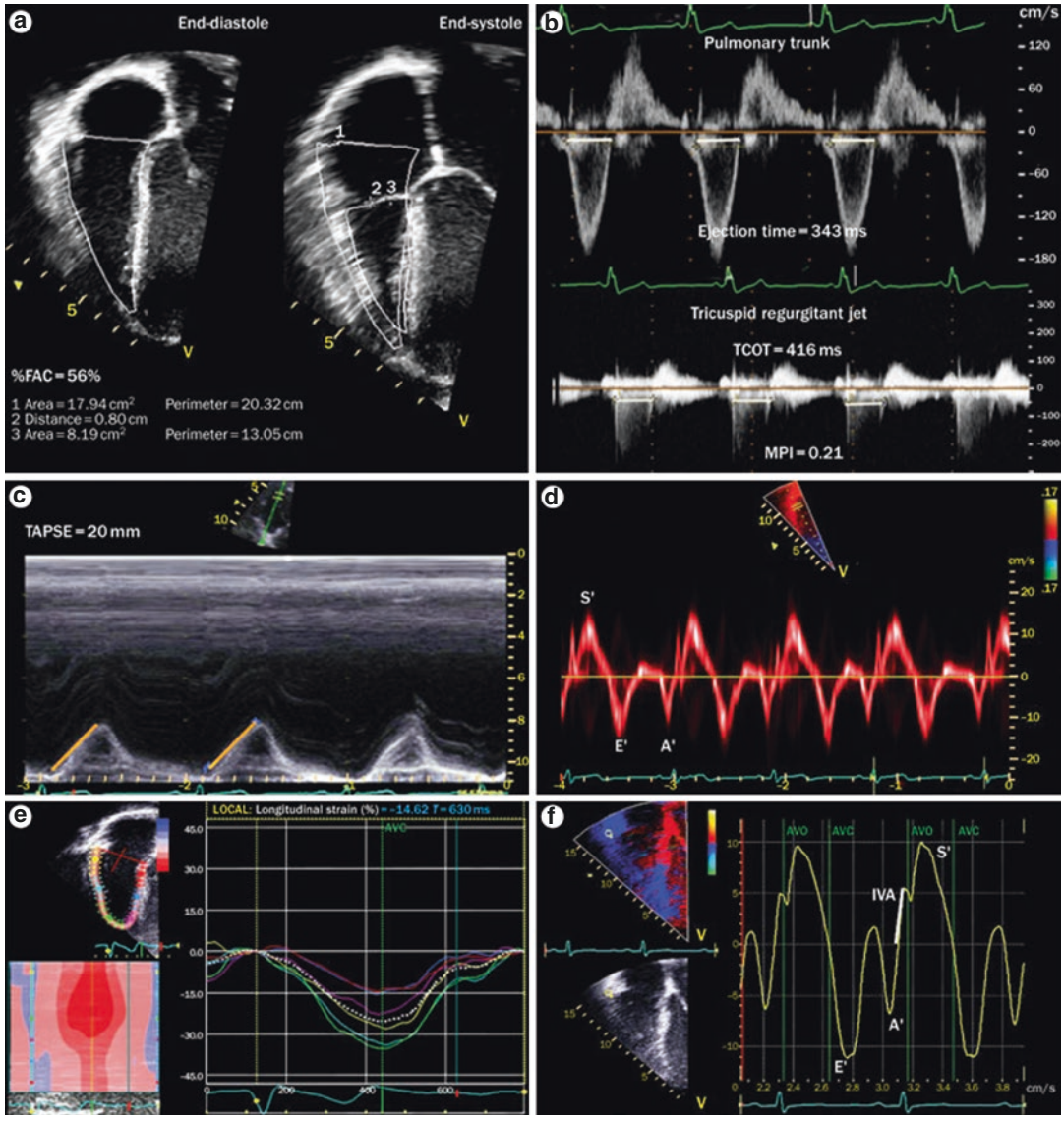

Fig. 22.2 Alternative ways to assess RV function by echocardiography: (**a**) %FAC which is calculated by using measurements from the apical four chamber view (End Diastolic Area-End Systolic Area/End Diastolic Area × 100). (**b**) MPI which is calculated by taking the difference of the tricuspid valve closure (found on the tricuspid inflow tracing) to opening time and the ejection time (found on pulmonary artery tracing) over the ejection time. (**c**) TAPSE is calculated by taking an M-mode echocardiogram through the tricuspid annulus. The excursion of the tricuspid annulus is measured in centimeters. (**d**) Tissue Doppler velocities of the tricuspid annulus can be used as a measurement of systolic longitudinal RV function. (**e**) Longitudinal strain measurements of the right ventricle, which are calculated by using speckle tracking technology. Systolic longitudinal shortening is shown as a negative value and are measured in six segments which are then used to trace a mean longitudinal strain curve (shown as the white dotted line). (**f**) Color tissue Doppler echocardiogram at the lateral tricuspid valve annulus is used to measure myocardial acceleration during isovolumetric period (*IVA*) which is independent of afterload. *Abbreviations*: *A'* late-diastolic tissue velocity, *AVC* aortic valve closure, *AVO* aortic valve opening, *E'* early-diastolic tissue velocity, *%FAC* percentage fractional area change, *IVA* isovolumetric acceleration, *MPI* myocardial performance index, *S'* systolic tissue velocity, *TAPSE* tricuspid annular systolic plane excursion (With permission from *Nat Rev Cardiol*. 2010;7(10):551–63. Macmillan Publishers)

is easy to measure and has excellent reproducibility, it is considered a useful quantitative parameter and is used regularly in clinical assessment [6].

More recently tissue Doppler ultrasonography has emerged as a method to directly measure myocardial velocities as a measure of geometry independent functional assessment. Tissue velocity, combined with myocardial deformation studies such as strain and strain rate index are obtained using tissue Doppler velocity imaging of the tricuspid annulus which measures the degree of myocardial shortening and lengthening reflecting longitudinal RV function (Fig. 22.2d–f). While tissue Doppler and myocardial deformation studies are attractive alternatives to TAPSE to assess RV longitudinal motion, and have excellent reproducibility, their dependence on angle of insonation and loading conditions of the RV have limited their clinical utility. Currently, these tools remain investigational with respect to RV systolic function assessment, but are promising with respect to the quantitative evaluation of RV performance [6–8].

In summary, echocardiography is a clinically meaningful and readily available tool for assessing the function of the forgotten ventricle [11]. Its ease of access combined with its relatively lower cost allows it to be used regularly and for serial assessment. A number of both qualitative and quantitative parameters have evolved in order to assess RV function using echocardiography, and while no individual parameter has demonstrated clear superiority in functional evaluation, the combined use of multiple parameters has allowed clinical echocardiographers to provide accurate and reproducible methods by which RV function can be seen and serially evaluated.

The Impact of Pectus Excavatum on Right Ventricular Function

Pectus excavatum is a common chest wall abnormality with a prevalence of 0.4–2 % in the general population [11, 12]. In fact, pectus excavatum accounts for 90 % of all congenital chest wall deformities and is six times more common in males than females [11, 12]. Patients with pectus excavatum have deformity of the chest wall in which the inferior portion of the sternum is invaginated causing the adjacent cartilage and structures to be displaced posteriorly. This structural aberration may cause the right ventricle and the right atrium to be compressed between the vertebral column and the depressed sternum, leading to effects on RV function [11]. Patients with pectus excavatum tend to experience symptoms of fatigue, shortness of breath upon mild exertion, chest discomfort and tachycardia [13]. Studies have also shown a higher incidence of mitral valve prolapse and dysarrhythmias in patients with pectus excavatum [13].

Although it is known that this chest wall deformity can cause RV dysfunction and affect the morphology of the RV, it has been unclear to what extent this occurs. While there have been numerous advances in cardiac imaging modalities, there have been limited studies to further investigate the impact of pectus excavatum on RV function. Mocchegiani and colleagues conducted a study in which 28 subjects with mild to moderate degrees of pectus excavatum underwent two-dimensional echocardiography to assess RV function and anatomy compared to 24 control subjects with normal chest wall architecture [11]. In subjects with pectus excavatum, mean RV outflow tract diameter at the aortic root was narrower (1.4 ± 0.3 cm/m^2) and end diastolic (10 ± 2.3 cm^2/m^2) and end systolic (5.8 ± 1.4 cm^2/m^2) areas were larger than those in normal controls (1.6 ± 0.3, 8.6 ± 1.7, and 4.5 ± 1.2 cm^2/m^2 respectively, $p < 0.013$). The magnitude of the abnormality was related to the level of chest wall deformity evaluated by chest radiography. They concluded that patients with pectus excavatum of moderate to severe degrees had associated alterations in RV function and morphology. Specifically, patients with pectus excavatum were found to have a higher prevalence of morphologic abnormalities including global dilatation, rounded apex, localized sacculations of the ventricular wall, trabecular

hypertrophy, and structural changes of the moderator band (Fig. 22.3) [11].

Given that patients with pectus excavatum present with non-specific symptoms including dyspnea, fatigue, chest discomfort and tachycardia, it is often difficult to distinguish a cardiac etiology from a non-cardiac etiology in these patients. Citing improvement in indices of cardiovascular function following corrective chest wall surgery in patients with pectus excavatum, some investigators have suggested that the deformed chest wall may contribute to cardiopulmonary impairment [14, 15]. Although cardiopulmonary impairment can contribute to symptoms in patients with pectus excavatum, and guide the need for and timing of corrective surgery, guidelines for establishing the role of impaired right ventricular function in informing the indication for surgery have yet to be established [14, 15].

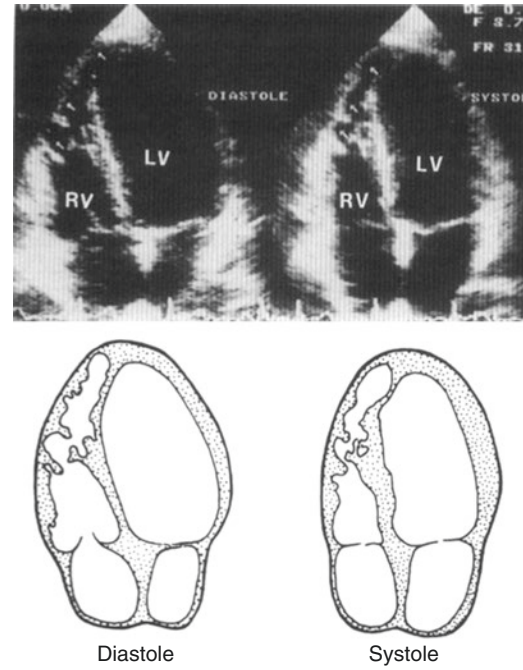

Fig. 22.3 Findings on four-chamber echocardiographic view of patient with pectus excavatum in diastole (*left*) and systole (*right*). The illustrations demonstrate morphologic changes including rounded apex and multiple sacculations found in the right ventricle (RV) (With permission from *Am J Cardiol*. 1995;76(12):941–6. Elsevier)

Principles of Echocardiographic Evaluation of Right Ventricular Function in Patients with Pectus Excavatum

As previously noted, assessing RV function through two-dimensional echocardiography is generally quite challenging given the location of the RV in the chest wall cavity and its complex morphology. Echocardiographic imaging in patients with pectus excavatum is even more limited, and is further hindered by geometric alteration to the already complex anatomy, including horizontal shift of the right ventricle within the chest, and overall abnormal cardiac motion. Visual estimation, as is most commonly used in RV functional assessment, often leads to even the most experienced echocardiographer to call a potentially normal right ventricle as abnormal. It is possible, that with the advent of three-dimensional echocardiography, that some of these limitations may be removed by quantitative assessment with volumetric analysis, but no such study in this population has yet been performed [13].

As qualitative assessment of right ventricular function is generally limited by echocardiography, and is likely further limited in patients with chest wall deformities, our group sought to identify if qualitative versus quantitative measures of RV function in patients with pectus excavatum were comparable to cardiac magnetic resonance (CMR) imaging. We compared the use of RV percent fractional area change and tricuspid annular plane systolic excursion in centimeters to qualitative echocardiographic assessment by trained experienced echocardiographers at a major academic medical center, and utilized CMR as a gold standard. In our small and limited cohort of patients with at least moderate pectus excavatum (pectus index by CMR, measured as maximum transverse diameter of the chest divided by the shortest sternospinal distance, 5.5 ± 2.5), quantitative measurements were found to be more accurate over qualitative measures when evaluating the right ventricle. In fact, all of the patients in our cohort were noted to have mild to severe RV

dysfunction by visual estimation, but in fact, had normal TAPSE (1.9±0.5 cm) and normal RV FAC (35.8±11.8%). They were further noted to have an overall normal RVEF as measured by CMR (53.5±5.4%). Given the overall normal RV function as assessed by CMR and quantitative echocardiography, none of the patients were recommended to have surgical correction, despite visual estimation of significant RV dysfunction. Given the lack of consensus as to the indications for surgery, an accurate assessment of RV function is necessary to guide decision-making in this complex patient population [15, 16].

Magnetic Resonance Imaging of Right Ventricular Function in Patients with Pectus Excavatum

Studies examining the changes on CMR imaging in patients with pectus excavatum are extremely limited. Saleh et al assessed RV function in patients with significant pectus excavatum (pectus index of 9.3±5) compared to control patients (pectus index of 2.3±0.4) using CMR. Their results demonstrated that compared to controls, patients with pectus excavatum have reduced RV short axis diameter, increased RV long axis diameter and approximately a 6% reduction in RV ejection fraction (53.9±9.6 versus 60.5±9.5, p=0.013). They also found that myocardial shortening and contractility was reduced along the short axis whereas increased on the long axis, which was hypothesized to be a compensatory change to preserve RV ejection fraction [17]. Further, Oezcan and colleagues compared echocardiography and CMR in 18 patients with pectus excavatum with a pectus index of >3.0 by CMR. They demonstrated that by CMR, cardiac leftward displacement was noted in 50% of patients, with minimal pericardial effusion noted in 56%, RV localized wall motion abnormality in 44%, and diminished RV systolic function in 44%. A completely normal cardiac examination was found in 33% of patients when assessed with echocardiography, but in only 11% when assessed by CMR, indicating improved sensitivity for the detection of RV functional abnormality by CMR imaging [18]. Despite its expense and limited accessibility, CMR is the ideal imaging modality for assessment of RV function in patients with pectus excavatum, as volumetric analysis is less limited by the chest wall deformity.

Conclusion

Imaging of the right ventricle is complicated and difficult, and every imaging modality has its own limitations. While CMR has evolved as a gold standard, echocardiographic evaluation is more clinically available, and considerably less expensive. RV functional assessment in patients with pectus excavatum is extremely important, as it may offer insight into symptomatic patients with potential cardiopulmonary impairment. Nonetheless, the evaluation of RV function is even more complicated in this unique population due to the inherent changes involved with the chest wall deformity, and is likely best performed using a multi-modality approach, including both quantitative echocardiography as well as CMR imaging to obtain the best possible assessment of RV performance to guide clinical decision making and the indication for surgical repair.

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Long Term Cardiopulmonary Effects After Surgery

23

David Sigalet

Introduction

Pectus excavatum is the most common chest wall deformity in children representing 90 % of all congenital chest wall deformities [1]. As outlined in Chaps 2 (Classification of Chest Wall Deformities) and 4 (Ultrastructural changes in cartilages), it has an incidence of approximately 1:300 to 1:400 in white males, In sporadic cases, although approximately 40 % of patients have a relative with the deformity, no specific genetic link has been established. Although the pathogenesis is unclear, the deformity results in a reasonably consistent pattern of an unbalanced overgrowth in the costochondral regions, which is typically more pronounced over the lower ribs [2]. Patients with pectus excavatum frequently complain of a combination of physical and psychological consequences of the defect. The subjective effects of the alteration on appearance are irrefutable; these may be rising with the increasing emphasis placed on appearance in modern culture (Chap. 8 Self and social perception in CWD). However, it is often more acceptable for patients and their families to focus on the effects of chest wall defects on cardiopulmonary physiology and physical activity. The effects of pectus carinatum on physical activity are highly variable; accordingly this discussion will focus on the effects on Pectus Excavatum (PE). Anecdotally, many patients with PE will complain of dyspnea on exertion and decreased ability to perform prolonged rigorous activity compared to their classmates [3–5]. These same patients often subjectively report an increase in exercise tolerance and improved self image post repair [4–7]. Quantification of the effects of surgical intervention would appear to be a straightforward matter, given the dramatic change in chest wall conformation which results from repair. However, there have been relatively few rigorous studies which have examined patients pre and post-operatively, and none which have included a comparator group of un-operated controls [8].

Today this topic has become much more commonly discussed because of the development of techniques utilizing a minimally access approach for repair of pectus excavatum (MARPE), as described by Nuss et al. [9] Patients and families are much more interested in discussing surgery for the repair of the pectus defect since the advent of this approach and there has been a dramatic increase in operations for the repair of pectus defects. Accordingly, health care providers are now routinely asked about the potential benefits of repair in pediatric patients with any variation of a pectus abnormality.

In order to understand the impact of repair, it is helpful to first assess the effects of pectus, if left uncorrected, on pulmonary and cardiac

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function individually, and the effects on overall aerobic exercise capacity. It is then useful to examine the effects of repair on these same parameters, and the added impact of the type of repair, the open (Ravitch) technique, or the MARPE technique.

The comparisons outlined are based on a review of the major published reports from the literature, combining studies from the past with more recent studies. The studies reviewed have the largest cohorts, with defined testing parameters, clear selection criteria, and normalization of values in growing children to age and sex matched controls.

Effects of Pectus Excavatum on Pulmonary and Cardiac Functioning

The natural history of pectus excavatum (PE) in the typical sporadic case is one of a noticeable depression prior to puberty. However, during the pubertal growth spurt there is typically a dramatic worsening of the depth of the depression, and with this, an increase in the perceived effects on exercise tolerance. Thus, in reviewing the documented changes associated with PE, it is also useful to consider the age, and maturational status of the patient, as well as the depth of the defect [10]. Examining the results of the largest studies in the literature, there is a clear effect of pectus excavatum in un-operated patients on pulmonary function. Subjects show a clear reduction in forced volume of expiration in one second (FEV1), the forced vital capacity (FVC), without a major effect on total lung capacity (TLC) (Table 23.1). However, none of the studies of PE patients have shown a reduction in cardiac output or stroke volume at rest, when normalized for age and weight (Cardiac index) (Table 23.2) [8, 11–13]. There is evidence that the effects of the pectus compression are dependent on the position of the patient; Zhao et al. showed that in patients with PE stroke volume decreased significantly during exercise when the exercise was done in the upright position, while when exercise was done in the supine position, there was no effect. This has been validated in a second study [14, 15]. Further there is strong evidence that patients with a significant pectus defect have a high rate of mitral valve prolapse, as well as increases in right sided atrial pressure which results in shunting through a patent foramen ovale [13, 14, 16]. Finally, results from autopsy studies, as well as long term population based studies suggest that PE may be associated with an increased risk of cardiac events in adulthood; this is of even greater importance in patients with Marfan's syndrome [17, 18].

Pectus Excavatum does appear to be associated with a modest reduction in the ability to perform aerobic exercise. Patients undergoing stringent exercise testing have been noted to have a reduction in VO_2 max, which is maintained into adulthood (Table 23.1) [8, 13, 19]. The cause of this is still uncertain, however, results of some studies suggest a relationship between a reduction in VO_2 max and a reduction in stroke volume during exercise [8, 13, 19]. In these studies the O_2 pulse, measured under the load of exercise, was decreased compared to age adjusted norms. The O_2 pulse is the ratio of VO_2 max divided by heart rate, and is directly related to stroke volume [20]. This has been replicated in studies using conventional 2-D echocardiography to directly visualize cardiac filling during exercise [14].

With significant pectus defects, recent studies have shown a significant decrease in the efficiency of the lungs in moving air during exercise, reflected in the volume of air moved at peak exercise (VE), in both adolescent and adult patients (Table 23.1).

In reviewing these studies, it is important to recall that these findings are primarily based on patients who present for repair of PE defects; this may introduce a significant selection bias. Patients who are motivated to seek intervention may have greater restrictions than those patients who are not, even with a similar degree of anatomic change. Further studies, based on population based cohorts, are required to explicitly examine the relationship between age, maturational status, the depth of the pectus depression and physiological effects on cardio-pulmonary function.

Table 23.1 Effect of pectus excavatum on pulmonary and cardiac function at different ages

	n	Age	Haller Index	FVC	FEV1	TLC	VO_2 max	O_2 pulse	VE
O'keefe et al.	67	13.9 ± 2.3	4.3 ± 1.3	91 ± 19	81 ± 17	99 ± 16	70 ± 15	76 ± 15	70 ± 16
Lawson et al.	408	13.4 ± 1.9	4.8	87 ± 14	86 ± 14	n/a	n/a	n/a	n/a
Castellani et al.	59	15.7 ± 4.5	–	91 ± 14	–	–	83 ± 12	–	–
Neviere et al.	120	27 ± 11	4.5 ± 1.1	92 ± 17	92 ± 18	95 ± 17	77 ± 2	91 ± 12	54 ± 11

Results: Values as % of expected normal values for age as mean ± SD

Table 23.2 Effect of repair of pectus excuvatum on pulmonary function

Operative technique		Preoperative				Post-operative			
Ravitch	n	Age	FVC	FEV_1	TLC	Time from OR	FVC	FEV_1	TLC
Quigley	15	16 ± 3	81 ± 17	–	–	0.7 years	80 ± 16 (−1)	–	–
Derveaux	32	17 ± 8	90 ± 10	93 ± 13	–	10	73 ± 13 (−17)	78 ± 16 (−15)	–
Morshuis	152	15.3 ± 5.5	78 ± 13	79 ± 15	84 ± 12	8.1	71 ± 13 (−7)	73 ± 16 (−6)	74 ± 12 (−10)
Kowalewski	34	13.4	80 ± 17	74 ± 17	–	4	84 ± 18 (+4)	78 ± 22 (+4)	–
Nuss									
Lawson	45	11.4	85 ± 12	84 ± 11	–	4.1	90 ± 11(+5)	89 ± 10 (+5)	–
O'Keefe	67	13.9 ± 2.3	91 ± 19	81 ± 17	99 ± 16	2.3	99 ± 23 (+8)	89 ± 20 (+8)	105 ± 19 (+6)
Borowitz	10	13.4 ± 3	91 ± 13	91 ± 12	91 ± 13	3	86 ± 17 (−5)	89 ± 14 (−2)	91 ± 10 (0)
Castellani et al.	59	15.7 ± 4.5	91 ± 14	–	–	5	88 ± 13 (−3)	–	–
Neviere et al.	70	27 ± 11	92 ± 17	92 ± 18	95 ± 17	1	90 ± 17 (−2)	89 ± 17 (−3)	93 ± 16 (−2)

Results: Values as % of expected normal values for age, () values are change from preoperative. Data: mean ± SD

Effects of Correction of Pectus Excavatum

Many groups have described the effects of repair of PE using a variety of measures related to cardiopulmonary function. Following the criteria for selection of articles outlined above, there have been a number of recent studies performed which have carefully documented the effects of operative correction of PE on cardiac and pulmonary function and extended these to examining the impact on maximal exercise capacity. In reviewing these findings it is important to recognize that there may be an additional effect of the surgical technique used in the correction of PE. Thus, it is useful to separate the studies of the effects of correction into the two major categories of operative techniques: variations of the open, or Ravitch technique and the MARPE techniques. The effects on pulmonary function are outlined in Table 23.2, the effects on cardiac function in Table 23.3, the overall effect on the peak aerobic exercise capacity are detailed in Table 23.4.

Effects of Correction on Pulmonary Function

In general, the methodologies used in these various reports are similar. Pulmonary function has typically been measured using a standard testing protocol at rest [21]. Importantly, in more recent studies, pulmonary function is also monitored as part of the exercise testing protocol, so that lung

volumes and the total volume of air moved while under the physiological stress of exercise can also be measured. The major metrics assessed have been the total lung capacity, forced vital capacity, and the volume of air expired over 1 s, or FEV_1. Interestingly in the studies reported, there was no improvement in pulmonary function in patients corrected using the Ravitch technique; moreover, there appears to be a mild decrease in lung function following open repair (Table 23.2). This was true for studies reported over a long time continuum, so it is likely that even with variations in the technique, such as the trend to reducing the extent of the dissection, there will not be an added benefit to lung function. However, in pediatric patients corrected using the Nuss technique, it does appear that PE repair improves FEV_1 significantly (Table 23.2). The effects on FVC appear to show a similar modest, but significant increase in function, while there does not appear to be an impact of repair on TLC. (which is not much affected by the PE defect primarily). It is important to note that these studies were done following removal of the bar; studies done with the bar in place show a clear reduction in pulmonary function [3, 22, 23]. In addition, the results of two recent studies suggest an interaction with the developmental stage of the patient at the time of repair; in adult patients undergoing repair of PE using the Nuss technique, there was no significant improvement in pulmonary function; similarly in patients under 11 years of age, there appears to be less of an effect [7, 13].

Effects of Correction of Pectus Excuvatum on Cardiac Function

Cardiac testing is typically done using echocardiography at rest. There are many small studies in the literature which are difficult to interpret because of variations in the endpoints reported. However, it would appear from the larger series, with cardiac testing done at rest (and standardized for height and weight) that there is no significant change following pectus repair (Table 23.3). Further, there does not appear to be a difference in the effects on cardiac function between the open repair, and the MARPE technique. However, there does appear to be an improvement in stroke volume at exercise following correction of the pectus depression; there also is strong evidence that correction improves mitral valve prolapse, and abnormalities of atrial pressure and flow [13, 14]. The very long term implications of these changes is not known, but in general optimization of cardiac functioning can only be viewed as a positive factor in maintaining health and well-being.

Effects of Correction of Pectus Excuvatum on Peak Aerobic Exercise Capacity

These studies have typically been done using either a cycling or treadmill ergometer, measuring the oxygen consumption of subjects as they exercise. The exercise load is increased, until the rate of CO_2 production shows a sudden increase, indicating that the subject has switched from aerobic to anaerobic metabolism (the anaerobic threshold) [24]. This indicates the upper threshold of aerobic exercise capacity, and at this time, the volume of oxygen that is being utilized by the patient is measured; this is the VO_2 max. The level of conditioning of the subject is an important factor in this. VO_2 max is a composite of the functioning of the lungs in gas exchange, the heart and vascular system in transporting, and the muscles in utilizing oxygen [24]. Aside from the amount of oxygen used, important measures relating to peak exercise capacity are the O_2 pulse (volume of oxygen/heart beat, a measure of stroke volume), maximal heart rate, respiratory reserve and the minute volume of breathing.

In the studies reviewed, following repair of PE, the VO_2 Max showed a modest trend towards improvement, while the maximal heart rate at exercise did not change (Table 23.4). Interestingly, there was a trend to a decrease in minute volume of breathing at maximal exercise, and an increase in the O_2 pulse. Although difficult to measure objectively, in those studies which reported the patient's subjective impression, the perceived capacity to exercise was increased [4, 19]. Similarly to the

Table 23.3 Effect of repair of pectus excuvatum on Cardiac function

Operative technique			Pre-operative				Post-operative				
Ravitch	n	Age	Ejection Fraction	Stroke volume	Cardiac output	Cardiac index	Time from OR	Ejection fraction	Stroke volume	Cardiac output	Cardiac index
Peterson	13					3.2					3.2 (0)
Kowalewski	12	13.4		9.8 ± 2.8	–	–	1	–	21.8 ± 5.1 (+12)	–	–
Nuss											
O'keefe	67	13.9 ± 2.3	–	68 ± 22	4.7 ± 1.6	3.2 ± 1.1	2.3	–	75 ± 21 (+7)	5.1 ± 1.3 (+0.4)	3.0 ± 0.8 (–0.2)
Neviere et al.	70	27 ± 11	63 ± 7	–	–	–	1	65 ± 7(+2)	–	–	–

Results: Values as % of expected normal values for age, ( ) values are change from preoperative. Data: mean ± SD

Table 23.4 Effect of repair of pectus excavatum on peak aerobic exercise capacity

Operative technique		Preoperative				Post-operative			
Ravitch	n	Age	VO_2Max	VE	O_2 Pulse	Time from OR	VO_2 Max	VE	O_2 Pulse
Quigley	15	16 ± 3	81 ± 17	58 ± 21	76 ± 14	0.7 years	80 ± 16 (−1)	62 ± 21(+4)	91 ± 15(+10)
Morshuis	35	15.3 ± 5.5	70 ± 4	79 ± 15	70 ± 12	8.1	74 ± 22 (+4)	73 ± 16 (−6)	74 ± 21 (+8)
Nuss									
O'keefe	67	13.9 ± 2.3	70 ± 15	70 ± 17	76 ± 14	2.3	72 ± 18 (+2)	64 ± 18 (−6)	81 ± 18 (+5)
Borowitz	10	13.4 ± 3	85 ± 15	60 ± 14	–	3	88 ± 1 (+3)	56 ± 12(−3)	–
Castellani et al.	59	15.7 ± 4.5	74 ± 14	–	–	5	71 ± 12(−3)	–	–
Neviere et al.	70	27 ± 11	77 ± 2	54 ± 11	91 ± 12	1	87 ± 2 (+10)	59 ± 13 (+5)	106 ± 8 (+15)

Results: Values as % of expected normal values for age, ( ) values are change from preoperative. Data: mean ± SD

findings related to cardiac functioning, there was no difference between the effects of the open and closed techniques of correction.

These results show that with long-term follow-up after repair of PE with the Nuss procedure in pediatric patients there is sustained improvement in pulmonary function, especially the FEV_1. This is more significant in those patients operated on during early-mid puberty, and does not appear to occur in patients operated on in adulthood, which suggests that for patients still completing their pubertal growth spurt some element of developmental remodeling with growth occurs following the repair [13]. However, there are no positive effects of the open repair on pulmonary function; indeed it appears that the long term effects of the open repair are deleterious [8]. The detailed observations from the studies showing an improvement in pulmonary functioning post repair suggest that the primary cause of improved pulmonary function post repair is an increased FVC [7, 19]. TLC consists of FVC + RV; the change in TLC and FVC was essentially the same, thus the increase in TLC was likely the result of increased FVC. Similarly, the ratio of mean FEV_1/mean FVC was essentially the same pre and post repair indicating no change in airflows and that the change in FEV_1 was also primarily related to a change in FVC [19].

These findings differ from the accumulated experience reported in a recent meta-analysis, which did not show an improvement in global pulmonary function following repair [25]. It is important to note that the majority of the patients reported in the meta-analysis underwent an open repair. Further studies are required to validate these findings, and to define the influence of such features as the timing of surgery relative to puberty, and the severity of the original defect on long term pulmonary capacity.

These factors may also play a role in the improvement in exercise capacity noted following repair. Normalization of the chest wall configuration should improve both the delivery of oxygen to the lungs and the ability of the heart to transport oxygenated blood. During ventilation, the thoracic cavity uses the diaphragm and extrinsic muscles of the chest wall to act like a bellows to drive air in and out of the lungs. When the normal configuration of the chest wall is distorted it has been postulated that the efficiency of the extrinsic musculature as well as the diaphragm is reduced [26]. There is evidence to suggest that with increased deformity (Haller index) pulmonary function is progressively decreased [27]. Therefore, upon correction of this abnormality it can be postulated that an improvement in chest wall musculature efficiency as well as the diaphragm function should occur. The interaction of repair on the dynamic function of the lungs at exercise is suggested by the observed decrease in Ve required at maximal exercise despite an increase in the amount of oxygen delivered. (i.e. the subjects are more efficient; they generate more useful work with a lower volume of air exchanged

following repair) [13, 19, 28]. This observation indicates that the patients' are not using as much of their maximum breathing capacity post repair for the same goals of exercise; there is an increase in respiratory reserve. This is a new twist on an old argument; with the increased sensitivity of modern exercise testing equipment more detailed studies of the pulmonary functioning under the load of exercise should be done to substantiate this in future studies.

Regarding the effects of correction on the second component of exercise physiology, oxygen delivery by the heart, the results suggest that both methods of repair appear to improve cardiac functioning, under load (Table 23.4). As described above, PE creates a fixed restriction in the chest volume which compresses the heart, decreasing stroke volume and hence cardiac output, especially at high load and with the patient in an upright position. The effect on stroke volume is best measured during exercise, which is technically challenging; in the studies quoted the O_2 pulse was used as a surrogate indicator of stroke volume. Indeed, the results show a modest, but significant increase in O_2 pulse, following correction in all the studies reviewed (Table 23.4). This finding is further supported by the report of Coln et al., who used echocardiography during exercise to directly visualize an improvement in cardiac chamber compression following pectus repair [14].

It is important to point out that a major limiting factor in many of the studies done in pediatric patients has been a failure to document the amount of exercise and sport the subjects are participating in. Activity and training will have a large additive effect on the VO_2 max and the general ability to exercise [24]. This may be further influenced by the length of time from the original operation, the timing of the removal of the bar, and the pattern of practice of the attending surgeon in allowing patients to resume full activities post-procedure. In general, many centers are finding that over the 3–4 year time from bar insertion to removal, the average level of aerobic activity of patients appears to decrease [19, 22]. It is important to document the level of ongoing sport and fitness training of the subject in any future studies. to gain more clarity about the relationship between exercise capacity and the long term effects of repair of PE. It is interesting to note that in a large study of adult patients with PE undergoing correction who had stable exercise patterns the greatest improvements in VO_2 Max was in trained athletes [13].

Overall, the results from several recent studies have added to our understanding of the effects of correction of PE. There appears to be a decrease in pulmonary function at rest following correction using the open technique but an improvement if correction is done using the MIRP. It would seem that the procedure should be done while the patients are still skeletally immature to effect a significant increase in pulmonary capacity. There does not appear to be an effect of correction of PE on stroke volume or cardiac function at rest, but there is an improvement in mitral valve prolapse, and in shunting at the atrium; these effects appear to be independent of the method used for surgery. There is modest evidence to show that correction of the pectus defect will increase the peak aerobic capacity of the patient; this also appears to be independent of the type of surgery used. However, there are a number of confounding factors to be examined in future studies, such as the effect of timing of the surgery, the relative severity of the defect, the impact of co-morbidities, and the impact of ongoing training and engagement in sport on the activity capacity of the individual patient. With these caveats in mind, the effects of a specific pectus configuration in an individual patient can be discussed, and the findings outlined used to guide the discussion of the potential benefits of correction for that individual.

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Rib Cage Anomalies in Severe Osteogenesis Imperfecta – Functional Investigations

24

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Abbreviations

E	expiration
f_B	breathing frequency
FEV_1	Forced Expiratory Volume in one second
FVC	Forced Vital Capacity
I	inspiration
OEP	Opto-Electronic Plethysmography
OI	Osteogenesis Imperfecta
RV	Residual Volume
S.D.	Standard Deviation
TLC	Total Lung Capacity
V_{AB}	Abdominal Volume
V_{CW}	Chest Wall Volume
V_E	Minute ventilation
V_{RCp}	Pulmonary Rib Cage Volume
V_{RCa}	Abdominal Rib Cage Volume
V_T	Tidal Volume
Φ_{RC}	phase shift angle between variations of $V_{RC,P}$ and $V_{RC,A}$
Φ_{TA}	phase shift angle between variations of $V_{RC,P}$ and V_{AB}
%pred	percentage of predicted values

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Structural Alterations of the Thorax in Osteogenesis Imperfecta

Osteogenesis Imperfecta (OI) is a complex genetically heterogeneous group of congenital disorders of collagen synthesis involving all the bones of the skeleton and characterized by brittle bones leading to different levels of deformities and frequent multiple fractures.

OI exhibits a broad range of clinical severity, ranging from multiple fracturing in utero to normal adult stature and a low fracture incidence [1, 2]. The traditional classification proposed by Sillence et al. [3] considers four main groups of OI syndrome according to the phenotypic variability, i.e. type I, II, III, and IV, however, new types have been recently introduced [4, 5]. OI type I is the mildest form characterized by blue sclerae. OI type II, also known as congenital OI, is perinatal lethal. OI type III is the most severe non-lethal form, characterized by multiple fractures with normal sclera, progressive long bones bowing, bone deformities and short stature [6]. OI type IV, finally, is a moderate form with normal sclerae [7].

In OI the most severe forms of the disease are commonly associated with severe and progressive deformities of the sternum, the chest and the spine. The most common sternal deformities are represented by pectus carinatum and pectus excavatum. An incidence of pectus carinatum and excavatum of 21 and 16% respectively, was reported on a population of 129 patients with different forms of

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OI [8]. Sternal bowing and rarefaction, especially at the manubrium, producing a tumor-like mass appearance on computed tomography scans can also be present in OI [8]. The chest is generally small, with thickened and shorter ribs. Each rib has characteristic "beading" patterns secondary to repeated fracturing. Biopsies of ribs revealed that a rapid surface turn-over of histologically well-organized but osteopenic bone, whose rate does not appear to decline with age in osteogenesis imperfecta, is present [9]. The radiographs show that an abnormal configuration of the ribs, namely a posterior angulation of the ribs and an elongation of the vertebral pedicles are consistent features of OI disease, particularly in type III [10]. A complication of OI in the newborns can be flail chest defined as two or more fractured adjacent ribs producing a loose segment of the rib cage capable of moving in paradox to the remaining one [11].

Spinal deformity in OI, namely thoracic scoliosis and kyphosis [8, 12], is exacerbated by collapsing of thoracic vertebrae resulting in a more horizontal position of the ribs [13] and multiple vertebral fractures resulting in a disproportionally distributed growth of the chest and trunk that lead to chest collapsing as the spinal deformity worsens [14, 15]. On its concave side, the chest cavity is undersized and the lung alveoli do not have space to properly develop and expand [12]; on the convex side of the chest, the ribs are deformed and displaced. This combined deformity of distorted thoracic spine and abnormal rib positioning, which has been termed a "windswept" chest deformity, limits thoracic movements and lung expansion [15]. This severe restricted lung disease, that has been coined thoracic insufficiency syndrome [16], is associated with lung infections due to improper ventilation and perfusion. In addition, the severe deformity augmented pressure on the great vessels with resulting cardiac failure [17].

In OI patients, respiratory deficiency can be worsened by rib fractures, muscles weakness, possible pulmonary hypoplasia and abnormal pulmonary fibroblast collagen [18–20]. As a consequence, patients with more severe OI are at significant increased risk for premature death secondary to pulmonary disease. In fact, life expectancy in OI type IV is similar to general population, while in type III is reduced [21, 22] with cardiopulmonary insufficiency and respiratory infections being the leading causes of death in these patients, and their prevention and treatment are important factors for prognosis [18, 23].

Relationships between Altered Thoracic Structure and Respiratory Function in Osteogenesis Imperfecta

Although it is reasonable to postulate that structural alterations of the chest wall, namely spinal and rib cage deformities, may contribute to cardiopulmonary problems in severe OI, few authors reported data on deformities and pulmonary function [12, 18, 24]. In the 70s, Falvo et al., studied pulmonary function in patients with severe, moderate and mild OI to characterize their pulmonary abnormalities to assess anaesthetic risk. Thoracic deformity, manifested by an increase in the anteroposterior diameter of the chest, and kyphoscoliosis were found in all the severely involved patients. They found that the reduction of vital capacity and increase in residual volume (RV) and RV/TLC ratio (TLC: total lung capacity) were present only in patients with kyphoscoliosis [18]. In 1999, a cross-sectional radiological and spirometrical study was performed on patients with congenital (i.e.: fractures at or before birth) and tarda (i.e.: fractures after birth) OI. Thoracic cage deformity was assessed using modification of a method previously described in adolescent idiopathic scoliosis [25]. It was found that increasing thoracic scoliosis was strongly correlated with decreased forced vital capacity (FVC) while there was no correlation between kyphosis, chest wall deformities and FVC [12].

However, the above cited papers, although importantly suggesting that a relationship between thoracic deformities and pulmonary function is evident in OI, have intrinsic limitations. These include the use of radiographic measurements, either x-ray or CT, that expose patients to ionizing radiation and therefore are problematic for longitudinal patients'

monitoring and that mild, moderate and severe OI patients were considered as an overall group, masking possible differences in the different forms of the disease.

To partially overcome these limitations, the authors of the present chapter recently took advantage of Opto-Electronic Plethysmography (OEP), a new method that does not expose the patients to ionizing radiation by studying respiratory kinematics in a population of OI patients during spontaneous quiet breathing in awake diurnal state both in seated and supine position [24]. Chest wall geometry and thoraco-abdominal volume variations were separately compared in two forms of OI, namely type III and type IV. The feasibility of using optical noninvasive techniques to quantify and assess the structural deformities of the torso in patients with OI type III and IV has been recently reported by other investigators [26]. Statistically significant agreement between the evaluation of truncal deformities based on digital stereophotogrammetry and those obtained by direct manual measurements is present.

Opto-Electronic Plethysmography (OEP) [27, 28], is a recently developed technique able to measure both chest wall shape and thoraco-abdominal kinematics (namely, volume variations of the rib cage and abdomen during breathing). OEP is based on a number (6–8) of special infrared video cameras working at a sampling rate of 60 Hz and several retro-reflective markers placed on the trunk of the subject according to specific anatomical points from clavicles to pubis. When patients are acquired lying supine on the bed, 52 markers are placed over the anterior chest wall surface [29, 30], while an 89 markers configuration was used for measurements in seated position [28]. From 3D marker coordinates measured by OEP in seated position at end-expiration several geometrical parameters can be calculated to characterize chest wall geometry [30]. These include trunk height, chest wall surface and total and compartmental trunk volumes (i.e. the volume enclosed by the surface obtained by triangulating all the markers). Several additional parameters including medio-lateral and antero-posterior diameter, perimeter and cross sectional area can be calculated at five vertical levels (Louis' angle, xiphoid process, lower costal margin, umbilical and iliac crest) (Fig. 24.1). In our the recent study, to quantitatively describe the geometrical deformity of the rib cage ('pectus carinatum') in OI patients, the angles subtended at the sternal level on the transversal plane (Fig. 24.1a, c) and on the sagittal plane (Fig. 24.1b, d) were calculated.

Starting from the 3D positions of the markers the system computes the volume variations of the total chest wall (V_{CW}) by Gauss's theorem. It also provides volume variations of the three chest wall compartments: pulmonary rib cage ($V_{RC,P}$), under the action of rib cage muscles, abdominal rib cage ($V_{RC,A}$), under the insertional action of diaphragm and abdomen (V_{AB}), under the action of the diaphragm and the expiratory abdominal muscles. From variations of V_{CW} during quiet breathing the ventilatory pattern, in terms of minute ventilation, breathing frequency, tidal volume and percentage contribution of each compartment to tidal volume can be determined on a breath-by-breath analysis. Moreover, dynamic thoraco-abdominal asynchronies can be quantified by determining the phase shift angle (Φ_{TA}) between volume variations of $V_{RC,P}$ and V_{AB} waveforms. Φ_{TA} is derived from the ratio of the distance m delimited by the intercepts of $V_{RC,P}$ (y-axis)-V_{AB} (x-axis) loop on a line parallel to the x-axis at 50 % of $\Delta V_{RC,P}$ divided by ΔV_{AB} (s) and computed as $\Phi_{TA} = \sin\text{-}1(m/s)$ [31–34]. In a similar way, dynamic asynchrony between the two rib cage compartments can be calculated as the phase shift angle between $V_{RC,P}$ and $V_{RC,A}$ waveforms (Φ_{RC}).

Anthropometric Data, Chest Wall Dimensions and Spirometry in OI

A hallmark of patients with OI is short stature. It has been shown that the reduced standing height seems to be correlated with the different OI types and the severity of collagen defect. A relatively short trunk, assessed by a stadiometer, and disproportional body length are typical in OI [35]. Body height and trunk height, assessed by OEP, were

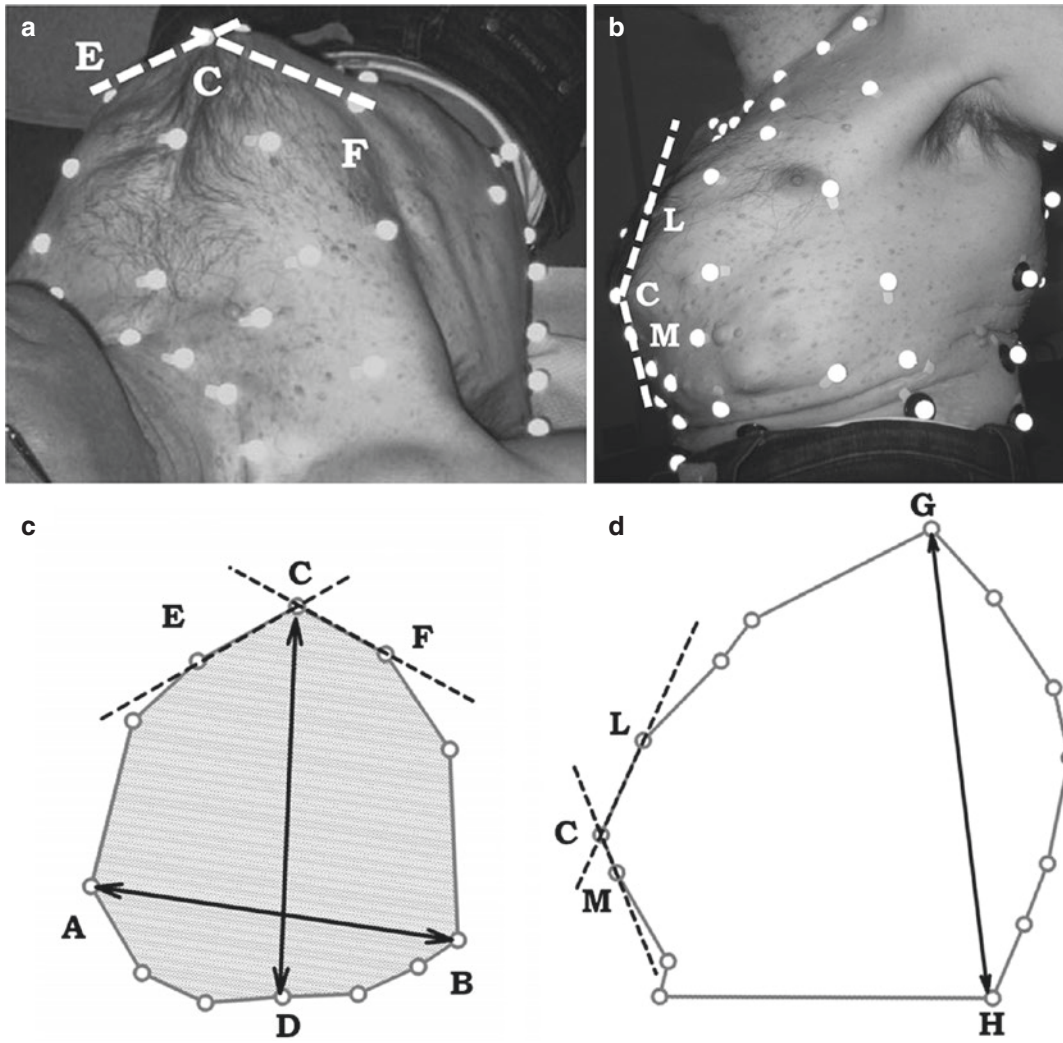

Fig. 24.1 Assessment of chest wall geometry and rib cage deformity by markers' projections – Experimental set-up for the analysis of chest wall volumes via optoelectronic plethysmography in supine (**a**) and seated (**b**) position on a representative OI type III patient. (**c**): schematic view of the markers on the transversal plane at the xiphosternal level, in which medio-lateral diameter (distance between markers *A* and *B*), antero-posterior diameter (distance between markers *C* and *D*), area (*grey area*) and transversal angle (angle formed between lines *CE* and *CF*) are shown; (**d**): schematic view of the markers on the sagittal plane in which trunk height (distance between markers *G* and *H*), and sagittal sternal angle (enclosed within lines *CL* and *CM*) are shown [24]

different among type III and type IV patients and a control group, being the lowest in type III OI form; but their ratio was not significantly different indicating that growth is disproportionally distributed in OI. This influences spirometric and lung volumes predicted values because they are computed using gender, age and height. According to the standard reference equations proposed by Quanjer et al. [36] and largely used in clinics, patients showed FVC and forced expiratory volume in one second (FEV_1) values (expressed as percentage of predicted values, %pred) within the normal range, with data of OI type III unexpectedly higher than predicted. Similarly, Takken et al. [37] showed that the reduced patients' height determines higher FVC and FEV_1 values even in children with OI type I, the mildest form of the disease. These high values of FVC and FEV_1 are

probably artifactual, at least in OI type III, because it is known that cardio-pulmonary insufficiency and respiratory infections represent the leading cause of death in these patients. In contrast, using more recent equations of Kuster et al. [38], FVC (%pred) and FEV_1 (%pred) became significantly lower in OI patients, particularly in OI type III (Fig. 24.2).

Chest Wall Deformity in OI

Among all the geometrical parameters that can be computed with OEP, only two resulted different among OI type III patients, OI type IV patients and healthy controls: trunk height, being lower in OI type III patients, and the xiphosternal angle. Patients with OI type III were characterized by lower angles (i.e., pectus carinatum) both in sagittal and transversal planes in contrast with the flatter sternum of both OI type IV and controls (Fig. 24.3). Severe thoracic scoliosis, indicated by an average Cobb angle values >40°, was present in both OI III and IV and no differences were observed.

Ventilatory Pattern in OI

In supine position, there were no differences in minute ventilation (V'_E), while tidal volume (V_T) and breathing frequency components were different, being lower the former and higher the latter in OI patients. No differences were found between type III and type IV. In seated position V'_E was lower in OI patients than controls due to lower V_T. Only in OI type III patients, breathing frequency was higher than controls. These results suggest that patients with severe Osteogenesis Imperfecta tend to breathe with a more rapid and shallow breathing pattern in both positions. The reduced expansion of the rib cage compartment (as described in the next paragraph) is probably the cause of the reduced tidal volume and the concomitant increased breathing frequency could represent the compensatory mechanism to guarantee a

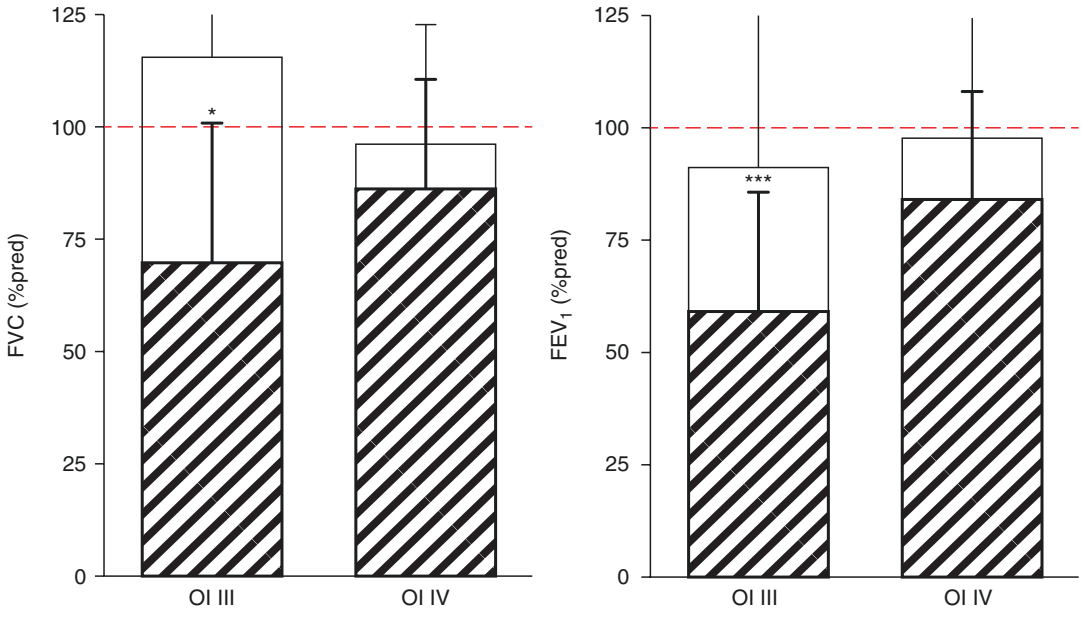

Fig. 24.2 Spirometry – Average values ± S.D. of forced vital capacity (*FVC*, *left panel*) and forced expiratory volume in 1 s (*FEV_1*, *right panel*) in OI type III and type IV patients. The volumes are expressed as percentage of predicted values considering both the reference values from Quanjer et al. [37] (*white bars*) and from Kuster et al. [38] (*dashed bars*). (*$p<0.05$, ***$p<0.001$, vs Quanjer et al. [36])

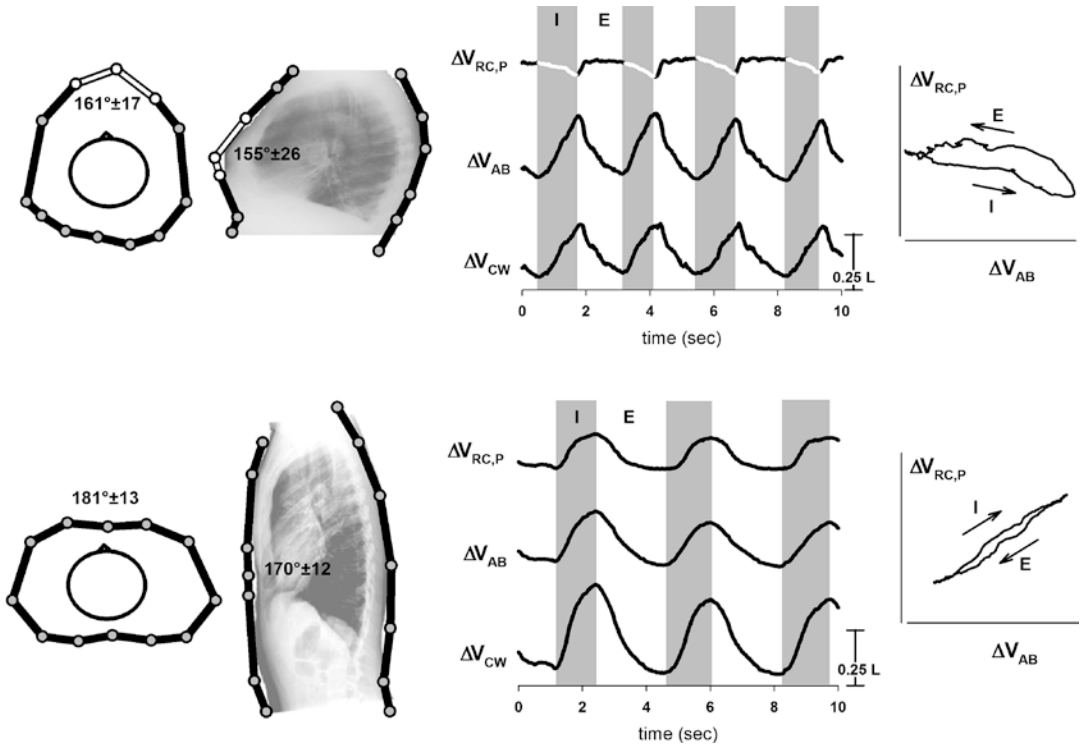

Fig. 24.3 Chest wall deformity and thoraco-abdominal volume variations during spontaneous breathing of OI type III and controls – Markers' projection on the transversal and sagittal views of the markers at xiphoid and sternal level (*left*); time courses of the volumes of the pulmonary rib cage, abdomen and total chest wall during 10 s of consecutive breaths at rest in supine position (middle); $\Delta V_{RC,P}$ versus ΔV_{AB} counterclockwise dynamic loop (*right*) on representative OI type III (*top panels*) and healthy control (*bottom panels*) subjects. (*I* inspiration, *E* expiration)

normal minute ventilation. However, apparently this mechanism is not fully effective in seated position, as shown by significantly lower minute ventilation in OI patients (Fig. 24.4).

Chest Wall Compartmental Volumes and Asyncrony in OI

In six out of seven patients with OI type III, paradoxical inspiratory movement of the pulmonary rib cage was detected during spontaneous breathing at rest in supine position. The percentage contribution of $V_{RC,P}$ to tidal volume in OI type III was reduced in seated position, being negative (i.e., paradoxical inward motion during inspiration) and lower compared to OI type IV and controls in supine. In seated position, no paradoxical motion was present, however, the percentage contribution of $V_{RC,P}$ to tidal volume was still significantly lower than controls. In OI type IV patients $V_{RC,P}$ contribution was lower than controls only in seated position. Similarly, the $V_{RC,A}$ contribution to V_T was reduced in OI type III patients compared to both OI type IV and controls in both postures. Hence, V_{AB} contribution to V_T was higher in patients with OI type III than the other two groups both in supine seated position (Fig. 24.5).

Asynchrony between pulmonary rib cage and abdomen (Φ_{TA}) was lower in OI type III patients than OI type IV and controls, both in supine and seated position. Within-rib cage asynchrony (Φ_{RC}) was lower in OI type III patients than OI type IV and controls, but only in supine position. In OI type III patients only, Φ_{TA} and Φ_{RC} were significantly different between seated and supine position. These results suggest that in OI type III

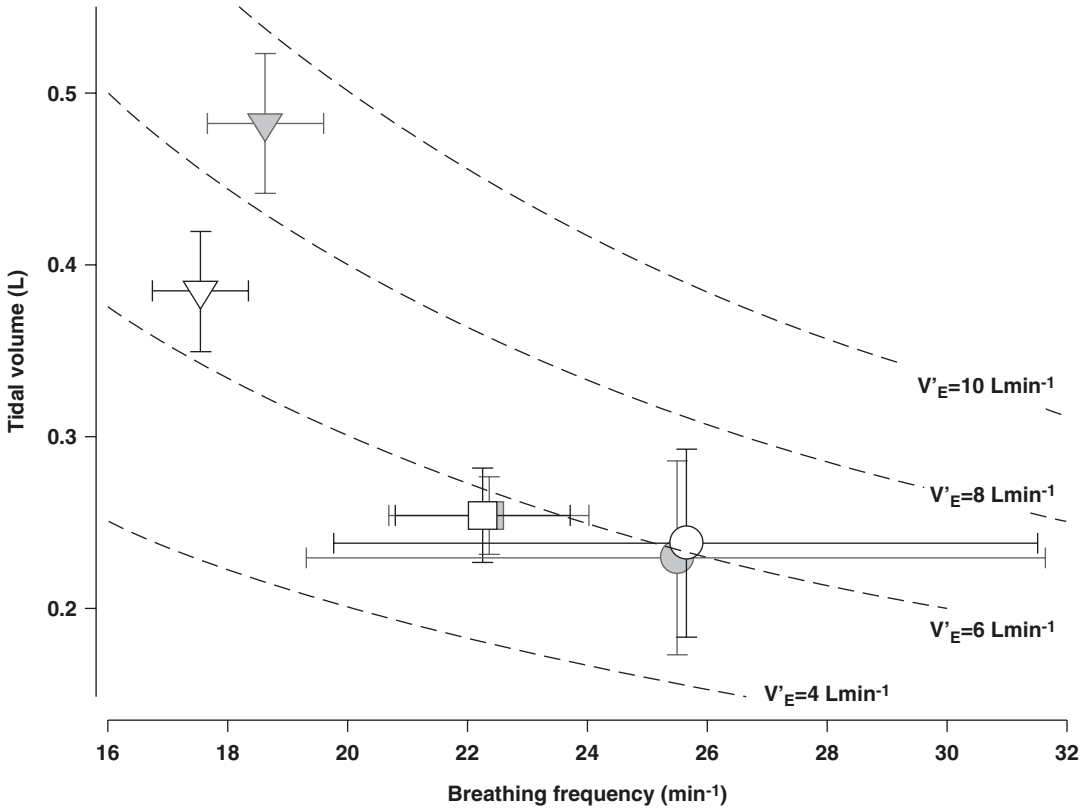

Fig. 24.4 Ventilatory pattern – Relationship between the average values ± S.D. of tidal volume and breathing frequency at rest in seated (*grey symbols*) and supine (*open symbols*) position of OI type III (*triangles*), OI type IV (*squares*) and healthy control subjects (*circles*). Short dashed lines represent isopleths of different levels of minute ventilation (V'_E) from 4 to 10 $Lmin^{-1}$, according to the relationship $V'_E = V_T \times F_B$ (V_T tidal volume, F_B breathing frequency)

patients the deformed and altered rib cage (i.e., pectus carinatum, more horizontal ribs and more compliant rib cage from brittle bones) alters the normal action of the intercostal muscles and makes the diaphragm to compensate for their reduced contribution to tidal volume. An additional indication that in these patients diaphragm function seems to be preserved is that they do not show significant nocturnal oxygen desaturation.

It can be hypothesized that the reduced angle between the ribs and the sternum modifies the action of the intercostal muscles and alters the "bucket-handle" motion of the ribs in relation to their fulcrum, the sternum [39]. Posture plays an important role not only in the distribution of tidal volume into chest wall compartments but also in the synchrony of their expansion. In our OI type III patients, not only the percentage contribution of $V_{RC,P}$ to tidal volume, but also thoraco-abdominal (Φ_{TA}) and rib cage (Φ_{RC}) asynchronies (Fig. 24.5) were significantly different between seated and supine position. The generally lower activity of the neck and rib cage muscles (scalene, sternocleidomastoid, and parasternal intercostal) in supine position [40], which in these patients is combined with the altered geometry of the rib cage, is likely the main reason of these differences.

Within-rib cage asynchronies and the paradoxical inward motion of the pulmonary rib cage observed in association with the expansion of the abdominal rib cage in OI type III in supine position also suggest that in these patients rib cage distortions are significant and therefore a substantial

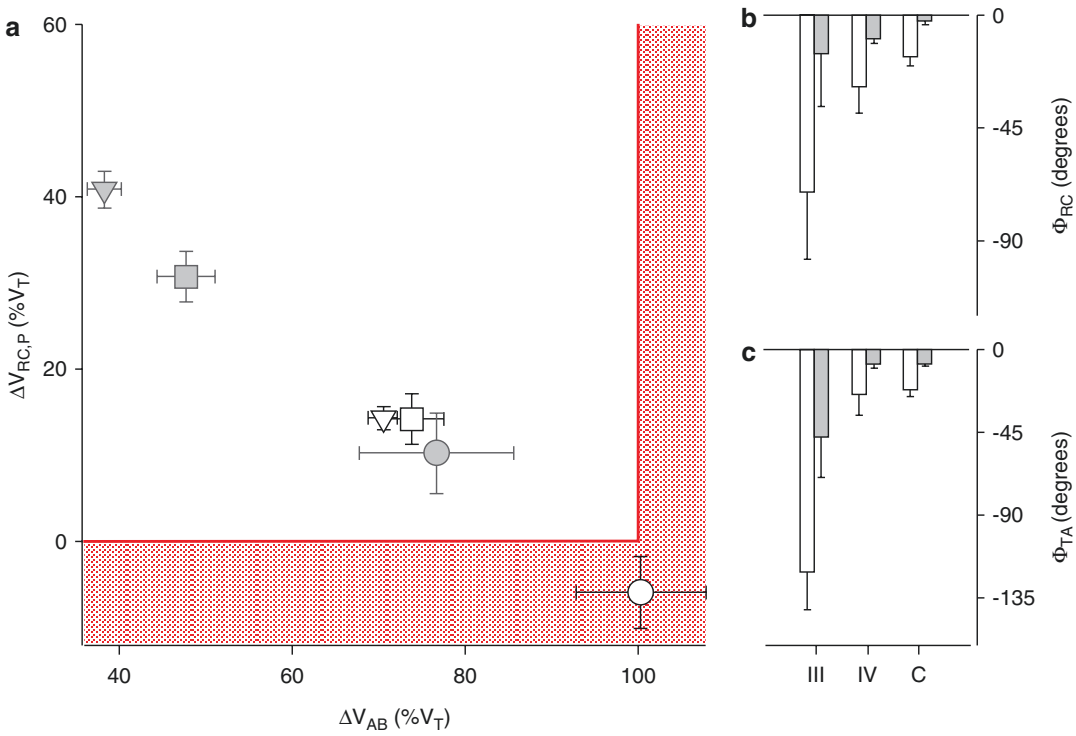

Fig. 24.5 Thoraco-abdominal contribution to tidal volume and asynchronies – (**a**) Relationship between the average values ± S.D. of the percentage contribution of pulmonary rib cage (*y-axis*) and abdomen (*x-axis*) to tidal volume in seated (*grey symbols*) and supine (*open symbols*) position of OI type III (*triangles*), OI type IV (*squares*) and healthy control subjects (*circles*). The red area represent the paradoxical motion area. (**b**) Average values ± S.D. of phase angle between pulmonary rib cage and abdominal rib cage volume (Φ_{RC}) during spontaneous quite breathing in OI type III, OI type IV and healthy control subjects (III, IV and C, respectively) in supine (*grey*) and seated (*white*) position. (**c**) Average values ± S.D. of phase angle between pulmonary rib cage and abdominal volume (Φ_{TA}) during spontaneous quite breathing in OI type III, OI type IV and healthy control subjects (III, IV and C, respectively) in supine (*grey*) and seated (*white*) position

fraction of the force developed by the diaphragm on the rib cage would go into distorting it, and only a small fraction into changing chest wall and lung volume. This is an inefficient way to breath [41] because rib cage distortions, minimal during breathing in normal subjects even during heavy exercise [42, 43], are costly.

Conclusions and Clinical Implications

It is known that patients with Osteogenesis Imperfecta are characterized by bone fragility, frequent fractures, bowing of the long bones, growth disproportionately distributed affecting predominantly trunk height more than the other trunk dimensions, and sternal deformities [1–8, 13]. In patients with severe OI a direct relationship exists between the structural modifications of the rib cage and the pattern of volume variations during breathing. Rib cage deformities, rather than scoliosis which was present in both OI types, is the determinant of the altered breathing pattern in type III. Patients with OI type III are characterized by pectus carinatum and inspiratory paradoxical inward motion of the pulmonary rib cage during quiet spontaneous breathing in supine position, associated to a high level of asynchrony between the three chest wall compartments. On the other hand, pectus carinatum, paradoxical motion and significant

asynchronies between chest wall compartments are not features of type IV patients. These findings suggest that functional assessment and treatment of OI should be differentiated in these two forms of the disease. Special attention should be paid on choosing the algorithm for the predicted spirometric values in Osteogenesis Imperfecta, to avoid overestimation of the spirometric volumes in the severe form of the disease. Finally, particular care should be given to early bracing to limit spinal deformities [8, 13], because it may compress the soft osteoporotic rib cage leading to exacerbate the deformation of rib cage geometry and therefore the correlated altered respiratory pattern shown in type III patients.

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Concepts in Allergy to Pectus Metal Implants

25

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Pectus excavatum, the most common deformity of the anterior chest wall occurs in 1:100 live births with a 3:1 male:female predominance. Three principal methods have been used to repair chest wall deformities, and include sternal turnover, sternal elevation and prosthesis implant [1, 2]. In 1998 Nuss described his 10 year experience with the use of a stainless steel implant utilized in a new technique for minimally invasive repair of pectus excavatum (MIRPE). The Nuss procedure has enjoyed wide-spread use and details of the procedure and review of complications including allergy have been reported in a number of sources [2–13].

Metal contact allergy is an acute or chronic skin inflammation stemming from repeated or prolonged exposure to metals in high concentrations, often in the form of jewelry, piercings, and clothing fasteners. Metal allergy is an important consideration when evaluating patients presenting with cutaneous complications following insertion of the pectus bar and device stabilizers. In a review of 1,015 pectus procedures, bar allergy accounted for 2.9 % of late postoperative complications [5]. Identifying patients with increased risk for developing cutaneous allergy to pectus implants is challenging. There is no definite genetic predisposition to metal allergy, although some research has linked mutations in the filaggrin gene complex with metal and jewelry allergy [14].Polymorphisms involving N-acetyltransferase, glutathione-S-transferase M and T, angiotensin-converting enzyme, tumor necrosis factor, and interleukin-16 have also been linked to predisposition to contact allergy [15].

The traditional stainless steel pectus bar is composed of 61 % iron, 18 % chromium, 14 % nickel, 2.5 % molybdenum, 2 % manganese, and small amounts (less than 1 %) of carbon, phosphorus, sulfur, nitrogen, copper, and silicon [16]. In the majority of cases metal allergy due to pectus implants is attributable to the nickel component of the bars. Nickel is the most frequent allergen found with diagnostic patch testing [17]. The prevalence of metal allergy for nickel is reported to be as high as 17 % for women and 3 % for men [18, 19]. Orthopedic implants associated with cutaneous or systemic hypersensitivity reactions include all forms of prostheses, stabilizers, rods, bars, plates and screws. Common allergens

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from these implants include nickel, cobalt, or chrome, or less likely materials in bone cement (e.g. methyl methacrylate, N,N,-dimethyl-p-toluidine and / or benzoyl peroxide) and sterilizing procedures (e.g. ethylene oxide) [20–23]. Although the prevalence of metal allergy among the general population is significant, most individuals will tolerate low levels of metals in prosthetic devices or implants without adverse reactions [20].

The pathophysiology of allergy to metal implants is thought to begin with an induction phase when the metal first comes in contact with the skin and stimulates the immune system to produce antigen specific T-lymphocytes. Repeat or continuous exposure to the allergen causes activation of theseantigen specific T-lymphocytes resulting in the cell mediated immune response that manifests clinically as allergic contact dermatitis [6, 24, 25]. Prior reports have shown that nickel ions can be leached in sufficient quantities from nickel containing braces which may induce nickel sensitization or elicit allergic contact dermatitis [26]. A similar mechanism could be the underlying process by which patients develop cutaneous or systemic manifestations of allergy to pectus implants [11].

Historically, reactions to a broad range of medical devices or implants have commonly been eczematous but may range in severity and presentation. Cutaneous findings have occasionally taken the form of urticaria, bullous eruptions, edema, or vasculitis. Potential systemic findings include: osteolysis, loosening, pain, burning, arthralgias, and myalgias. Prior reports specifically concerning pectus implants have reported nickel sensitivity to present with either cutaneous manifestations (eg. an erythematous patch along the chest wall) or in the form of non-cutaneous complications such as pericarditis and pleural effusions [5]. In 2007 Nuss's group wrote that in case of complications after the MIRPE procedure metal allergy should be considered after infection is excluded. In their retrospective review of pectus bar recipients 19 of 862 patients were diagnosed with nickel allergy. Ten of these patients presented with rash and erythema, five with pleural effusions, and one with a granuloma [6]. In a 2010 reivew of 1215 MIRPE cases Nuss's group reported that nickel allergy occurred in 35 (2.8 %) of patients [2].Pericarditis occurred in five of these patients, all prior to 2003, the year they began more rigorous screening for nickel allergy. The patients responded to indomethacin and prednisone and one required catheter drainage of the pericardium. Onset of symptoms was variable. In one small series, we reported that the mean and range of onset of cutaneous findings from the time of the procedure was 9.5 months, and 3–14 months respectively [11]. In another report a patient with recurrent pleural effusions had the bar removed after positive allergy testing [27]. In another case from the same study a female patient with nickel and chromate allergy received a stainless steel bar (which contains both metals) despite the allergy and did not develop pleural effusions until planned bar removal [27].

Making the diagnosis of allergy to a pectus implant is challenging – it is often a diagnosis of exclusion, supported primarily by a temporal relationship to device implantation, positive patch test results, and the absence of signs of infection (tenderness, fever, pain and response to antibiotics). Clinical suspicion is paramount and can be augmented by tools such as the guidelines revised by Thyssen et al. [20], summarized in Table 25.1.

A more straight forward approach to making the diagnosis of metal allergy couples clinical history and patch testing. Patch testing can be performed to determine if a type IV allergy exists to a specific metal allergen and is the gold standard for diagnosing allergic contact dermatitis. We patch test in accordance with the guidelines established by the North American Contact Dermatitis Group (NACDG) [28, 29]. First, the allergen is diluted in petrolatum or water to a concentration that does not produce active sensitization or irritation. A small amount of allergen is placed on to aluminum disks (8 mm), which is attached to strips of paper tape. Tests are applied to the upper back or midback, which must be free of dermatitis. The patches are left in place and kept dry for 48 h. Patch tests are removed after 48 h and read on day 2 or 3, and read again on

Table 25.1 Diagnostic criteria that support a causative association between metal release from an orthopedic implant and metal

Chronic eczema beginning weeks or months after the implant was placed
Eruption overlying the implant
Morphology consistent with dermatitis (erythema, induration, papules and vesicles) absence of other contact allergens or systemic cause
Histology consistent with allergic contact dermatitis
In rare instances, systemic allergic dermatitis (disseminated dermatitis typically localized in body flexures)
Positive patch test reaction to a metal used in the device or implant (often a strong reaction)
Serial dilution patch testing gives positive reaction to low concentrations of the metal under suspicion
Positive in-vitro test to metals, e.g. the lymphocyte transformation test for nickel, cobalt or chrome
Dermatitis is therapy resistant
Recovery after total removal of the offending implant

day 6 or 7. Reactions are graded on a scale from 0 (no reaction) to +3 (erythema in addition to vesicles/bullae). Patch testing should only be performed by those who have been trained to interpret patch test results.

In general when testing for putative orthopedic, endovascular and dental device allergy, we patch test with a series of metal allergens found in prostheses including titanium, vanadium, molybdenum, copper, aluminum, palladium, manganese, platinum, indium, gold, iron, tantalum, zirconium, and tin. Some device manufacturers may distribute patch test kits, which include materials that may not be found on the standard screening series. It may be difficult to distinguish implant allergy that is due to metal sensitivity from a reaction that stems from other materials encountered during the insertion of the device. For example, ethylene oxide gas, used in medical equipment sterilization, can elicit an immune response that mimics delayed-type hypersensitivity reactions. Allergies to latex, rubber, suture materials, antiseptic scrubs, wound dressings, adhesives, sterilizing agents, or antibiotics have all been reported and may mimic orthopedic implant allergy. For this reason when patch testing for prosthesis allergy we often evaluate for these potential allergies as well and also include other potential allergens such as surgical preps such as chlorhexidine and any topical or systemic antibiotics used intra- or post-operatively [27, 30].

Only a minority of patients will present with metal allergy following implant insertion, therefore we ordinarily do not recommend that clinicians perform routine pre-operative patch testing, unless the patient has a history of metal allergy or previous cutaneous or systemic reaction to an implant or device of concerning magnitude [20]. Further, the accuracy of pre-procedure patch testing is controversial. In one series of patients who developed suspected allergy to pectus implants pre-operative patch testing appeared to be an unreliable tool, as patients who were preoperatively tested with stainless steel alloy discs supplied by device manufacturer did not developed signs of allergy prior to implant insertion [11]. It is unclear if this is because the preoperative discs used were ineffective in producing reliable patch testing results, or if the patients were not sensitized to the allergen prior to insertion of the implant. Patch testing with metal salts rather than pieces of devices is generally preferred and better validated. In their 2010 report Nuss's group states "patients with a prior history of metal allergy, those with a family history of metal allergy, those who test positive to nickel, cobalt or some other constituent of stainless steel and patients who suffer from eczema should receive a titanium bar." [2] They recommend documentation of metal allergy with patch testing with TRUE test. Indications for patch testing in their 2007 article [6] included allergies to jewelry, orthodontic braces, metal buttons/snaps on clothing, and food, and also atopic patients with a history of eczema, allergic rhinitis and asthma. In April 2012, the Nuss's group required "all patients who will undergo pectus surgery to be tested for allergies to the metallic components of the implanted surgical stainless steel pectus bar" They recommended that this be performed with the following allergens: nickel sulfate 5 % pet, chromium chloride 1 % pet, potassium dichromate 0.25 % pet, copper sulfate 2 % pet, molybdenum chloride 0.5 % and manganese chloride 0.5 % pet [31].

While most patients with allergic dermatitis are patch test positive, some may have negative or indeterminate patch test results. In these instances other tests that may be of use include RAST testing, intra-dermal testing, in vitro lymphocyte transformation tests, or ELISA assays that measure cytokine activity.

Once a diagnosis of allergy to implant is made several therapeutic options exist. Because pectus implants are only in place for a few years some patients have been managed without early removal of the implants and stabilizers [11]. In the short term, dermatitis can be treated temporarily with topical corticosteroids and systemic antihistamines, both of which have a relatively benign side effect profile. Occasionally, a short-course of tapered systemic corticosteroids may be indicated; however these should be used with caution in pectus patients, given their young age. Removal of the implant is the most efficacious treatment option as metal allergy can only definitely be treated by avoidance of all contact with the allergen. Although contact allergy may be a chronic condition, most patients report resolution of symptoms within days to months once the allergen has been removed.

Titanium pectus implants are the recommended alternative for patients with suspected nickel allergy [6]. At some institutions titanium pectus bars have become the standard of care in order to avoid the complications of nickel allergy [11].Other advantages of the titanium bar include the possibility of undergoing MRI examination, its translucent appearance on X-rays, and its ability to pass through metal detectors [32]. Despite these advantages titanium bars are sometimes considered less practical as they must be bent into shape with a computer-assisted manufacturing technique to fit the patient preoperatively and are more expensive to manufacture [6].

In their 2010 report Nuss and his group [2] noted that of the 35 patients diagnosed with nickel allergy, 22 (2%) were diagnosed preoperatively received titanium bars. Of the 13 patients who received stainless steel bars and developed allergic reactions, ten were treated successfully with low dose prednisone until erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) returned to normal. Three required bar removal. Of these, one required no further treatment and two received replacement titanium bars.

It is interesting to speculate on the infrequency of cutaneous reactions among pectus bar implant recipients given the prevalence of nickel allergy. The vast majority of patients who receive pectus implants have satisfactory correction of the chest wall contours with few complications [3–5, 8–10, 21]. We suspect the discrepancy may be partly attributed to duration—since the bars are only in place for 2–3 years it is possible that implants are not in place long enough to elicit a response in a previously sensitized patient. Alternatively, some patients may develop mild reactions along the incision sites which may be simply overlooked or erroneously attributed to poor wound healing. Further prospective studies in device patients with putatitve metal allergy are warranted.

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Instruments for Correction of Chest Wall Deformities

26

Silvia Zötsch and Amulya K. Saxena

Introduction

The era of correction of chest wall deformities (CWDs) commenced a century ago with the first correction performed by Wilhelm Meyer in 1911 (see Chap. 1). Although the corrections in the initial five decades showed the emergence of techniques that involved various forms of cartilage resection, Matti Sulama from Finland in 1956 was the first to introduce the concept of using metal struts for the correction of CWDs. Since the advent of osteosythetic materials for reconstruction of CWDs in the mid 1950s, many techniques were developed using various forms of metal struts and fixation without any method gaining acceptance and popularity worldwide. This could also be attributed to the fact that most of the methods until the late 1990s involved open surgical approaches.

With the introduction of the minimal access surgical approach in 1998, this method gained popularity in the treatment of pectus excavtum deformities, with a global acceptance of this method. The instruments for performing the minimal access repair of pectus excvatum have thus been standardized with a few modifications that differentiate the products between the major suppliers. On the other hand, the large series of repairs of CWDs using the open Willital's technique and its refinement to the less invasive PLIER technique, (Pectus Less Invasive Extrapleural Repair) which is well suited for severely asymmetric forms of pectus deformities, combined forms of pectus excavtum-carinatum type of deformities, have utilized a standardized metal strut in over 2500 patients. This chapter will hence cover the instruments for minimal access repair of pectus deformities, the metal struts used for the Willital and PLIER technique as well as two other instrument systems for the stabilization of chest wall- the *Strasbourg Thoracic Osteosyntheses System* and titanium implants for ribs repairs.

Instruments for Minimal Access Repair of Pectus Deformities

The sets for minimal access repair of pectus deformities (pectus excavatum and carinatum) comprise of a series of special instruments. Two sets with varying accessories options presented in this section are the Pectus Security Implant and the Biomet Microfixation Pectus Bar.

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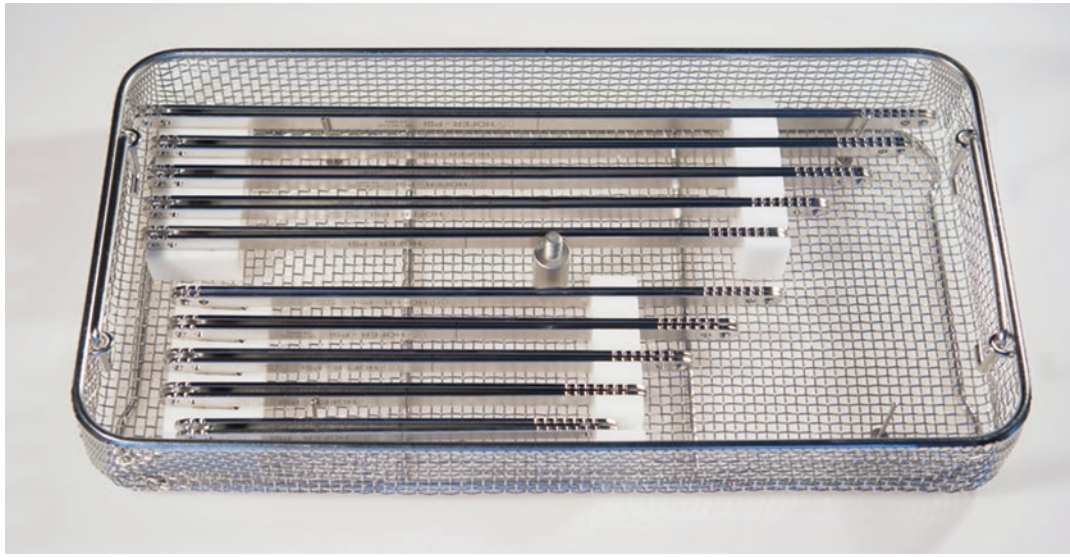

Fig. 26.1 View of a pectus bar tray consisting of pectus bars of various sizes (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

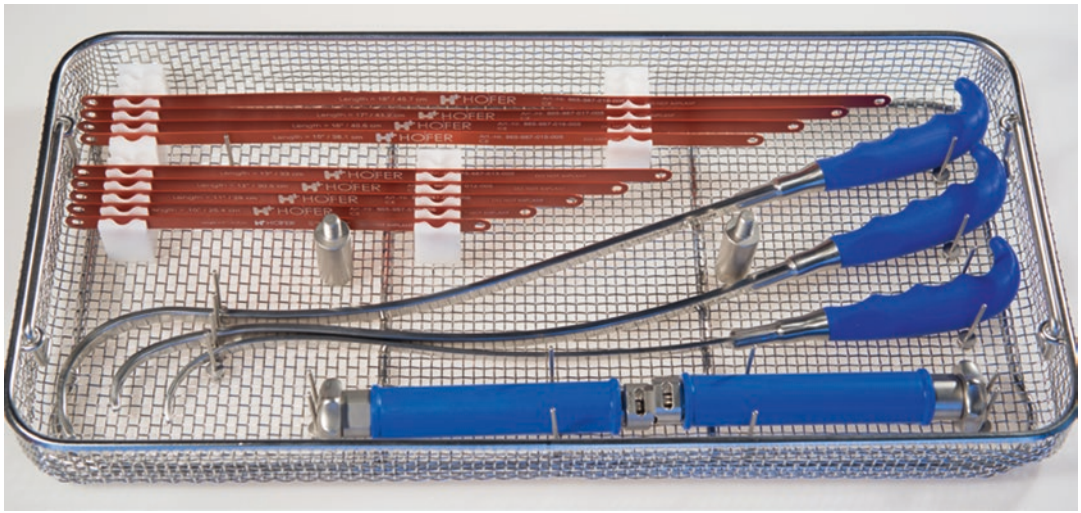

Fig. 26.2 View of the pectus instrument tray which comprises of pectus flexible ruler, pectus introducers, pectus stabilizers, pectus bar flipper and pectus benders (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Pectus Security Implant (Hofer Medical, Fürstenfeld, Austria)

(a) The Pectus Security Implant tray consists of an assortment of pectus bars of various sizes ranging from 22.8 to 45.7 cm (9–18 in.) in length. Pectus bars are not individually ordered, but must be part of the set since measurements are done on the patient under sterile conditions after which the appropriate pectus bar size is chosen for repair (Fig. 26.1). However, additional large size bars 48.26–55.88 cm (19–22 in.) which are not part of the standard set can be ordered.

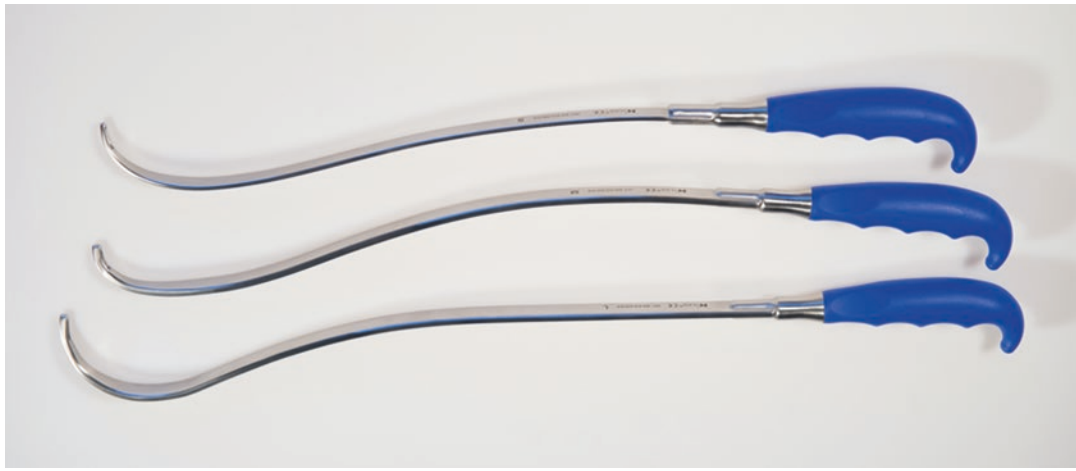

Fig. 26.3 View of the choice of pectus introducers with variation in the depth of the curved tips (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.4 Orthopedic plate bender press to bend the pectus bar (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

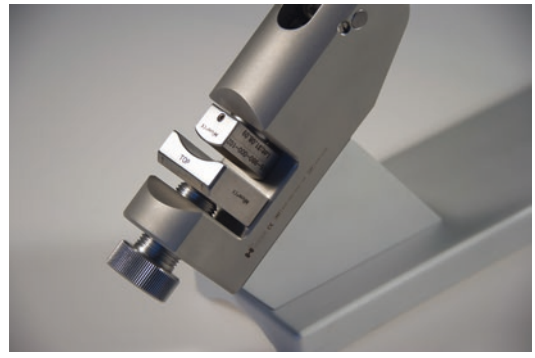

Fig. 26.5 Closer view of the orthopedic plate blender press shows the adjustment screw to set the depth of bends required on the pectus bar (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

(b) The pectus instrument tray comprises of a pectus template, pectus introducers, pectus stabilizers and pectus bar flipper (Fig. 26.2).
(c) Pectus introducers are available in three sizes. The choice of the introducer depends on the severity of the depression in pectus excavatum (Fig. 26.3).
(d) Pectus table top plate bender press is used to curve the pectus bar to fit the contours of the corrected chest wall (Figs. 26.4 and 26.5).
(e) Pectus templates are present in the set which are placed on the chest of the patient to measure the chest size and are shaped to the contours of the expected corrected result (Fig. 26.6).
(f) Pectus bars are 3 mm thick stainless steel bars which have rounded ends, blunt edges, notched end sides and perforations along with an integrated stabilizer plate (Figs. 26.7, 26.8, and 26.9).
(g) Pectus bar flippers are used to rotate the pectus bar once it is positioned in the chest. Bar flipper have a screw to secure the pectus bar to aid n controlled rotation of the pectus

Fig. 26.6 Pectus template is placed on the chest of the patient to measure the chest size and is shaped to the contours of the expected corrected result (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.7 Pectus bars used for minimal access repairs are stainless steel bars which have rounded ends, blunt edges, notched end sides and perforations (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

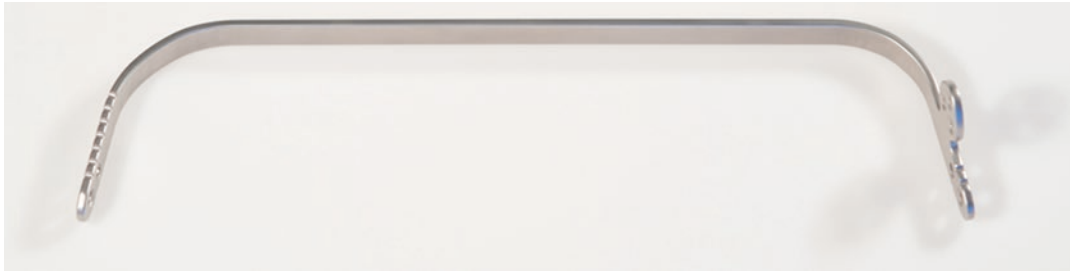

Fig. 26.8 Shape of the pectus bar after bending into shape for implantation (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.9 Some manufacturers supply pectus bars with integrated a stabilizer plate (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.10 Pectus bar flippers are used to rotate the pectus bar once it is positioned in the chest (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.11 A bar flipper could be used on one side or on both sides (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

bar (Fig. 26.10). A bar flipper could be used on one side or on both sides (Fig. 26.11).

(h) Bar stabilizer plates are not parts of the standard instrument tray, since the stabilizer is integrated on one end of the pectus bar. However, if stabilization on both sides is required, a separate stabilizer plate can be used (Fig. 26.12).

Biomet Microfixation Pectus Bar (Biomet Microfixation, Jacksonville, FL, USA) Formerly Known as The Lorenz Pectus System (W. Lorenz Surgical Inc., Jacksonville, FL, USA) and Biotech Pectus Correcting System (Biotech GmbH, Wiesbaden, Germany)

Instrument tray for the Biomet Microfixation Pectus Bar/ Biotech Pectus Correcting System consists is similar to the one described above with regards to the Pectus table top bender and pectus templates assortment of pectus bars, however other instruments which vary in design and method of application are listed below.

(a) Pectus introducers are also available in three sizes (small – 18.8 in., large- 20 in. and X-large-22.4 in.). The choice of the introducer used in repairs depends on the severity of the depression.

(b) Pectus bars are stainless steel bars which have rounded ends, blunt edges, serrated end sides and perforations along the ends of the plate (Fig. 26.13). The Biomet pectus bars come in sizes that range from 7 to 17 in. (17.8–43.2 cm) whereas the Biotech bars range from 20.5 to 45.5 cm. For patients with allergy to Nickel which is in the pectus bars,

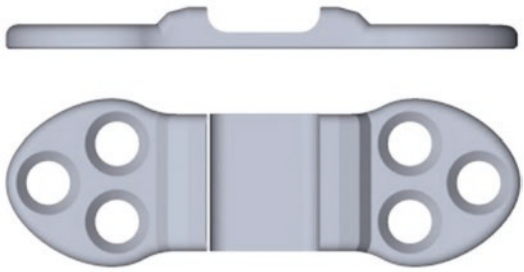

Fig. 26.12 Pectus bar stabilizer plate with three perforations each in both the flanges (Courtesy: Hofer GmbH, Fürstenfeld, Austria)

Fig. 26.14 Stabilizer plate with perforated edges (Courtesy: Biotech GmbH, Wiesbaden, Germany)

Fig. 26.13 Pectus bar is a stainless steel bar with perforations on the end. The pectus bar edges are rounded and smooth (Courtesy: Biotech GmbH, Wiesbaden, Germany)

Biomet Microfixation offers titanium bars for minimal access repair.

(c) Stabilizers plates are used to secure the pectus bar to the chest since these pectus bar do not have an integrated stabilizer plate (Fig. 26.14). In addition to the sliding plates offered by both manufacturers, Biotech offers stabilizer plates with screw fixation to offer better grip on the pectus bar (Figs. 26.15a, b). Stabilizer plates can be used on one side or both sides depending on the surgical protocol (Figs. 26.15c).

(d) Pectus bar bender is a hand held tool for bending the pectus bar offered by both manufacturers (Fig. 26.16). Unlike the Pectus table top bender press, this instrument can used to modify pectus bar contours when the pectus bar is placed in the patient before it is flipped into its final position.

(e) Pectus bar flipper is tool offered by both manufacturers. This tool comes with a handle and a slit on the top through which the end of the pectus bar can be slid (Fig. 26.17). The bar flipper tool can be used to flip from one end or a set of flippers may be used on both ends of the pectus bars.

Optical Devices and Video-Assisted Surgery Instruments

1. **Ports**: Ports with blunt tip trocars are preferred to gain access into the thoracic cavity (Fig. 26.18). Although, scopes can be directly inserted into the thoracic cavity without the use of a trocar, this is not advisable as smudging of the lens and loss of vision can occur which can jeopardize the safety of the procedure.

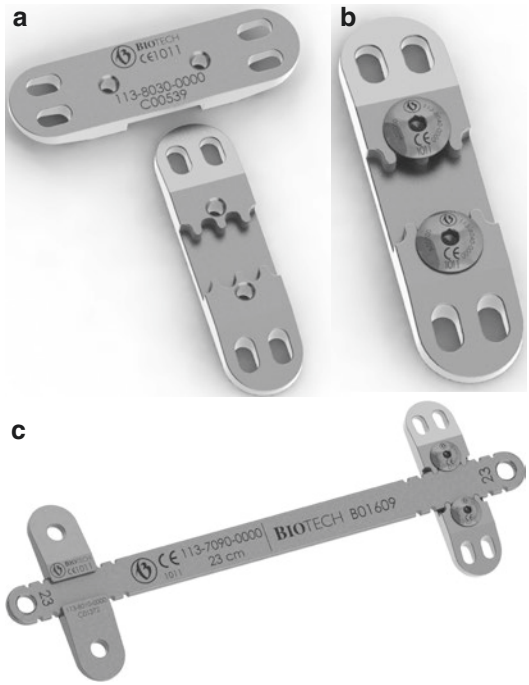

Fig. 26.15 Stabilizer plates can also be obtained with a screw-based securing system. (**a**) Stabilizer plate with screw-based system (*top- rear view* and *bottom-front view*). (**b**) Placement of the screw in the stabilizer plate. (**c**) Pectus bar with 2 stabilizer plates (*left- sliding stabilizer plate* and *right- screw-based stabilizer plate*) (Courtesy: Biotech GmbH, Wiesbaden, Germany)

Fig. 26.16 Pectus bar bender is a held tool with an adjustable wheel fitted below the jaws to control the degree of bar bending (Courtesy: Biotech GmbH, Wiesbaden, Germany)

Fig. 26.17 Pectus bar flipper with a slit in the top end of the tool to engage the pectus bar end and aid in its controlled rotation (Courtesy: Biotech GmbH, Wiesbaden, Germany)

2. **Scopes**: Scopes of 5 mm with 30° angle of vision are generally employed for the minimal access repair (Fig. 26.19). Turing the scopes within the ports allow better visualization of the retrosternal space when the introducer is used to create the precardial space. These scopes also give good vision when rib suture are placed which are used to secure the stabilizer.
3. **Operating scopes**: In case of prior surgery or adhesions, an operating scope has been found to be beneficial (Fig. 26.20). The operating scope offers a channel for the passage of a variety of instruments to perform tissue manipulation using the single access route (Fig. 26.21). The operation scope has a 0° angle of vision and is not recommended for use as a standard scope in minimal access repair of pectus deformities.

Fig. 26.18 Blunt trocars with 5 mm ports and an insufflation channel are preferred for gaining access into the thoracic cavity (Courtesy: Ethicon GmbH, Nordestedt, Germany)

Instruments for PLIER Repairs

The set for PLIER technique comprise of the following instruments (Firma Karl Lettenbauer, Erlangen, Germany):

(a) The chest wall correction tray consists of flat metal pectus struts of various sizes ranging from 10 to 30 cm (Table 26.1). The pectus struts of desired sizes can be ordered individually, with most of the pectus repairs of adolescents and adult requiring sizes between Nr. 6 and 9 (Fig. 26.22).
(b) Pectus struts are flat metal plates with perforations along the entire length of the strut.

Fig. 26.19 Optimal vision for minimal access repairs of pectus deformities is obtained by 5 mm scopes with a 30° angle of vision (Courtesy: Richard Wolf GmbH, Tuttlingen, Germany)

Fig. 26.20 Mini-operating thoracoscope of 5.5 mm with a 3.5 mm working channel. The scope offers an image of 50,000 pixels (Courtesy: Richard Wolf GmbH, Tuttlingen, Germany)

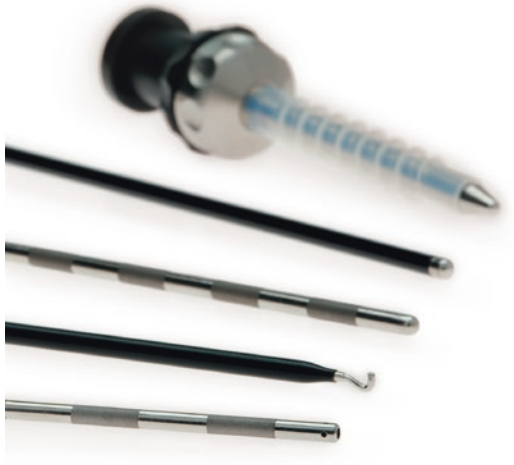

Fig. 26.21 Assortment of 3.5 mm instruments (flexible 5.5 mm port sleeve, coagulation button electrode, blunt probes, monopolar hook electrode, suction tube) that can be used with the mini-operating thoracoscope (Courtesy: Richard Wolf GmbH, Tuttlingen, Germany)

Table 26.1 Dimensions of metal struts sizes and dimensions for PLIER technique

Strut sizes	Dimensions (L×T×W)
Nr. 1	100×1.5×5.0 mm
Nr. 1a	115×1.5×5.5 mm
Nr. 2	130×1.5×6.5 mm
Nr. 2a	140×1.5×7.0 mm
Nr. 3	150×1.5×7.5 mm
Nr. 3a	150×1.0×7.5 mm
Nr. 4	175×2.0×8.5 mm
Nr. 4a	175×1.0×8.5 mm
Nr. 5	200×2.0×8.5 mm
Nr. 6	225×2.0×8.5 mm
Nr. 7	250×2.0×8.5 mm
Nr. 8	275×2.0×8.5 mm
Nr. 9	300×2.0×8.5 mm

Source: Firma Karl Lettenbauer, Erlangen, Germany

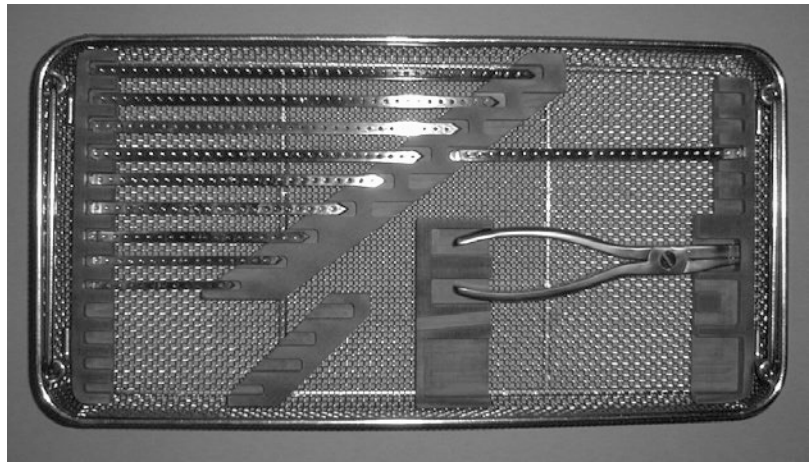

Fig. 26.22 The chest wall correction tray consists of an assortment of flat metal pectus struts of various sizes ranging from 10 to 30 cm (Table 26.1) (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

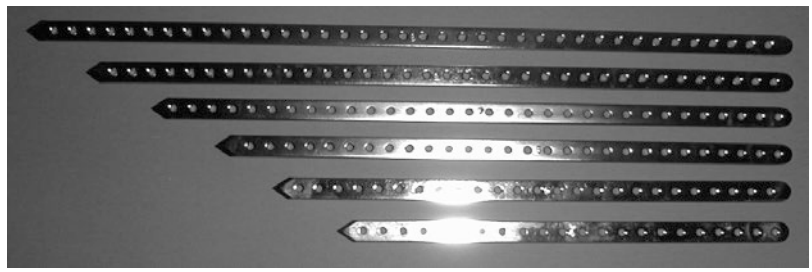

Fig. 26.23 One end of the strut is blunt and rounded, while the other end is shaped in the form a spearhead (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

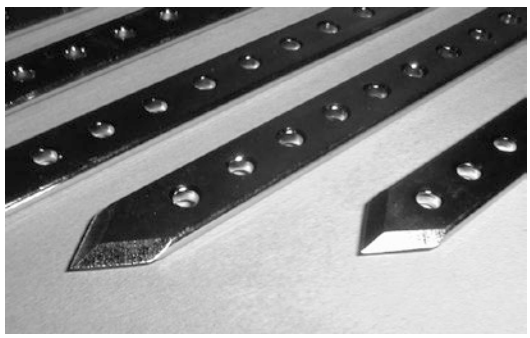

Fig. 26.24 The sharp spearhead end of the pectus strut enables easier passage of the strut through the sternum (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

One end of the strut is blunt and rounded, while the other end is shaped in the form a spearhead (Fig. 26.23). The sharp spearhead end of the pectus strut enables easier passage of the strut through the sternum (Fig. 26.24).

(c) Pectus strut cutter is an instrument used to the spearhead end of the pectus strut after it is passed through the sternum (Figs. 26.25 and 26.26). If additional struts are used that are not passed through the sternum, the spearhead end of these struts are cut using the pectus strut cutter prior to implantation.

(d) Pectus two-tooth plier is a special instrument to secure and control the pectus struts as it is passed through the sternum (Fig. 26.27). The plier has two teeth that safely lock the pectus strut to avoid slippage while it is manipulated.

(e) Pectus spanners are used as a pair. The pectus spanners are used to bend the pectus strut to the contours required to remodel the chest wall (Fig. 26.28).

Instruments for the Strasbourg Thoracic Osteosyntheses System

Strasbourg Thoracic Osteosyntheses System or STRATOS™ (MedXpert GmbH, Heitersheim, Germany) are implants which are made from pure titanium. The basic form of the implants of

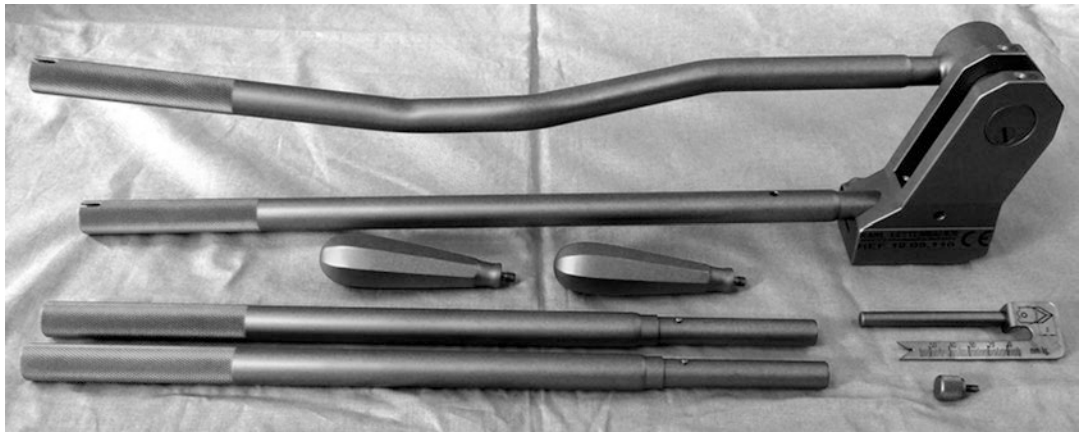

Fig. 26.25 View of pectus strut cutter used to the spearhead end of the pectus strut after it is passed through the sternum (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

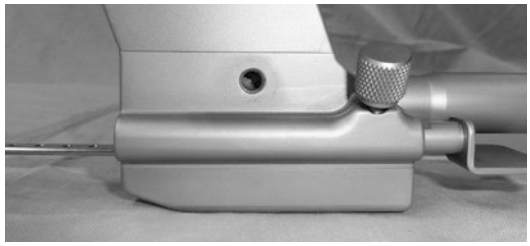

Fig. 26.26 Magnified view of the cutting tip of the pectus strut cutter (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

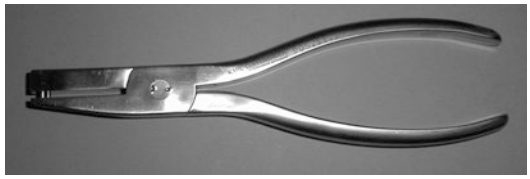

Fig. 26.27 Pectus two-tooth plier is employed to secure and control the pectus struts as it is passed through the sternum (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

the STRATOS™ system is computer calculated and permits three-dimensional shaping for optimal adaptation to the ribs. One complete implant consists of two rib clips (straight, 22.5° or 45° types) and one connecting bars (one of three lengths) (Fig. 26.29).

Fig. 26.28 Pectus spanners are used to bend the pectus strut to the desired contours (Courtesy: Firma Karl Lettenbauer, Erlangen, Germany)

(a) For easier identification all implants are color-coded; the blue implants are specifically designed for the correction of chest wall and sternal deformities, whereas the golden implants are designed for trauma surgery. Both types of connecting bars are scaled which allows the surgeon to determine the suitable bars as required in the anatomical situation. In addition, all bars can be cut to size (Fig. 26.30).
(b) The three different angulations of the rib clips can be adapted to any anatomical situation by means of a bending instruments and

Fig. 26.29 The assortment of ribs clips and connecting bars that comprise the STRATOS™ system (Courtesy: MedXpert GmbH, Heitersheim, Germany)

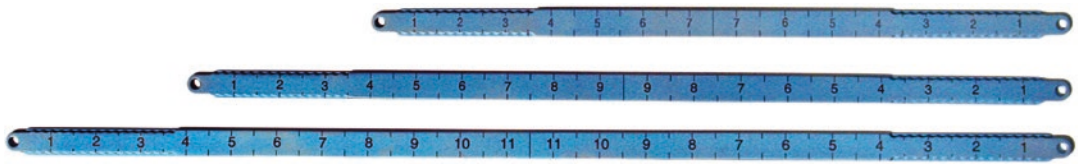

Fig. 26.30 Connecting bars of the STRATOS™ system are scaled which allows the surgeon to determine the suitable bars as required in the anatomical situation (Courtesy: MedXpert GmbH, Heitersheim, Germany)

Fig. 26.31 STRATOS™ blue implants are specifically designed for the correction of chest wall and sternal deformities (Courtesy: MedXpert GmbH, Heitersheim, Germany)

Fig. 26.33 Rib clips can be used for single rib correction STRATOS™ system (Courtesy: MedXpert GmbH, Heitersheim, Germany)

Fig. 26.32 STRATOS™ golden implants are designed for trauma surgery (Courtesy: MedXpert GmbH, Heitersheim, Germany)

cover a maximum range of −12° up to +57° (Figs. 26.31 and 26.32).

(c) Rib clips can be used in case a single rib correction is to be required (Fig. 26.33).

STRATOS™ can be used for a variety of corrective procedures that involve the ribs or the chest wall (Figs. 26.34, 26.35, and 26.36)

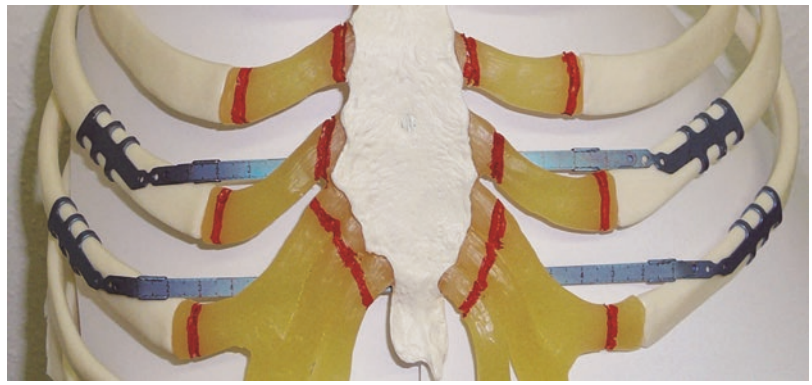

Fig. 26.34 View of a chest model showing the application of STRATOS™ for correction of chest wall deformities (Courtesy: MedXpert GmbH, Heitersheim, Germany)

Fig. 26.35 View of a rib model showing the application of STRATOS™ in rib trauma (Courtesy: MedXpert GmbH, Heitersheim, Germany)

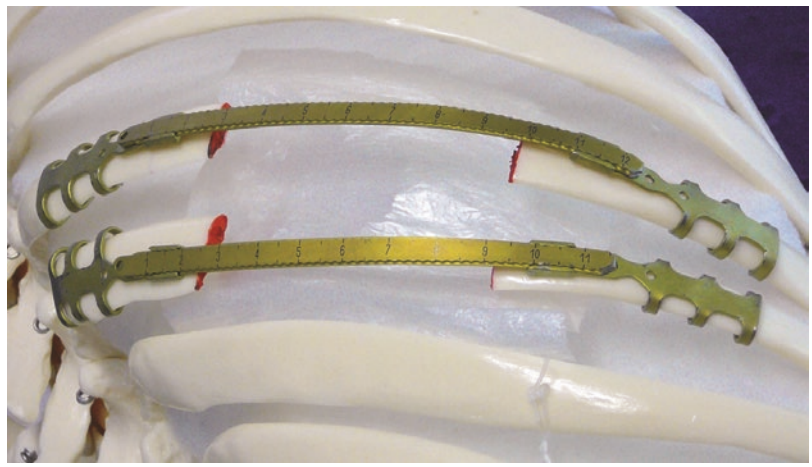

Fig. 26.36 View of a rib model showing the application of STRATOS™ for bridging rib defects after resection of tumors of the chest wall (Courtesy: MedXpert GmbH, Heitersheim, Germany)

Instruments for Rib Fracture Plating System

The Rib Fracture Plating System RibLock® (Acute Innovations®, Hillsboro, OR, USA) is a comprehensive system of implants and instruments specifically for repairing rib fractures. The plate's unique U-shape with locking screw technology provides excellent fixation. The plates in the system range from 46 to 76 mm in length. The 46 mm plate requires only four screws for fixation. The 61 cm and 76 cm plates are useful for repairing complicated frac-

ture patterns. The advantage of this system is that it allows for preservation of the neuromuscular bundles.

The plates are available in four sizes to match the anterior/posterior thickness of the rib. Color coding of the plates, screws and instrumentation ensures that the correct length of screw is used for the rib. The targeting guides aid in installing the plates in a straightforward, precise and repeatable manner. The plate's U-shape and locking screws allows fixation to be independent of bone quality and/or screw purchase in the bone.

Further Information on Instrument Manufacturers

1. Biomet Microfixation, Jacksonville, FL, USA *www.biomet.com*
2. Pectus Security Implant, Hofer GmbH, Fürstenfeld, Austria *www.hofer-medical.com*
3. Firma Karl Lettenbauer, Erlangen, Germany *www.karl-lettenbauer.de*
4. MedXpert GmbH, Heitersheim, Germany *www.medxpert.de*
5. Acute Innovations®, Hillsboro, OR, USA *www.acuteinnovations.com*

Part III

Pectus Excavatum: General Aspects- Pectus Excavtum

Overview of Repair of Pectus Excavatum Type of Deformities

27

Amulya K. Saxena

Introduction

Historically, surgical repair of pectus excavatum type of deformities was performed on patients who exhibited several sternal depressions, as the surgical procedure was associated with a high risk to justify it in patients with mild or moderate forms [1]. Unnecessary radical procedures to repair pectus excavatum were not recommended and even partial or non-esthetic repairs were proposed to be considered as acceptable outcomes [2]. Also, the primary intention of these surgical procedures was to address the symptoms of the patients with the aim of relieving cardiac compression, with cosmesis not being considered as the main aim of such repairs. With the changes in contemporary fashion trends and increased participation in sport activities, both of which are associated with larger exposure of the body, pectus excavatum repairs regained limelight in the past two decades. Patients with pectus excavatum type of deformities perceived their bodies as a possible source of embarrassment which further manifested as socio-psychological problem of varying degrees in these patients. With the increase in self-referrals in the absence of symptoms to general practitioners during puberty and early adulthood, the trend of further referring these patients to Centers who could evaluate these patients as well as offer surgical corrections was established.

The timing of the surgical intervention is an area where the surgical community is divided and the opinions are divergent. Surgical procedures are best performed in early adolescence when the thoracic skeleton is more pliable and accommodating [3]. This view has also been shared by other groups that have large experiences with correction of pectus deformities [4]. The concerns of developing acquired thoracic dystrophy that has been reported after repairs in early years needs to be addressed [5]. In large series reported with open pectus repairs on young adolescents, the acquired thoracic dystrophy complication has not been observed [4, 6, 7]. The main reasons for the absence of this complication in these series was that surgical procedures were performed in Centers of expertise and in the hands of experienced surgeons and the common denominator in these techniques being the preservation of part of the perichondrium and the growth centers at the ends of the cartilages. Another important aspect that also favors correction in early adolescence is the observation of the fact that in half of the patients with pectus excavatum and mitral valve prolapse, the mitral dysfunction disappeared after pectus excavatum repairs [8].

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Overview of Procedures

Prior to the advent of thoracic surgery, correction of pectus excavatum deformity was performed using by *external traction*, a method that was reintroduced in the 1920s by Zahradnicek [9]. The technique involved using traction on two wires introduced through operative perforations of the sternum.

The first surgical repair of pectus excavatum was performed by Wilhelm Meyer in 1911 who resected short segments of the right 2nd and 3rd costal cartilages with the intention of relieving cardiac compression [10] (Fig. 27.1). Meyer reported that the patient, who was significantly dyspneic before this intervention, improved significantly afterwards. Sauerbruch found this procedure to be of limited use and hypothesized that this limited resection could not be expected to relieve cardiac compression. In 1913, he performed a more radical procedure by resecting segments from the 5th down to the 9th costal cartilage, as well as the corresponding left hemisternum [11] (Fig. 27.2). Sauerbruch also advocated a more radical approach with bilateral resection of the 5th to 9th costal cartilage along with transverse resection of the sternum (Fig. 27.3).

Lexer in 1927 reported the treatment of a patient with severe pectus excavatum by resecting the sternum and replacing it in a turned-over position. Due to the resulting gap that occurred between the flipped sternum and the cartilages which caused difficulty in reattachment, the depressed sternal segment was removed [12] (Fig. 27.4).

Alexander in 1929 presented two different techniques on the two patients that were operated with trauma-induced pectus excavatum [13]. In the first patient, the sternum was divided transversely at the level of the third rib, then axially down to the xiphoid process (Fig. 27.5). In the second patient, the entire sternum was removed except the manubrium and the associated costal cartilages, leaving the heart exposed under the skin cover (Fig. 27.6). Alexander further used external traction after open repair and maintained it for various time intervals with the intention of achieving sufficient tissue fixation.

The next procedures advocated for repair involved the dissection as well as reattachment of the sternum. Nissen in 1943 resected the deformed portion of the sternum, after which it was flipped over and reattached horizontally to the costal cartilages [14] (Fig. 27.7). Judet and Judet in 1954 performed a complete sternal turnover procedure by dissecting the sternum and reattaching the resected sternochondral margin into the consequent gap in the anterior chest wall

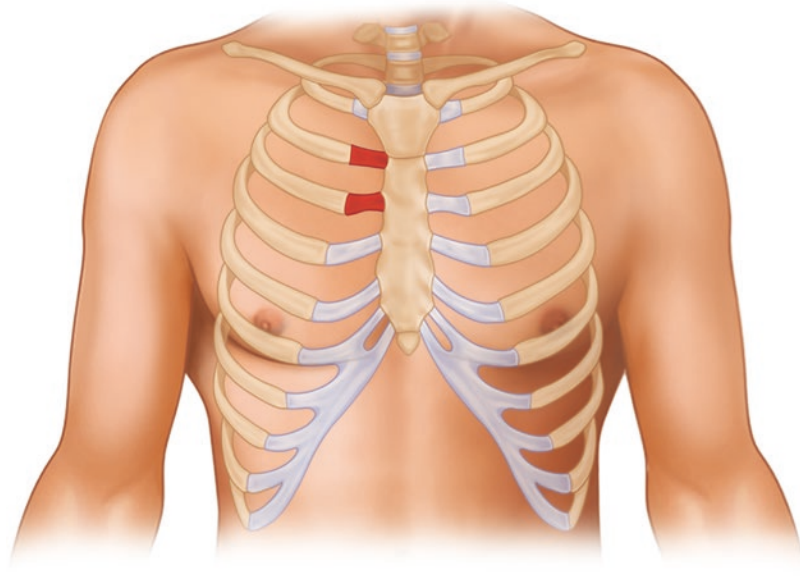

Fig. 27.1 Schematic view of pectus excavtum repair as performed by Meyer

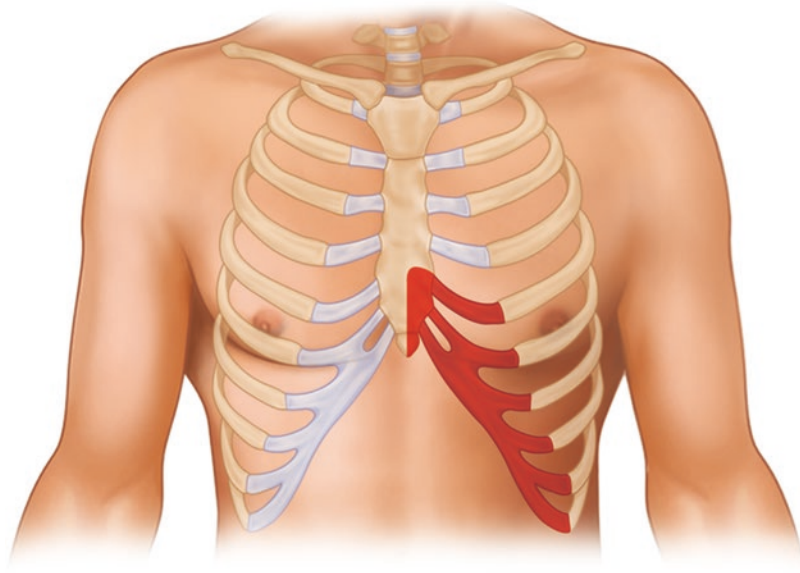

Fig. 27.2 Schematic view of pectus excavtum repair as performed by Sauerbruch

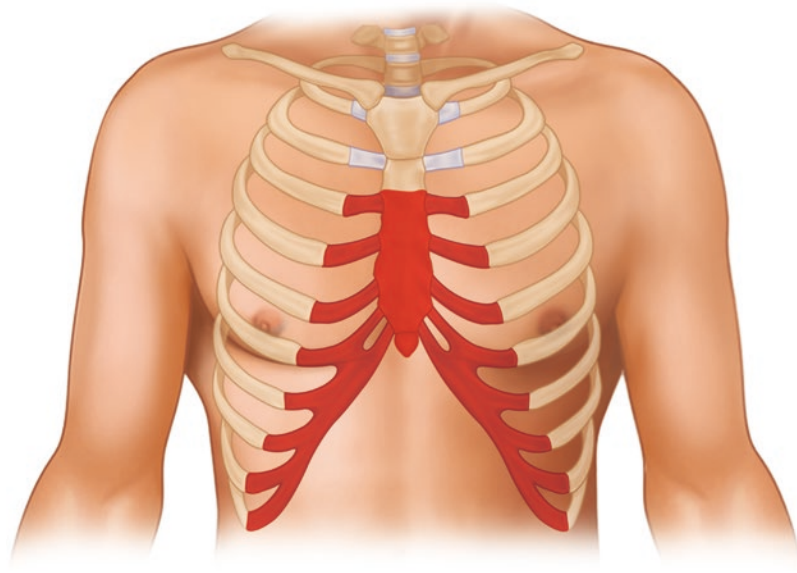

Fig. 27.3 Schematic view of pectus excavtum repair as performed by a further radical approach by Sauerbruch

[15] (Fig. 27.8). In these turn-over procedures the depressed portion of the sternum was explanted together with the deformed cartilages, turned over and reinserted into the resulting gap in the anterior chest wall. Any unwanted protuberance of the sternum was flattened using a hammer. This method was used extensively only in Japan by Wada, the shortcoming of this method being sternal necrosis and fistula formation [16, 17]. These complications prompted Taguchi to preserve the internal thoracic vessels. Ninkovic in 2003 revived the procedure by performing vascular anastomosis of the internal thoracic vessels (i.e. anastomosis of the the right internal thoracic vessels to the inverted left thoracic vessels) [18].

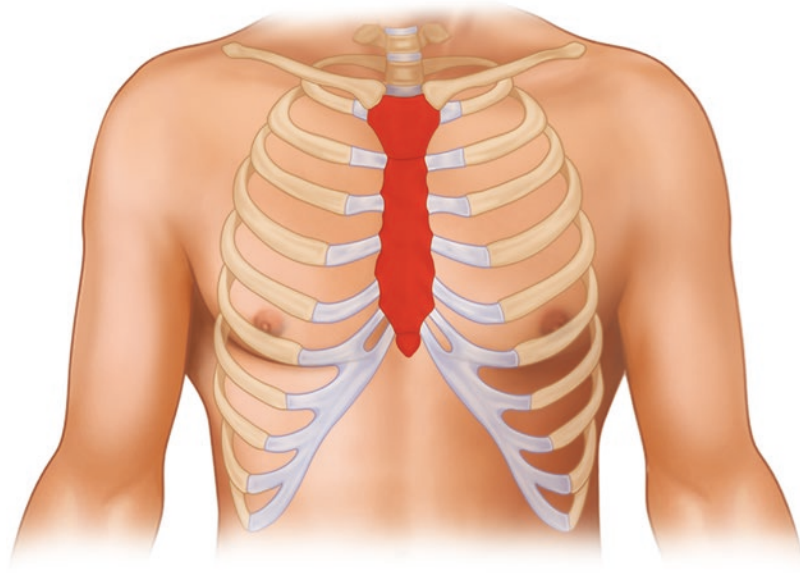

Fig. 27.4 Schematic view of pectus excavtum repair as performed by Lexer

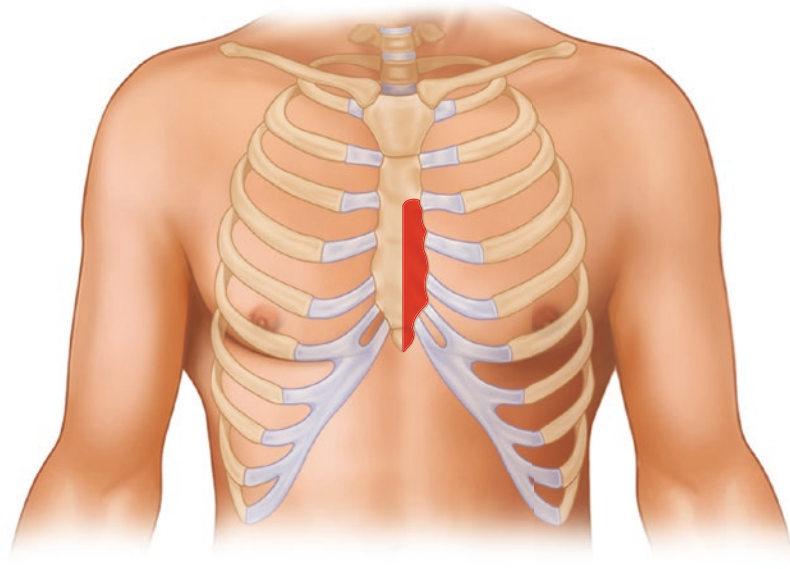

Fig. 27.5 Schematic view of pectus excavtum repair as performed by Alexander (Technique-1)

Lincoln Brown who proposed that pectus excavatum was caused by undue traction exerted upon the sternum by the diaphragmatico-sternal ligament, appropriately in 1940 designed two operative procedures in which all the tissue connection between the diaphragm and the lower sternum were divided [19]. The 1st procedure was indicated for infants or children. The basis of this procedure was to dissect the substernal ligament and/or the attachment to diaphragm under a small, vertical incision at level of the xiphoid in order to prevent the progression of pectus excavatum deformity in adulthood. The 2nd was indicated for older children, adolescents and adults (Fig. 27.9). The basis of this procedure was (a) resection of 2 cm costal cartilage segments of the 4th to 7th ribs near the junction with the sternum, and (b)

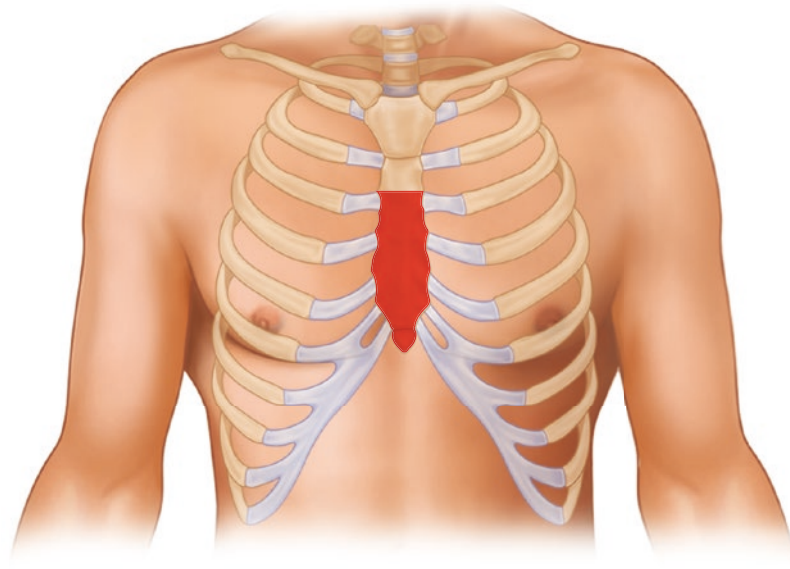

Fig. 27.6 Schematic view of pectus excavtum repair as performed by Alexander (Technique-2)

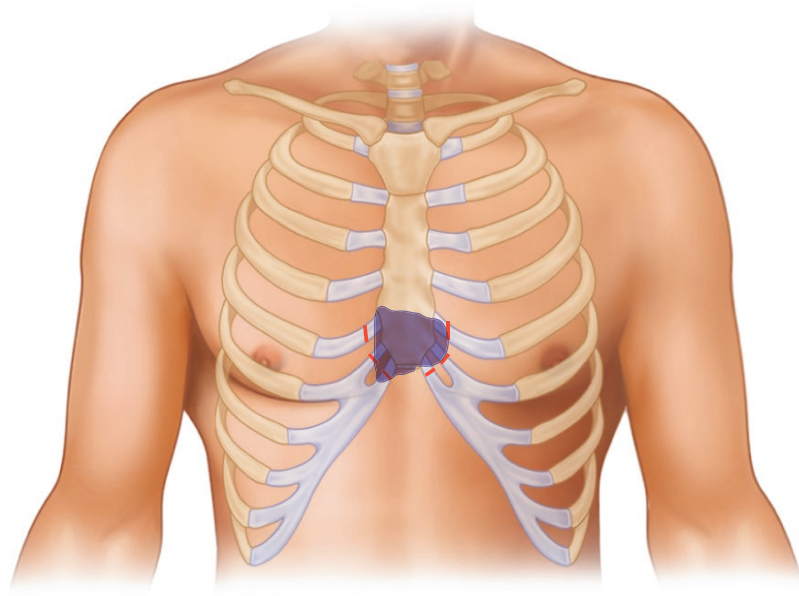

Fig. 27.7 Schematic view of pectus excavtum repair as performed by Nissen

wedge resection of the sternum at level of junction of the manubrium and sternal body. The elevated sternum was secured by wire sutures at the site of wedge resection. An external traction was added by fixation of a wire that penetrated the caudal part of the sternum and the 5th costal cartilage with a bridge placed on the anterior chest wall. The results of the operation were satisfactory only in young patients and this technique was unsuccessful in older individuals.

Sweet in 1943 and Brodkin in 1948 performed surgical repairs which was similar to the later but more popular Ravitch procedure [20, 21]. The Ravitch procedure consisted of (a) resection of deformed costal cartilages (b) dissection of the

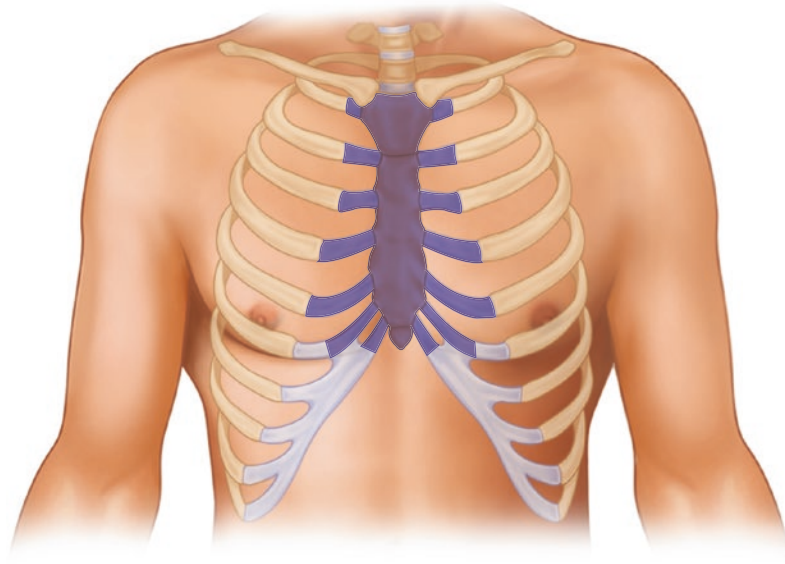

Fig. 27.8 Schematic view of pectus excavtum repair as performed by Judet

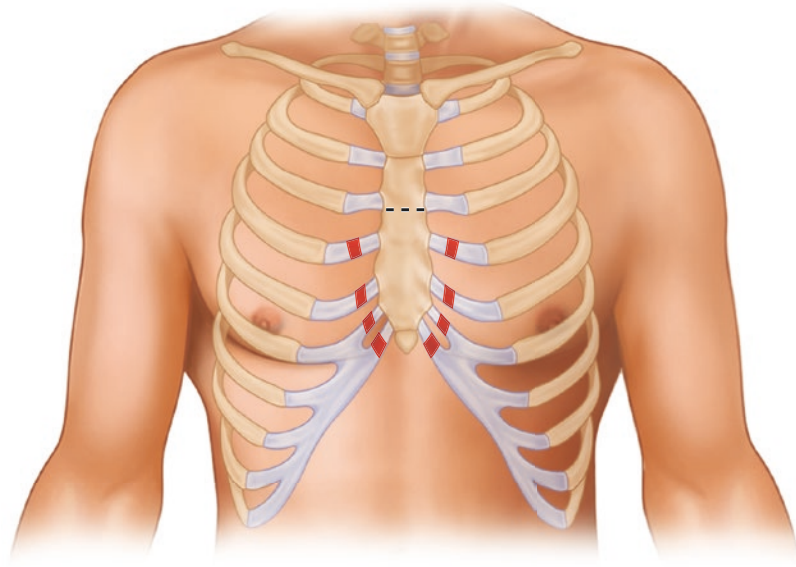

Fig. 27.9 Schematic view of pectus excavtum repair as performed by Brown

xiphoid-sternal junction, (c) cranial transverse sternal wedge osteostomy, and (d) refixation of the correctly positioned sternum [22] (Fig. 27.10). The wedge osteotomy was closed by interrupted silk sutures or wire sutures. However, the extensive defect resulting from excision of all of the deformed cartilages was left without sutures of any tissues. In 1965, Ravitch revised his original procedure in which he preffere an (a) oblique chondrotomy at 2nd or 3rd costal cartilages and overlap suture, sternal osteotomy on the posterior side of the sternum, instead of the anterior side as in the original procedure, and insertion of a small bone graft in the sternal opening. These modifications were done to offer better fixation to the mobilized sternum. A numerous modifications of the Ravitch procedure have been described by other surgeons and these modifications mainly

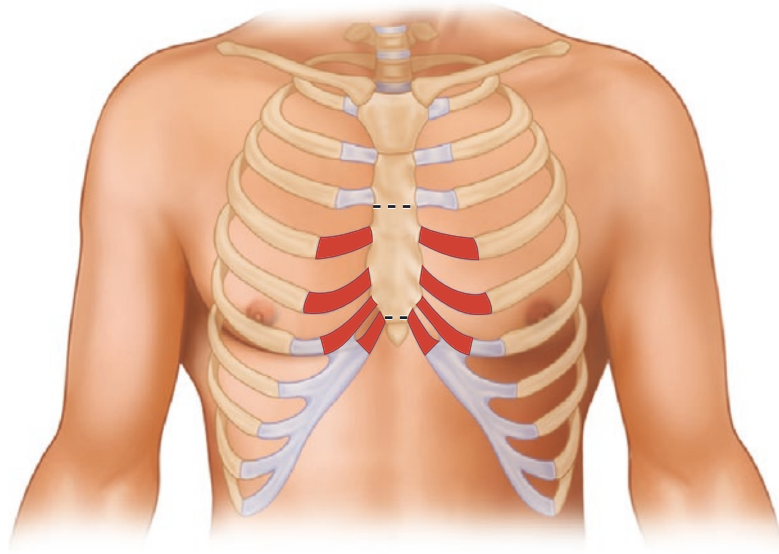

Fig. 27.10 Schematic view of pectus excavtum repair as performed by Ravitch

differ from each other on the techniques to fix the mobilized sternum.

Due to less favorable outcomes of the Ravitch procedure, several surgeons such as Alexander, Adams, Lester, and Dailey propagated the idea of using posterior rib struts for fixation of the newly positioned sternum [2, 13, 23, 24]. Adkins in 1958 was the first to use a methyl methacrylate substernal support for fixation of the sternum [25].

The following surgical techniques which were developed to fix the sternum without the use of support:

(a) Sicard in 1959 achieved the elevation and stabilization of the sternum by leaving the deformed cartilages in place and dividing them slanted towards the sternum [26]. These were then sutured together such that the divided ends overlap.
(b) Ravitch in 1961 propagated the placement of 2–3 nonabsorbable periostal sutures onto the edges of the sternal transverse osteotomy is applied to maintain sternal stability [27].
(c) Masson in 1970 utilized autogenous bone for concealment of the sternal depression [28].
(d) Ravitch in 1977 also proposed to keep the sternum elevated is by shortening the perichondrial strips after chondral resection using "ruffling" sutures [29].
(e) The Leonard procedure reported in 2003 which involves external traction applied for several months following open resection of the deformed cartilages and transverse wedge osteotomy using wires inserted through separate stab incisions has also been used to stabilize the sternum [30].

The following surgical techniques were developed to fix the sternum with the use of support using autogenous tissues:

(a) Robicsek in 1960 used the xiphoid process which is frequently dissected from the sternum during pectus repairs. He mobilized the xiphoid process and flipped it under the sternum to support the corrected position of the sternum [31] (Fig. 27.11). For this the xiphoid was partially freed of its attachment to the rectus fascia, pulled under the sternum and suspended with heavy wire sutures on the superior ribs or perichondrial strips. The long, vertical incisions essential for appropriate exposure left unsightly scars, hence were replaced by Baronofsky with the currently used vertical submammary incisions [32].
(b) Autogenous tibial grafts and ribs have also been used for retrosternal support [2, 13, 23,

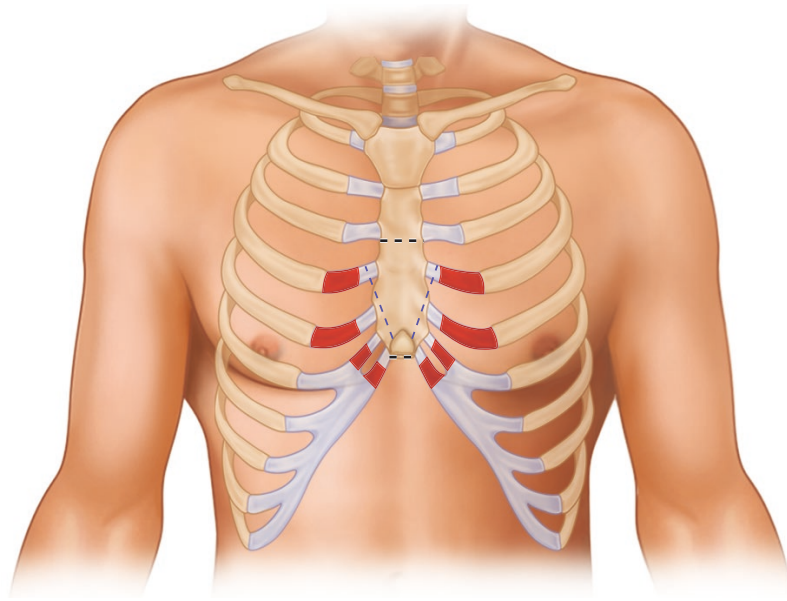

Fig. 27.11 Schematic view of pectus excavtum repair as performed by Robicsek

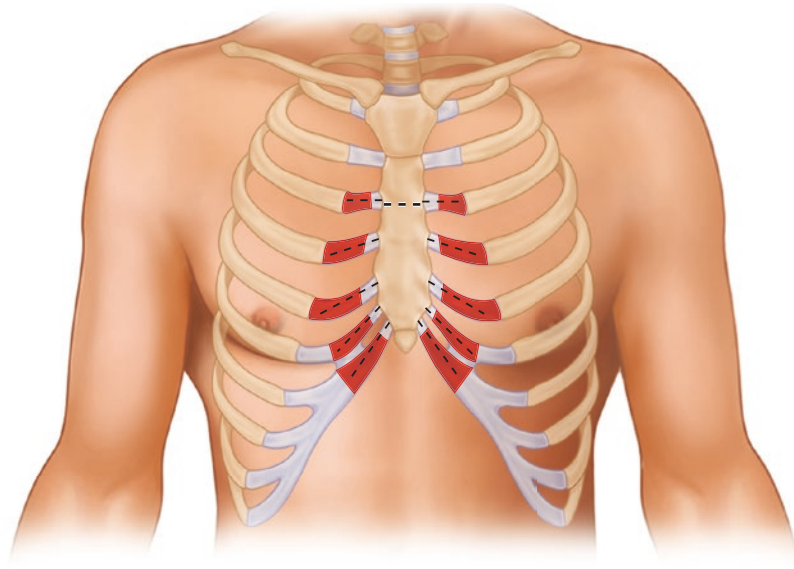

Fig. 27.12 Schematic view of pectus excavtum repair as performed by Welch

24]. The only drawback with this technique is that it may create a second scar at the place of procurement if the tibial graft is opted and prolong the operating time; however, it offers advantages to metal bars as it does not require a second intervention for removal of the bars.

(c) Suturing the perichondrial strips together behind the sternum in order to support the sternum should be avoided. This maneuver has not only been responsible for the decreases the thoracic diameter, but may retard chest growth and could lead to acquired thoracic dystrophy syndrome [33].

Welch in 1958 proposed a rather less radical procedure than Ravitch for the correction of pectus excavatum [34] (Fig. 27.12). Surgical cor-

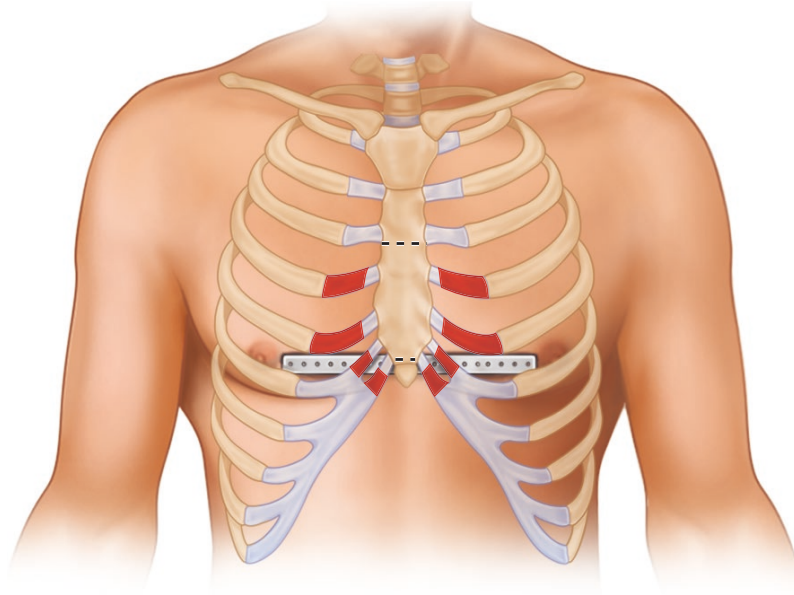

Fig. 27.13 Schematic view of pectus excavtum repair as performed by Haller

rection was performed using bilateral subperichondrial resection of the deformed costal cartilages and sternal osteotomy resecting a wedge of the anterior cortex along with the fracturing of the posterior cortex. In the early cases using this technique, the sternum was secured by intramedullary fixation with a Steinman pin. Later on, the procedure was modified and the anteriorly displaced sternum was maintained with silk sutures closing the osteotomy defect [35].

Metals in the form of Kirschner wires or metal struts or plates were utilized for the correction of pectus deformities and an assortment of these materials is available for repairs [25]. Plates have been used for anterior, trans-sternal and posterior sternal supports [1, 36, 37].

Haller adopted the Ravitch procedure but presented it with two modifications in 1989 [38] (Fig. 27.13). Haller realized that the sternum was not properly supported in the Ravitch procedure for which he advocated the placement of a temporary stainless steel strut beneath the sternum, and anchored the strut bilaterally to the 5th or 6th ribs. The strut was removed on an outpatient basis, 6–9 months after the primary repair. Haller also noticed that too extensive and surgical repair in the early years could induce constriction of the chest wall and the total resections of the deformed costal cartilages in Ravitch procedure resulted in the removal of growth centers of costal cartilages which he termed as "acquired Jeune's syndrome" [5]. His second modification was to shorten the length of the costal cartilages to 2.5 am and to exclude children below 4 years of age from surgery.

Fonkalsrud observed that in patients after the removal of deformed costal cartilages in the Ravitch's procedure, the regenerated cartilages are thin, irregular, and commonly rigid with calcification, even if the perichondrial sheaths are preserved. His approach was to resect short segments of costal cartilages (3–8 mm) on the medial and the lateral ends of the deformed cartilages [39] (Fig. 27.14). After this resection, the remaining costal cartilages were reattached to the sternum and the ribs. A transverse sternal wedge osteotomy was also performed and the sternum was fixed using a thin stainless steel Adkins strut which was placed posterior to both the sternum and the costal cartilages to elevate the sternum.

Rehbein in 1957 presented a procedure to correct pectus excavatum with rib splints [40] (Fig. 27.15). After completing the mobilization of the costal cartilages with minimal resection, narrow metal splints were pushed into the rib. The sternum and the cartilages were elevated, the

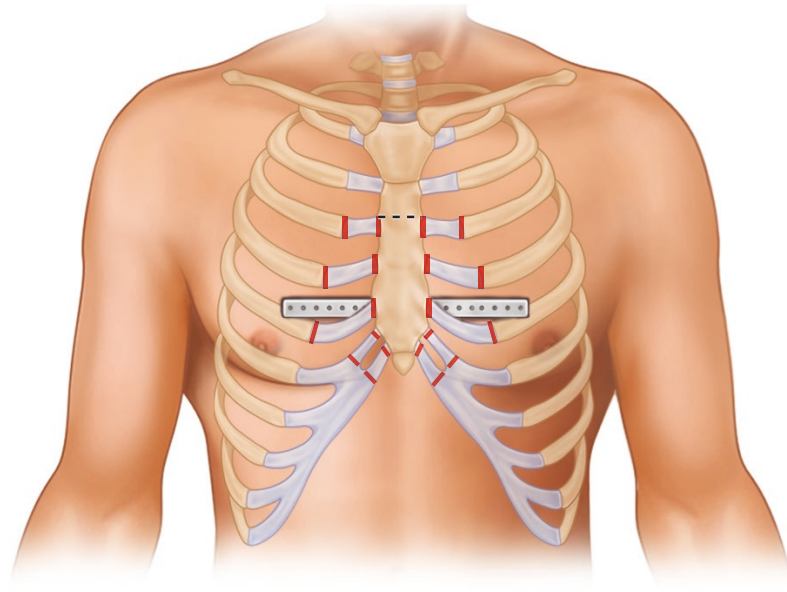

Fig. 27.14 Schematic view of pectus excavtum repair as performed by Fonkalsrud

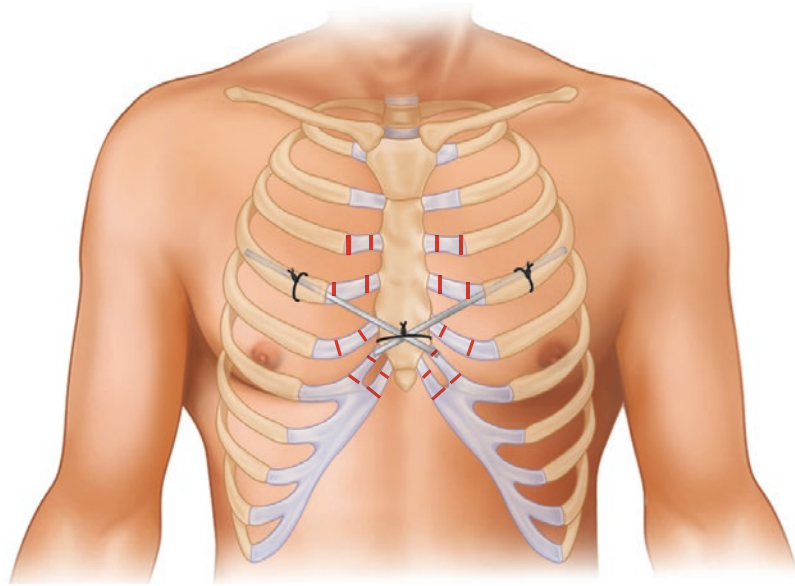

Fig. 27.15 Schematic view of pectus excavtum repair as performed by Rehbein

blades crossed and depressed. The sternum was fixed with a wire sling at the point of intersection. Costal cartilages which have been resected before to the necessary extent were secured by means of sutures. A stable fixation was achieved as the metal blades cross each other. In mild forms of pectus deformities one pair of rib splints were sufficient, however in moderate cases two pairs and in severe cases three pairs of ribs splints were necessary.

The Willital-Hegemann procedure has been used for the correction of pectus excavatum, pectus carinatum and other combined forms of protrusion-depression combined deformities since 1981 [37] (Fig. 27.16). In this procedure, repair is performed using a standard method of double bilateral chondrotomy parasternally and at points of transition to normal ribs. This is followed by a transverse sternal wedge osteostomy and detorsion of the sternum. The sternum edges

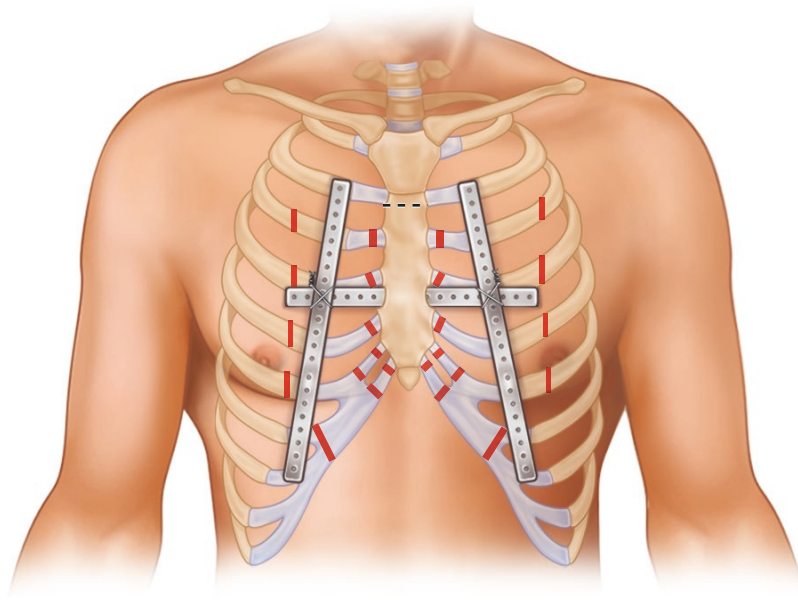

Fig. 27.16 Schematic view of pectus excavtum repair as performed by Willital-Hegemann

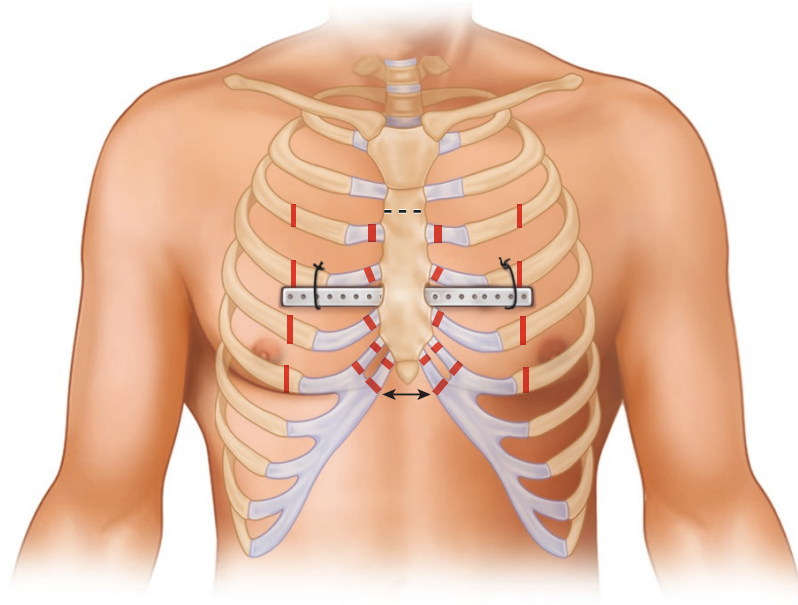

Fig. 27.17 Schematic view of pectus excavtum repair as performed by Saxena

after osteotomy are readapted by heavy absorbable sutures. Correction of the inverted ribs is performed using wedge osteotomy of ribs and readaptation using figure-of-8 sutures. Stabilization of the displaced sternum is performed using one trans-sternal strut. The two bilateral flail chest segments created are stabilized by and two parasternal metal struts. The metal struts are secured with each other by metal wires at the culmination points. Heavy absorbable sutures are used to secure the ribs to the struts at multiple points. Chest tubes are placed before completion of the procedure.

The PLIER procedure was reported in 2009 by the author and has been used for the correction of a variety of chest wall deformities [41] (Fig. 27.17). In this procedure, the repair is commenced with the dissection of the rectus muscle from the

xiphoid and the lowere rib attachments. After this a retro-sternal extra-pleural space is created. The repair is performed using a standard method of double bilateral chondrotomy parasternally of 1 cm in a caudal to cranial direction. A minimum length of 1 cm of cartilage is left on the sternum ends with the aim of maintaining an intact growth centre. A transverse sternal wedge osteostomy and the sternum is brought to the desired position. The sternum edges after osteotomy are readapted by heavy non-absorbable sutures. Correction of the inverted ribs is performed using wedge osteotomy of ribs. Stabilization of the displaced sternum is performed using one trans-sternal strut. The costal cartilages are readapted with absorbable suture. Heavy absorbable sutures are used to secure the ribs to the struts at multiple points, and two non-absorbable heavy sutures are that circumvent the ribs are used to secure the metal strut at the both ends. Only, in case of severe pectus deformities or the presence of a platythorax the bilateral flail chest segments created are stabilized by and additional parasternal metal struts. The strut(s) are removed after 15 months.

Robicsek in 2000 reported a large series extending over a period of three decades using a synthetic mesh which was applied as a *hammock* to maintain the position of the sternum [4] (Fig. 27.18). In this procedure, segments of the involved costal cartilages are resected subperichondrially, beginning at the level of the height of the sternal depression. A minimum length of 1 cm of cartilage at both ends with the aim of preserving the growth centres. The xiphoid process is detached from the sternum. The lower end of the sternum is raised and is freed of mediastinal attachments. Staying close to the sternal edges, the perichondrial and intercostal strips are disconnected. Effective drainage of the entire operative field is achieved by widely connecting the retrosternal space with the right pleural cavity and draining the right chest through an intercostal drain. A transverse wedge osteotomy is then done at the upper level of the sternal depression and the posterior lamina is broken by bending the sternum forward. A sheath of large-pore Marlex mesh (CR Bard Inc., Covington, GA, USA) is cut roughly to the size of the previously existing chest wall depression and placed under the sternum like a hammock. The edges of the mesh are spread tight and sutured to the remnants of the costal cartilages or bony ribs with heavy, non-absorbable filaments. The detached xiphoid process is mobilized and sutured to the lower edge of the mesh. The edges of the pectoralis major muscles are adapted and the incision is closed without additional drainage.

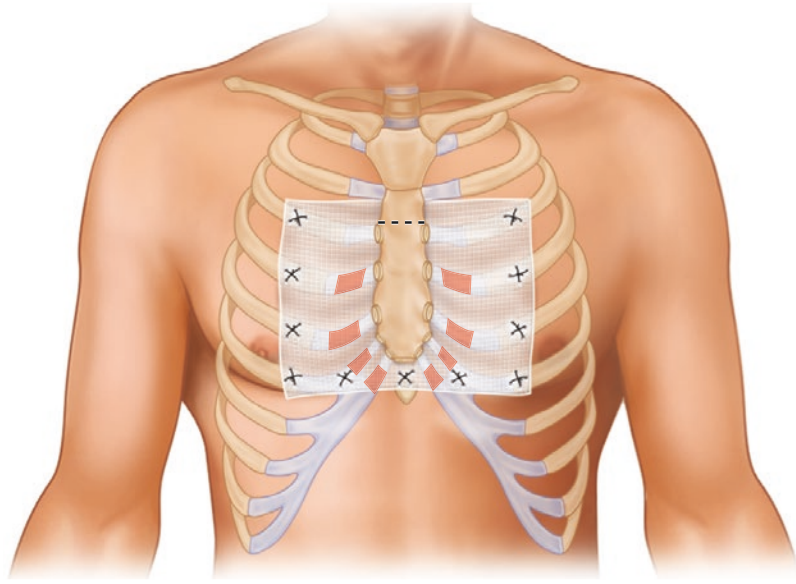

Fig. 27.18 Schematic view of pectus excavtum repair as performed by Robicsek

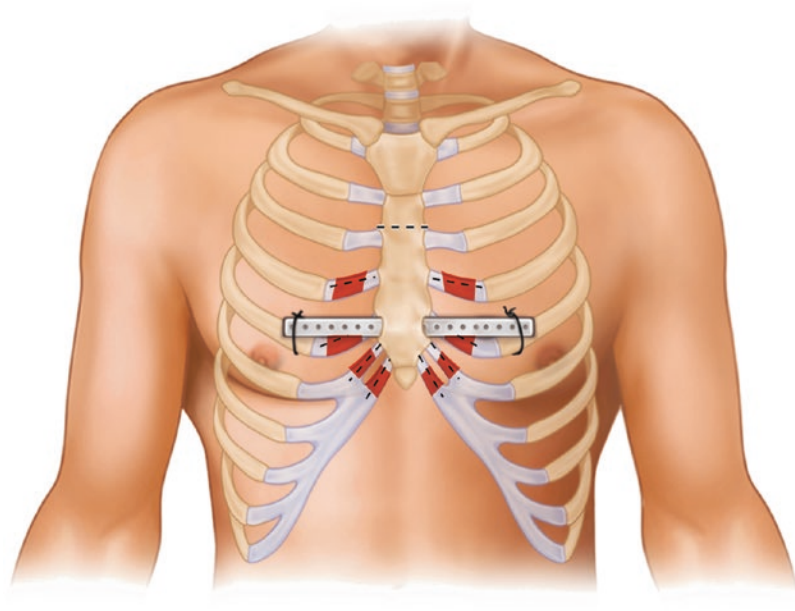

Fig. 27.19 Schematic view of pectus excavtum repair as performed by Hu

Hu in 2008 presented a procedure with subperichondrial resections and trans-sternal strut placement [42] (Fig. 27.19). A transverse incision is made at the level of the xiphoid and the pectoralis major muscle is elevated from the sternum with ribs. Subperichondral resection of all deformed rib cartilages and a transverse wedge osteotomy were made across the anterior table of the sternum at the level where the sternum angles to depress posteriorly. Nonabsorbable sutures were placed across the osteotomy. In this procedure when subperichondral resection of deformed costal cartilages is performed, it is recommended to preserve the costochondral junction. The incisions made through perichondrium should be clean, so that after removal of the deformed cartilage, the perichondral sheaths can be sutured to form a tube to enable regeneration of new cartilage tissue. The intercostal muscle bundles attached to the side of the sternum are left intact and attached to the sternum. The metal strut for sternal elevation is passed through the sternum and is bent to an "arch" shape, after which the perichondrial sheath is wired to the strut to provide better remolding for the chest wall.

Masaoka in 2012 presented a correction procedure without the use of metal struts with five different series modifications [43] (Fig. 27.20).

- In Series I the patients underwent the original Ravitch procedure (Fig. 27.20a), which was characterized by (a) resection of total deformed cartilages, (b) detachment of the sternum from the xiphoid, perichondria, and intercostal muscles, and (c) transverse sternal osteotomy and tripod fixation which involved fixation of the osteotomized sternum with a wedge of bone chip and overlap sutures of the obliquely transected bilateral 2nd or 3rd rib cartilages.
- In Series II the modifications involved (a) suturing of the sternum and rectus abdominal muscle and (b) plication of the perichondria of the removed costal cartilages and attachment to the sternum (Fig. 27.20b).
- In Series III two important modifications were adopted to strengthen the remodelled anterior chest wall. They were (a) shortening of the sternum by a 1–2 cm resection at the 5th intercostal level and (b) reduction of the length of the resected cartilages and suturing both stumps of the cartilages (Fig. 27.20c).
- In Series IV, the modifications involved an oblique osteotomy on the posterior surface of the sternum to correct the rotation of the sternum, and a bone chip was wedged in this slit. Tripod fixation remained unchanged (Fig. 27.20d). This procedure was planned for adults and a different procedure was devel-

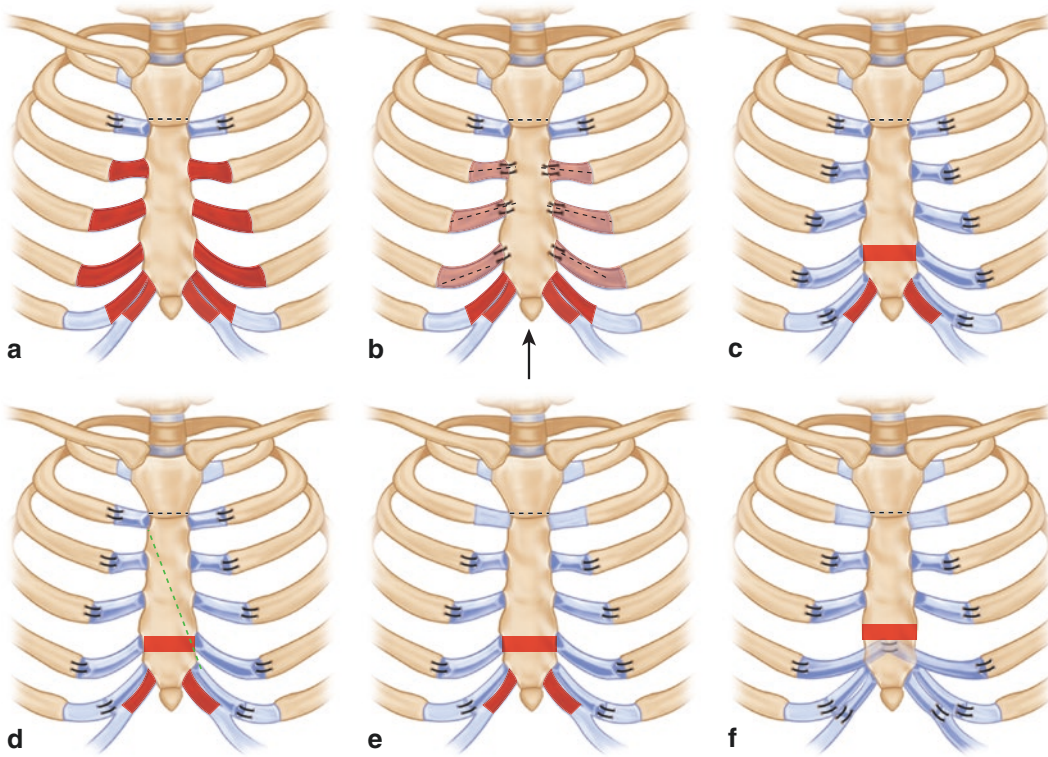

Fig. 27.20 Schematic view of pectus excavtum repair as performed by Masaoka (**a**) Series I, (**b**) Series II, (**c**) Series III, (**d** and **e**) Series IV, (**f**) Series V

oped for children. The tripod fixation was omitted because the sternum could be elevated effortlessly by resection of deformed cartilages (Fig. 27.20e). The adult-type procedure was abandoned because of difficulties in correction of platythorax and asymmetry pectus without the use of a metal strut.

- In Series V, sternal resection was performed at the 5th costal level, moving slightly to the cranial side, which was supported by a bridge constructed with bilateral 4th or 5th rib cartilages (Fig. 27.20f). This procedure was developed to strengthen the sternal support by autogenous tissue.

Nuss in 1998 reported on a minimal access procedure for the repair of pectus excavtum [44] (Fig. 27.21). The procedure involves bilateral chest wall incisions in the mid-axillary line and depending on the severity of the deformity, insertion of 1–3, arched metal bars through small lateral incisions, pushing them forward through both pleural cavities and the retrosternal space, then turning them with their convex side up, pushing forward the depressed anterior chest wall. The ends of the metal par are secured with stabilizer plates, with the trend to use a single stabilizer plate on the right side by most of the Centres. The bars are left in place for a period of 3 years. Nuss modified the original procedure, with the aim of avoiding injuries to vascular structures and to address the problem of bar dislocations. The modifications included (a) use of endoscopic visualization to observe the progression of the introducer tip in the thorax, (b) use of introducer to establish a tunnel for bar placement and (c) securing the bar with stabilizer with a polydioxanone (PDS) sutures around the bar and underlying ribs on the right side, and (d) use of two or more bars in the patients with asymmetry or platythorax. A variety of this technique was reported by Schaarschmidt in 2002, where the bars are positioned under the sub-muscular space rather than

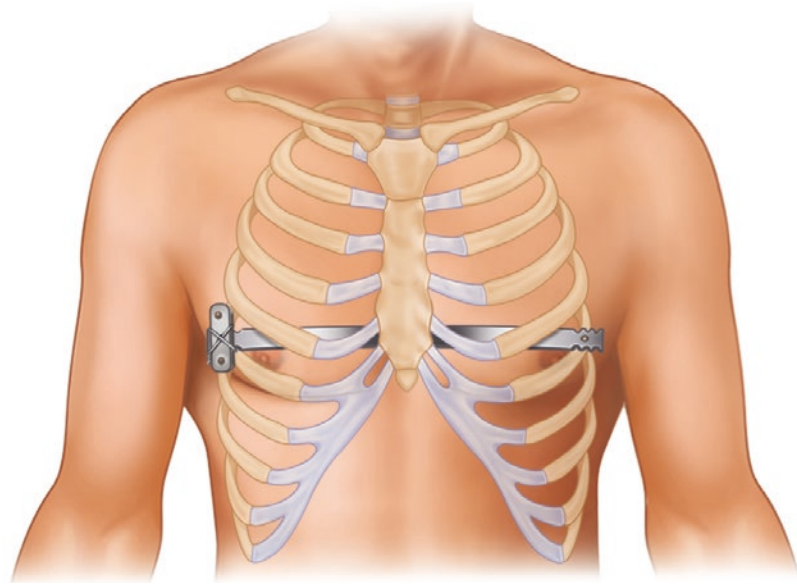

Fig. 27.21 Schematic view of pectus excavtum repair as performed by Nuss

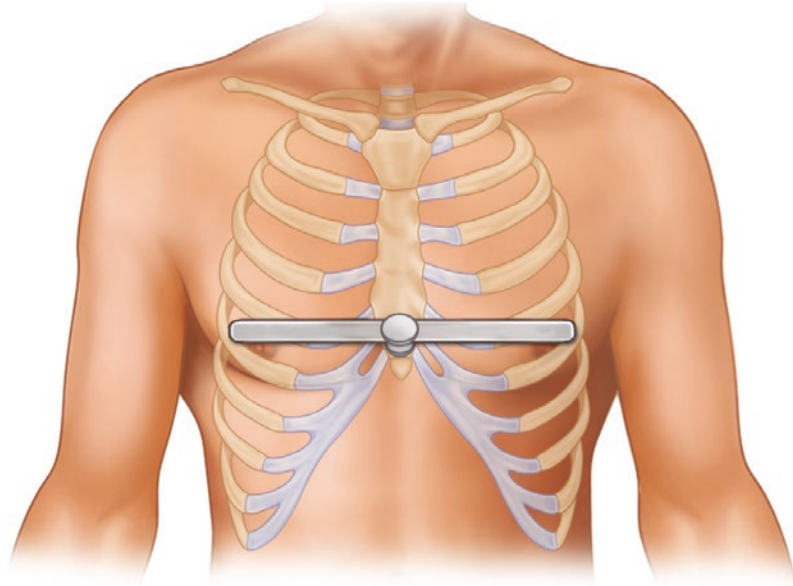

Fig. 27.22 Schematic view of pectus excavtum repair as performed by Harisson

through the pleural spaces [45]. Park in 2010 added three further modifications: (a) use of a 'crane' to elevate the depressed sternum before introducer is inserted and (b) use of morphology-tailored bar shaping system named "terrain contour matching" and (c) multipoint fixation of the bar (the bar is fixed at the hinge point which is done by passing a needle directly through the overlying skin, passing it around the rib, and back through the skin and tied) [46]. Pilegaard in 2008 proposed to use absorbable stabilizer in order to avoid chronic pain and necessity of removal. However, since these stabilizers break easily, further research is needed if they are to be used as part of the standard procedure [47].

Harrison in 2010 proposed the Magnetic Mini-Mover Procedure (3MP) for the correction of pectus excavutum [48] (Fig. 27.22). The 3MP uses a magnetic implant coupled with an external magnet to generate force sufficient to gradually

remodel the deformities. The magnimplant is set at surface of the sternum, and the magnatract is set on the rear side of the external brace, which is worn by the patient. The principle of this procedure is that the magnetic attraction between the magnimplant and the magnatract is used to lift up the sternal depression.

There are various procedures that have been suggested for the repair of pectus excvatum by plastic surgeons. In 1970, Masson and colleagues implanted a preformed silastic implant, with Dacron mesh patches on it, through a transverse incision just below the xiphoid [28]. Snel in 2009 presented a review of 75 patients with silastic implants in which complications occurred in seven patients (43 %) (Prolonged seroma in five, in which the implant was removed in one patient while repositioning was necessary in 2) [49]. Further, in two patients implants were removed due to pain and discomfort. Michlits in 2009 performed free fasciocutaneous infragluteal flap [50]. These plastic surgical procedures may provide better cosmesis; however, they cannot reduce compression on hearts and lungs from the sternal depressions. Hence, they are not suitable for symptomatic patients. Moreover, implantations of foreign body are not free from complications and always bear the risk of infection and displacement.

Outcomes

Since there are numerous surgical procedures that have been designed and performed with modifications for pectus excavtum, comparison between the procedures is not possible. Any new procedure that evolves is always '*mistakenly*' compared to the 'classical' Ravitch procedure of the 1950s; a procedure which has undergone hundreds of modifications and if performed is reported as 'modified' Ravitch procedure. The future course of pectus excavtum surgery will require well-controlled evidence based trials which should compare procedures with more effective and less radical ones.

Evaluation of Large Series with Open Repairs

In order to evaluate the complications, larger series in the hands of experienced surgeons need to be evaluated. Table 27.1 presents the list of authors with large experience in open repair of pectus excavutum deformities. However, the complications and the outcome in these series present large variations in patient populations are not uniformly presented.

In the series of operations reported by Ravitch, there were two patients with staphylococcal infection, in one of these patients the removal of the sequester was necessary. No other complications were mentioned in this series [51]. In the series reported by Haller, morbidity was reported to be less than 5 with 10 % patients requiring redo procedures for recurrences during the evolution of the technique [38]. After the evolution of the technique, unsatisfactory results were reported in 3 of 352 procedures and good to excellent results were achieved in 95 % of the patients.

In the series reported by Shamberger using the Welch procedure, the repairs were completed

Table 27.1 Authors that reported large single Centre experiences on "open" pectus repairs

Author	Publication year	Cases	Location	Technique
Ravitch	1965	164	Baltimore, USA	Ravitch
Shamberger et al.	1988	704	Boston, USA	Welch
Haller et al.	1989	664	Baltimore, USA	Modified Ravitch
Robiscek et al.	2000	800	Charlotte, USA	Marlex-mesh "hammock"
Saxena et al.	2007	1031	Münster, Germany	Willital-Hegemann
Hu et al.	2008	398	Sichuan, China	Strut
Fonkalsrud	2009	912	Los Angeles, USA	Strut
Masaoka et al.	2011	307	Nagoya, Japan	Metal free

with a low complication rate of 4.4 % [35]. Six complications were associated with Steinman pin fixation which was employed in the initial 28 patients. Major recurrence occurred in 17 patients (2.7 %) and with revisions necessary in 12. Satisfactory long-term results were achieved in the remaining 97.6 % of the patients, with follow-up ranging from 2 weeks to 27 years.

In the series reported by Robicsek [4], relevant postoperative complications were bleeding from the internal thoracic artery, infection, and seroma formations. The frequencies of these complications were not reported.

The series reported by Saxena et al. had a complication of 5.7 %. This series reported on long-term follow up and evaluation of late results which demonstrated a 1.4 % rate of major recurrences requiring surgical corrections. In this series mild recurrences were observed in 3.6 % patients, and these patients did not undergo surgical corrections. Furthermore, subjective complaints of the patients before the procedures were reported to be eliminated in 97 % of the patients.

In the series reported by Hu, early removal of metal bar due to dislodgement was necessary in four patients [42]. Whereas, recurrent depression was observed in three patients (0.75 %), a protrusion of 2nd or the 3rd costal cartilages was seen in five patients (1.26 %). Reconstructive procedures were successful in 98.7 % patients. Hu further reported the cardiac functions in these patients returned to normal level as observed by healthy individuals of comparable ages.

Fonkalsrud reported his series with a complication rate of 8 % [52]. Recurrences were reported in 5 (17 %) patients in Series-I, 15 (4.3 %) patients in Series-II, 7 (3 %) patients in Series-III and 4 (1.3 %) patients in Series-IV. From the four Series combined, a total of 22 (2.4 %) patients underwent surgery for recurrences. Surgeries for recurrences were reported to decrease during the learning curve. Satisfaction of surgical outcomes was evaluated and 94.2 % the patients or parents considered the results to be very good or excellent.

Masaoka reported 34 (11 %) patients with complications in his series of 307 patients [43]. He reported on six different series of procedural variations with variable groups of patients in each series, hence comparison of results is not possible.

Evaluation of Large Series with Minimal Access Repairs

With the advent of minimal access repair of pectus excavatum (MARPE) of 1998, complications and outcomes of this procedure are also difficult to evaluate, due to the evolution of this procedure over the past decade and the learning curve of the surgeons that performed such procedures.

Kelly in 2010 reported the outcomes of MARPE on 1215 patients in 2010 with an evaluation based on grades of satisfaction of the patients or the parents [53]. Excellent results were gained in 85.3 %. In their series, the majority of complication were categorized as minor, except for the single case of hemothorax that occurred due to injury of the internal mammary vessels. These complications were related to the period immediately during or early after repairs. The late complications were mainly associated with the displacement of the bar in 64 (5.7 %) patients in whom 45 required repositioning of the bar. It was also observed that the bar displacement rate was 12 % when no stabilizer plate was used, 6 % when wires were used to secure the pectus bar to the stabilizer plate and 2 % when pericostal sutures were placed to secure the bars. Allergy due to the implanted pectus bars was reported in 3.1 % of the patients.

Park compared the results of 1170 patients in two series with procedures performed between 1999 and 2002 to those performed between 2006 and 2008 [46]. A decrease in the total number of complications from 17 to 7.5 % was observed between the two periods. Decrease in the rates of pneumothorax from 7.5 to 0.8 %, bar displacement from 3.8 to 0.5 % and reoperation from 5.1 to 0.8 % were also evident in the two periods respectively.

Pilegaard reported on a series of 383 patients with 178 (49 %) pneumothorax, two bleeds, four pleural effusion, 11 seroma formations and in 11, and eight deep infection in eight [54]. Seven (1.8 %) patients were required surgical repositioning for displaced bars, whereas an additional 13 patients the stabilizer plate required removal because of pain.

The potential of injury to the heart or the great vessels that is associated with massive haemorrhage is the most dreaded complication of

MARPE [55, 56]. This complication could be life-threatening and require emergency surgery. This complication could occur during the surgical procedure, or in the late period of the postoperative course or at the time of bar removal [57, 58]. Despite the evolution of MARPE these complication still occur and are being increasingly reported.

Standardization of Results

The scenario in the repair of pectus excavtum type of deformities has had an enormous shift in the change of indication which has moved from the treatment of symptomatic patients in the past to the more contemporary esthetic and psycho-sociological indication for repair. The evolution of diagnostic and imaging technology such as computed tomography 3D reconstructions (that cannot be performed repeatedly due to associated concerns with radiation) to non-invasive non-radiation associated videostereoraster among others have to be standardized in order to compare results being reported (Figs. 27.23 and 27.24). Comparison of results will have to take into account the following factors prior to and after surgery before data interpretation based on evidence is possible:

1. Type of pectus deformity (symmetric/asymmetric)
2. Shape of the thorax (presence/absence of platytorax)
3. Presence of combined deformities
4. Lower rib eversion (Fig. 27.25)
5. Age of the patient at the time of surgery
6. Cardiovascular evaluations
7. Pulmonary evaluation
8. Imaging (Computed tomography/Videosteroraster/Caliper/Flexible ruler)
9. Evaluation of result by patient/parent
10. Evaluation of the result by surgeon
11. Psychological evaluation
12. Use of a standardized procedure
13. Complications during and immediately after the procedure
14. Late complications
15. Complications during bar removal (if metal is used)
16. Long-term follow up

The channeling of the variations in the presentation of pectus excavatum type of deformities, standardization of perioperative diagnostic investigations and clear guidelines for outcome criteria need to be established for the proper evaluation of results.

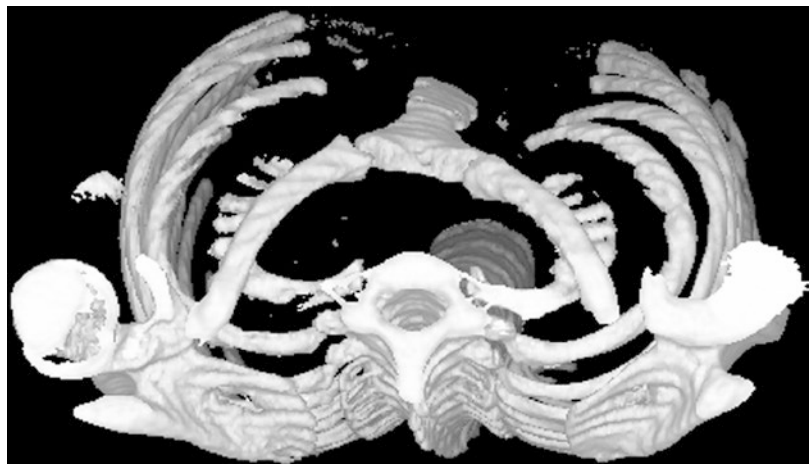

Fig. 27.23 Three-dimensional computed tomography of the pectus excavatum deformity showing the rotation of the sternum towards the right

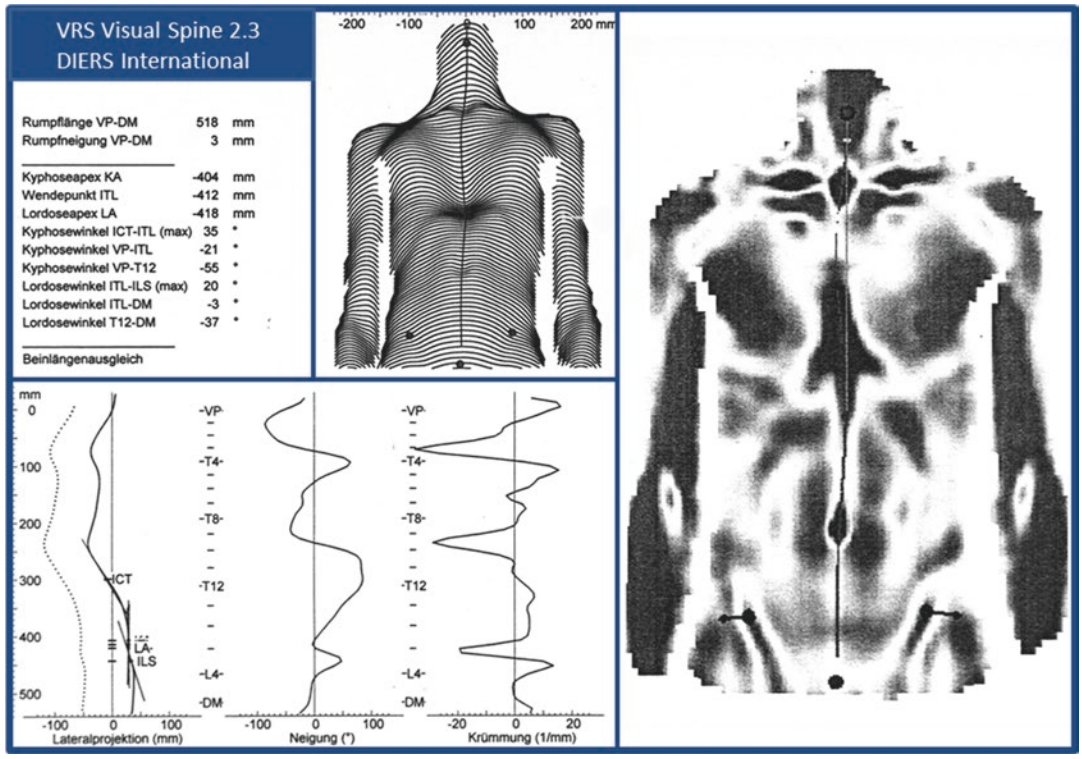

Fig. 27.24 Videostereoraster images of a patient with pectus excavatum demonstrating the contours of the chest wall

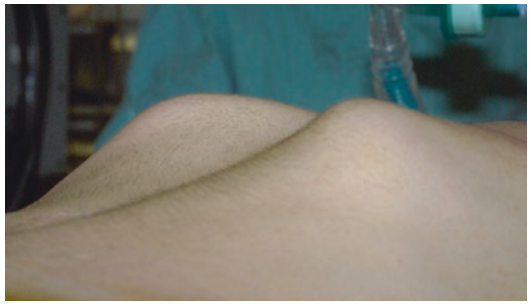

Fig. 27.25 Image of a patient with pectus excavatum associated with lower rib flaring or "winged ribs"

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The Vacuum Bell Treatment

28

Frank-Martin Haecker

Introduction

Surgical repair of Pectus excavatum (PE) in childhood and adolescents represents a well-established procedure in contrast to non-surgical treatment. Previously used operative techniques to correct PE were largely based on open procedures (e.g. Ravitch technique) and minimally invasive techniques. In 1998, Nuss et al. reported for the first time on their 10 year experience using a new technique of minimally invasive repair of pectus excavatum (MIRPE) [1]. Today, the MIRPE technique is well established and represents a common used technique [2–5]. More details are described in the following chapters of this book. However, conservative treatment of PE has a long tradition. The use of a vacuum bell to elevate the funnel in patients with PE, represents a potential alternative to surgery in selected patients.

With the widespread use of the MIRPE procedure the character and number of complications has increased [2–4, 6, 7]. Above all, recent studies report on an increasing number of near fatal complications [7–13]. Additionally, in many cases of PE, the degree of pectus deformity does not immediately warrant surgery. Some patients are reluctant to undergo surgery because of the pain associated with postoperative recovery and the risk of imperfect results. In this situation, the introduction of the vacuum bell for conservative treatment of PE has made this alternative therapy a focus of interest of patients and physicians.

The procedure of applying a vacuum to elevate the sternum was first used more than 100 years ago [14]. Spitzy and Lange reported their experience using a glass bell to correct PE. Inadequate material and relevant side effects eliminated the routine use of this method for conservative treatment of PE. Despite the risks and unsatisfactory results after operative therapy for some patients, there has been little progress in the therapeutic use of the vacuum therapy during the last few decades. In the meantime, materials have improved and the vacuum devices can now exert strong forces. In 1992, the engineer E. Klobe, who himself suffered from a PE, developed a special device for conservative treatment of PE [15]. Using his device during a period of 2.5 years, he was able to elevate the sternum and to correct his funnel chest to an extent that no funnel was visible any more [15].

Initial results using this method proved to be promising [16, 17]. Information on such new therapeutic modalities circulates not only among surgeons and paediatricians, but also rapidly among patients. In particular patients who refused operative treatment by previously available procedures, now appear at the outpatient clinic and request to be considered for this method.

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The Vacuum Bell

A suction cup is used to create a vacuum at the chest wall. The body of the vacuum bell is made of a silicon ring and a transparent polycarbonate window. A vacuum up to 15 % below atmospheric pressure is created by the patient using a hand pump (Fig. 28.1). Three different sizes (16, 19 and 26 cm in diameter) exist allowing selection according to the individual patients age (Fig. 28.2). The medium size model is available in a supplemental version with a reinforced silicon wall (type "bodybuilder"), e.g. for adult patients with a small deep PE. Additionally, a model fitted for young girls and women is available (Fig. 28.3). Pilot studies performed by Schier and Bahr [15, 16] showed that the device lifted the sternum and ribs immediately. We could also confirm this effect by thoracoscopy during the MIRPE procedure [18]. According to the user instructions and our experience, the vacuum bell should be used for a minimum of 30 min, twice per day, and may be used up to a maximum of several hours daily.

The vacuum bells by E. Klobe are CE certified and patent registered. In USA, the device was approved by the food and drug administration (FDA) in May 2012.

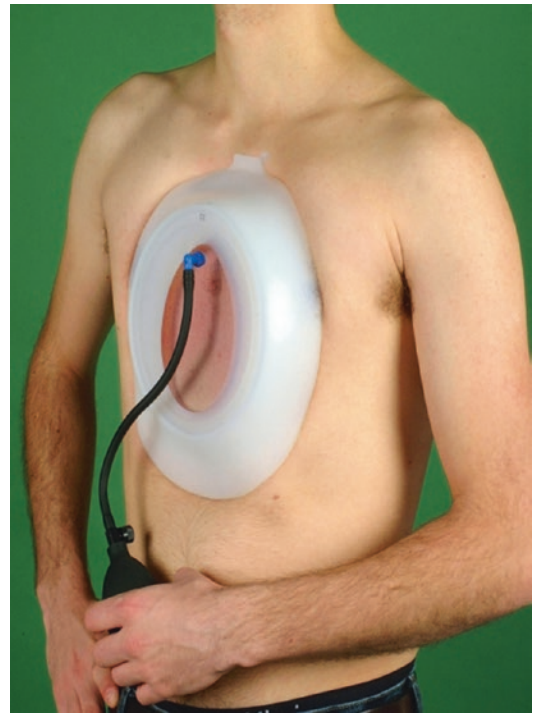

Fig. 28.1 Application of the vacuum bell

Indication and Contraindication

Indication for conservative therapy with the vacuum bell include patients who present with mild degree of PE, who want to avoid surgical procedure and who are reluctant to undergo surgery because of pain associated with the operation. Additionally, patients' concerns about imperfect results after surgery have to be noticed. Contraindications of the method comprise skeletal disorders, vasculopathies, coagulopathies and cardiac disorders [17]. To exclude these disorders, a standardised evaluation protocol was routinely performed before beginning the therapy.

Complications and relevant side effects include subcutaneous hematoma, petechial bleeding, dorsalgia and transient paresthesia of the upper extremities during the application. Rib fractures were reported elsewhere, but never seen in our patients' group.

Methods

In our unit, we offer a specialized outpatient clinic for chest wall deformities. Patients are referred by paediatricians, general practitioners or by orthopaedic surgeons. Since information on such new therapeutic modalities circulates not only among surgeons and paediatricians, but also rapidly among patients, we see an increasing number of patients who refer themselves. All patients are informed about the option of conservative vs. surgical therapy to correct PE. Standardised evaluation includes history of the patient and his family, clinical examination and photo documentation. The depth of PE is measured in a standardized supine position. Contraindications of the vacuum bell method comprise skeletal disorders, vasculopathies, coagulopathies and cardiac disorders. Meticulous family history is necessary to capture

Fig. 28.2 Vacuum bell in three different sizes (*left* 16 cm, *middle* 19 cm, *right* 26 cm in diameter)

Fig. 28.3 Vacuum bell, model for adolescent girls/women

these contraindications. To exclude cardiac anomalies, we do routinely cardiac evaluation with electrocardiogram and echocardiography.

When the patient and/or the parents decide to perform the conservative vacuum therapy, the first application of the vacuum bell occurs during the outpatient clinic visit under supervision of the attending physician. The appropriate size and model of the different type is defined. Patients learn the proper application of the device. In children under the age of 10 years, parents are instructed to use the device and children apply the vacuum bell under supervision of their parents or caregivers, respectively. The middle of the window should be positioned above the deepest point of the PE. The hand pump should be activated until the skin becomes red and the patient complains about local pain, respectively. Patients are in a supine position for the first application. During therapy, most adolescent and adult patients apply the device in an upright position whereas parents of children under the age of 10 years prefer to continue in a supine position. With the device in position, patients may move and walk around in their home environment.

In patients with a localized deformity, it could be helpful to apply the device using the small model (Fig. 28.4). In patients with an asymmetric PE or a grand canyon type PE, it could be useful to apply the device in changing positions.

When cardiac anomalies and other contraindications are excluded, patients may start with the daily application. All users are recommended to use the device twice daily for 30 min each. In fact, the length of time of daily application of the vacuum bell varies widely between patients. Some patients follow the user instructions and apply the device twice daily for 30 min each. However, some of the adult patients use the vacuum bell up to 8 h daily during office hours. A group of adolescent boys apply the device every night for 7–8 h. Since there does not yet exist a

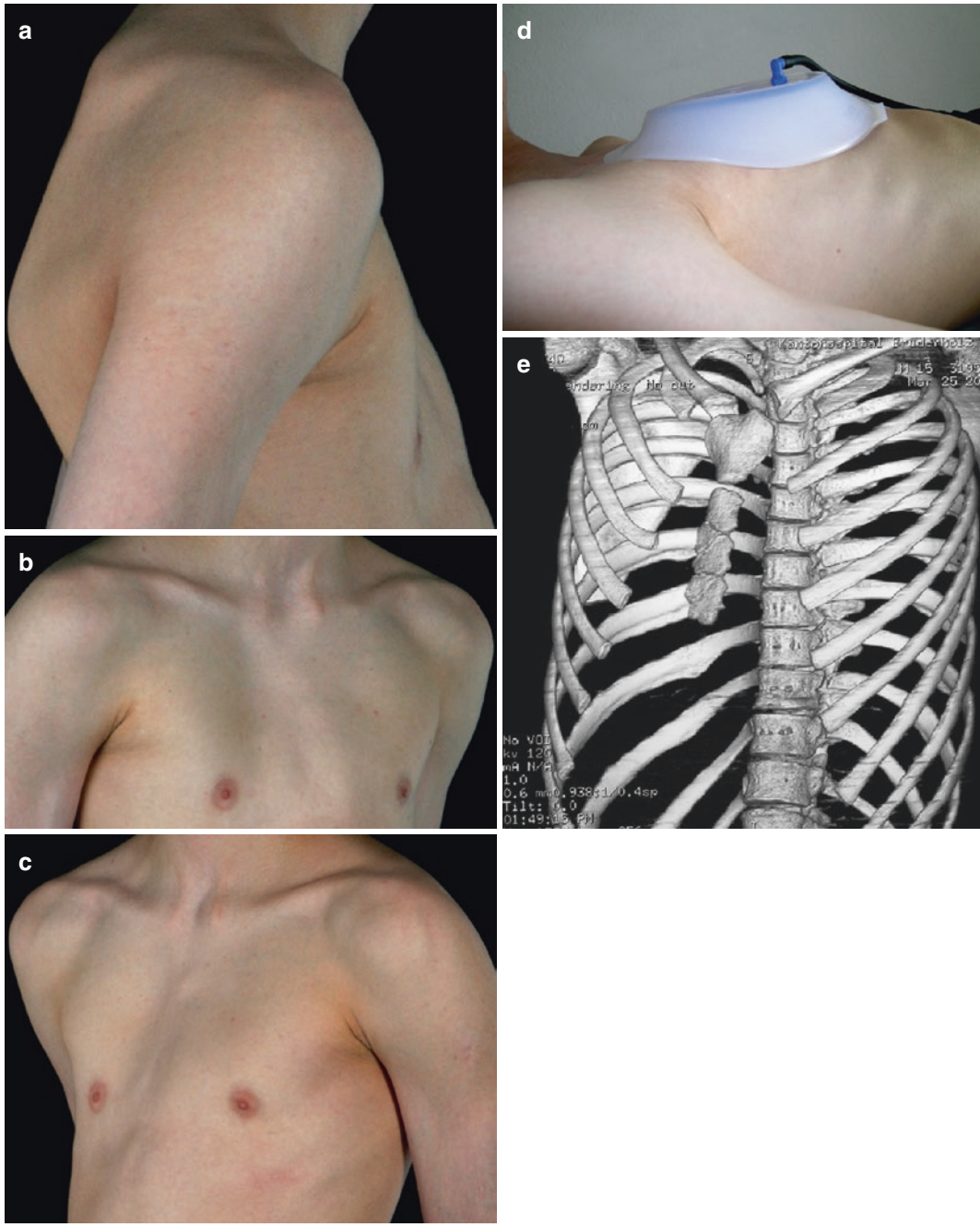

Fig. 28.4 Application of the vacuum bell in a patient with a localised deformity (**a**) lateral view, (**b**) oblique view from right side, (**c**) oblique view from left side, (**d**) application of the vacuum bell in supine position, (**e**) thoracal CT scan with 3D-reconstruction

detailed study protocol for the application, the duration and frequency of daily application depends on the patients individual decision and motivation.

Patients undergo follow-up at 3–6 monthly intervals including clinical examination, measurement of depth of PE and photo documentation. Tips and tricks to optimize the application

will be discussed. Clinical examination focuses on the improvement of depth of PE as well as on relevant side effects such as persistent hematoma and/or skin irritation. The endpoint of therapy is defined by the patient's individual decision which is confirmed by our clinical examination during the routine outpatient clinic visit.

Patients

Our patients group comprises applicants aged from 3 to 61 years. The "optimal" age for this treatment has still to be defined. Patients' perception is different and depends on applicants age in many aspects. As mentioned previously, we observe age specific differences of success [17]. During the first 1–5 applications, most of the patients experience moderate pain in the sternum and report a feeling of uncomfortable pressure within the chest. Adolescent and older patients develop moderate subcutaneous hematoma, which disappears within a few hours. Temporary side effects like transient paresthesia of the upper extremities during the application and/or mild dorsalgia are reported by some patients. These symptoms disappear when lower atmospheric pressure will be used during application. Analgesic medication should not be necessary and is not reported from any patient and parents, respectively. As mentioned above, the application of the vacuum bell in children aged 3–10 years should be supervised by parents or caregivers. In this age group, no relevant side effect is reported.

We summarized preliminary results of a subset of our patients group in a previously published study [19]. 133 patients (110 males, 23 females), aged 3–61 years (median 16.21 years) used the vacuum bell for 1 to maximum 36 months. When starting with the application, patients presented with a PE with depth from 2 to 5 cm. In 105/133 patients (79%), after 3 months of treatment an elevation of more than 1 cm was documented. In 18 patients (13.5%), the sternum was lifted to a normal level after 18 months (Figs. 28.5 and 28.6). The longest follow-up after discontinuation is 5 years, and the success until today is permanent and still visible (Fig. 28.6). Daily application of the whole group was 108 min/day in average. Application was terminated after approx. 22 months in average (range 6–36 months). In three patients with asymmetric PE, the depth of PE has decreased after 9 months, but the asymmetry is still visible (Fig. 28.7). Three patients were dissatisfied with the postoperative result (two patients after MIRPE, one patient after Ravitch procedure) and started treatment with the vacuum bell. All these patients were very well motivated and compliant which is a basic precondition for a successful therapy. At follow-up, all patients were satisfied and expressed their motivation to continue the application, if necessary. All our patients were recommended to carry on undertaking sports and physiotherapy, so that the accompanying improvement of body control was an important factor in outcome.

However, 13/133 patients stopped the application after 19.9 months in average, due to an unsatisfactory result (four patients) and decreasing motivation (nine patients). 12/13 patients underwent MIRPE.

A more differentiated analysis will enable us "to see behind the curtain". Age and gender specific differences, depth of PE, symmetry or asymmetry, concomitant malformations like scoliosis and/or kyphosis, etc. may influence the clinical course and the success of this therapy.

Intraoperative Use

Our preliminary experience with the vacuum bell method encouraged us to use the device intraoperatively during the MIRPE procedure to facilitate the dissection of the transmediastinal tunnel and the advancement of the Pectus introducer, the riskiest step of the MIRPE procedure. When creating the vacuum, the elevation of the sternum is obvious as demonstrated by Schier and Bahr for the first time [16], and persists for a distinct period of time. Therefore, we considered that the vacuum cup may also be useful in reducing the risk of injury to the heart and the mammary vessels during the MIRPE procedure. The device might also be applied for placement of the pectus bar. Since the manufacturer of the device does

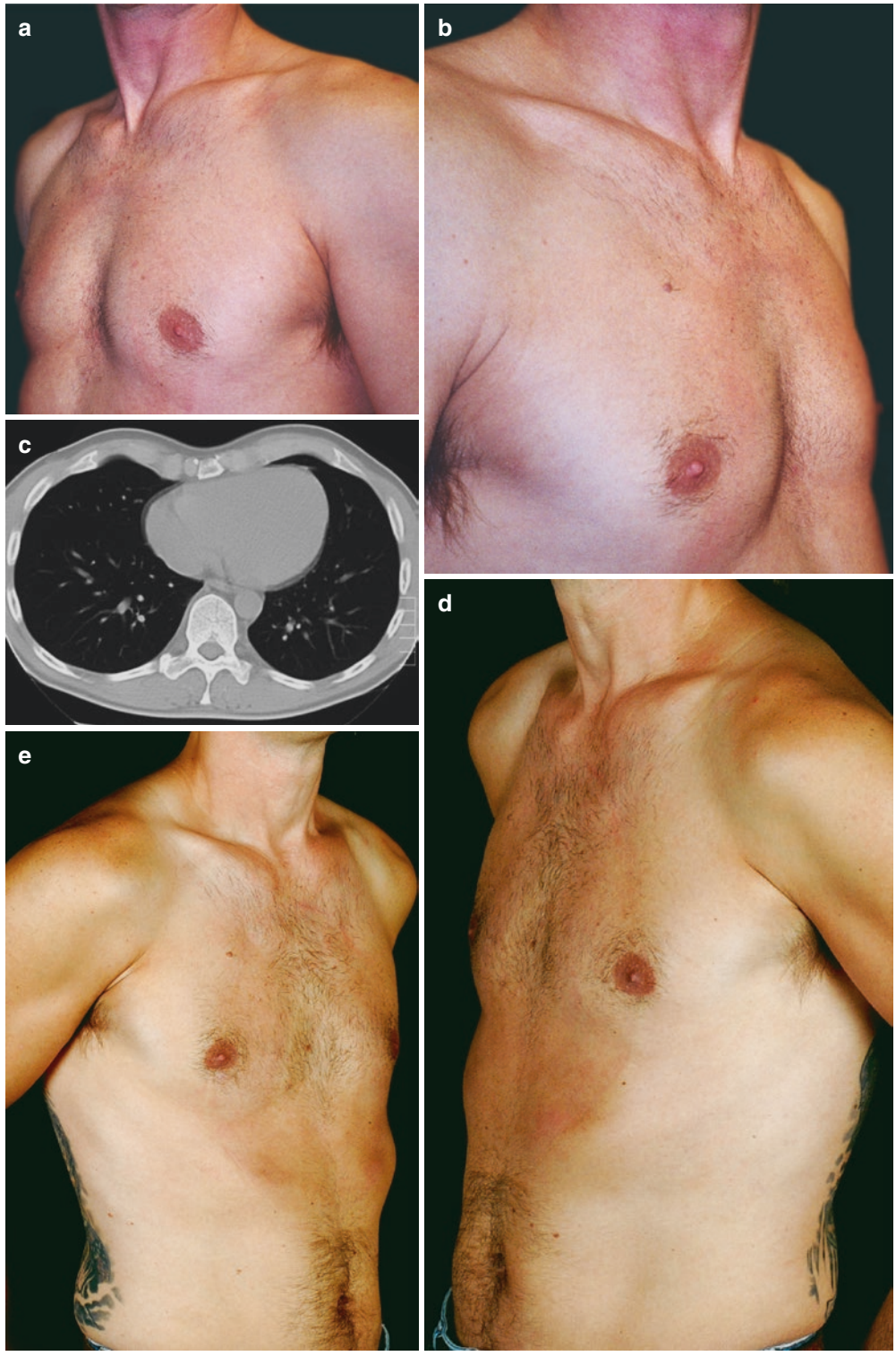

Fig. 28.5 Fourtyfive year old patient, before (*left*: depth of PE = 2.5 cm) vacuum bell therapy and after 12 months (*right*: depth of PE = 0.5 cm) (With kind permission from Springer Science + Business Media: Pediatric Surgery International, The vacuum bell for conservative treatment of pectus excavatum: the Basle experience. Volume 27, Issue 6. 2011. Frank-Martin Haecker)

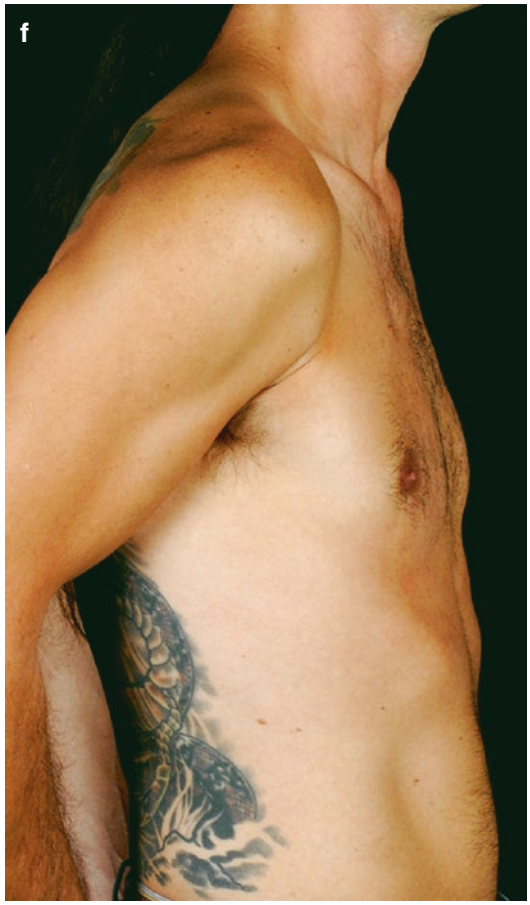

Fig. 28.5 (continued)

not have a license for sterilisation of the vacuum bell, this additional use had to be considered as "Off-label". In agreement with our hospital hygieneist and bearing in mind the nature of the material, we used gas sterilization for preparation of the device for intraoperative use.

In a pilot study performed from 2005 to 2010, 50 patients aged from 9 to 28 years (average 14.95 years; 39 males and 11 females) were operated on for PE using the MIRPE procedure. Thirty eight patients underwent primary surgery. Twelve patients (11 male, 1 female) used the vacuum bell for a period of 4–36 months (average 19.9 months) before surgery, and discontinued the application due to decreasing motivation and/or insufficient success. The vacuum bell was applied for retrosternal dissection and advancement of the Pectus introducer as well as placement and flipping of the pectus bar. The use of the vacuum bell led to a clear elevation of the sternum and this was confirmed by thoracoscopy (Fig. 28.8). Advancement of the Pectus introducer and placement of the pectus bar was safe, successful and without adverse events in all patients. No evidence of cardiac and/or pericardiac lesions or lesions of the mammary vessels was noted intraoperatively by using right sided thoracoscopy. Additionally, no midline incision to elevate the sternum with a hook was necessary [18].

Unanswered Questions

Optimal age for Vacuum Bell Therapy

As mentioned above, the optimal age for this treatment has still to be defined. We observe age specific differences of success. In our experience, growth spurt during puberty is the most important period to influence degree and depth of PE. Further studies have to evaluate whether beginning with the vacuum therapy before puberty will be more useful than starting during puberty or even later.

Quantitative Measurement of Pressure

The success of a therapeutic procedure not only requires a good technique, but also depends on an appropriate indication. It would be useful to measure the pressure that is necessary to lift the sternum during the first application. This measurement would enable us to divide patients into different groups, to identify "perfect" patients, and allow us to predict more accurately who of the users will benefit from this method and in whom the method will not work. We are working on such a device to measure the pressure under the vacuum bell.

Long-Term Results

Long-term results including 10 years and more are still missing. Further studies are necessary to elucidate these facts.

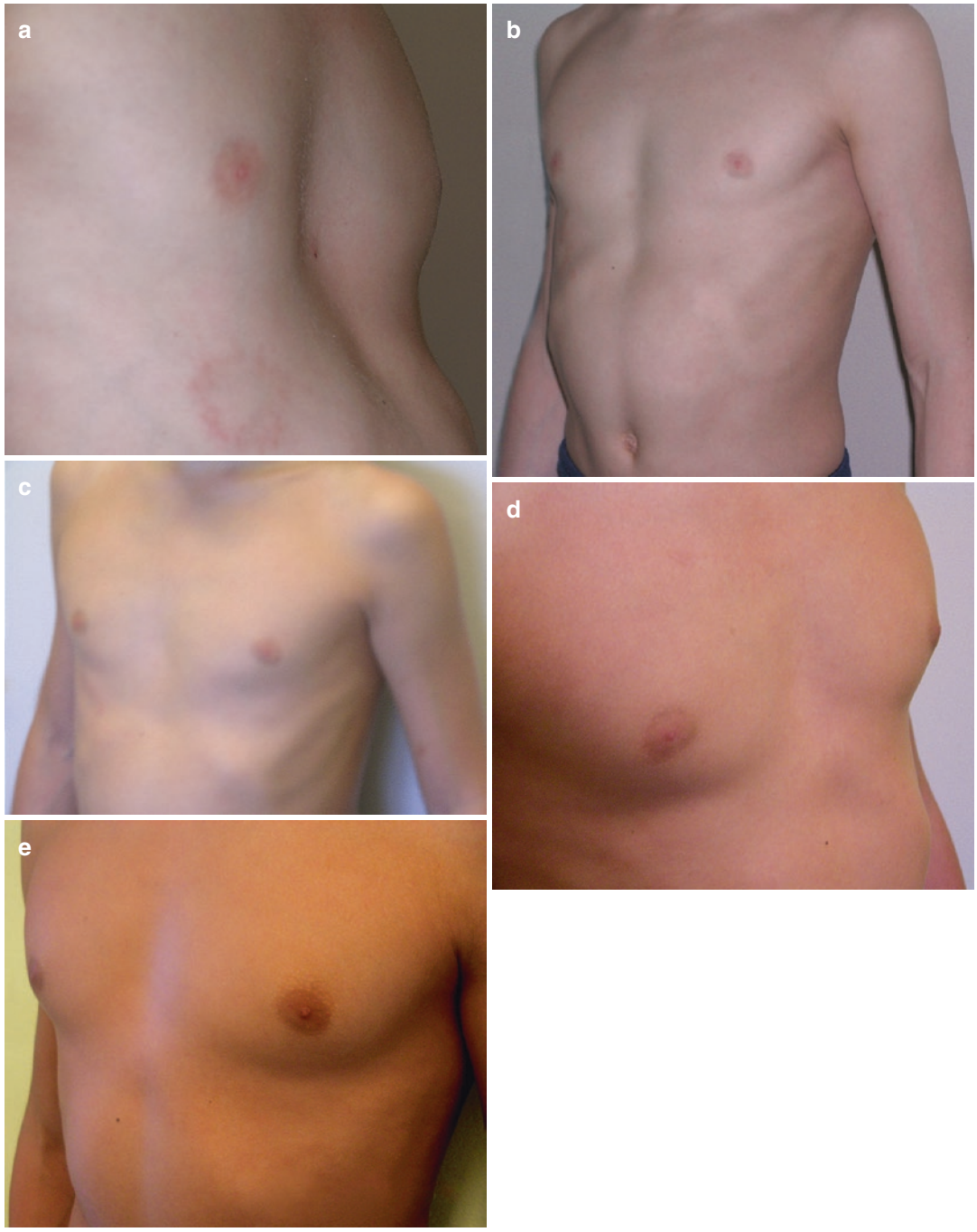

Fig. 28.6 Nine year old boy, (**a**) before (*left*: depth of PE = 2.8 cm) vacuum bell therapy, (**b**) after 10 months (*right*: depth of PE = 1.6 cm) and (**c**, **d**, **e**) 36 months after therapy (*below*) (With kind permission from Springer Science + Business Media: Pediatric Surgery International, The vacuum bell for conservative treatment of pectus excavatum: the Basle experience. Volume 27, Issue 6. 2011. Frank-Martin Haecker)

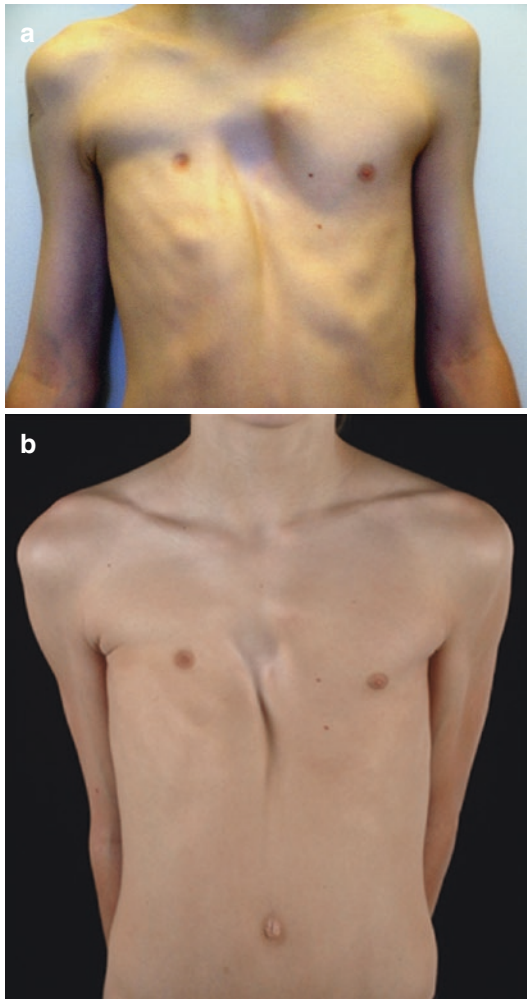

Fig. 28.7 Ten year old boy with asymmetric PE, (**a**) before (*left*) vacuum bell therapy, and (**b**) after 12 months (*right*) (With kind permission from Springer Science + Business Media: Pediatric Surgery International, The vacuum bell for conservative treatment of pectus excavatum: the Basle experience. Volume 27, Issue 6. 2011. Frank-Martin Haecker)

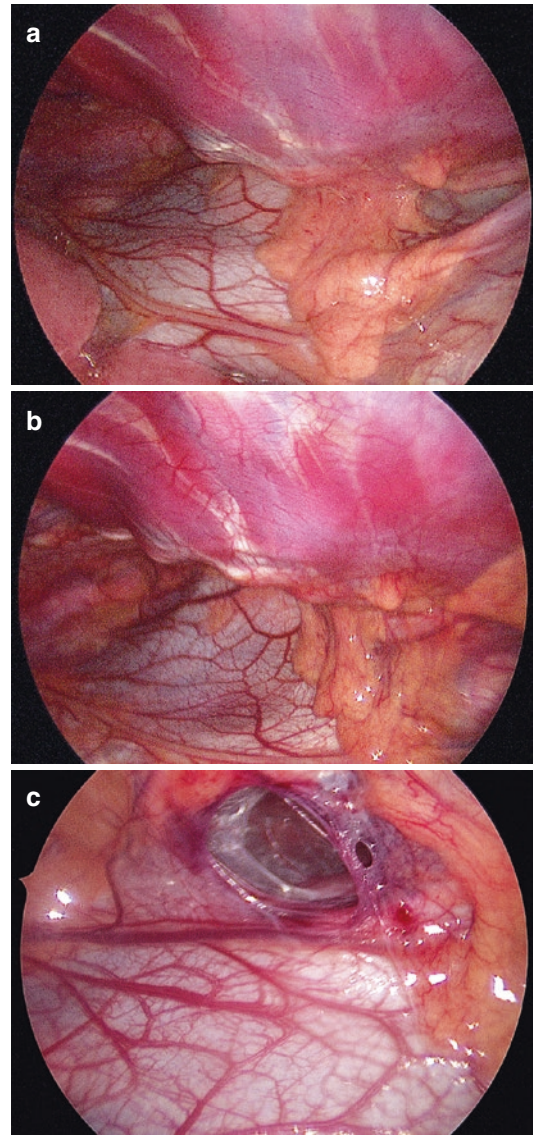

Fig. 28.8 Intraoperative use of the vacuum bell (**a**) Thoracoscopic view of the pectus excavatum (right thorax) (**b**) application of the vacuum bell and rising of the sternum and (**c**) elevation achieved between the sterum and pericardium to offer safe space for bar placement

Costs of Treatment

In most European countries, costs of treatment have to be paid by patients and parents, respectively. In some countries in South America, acquisition of the vacuum bell is covered by the individual national health care system or the local insurance. In USA, approval of the FDA was obtained in May 2012.

Pre-Treatment before Surgery

Physicians and patients discuss about the benefit to use the vacuum bell preoperatively prior to MIRPE procedure. Since in our country the majority of patients have to pay for the device, most of our patients are not interested in this "pre-treatment". Additionally, we observed no

significant difference between patients who used the vacuum bell before surgery, and patients who underwent primary surgery [18].

Objective Assessment of Depth of PE

To estimate the "objective" success of this treatment modality is very difficult. The definition of success may vary considerably between individuals. Depth of PE, symmetric vs. asymmetric deformity, as well as patients' age and sex represent important variables. Various scales and measurement methods including X-rays and computed tomography have been used to quantify the degree of deformity. Our method of assessment of depth of PE is not exact enough, esp. regarding the age specific differences. New methods for non-invasive assessment of chest wall growth may provide more detailed, objective information concerning the severity of PE. A 3-D laser scanner might help us to assess degree of PE and to follow-up our patients during vacuum therapy.

Conclusion

The vacuum bell therapy may allow some patients with PE to avoid surgery. Especially patients with symmetric and mild PE may benefit from this procedure. The application is easy, and we noticed a good acceptance by both paediatric and adult patients. However, the time of follow-up with a maximum of 10 years is still not long enough, and further follow-up studies are necessary to evaluate the effectiveness of this therapeutic tool. Additionally, more differentiated analysis must focus on age and gender specific differences. The intraoperative use of the vacuum bell during the MIRPE facilitates the introduction of the pectus bar. In any case, the method seems to be a valuable adjunct therapy in the treatment of PE.

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Thoracoscopic Aspects in Minimal Access Repair of Pectus Excavatum

29

Amulya K. Saxena

Introduction

Minimal access technique for the repair of pectus excavatum (MARPE) has been accepted as the procedure of choice in the treatment of young children, teenagers as well as adults with pectus excavatum (PEX) [1, 2]. Since its introduction, this technique has gained popularity among patients and surgeons mainly because it is perceived as a minimal access operation with good esthetic results. The procedure involves bilateral thoracic incisions, and a custom-modeled steel bar is guided below the sternum to elevate it to correct the deformity. The initial procedure described by Donald Nuss in 1998, did not employ thoracoscopy to visualize the preparation of the retrosternal space and placement of the pectus bar. Later, in 2002, an update and modificated version of the MARPE technique was reported by the same group using routine thoracoscopy to improve safety during mediastinal dissecton [3]. Although thoracoscopy gained acceptance after its introduction from this time and is now part of the MARPE procedure at most of the centers, it has been associated with complication in MARPE. The literature is scare with reports dealing with the complications related to thoracoscopy in patients undergoing MARPE and have been overshadowed by complications observed with the pectus bar [4, 5]. This chapter focuses on various aspects of the technology of thoracoscopy relevant to MARPE with the intention of identifying areas in which complications can occur as well as their prevalence along with the precautions and preventions to be taken to avoid them.

Most reports on MARPE have focused on minimizing the bar and stabilizer dislocation; as this still remains the most important issue as was evident from the initial report of Donald Nuss (8.8 %) [6]. The same report presented a 52 % complication rate resulting from residual pnuemothorax, with intervention required in 2.7 % of the patients. Despite this initial report, little attention has been paid in the literature to the aspects of thoracoscopy and its complications, which rank second to those of bar displacement in MARPE.

Aspects of the Technology

The Light Source

Minimal access surgery has over the past two decades used two main types of light source; Xenon and Halogen. When comparing the two, Xenon has a more natural color spectrum and a smaller spot size than halogen. The yellow light of the halogen bulb is compensated for in the video camera system by white balancing.

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The halogen light source is used in the medical field since last two decades but the temperature of these lights is 3200 Kelvin which makes it alter from natural light. Hence, the more suitable light source for endoscopic cameras involves the utilization of Xenon. The Xenon light source emits a spectral temperature of color of approximately 6000 Kelvin on average for a power of 300 W.

However, the endoscopic operating fields at present are being illuminated in a new light field. The Light Emitting Diode (LED) technology has replaced the Xenon lighting and is the key module for the future of endoscopic light sources (Fig. 29.1). These light sources are presently being supplied in different technologies, power classes and with a wide range of accessories. LED light sources have a very long lifetime and they virtually never need to be changed. The efficiency of LED lamps and modules is exceptionally high, i.e. the power consumption is low and very little heat is generated in the device. These new light sources are virtually inaudible, a feature that makes an important contribution to improved working conditions in the operating room. Furthermore, all devices have a glare shield if the light cable is not plugged in. The light color of the LED light sources is very constant and similar to the light generated by xenon lamps. The light power of the LED series is comparable with 180 watt xenon light.

Fig. 29.1 Endolight LED Source (Richard Wolf GmbH, Knittlingen, Germany) The dialog light control is a unique feature of the ENDOLIGHT light sources in conjunction with an ENDOCAM Logic HD camera. The camera controls the light volume of the light source fully automatically in dialog mode. This gives an optimum image quality while at the same time providing optimum protection for patients and equipment (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

Light Cable

Minimal access surgery depends on the artificial light that is available through fiberoptic cables. The principle of fiberoptic cable is based on the total internal reflection of light which enables the light beam to be conducted with all its intensity along a curved glass rod resulting from multiple total internal reflections.

Fiberoptic cables are made up of a bundle of optical fiber glass threads of sizes between 20 and 150 micron in diameter, which offer a very high quality of optical transmission. The multimode fiber maintains the intensity of light and offers the advantage of passing in a curved path through the light cable. With progressive use of light cables successive breakage in optical fibers occurs. Although this cannot be prevented, the coiling of light cables in acute diameters should be avoided as much as possible to minimize this breakage. Visualization in the hemithorax during MARPE and delicate manipulation of the introducer during this procedure require excellent vision. Care should be taken to have a light cable that provides sufficient illumination during MARPE, as loss of vision due to decreased illumination could result in serious intraoperative complications.

In order to overcome the shortcomings of conventional cables "armored light cables" (Richard Wolf Gmbh, Knittlingen, Germany) are gaining popularity. These cables have been engineered for reliability with three layers of protective coating to provide a robust light cable which is lightweight and extremely flexible. It consists of an innermost layer, which has a stainless stainless steel coil to prevents the fiber bundle from being crushed or broken. This is followed by a Teflon strain relief sleeve which encases the first layer and restricts the bundle from being pulled apart by extreme lateral forces. Medical-grade silastic rubber tubing covers the cable entirely and protects against the harsh environments of sterilization. These light cables are offered in straight, right angled, high temperature and dual cable models, ranging in 1.6, 2.5, 3.5 and 4.5 mm sizes. All cables can be steam autoclaved, gas sterilized or cold soaked.

Intensity Control Circuit

Manual adjustments allow the light source to be adjusted to a power level defined by the operating surgeon. The luminosity of most of the current light sources is adjustable to prevent excessive light during close-up viewing and darkness in more distant views. Contemporary light source systems however have automatic intensity adjustment technology. The images captured by a video camera are transformed into an electronic signal which is coded in order to be transmitted. The coding dissociates the luminance and color of the image. Optimal quality luminance signal are calibrated to 1 mV. Overexposed images surges the electronic signal pass >1 mV, and underexposed images make the signal drop <1 mV. Light sources equipped with adjustment analyze the luminance. These systems permit work to be performed at different distances from the target in desirable viewing conditions. However, the requirements of these systems in no longer necessary as the cameras presently available are often equipped with systems which are capable of automatic gain control in poor light condition which can overcome issues associated with luminance fluctuations.

Telescope

The rigid scopes are based on the Hopkins rod lens system for endoscopic surgery provides good resolution and satisfactory depth perception. There are three area of variations when it comes to the choice of endoscopes: (1) number of rod lens systems (6–18 Rods), (2) Directions of view (0°–120°) and (3) size (1.5–15 mm). The endoscope most commonly used in the pediatric age group is the 30° 5 mm and 33 cm length scope. Various scopes mainly varying in their directions of view have been proposed for MARPE, an area of importance that has been discussed later in a forthcoming section of this chapter. However, a operating scope may be an useful instrument in MARPE especially if the patient has had prior surgery or pulmonary conditions that may have led to adhesions (Fig. 29.2). This

Fig. 29.2 Operating scope Set "Leipzig Egg" (Richard Wolf GmbH, Knittlingen, Germany) possess a 5.5 mm Operating scope with a 3.5 mm working channel and a 0° direction of view 0°,and working length of 215 mm (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

instrument for single-port thoracoscopy permits minimal access to perform adhesion removal through a working channel in the operating scope. Various instruments can be introduced through this working channel or liquid can be suctioned off (Fig. 29.3). The salient features of the operating scope (Richard Wolf Gmbh, Knittlingen, Germany) are that it possess a diameter only 5.5 mm diameter, offers an optimum image quality with 50.000 pixels, has no rod lens breakages caused by overload, provides working channel for 3.5 mm instrument, working instruments can be quickly changed and are always in the visual field of view, and suctioning liquid even works when the instrument set has been inserted.

Endoscopic Camera

Endoscopic camera are available either in single chip or three chip versions based on the charged couple device (CCD) for the mixing of the three primary colors red, blue and green) in different proportions (Fig. 29.4). The CCD is located in the head of camera to capture an image taken by the scope. The camera head also consists of an objective zoom lens that focuses the image of

Fig. 29.3 Assortment of instruments that can be used along with the Operating scope set "Leipzig Egg" (Richard Wolf GmbH, Knittlingen, Germany) (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

the object on the CCD which then converts optical image into an electrical signal that is sent through the camera control unit. The number of pixels determines the resolution with the average chip containing 250000–380000 pixels. In single chip camera, these three primary colors are sensed by single chip. In three chip camera there are 3CCD chips for separate capture and processing of three primary colors. In three chip cameras color separation is achieved with a prism system that overlies the chips. This system gives a higher resolution and better image quality because the pixel number is three times greater. Present systems available such as the Endocam® Logic HD (Richard Wolf Gmbh, Knittlingen, Germany) features fast and easy control of all functions facilitated by the touch display on the unit and an on-screen display with intuitive and smart menu navigation (Fig. 29.5) They form the centerpiece of the CORE (Complete Operation Room Endoscopy) system (Richard Wolf Gmbh, Knittlingen, Germany) (Fig. 29.6).

Focusing of Endoscopic Camera

Endoscopic camera need to be focused before inserting inside the thoracic cavity for MARPE. At the time of focusing, these camera tips should be placed at a distance of 5 cm away

Fig. 29.4 Three chip CCD camera head (Richard Wolf GmbH, Knittlingen, Germany) (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

Fig. 29.5 ENDOCAM Logic HD (Richard Wolf GmbH, Knittlingen, Germany) camera forms the central module in a new generation of digital camera heads, intelligent LED light sources, efficient light cables, brilliant monitors and advanced chip-on-the-tip technology (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

from the target. This distance is optimum for setting the focus adjustments because during endoscopic surgery this is the distance maintained for performing procedures.

Endoscopic Insufflator

The electronic CO_2 insufflattor is an insufflation unit for use in endoscopic procedures which offers controlled pressure insufflation of the cavity to achieve the necessary work space. Carbon dioxide is the preferred gas because it does not support combustion, is soluble which reduces the risk of gas embolism, and is cheaply sourced. Automatic insufflators enable to preset the insufflating pressure, and pump gas until the required pressure is reached within the cavity. The insufflator activates and delivers gas automatically when the pressure within the cavity falls because of gas leakage from the port site or the port valve. Patient safety is ensured by optical and acoustic

Fig. 29.6 Complete operation room endoscopy (CORE) system (Richard Wolf GmbH, Knittlingen, Germany) provides an operation suit with excellent ergonomics to perform endoscopic surgical procedures (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

alarms as well as several mutually independent safety circuits. The pressures safely used for pediatric and adolescent video-assisted thoracic surgery can be seen in Fig. 29.7 (Adapted from Saxena et al. 2007) [7].

Presently, high-performance insufflators are available that offer excellent advantages for endoscopic procedures. The Insufflator High Flow 45 (Richard Wolf Gmbh, Knittlingen, Germany) has the capability to provide enormous flow capacity of max. 45 l/min and comes in four hardware versions offering an intuitive operating concept with a large touchscreen (Fig. 29.8). This saves time, improves control and provides outstanding support for the surgeon when carrying out interventions. The four versions include (a) Basic, (b) EVAC with smoke evacuation, (c) HEAT with gas heating and (d) EVAC + HEAT with smoke evacuation and gas heating.

Ports and Trocars

The trocar is the stylet which is introduced through the port as a system and are available with different type of tips. The cutting tips of these trocars are either in the shape of a three edged pyramid or a flat two edged blade. Conical tipped trocars are less traumatic to the tissue. Ports are in general made from plastic or metal, with the plastic devices designed in such a way as to minimize the reflection of light from the endoscope. Most of the disposable plastic trocar have a spring loaded mechanism that withdraws the sharp tip immediately after it passes through the chest wall to reduce the incidence of injury of viscera. Trocar and ports are of different sizes and diameter depending upon the instrument for which it is used. The diameter of ports ranges from 3 to 30 mm; the most common size for MARPE being 5 mm.

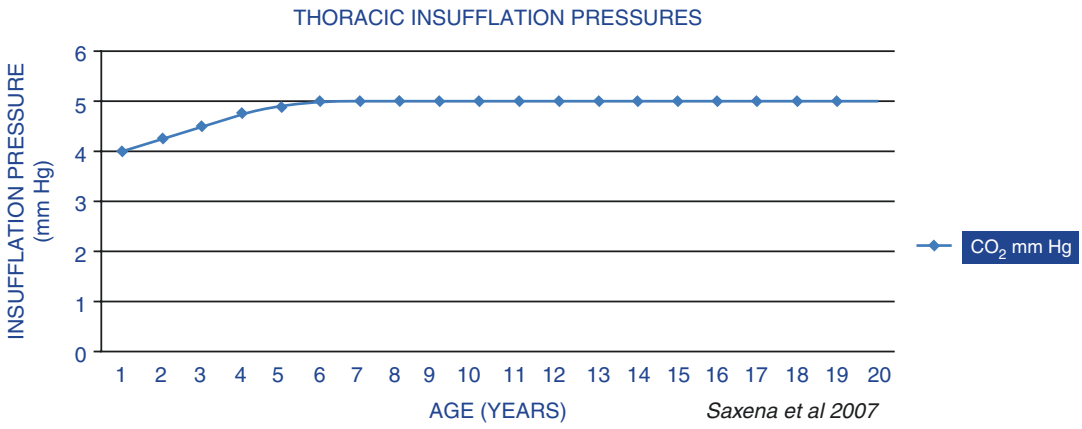

Fig. 29.7 Insufflation pressures recommended for the pediatric and adolescent population for video-assisted thoracic surgery (VATS) procedures

Fig. 29.8 Insufflator High Flow 45 (Richard Wolf GmbH, Knittlingen, Germany) offers a large touchscreen for better overview and efficiency, intuitive menu navigation, and automatic adjustment of display brightness to ambient light (Image: Courtesy of Richard Wolf GmbH, Knittlingen, Germany)

Metal trocars have different type of tips (pyramidal tip, eccentric tip, conical tip or blunt tip). Since a single port is used in MARPE, these ports have a valve mechanism at the top. Port valves provide internal air seals, which allow instruments to move in and out within a port without the loss of the insufflated gas. These valves are manually or automatically retractable during instrument passage.

Procedural Details with Regards to Thoracoscopy

The MARPE procedure is performed by making small bilateral transverse incisions on either side of the chest. A pectus bar of appropriate size is selected and custom modeled to the desired chest contour. Right-side thoracoscopy is performed by insertion of a 5-mm port through the right lateral chest wall just above the estimated level of the diaphragm in the anterior axillary line. The intrathoracal placement of the port is confirmed by observing lung tissue with a 5-mm scope after which a pressure of 4–5 mmHg is employed for insufflation. The introducer is inserted into the thorax from the right side (Fig. 29.9) and using the tip the introducer is carefully passed in the retrosternal space to the left hemithorax (Fig. 29.10). An umbilical tape is attached to the tip of the introducer after which it is withdrawn. Using the umbilical tape as a guide, the pectus bar is introduced and placed in the desired position (Fig. 29.11). After the pectus bar is flipped, a single stabilizer plate is used on the right side and polydioxanony sutures (PDS, Ethicon, Somerville, NJ, USA) are placed around the pectus bar to anchor to the underlying ribs (Fig. 29.12). At the end of the procedure after closure of the thoracic incisions and before removing of the port, the anesthetist applies positive pressure ventilation to

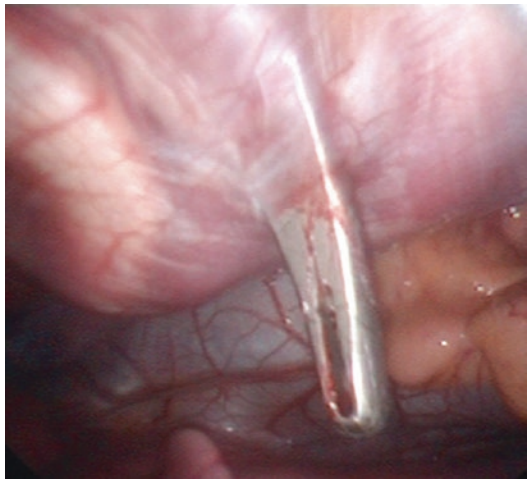

Fig. 29.9 Endoscopic view of the right hemithorax with a 5 mm 30° scope demonstrating optimal vision of the pectus introducer tip entering the right thorax during MARPE

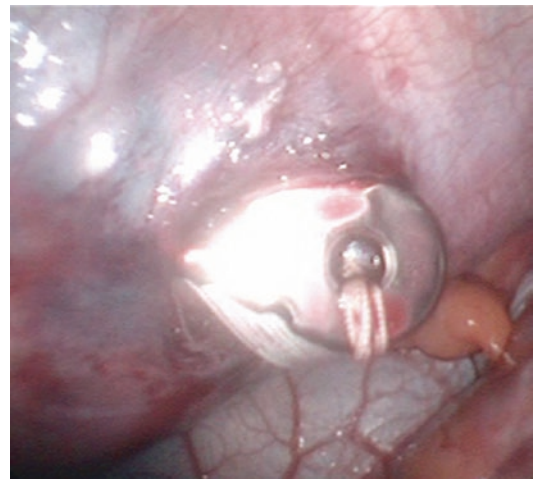

Fig. 29.11 Endoscopic view of the right hemithorax with a 5 mm 30° scope demonstrating optimal vision of the pectus bar being introduced through the right thorax during MARPE

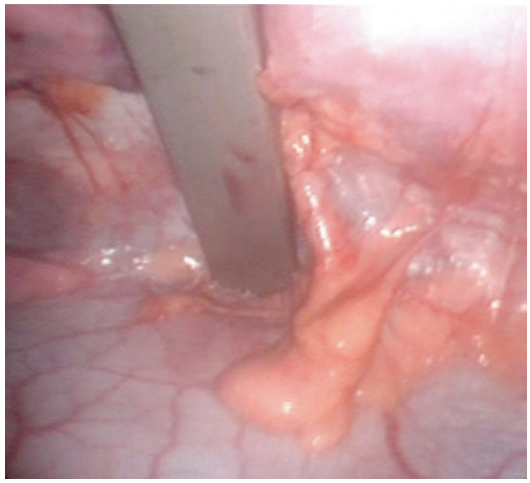

Fig. 29.10 Endoscopic view of the right hemithorax with a 5 mm 30° scope demonstrating optimal vision of the pectus introducer passed through the restrosternal space during MARPE

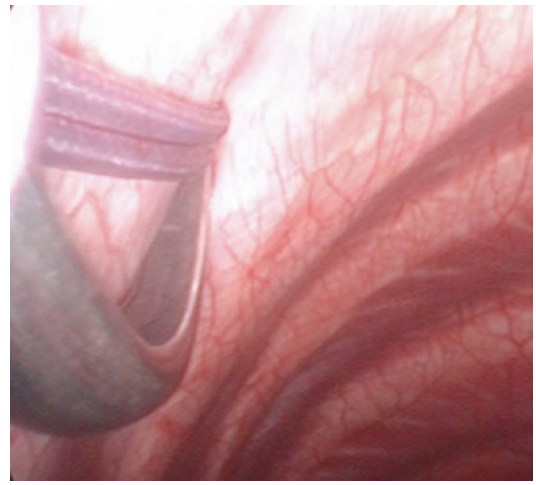

Fig. 29.12 Endoscopic view of the right hemithorax with a 5 mm 30° scope demonstrating optimal vision of the placement of a PDS cord around the rib for securing the pectus bar end during MARPE

evacuate the accumulated carbon dioxide (CO_2). At this time, the patient is brought into left semi-lateral head drop position so as to raise the level of the port to the highest position and facilitate maximum gas evacuation from the thorax. Routine postoperative chest films are part of the protocol and are taken to evaluate the position of the bar as well as to evaluate the thoracic cavity with regard to pneumothorax.

Issues with Port Placement

The most important aspect in thoracoscopy is the determination of the suitable point of insertion of the port. Port placement site in MARPE differs to a large extent from those sites chosen for other video-assisted thoracoscopic surgery (VATS) procedures. Optimum visualization during MARPE requires port placement almost at

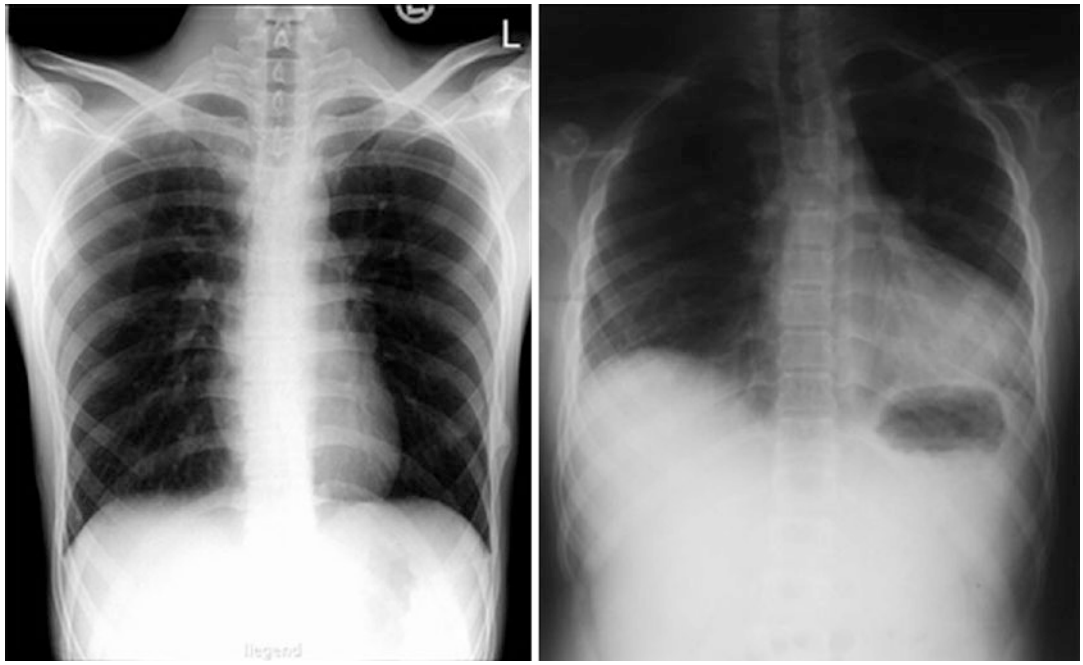

Fig. 29.13 Chest films demonstrating the variations in the level of the diaphragm of 2 patients that underwent MARPE. Correct estimation preoperatively allows correct port placement and prevent injury to the diaphragm or liver

the level of the diaphragmatic recess which is markedly lower than the mid thoracic placement of ports for VATS. Right side port insertion in MARPE carries a risk of injury to the diaphragm or the liver. Chest films and computed tomography (CT) of patients with pectus deformity have mainly evaluated the severity of the deformity as well as the relation of the depression to the heart [8, 9]. However, if thoracoscopic assisted MAPRE is to be performed, equal emphasis must be placed on the evaluation of the diaphragm and the upper abdominal parenchymatous organs, to avoid injury during port insertion. Assessment of the diaphragm should be included as part of the preoperative chest radiograph protocol (Fig. 29.13).

Insufflation Pressures

After the port is positioned in the right hemithorax, examination of the hemithorax is the first step before insufflation is started. CO_2 insufflation utilizing positive pressure has been advocated to aid in the creation of an artificial pneumothorax when single lung ventilation is not used [10]. Insufflation CO_2 pressures in MARPE have been reported in a wide range and vary from 4 mmHg (standard at our center) up to as high as 10 mmHg reported by others [11, 12]. Although CO_2 insufflation has been carried out safely under various pressures; insufflation pressures as low as 5 mmHg have been reported to result in significant decrease in cardiac index, mean arterial pressure, stroke volume, and left ventricular stroke work index [13]. Hence, precautions to use minimum pressure needed to be respected, especially after the substernal tunnel has been created; because the ipsilateral insufflation can be transferred to the contralateral chest and lead to difficulty in ventilation, a phenomenon not associated with other general VATS procedures. We recommend insufflations pressures in the range of 4–5 mmHg for thoracoscopy in MARPE (Fig. 29.7) [7].

Scopes for MARPE

The use of the type of scope has not been reported uniformly and its importance has been underesti-

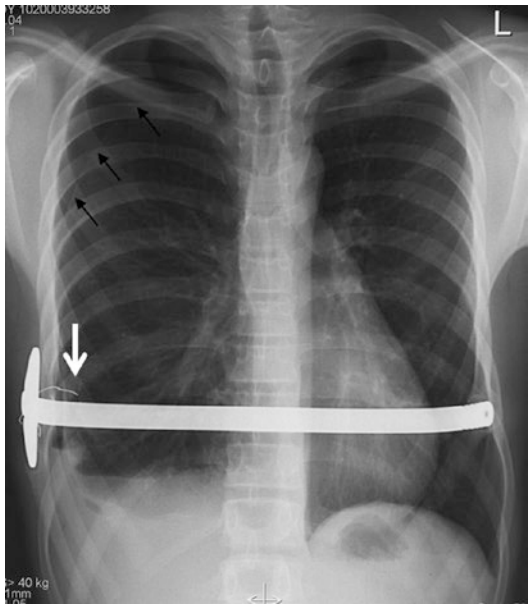

Fig. 29.14 Erect chest film demonstrating a broken wire (*white arrow*) that was used to secure the stabilizer plate in a patient with MARPE. The black arrows demonstrate the lung contours to demonstrate pneumothorax in the left hemi-thorax due to lung injury from the broken wire

mated in reports on thoracoscopic assisted MARPE [14, 15]. Scopes with an angle of vision of 30° are most widely used at Centers worldwide in the field of endoscopic surgery; and have also been used for MARPE because of their ready accessibility. Zallen et al. have reported, even the use of separate 45° and 70° scopes for mediastinal dissection as well as placement of wires to secure the bar respectively [12]. After our experience with wire breakage complications (Fig. 29.14), pectus bars have been secured to the ribs with stabilizer plates using PDS cords. Suturing of PDS cords does not require special scopes, and no complication with the use of PDS cords securing the bar and stabilizer on the underlying rib, have been observed in our series. Scopes with an angle of vision of 30° have provided an optimum operative field of vision and we recommend their use in MARPE (Figs. 29.9, 29.10, 29.11, and 29.12). Investing in scopes of different sizes such as 45° and 70° does not offer any added advantages to the procedure if proper port site selection is chosen.

Options in Thoracoscoscopy

Thoracoscopy was effectively applied only on the right side of the chest in our experience for the placement of the pectus bar. Some authors have suggested the use of left thoracoscopy for MARPE, stating advantages of eliminating the risk of cardiothoracic injury [16]. However; it is easier to perform the mediastinal dissection of the substernal tissue from the pericardial tissue from the right side, when compared to the limited view achieved with left side approach. Bilateral thoracoscopy has been suggested by other authors, but significant advantages of employing it have not been elaborated [15, 17, 18]. Determination of pulmonary tissue entrapping, between the pectus bar and the chest at the time of flipping, could be a reason for the use of bilateral thoracoscopy; however pulmonary tissue entrapping is theoretically not possible with positive insufflation pressures. Furthermore, bilateral thoracoscopy could increase the chances of chest wall paresthesia due to port injury of the intercostals nerves; which is a complication of thoracoscopy in VATS, but has not been reported in MARPE to date [19]. Since, the efficacy and safety of single-port technique for VATS has been advocated [20], only right side thoracoscopy can be considered to be sufficient for mediastinal exposure and dissection in MARPE.

UderesTimating Residual Pneumothorax in MARPE

Pneumothorax is the most common complication of thoracoscopy in MARPE (Table 29.1). Reports of pneumothorax associated problems have decreased remarkably after it was became more evident to evacuate all insufflated gas from the thorax after completion of the procedure. The trapped CO_2 can be evacuate when the anesthetist applies positive pressure ventilation and a positive end-expiratory pressure (PEEP) at the end of the procedure [21]. For this, the patient is kept the patient in left semilateral head drop position and evacuation of the gas is achieved under positive PEEP at the end of MARPE procedure, before final closure.

Table 29.1 Incidence of pneumothorax due to thoracoscopy in MIRPE reported in the literature

Author	Publication Year	Patients	Pneumothorax		Pneumothorax requiring intervention	
			Patient	Percent	Patient	Percent
Hebra [4]	2000	251	–	–	12	**4.8**
Nuss [6]	2002	329	74	52	9	**2.7**
Croitoru [3]	2002	303	155	59.7	8	**2.6**
Uemera [15]	2003	107	1	0.9	1	**0.9**
Park [14]	2004	335	23	6.9	13	**3.9**
Boehm [11]	2004	21	2	9.5	2	**9.5**
Futagawa [21]	2005	21	7	33.3	–	–
Hendrickson [16]	2005	51	20	39.2	2	**4**
Croitoru [22]	2005	50	35	70	12	**24**
Saxena [7]	2007	160	15	9.4	7	**4.4**
Zganjer [23]	2011	128	36	28.1	8	**6.3**
Shu [24]	2011	406	9	2.2	DNA	**DNA**
Hanna [25]	2013	73	3	4.1	DNA	**DNA**
Knudsen [26]	2013	938	16	1.7	16	**1.7**
Li [27]	**2014**	**30**	**5**	**16.7**	**DNA**	**DNA**

DNA data not available

However, repair of recurrent failed funnel chest repair in a large series published recently, has again demonstrated this problem in 70 % patients with 24 % requiring interventions [22]. An alternative to address this problem would be to place a 16 Fr or 20 Fr chest catheter during surgery that can be pulled out in the recovery room or later on the wards.

Patient Positioning During MARPE

Patient positioning before the implementation of thoracoscopy in MARPE did not play a major role in influencing the complications. With thoracoscopy, it is necessary to have the surgeon and the cameraman on the same side of the patient. Under anesthesia, the patients have decreased normal resting tones which allow the patient to assume non-physiologic positions. This could result in nerve ischemia when the vessels supplying the nerves become compressed [28]. If the angle of upper extremity abduction exceeds 90°; stretching and compression of the brachial plexus can occur [29]. Hence; it is recommended to position the patient on the right edge of the table with both the arms extended 90° at the shoulder joint. MARPE with the right side thoracoscopy approach allows the cameraman to be positioned close to the right thigh area and provides sufficient space for the surgeon on the right side of the patient's chest to operate, without having the need to overextend the upper extremity. Positioning of the operation team and the patient as mentioned above helps to diminish the possibility of injury to the brachial plexus.

Conclusions

Thoracoscopy is an incorporated part of the MARPE procedure. Though it has not been given importance in the literature and has rendered the MARPE safer, it is associated with pitfalls. Right sided thoracoscopy is an effective adjuvant to MARPE and is the most commonly practiced approach in MARPE. The possible complications arising from thoracoscopy in MARPE have been elaborated in this chapter with options to avoid them.

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Complications of Minimally Access Pectus Excavatum Repair

30

Frazier Frantz and Michael J. Goretsky

Introduction

Minimally access pectus excavatum repair (Nuss procedure) has been performed in over 1600 patients with severe pectus excavatum at the Children's Hospital of The King's Daughters in Norfolk, Virginia. Clinical analysis of this large patient series has allowed for identification and quantitation of intraoperative, early- and late-postoperative complications unique to this surgical procedure [1]. Based upon this information, treatment recommendations and preventive strategies for these complications have been generated. Many of these have now been incorporated as modifications to the original surgical technique and have improved overall safety and efficacy.

Intraoperative and Early Postoperative Complications

Thoracoscopy Related Complications

With adherence to a standard operative protocol, which includes thoracoscopy with CO_2 insufflation to optimize intrathoracic visualization during substernal dissection to avoid cardiac injury and three-point pectus bar fixation to minimize bar displacement, the intraoperative and early-postoperative complications of the Nuss procedure have been minimal. There have been no operative deaths. A single case of cardiac injury occurred in a patient with a history of median sternotomy and open heart surgery. Early postoperative complications in our series through 2012 have included pneumothorax requiring tube thoracostomy (2.5 %), suture site infections (0.7 %), pericarditis (0.3 %), pleural effusion requiring tube thoracostomy (1.8 %), and temporary paralysis (0.1 %), unpublished data.

Cardiac Injury Risks

Cardiac injury, the most serious potential complication of the Nuss procedure, has been reported rarely [2]. This has occurred most frequently in cases involving patients who have undergone previous open heart surgery or pectus excavatum repair. In these patients, the substernal dissection is complicated by the presence of adhesions and fibrosis that weld the tissue layers together, including the pericardium and heart. The other group of patients in whom this complication is more likely to occur is those with very high Haller indices, especially those with stiff chest walls, in whom clear visualization of the sternum and heart may be difficult to maintain. When there is frank evidence or high

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suspicion of cardiac perforation, the most important initial step is to stop the dissection and leave the pectus introducer in place. Removal of the introducer could result in further tearing of the heart or release of tamponade. The cardiac surgery team should be called and preparations should be made for blood transfusion, median sternotomy, and cardiopulmonary bypass. If the presence of cardiac perforation is unclear and the patient is stable, confirmation of injury may be facilitated with transesophageal echocardiography (TEE) to detect evidence of pericardial fluid/tamponade or encroachment of the introducer into the heart chambers. When recognized in a timely fashion, cardiac perforation can be treated successfully using cardiopulmonary bypass and direct suturing of the heart. It has become our practice to alert the cardiac surgery team preoperatively when these high risk patients are scheduled for chest wall surgery. In some instances, where patients have had previous open heart surgery, the cardiac surgeons have participated at the beginning of the Nuss procedure to release the adhesions between the heart and sternum through the inferior aspect of the previous sternotomy incision. For all cases involving these high risk populations, it is imperative to have blood available for transfusion, a sternal saw, and access to cardiopulmonary bypass, if needed.

Prevention of cardiac perforation is focused on ensuring optimization of exposure of the substernal region as well as temporary sternal elevation maneuvers to increase the space between the sternum and heart and subsequently make substernal dissection safer. Thoracoscopy can be approached via the right chest, left chest, or both chest cavities, utilizing 30- or 45-degree thoracoscopes to optimize exposure. During substernal dissection with the pectus introducer, it is imperative that the introducer tip is in view at all times. If multiple bar placement is planned, the superior tunnel should be created first. The pectus introducer should then be left in place to elevate the sternum during dissection of the inferior tunnel, which is typically at the maximal point of sternal depression. Multiple techniques are available to achieve temporary sternal elevation during difficult pectus repair. These include utilization of the Vacuum Bell [3], manual sternal retraction via a subxiphoid incision, and the crane technique [4].

Residual Pneumothorax

Pneumothorax requiring tube thoracostomy may be detected intraoperatively or during the postoperative period. These occur by one of three mechanisms: (1) inability to evacuate residual insufflated CO_2 from the pleural spaces, (2) intraoperative lung trauma with alveolo-pleural leak from lysis of adhesions present from previous lung infections or chest surgery, and (3) air leaks into the pleural space through the surgical incisions or trocar sites, particularly if the patient is breathing spontaneously prior to incision closure. CO_2 insufflated during surgery is evacuated at the end of the procedure by placing the insufflation tubing to water seal and administering multiple, large, positive-pressure breaths. Confirmation of evacuation of the insufflated CO_2 is obtained via intraoperative chest x-ray at the end of the procedure. Pneumothoraces requiring tube thoracostomy are detected intraoperatively by evidence of continued air leak (bubbling) under water seal or by the presence of a moderate or large pneumothorax on chest x-ray. Postoperatively, pneumothoraces are detected on routine chest x-rays obtained on days 1, 3, and 5 after surgery by the presence of serial enlargement. With regard to prevention, the surgical incisions should be closed prior to completing evacuation of insufflated CO_2, and spontaneous ventilation should be avoided during this portion of the procedure. The port and insufflation tubing unit should be verified to be airtight and should not be removed until the surgical incisions are closed and the air leak under water seal has ceased. Pneumothoraces that result from lung trauma are not preventable in most cases because they result from necessary lysis of pulmonary adhesions. If lung injury does occur, it is important to recognize this and to place a chest tube at the time of initial operation because this pneumothorax will almost certainly expand postoperatively.

Infection Related Complications

Early suture site infections have occurred at an incidence below national averages for clean surgical cases. By definition, these infections are limited to the skin and subcutaneous tissues, and they typically present with fever and perincisional erythema with or without drainage. Because of the risk of these infections spreading to the deeper soft tissues and potentially involving the implants, aggressive treatment with parenteral antibiotics is undertaken. Preventive measures implemented for these patients include preoperative skin scrubs with 2 % chlorhexidine, prophylactic antibiotic coverage for 24 h, and standard, sterile intraoperative techniques. Patients who present on the day of surgery with evidence of skin (i.e., folliculitis) or remote site infections are typically treated for these and rescheduled for repair due to the inherent, increased risk of surgical site or bar infection.

Pericarditis after the Nuss procedure mimics post-pericardiotomy syndrome and includes a symptom complex of retrosternal chest pain, lethargy, and fever. A friction rub may be present on physical examination, and chest x-ray may reveal an enlarged cardiac silhouette. The etiology of this is unclear, but we believe it to be attributable to metal allergy or pericardial injury that occurred at the time of bar insertion. The diagnosis is based on the presence of clinical symptoms in association with echocardiographic findings of pericardial thickening or effusion. Inflammatory parameters, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), are typically elevated. If the diagnosis is suspected, cardiology consultation should be obtained. In our series, medical treatment with steroids or non-steroidal anti-inflammatory drugs has been effective. One patient did require placement of a pericardial drain. It is noteworthy that, since implementation of preoperative metal allergy testing in 2003, the incidence of pericarditis has dropped dramatically and has not occurred over the past 4 years. The avoidance of placing stainless steel bars in metal-allergic patients, thus, appears to be an effective prevention strategy for pericarditis. An additional prevention strategy is avoidance of pericardial trauma during bar insertion. This can be accomplished via adherence to the principles outlined earlier with regard to thoracoscopic visualization of the introducer tip throughout substernal dissection and utilization of temporary sternal elevation techniques. If pericardial injury is witnessed during substernal dissection or bar placement, the hardware should not be left in the pericardial sac. In addition to the risk of pericarditis, this bar position could lead to erosion of the bar into the heart, which could manifest as significant hemorrhage at the time of bar removal. Violation of the pericardial sac should be addressed by withdrawing the hardware and creating a second substernal tunnel in the appropriate plane for bar insertion. Postoperatively, the patient should be watched closely for the development of a friction rub.

Complications Related to Bleeding

Hemothorax in the early postoperative period is infrequent and is not typically associated with vital sign changes or distinct clinical symptoms, other than pain on the involved side, which is difficult to differentiate from routine postoperative pain. It is usually detected on routine postoperative chest x-rays. All patients have been treated with thoracoscopy, clot evacuation, and tube thoracostomy. While it is clear that hemothorax results from a bleeding site that was not appreciated at the time of bar insertion or occurred afterwards, no active bleeding sites have been found in any of the patients in our series. The potential sites of hemorrhage include: (1) intercostal vessels which could be injured with instrumentation at thoracostomy sites or with pericostal suture placement, (2) mediastinal vessels anterior to the heart which could be torn during substernal dissection, and (3) internal mammary vessels. Presumably, any of these vessels could vasospasm at the time of injury and then hemorrhage postoperatively. Prevention entails optimizing thoracoscopic visualization of areas of potential hemorrhage at the time of bar insertion. If bridging vessels are visible anterior to the heart at or near the site of proposed substernal tunnel creation, it is safest to control these vessels with

electrocautery or the Harmonic scalpel prior to instrumentation.

Pleural Effusion

Pleural effusion is common after the Nuss procedure and will be apparent to some extent in most patients on routine postoperative chest x-rays. No intervention is typically required, as this fluid will be reabsorbed in the first 2–3 weeks after surgery. Spontaneous resolution is facilitated by ensuring adequate analgesia and encouraging pulmonary toilet. For a small percentage of patients, these effusions will enlarge and cause pain and impaired ventilation. Compressive atelectasis or pneumonia may occur. Tube thoracostomy will be necessary in these situations. The etiology of pleural fluid accumulation is not completely clear but is likely secondary to reaction of the pleural surface to surgical trauma and the presence of an indwelling substernal bar. Like pericarditis, this is another early complication that has dwindled in incidence since the implementation of preoperative metal allergy testing, so any allergic etiology may play some role here.

Temporary Paralysis

One of the least common yet most devastating complications in our series has been temporary paralysis which was present postoperatively in two patients. Both patients had thoracic epidural catheters placed for perioperative analgesia, a routine procedure for all patients undergoing the Nuss procedure at the time. The clinical presentation was that of anterior cord syndrome with lower extremity paralysis and loss of bowel and bladder control. With inpatient rehabilitation, significant recovery of neurologic function was observed. Despite extensive radiographic and neurologic evaluation to determine a cause, no identifiable injury or pathology of the spinal cord was demonstrated. We have postulated that these cases may have resulted from an underlying, congenital anomaly of spinal cord perfusion, which may be more prevalent in patients with pectus excavatum. Regardless of the etiology, however, the magnitude of the complication prompted discontinuation of thoracic epidural utilization for our patient population in March 2011.

Late Postoperative Complications

As with early postoperative complications of the Nuss procedure, late complications do occur but should be rare when proper techniques and principles are followed. In our experience bar displacements, overcorrection, bar allergy, wound infection, and recurrence have primarily been the late postoperative complications and all should be rare (<5 %).

Pectus Bar Dislocation

Bar displacements have historically been the most common complication we have seen and have dramatically decreased with various modifications since the initial presentation in 1998 [5]. We define bar displacement as any movement that occurs after the initial surgery. However, a shift of less than 15° (mild shift), as long as the bar is still providing the same support under the sternum, and the external result looks the same does not often require surgical intervention. If the shift occurs in the first 30 days or is greater than 15° then strong consideration for surgical revision should be done. Of 84 bar displacements, 54 (3.7 %) required repositioning. Of these 84 displacements, 16 (15.2 %) of 105 occurred before stabilizers were available, a time period covering our first 105 repairs. After the introduction of stabilizers, the incidence of bar displacement decreased from 15.2 to 6.5 % or 27 of 398 patients. When the bar and stabilizers were wired together, the incidence of bar displacement decreased to 4.3 % or 14 of 322 patients. Since we combined placing a stabilizer on the left, polydioxanone (PDS™ II (polydioxanone) Suture, Ethicon, Inc) sutures around the bar and underlying rib on the right, and the liberal use of

two bars the incidence of bar displacement has dropped to 1 % where any further intervention is required [1].

There are multiple factors that can be done to minimize bar displacement. Bar configuration is the first principle in minimizing displacement. The correct length is important since bars that are too long or bent too much can cause significant long term pain and discomfort. We recommend a length that is measured from mid axillary line to mid axillary line and subtracting 1 in.. Slightly shorter bars are equally efficacious as long as they are positioned correctly. The bar should be bent so that it fits comfortably on the side and there is a short flat section in the middle to support the sternum. Proper bar positioning is one of the most important factors in minimizing displacement and the most frequent cause of movement that we have seen from other centers. It is critical that the bar enters and exits the chest medial to the top of the pectus ridge. Bars that enter lateral to this ridge frequently strip the muscle and are unstable. Sternal elevation with the introducer prior to bar placement can assist in stretching the chest cage prior to flipping the bar and thus minimize stripping of the muscle. The liberal use of two bars and in rare occasions three bars can dramatically improve the end result and decrease bar shift. We will routinely place two bars in patients who have any of the following factors – a stiff chest, severe torsion, saucer shaped defect, or any patient where we feel the 1 bar is unstable. Our stabilization technique uses multiple PDS (Ethicon, Inc) sutures around the bar and rib on the right and a metal stabilizer on the left secured with a permanent type suture to the bar. If using two bars we alternate sides of the stabilizer. It is satisfactory to use bilateral stabilizers in older patients, but should be avoided in younger patients who are still growing. We restrict our patients for 6 weeks with nothing but activities of daily living and they can then resume all activities and sports at 3 months. However, any major activity that entails repetitive direct blows to the chest such as American football and boxing we instruct them to avoid while the bar is in place.

Concerns with Overcorrection

Overcorrection and development of a carinatum type defect can occur. We have seen a 3.1 % incidence of overcorrection and of those only .3 % developed a true carinatum defect. Factors that put the patient more at risk for this complication are those patients with Marfan's syndrome and those patients with very severe pectus deformities with indexes greater than ten. Although the incidence is still low it is prudent to counsel the patient on this condition preoperatively. Factors that can prevent this are using the appropriate length of the bar and using multiple bars to help distribute the pressure. Although we strongly encourage and recommend exercise and posture programs for all our patients, it is imperative for those patients with a marfanoid body habitus. In our early experience it was discovered that a slight overcorrection was better to minimize recurrence. The use of a carinatum brace can be done for those that develop a true carinatum, but it is usually not tolerated while the Nuss bars are still in place. Our judgment is it is better to leave the bars in place for 2 years and then deal with a carinatum as opposed to a recurrent pectus excavatum, however if a severe overcorrection is developing early bar removal may be considered. It is rare to have to perform a cartilage resection type surgery.

Issues Related with Metal Allergy

Bar allergy in our initial experience was thought to be an infection. However with knowledge of this condition an incidence of close to 3 % was encountered [6]. Metal allergy is a well-recognized condition in the neurologic, orthopedic, and craniofacial literature. In our experience the clinical presentation can vary tremendously if an allergy exists once a bar is placed. Patients can present with persistent pain, fever, rash, erythema, and wound issues with negative cultures. A persistent pleural effusion, pericarditis, and chronic fatigue syndrome may also be the only symptoms that develop. Patients will tend to have an atopic history with eczema, asthma, or history of metal/jewelry reaction.

However, some patients may have absolutely no history and still have a metal allergy. We have begun a routine policy of screening all patients for metal allergy. The TRUE® (Thin layer Rapid Use Epicutaneous) test patch tests for nickel and chromium, and the allergEAZE® adds some of the more minor components (potassium dichromate, copper sulfate, molybdenum, manganese) found in the stainless steel bar. With the practice of routine testing we have recently discovered that 8 % of our patients have a metal allergy (Data in press). If a patient has a metal allergy a custom made titanium bar with titanium stabilizer should be used. It is important to remember to not use a metal wire to secure the stabilizer if the patient has an allergy. If a patient is shown to have a metal allergy or there is a suspicion of an allergic reaction after a stainless steel bar is placed it is not mandatory to remove the bar. We have had success using nonsteroidals and/or low dose prednisone in treating the symptoms. We have only had to remove the bar in three patients who had intractable symptoms with continued wound problems. In those three, only one to date required a bar exchange with a titanium bar.

Wound Infections

Wound infections should also be rare in a clean case such as a pectus repair [7]. Our overall incidence is less than 2 % and most of these have been superficial. Deep infections involving the bar have only occurred in 0.3 % and in these patients we have only had to remove the bar four times. We classify our infections as either superficial (intact skin with evidence of infection or abscess limited to superficial tissues) or deep (exposed bar). If an infection occurs we utilize inflammatory markers such as WBC, ESR, and CRP to guide therapy. We have only had one incidence of MRSA (methicillin resistant staph aureus), and the rest of the infections have been Staph aureus or staph epidermidis. We use antibiotics till the inflammatory markers have normalized. Abscesses are aggressively opened and drained and the use of a wound V.A.C. ® (KCI) may be beneficial. Long term suppressive antibiotics may be necessary and the goal is to maintain the bar for 2 years.

We utilize a variety of measures to prevent infections. The case will be postponed if there is any respiratory infection or superficial skin infection. We have our patients wipe their chests with Sage® (2 % chlorhexidine gluconate) the night before and morning of surgery. A 1st generation cephalosporin is used for 24–48 h for prophylaxis. The use of double gloving may be beneficial to minimize glove tears that can infect the bar during insertion. Aggressive pulmonary toilet and ambulation postoperatively is critical to minimize any pulmonary infections.

Late Hemothorax

We have had only two patients who developed a late hemothorax secondary to trauma. Both underwent thoracoscopy with drainage of the hemothorax. No active bleeding was found, with a presumed etiology therefore being an injury to an intercostal vessel. Whether the hemothoraces would have developed in these patients as a result of their thoracic trauma if they had not had a pectus bar in situ is unknown. We do have several patients who were involved in major automobile accidents who sustained head and musculoskeletal trauma but no chest injuries.

Recurrence

Recurrence after the minimally invasive Pectus repair should be minimal. In our series we have a less than 1 % recurrence risk which we define as deformities of sufficient severity that require reoperation due to significant depression with return of symptoms. The majority of the recurrences were in our first 10 years of experience. Although younger patients tended to have a higher recurrence rate, this was most likely due to early removal of the bar and not their age. We learned that the bar needs to be left in place for a minimum of 2 years. For complicated pectus repairs (syndromic patients, redo repairs) we promote leaving the bar in place for 3 years.

We have an extensive experience with the use of the Nuss technique to repair recurrent pectus excavatum [8]. Of over 110 patients, only 11

have had their primary repair done at our institution. There is a similar distribution of patients who have had recurrences from both the Nuss repair and Ravitch repair. The minimally invasive repair has been useful in both patients who have had a failed previous open as well as minimally invasive correction. There are definitely technical issues associated with both repairs that one needs to be familiar with. Our approach to the preoperative evaluation of the patient with a recurrent pectus excavatum is very similar to that of any patient that presents with a pectus excavatum. However since there definitely is an increased morbidity with redo repairs we feel that all patients considering repair after a failed pectus surgery should have return of preoperative symptoms, along with a higher CT index (HI > 3.7). Every patient needs to be individualized, and some patients may be good surgical candidates that may only have a HI > 3.2.

Patients who have had a previous Ravitch repair will tend to have stiffer chest due to the cartilage resection and may have varying degrees of acquired thoracic chondrodystrophy. Most patients will require two bars and some even three due to the atypical stiffness of the chest. Although the Ravitch repair is extrapleural we have found patients can have significant pulmonary adhesions and may need a chest tube depending on the degree of dissection. It is imperative to counsel the patients that due to the stiffness of the chest from the cartilage resection, they will tend to have a physiologic and cosmetic improvement after a Nuss repair but to a lesser extent than those patients who had undergone a primary Nuss procedure. Rib and sternal fractures may also occur due to the atypically stiff chest from the cartilage resection.

The only patient population that we feel is not appropriate for the Nuss technique is the patient who as a result of the Ravitch procedure has severe debilitating thoracic chondrodystrophy. There will be varying degrees of acquired thoracic chondrodystrophy after a failed Ravitch procedure, however those patients with a frozen chest and PFT's less than 30 % predicted have a higher risk of life threatening vagal events during the procedure as a result of the physiology created from the thoracic chondrodystrophy. The cosmetic and physiologic benefits are negligible after Nuss repair; and this combined with a significant potential morbidity we cannot recommend the Nuss repair for this rare patient population.

There are many modifications with any surgical procedure; however, there are distinct technical issues that we feel make recurrence higher if not adhered to with the Nuss repair. It is imperative that the entry and exit sites into the chest should be medial to the top of the pectus ridge for bar placement. If these sites are lateral there is a tendency for muscle stripping leading to a failed repair. Bending the bar with too much convexity will cause the bar to be too tight against the chest and lead to increased and prolonged pain and discomfort along with muscle erosion. The pectus bar must stay in a minimum of 2 years, and will definitely lead to an increased risk of recurrence if removal is done prior to 2 years.

The patient who has a recurrence after a Nuss repair has a set of problems that are distinct from those patients with a failure after a Ravitch repair. The patient who had a previous Nuss repair may have extensive "toxic" intrathoracic adhesions. One must be prepared with vessel sealing devices to assist in taking down these vascular adhesions to allow safe bar placement. Chest tubes are almost always needed due to this extensive dissection and the increased risk of air leak and pleural effusions. Any mechanism to lift the sternum is beneficial to get across the chest in this previously dissected plane with adhesions. Subjectively we feel that those patients with a failed Nuss procedure will have a better cosmetic and physiologic outcome compared to those patients with a failed Ravitch repair due to the more flexible chest.

Chronic Pain

While most late complications after the Nuss procedure occur with a definable physical finding or symptom complex, there are occasional patients who present months after surgery with a chief complaint of chronic pain. Many of them may have a neuropathic component to their pain, but it is inherent upon the surgeon to ensure that there is no underlying pathologic source.

Consideration should be given to potential causes, with a differential diagnosis including possible bar allergy, bar displacement, stabilizer fracture or displacement, bar contour tightness/pinching of lateral chest wall, excessive bar length, bar erosion into the sternum, chronic low grade infection, and narcotic dependence/withdrawal. The typical work-up should start with physical examination to assess localization of the pain and changes in chest wall appearance, as well as abnormalities on lung auscultation. Chest x-rays will allow for comparison with initial postoperative films and will demonstrate lung infiltrates, pleural effusion, hardware status, and bony changes. Laboratory evaluation to detect evidence of underlying allergy or infection should include CBC, ESR, and CRP. Once this evaluation is complete, identified abnormalities can be addressed, or the patient can be referred to physical therapy or pain management specialists for further treatment.

Complications Related to Pectus Bar Removal

Once permanent chest wall remodeling is deemed complete after having the Nuss bars in place for 2–4 years, the bars should be removed. The procedure to remove these is typically an outpatient procedure and has been performed in 1179 patients in our series through December 2012 with an overall complication rate of 2 % (unpublished data). These complications have included pneumothorax requiring tube thoracostomy (0.4 %), retained residual wire fragments (0.6 %), surgical site infections (0.3 %), and intraoperative hemorrhage (0.1 %). Our current, standard operating procedure for bar removal entails: (1) opening the surgical incisions on both sides of the chest wall, (2) mobilizing both ends of the bar, (3) removing the wire/suture attaching the bar and stabilizer and then disengaging the stabilizer, (4) removing any heterotopic calcification which may be tethering the bar to the underlying rib, (5) straightening the ends of the bar, (6) ensuring the absence of bar tethering in the fibrous capsule, and (7) removing the bar from the patient's right side with a slow, downward pull.

Pneumothorax

Pneumothorax after bar removal likely occurs as the fibrous capsule around the bar is disrupted and air enters the pleural space. This is often not apparent intraoperatively and is typically detected on the immediate postoperative chest x-ray. If a small pneumothorax is present, the patient can be observed in the PACU, and a repeat chest x-ray can be obtained in several hours to ensure the absence of enlargement of the pneumothorax. Large or expanding pneumothoraces require tube thoracostomy. The best chance for preventing this complication is to remove the bar with large positive-pressure breaths held by the anesthesiologist and to avoid spontaneous respiration until the incisions are closed.

Wire Fracture

It is common to find the wire attaching the bar and stabilizer fractured at the time of bar removal due to the routine forces exerted on the chest wall after bar insertion. Efforts should be made to recover all of the wire fragments, if feasible. Intraoperative fluoroscopy can facilitate identification of these fragments. In some instances, these small, 2–3 mm fragments will be embedded in the fibrous capsule that surrounded the bar or in heterotopic calcification. In these circumstances, the fragments are likely best left in situ, as they are unlikely to cause problems, and continued dissection is likely to be bloody and time-consuming. Our practice has recently converted to using Fiberwire for securing the bar and stabilizer, so the incidence of retained wire fragments will eventually stop.

Surgical Site Infections

Surgical site infections after bar removal have typically been preceded by wound seroma or hematoma that has contributed to would disruption. All

cases have resolved with treatment with oral antibiotics. The most effective preventive measure is likely ensuring meticulous hemostasis, especially considering the large residual dead space left in the would after removal of the bar and stabilizer.

Hemorrhage

Hemorrhage encountered during bar removal is a rare but potentially devastating complication. At least one mortality has been reported in the literature [2]. Hemorrhage can occur anywhere along the fibrous capsule and involve abutting structures, such as periosteum, internal mammary vessels, pericardium, or the heart (especially if the initial bar placement was trans-pericardial). In instances when hemorrhage is obvious from the fibrous tract after bar removal, direct pressure over the tract will usually facilitate hemostasis and cessation of hemorrhage. While holding pressure, blood should be typed and crossed for possible transfusion. Persistent bright, brisk hemorrhage or any changes in vital signs mandates urgent cardiac surgery consultation and preparations for urgent thoracotomy or sternotomy. If the patient remains hemodynamically stable but hemorrhage persists, TEE can facilitate identification of a cardiac injury. In some instances where there is an incomplete fibrous capsule around the bar or where capsule disruption occurs with bar removal, hemorrhage may not be apparent externally but will accumulate in the pleural space. In this circumstance, evidence of hemorrhage will be apparent on the postoperative chest x-ray, and the patient may manifest vital sign changes if the hemorrhage is ongoing. The best surgical approach to this scenario would likely be thoracotomy on the affected side to identify the source of hemorrhage. Thoracoscopy could be considered as an alternative, but the likely presence of pulmonary adhesions around the fibrous capsule would preclude prompt or clear visualization of the mediastinal region. Preventive measures incorporated into our bar removal protocol included straightening of the bar ends (to avoid lacerating the heart with the curved bar end) and bar removal from the patient's right side (to minimize the length of bar that passes across the anterior surface of the heart).

Conclusions

Over the last 25 years, we have evaluated over 3200 patients for pectus disorders and have performed the Nuss procedure in over 1600 patients. Lessons learned over this period have led to treatment and prevention strategies for potential surgical complications. Implementation of these techniques has helped to dramatically decrease the morbidity of surgical repair and to ensure the safe placement and removal of the Nuss bar. Short- and long-term complications should be minimal with the proper use of the Nuss repair.

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31 Pleural and Pericardial Associations After Minimal Access Pectus Repair

Christoph Castellani and Amulya K. Saxena

Pleural Effusions

Pleural effusions are among the most common post-operative findings after pectus repairs. The overall the incidence of pleural effusions is reported between 1 and 29 % in the literature with surgical drainage required in 1–8 % (Table 31.1). The etiology possibly involves vascular erosion during surgery or by implant components, infection and allergic reactions.

Etiology

Vascular Erosion

During minimal access repair of Pectus Excavatum (PE) injuries to the intrathoracic vessels is always possibility. Whereas arteries present with an active bleeding which can be observed during the surgery; lacerated veins may be compressed from the intrathoracal pressure achieved by carbon dioxide insufflation that is used for performing video assisted thoracal surgery (VATS). Venous bleeding hence can be obscured and cannot be initially observed and lead to delayed hemothorax within the first days after surgery. In a large series of 1215 pectus excavatum patients that underwent MARPE, a hemothorax incidence of 0.5 % has been reported [1]. In 4 of these patients, a VATS was performed and identified intercostal veins, mediastinal veins or internal thoracic vessels as the source of bleeding [8]. In our own patient cohort, hemothorax was observed in 3.9 % patients – all of which required needle aspiration for evacuation of the hemothorax [5]. It is important to bear in mind that vascular lesions may also occur in the postoperative course and at the time of Pectus bar removal. Furthermore in our cohort, one patient was observed to present with a late occurrence of hematopneumothorax caused by a broken fixation wire – an incident that prompted us to switch from metal wires to PDS® cords (Polydiaxonon, Ethicon GmbH, Norderstedt, Germany) for Pectus bar fixation [5]. Another three patients sustained a thorax trauma also leading to a hemothorax. All four of the above mentioned patients required evacuation of the chest through drains [5]. Vascular lesions may also be encountered at the time of pectus bar removal, although the incidence of such events is low. In another report, a post-operative hemothorax was reported in three of 343 patients (0.9 %); two of who requires the placement of a chest tube and one required open surgical explorations [10]. In has to be noted during that any intra- or postoperative clinical deterioration (loss of blood pressure, respiratory compromise) has to

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Table 31.1 Incidence of pleural effusions (including hemothorax) after Nuss procedure

Author	n	Incidence pleural effusion (%)	Incidence drainage required (%)
Boehm et al. [1]	21	4.8 %	4.8 %
Molik et al. [2]	35	11.4 %	2.9 %
Croitoru et al. [3]	50	Not mentioned	6 %
Engum et al. [4]	20	15 %	5 %
Castellani et al. [5]	180	29.4 %	8.3 %
Shu et al. [6]	406	1.5 %	1.5 %
Zganjer et al. [7]	128	0.8 %	0.8 %
Kelly et al. [8]	1215	Not mentioned	0.8 % (0.5 % hemothorax underwent thoracoscopy)
Hosie et al. [9]	172	1.7 %	Not mentioned

prompt awareness during surgery and thorough clinical examination with chest x-ray during the postoperative course. If an intervention becomes necessary, the preference is to utilize a large caliber drain or chest tube with the intention to prevent obstruction of the drain or chest tube by clots of blood during the course of drainage.

Infection

Pneumonia is an infrequent finding after pectus repairs and the procedures may be responsible for concomitant pleural effusion and pleuritis. Reported incidences for these pathologies range from 0.5 to 4.7 % in the reported literature [7, 8]. Managements for these patients is pursued through administration of antibiotics and physiotherapy. In our series, two patients suffered postoperative pneumonia with concomitant effusion – one of whom was treated by systemic antibiotics, whereas the other required drainage for the effusions. In case a chest tube is placed, it is mandatory to obtain a swab of the effusion for microbiological investigation. In case of bacterial involvement, antibiotic resistograms should dictate the choice of antibiotic and the treatment strategy.

Allergic Reaction

The Pectus bars used for the MARPE (Minimal Access Repair of Pectus Excavatum) are fabricated from stainless steel which contains small amounts of Nickel which might trigger allergic reactions. Nickel as a metal is well known to be responsible for contact allergies in susceptible humans. In 2007, this problem was recognized in context of MARPE and investigations with regards to metal allergies were conducted in 862 pectus patients [11]. In these patients, dermal contact tests were performed and nickel allergy was confirmed in 19 patients (2.8 %). In this group, 63 % patients presented with skin rash and erythema, 32 % with pleural effusion and 5 % with granuloma formation. From this data, a specific pre-operative screening questioning the patients for allergies (jewelry, orthodontic braces, metal buttons on clothing and food) and for signs of atopy (allergic rhinitis, eczema or asthma) was conducted. Any positive finding in this questionnaire prompted allergy screening by skin test and if reaction a positive reaction was documented, a switch from steel to titanium bars was opted. A later series of 1215 patients demonstrated the positive aspects of this screening method [8].

Clinical Presentation and Diagnostics

Clinically, patients with pleural effusions present with (increasing) chest pain, dyspnoea, tachypnoea, orthopnoea and respiratory distress, often requiring oxygen supplementation. Clinical examination of these patients may exhibit decreased thorax movements. Especially, if only one side of the thorax is involved, differences can be observed by percussion and/or auscultation. Percussion shows hyposonic echoes, and auscultation reveals diminished breath sounds at the level of the effusion. These findings should prompt further diagnostics by pleural ultrasound and/or chest films.

Table 31.2 Incidence and treatment of pericardial effusions

Author	n	Incidence pericardial effusion	Treatment
Ong et al. [12]	78	1 pericarditis (1.3 %)	1× pericardiocentesis
Croitoru et al. [3]	50	2 pericarditis (4.0 %)	Both conservative
Castellani et al. [5]	180	4 effusions (2.2 %)	1× pericardiocentesis
Nuss et al. [13]	329	8 pericarditis (2.4 %)	2× pericardiocentesis
Kelly et al. [8]	1215	5 pericarditis (0.5 %)	1× drainage
Hosie et al. [9]	172	1 pericarditis (0.6 %)	Not mentioned

Therapy

In case of respiratory compromise, a single puncture or chest tube placement for drainage is indicated. Ultrasound guided drainage is the preferred approach to reach the target and minimize the chance of injury to the neighboring structures. In case of exudates, recurrent effusions or haematopneumothorax, placement of a chest tube should be considered instead of a single puncture aspiration. Aspirates should be sent for bacteriological examination. If a patient suffers from recurrent pleural effusions or combinations of pleural and pericardial effusions, postpericardiotomy syndrome should be considered which is explained in details in a forthcoming section in this chapter.

Pericardial Morbidity

The incidence of pericardial effusions after MARPE ranges between 0.5 and 4.0 % (Table 31.2). Clinical symptoms involve retrosternal pain, tachypnoea and fever. At auscultation, a pericardial rub maybe encountered during examination of these patients. In case of clinical suspicion of pericardial effusion, the diagnosis should be confirmed by cardiac ultrasound. Even if confirmed, only a small subset of patients reported with pericardial effusions will require pericardiocentesis. The main reason for pericarditis is postpericardiotomy syndrome which has been discussed in the section below. With increasing awareness of this entity, therapy successfully tailored to this condition.

Postpericardiotomy Syndrome

Recurrent pleural or pericardial effusions, fever or chest pain must raise suspicion of a postpericardiotomy syndrome (PPS). Although this condition has been found predominantly to affects patients after cardiac surgery, this has also been reported after pulmonary surgery and surgical procedures on the thoracic wall. Intra-operative exposure of mesothelial cells to the immune system has been discussed to be the possible patho-physiological etiology for a combined humoral-cellular auto-immune response leading to recurrent sterile inflammation.

When the precise mechanism are investigated, PPS patients have been found to often show an increase in anti-sarcolemma-antibodies for heart and skeletal muscle as well as anti-endothelial-cell-, anti-smooth-muscle- and anti-nuclear-antibodies [14]. Another report on this condition has demonstrated elevated circulating anti-actin and anti-myosin antibodies [15]. Also with regards to cellular pathways, elevated CD25 positive T-helper cells and cytotoxic T-cells have been found to be associated in the development of PPS [16]. However, to date, there is no single reliable laboratory parameter to confirm PPS; the diagnosis of which still depends on a combination of clinical symptoms and lab investigations.

In a series of adult patients after cardiac surgery who presented with PPS, the predominant symptoms exhibited were pleuritic chest pain (56 %), fever (54 %) and pericardial rub (32 %). Furthermore, 74 % of these patients had an elevated C-reactive-protein, of which 93 % presented with pleural and 89 % with pericardial effusions [17].

Table 31.3 Therapy regimes and patient data in different cases of PPS after surgical repair of pectus excavatum

Author	n	Age	Int[a]	Effusion	Laboratory[b]	Therapy
Berberich et al. [19]	1	17	6w	Bilateral pleural, pericardial	CRP↑, WBC↑, AHA + VIR neg.	1× drainage, Prednisone, Diclofenac
Boehm et al. [1]	1	14	10d	Not stated	Not stated	NSAR, steroids
Muensterer et al. [21]	1	14	10d	Right pleural, pericardial	CRP↑, WBC↑ VIR neg.	1× drainage, ASS change to methylprednisolone and ibuprofen.
Castellani et al. [5]	2	16 28	8 m 5d	Bilateral pleural, pericardial (recurrent) Bilateral pleural (recurrent)	CRP↑ Rheuma + VIR+ AHA neg. Not stated	Amoxicilline change to steroid 2× drainage, clindamycin + diclofenac change to steroids + diclofenac

[a]Interval between operation and first symptoms
[b]*CRP* C-reactive-protein, *WBC* white blood cell count, *AHA* anti-heart-antibody, *VIR* virology (coxsackie, cytomegaly)

Since the immune response is unspecific and diagnosis also relies on clinical findings, the presence of at least 2 of the below mentioned conditions have been present to raise the suspicion of postpericardiotomy syndrome [18]:

- Fever without alternative causes
- Pleuritic chest pain
- Friction rub
- Evidence of new or worsening pleural effusion
- Evidence of new or worsening pericardial effusion

Routine laboratory investigations should involves white blood cell count, C-reactive protein and erythrocyte sedimentation rate in the work up to suspected PPS. Besides these, viral infections especially Cytomegalovirus and Coxsackie virus should be investigated and ruled out. Anti-heart-antibodies (AHA) can be determined but have no influence for the further treatment. Bacterial infections should be ruled out by microbiological examinations of the fluids aspirated.

Cases of pericarditis responding to Indometacin or acetyl-salicylic acid have been reported after surgical repair of pectus excavatum [3, 13]. Proven PPS however is limited to case reports [1, 19–21]. Symptoms described by these reports were similar to those reported after cardiac surgery. In all cases, recurrent effusions combined with clinical symptoms and lack of other causes lead to the diagnosis of PPS. Treatment has consisted of drainage in case of acute respiratory compromise, non-steroidal anti-inflammatory drugs (NSAID), steroids (prednisolone 2 mg/kg/day) and in some cases antibiotics. Colchicine, which is proposed in the management of adult PPS has possible severe side effects and has not yet been administered and/or reported in PPS after pectus repairs. Colchicine administration is recommended at onset of clinical symptoms for a period of 14 days [22] with slow reduction in the administration of the amount of steroids over another 14 days [23]. Neither prevention nor modification of the clinical course is possible by prophylactic steroids [19]. In case of recurrence, a longer steroid course of 4–6 weeks is recommended [19]. Therapy regimens as reported by various centers are summarized in Table 31.3.

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32 Management of Postoperative Infections Following the Minimally Access Repair for Pectus Excavatum

Sohail R. Shah and George W. Holcomb III

Since the first description of the minimal access technique for repair of pectus excavatum (MARPE) in 1998 [1], the procedure has gained widespread acceptance with minor modifications to decrease the incidence of complications. One such complication, that is inherent with the implantation of any prosthesis, is the potential for infection. Although reported infection rates are low, ranging from 1.4 to 6.8 % [2–5], the management of infectious complications can be complex. A significant concern is the possible need to remove the substernal bar prematurely, as early bar removal could lead to recurrence of the defect. Fortunately, the reported rate of bar removal secondary to infection is low and ranges from 0.3 to 1.6 % [2–7]. In order to continue to decrease the incidence of infectious complications, surgeons need to have a good understanding of prevention and treatment strategies. Infectious complications after MARPE can be divided into two primary types: primary pulmonary infections, and skin and soft tissue infections. The pulmonary infections include infected pleural effusions, pneumonia, and empyema. These postoperative infections occur in less than 1 % of patients [3], but can lead to significant sequelae including mediastinitis, pericarditis, and bar infections. The best preventive measures for primary pulmonary infections include adherence to sterile technique, aggressive pulmonary toilet, good postoperative pain control, and the use of perioperative prophylactic antibiotics. In comparison, skin and soft tissue infections are more common and often more challenging to diagnose and manage. To better understand skin and soft tissue infections after MARPE, we will focus on perioperative prevention strategies, diagnosis, and treatment recommendations.

Perioperative Strategies for Decreasing Skin and Soft Tissue Infection

As with any operative procedure, the prevention of postoperative skin and soft tissue infections begins with the preoperative management strategy. Such strategies are especially important with the implantation of a prosthesis, such as a pectus bar. During the preoperative evaluation, any upper respiratory or skin infection must be evaluated and treated prior to proceeding with the pectus repair. Additionally, if the patient has acne over the thorax, this should be further evaluated and treated by a dermatologist. All patients should also receive a preoperative antibiotic (i.e., cefazolin) for

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prophylaxis. The prophylactic antibiotics are continued postoperatively for the first 24 h

During surgery, there must be adherence to meticulous sterile technique. We recommend a strict surgical scrub, use of a "double-gloving" technique, and appropriate surgical prep with the addition of an iodine impregnated drape covering the exposed thoracic cavity (Fig. 32.1) [6].

Evaluation and Management of Postoperative Surgical Site Infection

After MARPE, skin and soft tissue infections can have extremely variable presentations. These presenting symptoms can become evident within 24–48 h or as late as 1–2 years after bar placement. Initial symptoms may include erythema, warmth, fevers, drainage, and/or chest pain, especially over the bar. The first step in management is to determine the extent of the surgical site infection (SSI), which will often dictate treatment.

Shin et al. previously classified postoperative wound infections after MARPE into three categories: cellulitis, stitch abscess, or bar infections. Cellulitis was described as erythema and warmth without induration, fluctuance, or drainage. A stitch abscess was described as a superficial abscess without extension to the bar, and a bar infection was described as an abscess in direct contact with the bar and having associated purulent drainage [4]. While these categories are useful for comparative discussion, one must remember that manifestations of these SSIs can present anywhere along this spectrum and not necessarily within a discrete category.

As reported in the literature, and from our own experience, most of these skin and soft tissue infections can be managed conservatively [3–6]. If a patient presents with cellulitis alone, he/she can be placed on oral antibiotics and followed closely as an outpatient. If the oral antibiotics are unsuccessful in clearing the cellulitis, or if there is progression to an abscess, then hospital admission is recommended with initiation of intravenous antibiotics (Fig. 32.2). Initial antibiotic choices should cover standard skin infections based on local infectious disease experience. Our initial choices are usually cefazolin or vancomycin. Any suspicion for an abscess should prompt further evaluation with ultrasound and, if needed, drainage with collection of wound cultures. For a simple abscess in a clinically-well patient, percutaneous drainage of the abscess may be considered with fluid samples sent for Gram stain and culture. Antibiotic coverage can then be tailored to specific isolates. The most common reported species are *Staphylococcus aureus* and *epidermidis* [4–6, 8].

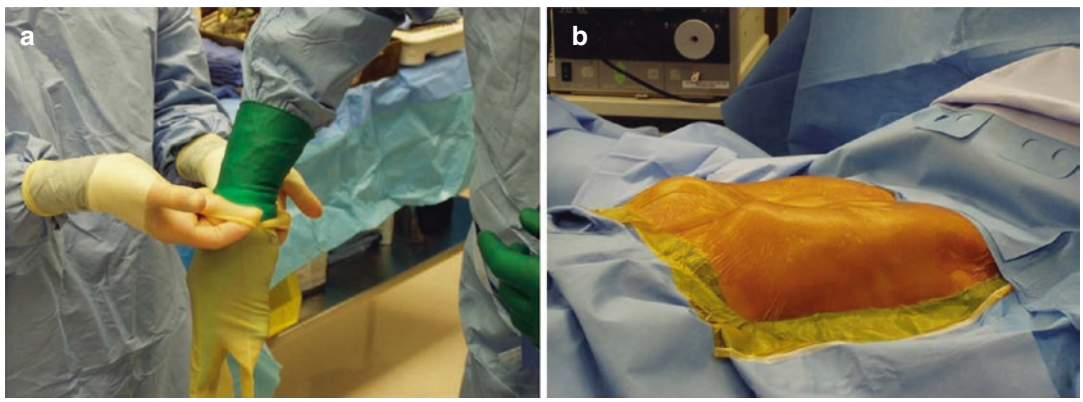

Fig. 32.1 In order to prevent postoperative infections following the minimally invasive pectus excavatum repair, adherence to sterile technique is important. In addition, we utilize a "double-gloving" technique (**a**). If there is a break in the outer glove, then, hopefully, the inner glove will help prevent the development of an infection. Also, following surgical prep, we utilize an iodine impregnated drape to cover the exposed thoracic cavity (**b**)

If there is persistent wound drainage or progression of the infection on appropriate intravenous antibiotics after abscess drainage, then further debridement may be necessary (Fig. 32.3). With persistence of an abscess over a lateral chest wall stabilizer, the stabilizer may need to be removed. If the lateral stabilizer is removed due to infection within the subcutaneous pocket, the soft tissue can be re-approximated over the retained bar while the skin is left open and treated with wet-to-dry dressing changes.

Once intravenous antibiotics are initiated, they are continued for at least 14 days or until the infection clears. If clinical improvement is noted, we recommend placement of a peripherally inserted central venous catheter (PICC) and completion of intravenous antibiotic therapy at home. In some cases, we have continued parenteral therapy for 3–6 weeks.

Upon completion of the parenteral antibiotic therapy, patients are started on prolonged oral suppressive antibiotics. The duration of oral suppressive antibiotics is not well defined in the literature, but has been as long as 6–12 months in our experience. Long-term suppressive antibiotics may allow the bar to remain in place until appropriate correction of the defect is achieved [4–6].

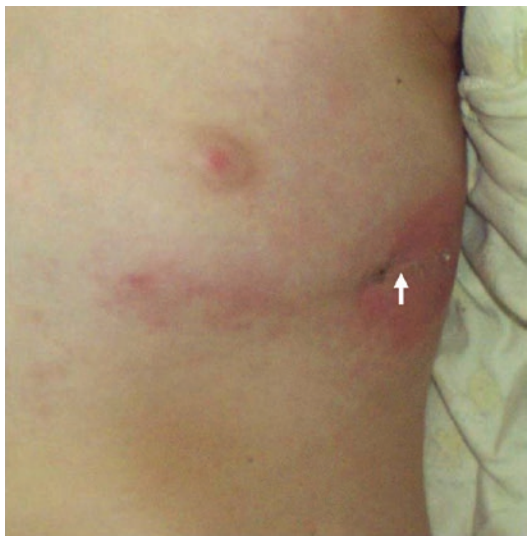

Fig. 32.2 This patient has developed cellulitis around the left lateral incision (*arrow*). Erythema is noted to be tracking along the bar towards the midline of the chest. This patient responded well to intravenous antibiotics followed by oral antibiotic suppression and did not require removal of his bar

If there is persistent wound infection with drainage, or clinical deterioration with these previously discussed measures, then bar removal may be unavoidable. However, bar removal should be reserved for multiple recurrent infections or complicated infections that do not respond to conservative measures or removal of the lateral chest wall stabilizer.

Special Considerations in Minimally Invasive Pectus Repair

Metal Allergies

In the treatment of postoperative infections after MARPE, physicians must be prepared to address nonspecific symptoms that make the diagnosis difficult. One such confounding diagnosis is a metal allergy. In the United States and Europe, metal allergies, specifically nickel, are the most common contact allergy. Common symptoms include erythema and rash.

In the orthopedic literature, as many as 13 % of patients undergoing prosthesis implantation have a metal allergy [9]. When this was evaluated in patients undergoing pectus repair, Rushing et al. found that 2.2 % of patients were sensitive to nickel, cobalt, or chromium [10]. Therefore, nonspecific symptoms of erythema or rash over the standard stainless steel pectus bar that do not respond to traditional therapy with antibiotics may warrant evaluation for a metal allergy [10, 11].

Additionally, we now screen all patients preoperatively for possible metal allergies. We recommend that any patient with a metal or food allergy, or atopy (eczema, asthma, or allergic rhinitis), should undergo a dermal patch test for evaluation of a metal allergy. If a metal allergy is identified preoperatively, a custom-made titanium bar, which is manufactured based on specific computed tomography scan chest measurements, should be used.

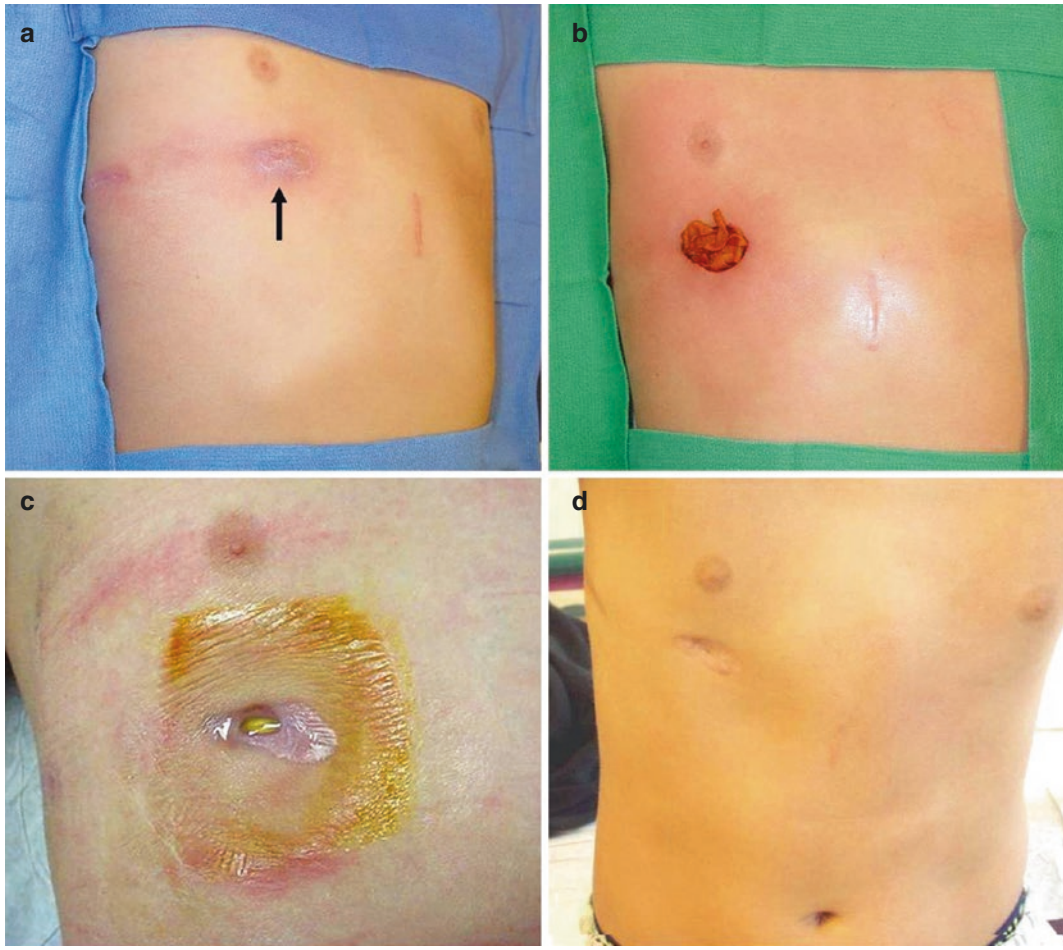

Fig. 32.3 This patient developed an abscess (*arrow*) overlying the right aspect of the subcutaneous track for the bar (**a**). He had developed mild trauma to the area which may have precipitated the development of the abscess. The infection did not respond to intravenous antibiotic management so the area was opened and packed (**b**). Several months later, the cavity is continuing to close by secondary intention and was being packed at home. Note the bar can be seen through the open incision (**c**). Finally, the open wound has closed and the infection has resolved (**d**). The appearance of the chest wall is approximately 6 months following development of the infection. He recovered uneventfully and underwent bar removal at the usual 3 year time period following its insertion

The Use of Lateral Stabilizers

In our practice we routinely place bilateral chest wall stabilizers to prevent displacement of the pectus bar. However, there are reports that suggest that lateral stabilizers may increase the incidence of wound infections [12]. In a previous report of our institutional experience with postoperative wound infections after MARPE, we were successful in avoiding bar removal in all cases that presented with infection over the hardware with the conservative management strategies described [6]. Therefore, we believe the routine use of lateral stabilizers is important as the small risk of wound infection is outweighed by the risk of bar migration or slippage.

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Recurrent Pectus Excavatum Repair

33

Michele L. Lombardo and Donald Nuss

Introduction

Although Bauhinus described a classic case of pectus excavatum in 1595 [1] and isolated cases were done in the first part of the twentieth century, it was not until after the Second World War when Ravitch wrote his seminal paper in 1949 on radical resection of all the cartilages with complete isolation of the sternum (in order to avoid using external traction) that pectus excavatum repair became commonly accepted as a treatment for this condition [2]. However, despite Ravitch's claim to have almost no recurrences [3], other surgeons such as Wallgren and Sulamaa introduced the concept of internal support to prevent recurrence by passing a support bar through the sternum in 1956 [4], followed shortly thereafter by Adkins and Blades in 1961 with the proposal of placing the bar behind the sternum [5]. There were few long-term studies published during the era of extensive open resection; those that were published had a recurrence rate that varied from 1 to 22 % [2, 6–8]. The pediatricians who saw the patients on a regular basis were not impressed and Sidney Gellis stated in his Weekly Pediatric Notes "that it has been evident for a long time that far too many children have had surgery for pectus excavatum" [9]. In 1990, Pena drew attention to the danger of recurrence and asphyxiating thoracic chondrodystrophy developing in young children after wide resection. He wrote, "it appears necessary to develop alternative techniques that avoid the removal of costal cartilages and to re-evaluate the optimal age for repair of these malformations" [10]. This concern was further reiterated by Haller and Columbani et al in a paper entitled "Chest Wall Constriction After Too Extensive and Too Early Operations For Pectus Excavatum." They concluded that "these extensive procedures have removed or prevented growth center activity, which resulted in restriction of chest wall growth" [6]. These findings were recently corroborated by Chen et al [8]. As a result, the open technique was modified to a minimal cartilage resection technique and is now performed in much older patients with an improved recurrence and reoperation rate of only 2.4 % [11].

In 1998, Nuss and Kelly et al published their ten year experience using a novel new concept of excavatum repair, which required no cartilage or sternal resection. The closed technique relies on the flexibility of the anterior chest wall and is effected by placing a pre-bent retrosternal bar through bilateral thoracic incisions. The bar is inserted into a transthoracic tunnel with the concavity facing posteriorly and when it is in

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position it is turned over, thereby pushing the anterior chest wall outward and correcting the pectus excavatum [12]. Initial long term results were affected by the "early learning curve," during which time it was determined which materials were of sufficient strength to support the anterior chest wall in its corrected position, how long the support bar needed to stay in place, how to stabilize the bar and how many bars were needed to give excellent long term correction. Despite the learning curve, Kelly et al reported a recurrence rate requiring re-do repair over the first twenty one years of only 1.4 % of the patients [13].

Causes of Recurrence

The causes of recurrence vary according to the different procedures. In general, immediate recurrence in both the open or closed techniques is a result of a technical error, while rapid growth during puberty can account for later recurrences.

Recurrence after the open procedure depends on whether a sub-sternal support bar was used, how long it was left in place, the age of the patient at the time of repair, how well the excavatum was corrected at the time of surgery, and whether there was excessive bleeding or post operative infection. Adkins and Blades were the first to place a substernal bar after the open procedure and recommended a smooth steel strut which they left in place for eight to twelve weeks, although now most surgeons would leave it in place for a minimum of six months [5]. If it is removed too soon recurrence is more likely. A wide resection in prepubertal patients, especially very young patients, has a markedly increased risk of interfering with the growth centers. In turn, this can lead to recurrence with a constricted and rigid anterior chest wall. Under correction will set the patient up for recurrence since all the forces present before surgery still exist; as the patient goes through a growth spurt, a minor under correction may become quite severe. Postoperative complications, such as hematoma formation and infection, may not only damage the repair but lead to excessive fibrosis and calcification.

Recurrence after the minimally invasive or closed procedure is due to early bar removal, bar displacement, incomplete or under correction, lack of regular exercise or rapid growth. Long-term studies have shown that the bar should remain in situ for at least two years after repair and preferably for three years (see Fig. 33.1). Occasionally, it is necessary to remove the bar (s) early because of infection, allergy, or trauma and this leads to a higher risk of recurrence if the bar is removed before two years. Bar displacement usually occurs within the first month or two of the postoperative period. This almost always results in immediate recurrence and is due to inadequate stabilization of the bar (s) at the time of repair. Incomplete or under correction is a technical error which leads to recurrence as the patient grows. This occurs if the bar is not properly placed under the deepest point of the depression, if the bar is placed too far laterally, if the bar is not bent sufficiently, or if two or more bars were needed and only one was placed. All patients should do daily deep breathing and breath holding exercises to expand the chest wall, open up the bony portion of the rib cage and help stretch the chest wall ligaments. Failure to do these chest wall exercises in sedentary and young patients who have not yet gone through puberty may result in recurrence during rapid growth spurts. Prepubertal patients have a slightly higher risk of recurrence when they go through a rapid growth spurt; however, the risk is not as high as was initially expected (Fig. 33.2). This younger age group should especially be encouraged to participate in aerobic activities that include running, dancing or swimming, as well as doing the aforementioned breathing exercises.

Criteria for Surgical Correction

The criteria for surgical correction are similar to those of the primary repairs but require additional evaluation to assess rigidity of the anterior chest wall. Determination of a severe pectus excavatum

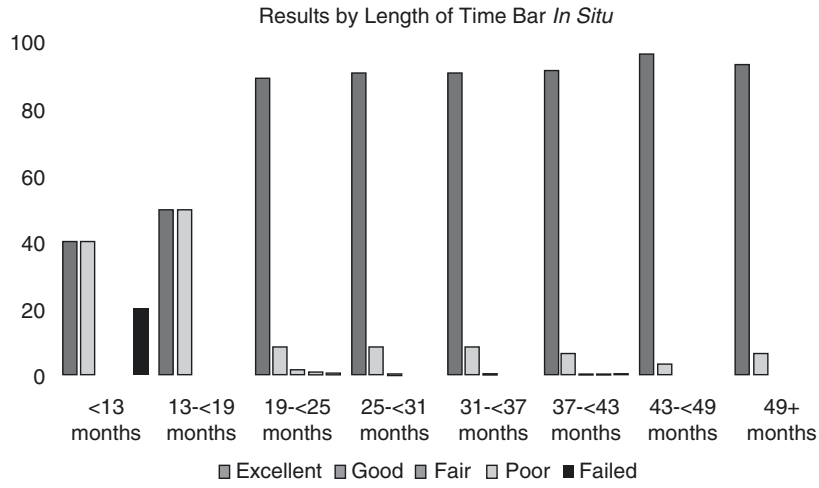

Fig. 33.1 Duration of pectus bar placement

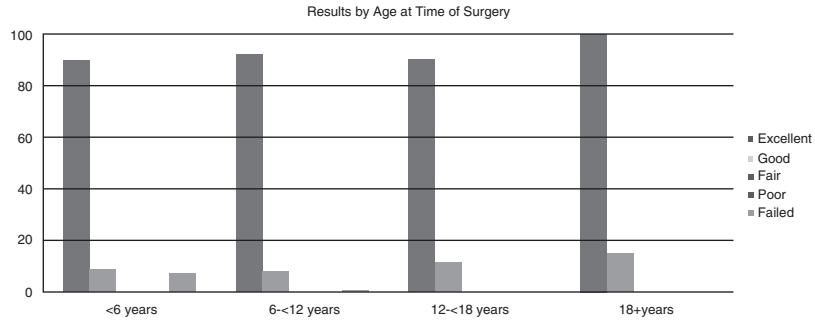

Fig. 33.2 Age at time of pectus excavatum repair

and the need for primary repair includes two or more of the following:

- Documentation of progression of the deformity with associated symptoms.
- A CT index greater than 3.25 showing cardiac compression and or displacement.
- Pulmonary function studies that indicate restrictive or obstructive airway disease.
- Cardiac compression or displacement, mitral valve prolapse, right bundle branch block, murmurs.

When evaluating a recurrence, it is not only important to decide whether the patient fulfills the aforementioned criteria, but in the case of a re-do procedure it is even more important to assess the patient with regard to the type of procedure that will be needed to effect the repair. Patients with simple recurrence and residual flexibility after either the open or closed technique do well with a closed repair. Whereas, a patient with a complex recurrence, as in the case of patients with chondrodystrophy or a rigid anterior chest wall, are definitely not candidates for closed repair, since placing a bar under a rigid chest wall will not serve to achieve repair. In fact, it is dangerous to attempt a closed re-do repair on a patient who has a severe recurrent pectus excavatum with cardiac compression, adhesions and a rigid anterior chest wall. In these cases, one may not only perforate the heart during dissection, but the pressure on the heart, when passing the introducer under the rigid anterior chest wall, may cause cardiac arrest. Patients with a severe recurrence and a rigid anterior chest wall are very difficult to repair even by the open technique and may require a combined approach consisting of transection of the calcified bony portion of the ribs and placement of substernal bar support. Various techniques have been described for the management of patients with asphyxiating osteochondrodystrophy with limited success. [6, 8, 14].

Types of Recurrences and Selection of re-do Technique

The recurrences should therefore be divided into two groups: simple recurrence and complex recurrence [15]. In a simple recurrence the sternum has simply sunken back into the thoracic cavity but there is still some flexibility of the anterior chest wall with respiration or on palpation, the surface of the anterior chest wall is still smooth and the deformity is relatively symmetric. Conversely, in a complex recurrence the anterior chest wall is corrugated, asymmetric and completely rigid. These patients exhibit minimal chest movement on deep inspiration but have significant abdominal movement, as they rely almost entirely on the diaphragm for respiration [16]. If the repair was done at an early age there may also be overall chest wall growth failure; these patients fall into the category of acquired asphyxiating thoracic chondrodystrophy as mentioned above.

In addition to assessing the external characteristics of the chest wall, it is important to take into account the fact that all recurrent pectus excavatum patients will suffer from pulmonary, pericardial and mediastinal adhesions, which will make the second operation much more challenging. These adhesions will vary from mild to very severe, depending on the number and type of previous procedures, post-operative complications such as infection, hemothorax, bar displacement, and the time elapsed since the last thoracic operation.

If the patient has had only one previous thoracic procedure and especially if more than six months have elapsed, adhesions may be relatively easy to take down by using a thoracoscopic approach. However, if the patient has had multiple previous procedures or open heart surgery, it may be safer to use the open technique. Patients should be warned that the re-do operation is more challenging and that thoracoscopy may be required to determine whether the closed or open technique will be best suited for that particular patient. The majority of the cardiac perforations reported in the literature have occurred in patients who have had previous pectus excavatum repair or cardiac surgery [17].

We have found that recurrence after an open procedure can be simple, as described above, but usually the recurrences are more complex. The ossification that occurs in the cartilage bed after cartilage resection leads to an unusually stiff chest wall and may result in laceration of the intercostal muscles at the time of tunnel creation or bar rotation, lack of visibility due to adhesions, and of course, increased risk of cardiac and pulmonary injury. If the primary repair was done at an early age (less than five years) and more than four cartilage pairs were resected, acquired thoracic chondrodystrophy may also be present [6]. Such patients have abdominal breathing patterns and exhibit a severe restrictive pattern on pulmonary function testing.

Patients with connective tissue disorders are more likely to have recurrence, irrespective of the type of initial repair. Additionally, since the pectus excavatum tends to be more severe at an earlier age in these patients, they also have undergone repair at an early age, further complicating the issue [18].

Pre-operative Evaluation of the Patient with Recurrence

The standard preoperative workup for primary pectus excavatum repair should be completed for recurrent cases. This includes a thorough history and physical examination to determine whether the patient is symptomatic and whether the deformity is progressing, CT scan, pulmonary function studies and full cardiology evaluation including echocardiogram and ECG (Fig. 33.3). If the patient fulfills the criteria for pectus repair, allergy testing should also be discussed.

In preparation for surgery, patients and their families should be counseled of the risks and challenges inherent in reoperative surgery. We counsel patients that operative time, risk of pneumothorax or hemothorax, and risk of cardiac and pulmonary injury are all increased in comparison to primary pectus excavatum repair because of pulmonary and mediastinal adhesions, chest wall rigidity and thoracic chondrodystrophy, if present. Patients are informed that a chest tube will

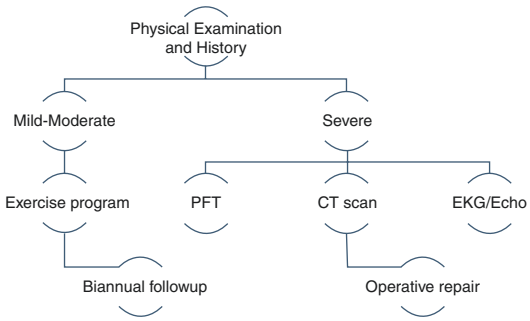

Fig. 33.3 Patient selection and preoperative workup for pectus excavatum repair

likely be placed at the end of the procedure since taking down pulmonary adhesions is likely to lead to pleural surface oozing and air leakage. Additionally, patients should be aware that the cosmetic results are less favorable when compared to outcomes from primary repairs.

Technical Considerations, Risks and Challenges in Reoperative Pectus Surgery

In our experience, the minimally invasive approach can be utilized for the majority of patients with recurrent pectus excavatum, irrespective of initial approach [19, 20]. However, because reoperations are much more challenging than the primary procedures, we do not recommend that surgeons undertake the re-do repair until they have considerable experience with the primary closed repair. The contraindications to minimally invasive reoperation include acquired asphyxiating thoracic chondrodystrophy, a rigid anterior chest wall (especially if there is associated severe cardiac compression) and a very high or superiorly placed deformity (Figs. 33.4 and 33.5). In these patients, there is no ability to increase thoracic domain by conventional methods for pectus excavatum repair.

There are no specific recommendations regarding timing of redo pectus operation. In general, reoperative repair is indicated in symptomatic patients with severe pectus excavatum, following similar guidelines to the criteria for initial repair.

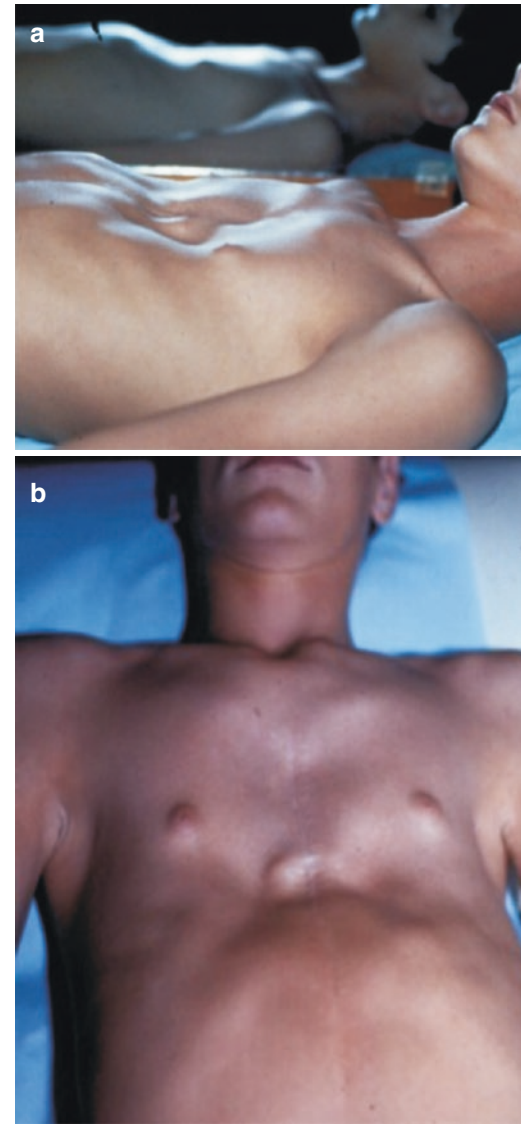

Fig. 33.4 Acquired thoracic chondrodystrophy. This young adult underwent open repair of pectus excavatum at 5 years of age. Note decreased thoracic domain, rigid and corregated chest wall. This patient would not be a candidate for re-do repair by the closed technique

A median sternotomy instrument tray should be available and the cardiac surgeons should be placed on alert that they could be needed in case of a cardiac perforation. Type-specific blood should be readily available. Operative preparation should include large bore intravenous access, arterial blood pressure monitoring, bladder catheter for measurement of urine output and nasogastric decompression.

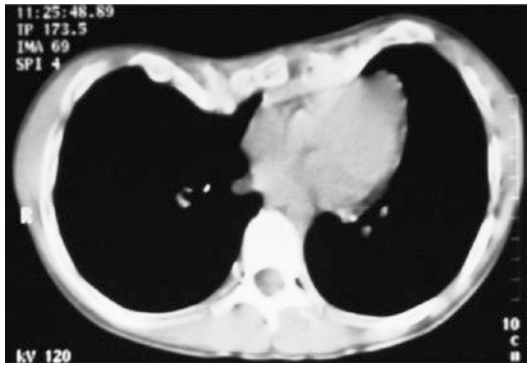

Fig. 33.5 Osteochondrodystrophy. Osteochondrodystrophy after primary open repair. Note complete ossification of costal cartilages

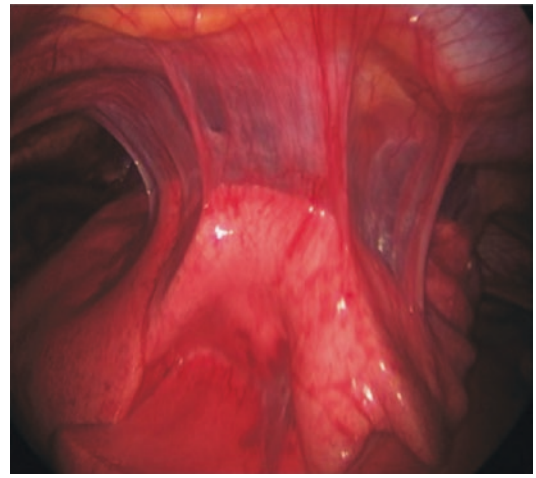

Fig. 33.6 Pulmonary adhesions. Significant vascular pulmonary adhesions after primary closed repair

Significant intrathoracic and mediastinal adhesions should be anticipated in re-operative situations. Dense and sometimes vascular adhesions can be encountered in patients who had previous open or minimally invasive initial repairs (Fig. 33.6). Adhesiolysis can be associated with significant blood loss; however, in our experience no patients have required transfusion. We perform adhesiolysis with endoscopic shears or the Harmonic scalpel (Ethicon, Somerville, NJ, USA). In some cases, bilateral thoracoscopy may be helpful. A small air leak that resolves in one to three days is to be expected after pulmonary adhesiolysis and requires chest tube placement at the end of the repair to prevent pneumothorax, pleural effusion and hemothorax. Chest tube placement does not significantly alter the time to discharge and in fact may even shorten the length of stay.

Sternal elevation should be used in patients with deep excavatum deformities to minimize the risk of cardiac and pulmonary injury. Furthermore, sternal elevation decreases the risk of intercostal muscle laceration. This can be accomplished using a variety of techniques including the vacuum bell, sutures through the sternum placed on anterior traction, retractors placed either through the thoracostomy wounds or through an inferior sternal incision [21–23].

Intercostal muscle laceration is more common in patients who have had previous pectus excavatum repair due to the rigidity of the chest wall. Therefore, it is very important to make the thoracostomy sites more medial than for primary repairs and to take care to avoid posterior pressure on the intercostal space when during substernal tunnel creation and bar rotation. After successful introducer placement and prior to insertion of the bar, it is essential to elevate the anterior chest wall numerous times in order to loosen up the anterior chest wall, thereby decreasing the amount of pressure that will be exerted on the bar. If the chest wall is too rigid and does not respond to the elevation maneuver, one should give strong consideration to conversion to the open or combined technique.

Re-do patients more frequently require two or more bars for proper sternal elevation and to obtain a satisfactory result. There is a slightly increased rate of bar dislocation in re-do cases and therefore it is essential that the bars are properly stabilized at the end of the repair.

Results

The vast majority of repairs for recurrent pectus excavatum at our institution have utilized the closed technique. Surgeon-grading of postoperative result is somewhat less favorable than for primary procedures, yielding an excellent result in 62.5 % of re-do patients after primary open repair and 74.5 % of re-do patients after primary closed repair, in comparison to 85.3 % after a pri-

mary closed repair [13, 20]. In patients who have undergone a primary open repair, this may be due to chest wall stiffness associated with chondrodystrophy. For the same reason, bar displacement is more common in re-do patients after a primary open repair, occurring in 12 % of patients versus 7.8 % after primary closed repair (for comparison, 3.4 % of patients suffer bar displacement after primary repair) [13, 20].

Cardiac complications occur with a higher frequency in reoperative cases. We have reported non-fatal arrhythmias and arrests [20]. In our two cases of cardiac arrest, intra-operative echocardiogram revealed right ventricular dysfunction that was not appreciated on preoperative echocardiography. The ventricular dysfunction is thought to have increased susceptibility to vagal stimuli from hypovolemia and anesthesia. Anatomic cardiac injuries have been reported by others; some have been fatal [17].

Conclusion

Recurrent pectus excavatum is more challenging to repair than primary pectus excavatum and requires significant knowledge of and experience with the advantages and disadvantages of the various techniques available. There is an increased risk of cardiac and pulmonary injury because of intrathoracic adhesions and a much less flexible chest wall. Patients must be carefully evaluated preoperatively to determine not only their fitness for surgical repair, but also with a view to selecting which technique will be most beneficial for the individual patient. Preoperative counseling and preparation are crucial in this setting. In experienced hands, the majority of patients can be successfully repaired with either the open or closed techniques.

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Techniques and Instruments for Pectus Bar Removal

34

Amulya K. Saxena

Introduction

Both open repair and minimal access surgical procedures employ metal struts or bars for the correction of Chest wall deformities (CWDs). The metal of choice for these procedures is stainless steel, although titanium is also used, its application has been reserved for patients with known allergy to Nickel which is a component in stainless steel [1]. The metal bars and struts for CWD repairs are not permanent implants and are removed based on the type of repair for which they were originally conceived. In open repairs which involve resection and anastomosis of the cartilage and remodeling of the sternum, the implants are removed after a period of 15 months. On the other hand, the minimal access repair which applies the concept of chest wall remodeling using pressure, correction of the deformities is best achieved when the pectus bar is left in position for a period of 3 years. There are large differences between the removal techniques of pectus struts for open repairs and pectus bars for minimal access repair, both of which will be highlighted in this chapter. Since the removal of the pectus bar after minimal access repairs still remains an area of concern, this chapter will also focus on the published techniques for pectus bar removal as well as instruments developed to aid pectus bar removal.

Pectus Strut Removal After Open Repairs

In open repairs, such as the PLIER technique, metal struts are passed through the sternum and the metal does not enter the thoracic cavity [2]. Also, in this technique, when additional metal struts are employed, for example when a severe combined deformity or a platythorax is corrected, these struts are placed on the thoracic wall with sutures used to secure the ribs to these struts. Removal of these struts after 15 months of open repairs, involves the opening of the previous incision and extraction of the metal implants without entering the thoracic cavity. Since these struts, do not have contact with intra-thoracic organs, complications relating to intra-thoracic organ lesions, such as injury or erosion into major cardiovascular structures, have not been reported in large series using similar techniques during the removal of the metal struts [3–5].

Removal of metal struts after correction of CWDs using the PLIER technique requires incision of the prior surgical incision scar and exposure of the metal struts. Two instruments that are necessary for the removal of the metal struts are the pectus spanners and pectus two-tooth pliers

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(Figs. 34.1, 34.2, and 34.3). Once the metal is exposed, a rasp is inserted in the plane between the metal and the chest wall to create a space to position the pectus spanners. The pectus spanners are placed on the strut approximately at a distance of 2 cm from each other and are moved towards and away from other until the in between them strut are broken. The pectus two-tooth pliers are then used to grasp the perforation on the edge of the broken metal strut, which are then removed in parts. A hammer is used to exert force on the pectus two-tooth pliers to aid dislodgement and removal of the strut placed through the sternum. In case of bone formation on the other metal struts, if employed, hammer taps on the pectus two-tooth pliers is again used to aid in removal of these struts.

Varying degrees of reaction to the metal strut can be observed as capsule formation around the implanted metal at the time of removal. If the capsule formed along the metal strut is soft tissue scar, it is leveled with a scalpel; however, in case of bone formation, a chisel is used to remove the hardened bony tissue to level the site of the implants. In order to decrease the possibility of seroma formations, 2 subcutaneous drains are placed which are removed on the 3rd day after surgery.

Fig. 34.1 Pectus spanners that are employed to break the metal struts at the time of pectus strut removal

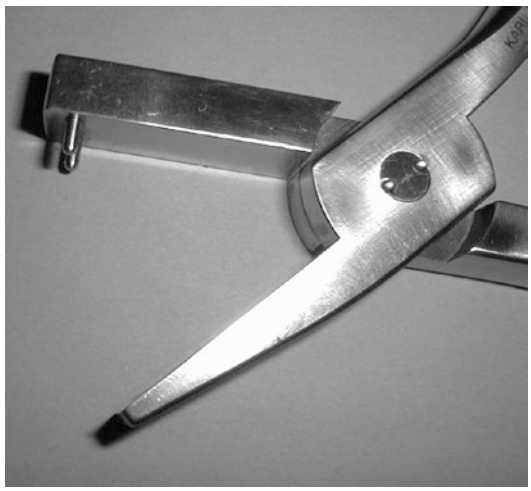

Fig. 34.2 Pectus two-tooth pliers demonstrating the construction of the instrument at the tip

Pectus Bar Removal After Minimal Access Repair

In the minimal access repair of pectus excavatum, the pectus bar is positioned below the sternum within the thorax. The placement of the pectus bar below the sternum brings it into close proximity of intra-thoracic structures, cardiovascular and pulmonary, within the anterior mediastinum. The effects of the pectus bar which comes into contact

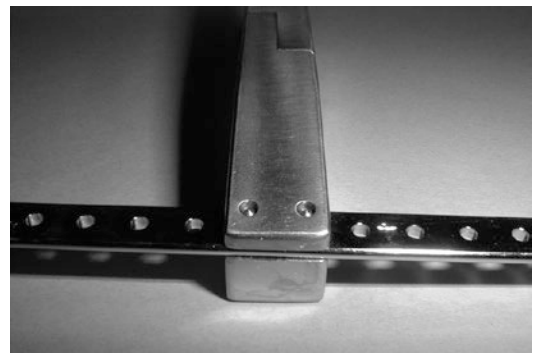

Fig. 34.3 View of the pectus two-tooth pliers clutching the perforation of the metal strut

with the lung that is in the state of constant motion, could lead to formation of effusions; but erosion associated complications of the lung associated with the pectus bar or its removal have not been reported. However, unlike the lungs, major cardiovascular structures that are in direct contact with the implanted pectus bar could be directly affected, with erosion of these structures that have manifested themselves with severe hemorrhage at the time of pectus bar removal [6–8]. In a large series on pectus bar removal in 343 patients after minimal access repairs from Aarhus, Denmark, eight patients (2.4 %) had complications after the surgery [9]. Whereas, five patients had pneumothorax, three had a hemothorax with one of these patients requiring open surgery.

Beside the possibilities of pectus bar complications due to their close proximity to the major cardiovascular structures, attention is also being focused now towards the role of implanted pectus bars on the internal thoracic vessels. Reports are emerging on the alteration of blood flow in internal thoracic vessels being as high as 44 % in patients who have had minimal access repairs [10]. The pressure exerted on these vessels could lead to occlusion and thrombosis or even erosion. Erosion of these vessels and severe hemorrhage in a patient at the time of pectus bar removal was observed in 1 patient at our Center; with this patient requiring emergency open surgery to manage the bleeding (unpublished data).

A. **Techniques for pectus bar removal**
Pectus bar have perforations on their edges and the basic principal in their removal involves locking their edge with either a bone hook, Kocher–Ochsner clamps, bar crank or even a towel clip and pulling them out through the chest, bearing in mind the natural curvature of the bar to prevent injury to the thoracic organs (Fig. 34.4). With this principle in mind, the following 5 technical options are known for pectus bar removal:

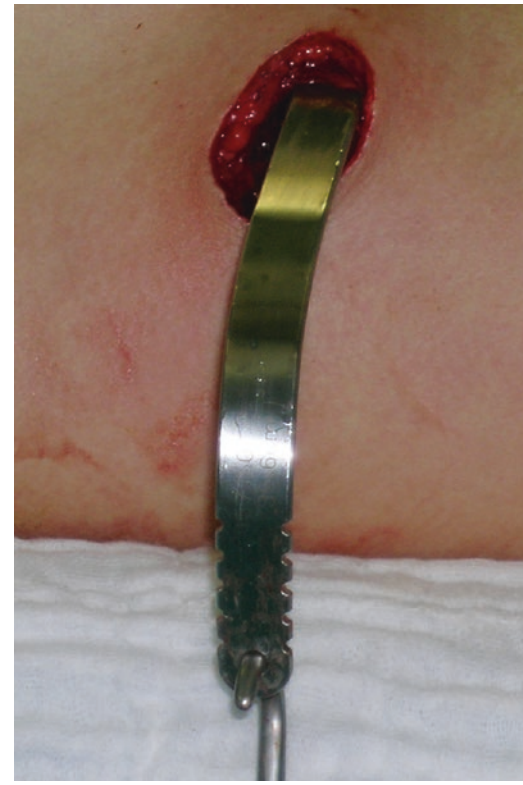

Fig. 34.4 Operative view showing the bone hook locking the pectus bar to aid pectus bar removal

1. ***Bar straightening technique***
In this technique, the prior skin incisions are opened on both side of the chest. After exposing the pectus bar, bar benders are inserted subcutaneously into the wound at both ends of the bar. The pectus bar is then immobilized and straightened [11]. The straightened bar is then removed (Fig. 34.5).
2. ***Split operation table or pull down technique***
In this technique, two operating tables are placed perpendicular to each other forming a T [12]. The patient head is placed on the transverse table on which the arms are extended 90°. The body is positioned on the vertically oriented table. The longitudinal table is then shifted away to create an open space around

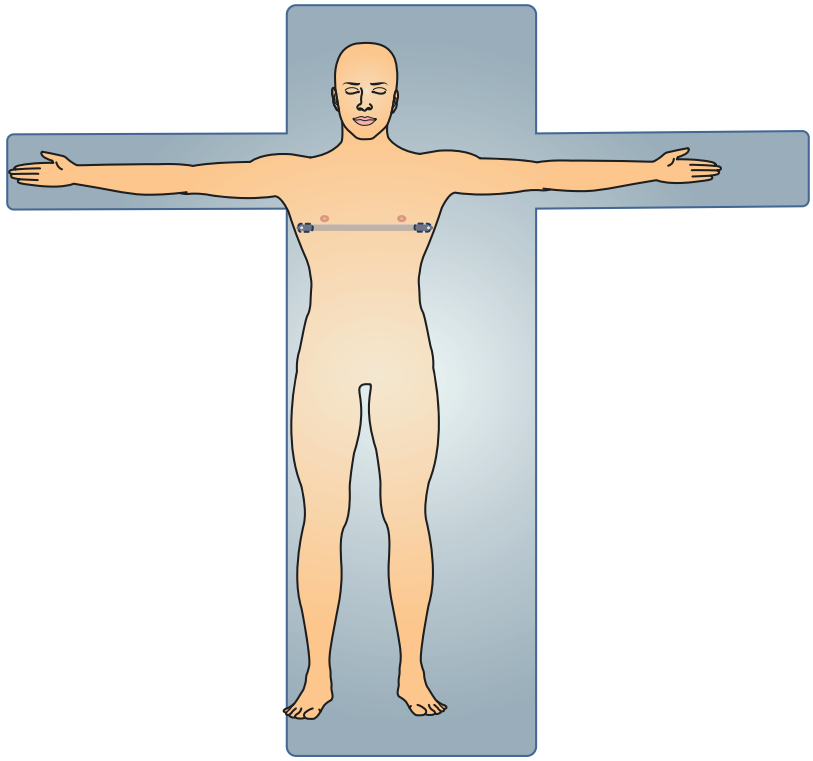

Fig. 34.5 Schematic view of the "Bar straightening technique" for pectus bar removal

the chest, leaving only the thoracic spine is unsupported (Fig. 34.6). The lateral chest incisions are incised simultaneously to remove the lateral stabilizers after which the end of the bar is grasped on one side and removed with a single pull taking advantage of the open space to rotate the bar around the torso between the operating tables.

In the pull down technique, the patient is placed on the edge of the single operating table and the procedure is performed as with the single pull of the bar described above; however in this technique taking advantage of the open space below the operating table to rotate the bar around the torso.

3. ***Prone position technique*** [13]
   The patient is placed in the prone position with the knees flexed, chest supported with two long pillows, arms abducted at the shoulder, and head slightly tilted upward (Fig. 34.7). The lateral incisions are then made to remove fixation sutures or wires. The pectus bar is grabbed and remove with a single pull with the natural curvature of the bar in an upward directed circular motion to remove the pectus bar.
4. ***Bar flipper technique*** [14]
   The patient is placed supine close to the right edge of the operating table (Fig. 34.8). The lateral chest incisions are incised simultaneously and the stabilizer plate is removed. The pectus bar flipper is then placed through the tip of the pectus bar on the left side. The pectus bar flipper is turned several times in the clockwise and counterclockwise direction to loosen the pectus bar from the

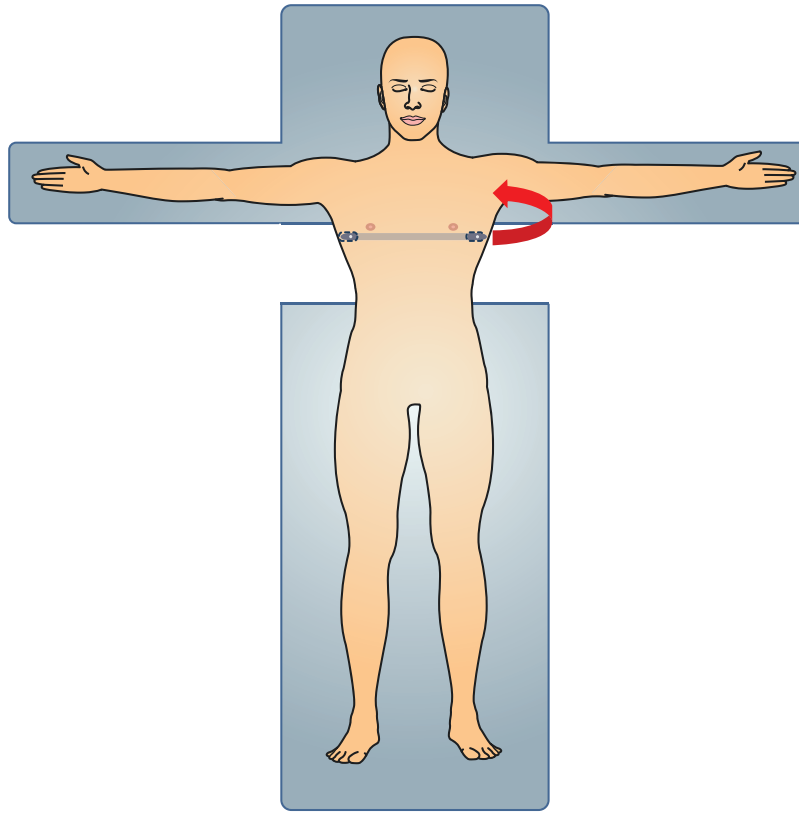

Fig. 34.6 Schematic view of the "Split operation table technique" for pectus bar removal

surrounding fibrous capsule, after which the pectus bar is then slowly pulled out of the thorax along the natural curvature of the chest wall. After half of the bar is pulled out of chest, the flipper is turned in the counter-clockwise direction, parallel to the lie of the patient, after which the bar is completely removed with a C-like curved motion towards the head of the patient.

B. **Instruments for pectus bar removal**

(a) ***Pectus Removal Benders***

Pectus Removal Benders (Biomet Microfixation, Jacksonville, FL, USA) are a pair of instruments the tips of which resemble spanners. The instrument tips are employed to latch on to the pectus bar and allow bending of the petcus bar outwards to enable it to detach from its area of attachment to the chest wall. If straightening of the pectus bar is preferred at the time of removal, the Pectus Removal Benders can be used on both ends to straighten the bar.

(b) ***Pectus Bar Stabilizer Anvil***

In patient with less scar tissue or bone formation around the bar, the *bar stabilizer plate* can be slid off from the *pectus bar* end with minimal effort. However, in moderate or serve forms this could be challenging. Increasing the size of the incision to gain more access is an option, but that still does not solve the problem

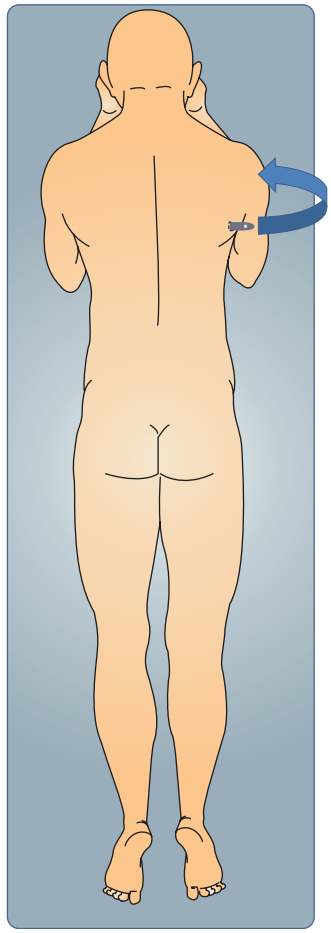

Fig. 34.7 Schematic view of the "Prone position technique" for pectus bar removal

of instrumentation; even more if the repair was a minimal access procedure. Utilization of improper instruments to dislodge an impacted bar stabilizer plate increases the time required for the procedure and can lead to unnecessary damage of the makeshift instruments used.

Since the reaction of the tissue to the implanted *pectus bar* can vary from simple scar to significant bone formation, a *Pectus Bar Stabilizer Anvil* tool was designed for the purpose of detaching the stabilizer plate from the pectus bar (Fig. 34.9) [15]. According to the design provided the tool was manufactured by *Walter Lorenz Surgical Inc., Jacksonville, FL (now Biomet Microfixation, Jacksonville, FL, USA)*. The *Pectus Bar Stabilizer Anvil* is a rod shaped tool with a slightly splayed tip which is designed to fit over the medial segment of the exposed *pectus bar* (without gripping the *pectus bar*) to contact the *bar stabilizer plate* (Fig. 34.10). Once the suture or wire around the *bar stabilizer plate* has been removed, the *Pectus Bar Stabilizer Anvil* tool is placed on the *bar stabilizer* (Fig. 34.11). The tool is tapped with a hammer to enable the transmission of force onto the flanges of the *bar stabilizer*. This facilitates the *bar stabilizer plate* to slide over the *pectus bar* resulting in its detachment (Fig. 34.12).

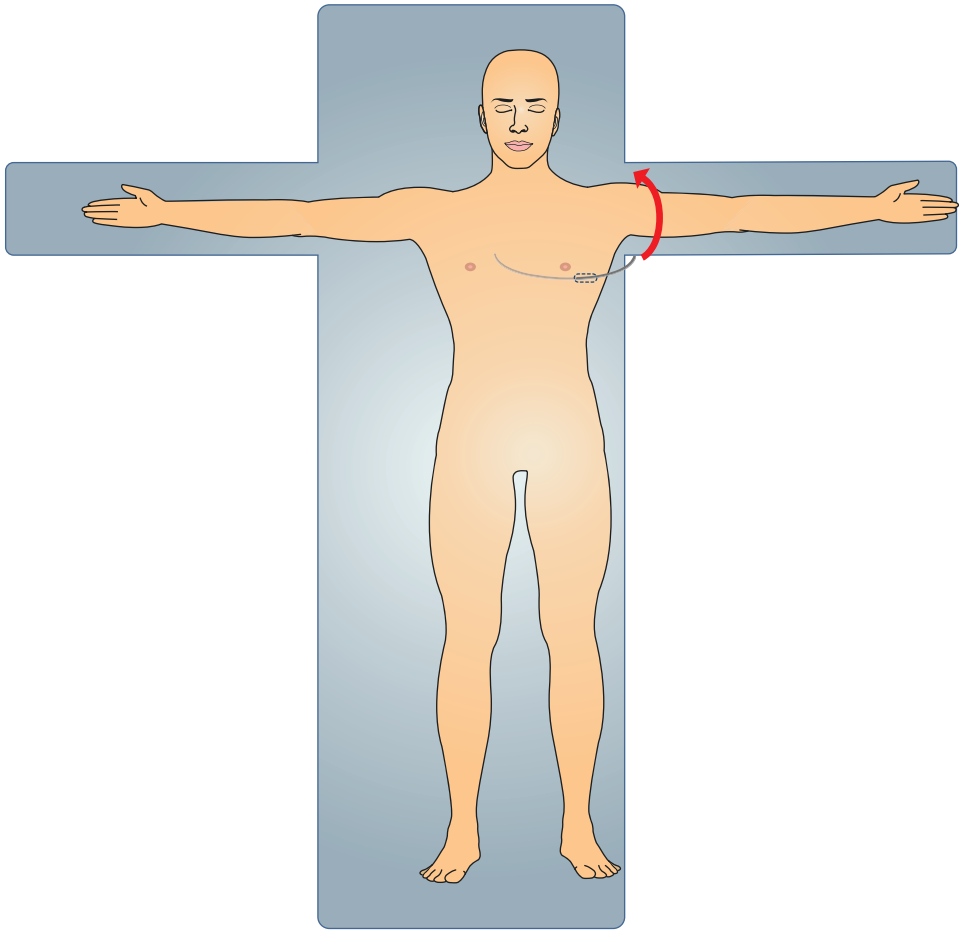

Fig. 34.8 Schematic view of the "Bar flipper technique" for pectus bar removal

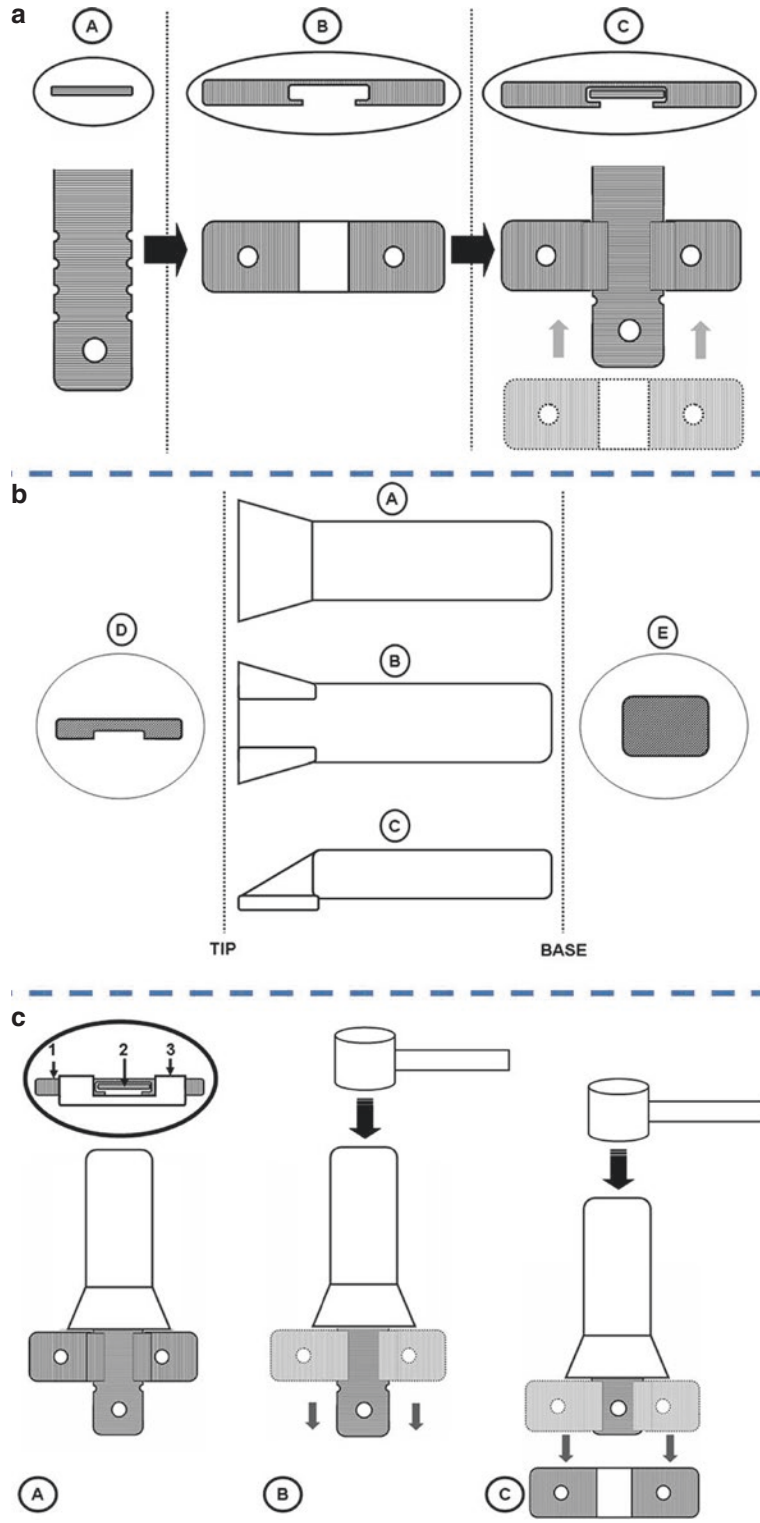

Fig. 34.9 Design diagrams for the development of *Pectus Bar Stabilizer Anvil* tool. **Section A***:* line diagrams of the stabilizer placement: (*A*) pectus bar end, (*B*) bar stabilizer and (*C*) attachment. **Section B***:* line diagrams of the *Pectus Bar Stabilizer Anvil* tool (*A*) front view (*B*) back view and (*C*) side view, (*D*) tapering tip with the groove to accommodate the bar tip and (*E*) rear end. **Section C***:* line diagram of *Pectus Bar Stabilizer Anvil* tool application (*A*) The tip of the tool is placed on the stabilizer plate (1) Stabilizer plate (2) Pectus bar and (3) Pectus Bar Stabilizer Anvil tool tip; (*B*) Using a hammer the tool applies selective pressure on the stabilizer plate to aid its removal; and (*C*) dislodgement of the bar stabilizer

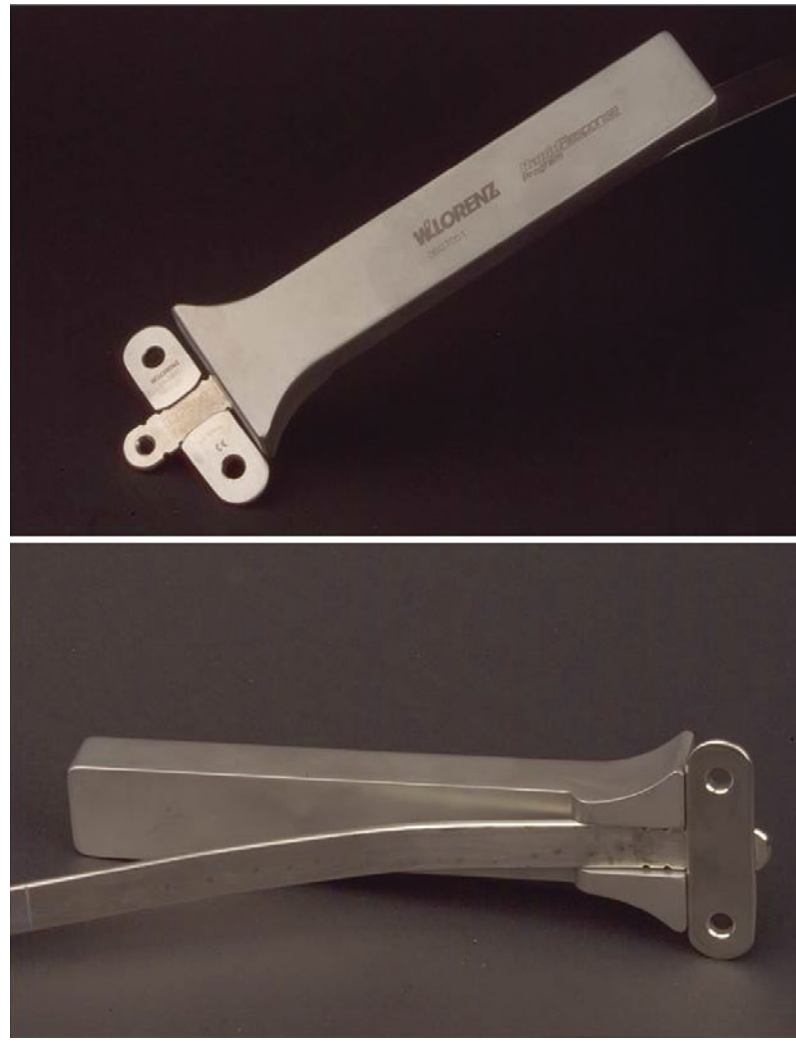

Fig. 34.10 *Pectus Bar Stabilizer Anvil* tool- front view showing placement on a pectus bar and stabilizer plate (*top*) and back view showing the groove in the tip to accommodate the pectus bar and positioning on the stabilizer plate (*bottom*)

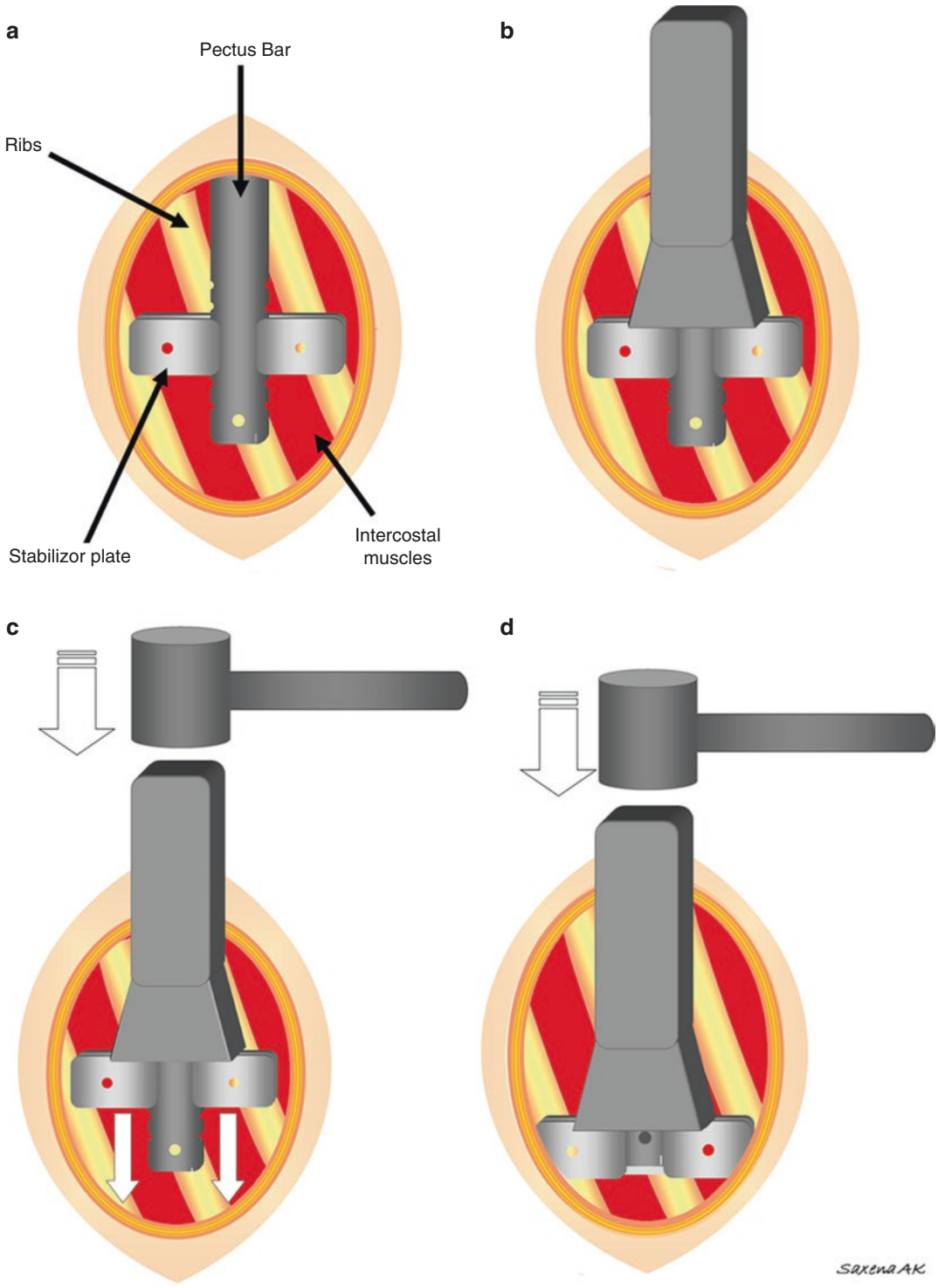

Fig. 34.11 Schematic view of the site of the operation showing the application of the *Pectus Bar Stabilizer Anvil* (**a**) Stabilizer plate attached to the pectus bar and chest wall, (**b**) placement of the *Pectus Bar Stabilizer Anvil* tool on top of the stabilizer plate, (**c**) using a hammer the rear of the anvil too is tapped to (**d**) detach the stabilizer plate from its attachments the pectus bar

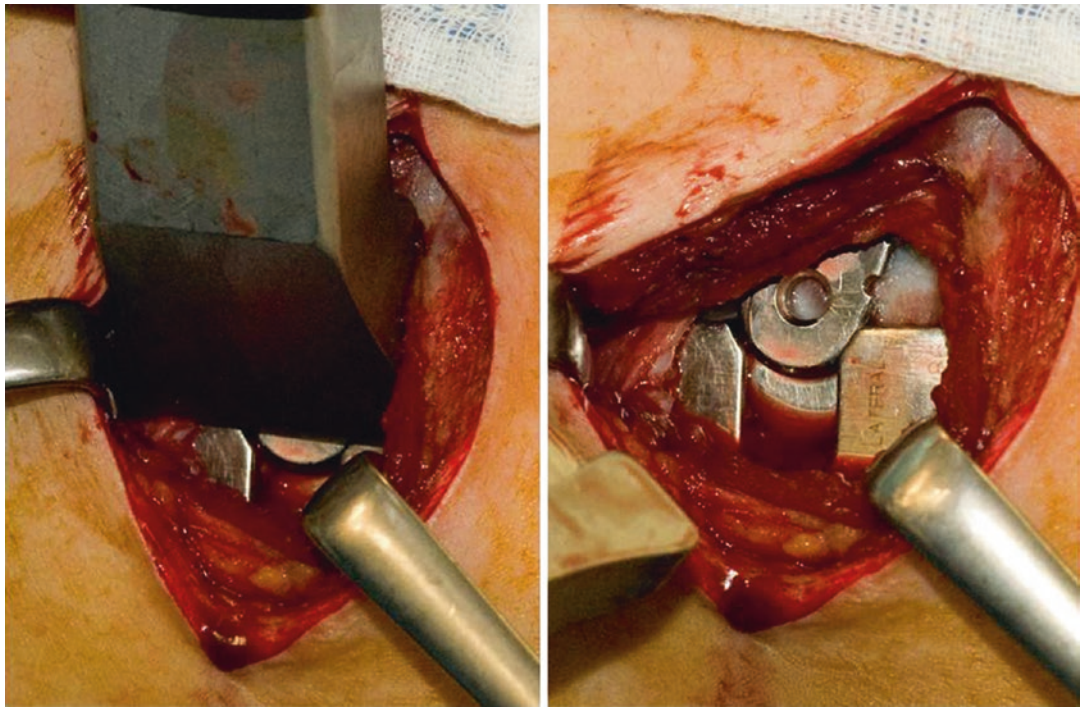

Fig. 34.12 Operative view: (*left*) Placement of the *Pectus Bar Stabilizer Anvil* tool on top of the stabilizer to dislodge the stabilizer plate and (*right*) detached stabilizer plate

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Part IV

Pectus Excavatum: Surgical Techniques

Minimal Access Repair of Pectus Excavatum

35

Michael Höllwarth and Amulya K. Saxena

Technical Highlights

The Minimal Access Repair of Pectus Excavatum (MARPE) is a video assisted thoracic surgery procedure for the correction of chest wall depression type of deformities.

The main technical highlights of this procedure are:

(a) Mapping of the thoracic contours using the pectus template and modeling the template
(b) Selection of an appropriate size pectus bar
(c) Bending of the pectus bar with an orthopedic metal plate press
(d) Bilateral transverse incisions in the midaxillary line and muscle preparation
(e) Insertion of a 5 mm port below the right transverse chest incision
(f) Thoracic insufflation with 4–5 mmHg $C0_2$
(g) Insertion of the pectus introducer through the right chest
(h) Introducer tip is used to dissect the intercostal muscle and enter the right thorax
(i) Rotation of the introducer tip and preparation of the retrosternal tissue behind the sternum
(j) Exiting of the introducer tip through the intercostal muscles in the left thorax
(k) A cotton umbilical tape is tied to the slit in the introducer tip
(l) The introducer is retrieved on the right side with the tape after which the tape end is cut
(m) The perforation on the edge of the pectus bar is secured to the tape
(n) The pectus bar is introduced into the right chest right guided by the tape
(o) The end of the pectus bar is retrieved out of the left thorax and the tape is cut.
(p) Using a pectus bar flipper, the bar is secured on the right edge and flipped into position
(q) A single stabilizer plate is used on the right side secured by a PDS suture around the rib
(r) The muscles and skin are adapted and the wound is closed
(s) The patient is tilted to a head down position with a left body tilt
(t) Insufflated gas is evacuated through the port aided by lung insufflation by the anesthetist
(u) After complete evacuation the port is removed and the insertion site closed.

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Indications

MARPE is suitable for correction of chest deformities with depression:

- Symmetric Pectus Excavatum
- Mild or moderately asymmetric Pectus Excavatum
- Pectus Excavatum with Platythorax

Preoperative Considerations

1. The procedure should be offered and performed in patients>14 years of age. Due to variations in body growth among teenagers, surgery should be reserved for teenagers whose body sizes have reached adolescent or adult proportions.
2. Adult patients of any age with relatively stiff chest wall are well suited for this procedure.
3. Chest radiographs are taken to access the extent of the deformity.
4. Lung function tests are performed to evaluate the pulmonary function.
5. Electrocardiograms and or echocardiography to diagnose or rule out cardiac pathologies.
6. Computed tomography scans are performed to plan surgery with regards to the level of depression, sternal rotation and the degree of cardiac displacement. Three-dimensional images are helpful in planning the procedure.
7. Photographic documentation of the deformity
8. Coagulation tests are performed to rule out disorders in bleeding disorders
9. Since nickel is present in stainless steel struts, patient history is taken to rule out nickel allergy.
10. Antibiotics are administered on the day of the procedure and are continued for a period of 3 days after the procedure. Cefuroxime is the choice of antibiotic.

Special Instruments

MARPE requires the following list of instruments which are specific for this procedure:

1. Pectus bar
2. Orthopedic metal plate bender press
3. Pectus introducer
4. Pectus bar bender
5. Pectus bar flipper
6. Bar stabilizer plate
7. Cotton umbilical tape
8. Scope for video assisted thoracic surgery (5 mm, 30°)
9. Port with blunt trocar tip and insufflation channel 5 mm
10. Endoscopy tower/suite

Surgical Technique

The patient is positioned supine towards the right edge of the operation table with both arms extended perpendicular to the torso. The extended arms are positioned and rested on side tables which are attached to the main operating table. After the patient is prepped, the thoracic contours of the deformity are mapped at the point of maximum depression using a pectus template. The template is then further molded to conform to the desired repair contours. An appropriate size pectus bar is then selected and bent appropriately using an orthopedic metal plate press. The bent pectus bar is kept in a solution of gentamycin until it is implanted.

Transverse skin incisions in the mid-axillary line are done on both sides of the chest and the muscles are preparation until the rib cage is exposed. Blunt direction is carried out in the plane between the pectroalis major muscle and the rib cage; with the preparation directed medially towards the sternum. A 5 mm port is inserted below the right transverse chest incision just above the level of the diaphragm and insufflation is maintained with a pressure of 4–5 mmHg $C0_2$. This is followed by the insertion of the pectus introducer through the right chest. Keeping the tip of the introducer pointing downward, the intercostal muscles are bluntly dissected to enable the pectus introducer to enter the right thorax. The introducer tip is then rotated 180° and with the tip now pointing upwards and the retrosternal tissue behind the sternum is prepared bluntly. After this the pectus introducer tip is passed through the intercostal muscles in the left thoracic cavity to enable the tip of the introducer

to exit the left thoracic cavity. Care should be taken to prevent large lacerations or tears in the intercostal muscles as the pectus introducer is manipulated within the intercostal spaces. Two cotton umbilical tapes are tied to the slit in the introducer and the introducer is retrieved on the right side with the tape after which the tape end is cut. The second tape is kept as a back-up in case the first tape dislodges or snaps during pectus bar positioning. The perforation on the edge of the pectus bar is now secured to the tape and the pectus bar is introduced into the right chest guided by the tape. The end of the pectus bar is retrieved out of the left thorax and the tape is cut. With the pectus bar now placed behind the sternum and its ends facing upwards like a U, the pectus bar flipper is attached securely to the right end of the pectus bar. The bar is then flipped 180° into position. The bar ends should be in close proximity to the rib cage on the sides. Although two stabilizer plates can be applied on both ends of the pectus bar, a single stabilizer plate used on the right side is sufficient to secure the pectus bar. Pectus bar with integrated stabilizer plate on one end is preferred, as this one piece unit does away with the problems of stabilizer plate and pectus bar disconnections. Using a large diameter Deschamps ligature carrier a heavy polydiaxone (PDS suture Ethicon, Somerville, NJ, USA) is placed around the rib which passed through the perforations of the stabilizer plate and tied. The muscles at the site of the incision are adapted and the skin incision closed with a monofilament non-absorbable running suture. At this time, the patient is brought into left semi-lateral head drop position so as to raise the level of the port to the highest position and facilitate gas evacuation from the thorax. After complete evacuation of the insufflated gas, the port is removed and the port-site is closed with a single suture.

In adult patients, with severe depression, it is advisable to use 2 pectus bars. The first bar is placed cranial to the level of maximal depression. The placement of the first cranial bar aids in the partial elevation of the depression and also allows for easier placement of the second bar placed caudal at the level of maximal depression. The placement of 2 pectus bars also distributes the forces of the chest wall and has been found to be associated with decreased postoperative pain in adults with severe pectus excavatum deformities. Two pectus bars are also recommended for correction of pectus deformities associated with platythorax in the both the adolescent and adult age groups. In case the two pectus bar option is chosen, the second stabilizer plate is placed on the other side of the chest.

Procedure Overview (Figs. 35.1, 35.2, 35.3, 35.4, 35.5, 35.6, 35.7, and 35.8)

Postoperative Management

Perioperative antibiotic therapy is administered and continued for 3 days; with Ceftriaxone being the drug of choice. Postoperative chest films are taken in all patients. All patients are also administered strong analgesics for first 72 h. Depending on the age and response of the patient, sedation is maintained for the first 1–3 days for postoperative pain. Further administration of medications and therapies depends on the use of an epidural catheter, patient-controlled analgesia (PCA) or oral analgesia. The patients are mobilized on the 2nd day after surgery with assistance. Attention is paid that there is no chest or waist bending and no twisting during mobilization. Deep breathing and incentive spirometry is advised at regular intervals. Pain medications are reduced successively and the patients are discharged on the 5th day on oral pain medications. Patients who preferred to be discharged earlier are permitted to leave the hospital. At the time of discharge patients were advised to avoid body contact sports. Once discharged, the patients are advised to maintain good posture with return to regular activities recommended after a period of 4–6 weeks. During these initial 4–6 weeks patients are advised to:

- Take regular walks of short intervals and gradually increase the duration of walks

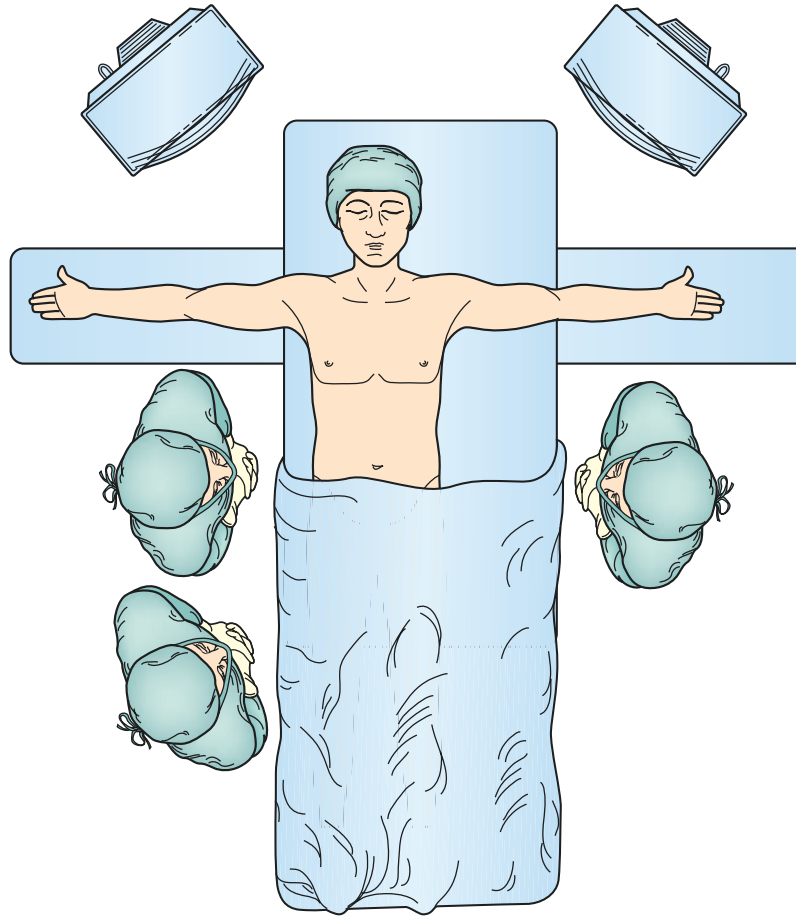

Fig. 35.1 Operation room layout with positioning of the patient on the right edge of the table. The surgeon is positioned near the right arm of the patient; the cameraman stands next to the surgeon towards the torso of the patient and the first assistant on the left side

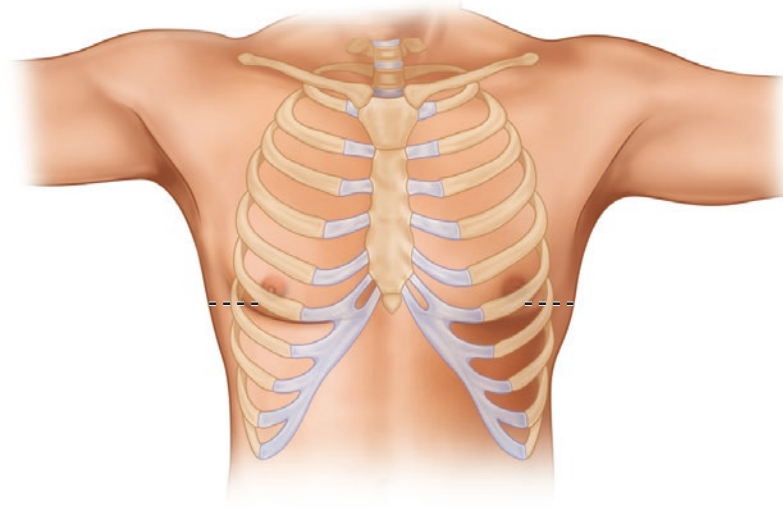

Fig. 35.2 Transverse skin incisions in the mid-axillary line are done on both sides of the chest and the muscles are preparation until the rib cage is exposed

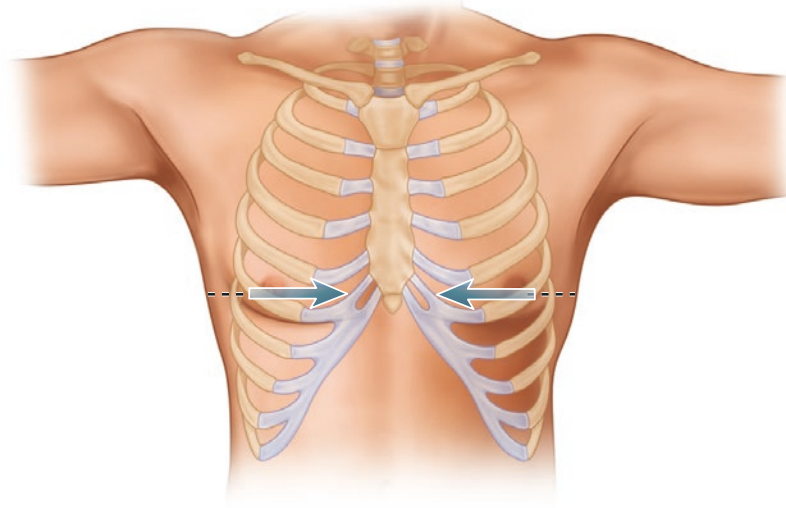

Fig. 35.3 Blunt direction directed medially towards the sternum is done in the plane between the pectoralis major muscle and the rib cage

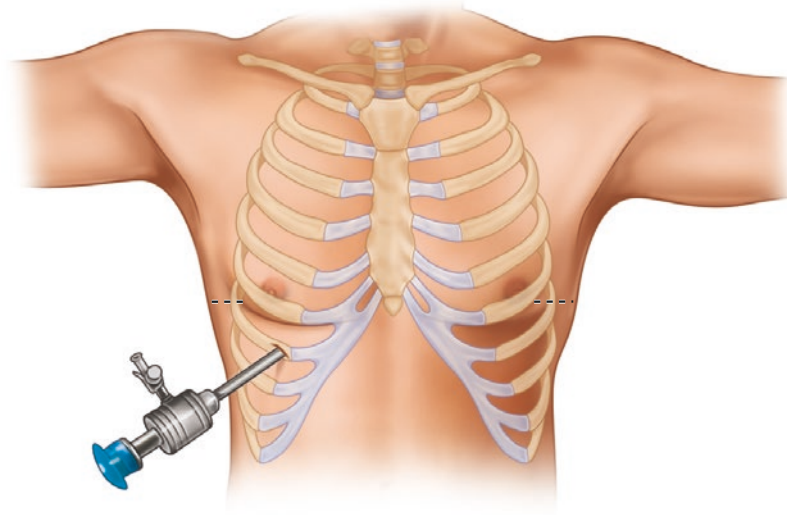

Fig. 35.4 A 5 mm port is inserted below the right transverse chest incision just above the level of the diaphragm

- Avoid waist bending or twisting
- Perform breathing exercises further that were initiated after surgery
- Shower or bathe after 1 week
- Avoid lifting heavy objects or weights for 8 weeks
- Avoid sports or sport activities for 12 weeks

The patients are followed-up 2 weeks after discharge and with further follow-up planned after 3, 6, 12 months and then after 3 years at the time of bar removal.

Results

MARPE gained popularity in the correction of pectus excvatum types of chest wall deformities in the late 1990's and the technique has evolved over the years. MARPE has a learning curve and

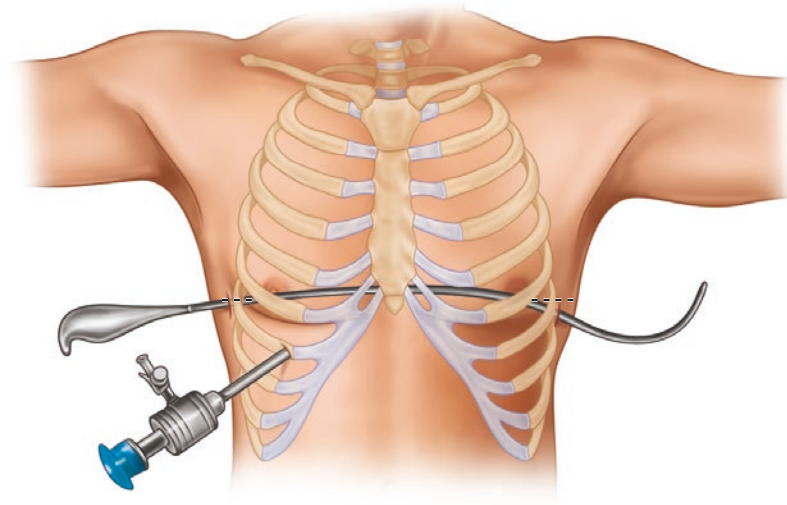

Fig. 35.5 The pectus introducer is inserted into the chest from the right incision which circumvents the sternal depression and exits the thorax on the left side. Cotton umbilical tapes are tied to the tip of the pectus introducer after which it is retracted back out of the right chest incision with the tapes

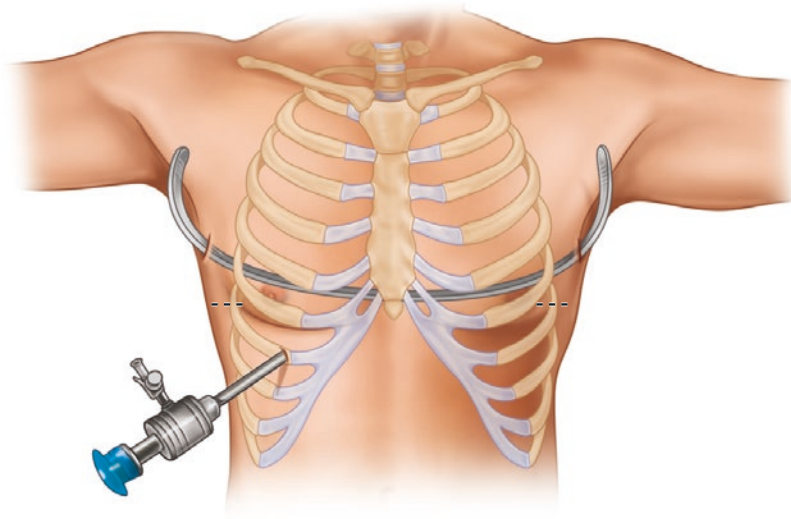

Fig. 35.6 The pectus bar is then placed behind the sternum and its ends facing upwards like a U using the cotton umbilical tapes to guide it through the thoracic cavity and the retrosternal space

surgical related complications and morbidities have declined over the past decade with increasing experience in the procedure and the comparison of technical modifications between Centers.

Results from our Center reported in 2008 which were part of the learning curve presented the experience with the 167 patients (136 males and 31 females) with a mean age of 16.3 (range 5–40 years). Minor complications were seen in 122 patients (73.1 %) and consisted of breakage of wires used to secure the lateral stabilizer plate (n=48), pleural effusions (n=28), intraoperative rupture of the intercostal muscle (n=15), pericardial tears without clinical significance (n=7) and lung atelectasia (n=4). Most minor complications were avoided in the subsequent patients with modification in the technique. These included fixation of the bar and the stabilizer plate to the underlying rib, use of PDS cords instead of metal wires to fix the bar and the stabilizer plate, and entry and exit into the thorax with the introducer in closer proximity to the sternum. Complications such as residual pneumothorax were minimized by near to total evacuation of the insufflated gases

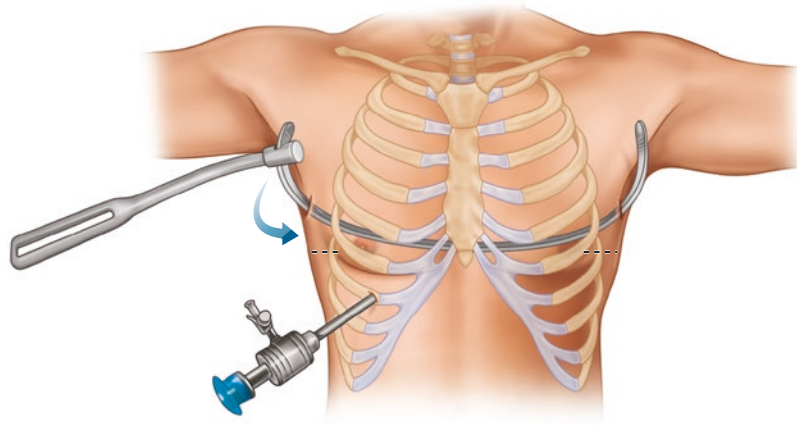

Fig. 35.7 The pectus bar flipper is attached securely to the right end of the pectus bar after which the pectus bar is flipped 180° into position

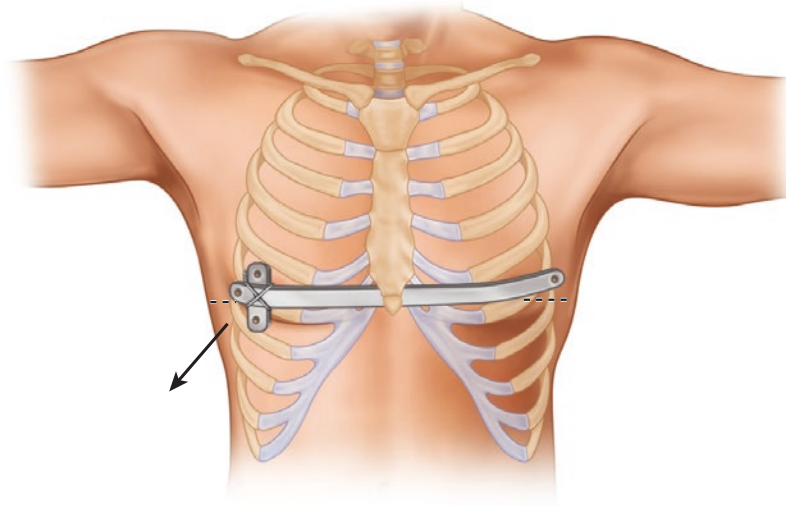

Fig. 35.8 The pectus bar is stabilized with a single stabilizer plate on the right side secured with PDS cords and wound is closed. After a head low left semi-lateral positioning of the patient the insufflated carbon dioxide are evacuated from the thorax through the port, after which the port is removed

through the port with head low tilt in the left semi-lateral position of the patient aided by controlled inflation of the lungs through the anesthetist.

Major complications occurred in seven patients (4.2 %) and consisted of one intraoperative heart perforation, one piercing of the liver with the trocar, bar infections (n = 2) and significant bar displacement (n = 3). Despite the application of intra-thoracic visualization using scopes, possible injury to the heart is extremely rare but cannot be ruled out during MARPE. Although the scopes can follow introducer tip in the right thorax, there is a blind-spot on the contralateral side of the sternal depression as the introducer tip is advanced in the left side of the thorax. In severe pectus deformities, the use of a vacuum bell or a sub-sternal incision could be other options to elevate the sternum during insertion of the introducer tip. Estimation of the level of the diaphragm is important in preoperative workup as the level of the diaphragm is variable in patients. Injury to the liver with trocars during the placement of ports can be avoided by careful evaluation of chest films in patients prior to surgery. With regards to bar displacement, changes in fixation of the bar as described in the previous paragraph have shown to overcome this complication. Major complications related to the

MARPE are rare and are mainly attributed to the learning curve.

Infection in the implanted bars could pose a major complication after MARPE. If a bar is infected, there is no need for immediate bar removal and it is recommended to treat the patient with antibiotics first. In order to minimize the chance of bar infections, after bar bending the bar is placed in a solution of gentamycin until the chest is prepared for implantation. The chest incisions are packed with betadine gauze intermittently during other the procedural steps. Furthermore, the contact of the bar to the skin of the patient must be minimized during the implantation process. As a standard part of our protocol, the operating teams gloves are changed after the chest wall incisions and the retrosternal tunnel has been established and before the bar is implanted.

Procedure Related Images

(Figs. 35.9, 35.10, 35.11, 35.12, 35.13, 35.14, 35.15, 35.16, 35.17, 35.18, 35.19, 35.20, 35.21, 35.22, 35.23, 35.24, 35.25, 35.26, 35.27, 35.28, and 35.29)

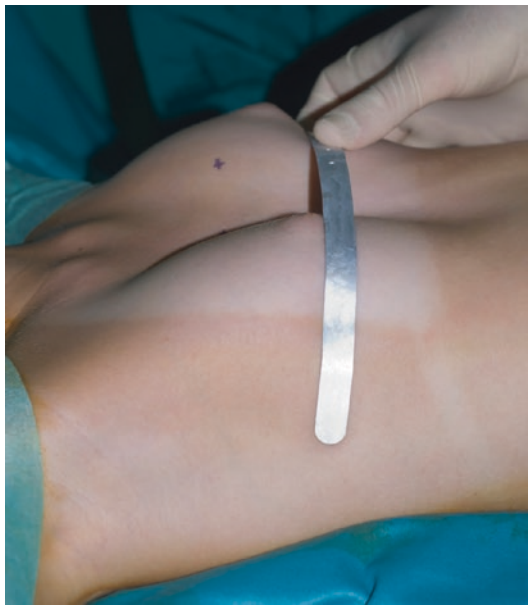

Fig. 35.9 After the patient is prepped, the contours of the deformity are mapped using a pectus template at the point of maximum depression

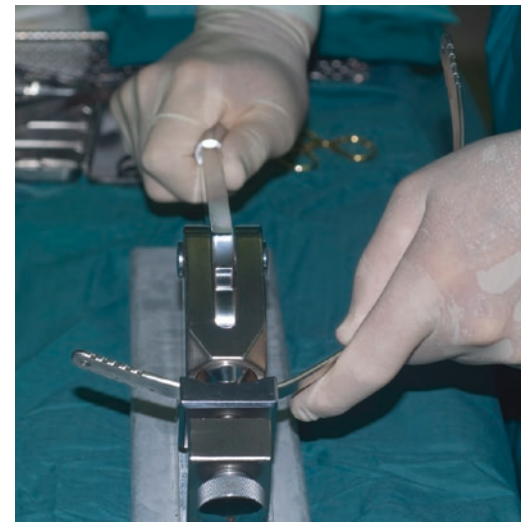

Fig. 35.10 The pectus bar is bent appropriately using an orthopedic metal plate press

Fig. 35.11 The bent pectus bar is placed in a gentamycin solution until the time of implantation

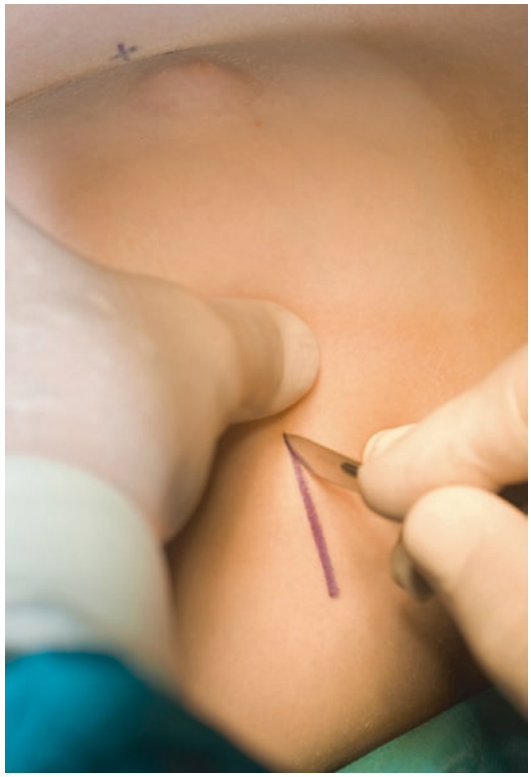

Fig. 35.12 Transverse skin incision lines are drawn in the mid-axillary line on both sides of the chest

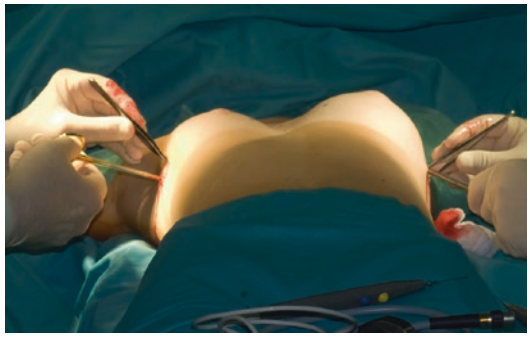

Fig. 35.13 The skin is incised simultaneously on both side of the chest

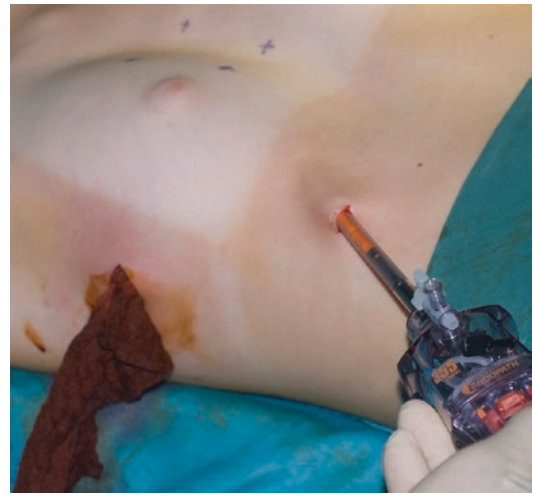

Fig. 35.14 Sub-muscular tunnels are created directed medially between the pectoralis muscle and the rib cage towards the sternum after which betadine gauzes are placed in the wounds. A 5 mm port is inserted below the right transverse chest incision above the estimated level of the diaphragm

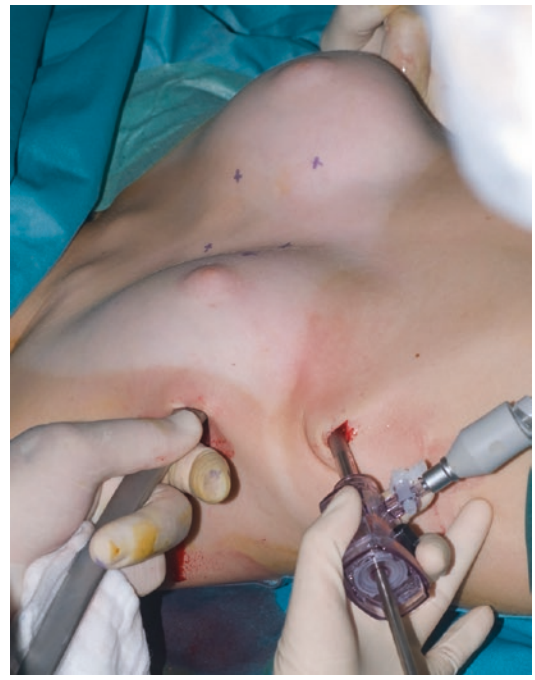

Fig. 35.15 The pectus introducer is then inserted through the right chest incision in to the submuscular tunnel-

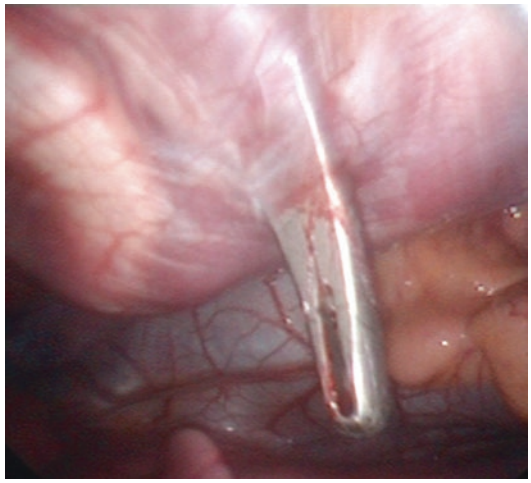

Fig. 35.16 Endoscopic view of the pectus introducer tip entering into the right thoracic cavity through the intercostal space after blunt dissection

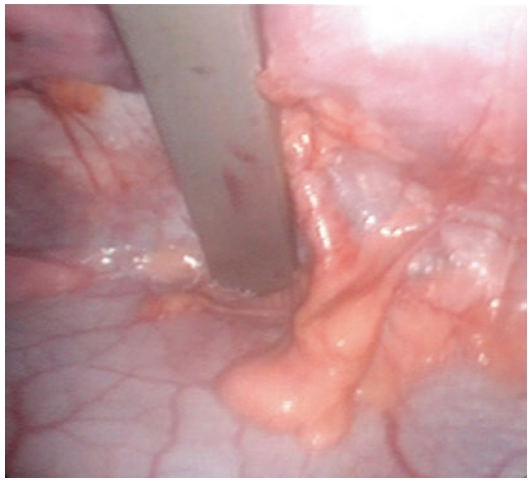

Fig. 35.17 Endoscopic view of the introducer traversing the retrosternal space below the sternal depression

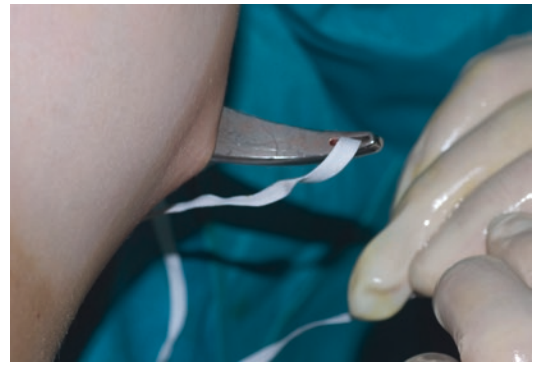

Fig. 35.18 Cotton umbilical tapes are tied on the tip of the inserted pectus introducer

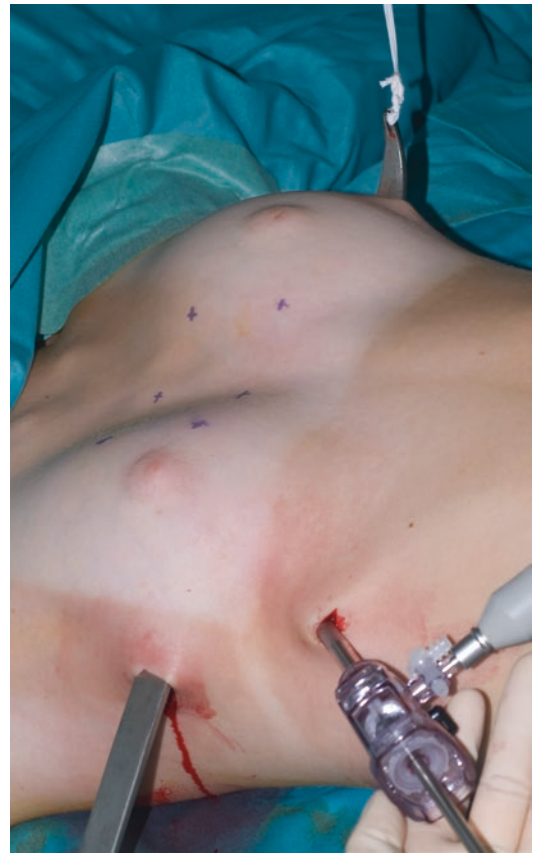

Fig. 35.19 After the cotton umbilical tapes are tied the pectus introducer is retracted

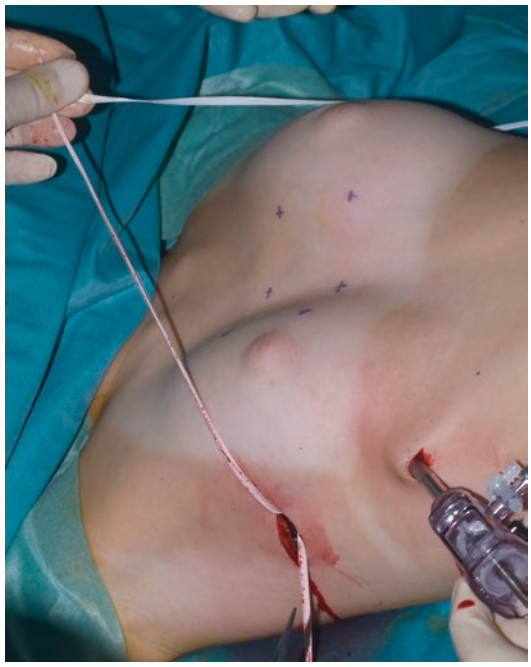

Fig. 35.20 The ends of the cotton umbilical tapes are identified and secured

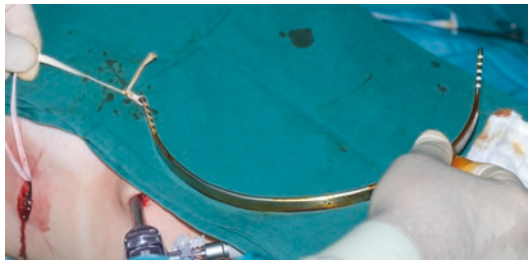

Fig. 35.21 The end of one tape is tied securely to the perforation on the pectus bar

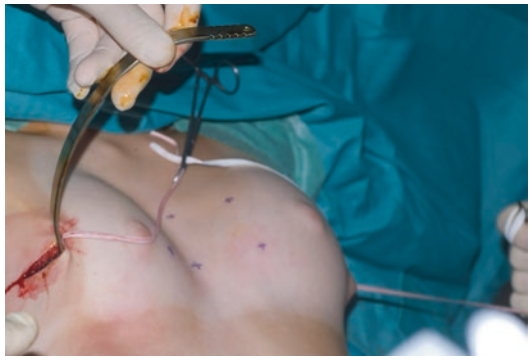

Fig. 35.22 The pectus bar is guide through the retrosternal space using the cotton umbilical tape to aid its placement

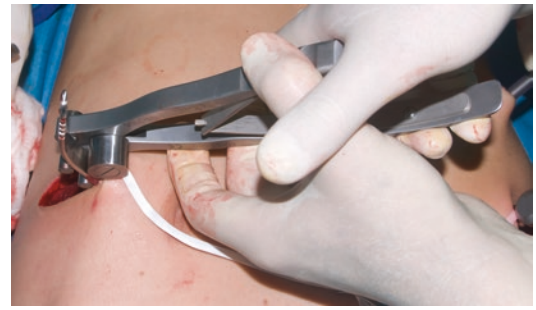

Fig. 35.23 Final corrections to the pectus bar contours are made is necessary using a pectus bar bender

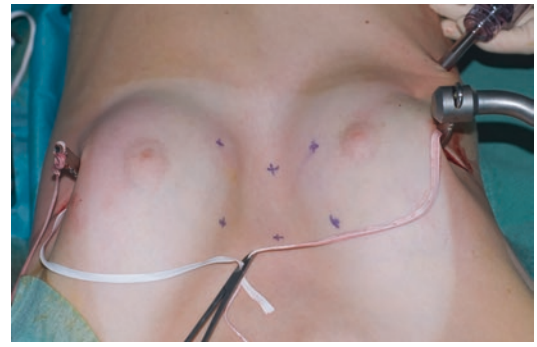

Fig. 35.24 The pectus bar flipper is latched to the right end of the pectus bar to rotate the bar 180° into its desired position

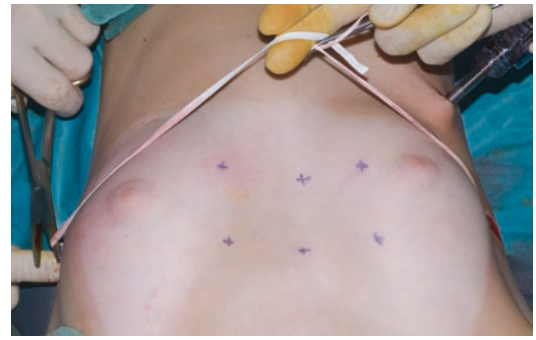

Fig. 35.25 The second cotton umbilical tape if not used is cut and removed

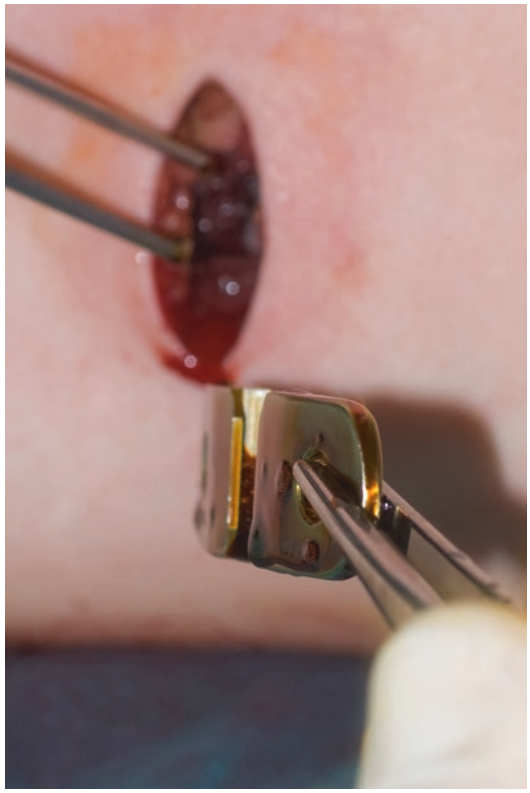

Fig. 35.26 A single stabilizer plate is placed on the right side to secure the pectus bar

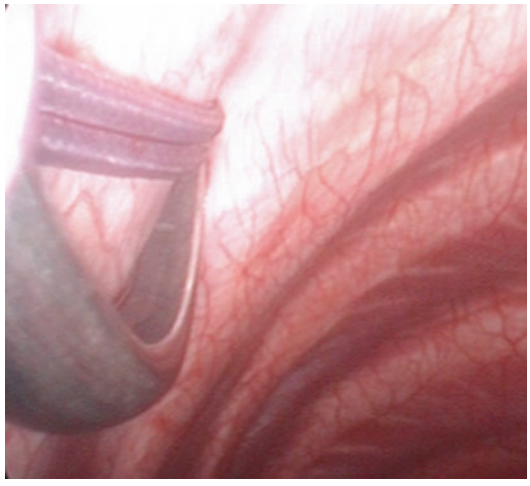

Fig. 35.27 Using a heavy PDS suture on a Deschamps ligature carrier, the cord is passed behind the rib and through the hole in the stabilizer plate after which it is tied. Endoscopic view of the Deschamps ligature carrier being passed behind the rib

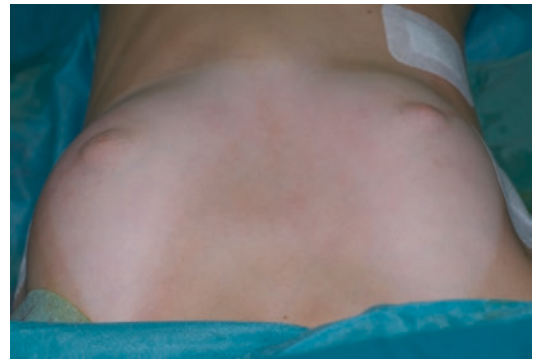

Fig. 35.28 Result of the minimal access repair after completion of the procedure

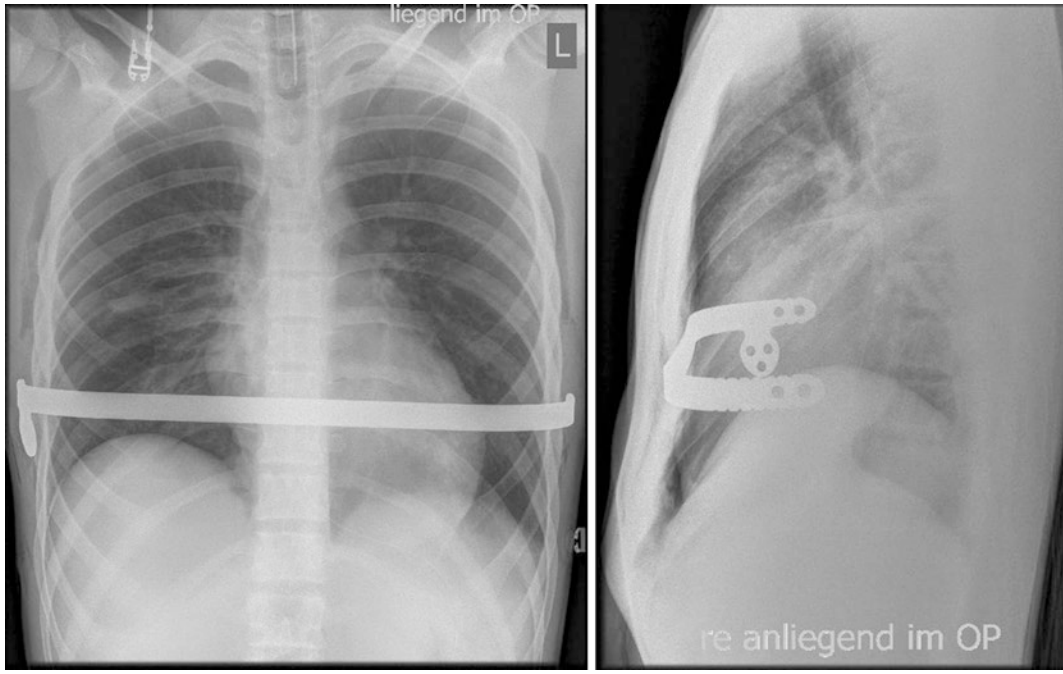

Fig. 35.29 Chest films (left-posterior anterior and right-lateral) showing the placement of the pectus bar in a male patient. The pectus bar used in this patient has an integrated stabilizer on the right end

Suggested Reading

1. Castellani C, Schalamon J, Saxena AK, Höllwarth ME. Early complications of the Nuss procedure for pectus excavatum: a prospective study. Pediatr Surg Int. 2008;24:659–66.
2. Nuss D, Kelly Jr RE, Croitoru DP, Katz ME. A 10-year review of a minimally invasive technique for the correction of pectus excavatum. J Pediatr Surg. 1998;33:545–52.
3. Saxena AK, Castellani C, Höllwarth ME. Surgical aspects of thoracoscopy and efficacy of right thoracoscopy in minimally invasive repair of pectus excavatum. J Thorac Cardiovasc Surg. 2007;133:1201–5.
4. Schalamon J, Pokall S, Windhaber J, Höllwarth ME. Minimally invasive correction of pectus excavatum in adult patients. J Thorac Cardiovasc Surg. 2006;132:524–9.
5. Van Renterghem KM, von Bismarck S, Bax NM, Fleer A, Höllwarth ME. Should an infected Nuss bar be removed? J Pediatr Surg. 2005;40:670–3.

Surgical Techniques- Pectus Excavatum: The Modified Ravitch Procedure

36

Julia Funk and Christian Gross

Technical Highlights

The main technical highlights of this procedure are:

(a) Vertical incision in males and females
(b) Dissection of the pectoral muscles to expose the sternum and ribs
(c) Bilateral parasternal chondrotomy to release the sternum from the ribs
(d) Digital separation of the sternum from the pericardium and pleura
(e) Proximal sternotomy
(f) Undermining of the sternum and insertion of bone strut
(g) Fixation of the bone strut to the osseous portion of the ribs
(h) Suture fixation of the rectus muscle to the sternum
(i) Suture fixation of the pectoral muscles

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Indications

Since Ravitch's description of this technique for the correction of pectus deformities, most of the repairs performed by this procedure are modifications of the original technique. The modification used for repairs at our Centre utilizes a bioresorbable bone strut which avoids subjecting the patient to a second surgical procedure.

- All symmetric and asymmetric forms of Pectus Excavatum (PEX) and Pectus Carinatum (PEC)

Preoperative Considerations

1. As a general rule, the procedure should be offered to and performed in patients >12 years of age. The preferred age is about 14 years for both genders.
2. This procedure is also well suited for adult patients of any age with relatively rigid chest walls.
3. Lung function tests are performed to evaluate pulmonary function.
4. Electrocardiograms should be performed to diagnose or rule out cardiac pathologies.
5. Magnetic Resonance Imaging of the chest is required to calculate the Haller-Index.
6. Photographic documentation of the deformity and postoperative results.
7. Standard preoperative blood-tests including blood count, coagulation tests, CRP.

A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_36

8. Antibiotics are administered on the day of the procedure and are continued for a period of 3 days after the procedure. Cefuroxime is the antibiotic of choice.

Special Instruments

1. An allogenic bone strut from either the tibia or femur is required with a minimum length of 250 mm.
2. Saw for adjusting the length of the bone graft
3. Bone File for grinding the edges of the bone graft to avoid pleural or pericardial perforation
4. Luer Bone Ronguer
5. Periosteal elevators

Surgical Technique

The length of the vertical midline incision depends on severity and localisation of the deformity. In general, an incision of 7–10 cm length is sufficient for both genders. The skin incision begins proximally on the level of the first deformed rib and extends just short of the level of the xiphoid. The subcutaneous tissue is dissected until the periosteum of the sternum is reached. The preparation is then continued laterally. The pectoral muscle is exposed and detached from its sternal insertion to the sternocostal joints using electrocautery. The dissection is continued in the lateral plane and the muscles are separated from the anterior surface of the ribs using blunt dissection and electrocautery until the costal perichondrium is fully exposed. Only in exceptional cases it is necessary to detach the intercostal muscles completely. After preparing one side the edges and the base of the wound are covered with soaked gauze which is followed by similar preparation on the contralateral side. The rectus abdominis muscle is then released partially from xiphoid and ribs; and complete detachment is usually not necessary. Hemostasis is performed thoroughly during soft tissue preparation.

After exposing the cartilaginous sternal rib attachments over a length of about 5 cm, their perichondrium is incised longitudinally over the entire exposed surface with a scalpel. The cartilage is then dissected from the perichondrial sleeve with great care. It is technically easiest to start at the sternocostal joint with a *Luer Bone Ronguer* and proceed with a small *Periosteal Elevators* to "peel" the perichondrium from the cartilage without lacerating the perichondrium. Generally 5–8 costal cartilages have to be removed on each side in order to facilitate the mobilization of the sternum.

During the separation of the sternum from the ribs, it important to assess the mobility of the detached sternum. This can be performed using a sharp clamp which is used to grasp the distal end of the sternum or the xiphoid and evaluated to access the degree of mobilisation. Injuries to the pleura are a rare occurrence during these manipulations. The integrity of the pleura can be confirmed if in doubt by having a Valsalva manoeuvre performed through the anaesthesiologist after sealing the operation site with warm sterile solution. If a pleural lesion is established, the pleura can be closed with 5–0 or 6–0 Vicryl sutures.

When the sternum is sufficiently mobilized to compensate the deformity, established through elevation of the sternum as described above, the liberation of the sternum from pleura and pericardium can be performed. At the level where the deformity is most pronounced, the sternocostal joints are resected bilaterally. The sternum is then elevated with a sharp clamp that is attached to the sternum at the sites of resection. This is followed by the detachment of the sternum from the pleura and pericardium through blunt and preferably digital dissection. Digital preparation is best achieved when commenced from the sternum edges. Using this manoeuvre the retrosternal space can be separated progressively until the bone strut can be passed through it with ease.

The next step involves the preparation of the bone graft. For this allogenic human frozen femoral or tibial bone grafts are used which has been prepared according to local, national, and European regulations in a certified bone bank. The preparation of these grafts involves a standardized chemical process for cold sterilization through the application of peracetic acid-ethanol under negative pressure. According to the medical drug law (§105 AMG) the conditioned bone is a licensed pharmaceutical product. A pharmacological interaction of the graft with other substances is not known of and

also the probability of transmission of infectious diseases is minimized or excluded. The struts are obtained as cortical bone grafts of different lengths with a width of about 1.5–2 cm. The length of the strut should be approximately equal the distance between the two nipples, and is shortened to the required length size with an oscillating bone saw. All the edges are then thoroughly ground with a file until they are smooth throughout without any burrs.

If the mobilization of the sternum does not lead to a full correction of the deformity, a sternotomy has to be performed and is carried out before the insertion of the bone strut. The site for the osteotomy is the lower third of the manubrium and 2–4 cm cranial of the most proximal chondrotomy/rib resection. The osteotomy is carried out with an oscillating saw in a transverse direction with the saw blade held at a 45–60° angle in a caudal to cranial direction. The osteotomy is performed on the outer periosteum leaving the inner periosteum intact, and a wedge of appropriate size to correct the deformity is excised. Closing wedge correction of the deformity can be achieved without complete destabilization of the sternum hence the integrity of the periosteum is maintained on one side. This will play an important role in achieving a good outcome.

In order to avoid skin and soft tissue damage during the passage of the bone strut, the wound edges are covered with moistened gauze. The bone strut is inserted in a transverse plane of the retrosternal tunnel from left to right during which the sternum is held in an elevated plane. The lateral ends of the strut are then rested on top of osseous surface of the ribs. Through the Valsalva manoeuvre, the pleura is again tested for integrity. As the pleura remains intact, no placement of chest tube is necessary. The bone strut is fixed to the sternum with heavy sutures 1–0 or 2–0 vicryl or PDS sutures which are tied over the sternum after passing them interlacing beneath the bone strut and sternum.

Sometimes it is necessary to smooth the deformed anterior part of the sternum for best esthetic results. This is achieved either with the oscillating saw held in a tangential direction towards the sternum or with a *Luer Bone Ronguer* for more selective corrections. The resulting areas of cancellous bone are closed with bone wax to avoid the development of wound hematoma. The perichondrial sleeves are readapted and the pectoralis muscle is reattached to its sternal insertion. A subcutaneous wound drainage is inserted and the subcutaneous and cutaneous tissue is adapted conventionally (Figs. 36.1, 36.2, 36.3, 36.4, 36.5, 36.6, and 36.7).

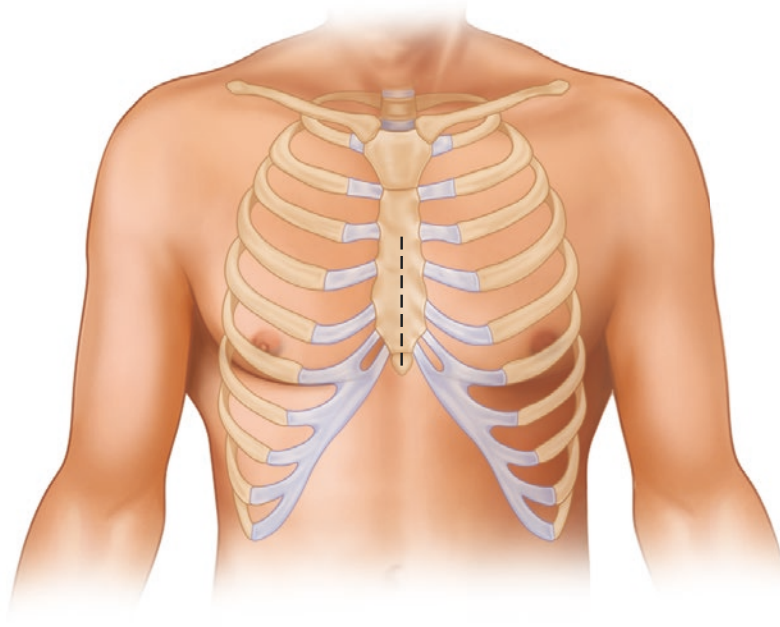

Fig. 36.1 Vertical midline incision is preferred in both genders

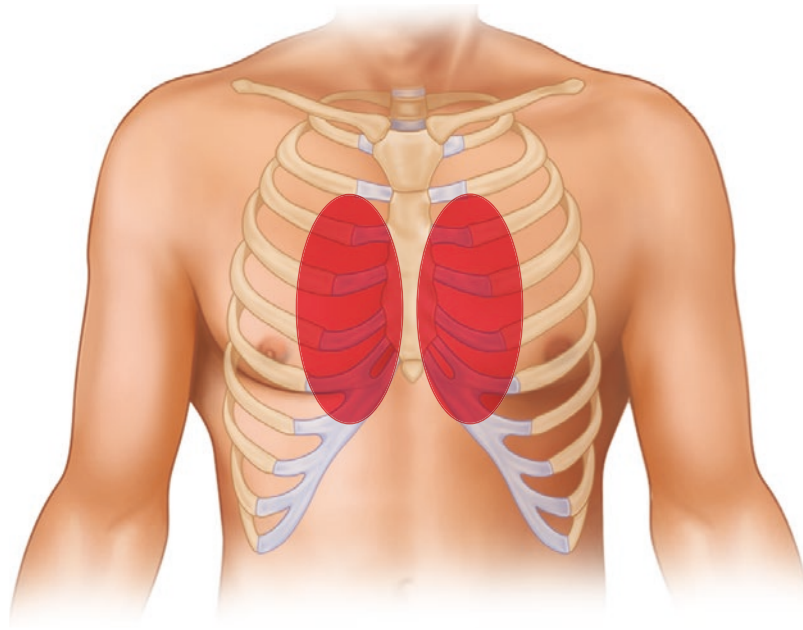

Fig. 36.2 Dissection of the pectoral muscles is then performed to obtain maximum lateral exposure of the sternum and the ribs. The wound edges are secured by wrapping them with gauze

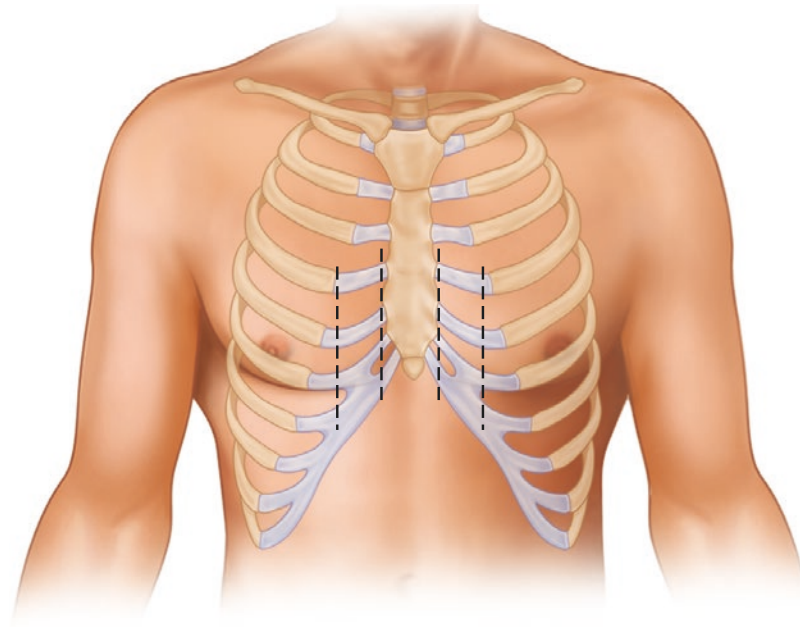

Fig. 36.3 Bilateral parasternal chondrotomy is then performed for which a scalpel is used to incise the perichondrium and 4 cm segments of cartilage are removed. The dissection proceeds from caudal to cranial with precaution taken to avoid injury of the pleura and the internal thoracic vessels

Postoperative Management

Perioperative antibiotic therapy is administered as described above with Ceftriaxone being the drug of choice. The wound dressing that has been applied in the operating room is left in place for 3 days. The subcutaneous wound drainage is removed on the second postoperative day. Postoperative analgesia includes administration of opioid drugs for at least the first 24 h. The patients are mobilized on the first day after surgery. Respiratory exercises under

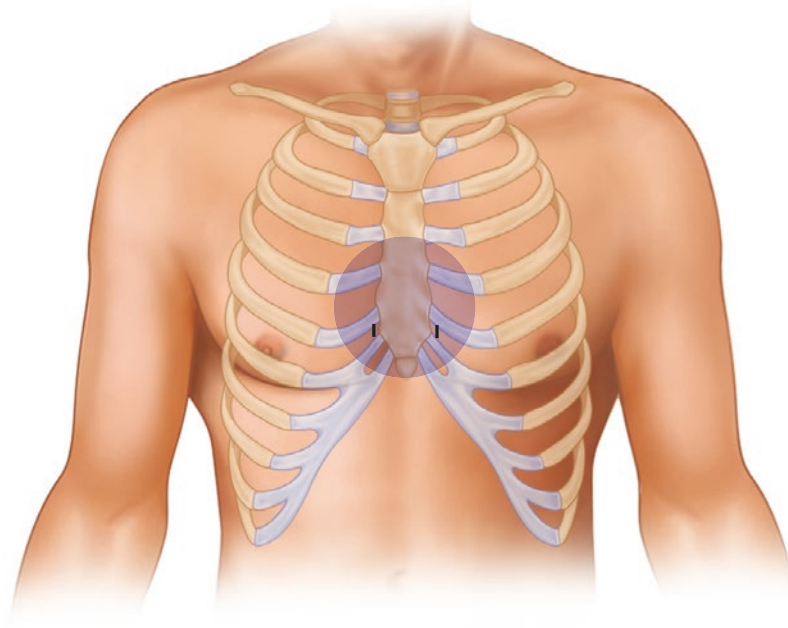

Fig. 36.4 The retrosternal tissue preparation is then performed. The xiphoid is secured with a clamp to provide retraction and facilitate digital tissue preparation. Precaution is taken to keep the pleura closed

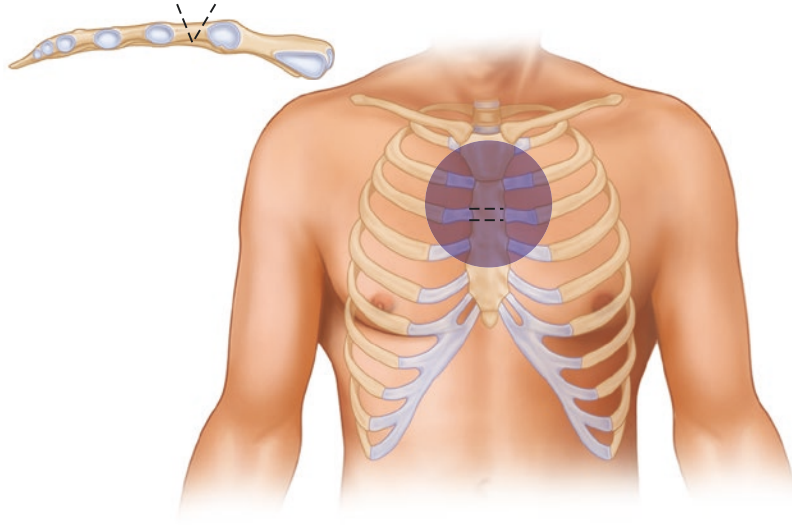

Fig. 36.5 Level of sternal osteotomy for at the cranial part of the sternum

physiotherapeutic guidance and with deep breathing exercising devices is also commenced on the first postoperative day. The patients are discharged from hospital as early as the third postoperative day.

The patients are followed-up at regular intervals (3, 6 and 12 months) after the procedure. Light exercises such as jogging is permitted 4 weeks after surgery and swimming is permitted after 3 months. Contact sports and excessive exercises are to be avoided during the 1st year after surgery.

The advantage of this modification of the procedure is that the bone graft is bioresorbable and does not have to be removed. Thus, a second procedure after correction of symmetric or asymmetric pectus excavatum deformities can be avoided. The mobilization of the sternum from

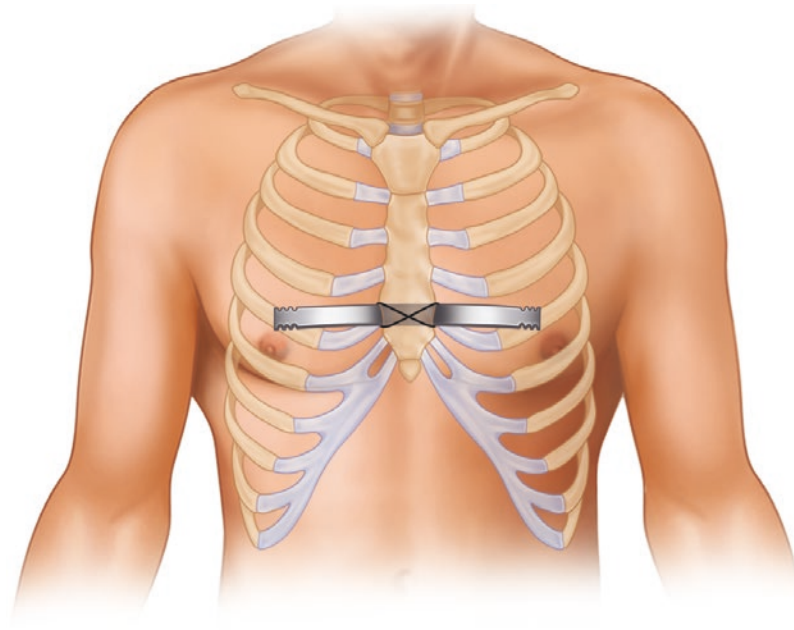

Fig. 36.6 The bone strut is inserted from left to right via the prepared retrosternal tunnel and is then tied to the sternum using interlacing absorbable heavy sutures

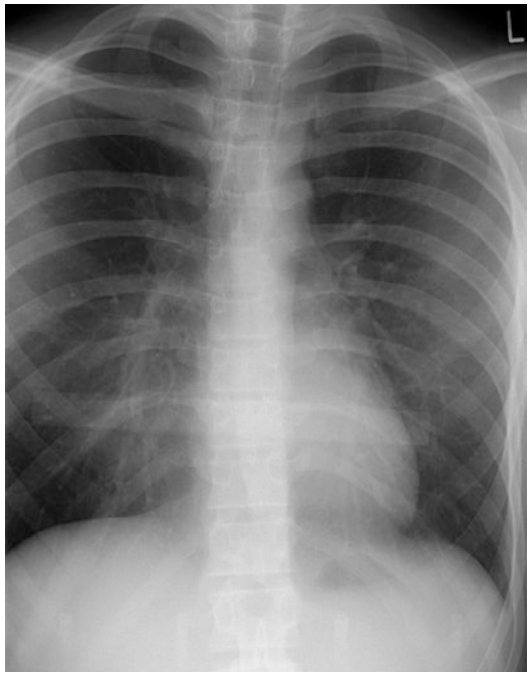

Fig. 36.7 The allogenic bone graft can be seen on a postoperative x-ray at the level of the 10th thoracic vertebra

the embedding ribs as described here is also suitable for the correction of pectus carinatum deformities. In adolescents and adults good long-term corrections can be achieved with a low rate of morbidity. Our patients are content with their operative results.

Suggested Reading

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2. Ravitch MM. The operative treatment of pectus excavatum. Ann Surg. 1949;129:429–44.
3. Ravitch MM. Congenital deformities of the chest wall and their operative correction. Philadelphia: WB Saunders; 1977. p. 145–58.
4. Willital GH. Indication and operative technique in chest deformities (author's transl). Z Kinderchir. 1981;33(3):244–52.

Welch Procedure

37

Juan C. de Agustín-Asensio

Technical Highlights

The Welch procedure is an open technique developed in 1958 to reduce the morbidity associated by the Ravitch procedure without reducing the efficacy for the correction of Pectus Excavatum.

The main technical highlights for this procedure are:

(a) Limited transverse sub-mammary skin incision within the nipple lines.
(b) Elevation of the pectoralis major muscle from the sternum.
(c) Elevation of portions of the pectoralis minor and serratus anterior muscles.
(d) Subperichondrial resection of the costal cartilages.
(e) Total preservation of the perichondrial sheaths.
(f) Incision of the perichondrium at junction to the sternum at a 90° angle.
(g) Division of cartilages at the junction of the sternum by knife.
(h) Welch perichondrial elevator are used to lift the cartilage.
(i) Avulsion of the cartilage.
(j) Elevation of the sternum at the junction of the xiphoid process.
(k) Division of the xiphoid process and the rectus muscles insertion.
(l) Anterior wedge sternal osteotomy above the level of the first deformed cartilage.
(m) Closure of the osteotomy with heavy silk sutures or titanium miniplates.
(n) Position of flat drain over the sternum.
(o) Pectoral muscle flaps are reattached to the midline of the sternal periosteum.
(p) Rectus muscles are reattached to the pectoral muscle flaps.

Indications

All types of Chest Wall Deformities can be corrected using this technique. However current indications for this technique are:

- Severely asymmetric Pectus Excavatum
- Severely asymmetric Pectus Carinatum
- Combined forms of Pectus Excavatum and Pectus Carinatum
- Chest Wall Deformities associated with Poland's Syndrome
- Other complex sternal malformations

Preoperative Considerations

1. The procedure should be offered and performed in patients >12 years of age as a general rule. Due to variations in body growth

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among teenagers, surgery should be reserved for teenagers whose body sizes have reached adolescent or adult proportions.
2. The procedure should be performed in girls only after demarcation of the breast contours. This is mainly to avoid potential breast deformation that may result from submammary skin incisions in females operated before reaching pubertal changes.
3. Adult patients of any age with relatively stiff chest wall are well suited for this procedure.
4. Chest radiographs are taken to access the extent of the deformity.
5. Lung function tests are performed to evaluate the pulmonary function.
6. Echocardiography and electrocardiograms to diagnose or rule out cardiac pathologies.
7. CT scan in severe thoracic asymmetry
8. Photographic documentation of the deformity
9. Coagulation tests are performed to rule out disorders in bleeding disorders
10. Blood reservation for auto-transfusion.
11. A single preoperative dose of antibiotics (cephazolin, 30 mg/Kg) is used as chemoprophylaxis.

Special Instruments

Besides the instruments used for general thoracic surgical procedures, the following instruments are additionally required for this procedure:

1. Strong towel clips for elevation of the sternum
2. Welch perichondrial elevators
3. Surgical oscillating saw (Aesculap)
4. Titanium miniplates and screws (Tarma)
5. Surgical drill (Aesculap)
6. Argon beam coagulator (Covidien)

Surgical Technique

The patient is in a supine position on the operating table with fully extended arms. No dorsal elevation is necessary for this procedure. A transverse sub-mammary skin incision is preferred below and within the nipple or clavicular midlines. Skin and subcutaneous fat is elevated from the muscular plane which should be enough to expose the pectoralis major and rectus muscle insertions. Parts of the pectoralis minor and serratus anterior muscles are elevated to gain access to the sternum and costal cartilages. Subperichondrial excision of the most evident malformed costal cartilages is performed. Bilateral excision from the third to the seventh cartilages is performed with care taken to preserve the perichondrial sheath. The pericondrium is dissected 90° in each direction at its junction with the sternum to facilitate visualization of posterior wall of the costal cartilage. The cartilages are divided at the junction of the sternum using a scalpel. Welch perichondrial elevators are used to facilitate this manoeuvre and protect the mediastinum. The divided cartilages are then held with an Allis clamp and avulsed from the costochondral junction. Avulsion of the cartilage produces more haemorrhage than clean cut by knife just before cartilage insertion. However, this leaves a thin cartilage pellet close to the costochondral junction.

Once the costal cartilages are removed, the rectus abdominis muscle insertions are divided from the xiphoid process sternum and the retrosternal space is created using blunt dissection. A sternal transverse osteotomy is performed just above the highest resected cartilage removing a small wedge of anterior cortex. An oscillating surgical saw is used to facilitate this manoeuvre. The cephalic and caudal fragments of the sternum are maintained in an overcorrected position using heavy silk sutures or titanium miniplates. These small plates are 20–30 mm long, and a single “T” shaped miniplate is used for children <8 years of age whereas 2 or 3 “I” shaped miniplates are preferred for the older patients. The plates are bent in a curve at their centre to create a 30° angle above the horizontal plane. Small screws fix the miniplates to the anterior surface of both sternal fragments. The miniplates are not removed and remain in-situ.

When sternal rotation is present, which is generally with a tilt to the right side, the sternum is completely transected with preservation of the posterior periosteum. In these patients, the miniplates are bent to lift the sternum from the more depressed side.

In children <10 years of age, screws of 9–10 mm length and 2.3 mm width are used. In older children and young adults the 12–13 mm long screws are preffered. Once the plate is fixed, irrigation with saline solution is performed and the perichondrial sheaths are closed.

Pectoral muscle flaps are secured to the midline of the sternal periosteum with absorbable sutures, after which the rectus abdominis muscles and the pectoral muscle flaps are reattached. A suction drain is placed at the presternal space after which the skin is closed using monofilament absorbable suture.

Postoperative Management

Perioperative antibiotic therapy is administered in a single dose or a maximum of three doses; with Cephazoline being the drug of choice. Alternating administration of intravenous metamizole and paracetamol at high doses is adequate to maintain good analgesia. Pain is less when there is good stabilization of the sternal fragments achieved during surgery. Patients are mobilized on the 2nd day after surgery and pulmonary exercises are commenced on the 4th day. Patients are kept in hospital mainly to treat pain and initiate respiratory therapy and are discharged when the drain tube is removed, usually at the 4th or 5th day. Patients are advised to avoid full body contact sports for 3 months. No sports restrictions are recommended after 6 months. The patients are followed-up at regular intervals (3, 6, 12 and then annually) after the procedure.

Results

The Welch technique is one of the most common procedures performed in paediatric thoracic surgery. The use of miniplates described in this chapter were introduced at our Centre in 1995 and have been used in 41 patients (24 males and 17 females) with a mean age of 10 years (4–20 year) with a follow-up of 12 years (10–15 years). Five of these patients were reoperations who were repaired in the past corrected with Welch-Ravitch operations. Our series had only one patient with one Marfan syndrome and the pectus deformity was corrected in this patient at the time of her cardiac transplantation. Another 3 patients had their Pectus excavatum repairs also simultaneously at the end of their cardiac malformation surgical repair (2 interatrial septal defect and 1 interventricular septal defect). With an exception to these patients, operations are not recommended in patients <13 years of age.

Assessing our patients, the depth of the defect in supine position was 2.45 ± 0.51 cm (mean ± SD) and 35% showed right sternal rotation with a Shamberger-Welch Pectus index was 6.25 ± 0.91 and the Haller Pectus Index of 5.4 ± 2.98.

In this series, the miniplates have maintained their position and only three severe and four mild recurrences have been observed. Thirty-four patients had excellent results following repairs with this method. In all the patients, the cranial part of the sternum demonstrated some degree of depression over time, which was a consequence of sternal anterior angle overcorrection at the level of the miniplates. Four patients had an anterior displacement of a single screw from the proximal part of the sternum; however since these screws were palpable beneath the skin they were removed under local anaesthesia in the outpatient clinic. Two miniplates fractured, but that did not influence the long-term results of the operation. There were no major complications. Pain was reduced when the sternal fragments were compact and secured. Drains were in place for a mean of 3.5 ± 2 days; however two patients presented with seroma formation which were aspirated with a syringe in the outpatient setting. The mean hospital stay was 5 ± 1.8 days.

One of the main differences with the techniques that use permanent AO plates is that these plates could interfere with the growing sternum. Our technique with small titanium miniplates bent with 30° angle have been found to have the advantage as these small plates fix a limited portion of the sternum through the osteotomy site, and hence they do not interfere with the growing sternum.

Welch technique, with or without titanium miniplates, is very effective in correcting the asymmetric and complex deformities, because the sternum can be modelled to restore the horizontal level very efficiently (Figs. 37.1, 37.2, 37.3, 37.4, 37.5, 37.6, 37.7, 37.8, 37.9, 37.10, 37.11, and 37.12).

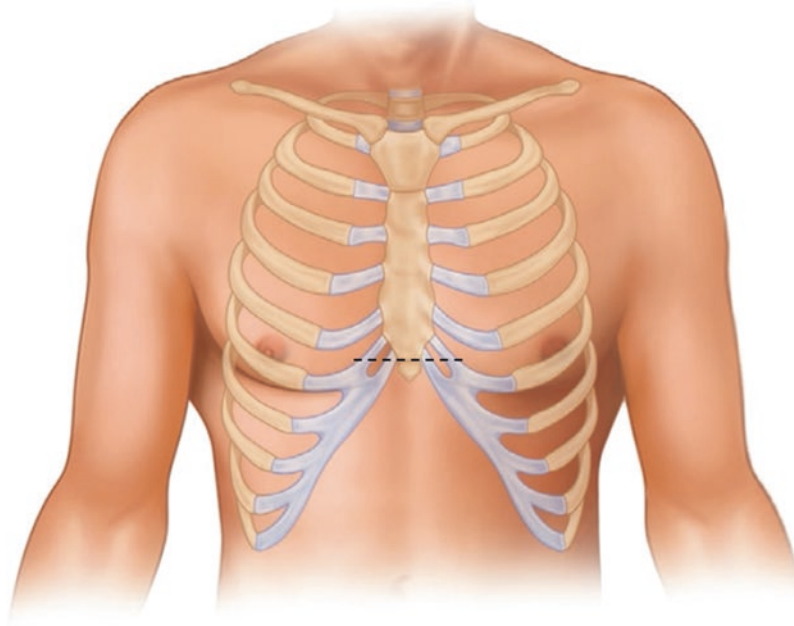

Fig. 37.1 Limited transverse submammary incision between clavicular midline along subcutaneous dissection. The wound edges are secured with towels hitched to subcutaneous tissue

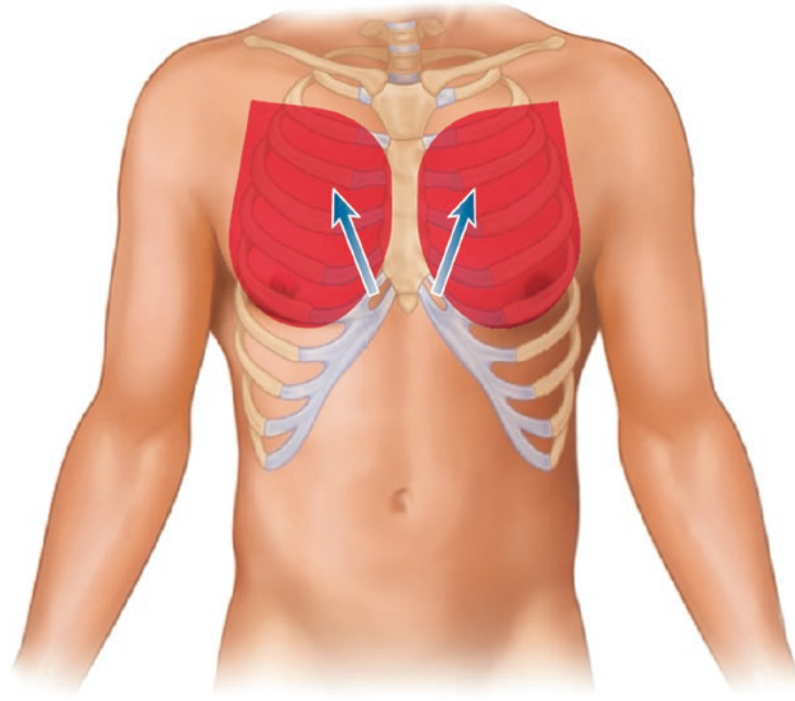

Fig. 37.2 En-bloc dissection of the pectoralis muscles is then performed to obtain enough lateral exposure of the sternum and the ribs

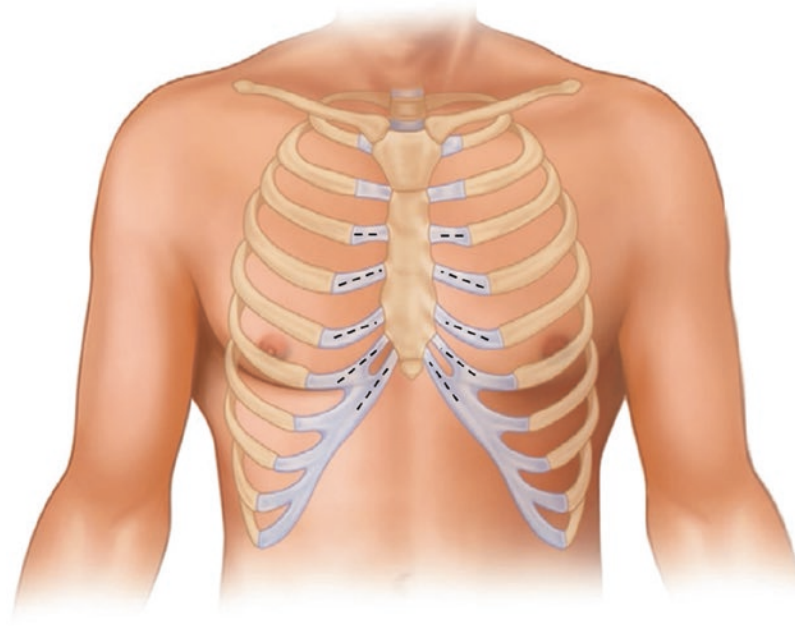

Fig. 37.3 Bilateral parasternal total chondrotomy is performed from the third to the seventh ribs. The cautery is used to incise the perichondrium and Welch elevators used to isolate the cartilages, preserving the pericondrium. Precaution is taken to avoid injury to the Internal Thoracic Vessels

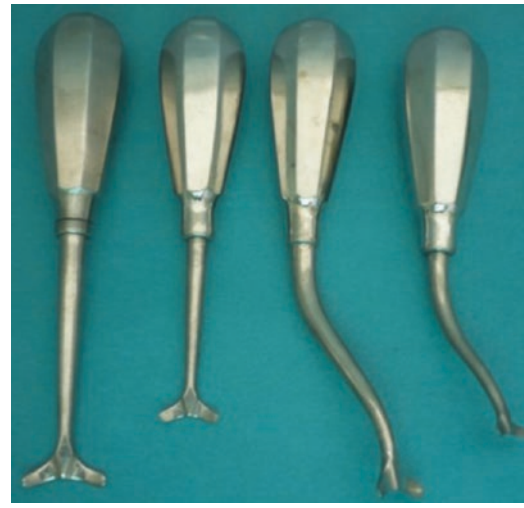

Fig. 37.4 Welch elevators facilitate perichondrial dissection. Another dissectors can be also used, as the double face Obwegeser freer periosteal elevator

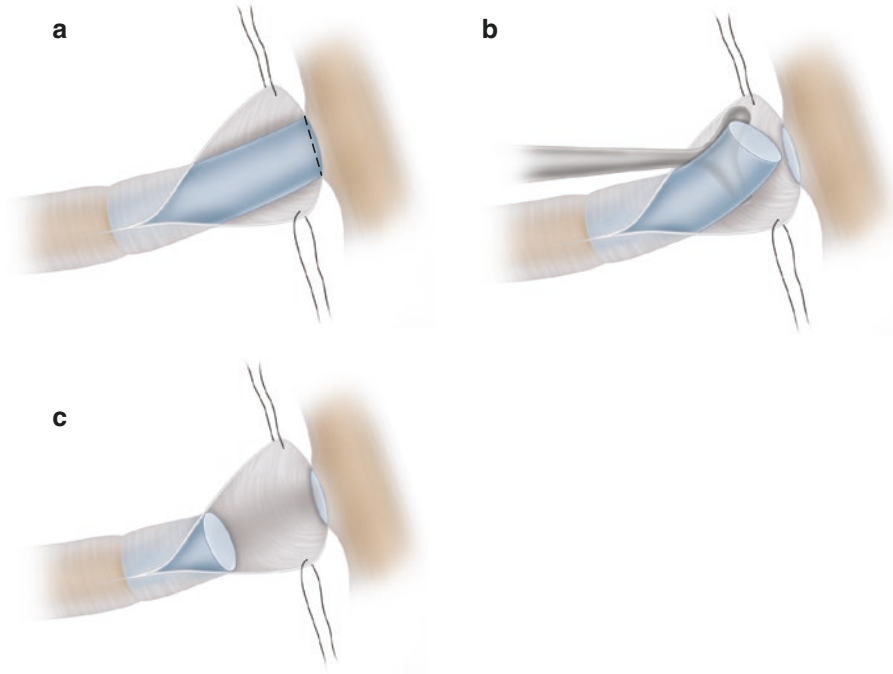

Fig. 37.5 Bloodless plain between perichondrium and costal cartilage is found by cutting the perichondrium 90° upwards and downwards at its juction to the sternum (**a**). Welch elevators facilitates this disection and protects the section of the costal carilage to the sternum (**b**). Cartilage is elevated by an Allis clamp and finally cut at its juction at the costal edge (**c**)

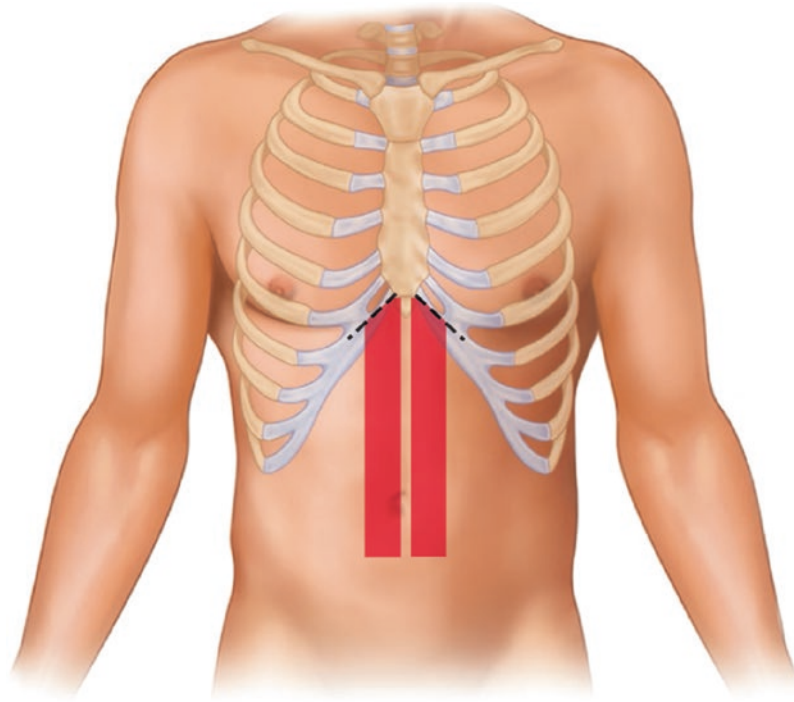

Fig. 37.6 The Xiphoid and rectus muscle insertions are severed from the sternum

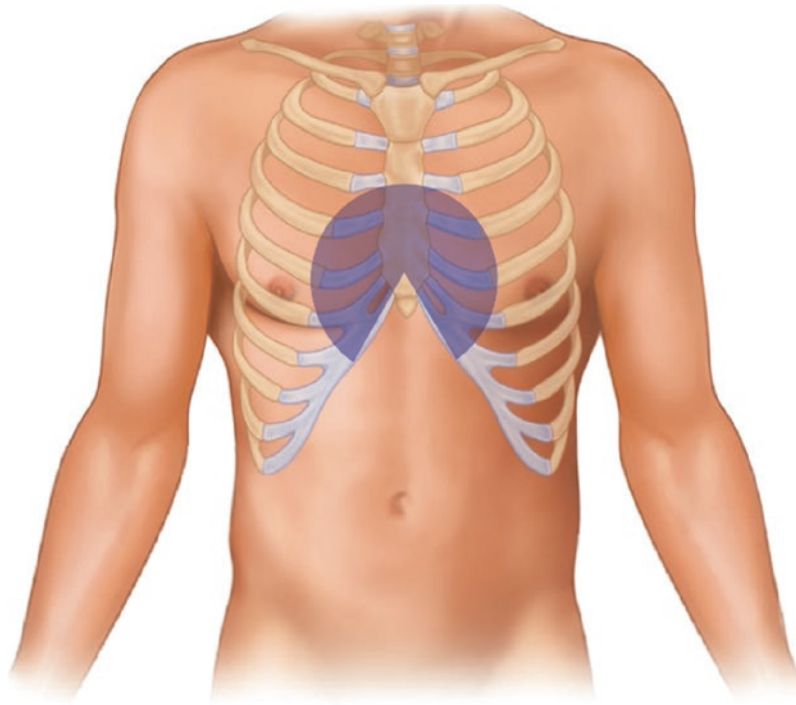

Fig. 37.7 The sternum is elevated to facilitate digital retrosternal dissection. Precaution is taken to keep the pleura closed

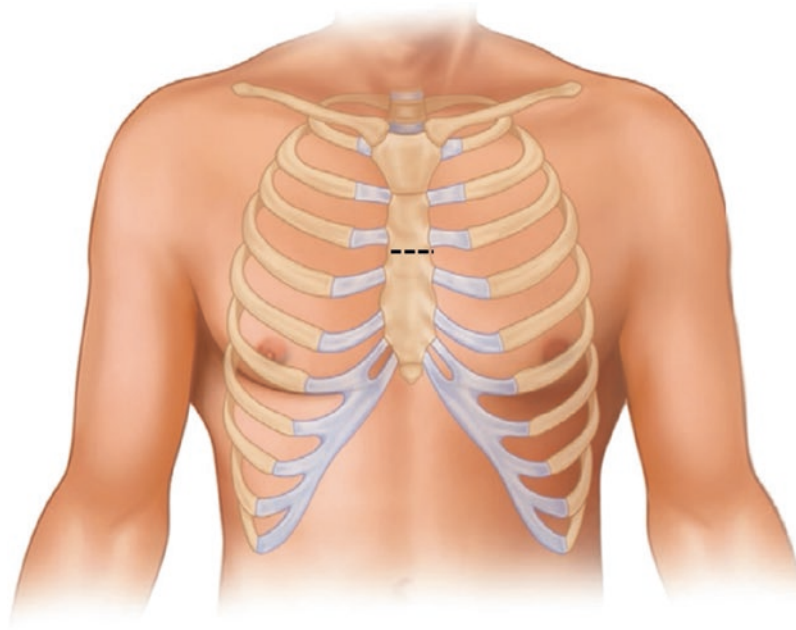

Fig. 37.8 Wedge sternal osteostomy is performed above the level of the first deformed cartilage. The sternal fragments are secured using braided non-absorbable suture or titanium miniplates

Fig. 37.9 Two different titanium miniplate outlines are used to fix the sternal fragments. The "T" miniplate is used in children under 8 years old. Two or three straights minplates are required for older children (From de Agustín-Asensio et al. [1]. With permission from Elsevier)

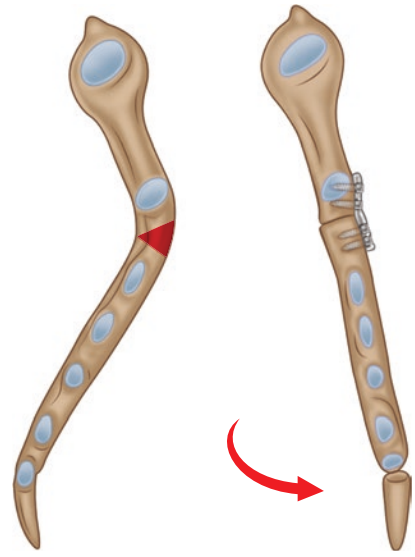

Fig. 37.11 Change on the sternal position after osteotomy and further fixation of the sternal fragments (From de Agustín-Asensio et al. [1]. With permission from Elsevier)

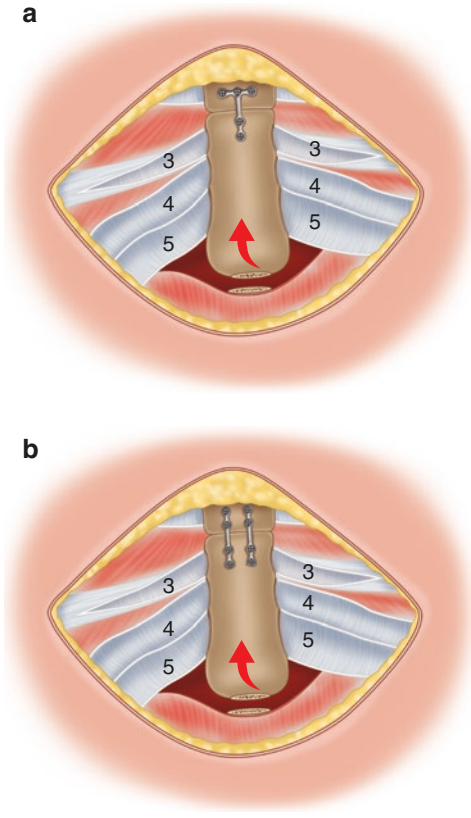

Fig. 37.10 Frontal view of the corrected position of the sternum using (**a**) one and (**b**) two titanium miniplates (From de Agustín-Asensio et al. [1]. With permission from Elsevier)

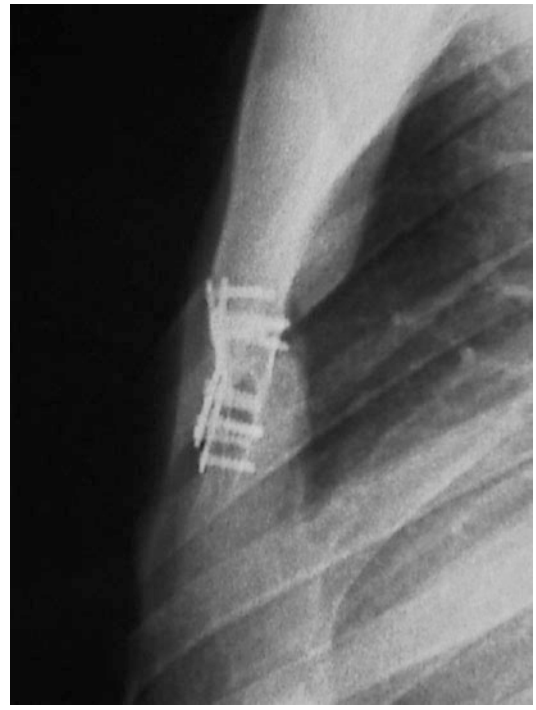

Fig. 37.12 Postoperative lateral view of a corrected position of the sternum using three titanium miniplates in an 18 year old child

Suggested Reading

1. de Agustín-Asensio JC, Bañuelos C, Vázquez JJ. Titanium miniplates for correction of pectus excavatum. J Am Coll Surg. 1999;88:455–8.
2. Schamberger RC, Welch KJ. Surgical repair of pectus excavatum. J Pediatr Surg. 1988;23:615–22.
3. Welch KJ. Satisfactory surgical correction of pectus excavatum deformity in childhood: a limited opportunity. J Thorac Surg. 1958;36:697–713.

The Willital-Hegemann Procedure

38

Günter H. Willital and Amulya K. Saxena

Technical Highlights

The Willital-Hegemann procedure is an open technique for the correction of the various forms of Chest Wall Deformities (CWD).

The main technical highlights of this procedure are:

(a) Sagittal incision in males and sub-mammary incision in females
(b) Dissection of the rectus muscle to release the Xiphoid process
(c) Dissection of the pectoral muscles to expose the sternum and ribs
(d) Bilateral parasternal chondrotomy to detach the sternum from the ribs
(e) Bilateral parasternal rib fractures in severe forms deformities
(f) Sternal wedge osteotomy and stabilization of the sternum using sutures
(g) Using of 1 trans-sternal metal strut and 2 parasternal struts to stabilize the rib cage
(h) Fixation of the metal struts at the culmination points with stainless steel wires
(i) Placement of chest tubes
(j) Re-anastomosis of sternum to the rib cartilages
(k) Stabilization of the fractured ribs using figure-of-eight sutures
(l) Suture fixation of the rectus muscle to the sternum
(m) Suture fixation of the pectoral muscles

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Indications

All types of Chest Wall Deformities can be corrected using this procedure:

- Severely asymmetric Pectus Excavatum
- Severely asymmetric Pectus Carinatum
- Combined forms of Pectus Excavatum and Pectus Carinatum
- Chest Wall Deformities associated with Poland's Syndrome

Preoperative Considerations

1. The procedure is offered in patients >12 years of age as a general rule.
2. The procedure is preferably performed in girls after demarcation of the breast contours.
3. Adult patients of any age are well suited for this procedure.

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4. Chest radiographs are taken to access the extent of the deformity.
5. Electrocardiograms to rule out cardiac pathologies.
6. Video-stereo-raster investigations.
7. Chest perimeter plots to quantify the deformity at three different points: manubrium, sternum and xiphoid.
8. Measurement of thorax diameter using a pelvimeter
9. Photographic documentation of the deformity
10. Coagulation tests are performed to rule out disorders in bleeding disorders
11. Careful patient history is taken to rule out nickel allergy.
12. Antibiotics are administered on the day of the procedure and are continued for a period of 7 days after the procedure. Ceftriaxone is the choice of antibiotic.

Special Instruments

The instruments used for thoracic surgical procedures are required. Additional instruments required for this procedure (see Chapter on Instruments):

1. Stainless steel metal struts (Karl Lettenbauer GmbH, Erlangen, Germany)
2. Surgical toothed Pliers (Karl Lettenbauer GmbH, Erlangen, Germany)
3. Surgical Spanners (Karl Lettenbauer GmbH, Erlangen, Germany)
4. Stainless steel wires

Surgical Technique

A vertical midline incision 15–20 cm long is preferred in males, whereas a submammary incision is opted in females. In the incision, skin, fat and pectoral muscles are prepared as a single flap for which the dissection is performed using a needle-tipped electrocautery. The costal cartilages are exposed and parasternal wedges of the deformed cartilages are resected after placing a perichondral elevator around the cartilage and incising it with a scalpel. Bilateral cartilage blocks are resected as the dissection proceeds cranially successively freeing the sternum from its caudal to cranial attachments. Dissection is ceased once sufficient sternum has been freed from the distal costal cartilages, while still remaining attached on the cranial end. Precaution should also be taken to avoid injury to the internal mammary vessels. The attachment of the rectus abdominis muscle to the xiphoid is exposed and the rectus muscle is severed to free the xiphoid. The attachment of rectus abdominis muscle to the lower costal cartilages is also dissected free. The xiphoid is grasped using a Kocher clamp and retracted. Under traction, the sternum is freed from its anterior mediastinal attachments using careful digital preparation. The deformed costal cartilages are additionally resected at the level of transition to the normal ribs to obtain acceptable thoracic contours.

A partial transverse sternal wedge osteotomy is performed using a *Luer Bone Ronguer*. Once the sternum is dissected free, a perforated Hegemann steel strut (Lettenbauer, Erlangen, Germany) is passed through the sternum. To facilitate this, the distal part of the sternum is elevated above the level of the thorax so as to avoid injury to any intra-thoracic structure while passing the strut through the sternum which is inserted using a hammer. The strut is then bent with spanners so that it fits the thorax wall perfectly at the edge of the impression. On achieving the desired shape, the strut and the sternum are returned back into the thoracic cavity. Two parasternal metal struts are also employed, with the points of attachment being the second rib and the lowest end of the rib cage. Heavy sutures are used to suture the cartilages as well as the trans-sternal strut to the ribs. Two parasternal struts are employed to provide anchorage to the lateral flail chest segments, which are formed as a result of double bilateral chondrotomy. The trans-sternal strut is secured to the two parasternal struts using stainless steel wires. Chest tubes are placed before completion of the procedure. The pectoral muscle flaps and the severed rectus muscles were then sutured and fixed to the sternum. The

overlying subcutaneous and cutaneous structures are closed in the conventional manner with interrupted subcutaneous sutures and monofilament non-absorbable running skin suture.

Procedure Overview (Figs. 38.1, 38.2, 38.3, 38.4, 38.5, 38.6, 38.7, 38.8, 38.9, and 38.10)

Postoperative Management

Perioperative antibiotic therapy is administered; with Ceftriaxone being the drug of choice. All patients are also administered strong analgesics for 72 h. Chest drains are removed on the 3rd day after surgery. On the 4th day pulmonary exercises are commenced. Patients are kept in hospital to follow pain and recovery after this procedure. At the time of discharge patients are advised to avoid body contact sports until the struts had been removed. Regular swimming is encouraged along with light athletic exercises. The patients are followed at regular intervals (3, 6, 12 and 15 months) after the procedure. The struts are removed after a period of 15 month.

Results

The Willital-Hegemann procedure was carried out over a 20 year period on 1262 patients with pectus deformities (968 male and 294 female patients). The corrections were completed with successful repair in 1244 (98.6 %) patients, along with a low complication rate of 5.7 %. The median age of the patients was 14.9 years (range, 2–53 years). The follow-up period ranged from 2 to 12 years (mean, 5.4 years). Major recurrences were observed in 18 (1.4 %) patients, and mild recurrences were observed in 46 (3.6 %) patients. The struts were removed after a period of 24–36 months and were associated with a complication rate of 2.6 % at the time of removal.

Custom-tailored molding of the chest wall can be achieved by using this method, which is not possible with minimal-access techniques. Open repair is effective for all variations of chest wall deformities and in patients of all ages, causes only mild pain after the procedure but no prolonged episodes of pain as seen in minimal-access techniques, and produces good physiologic and cosmetic results. Improvement of subjective complaints, satisfactory long-term results, and improvement in psychological problems indicate the need to offer this procedure among other surgical correction options for low-risk children.

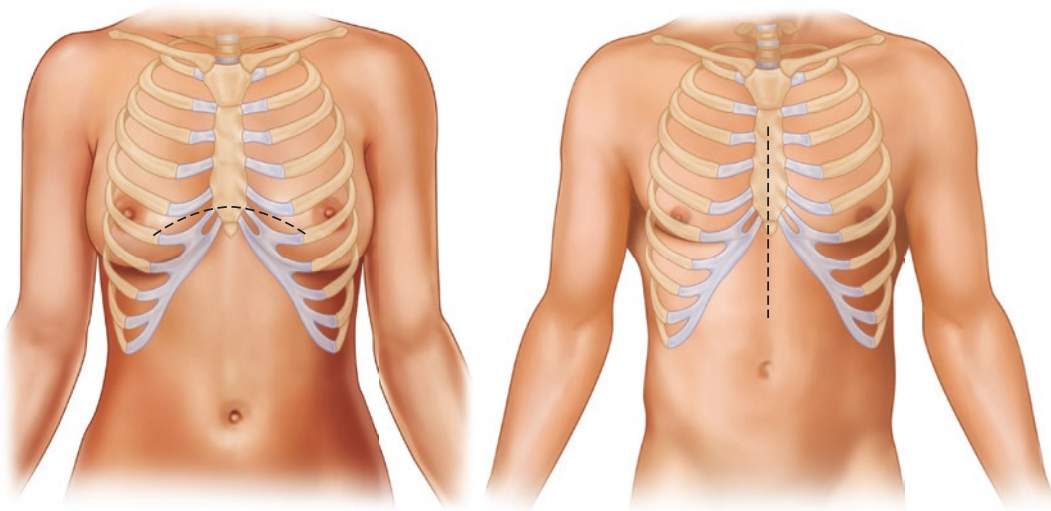

Fig. 38.1 Submammary incisions is preferred in females (*left*) and vertical midline incisions in males (*right*)

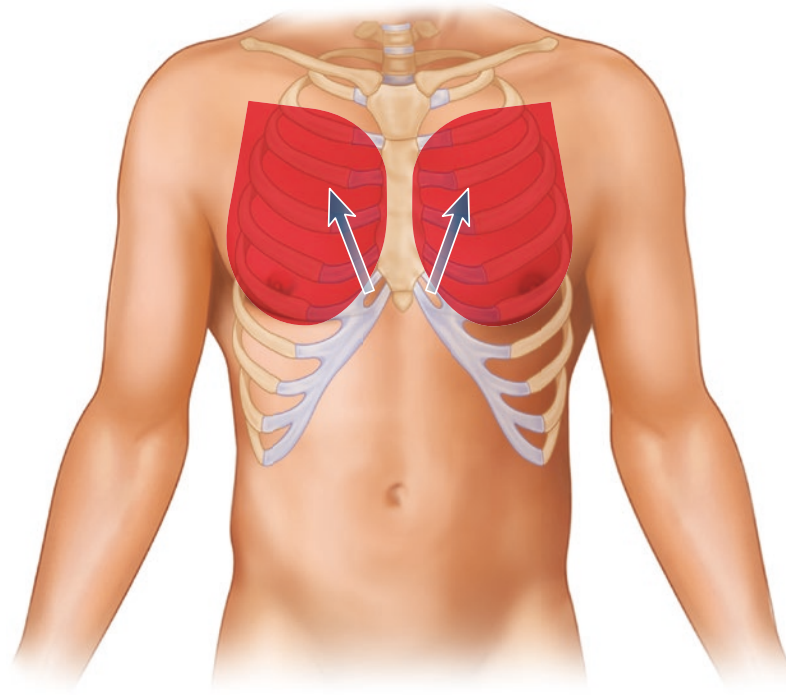

Fig. 38.2 Dissection of the pectoralis muscles to allow lateral exposure of the sternum and the ribs. The wound edges are secured with towels hitched to subcutaneous tissue

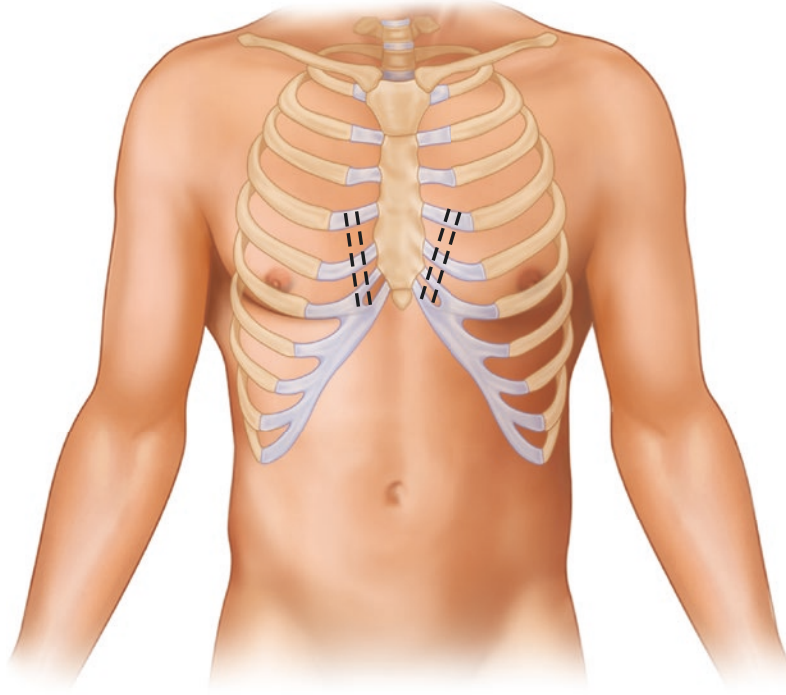

Fig. 38.3 Bilateral parasternal chondrotomy is achieved with the use of an electrocautery to incise the perichondrium and the removal of 1 cm segments of cartilage. The dissection proceeds from caudal to cranial with precaution taken to avoid injury to the Internal Thoracic Vessels

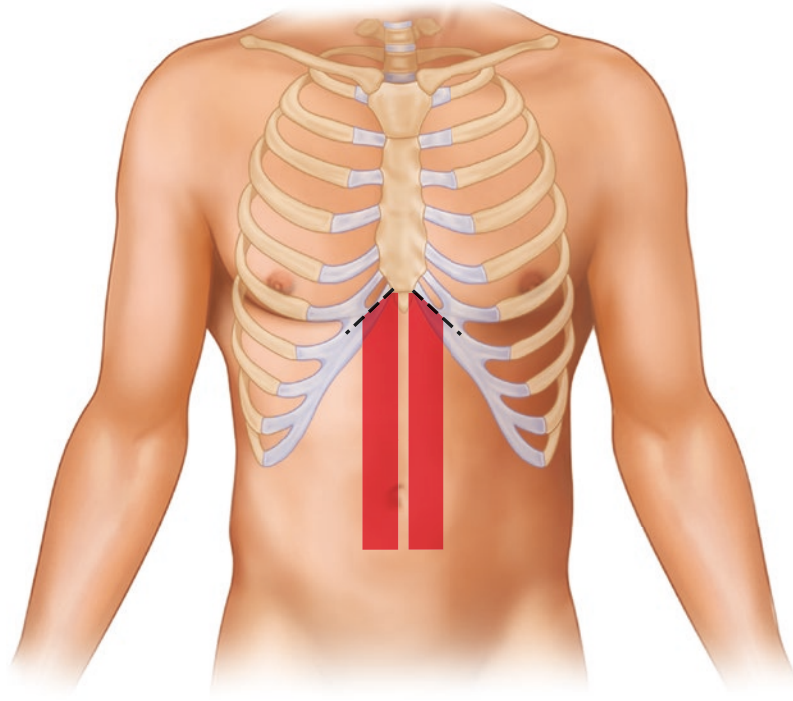

Fig. 38.4 The rectus abdominis muscle is detached from the sternum to expose the xiphoid. The dissection is enlarged along the length of the lower ribs

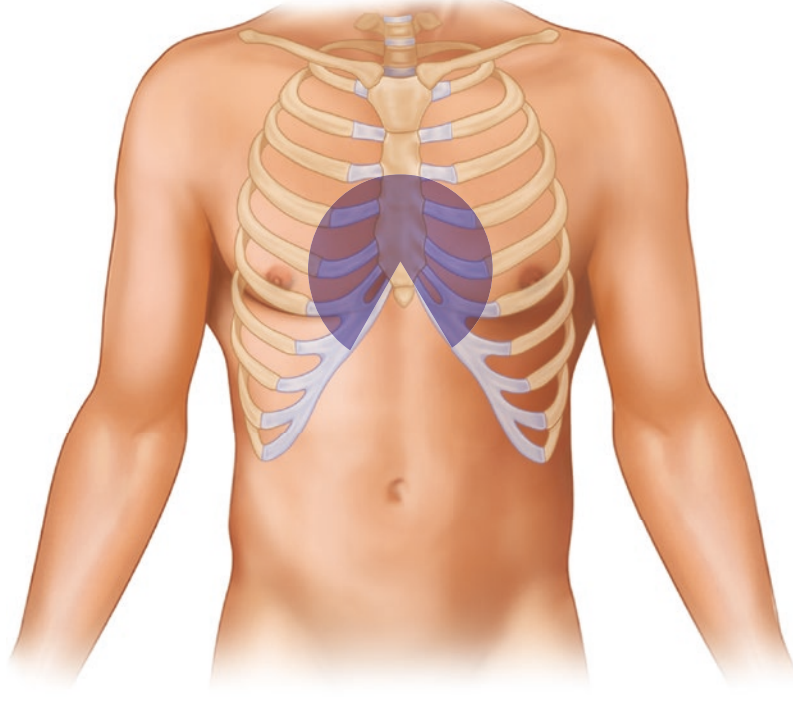

Fig. 38.5 A retrosternal space is then prepared aided by the xiphoid secured with a clamp to provide retraction

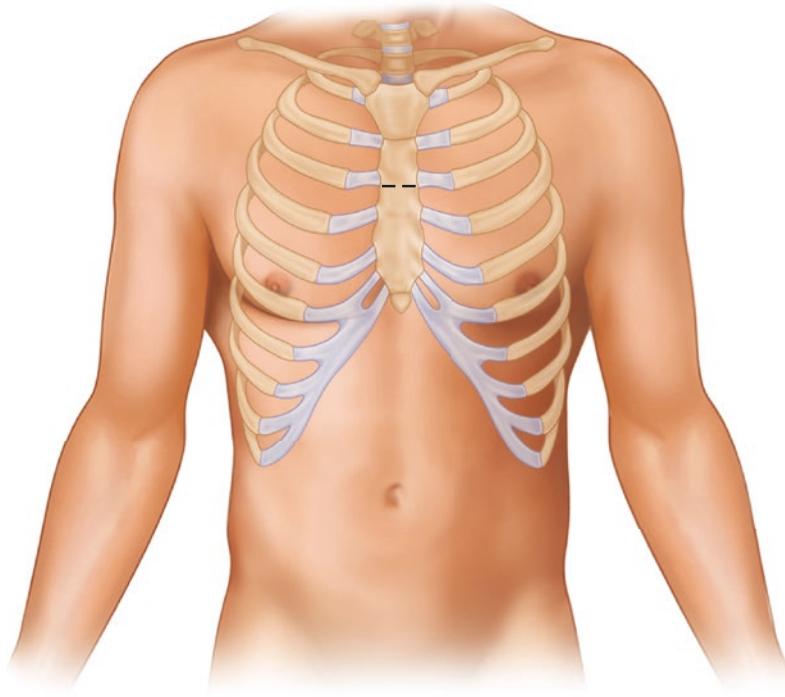

Fig. 38.6 Partial wedge sternal osteostomy is performed. The edges of the sternum after partial osteotomy are secured using heavy absorbable suture

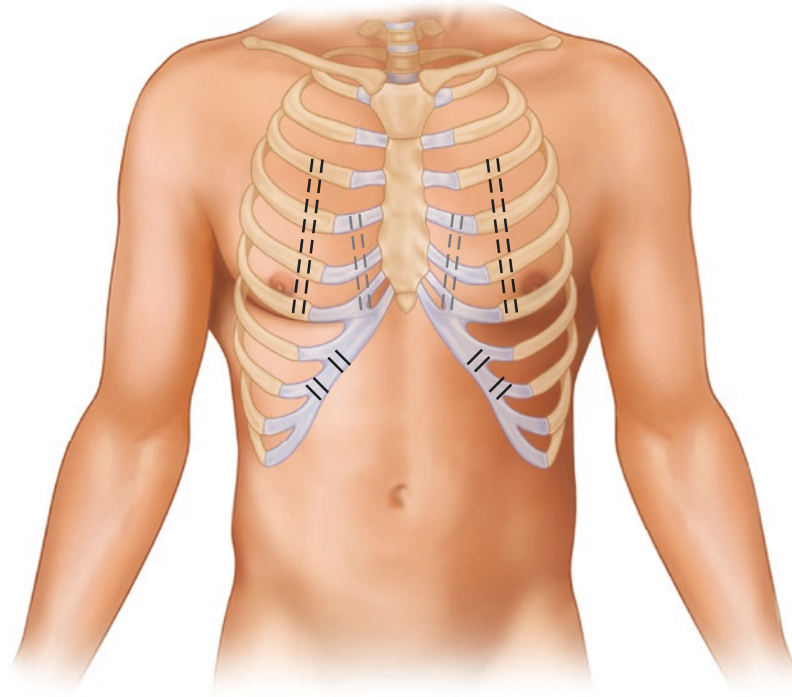

Fig. 38.7 Ribs are fractured in patients with platythorax or lower rib eversions. Lower rib eversions are corrected by chondrotomy and re-anastomosis

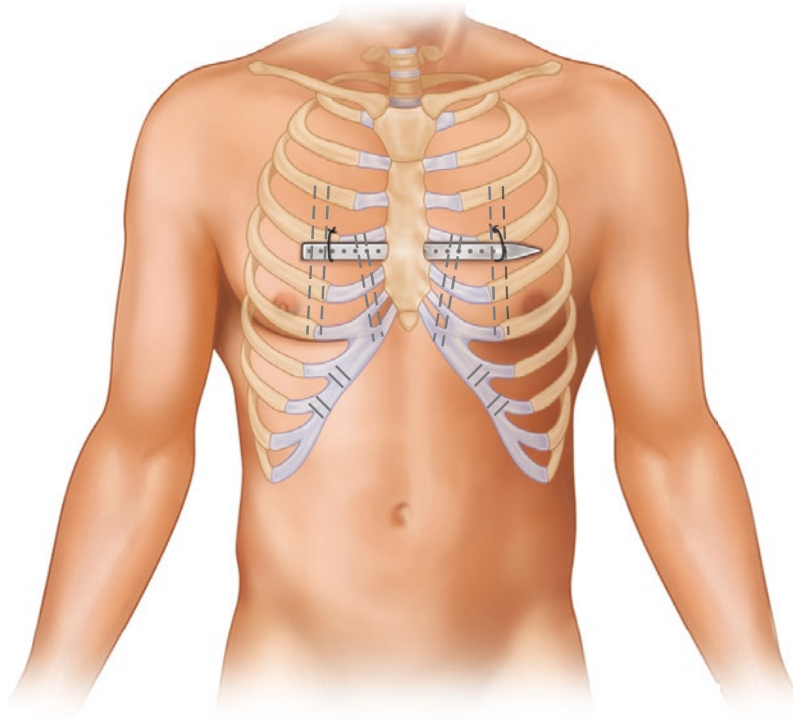

Fig. 38.8 A metal strut with a sharp and pointed edge is passed through the sternum using a hammer. Once positioned the sharp edge of the strut is cut using pliers

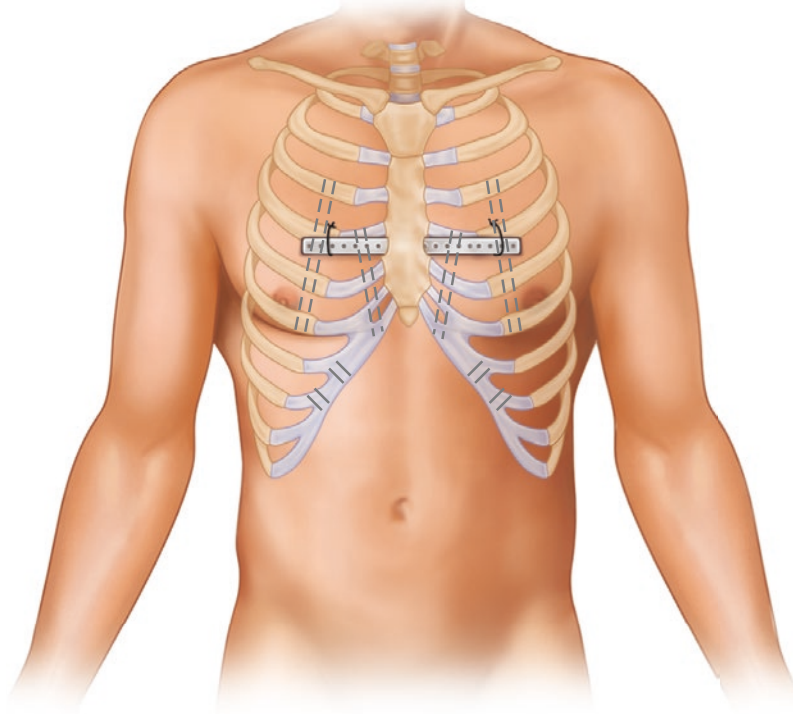

Fig. 38.9 The transsternal strut is secured to the underlying rib using heavy absorbable sutures, which are also used to re-attach the cartilages to the sternum

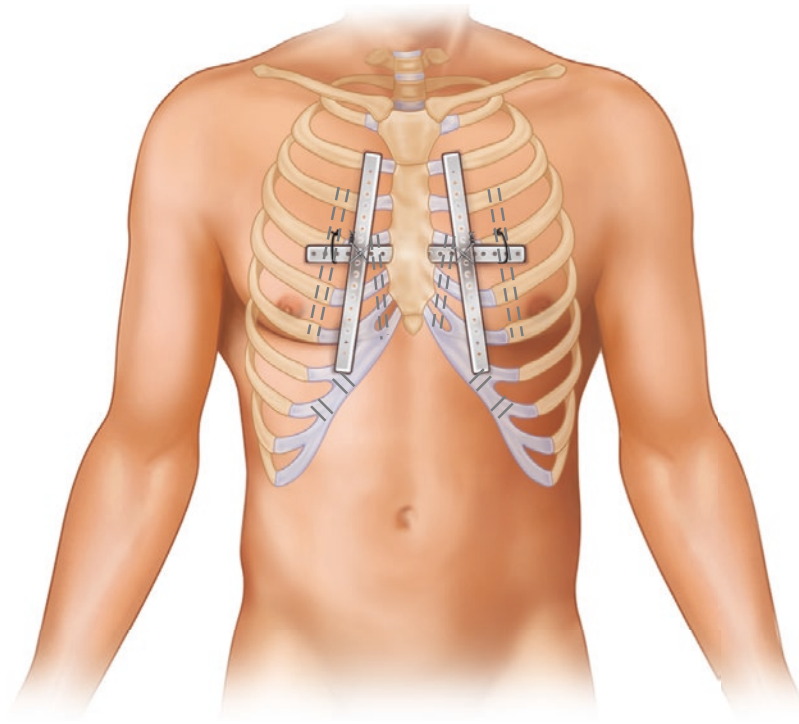

Fig. 38.10 Longitudinal struts are placed on the flail chest wall segments to which they are secured with heavy absorbable sutures. Stainless steel wires are used to secure the parasternal struts on the culmination points to the trans-sternal struts

The follow up period ranged from 6 months to 12 years. The struts were removed after a period of 15 months and no recurrences after strut removal were observed. Hypertrophic scars were corrected at the time of strut removal. All patients were followed up 3, 6 and 12 months after the procedure. The follow up investigations included (a) measurement of thorax diameter using a pelvimeter (b) electrocardiogram (c) pulmonary function tests, (d) chest ultrasound, (e) clinical photograph and video-stereo-raster examinations.

Procedure Related Images
(Figs. 38.11, 38.12, 38.13, 38.14, and 38.15)

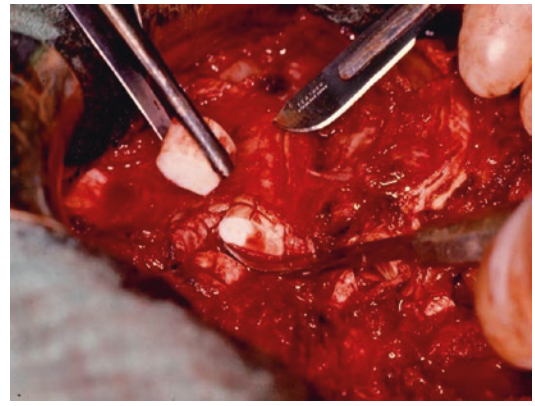

Fig. 38.11 En-bloc bilateral resection of costal cartilages is performed using a scalpel with removal of cartilages from the caudal to cranial direction

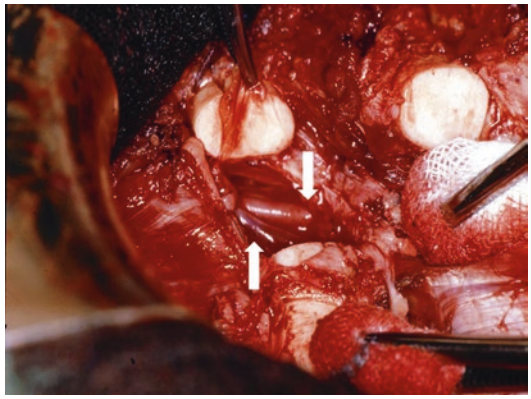

Fig. 38.12 Precaution should be taken to identify the internal thoracic vessels (white arrows) which run just below the line of dissection so as to avoid injury to them during cartilage resection

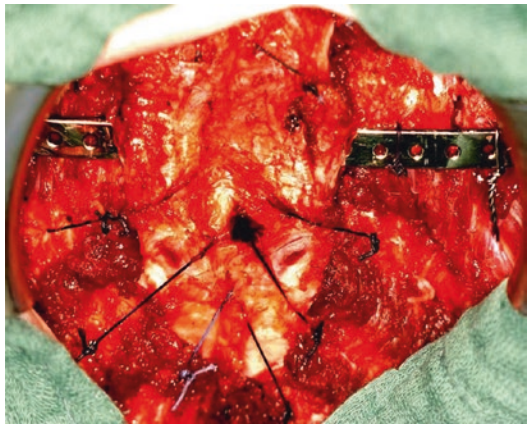

Fig. 38.13 Images showing the trans-sternal strut in position. The caudal end of the sternum is sutured to the adjacent ribs using heavy absorbable suture

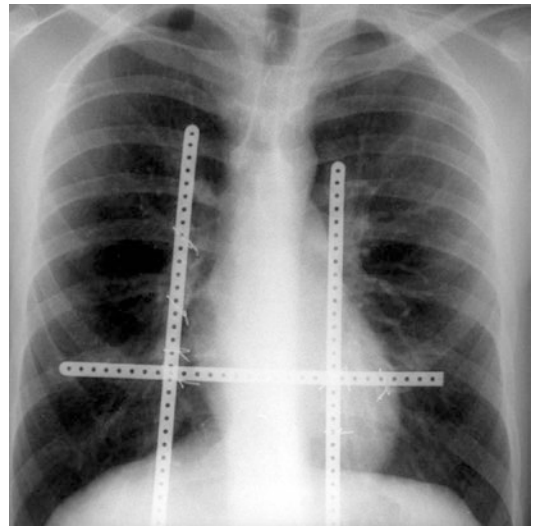

Fig. 38.14 Chest film demonstrating the position of the struts. Multiple stainless steel wires were used in the first decade of the series. During the second decade stainless steel wires were employed to secure the struts at the culmination points

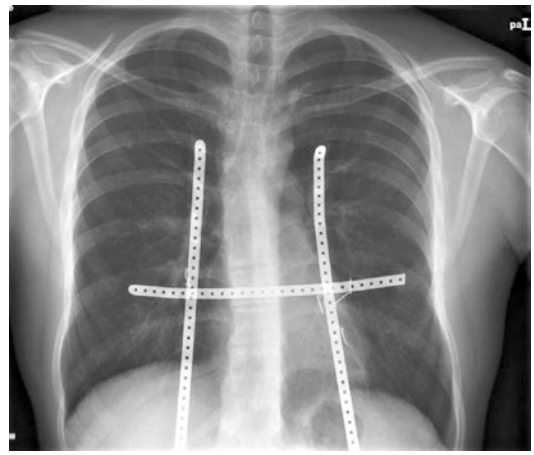

Fig. 38.15 Chest film demonstrating the position of the struts. Multiple stainless steel wires used in the first half of the series, whereas stainless steel wires were employed to secure the struts at the culmination points in the second half of the series

Suggested Reading

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Minimal Access Repair of Pectus Excavatum (MARPE)- Pilegaard Modification

39

Hans K. Pilegaard

Technical Highlights

Minimal Access Repair of Pectus Excavatum (MARPE) modified by Pilegaard [7, 8] is a minimal access procedure to correct pectus excavatum by using small incisions and short pectus bar (s).

The main technical highlights of this technique are:

(a) Only right side usage of a scope is preferred in this technique.
(b) The deepest point under the sternum is identified
(c) The position (entrance and exit points) of the bar through the chest wall is defined
(d) Bilateral small skin incisions, which in females are positioned in the sub-mammary sulcus
(e) If the pectus bar is placed above the level of the nipples, the left incision might be periareolar
(f) A proper size introducer is used to ensure close contact to the chest wall.
(g) Bars are normally placed subcutaneously, but if a cranial positioned bar is used, it is placed beyond the pectoral muscle.
(h) The pectus bar is placed above two ribs on each side. This is mandatory on the side with no stabilizer
(i) The pectus bar is secured to the stabilizer with a steel wire or by simple bending of the pectus bar end to fit snug to the chest wall.
(j) The skin is closed in two layers

Indications

This technique can be used to correct pectus excavatum without an upper age limit [6]. The indications are esthetic concerns, symptoms of decreased capacity under physical activity and a Haller index > 3.25.

- All forms of pectus excavatum

Preoperative Considerations

1. The operation should be offered the patient after the initialization of the growth spurt just in the beginning of the puberty to have the best esthetic result (11–13 years old).
2. In girls, it is an advantage to have a demarcation of the breast contours, because the skin incisions then can be placed correctly in the submammarian sulcus.
3. Adults should be consented as they often need two bars.

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A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_39

4. The depth of the pectus deformity, is measured using a slightly bended template, and should be more than 3 cm in adults in symmetric cases.
5. It is mandatory to obtain chest films before surgery
6. If there is a suspicion of a cardiac condition (e.g., Marfan's syndrome), a Computed Tomogram (CT) and echocardiography should be performed.
7. Documentation by photos of the deformity before and surgery.
8. Although the bar contents nickel, it should not cause any symptoms as nickel causes contact skin allergy and not when implanted.
9. Antibiotics are administered prior to surgery and continued for 3 days. Cefuroxime and Gentamycin are the antibiotics of choice.
10. An epidural catheter is placed in every patient if possible and removed after 2 days
11. Single lumen tube is used and removed on the table. CO_2 insufflation is not necessary.

Special Instruments

Along with the instruments used for general thoracic surgery, the following instruments are required:

1. Pectus bar
2. Orthopedic metal plate bender press
3. Pectus introducer
4. Pectus bar bender
5. Pectus bar flipper
6. Bar stabilizer plate
7. Cotton umbilical tape
8. Scope for video assisted thoracic surgery (5 mm, 30°)
9. Port with blunt trocar tip and insufflation channel 5 mm
10. Endoscopy tower/suite

Surgical Technique

The patient is placed in the supine position on the table, approximated to the right edge of the operating table to facilitate free movement of the scope. The right arm is placed above the head, with a small pillow under the shoulder to support it. A 5-mm 30° thoracoscope is used and is placed just below the midaxillary line around the level of the nipple. The deepest point under the sternum is defined. The points for entrance and exit of the bar through the chest wall should be just medial to the highest points of the excavation. These points are marked at the skin and the right point may be checked by pressing at it with a fingertip and looking in the scope if it is in the line of the deepest point.

If the depression is excessively deep it may be necessary to use two bars. In such a case, the location of the bar under the deepest point will be facilitated if the first introducer is placed more cranially and left in position while the next introducer is inserted under the deepest point. This makes it easy to replace the first introducer with a pectus bar which is followed by the second introducer replacement with a pectus bar.

A template is used to determine the correct shape of the pectus bar. The pectus bar should have sufficient length so the opposite end where the stabilizer plate is positioned covers two ribs, which means that the bar often is placed asymmetric on the chest wall. Because the bar will be straightened by the force of the sternum, the shape of the template should overcorrect the excavation. The bar is bent in the same shape as the template, but care should be taken not to bend the end where the stabilizer plate will be positioned too much. The bar is now placed on the anterior chest wall to mark the ends where the incisions should be made.

Once the incisions have been made, the introducer is inserted under the guidance of the scope. The large introducer may be used in most cases but if there is uncertainty about the tip not being in close contact with the posterior wall of the sternum/chest wall, it should be changed for the x-large introducer. When the tip of the introducer has passed the deepest point, there is only a very short distance to the point of exit through the chest wall. Elevate the chest wall by lifting the introducer and press on the curvatures to make a sort of pre-correction. If the x-large introducer is used it is advisable to insert either a hook or a steel wire through

the introducer eyelet to pull the long tip through the chest wall before the introducer is pushed through the patient.

To guide the bar through the chest a suture or a tape is used which is attached to the eyelet of the introducer before it is removed. The bar is placed like the shape of U and then rotated 180° with the use of one or two flippers. The stabilizer is placed on one end and preferrably on the left side and fixed to the bar either with a steel wire around the bar or by bending the end more by the flipper if the end of bar is not close to the chest wall.

The air in the chest cavity is evacuated with a small tube placed through the 5 mm port to the scope and using a water-seal. The air from the left side can be redirected by fixing that side with the right hand while the anaesthesiologist ventilate with positive PEEP.

The skin incisions are closed in two layers. The skin is closed by an intracutaneous absorbable suture.

Procedure Overview (Figs. 39.1, 39.2, 39.3, 39.4, 39.5, 39.6, and 39.7)

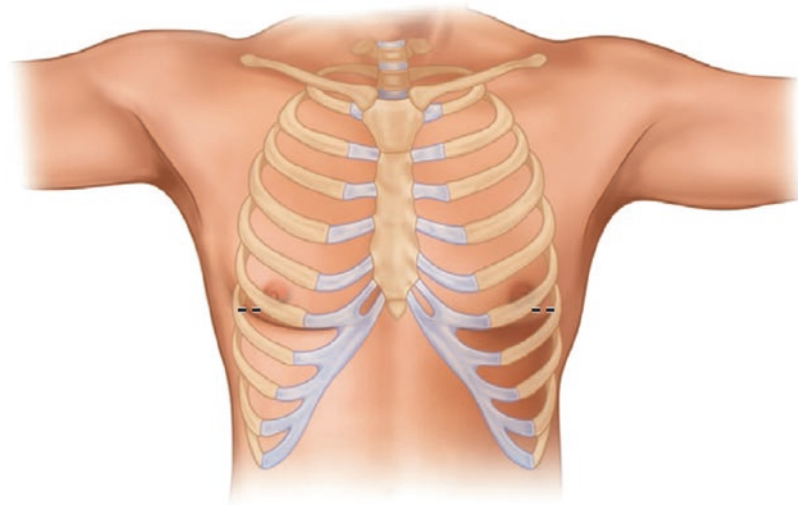


Fig. 39.1 Transverse skin incisions in the mid-axillary line. In girls with demarcation of the breast contours, the skin incisions are placed in the submammarian sulcus

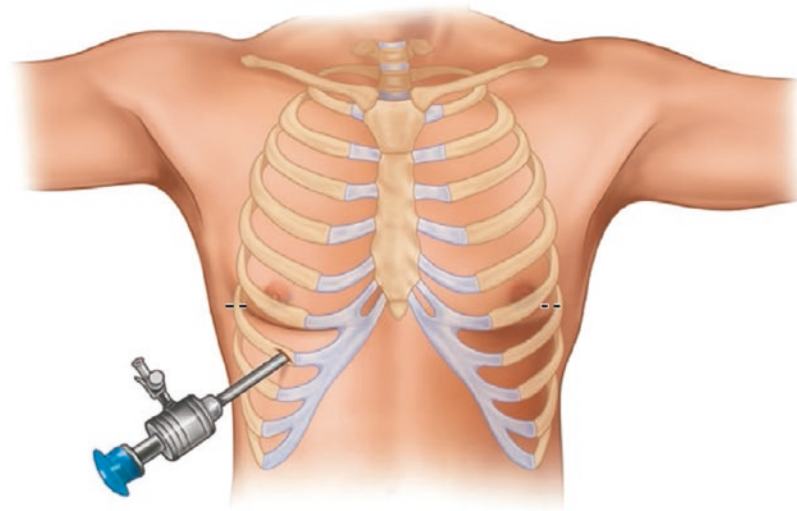


Fig. 39.2 A 5 mm port is inserted just above the level of the diaphragm

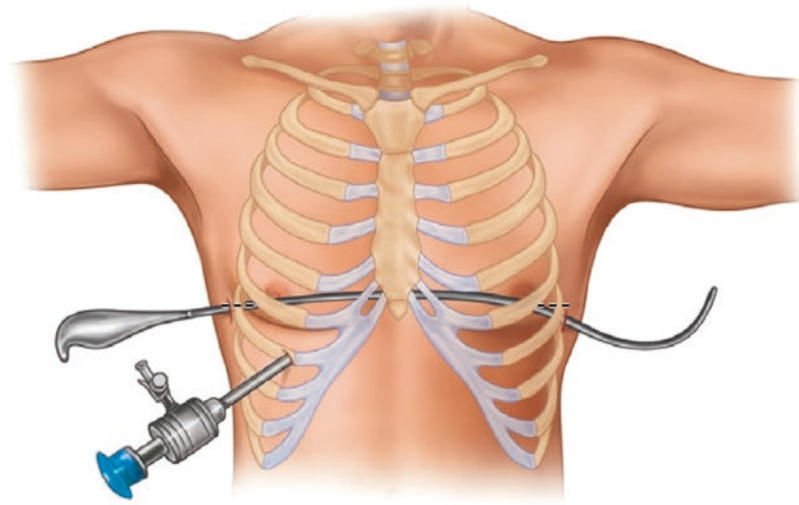


Fig. 39.3 Using the correct size pectus introducer, it is inserted into the chest in close contact with the posterior sternal wall and retrieved from the opposite side

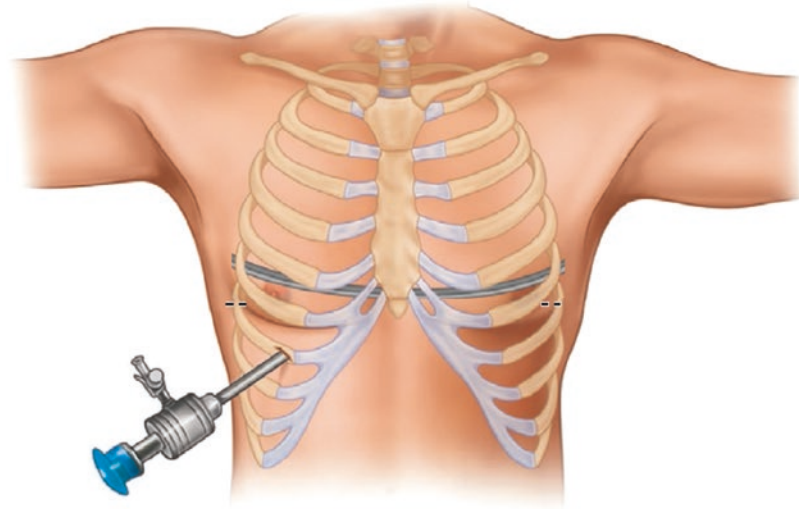


Fig. 39.4 The pectus introducer is removed leaving a guide cotton tape to which the pectus bar is attached and brought into position in an inverted position

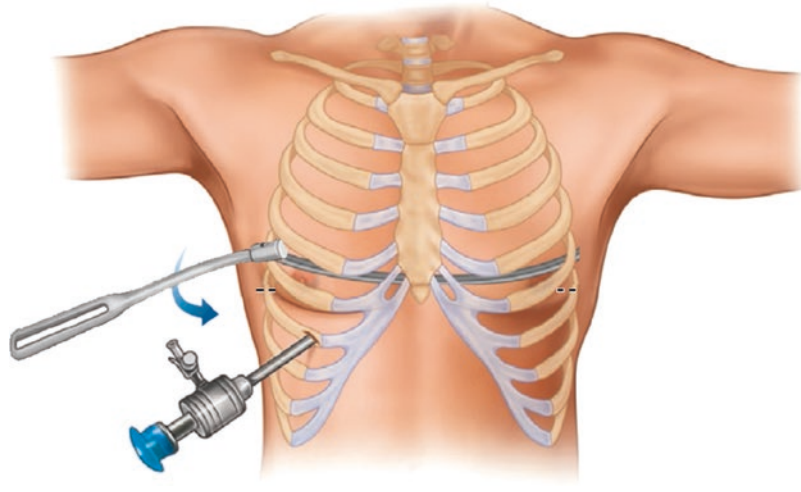


Fig. 39.5 The bar is placed like an U and then rotated 180° with the use of one or two flippers

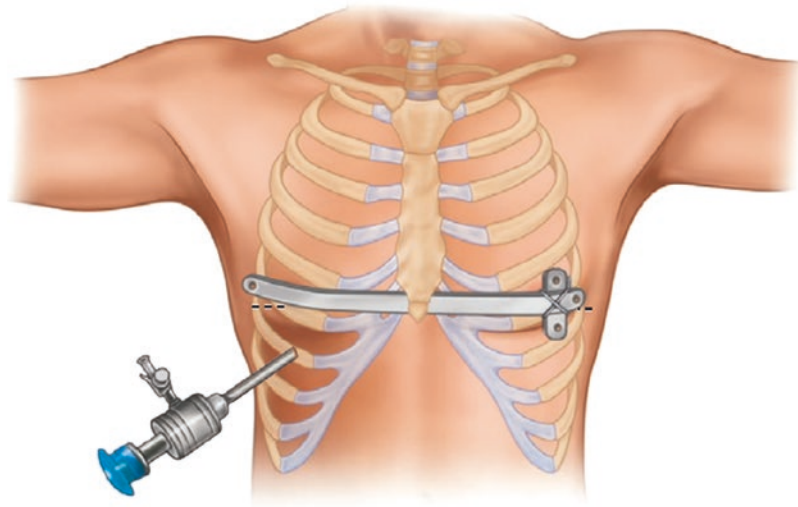


Fig. 39.6 The pectus bar is secured on the left side with a single stabilizer plate

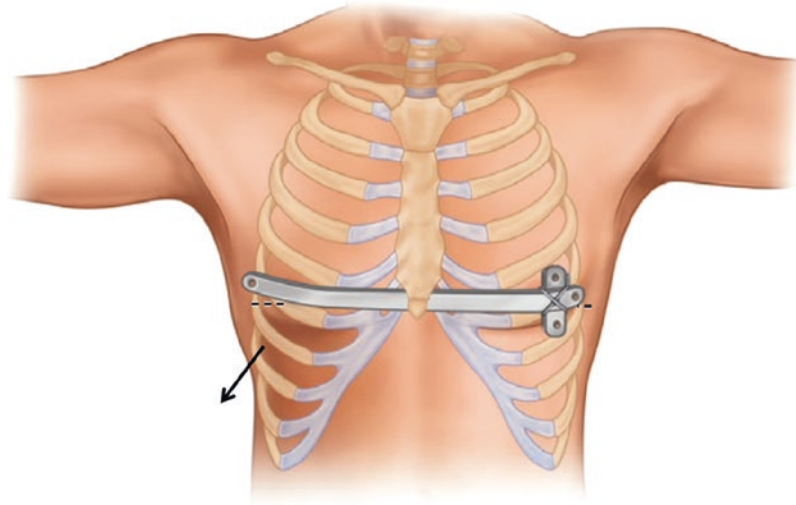


Fig. 39.7 The air from within the chest is evacuated using an underwater seal and the incisions are closed

Postoperative Management

Antibiotics are given for 3 days after the procedure. The epidural catheter is kept for 2 days and oral pain medications is started the day after surgery: These consist of morphine for 2 weeks, non-steroidal anti-inflammatory drugs for 4 weeks and paracetamol for 5 weeks. The patient is mobilized as soon as possible, and most of the patients commence with walking on the day of surgery. Approximately 75 % of the patients are discharged from the hospital on day 2 after surgery and less than 10 % stay more than 3 days. Postoperative x-ray are performed on the second day. In around half the patients a pneumothorax may be present, but less than 1 % will require drainage as the pneumothorax will resolve spontaneously [2]. Patients are seen in the outpatient clinic 6–8 weeks after surgery for a clinical evaluation. An x-ray of the chest is performed at this time and information about further rehabilitation is again given to the patient. Then the patient is

seen at the time for bar removal, which is performed after 3 years. If there any problems in between these intervals, the patient is informed to contact the department.

At the time of discharge from the hospital, the patients are informed about the restrictions for the first 6 weeks which include: not to carry more than 2 kg in front of the body or 5 kg on the shoulders, not to use the bicycle, not to twist the upper body too much and sleep on the back. For the entire course where the bar is in situ the patient is informed to avoid heavy contact sport such as American football, rugby, ice hockey and other self-defense sports. The bar(s) are removed after 3 years.

Results

The procedure has been used in more than 1600 patients with a median age of 16 years (range 7–58 years). The ratio between males and females has been 6:1. The mean operation time has been 36 min (12–270 min). Approximately 74 % have been treated with a single pectus bar, but the number of pectus bars are dependent of the age of the patient (Table 39.1). There have been few complications through the time since the procedure has been offered in 2001. Since 2011, more than 700 patients have been corrected and 30 % have corrections with more than one pectus bar. The median length of bar has been 10 in. (range 8–14). The distribution is shown in Table 39.2. When the procedure was commenced, the median stay after surgery was 7 days, however at present it has decreased to 2 days (Table 39.3). This can be mainly attributed to the change in pain management, providing better information to the patients prior to surgery and faster mobilization after surgery.

Of the last 720 patients, seven patients had redo corrections, three have had previous heart surgery, two have had a concomitant heart operation of the total four done without complications. Twelve patients (1.7 %) have been re-operated because of: deep infection (n=1), dislocation from side to side (n=2) and rotation of the bar (n=9).

The pectus bar is routinely removed after 3 years [5]. The recurrence rate is very low. The pectus bars have been removed in 1000 patients and in less than ten patients there has been a recurrence; with three opting for a redo procedure.

The quality of life is significantly increased after surgery both assessed by the patients and the parents [1]. The compromised cardial performance has been found to improve significantly after surgery; after bar removal there is no difference between these patients treated for pectus excavatum and a comparable group [3, 4, 9].

Table 39.1 Shows the number of patients with percentages in brackets since 1.1.2011

	Total	<18 years	18–29 years	≥30 years
1 bar	473 (66 %)	359 (74 %)	99 (60 %)	15 (21 %)
2 bars	238 (33 %)	126 (26 %)	64 (39 %)	48 (69 %)
3 bars	9 (1 %)		2 (1 %)	7 (10 %)

Table 39.2 Shows the number of bars used in different sizes since 1.1.2011

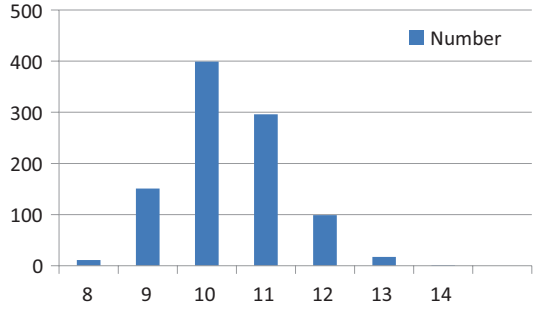
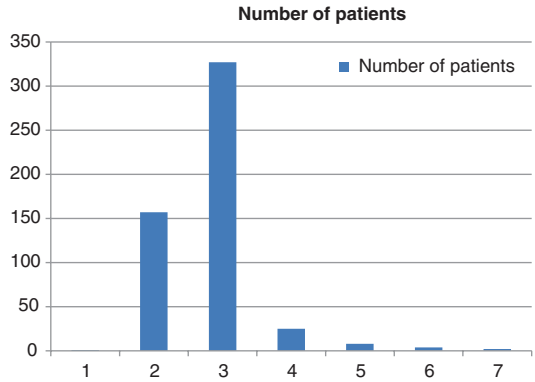


Table 39.3 Show the length of stay in days after surgery for the last 524 patients



Procedure Related Images (Figs. 39.8, 39.9, 39.10, 39.11, 39.12, 39.13, 39.14, 39.15, 39.16, 39.17, 39.18, and 39.19)

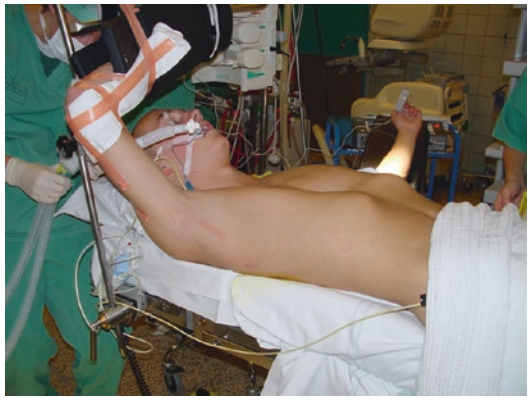


Fig. 39.8 The patient is positioned as shown towards the right edge of the table with the right arm raised above the head

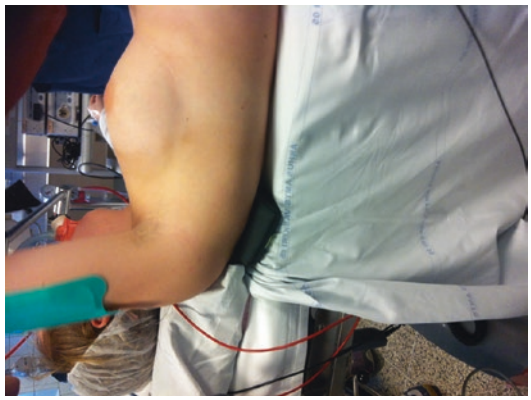


Fig. 39.9 A pillow is placed under the shoulder to elevate the right hemithorax

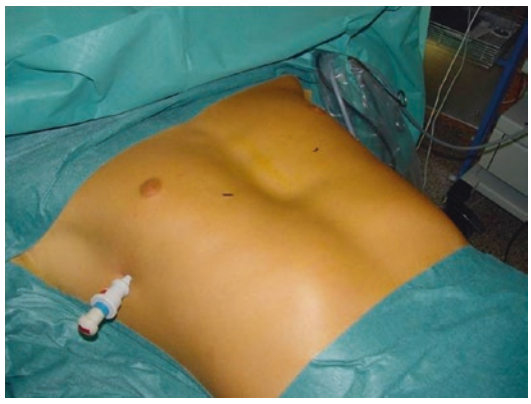


Fig. 39.10 An optic port is positioned in the right thorax and the entrance and exit of the pectus bar through the chest wall are marked

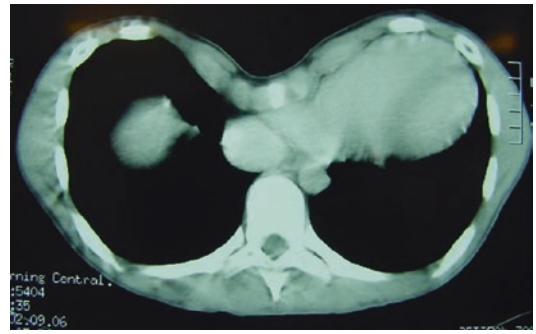


Fig. 39.11 Computed tomogram showing the entrance and exit points through the chest wall

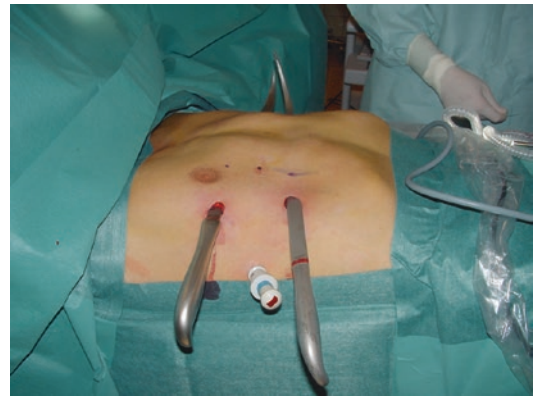


Fig. 39.12 Intraoperative situ of a patient requiring two bars to correct the pectus deformity

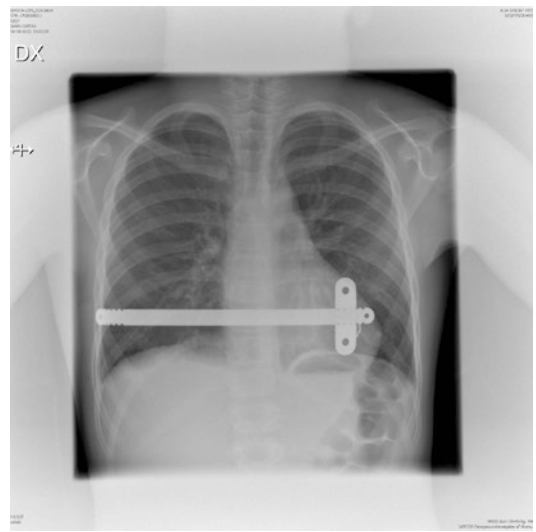


Fig. 39.13 The asymmetric position of the bar which is the hallmark of this technique can be seen on this postoperative chest film

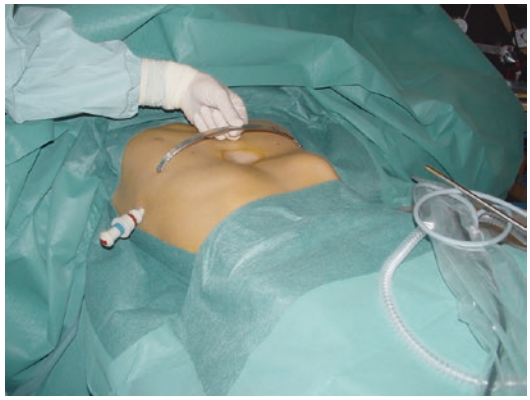


Fig. 39.14 The overcorrection of the pectus bar as shown is preferred in our technique

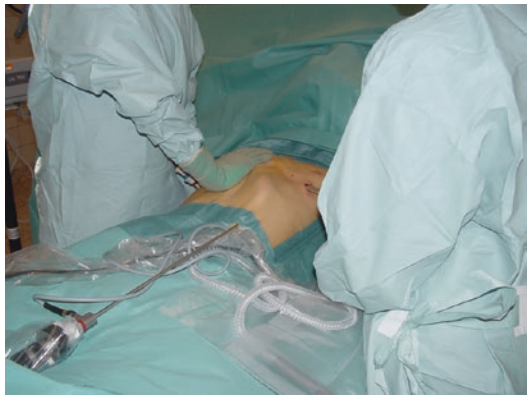


Fig. 39.15 Pressing on the curvature is performed to make a pre-correction of the deformity

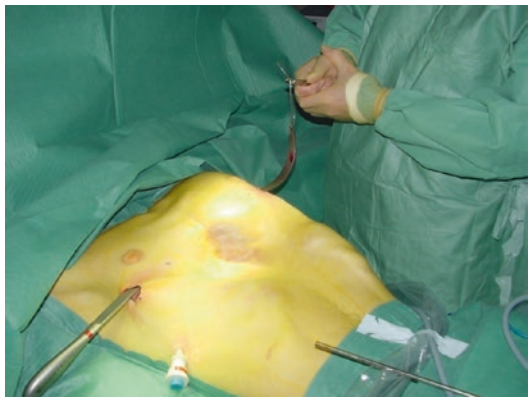


Fig. 39.16 Operative image showing the steel wire being placed through the eyelet of the extra large size introducer

Fig. 39.17 The position of the pectus stabilizer on the pectus bar is done with a steel wire

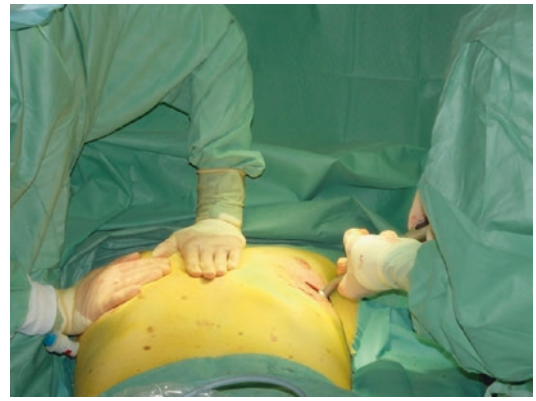


Fig. 39.18 The pectus bar end is bent to lock the stabilizer plate

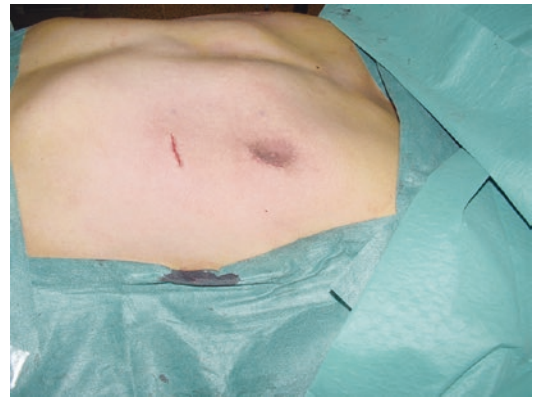


Fig. 39.19 One of the two periareolar incisions after closure

Recommended Reading

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Sternum Elevators for Minimal Access Pectus Excavatum Repair

40

Satoshi Takagi and Hiroyuki Ohjimi

Technical Highlights

During minimal access repair of pectus excavatum (MARPE), cardiac or pericardial injury with the introducer is one of the most serious complications which can be life-threatening [1]. Although a scope is used as a precaution to monitor the areas at risk, the view is quite limited, particularly at the point where the depressed sternum and right atrium remain in close contact. The use a novel sternum elevator which efficiently opens the anterior mediastinal space for the safe completion of MARPE is presented in this chapter.

The technical highlights of MARPE have been described in Chap. 35. The main technical advantages of the sternum elevator as follows:

- This device does not require any extra skin incisions.
- It does not interfere with an introducer or a scope during the surgical procedure.
- Is also effective in adults, whose rib cages are firmer than those of the young patients.
- Will help decrease the risk of life-threatening cardiac and pericardial injuries.

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Indications

MARPE is suitable for correction of chest defromities with depression:

- Symmetric Pectus Excavatum
- Mild or moderately asymmetric Pectus Excavatum
- Pectus Excavatum with Platythorax

Preoperative Considerations

For preoperative consideration please refer to the Section described in Chap. 35 on MARPE.

Special Instruments [2]

For the list of special instruments please refer to the Section described in Chap. 35 on MARPE. Instruments specific for this procedure are as follows:

- **Introducer:** A pectus introducer is an instrument that is used to create a tunnel in the anterior mediastinal space for the leading of a tape through it. Three introducers with different curvatures are prepared specially for the authors and one is selected according to the depressed shape of the pectus excavatum.

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- **Sternum elevator:** The sternum elevator is designed to hold up a depressed sternum during MARPE. The handle side and the elevator side are connected in a horseshoe shape Fig. 40.11. The elevator side is not straight, but is gently curved. The curvature is similar to that of a pectus plate introducer; therefore, these two instruments can be passed through the same skin incision site and manipulated in the thoracic cavity with minimal interference. The sternum-touching side of the elevator has a rough surface to prevent it from slipping off of the sternum. An assistant standing on the patient's left side holds the elevator up and creates sufficient space between the sternum and heart. This elevator is now commercially available from Solve Co., Yokohama, Japan.

Surgical Procedures

For the repair of pectus excavatum, general anesthesia is first administered to a patient; if possible using a double-lumen endotracheal tube whenever possible. The patient is positioned on the right edge of the operating table to ease the handling of a scope that is inserted into the right chest cavity. The patient's shoulders are abducted outwards and the elbows are flexed upwards. Padding is placed under the forearms and hands to prevent neurological injury. The video monitor for the scope is positioned at the left of the patient's head. The chest is prepped and draped and surgical marking proceeds.

The deepest section of the sternum at the midline is marked. If the deepest point is beyond the sternum, the lower end of the sternum is marked. Through that marked point, a horizontal line is drawn on the chest. The marking continues to the top of the pectus ridge, the point of reflection towards the costochondral chest cage. Thoracic entry (right side) and exit (left side) points for the pectus bar are located at the intercostal space near the intersection of the horizontal line and the pectus ridge. The fifth intercostal space is generally the region that is often used for those two points. The entry point for the sternum elevator is also marked at 1 cm medial to the Nuss plate entry point in the same intercostal space.

The operative procedure starts with bilateral lateral thoracic incisions that are positioned posterior to the anterior axillary line for an inconspicuous scar. Up to a 3 cm incision permits the surgical procedures on the right side, where an introducer and sternum elevator are passed through simultaneously. A subcutaneous dissection around the site and to the pectus ridge is performed through each incision. On the right side, while the chest skin is hitched forward with a finger and in isolated cases selective lung ventilation of the left lung is performed, a scope is inserted into the pleural cavity through a third small incision. The seventh intercostal space on the middle axillary line is commonly selected for this purpose. Hereafter, all of the maneuvers within the right chest cavity are monitored through the scope.

The sternum elevator is then brought through the right skin incision. After penetrating the thoracic wall at the point just inside the pectus ridge, the elevator instrument is flipped around and advanced to the back of the sternum. An assistant standing on the patient's left side holds up the instrument. In the endoscopic view, the sternum is well elevated and apart from the heart. An introducer follows the elevator, enters the chest cavity at the pectus ridge, passes across the already widened mediastinal space, and finally comes out through the left incision. Due to the distinctive curvature of the elevator, these 2 instruments can be passed through the same skin incision without much interference.

The introducer leads a cotton tape from left to right and the sternum elevator is removed. A preformed pectus bar is placed with guidance of the tape. The concave deformity of the pectus excavatum is corrected after a 180° rotation of the plate. In most cases, the plate is stabilized by the application of a stabilizer plate. If the concave deformity remains significant, a second pectus bar can be placed, most often in the third

intercostal space. The surgical steps are the same as those of the first plate insertion, but by then the sternum elevator is not needed because the sternum is already elevated by the first plate.

Differential lung ventilation, if used, is then discontinued, the thoracoscope is removed, and the incisions are closed up. Chest x-rays (antero-posterior and lateral views) are obtained to check for a pneumothorax and to verify final plate placement.

Procedure Overview (Figs. 40.1, 40.2, 40.3, 40.4, 40.5, 40.6, 40.7, 40.8, 40.9, and 40.10)

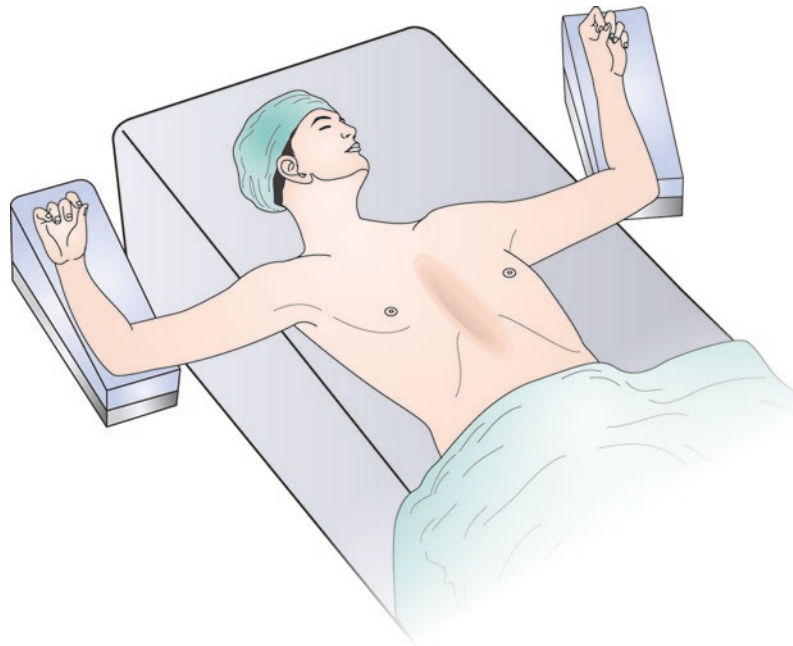

Fig. 40.1 Surgical positioning of the patient for MARPE

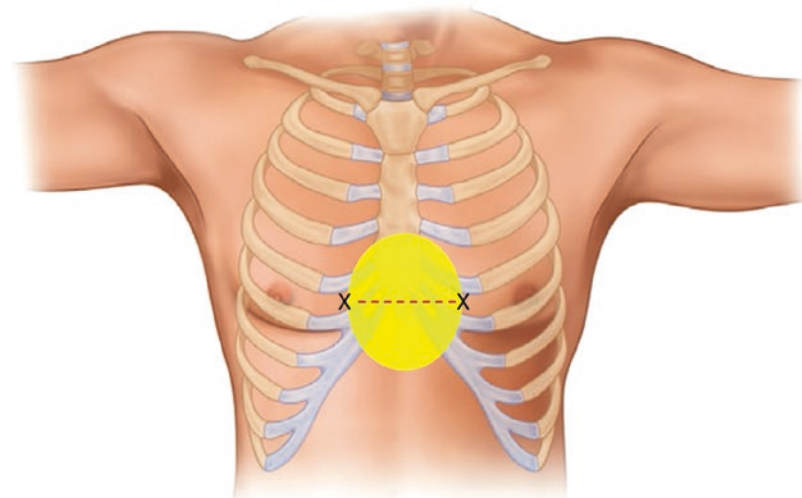

Fig. 40.2 The intersection of the *horizontal line* and the pectus ridge (*shaded area*) indicates the thoracic entry (*right side cross*) and exit (*left side cross*) points for the pectus bar. The entry point for the sternum elevator is marked at 1 cm medial to the pectus bar entry point

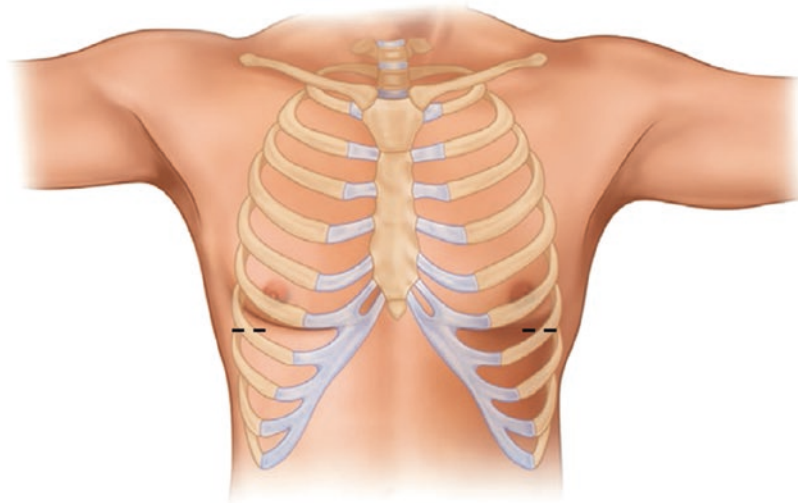

Fig. 40.3 Skin incisions positioned posterior to the anterior axillary line

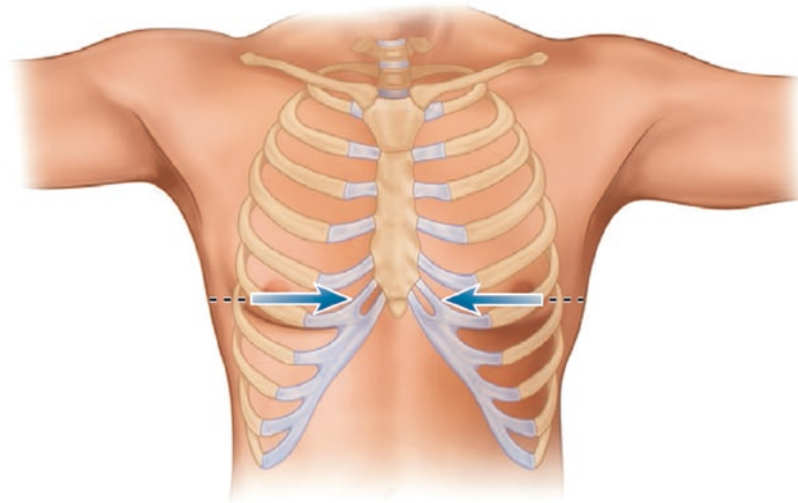

Fig. 40.4 Blunt dissection is carried out to create a tunnel to the ridge of the depression

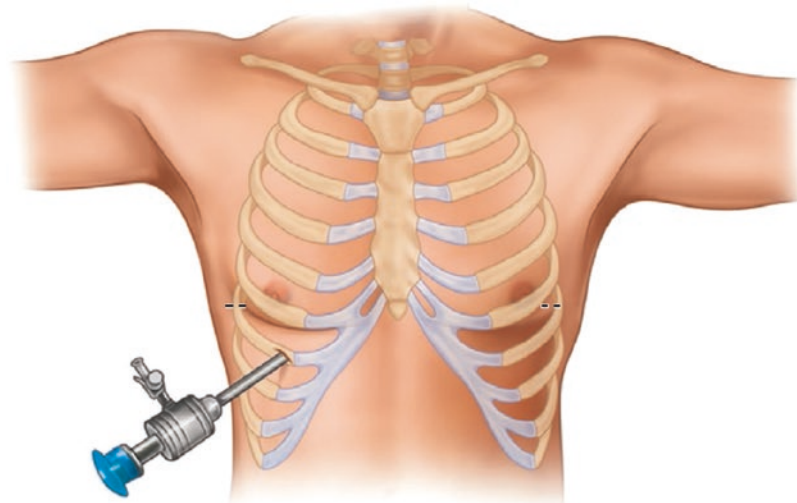

Fig. 40.5 A 5 mm port is inserted below the right transverse chest incision just above the level of the diaphragm

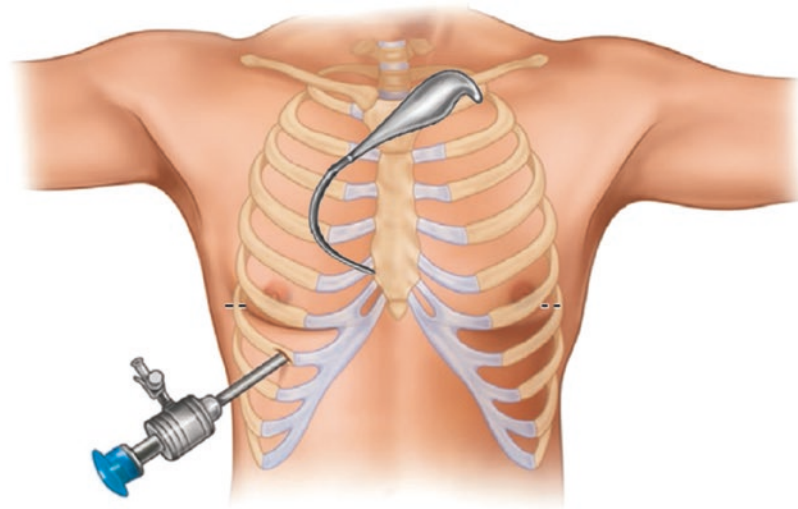

Fig. 40.6 An elevator upholding the sternum and widening mediastinal

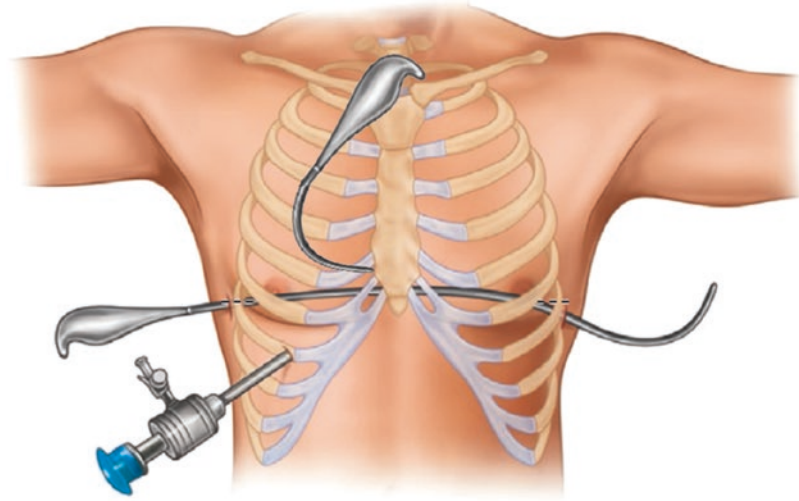

Fig. 40.7 With the elevator upholding the sternum and the widened mediastinal, an introducer follows the elevator through the same skin incision and comes out to the left side. A cotton tape is tied at the end of the introducer

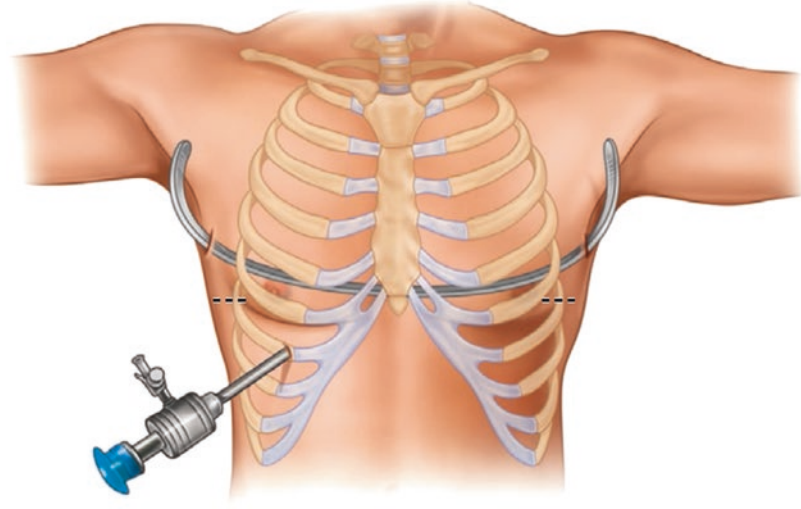

Fig. 40.8 The pectus bar is then placed behind the sternum and its ends facing upwards like a U using the cotton umbilical tapes to guide it through the thoracic cavity and the retrosternal space

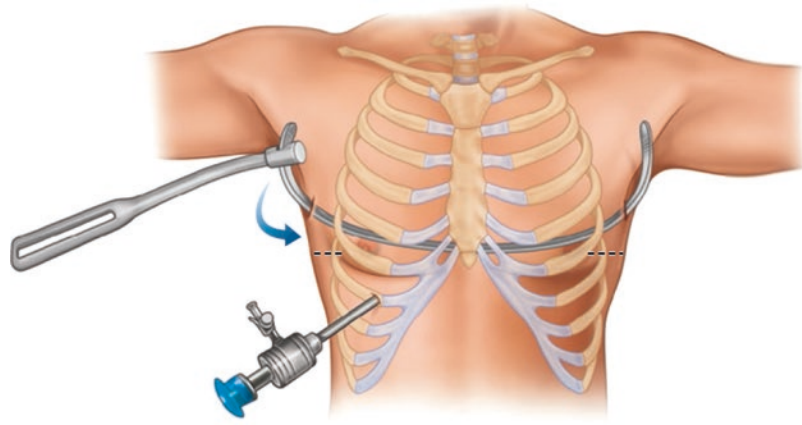

Fig. 40.9 The pectus bar flipper is attached securely to the right end of the pectus bar after which the pectus bar is flipped 180° into position

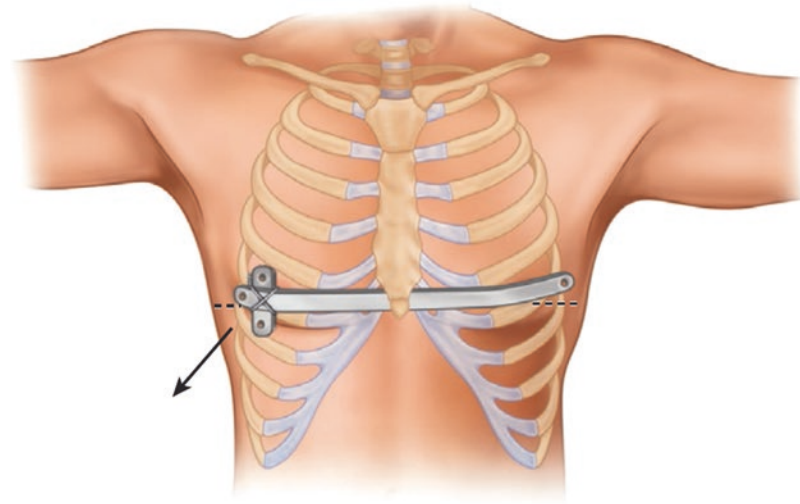

Fig. 40.10 The pectus bar is stabilized with a single stabilizer plate and the wound is closed. After the insufflated gases are evacuated from the thorax, the port is removed

Results

The procedure was conducted in 100 patients (79 males and 21 females) undergoing pectus excavatum repair with a mean age of 12.9 (range, 5–36) years. The mean Haller's computed tomography index was 5.3 (range, 1.8–17.7). No cardiac or pericardial injury was encountered. Use of the elevator was always helpful for the process of substernal tunnelling with the introducer, even in elderly patients, whose rib cages were usually firmer than those of younger patients. A second pectus bar was applied in 50 patients, and only a few required use of the elevator for the second substernal tunneling process.

Because of its characteristic shape, the elevator does not interfere with the MARPE, and it allows the introducer to pass through next to it, meaning that the introducer goes through the widest retrosternal space, which helps reduce the risk of life-threatening cardiac and pericardial injuries. The following dissection is relatively safe and straightforward. Compared to the other sternum-elevating devices reported to date, the most distinctive feature of our elevator is that its insertion requires no extra skin incisions.

Procedure Related Images

(Figs. 40.11, 40.12, 40.13, and 40.14)

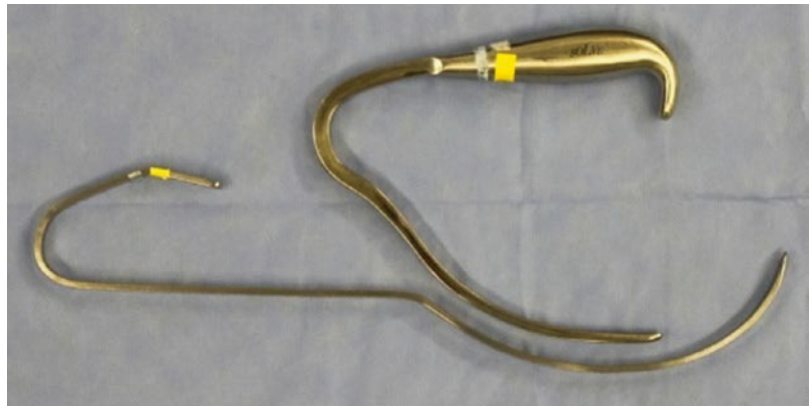

Fig. 40.11 A sternum elevator (*above*) and an introducer (*below*), that have a similar curvature

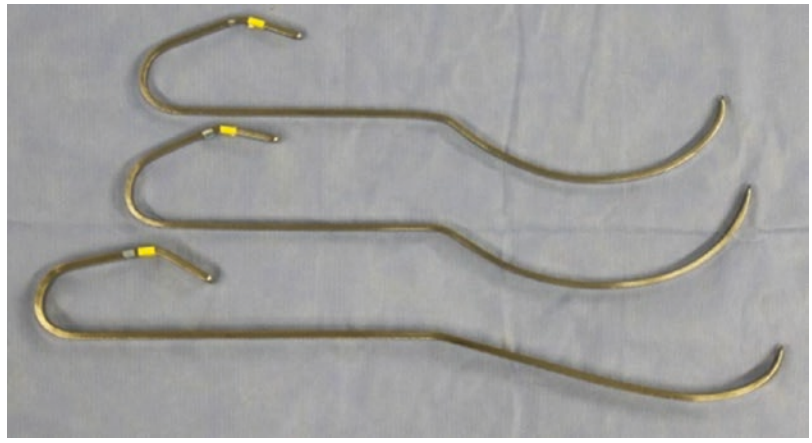

Fig. 40.12 The assortment of three types of introducers available for MARPE at the authors Centre

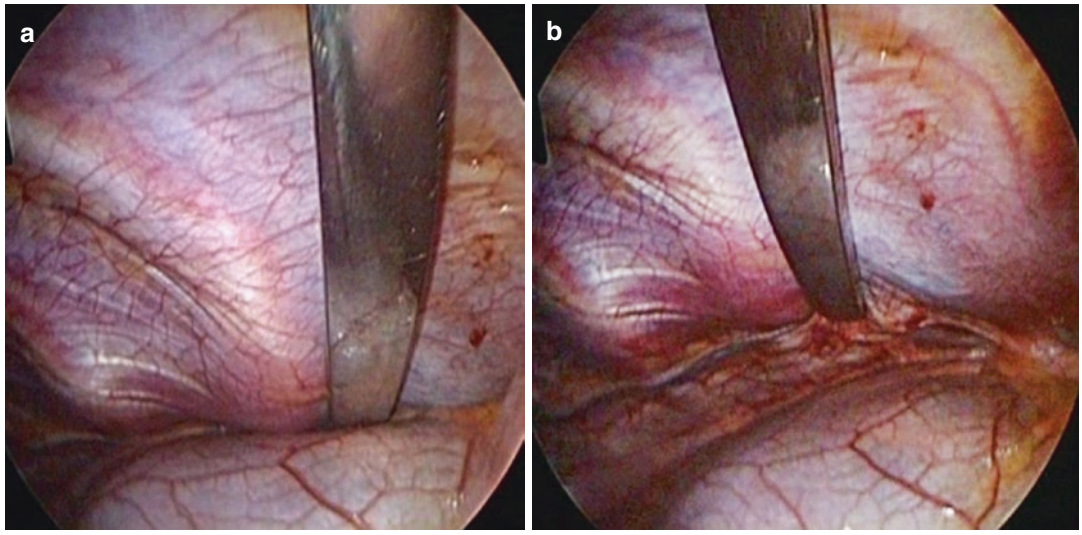

Fig. 40.13 Intrathoracic views of the retrosternal space without (**a**) and with (**b**) sternum elevator

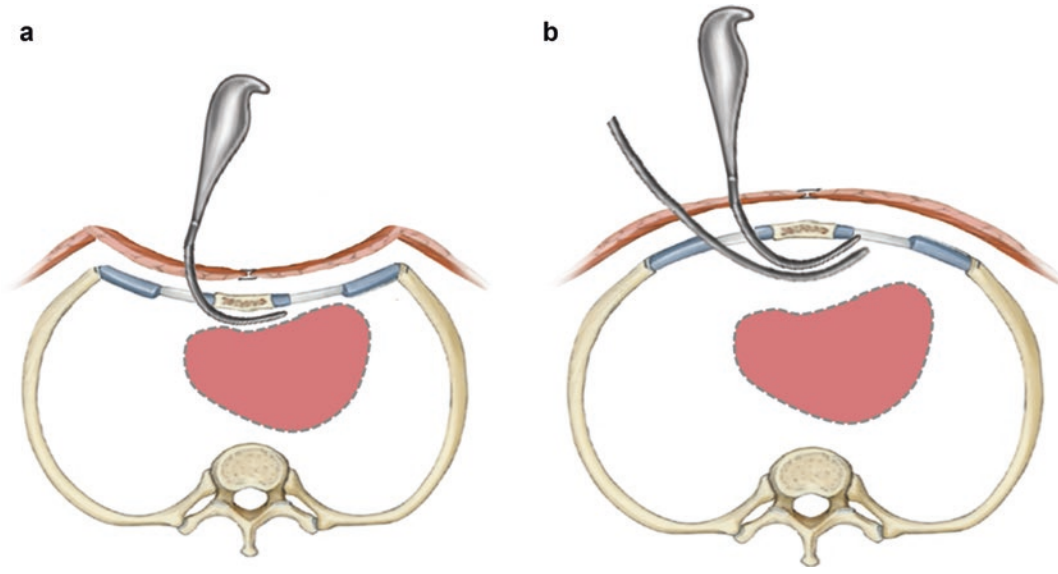

Fig. 40.14 Schematic view of the intrathoracic space without (**a**) and with (**b**) sternum elevator used for MARPE

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Minimally Access Repair of Pectus Excavatum- with Sternotomy in Adults

41

David Pérez, Gara Torrent, and Santiago Quevedo

Technical Highlights

The main technical highlights of this procedure are:

(a) Short and flat bars are used (curved only at the ends).
(b) Bilateral perimammilar incisions in males and sub-mammary incisions in females are used for insertion of both the introducer and the pectus bar, which provide good esthetic results. Incisions at the axillary lines are requuired in cases of depression of the lower end of the sternum.
(c) Use of angular lens to examine the pleural cavities and to monitor the whole procedure, which enhances the safety of this intervention.
(d) Transverse sternotomy performed with endoscopic monitoring of the mammary vessels. This section allows for immediate correction of the deformity and reduces postoperative pain as well as the probability of implant migration.
(e) Instruments and implant are inserted according to the steps described for the standard MARPE technique.
(f) The bar is bilaterally fixed to the costal arcs by means of steel wire, without stabilizer plates.

Indications

- Symmetric Pectus Excavatum in adults
- Symmetric Pectus Excavatum in older adolescents
- Very severe Pectus Excavatum in adults, relatively asymmetric or complex (utilization of longer bars. Incisions at axillary lines).

Preoperative General Considerations

1. The basic preoperative work-up includes: anteroposterior and lateral chest films, electrocardiogram and general laboratory investigations.
2. Quantitative estimations of the deformity, such as the Haller index or volumetric studies have low impact on the decision of establishing the indication for surgery or on the planning of the intervention.
3. Computed tomogram (CT) with three-dimensional reconstruction offers attractive images which are interesting from the

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academic point of view, but do not provide additional relevant information.
4. Photographic documentation of the deformity before and after correction is recommended, taking frontal, lateral and oblique pictures from both sides of patients while they are in the supine and standing positions.
5. Procedures are conducted under general anesthesia with selective intubation with double lumen tube. In case that this intubation procedure is not feasible, simple intubation can be used, with low ventilation volumes is sufficient.
6. Although sternotomy markedly reduces postoperative pain, postoperative analgesia protocols are recommended, which include epidural analgesia during the first few days and intravenous analgesic treatment on subsequent days.
7. Antibiotic prophylaxis with cephalosporin, is administered 30 min before skin incision and maintained until the fifth postoperative day
8. Antithrombotic prophylaxis should be administered with low molecular weight heparin at standard doses.

Special Instruments

The surgical instruments for general thoracic surgery and following special intruments:

1. Pectus surgical instruments set with bars, bar bending tools and dissectors (Biomet Microfixation Pectus Bar, Jacksonville, Florida). Stabilizer plates are not needed.
2. Scope of 5 mm or 10 mm with 30° and flexible endoscopic ports (Flexipath 12 mm).
3. Costotome and/or Lebsche knife (Codman and Shurtleff, Inc, Bridgewater, MA, USA).
4. Multifilament Dall-Milles steel cable system and cable grip (Stryker Orthopaedics, Mahwah, NJ, USA) or monofilament stainless steel sternotomy wire.

Surgical Technique

The patient is placed in the supine position with a small interscapular roll and with the arms in abduction.

Incisions 2–3 cm long are performed on the internal margins of both areolas in male patients or on the submammary creases in female patients. The distance between both areolas is measured and a metal bar is selected that is four centimeters longer than that distance. The bar is then bent with a mild curvature at the both ends.

The mammary fat and pectoral muscles are displaced and flexible ports are inserted in both the hemithorax. Flexible ports allow for simultaneous parallel insertion of the scope and instruments. Both pleural cavities are then explored with a 30° scope.

A skin incision of 2 cm is made on the sternum, through which the pre-sternal tissue is dissected until the surface of the sternum is reached. Then the entire sternum thickness is sectioned with a costotome and/or a Lebsche knife. The right mammary vessels are monitored through scope, while the left vessels are preserved by stopping the sectioning procedure as soon as bone resistance is perceived to drop.

The pectus introducer is inserted through the right skin incision into the subpectoral layer and is moved forward until the highest point of the anterior arch is reached. At this point, it enters the right pleural cavity and is moved towards the contralateral hemithorax. The midline is crossed behind the depressed sternum ventral to the pericardium under endoscopic vision. At that moment, the scope is passed to the left hemithorax to monitor the entrance of the introducer in the left pleural cavity and its exit to the subpectoral space. At this point the introducer is advanced and a corresponding submuscular tunnel is created on the left side.

A cotton tape is tied to the end of the dissector and the dissector is withdrawn back to exit the ribcage. This cotton tape preserves the trajectory and facilitates insertion of a second broader introducer that widens the tunnel. The pectus bar is pulled through the anterior mediastinum from one incision to the other into the submuscular pockets and is finally turned and fixed to the ribs using steel wires without stabilizer plates. Our preference is for the Dall-Milles steel cable system and cable tensor.

After evacuation of the thoracic cavities, the wound is closed in layers.

Procedure Overview (Figs. 41.1, 41.2, 41.3, 41.4, 41.5, and 41.6)

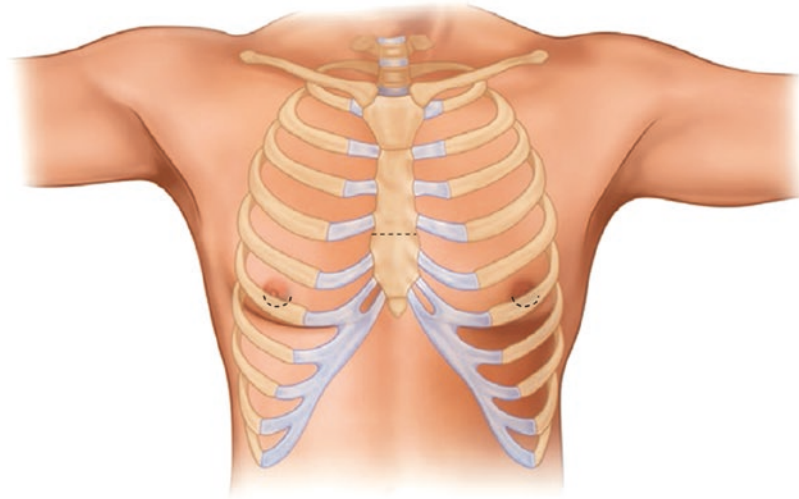


Fig. 41.1 Peri-mammilary incisions in male patients or in the submammary creases in female patients and an additional 2 cm incision to perform the sternotomy are made

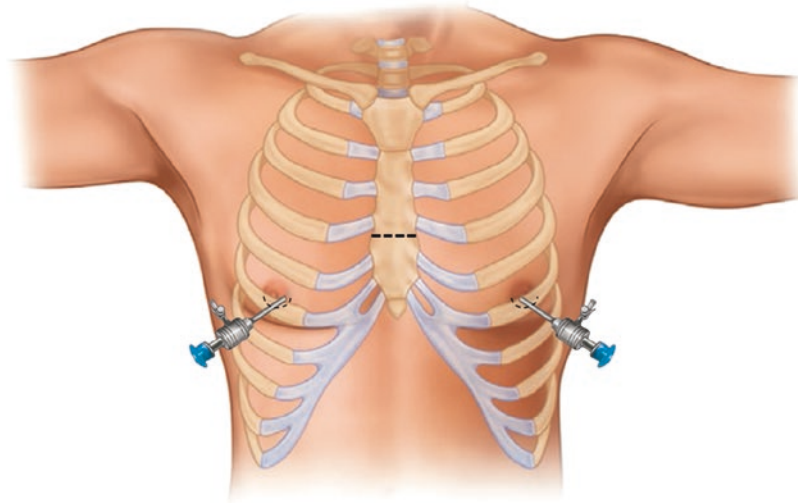


Fig. 41.2 Bilateral insertion of ports after which a sternotomy is performed under endoscopic vision of the right mammary vessels

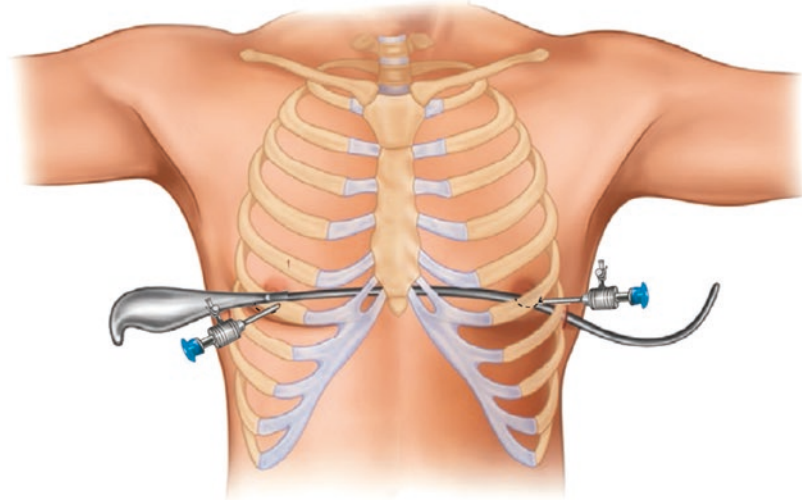


Fig. 41.3 The introducer is inserted through the right perimammilary incision. The introducer has crossed midline and exits the left pleural space and left chest wall in a symmetric manner. The scope is switched from one side to the other for this maneuver. A cotton tape is tied to the introducer and retrieved

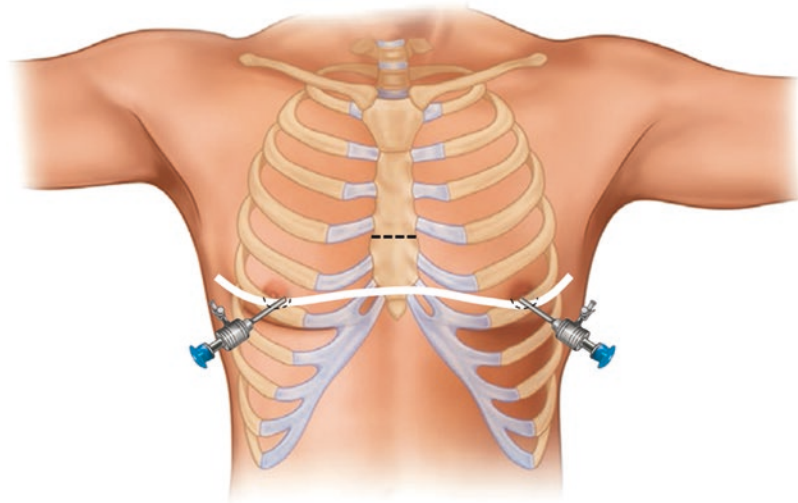


Fig. 41.4 The cotton band is used to help guide the bar into its final position

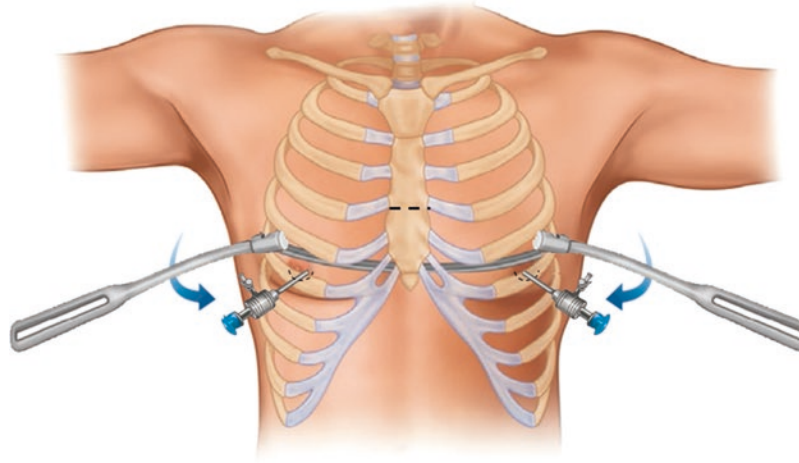


Fig. 41.5 The pectus bar is flipped and set into the desired position

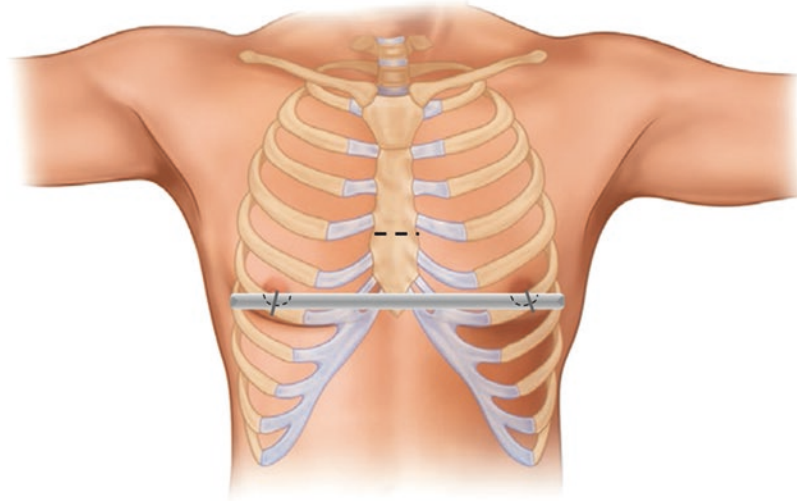


Fig. 41.6 Both ends of the pectus bar are attached to the ribs with multifilament wires using a Dall-Milles steel cable system tied around the rib

Postoperative Management

Patients are extubated in the operation room and transferred to the recovery room for the first few hours, where the analgesic treatment is adjusted and a chest film is taken. After transfer to the wards, respiratory physiotherapy is provided after 24 h and mobilization after 48 h postoperatively. Patients are discharged on the 5th day with pain medication which consists of non-steroidal anti-inflammatory drugs. Relative rest for 2 weeks is recommended. Patients are advised to avoid sport activities in general for 6 months and competitive or contact sports for 1 year.

Removal of the pectus bar after 3 years is recommended; however, it can be removed earlier, once the sternal fracture is consolidated in its new position.

Comments

Correction of Pectus Excavatum by using the Minimal Access Repair of Pectus Excavatum (MARPE) produces excellent thoracic remodeling results in children and adolescents for whom this technique was originally designed. Placing a retrosternal bar in older adolescents or adults, who have less malleable ribcages, often fails to elevate depressed sternum enough thus producing unsatisfactory outcomes. Moreover, placing an implanted bar in the ribcage generates tension forces that may lead to complications such as severe or prolonged postoperative pain and/or bar displacement or rotation. Using two or more metal bars to counteract the sternum resistance may enhance thorax remodeling but requires more surgical wounds or larger ones, which worsens the cosmetic outcome of this intervention.

The modified MARPE technique for correction of Pectus Excavatum in adults, is based on the idea that in order to achieve similar results to those attained in younger patients, the rigidity of the ribcage should be reduced before inserting the pectus bar; a goal that can be easily achieved by performing transverse section of the sternum. Osteotomy reduces thoracic rigidity thus facilitating immediate correction of the deformity, while reducing postoperative pain and implant displacements. Further relevant differences between our technique and the standard procedure are: use of short and mildly bent pectus bars and their insertion with bilateral endoscopic control through minimal periareolar or submammary skin incisions, with yields good esthetic results.

For the success of this technique patients with chondromanubrial sternal depression or inelastic depression extensively involving the whole sternal body and both anterolateral rib cages (platythorax) are not eligible candidates for this intervention and should therefore be considered for an open surgical repair. Patients with rigid though asymmetric or extensive deformities however, can benefit from transverse sternotomy but need longer bars, which can only be inserted through lateral wounds.

Procedure Related Images

(Figs. 41.7, 41.8, 41.9, 41.10, 41.11, 41.12, 41.13, 41.14, 41.15, 41.16, 41.17, 41.18, 41.19, 41.20, 41.21 and 41.22)

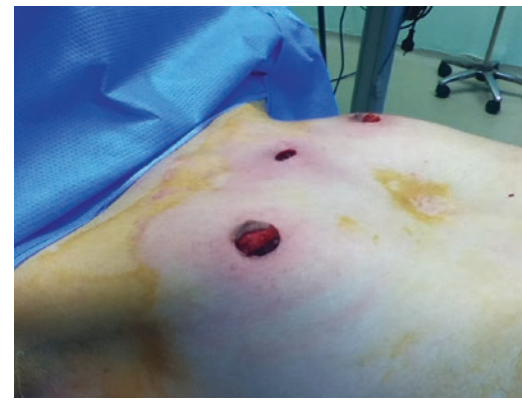


Fig. 41.7 The 2–3 cm long perimmamilar incisions made in male patients or on the submammary creases in female patients provide easy access to insert the bar. An additional 2 cm incision is made in order to perform the sternotomy

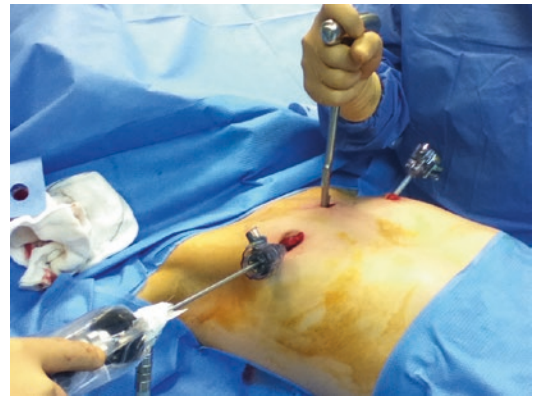
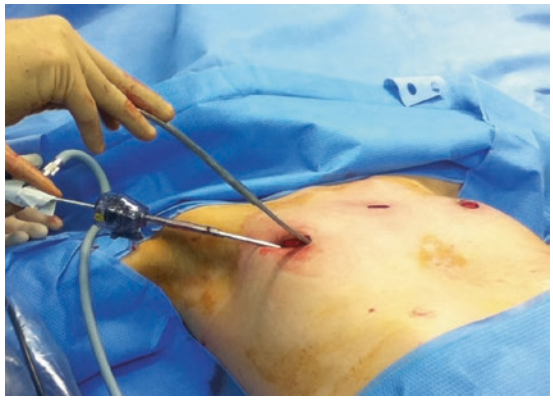
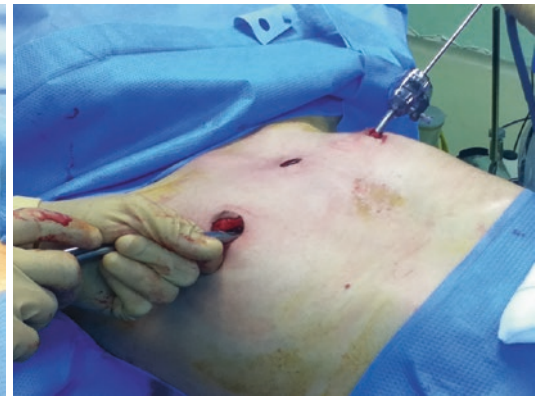


Fig. 41.8 The sternotomy is safely performed under endoscopic monitoring of the right mammary vessels. Transverse sternal osteotomy reduces tension of the rib cage if the sternum is rigid and depressed





Figs. 41.9 and 41.10 The introducer is inserted through the right perimammilary incision in the subpectoral layer and is moved forward just above the anterior ribs until the location to insert the bar is reached. At this point it is inserted into the intrapleural space and is moved in a transverse direction into the mediastinum in order to create the retrosternal tunnel

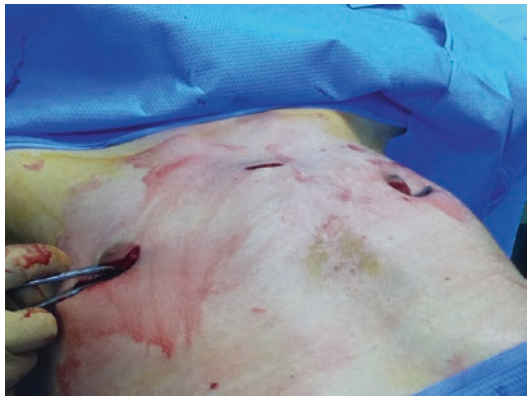


Fig. 41.11 The introducer has crossed midline and exits the left pleural space and left chest wall in a symmetric manner

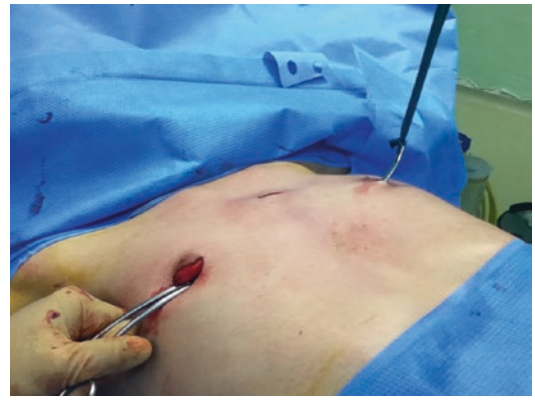
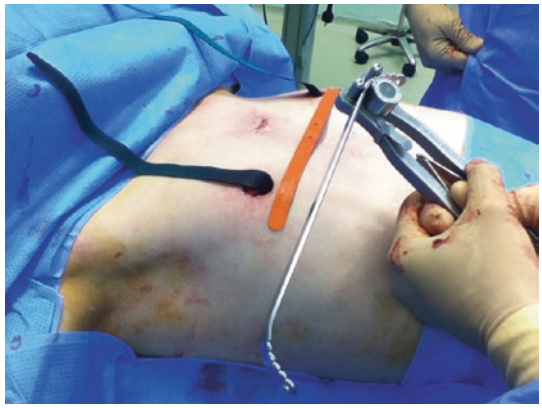
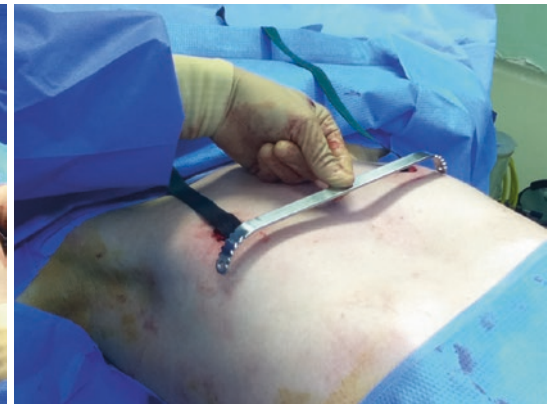
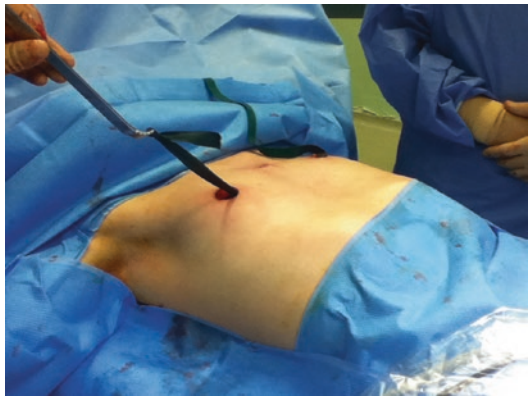
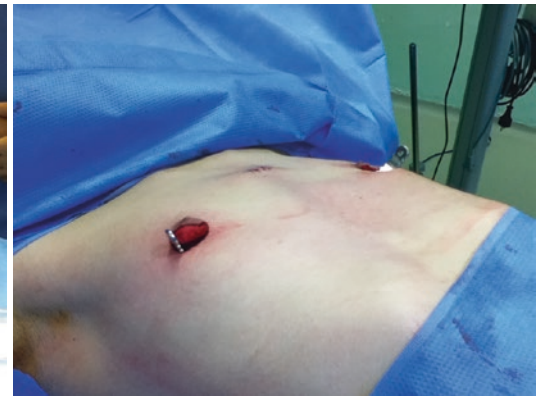


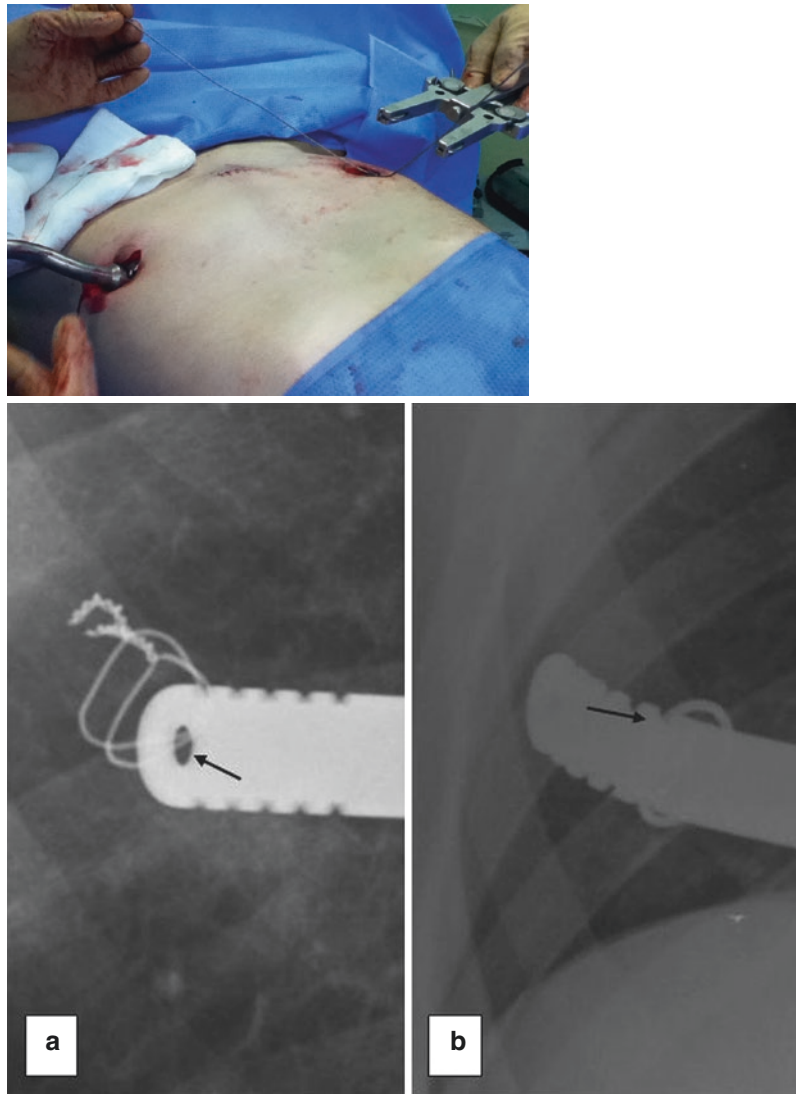
Fig. 41.12 The cotton tape preserves the trajectory and guides insertion of a second thicker dissector that widens the tunnel and of the metal implant

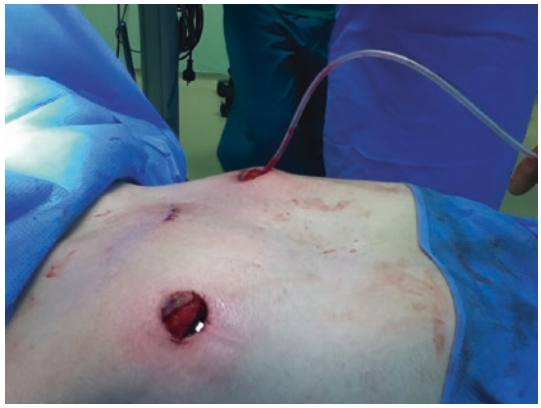
Figs. 41.13 and 41.14 A short metal bar is selected and bent with a mild curvature at the ends

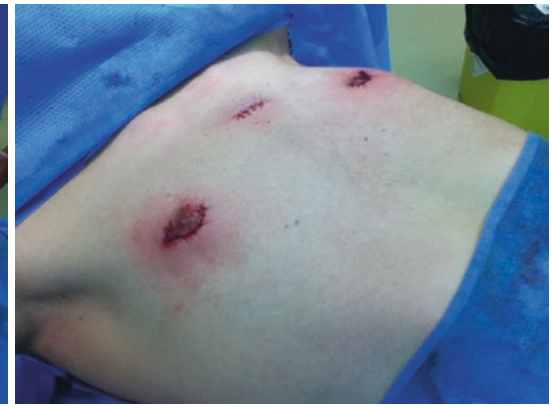



Figs. 41.15 and 41.16 The cotton band is used to help guide the bar into its final position

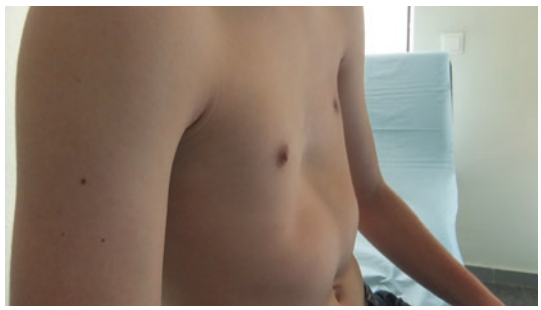


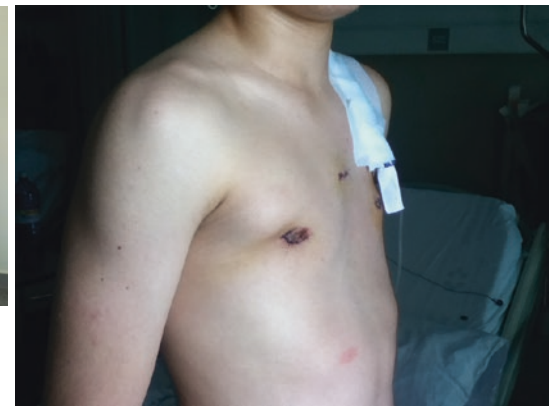
Figs. 41.17 and 41.18 The pectus bar is rotated and secured with steel wires. Both wings of the pectus bar are attached to the ribs with multifilament wires using a Dall-Milles steel cable system tied around the rib. Postoperative X-rays demonstrating the bar secured with a monofilament metal wire (**a**) -which was introduced through the bar hole (*see arrow*)- or with multifilament wire (**b**) embracing rib and prosthesis at its serrated end border (*see arrow*)





Figs. 41.19 and 41.20 Once the pleural cavities are aspirated, the wound is closed in layers using simple interrupted sutures, such as 3–0 absorbable and 4–0 non-absorbable monofilament for subcutaneous tissue and skin, respectively





Figs. 41.21 and 41.22 Pre- and postoperative (4th postoperative day) images of the deformity repair in an adult male patient. Note that this patient has Poland's syndrome with absence of the left pectoralis major and minor muscles. Although this anomaly limits the final esthetic result of the intervention, adequate contour of the chest wall is achieved and scars are well concealed

Suggested Reading

1. Pérez D, Cano JR, Quevedo S, López L. New minimally invasive technique for correction of pectus carinatum. Eur J Cardiothorac Surg. 2011;39(2):271–3.

2. Pérez D, Cano JR, Quevedo S, López L. Videothoracoscopic repair of pectus excavatum with sternal transection for adult patients with nonelastic deformity. J Thorac Cardiovasc Surg. 2011;142(4):942–3.

Free Fat Transplantation for the Aesthetic Correction of Mild Pectus Excavatum

42

Aris Sterodimas and Luiz Haroldo Pereira

Technical Highlights

The authors have introduced a new method for transplantation of Stromal Enriched Lipograft (SEL) in order to restore the visible and non-functionally disturbing deformation of mild funnel chests.

The main technical highlights of this procedure are:

(a) In Stromal Enriched Lipograft (SEL), freshly isolated Stromal Vascular Fraction (SVF) is attached to the aspirated fat, with the fat tissue acting as a living bioscaffold before transplantation.
(b) The donor site for fat harvesting can be chosen according to the preference of the surgeon and the patient.
(c) An important consideration for harvesting and refinement in preparation for grafting is to respect and maintain the tissue architecture of living fat.
(d) Fat is aspirated using the syringe method.
(e) The 2/3 of the aspirated fat is used in order to isolate the SVF containing Adipose Derived Stem Cells (ADSCs).
(f) The remaining 1/3 of the aspirated fat is used for the ADSCs to be attached.
(g) The SEL is inserted into tunnels beginning at the deep layer and working up into the more superficial. Different trajectories are created. The SEL is injected as the cannula is withdrawn.

Indications

- Mild types of pectus excavatum [6]
- Mild chest wall deformities associated with Poland's Syndrome [9]

Preoperative Considerations

1. Candidates are patients over 18 years old with mild pectus excavatum (Fig. 42.1). Preoperative symptoms such as dyspnoea on exertion, shortness of breath at rest, chest pain, and palpitations are indications for further exams and possibly more complex surgical repairs.
2. A full preoperative series of laboratory exams, CBC, coagulation tests, glucose, urea and creatinine tests are performed.

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A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_42

3. Electrocardiograms are performed in order to diagnose or rule out cardiac pathologies.
4. Chest X-Rays are performed in order to access the extent of the chest deformity.
5. Lung function tests are performed in order to evaluate the pulmonary function.
6. Chest perimeter plots to quantify the deformity at three different points: manubrium, sternum and xiphoid.
7. Preoperative photographs of the deformity are taken.

Special Instruments

The following instruments and reagents are required for this surgical procedure:

1. Celltibator (Medikan, Los Angeles, CA).
2. Lipokit Centrifuge (Medikan, Los Angeles, CA)
3. Collagenase (Sigma, St. Louis, MO)
4. Syringe assisted liposuction tools (Tulip Medical, San Diego, CA)

Surgical Technique

The pectus excavatum area to be treated is marked with the patient standing up, sitting down and in supine position. The areas to be liposucked are also marked. Preoperative sedation in is administered at the surgical suite. Anesthesia consists of intravenous sedation and epidural anaesthesia. The patient is placed in supine position with the arms spread out.

After the injection of normal saline wetting solution containing 1 : 500,000 of adrenaline by a small bore cannula and waiting 15 min, a 10-cc syringe attached to a 2.5 mm blunt cannula is inserted through a small stab incision in the abdomen. Fat is aspirated by using the syringe method [12]. The 2/3 of the aspirated fat is used in order to isolate the SVF. Digestion is done with 0.075 % collagenase (Sigma, St. Louis, MO) in buffered saline and agitated for 30 min at 37 °C in Celltibator (Medikan, Los Angeles, CA). Separation of the SVF containing ADSCs is then done by using centrifugation at 1200 × g for 5 min [10, 13]. The Lipokit Centrifuge (Medikan, Los Angeles, CA), is used. The SVF is located in the pellet derived from the centrifuged fat at the bottom of the lipoaspirate.

The remaining 1/3 of the aspirated fat is treated in the following manner in which the syringe is held vertically with the open end down through which the fat and fluid are separated. Isotonic saline is added to the syringe, the fat and saline are separated and the exudate discarded. The procedure is repeated until the fat becomes yellow in colour, free of blood and other contaminants [2, 4, 5, 14]. In Stromal Enriched Lipograft (SEL), freshly isolated SVF is attached to the aspirated fat, with the fat tissue acting as a living bioscaffold before transplantation [11] (Fig. 42.4). This whole procedure is done manually inside the operating theatre by 2 experts in tissue engineering for which they require about 90 min.

A 10-cc syringe attached to a 2.5 mm blunt cannula filled with the prepared fat is used. The adipose tissue graft enriched with SVF is woven in to the targeted chest area, injecting only a tiny amount with each pass as in order to obtain the most reliable clinical outcome. Tissue planes are created by using specific cannulas in different trajectories, always from the deeper aspect to more superficial areas. The fat is injected as the cannula is withdrawn in order to avoid intravascular fat injection.

Procedure Overview (Figs. 42.1, 42.2, and 42.3)

Postoperative Management

Intravenous antibiotics and analgesics are administered perioperatively and during the first 24 h. The patients are mobilized on the 1st day after surgery and discharged 24 h after the surgical procedure. At the time of discharge patients are advised to avoid lifting heavy objects and con-

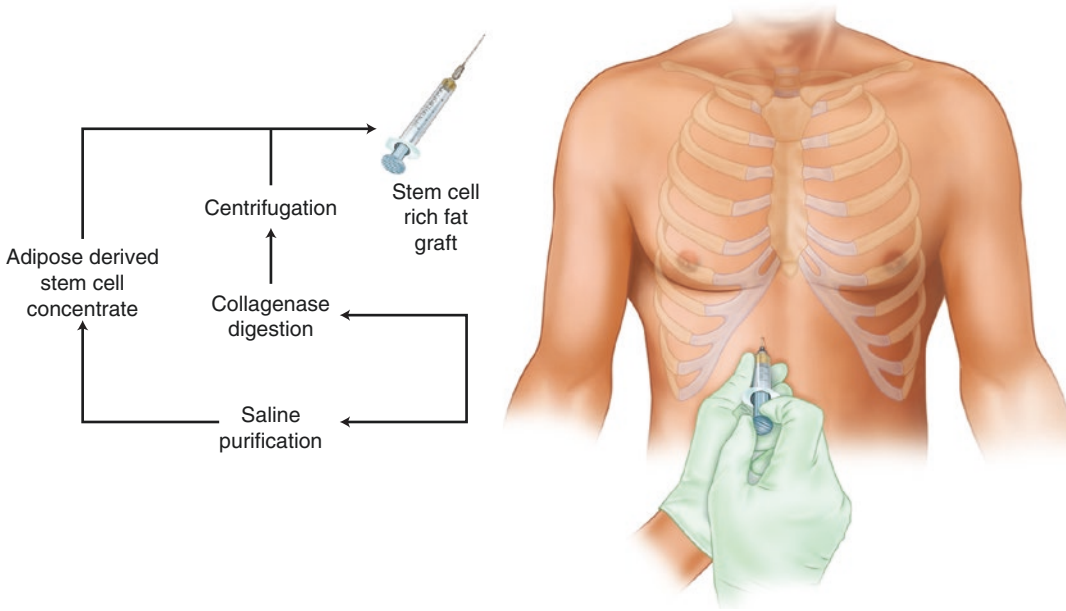

Fig. 42.1 Overview of the process to obtain stem cell rich fat graft

tinue for 7 days antibiotics and analgesic treatment. The patients are followed-up at 1, 2, 6 and 12 months after the surgical procedure. Patients overall satisfaction with their appearance of the treated areas after undergoing SEL is rated on a scale of 1–5, whereas 1 is 'poor', 2 is 'fair', 3 is 'good', 4 is 'very good' and 5 is 'excellent'. The patient satisfaction scale has been already used in published papers and has been peer reviewed [1, 7, 8]. Evaluation of patient satisfaction is done at least 24 months after the initial surgical procedure.

Results

A total of 13 female and 27 male patients were operated from January 2008 to January 2010. Age distribution of patients ranged from 19 to 52 years, with a mean of 22.3 years. The total amount of SEL transplanted to the chest area varied from 25 to 145 ml (mean, 62 ml). There were no intraoperative complications and no blood transfusions were required. There were no cases of liponecrosis and no liponecrotic lumps were palpated on postoperative evaluation. There were no cases of cellulitis of donor and grafted areas, no deep vein thrombosis and no pulmonary embolism. All the patients achieved satisfactory results undergoing just one session of SEL. At 24 months 70 % percent reported that their appearance after thoracic SEL contouring was 'very good' to 'excellent' (32 % 'excellent' and 38 % 'very good') and 23 % responded that their appearance was 'good'. Only 7 % of patients thought their appearance was less than good, (6 % 'fair' and 1 % 'poor').

Autologous fat tissue transfer has recently become extremely popular in the field of plastic and reconstructive surgery. Stromal Enriched Lipograft technique which was firstly described by the author and his team is based on autologous fat grafting enriched with ADSCs and produces superior results without the need for repeat treatment sessions, which are usually necessary with the non-stem cell enriched autologous fat transplantation [13]. The presence of adipose-derived stem cells in adipose tissue transplantation may contribute to neoangiogenesis in the acute phase by acting as endothelial progenitor cells or angiogenic-factor-releasing cells [17]. The number of functional ADSCs is likely to be important

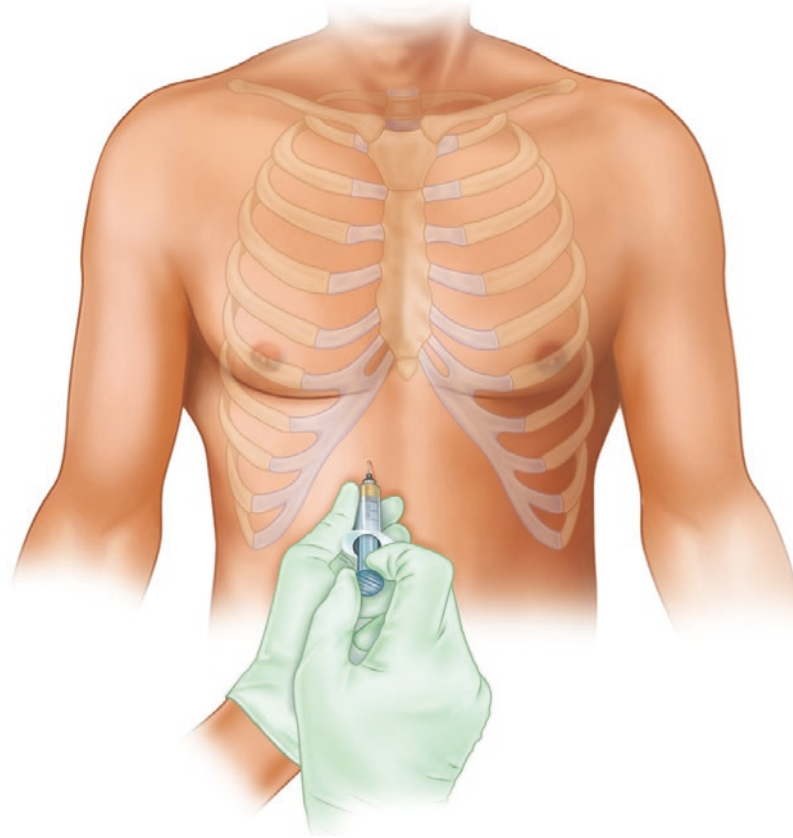

Fig. 42.2 After the injection of normal saline wetting solution containing 1:500,000 of adrenaline is administered by a small bore cannula and blunt cannula is inserted through a small stab incision in the abdomen. Fat is aspirated by using the syringe method

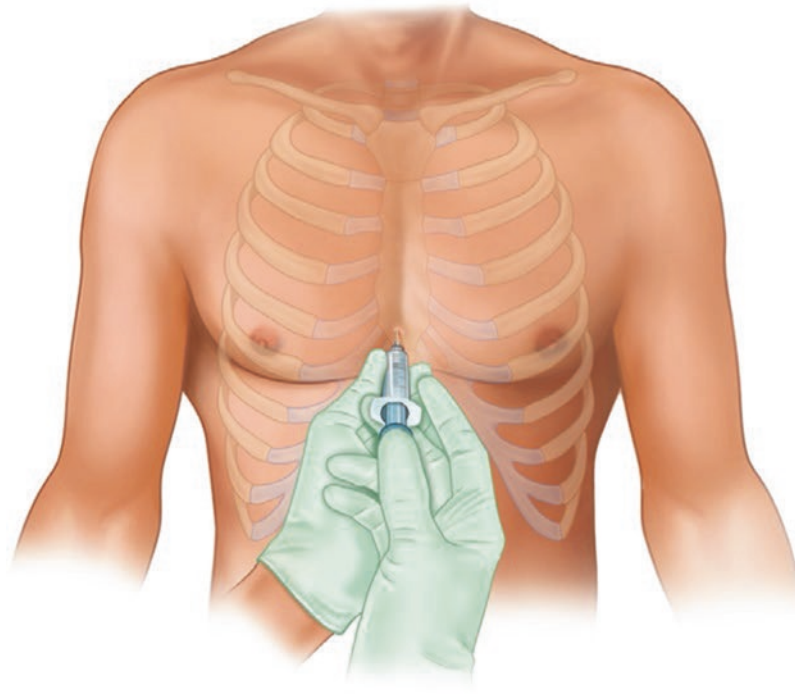

Fig. 42.3 The adipose tissue graft enriched with SVF is woven in to the targeted chest area, injecting only a tiny amount with each pass

for tissue repair and remodelling and ADSCs differentiate into vascular endothelial cells which contribute to neoangiogenesis in the acute phase of transplantation. ADSCs up regulate also their proneovascular activity in response to hypoxia, and may harbour the capacity to home to ischemic tissue and function cooperatively with existing vasculature to promote angiogenesis [3]. Successful SEL graft requires attention to patient preparation, meticulous planning, and optimizing the harvesting, storage, and transplantation of adipose tissue [16]. SEL graft is a living tissue that must be in close proximity to a nutritional and respiratory source to survive [15]. The SEL technique for mild pectus excavatum correction provides a safer alternative to allogenic implant use, resulting in the creation of a functional tissue which has a more natural look and carries fewer risks than currently available augmentation options. There were no major complications reported in this series of patients. In female patients, mild pectus excavatum can cause modified breast morphology, resulting in mammary asymmetry, which may be increased by placing mammary implants alone. Stromal Enriched lipograft is simultaneously applied for the correction of the pectus excavatum deformity. Fat transfer using the SEL technique for treatment of mild pectus excavatum achieves satisfactory aesthetic results with high patient satisfaction rates in just one surgical procedure.

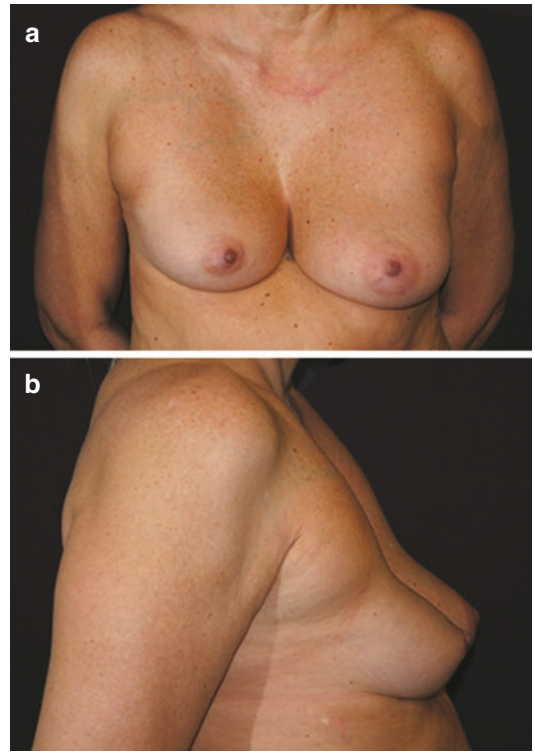

Fig. 42.4 (**a, b**) Preoperative views of a female patient with mild pectus excavatum

Procedure Related Images

(Figs. 42.4, 42.5, 42.6, 42.7, 42.8, 42.9, 42.10, 42.11, and 42.12)

Patient 1

A 33 year old woman with mild pectus excavatum requested aesthetic correction, combined with breast augmentation (Fig. 42.10a, b). Initially bilateral breast augmentation by insertion of silicone implants was performed (Fig. 42.8a, b). SEL chest contouring was performed with insertion of 65 ml of fat (Fig. 42.8c, d). SEL breast contouring of 35 ml was also performed in order to complement the breast augmentation (Fig. 42.9a). The follow-up for the patient has been 32 months with no complications and an excellent aesthetic result (Fig. 42.10c, d).

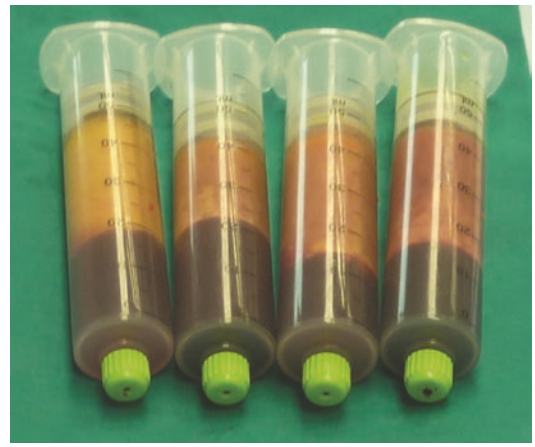

Fig. 42.5 Lipoaspirated fat using the syringe method

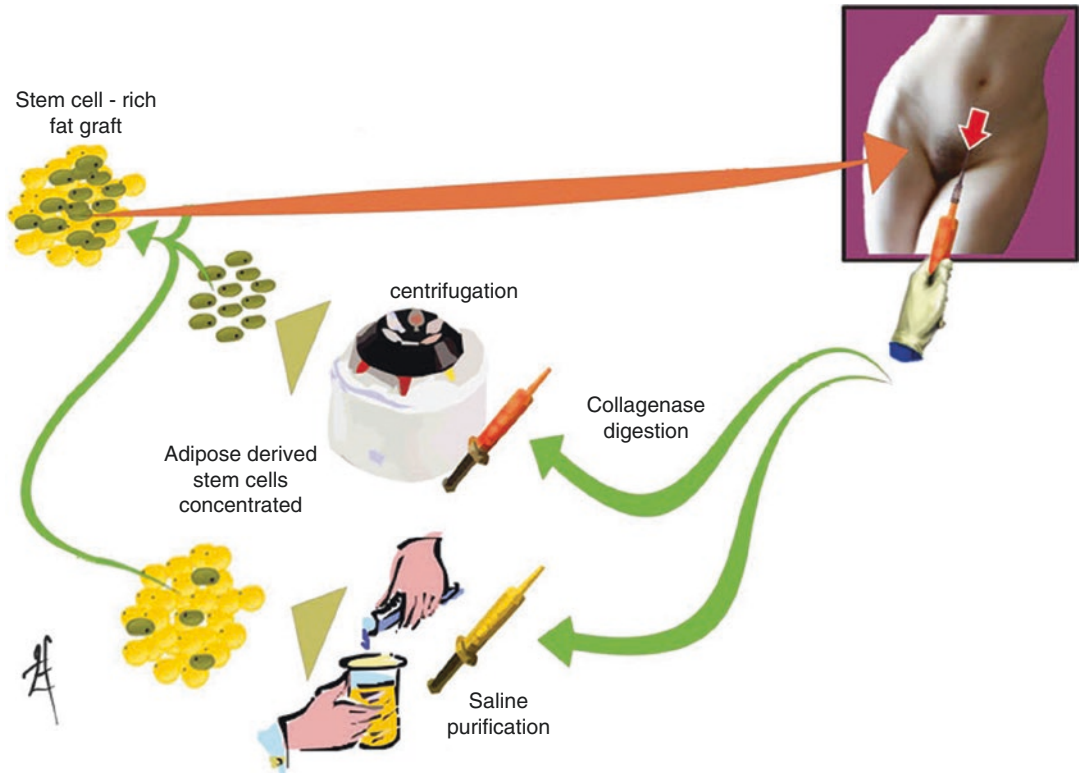

Fig. 42.6 Schematic representation of the stromal enriched lipograft technique

Patient 2

A 29 year old male was referred to our clinic because he was highly concerned about the aesthetic appearance of his pectoral area (Fig. 42.11a, b). The patient expressed the desire to undergo the least invasive intervention that could produce an aesthetically acceptable result. It was decided to perform the SEL chest contouring. A total of 110 ml was injected and the follow-up for the patient has been 35 months with no complications and a very good aesthetic result (Fig. 42.11c, d).

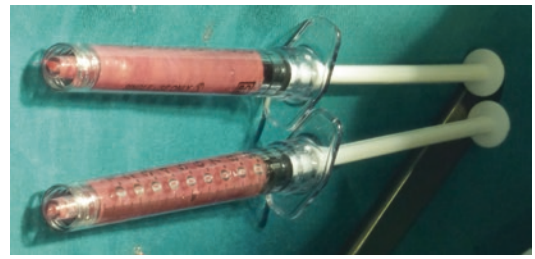

Fig. 42.7 Stromal enriched lipograft in 1 ml syringes for injection

Patient 3

A 33 year old woman with mild pectus excavatum requested aesthetic correction and simultaneous breast augmentation (Fig. 42.12a, b). Bilateral breast augmentation and SEL chest contouring was performed with insertion of 35 ml of fat. The follow-up for the patient has been 32 months with no complications and an excellent aesthetic result (Fig. 42.12c, d).

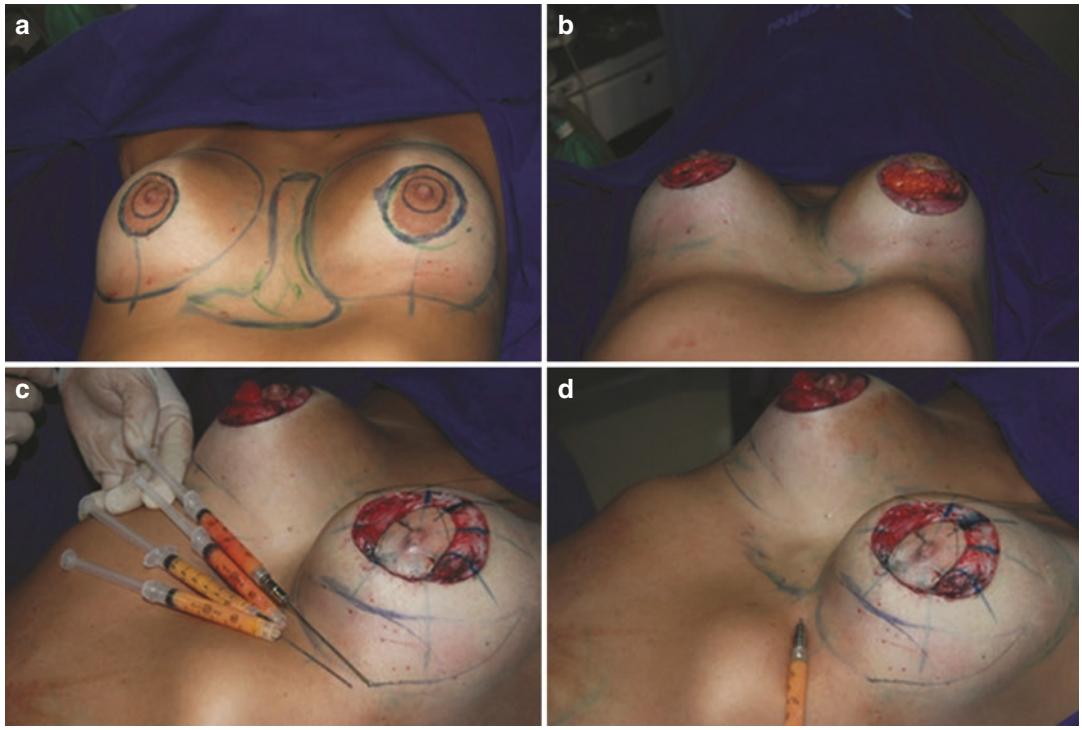

Fig. 42.8 (**a**,**b**) Intraoperative photos of mild pectus excavatum in a female patient after breast augmentation has been performed. (**c**, **d**) Intraoperative photos of stromal enriched lipograft for the correction of the mild pectus excavatum

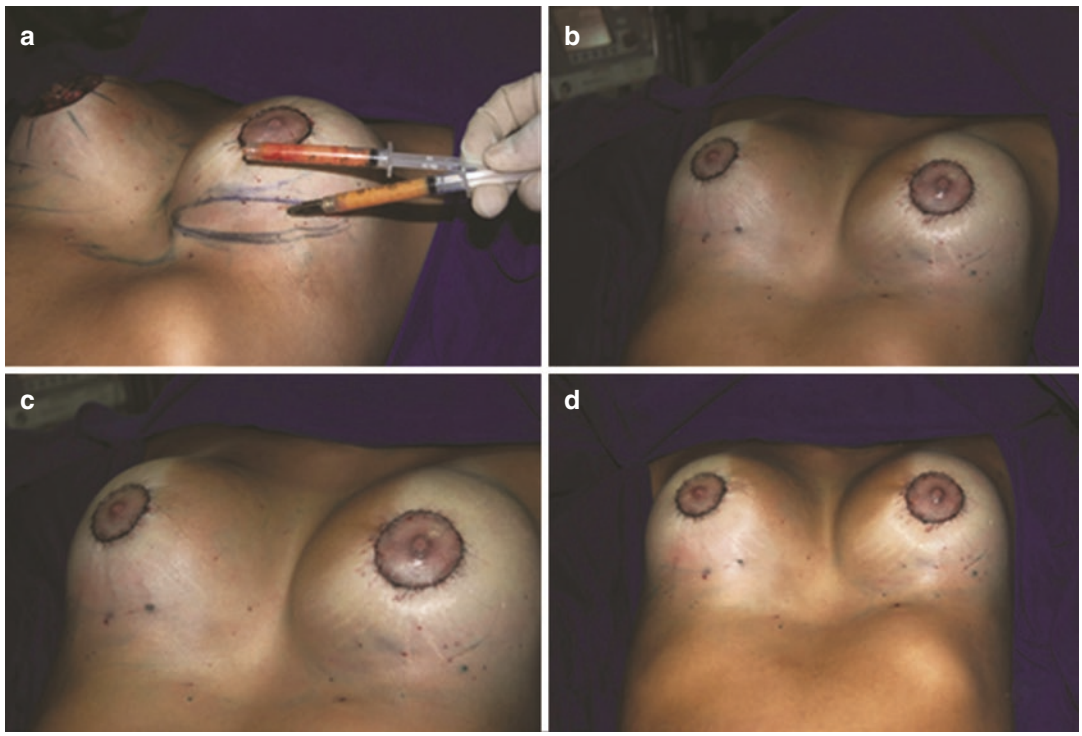

Fig. 42.9 (**a**) Intraoperative photo of Stromal Enriched Lipograft to the breast. (**b–d)** Intraoperative photos of the corrected mild pectus excavatum in a female patient

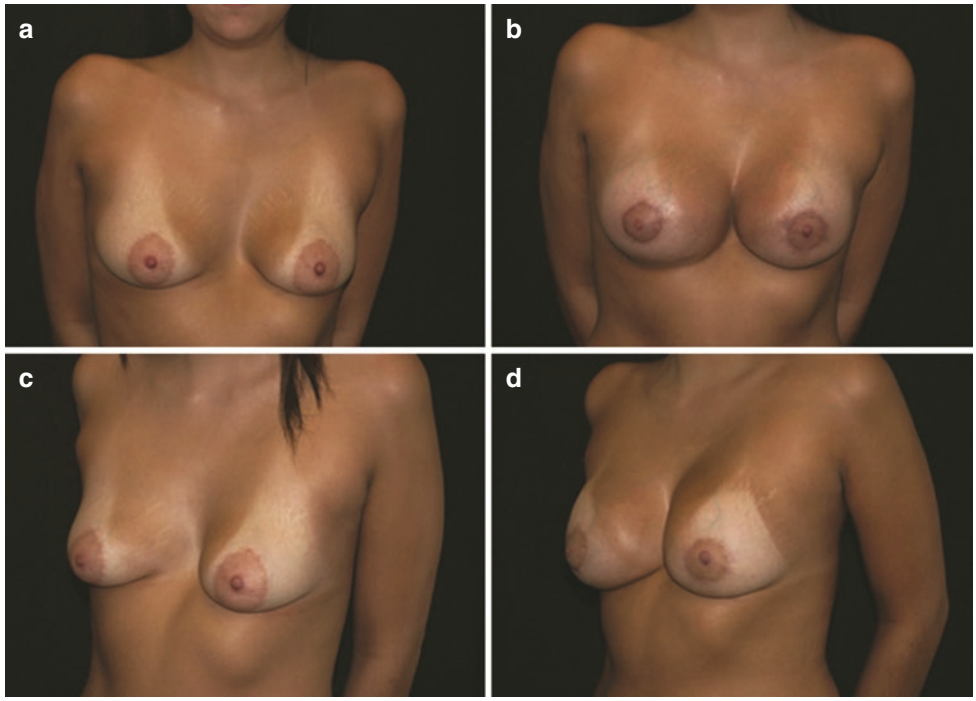

Fig. 42.10 (**a**, **b**) Preoperative photos of a female patient with mild pectus excavatum. (**c**, **d**) Postoperative photos of a female patient who underwent bilateral breast augmentation and Stromal Enriched Lipograft for the correction of mild pectus excavatum

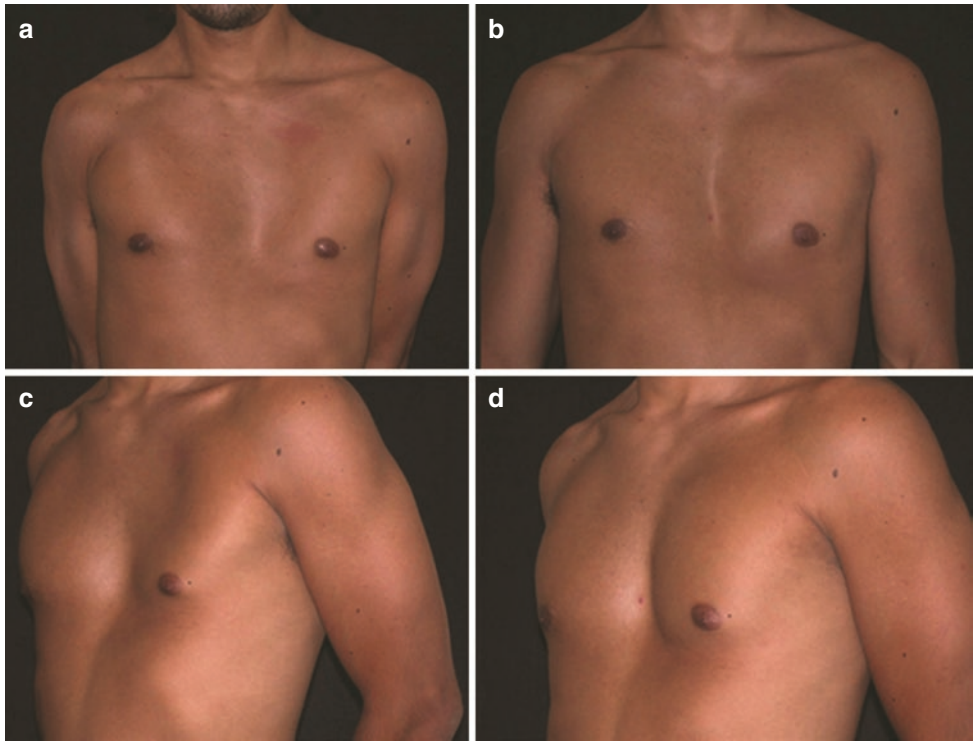

Fig. 42.11 (**a**, **b**) Preoperative photos of a male patient with mild pectus excavatum. (**c**, **d**) Postoperative photos of a male patient who underwent Stromal Enriched Lipograft for the correction of mild pectus excavatum

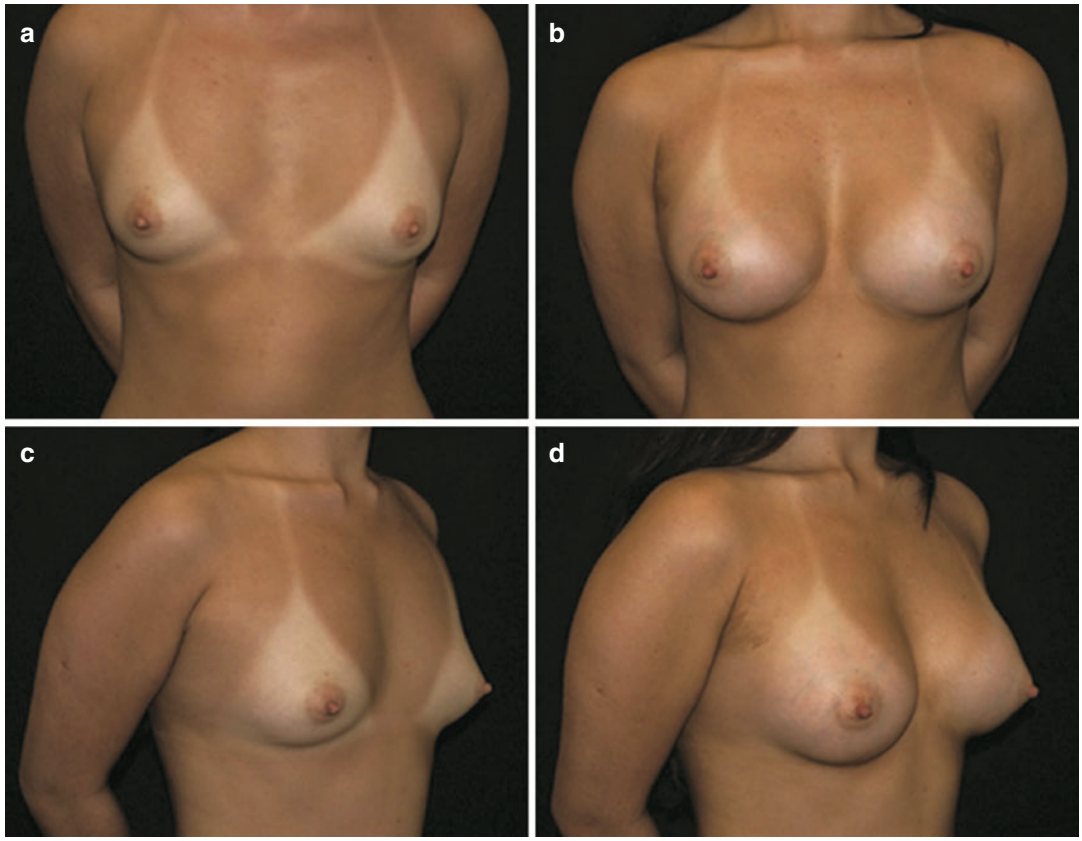

Fig. 42.12 (**a**, **b**) Preoperative photos of a female patient with mild pectus excavatum. (**c**, **d**) Postoperative photos of a female patient who underwent bilateral breast augmentation and Stromal Enriched Lipograft for the correction of mild pectus excavatum

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Customised Silicone Prostheses

43

Samer Saour and Pari-Naz Mohanna

Technical Highlights

Silicone prostheses have evolved as successful alternatives to correct chest wall deformities.

The main technical highlights of this procedure are:

(a) Careful patient selection
(b) Manufacture of the customised Silicone prostheses using either an alginate impression or a 3D CT scan reconstruction
(c) Choosing the best approach for the patient – an epigastric, axillary or infra-mammary incision can be used, with an infra-mammary incision being preferred in female patients.
(d) Careful dissection of skin and subcutaneous tissue off the chest wall to identify the defect without making the pocket too large.
(e) Careful haemostasis.
(f) Insertion of the customised silicone implant into the dissected pocket ensuring correct placement.
(g) Anchoring of the customised implant to the chest wall with sutures to prevent rotation, malposition and displacement.
(h) The use of suction drains to reduce fluid accumulation around the implant.
(i) Meticulous tension free skin closure in two layers.

Indications

Most types of Chest Wall Deformities can be corrected using this technique. However special indications for this technique are:

- Chest Wall Deformities associated with Pectus Excavatum
- Chest Wall Deformities associated with Poland's Syndrome
- Acquired post surgical chest wall deformities

Preoperative Consideration

1. A full history and examination.
2. Surgery should be delayed until the full growth potential has been achieved.
3. Full explanation of the customised silicone implant manufacturing process and surgery together with the associated complications.
4. Photographic documentation of the deformity
5. Anaesthetic review with assessment of any cardiovascular or respiratory sequelae.
6. Preoperative blood tests, chest X-Ray and lung function tests if indicated.

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7. Antibiotics are given at the start of the procedure and continued for 7 days post operatively.

Special Instruments

Besides the instruments used for general plastic surgical procedures, the following instruments are additionally required for this procedure:

1. A lighted retractor to facilitate dissection of the subcutaneous pocket
2. The customised silicone prostheses – the manufacturing of this is discussed below

Manufacture of the Customised Silicone Implant

Silicone implants are manufactured using a template of the chest wall defect, which can be created by either using an alginate impression or a 3-D CT scan reconstruction.

1. Alginate Impression
   An alginate impression is taken of the chest wall defect and then cast in hard dental stone thereby creating a working model of the chest wall. A wax pattern of the desired implant is constructed using the dental stone model (Fig. 43.1a) which is then tried on the patient and adjusted if needed. It is essential to keep the edges of the wax pattern as thin as possible to prevent subcutaneous visibility and palpability (Fig. 43.1b).
2. 3-D CT Scan Reconstruction
   A 3-D CT scan reconstruction software is used to create a model of the chest wall defect. Although this is a highly accurate technique it is more expensive than using an alginate impression. This method allows production of a silicone implant which is nearly identical to the chest wall defect and is particularly useful when treating female patients.

This technique involves taking a CT scan of the affected area. The actual chest wall defect is then marked on the CT scan (Image 2a) and then rapid prototyping is used to produce a 3-D working model of the deformity (Image 2b). Rapid prototyping is a process that builds models directly from computer data in a layered manner, however this process is very time consuming and can take up to 12 h to complete.

Regardless of which method is used to prepare the template, the mould construction remains the same. The mould is created by removing the wax template, leaving a void the surface of which is roughened to texture the implant in order to prevent capsular contracture. The mould is then lined with foil before injecting implant grade silicone (NuSil Med 4805, Nusil, Sophia Antipolis cedex, France) into the void.

The silicone implant is then removed from the mould and holes incorporated into the implant to allow both tissue integration and facilitate suturing of the implant to the chest wall. These refinements help to prevent malposition and displacement of the silicone implant. It is important that the implant is soft and flexible thereby accommodating for changes in body posture.

Surgical Technique

The choice of incision depends on the type of chest wall defect and the gender of the patient. Commonly used incisions are infra-mammary, epigastric and axillary with infra-mammary being preferred in female patients. In male patients requiring a customised silicone implant for chest wall deformities associated with Poland's syndrome an axillary or epigastric approach is often used. Preoperative markings are made of the defect with the patient supine.

An incision is made through the skin and subcutaneous tissue down to pectoralis major fascia or chest wall depending on which incision has been selected. The desired pocket is dissected using diathermy to reveal the defect requiring reconstruction. A lighted retractor is ideal as it allows elevation/retraction of the skin and subcutaneous tissue whilst providing illumination of the pocket.

Meticulous haemostasis is crucial to reduce the risk of haematoma. It is essential that the pocket is not made larger than the prostheses as this will increase the likelihood of rotation, malposition and displacement. Once the desired

pocket has been created it is washout out with betadine and then normal saline.

A 16 gauge suction drain is then inserted into the pocket. A change of surgical gloves is recommended prior to handling the silicone implant. The implant is removed from the sterile packaging and covered completely with betadine. It is then inserted into the pocket manually whilst the assistant uses a deaver surgical retractor to elevate the overlying skin and subcutaneous tissue. The implant is then adjusted until it is in the correct position thereby correcting the chest wall defect.

2/0 Vicryl sutures are then used to secure the silicone implant to the chest wall through the holes that were incorporated into the implant during the production.

A tension free closure of the incision is carried out in two layers, using 3/0 Vicryl for the dermal layer and a 3/0 Monocryl subcuticular suture for the skin. Skin glue is applied to the incision and the patient is placed into a vest compression garment.

Procedure Overview (Figs. 43.1, 43.2, and 43.3)

Postoperative Management

A dose of perioperative intravenous antibiotics are administered; with Co-Amoxiclav being the drug of choice. These are then continued orally for a period of 7 days post operatively. All patients are given appropriate analgesics and encouraged to mobilise that same evening. The drain output is monitored on a daily basis and only removed when the

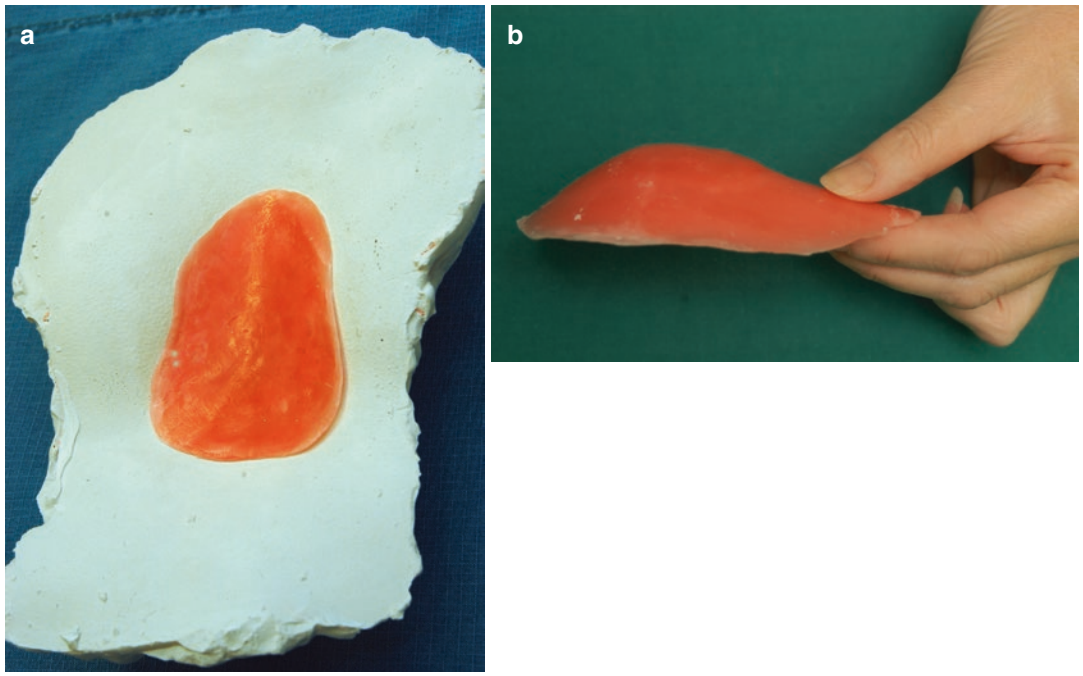

Fig. 43.1 (**a**) A wax pattern of the desired implant shape is constructed using the working model of the chest wall defect. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. *Journal of Plastic, Reconstructive & Aesthetic Surgery*. October 2008. Vol 61, Issue 10. With permission from Elsevier. (**b**) The wax pattern showing very thin edges ensuring no subcutaneous show of the implant. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. *Journal of Plastic, Reconstructive & Aesthetic Surgery*. October 2008. Vol 61, Issue 10. With permission from Elsevier

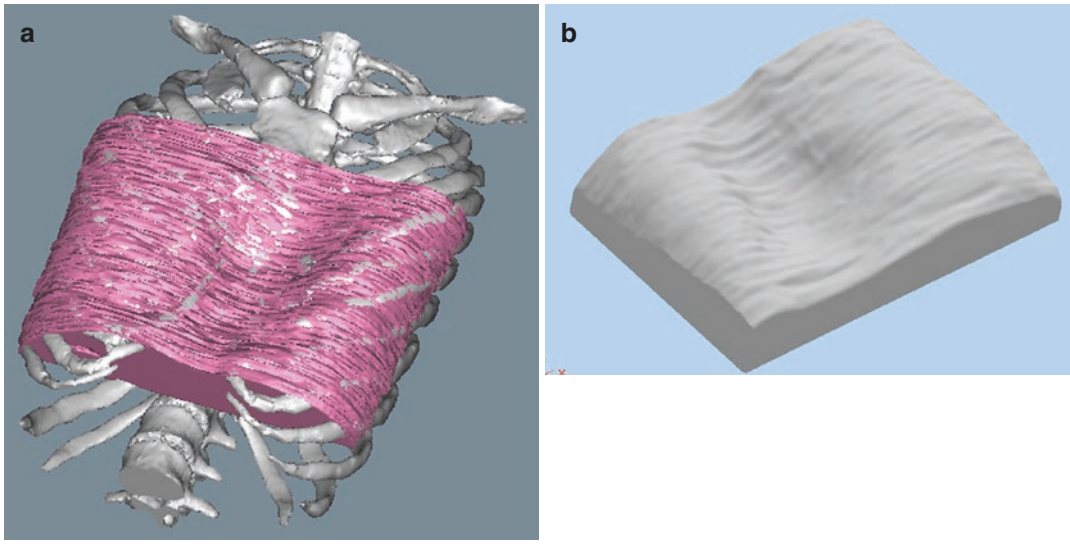

Fig. 43.2 (**a**) The size of the model required is marked on the CT data and is represented by the pink area. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. *Journal of Plastic, Reconstructive & Aesthetic Surgery*. October 2008. Vol 61, Issue 10. With permission from Elsevier. (**b**) This computer generated model represents the area chosen and this will then be used to construct an exact working model of the chest wall defect by rapid prototyping. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. *Journal of Plastic, Reconstructive & Aesthetic Surgery*. October 2008. Vol 61, Issue 10. With permission from Elsevier

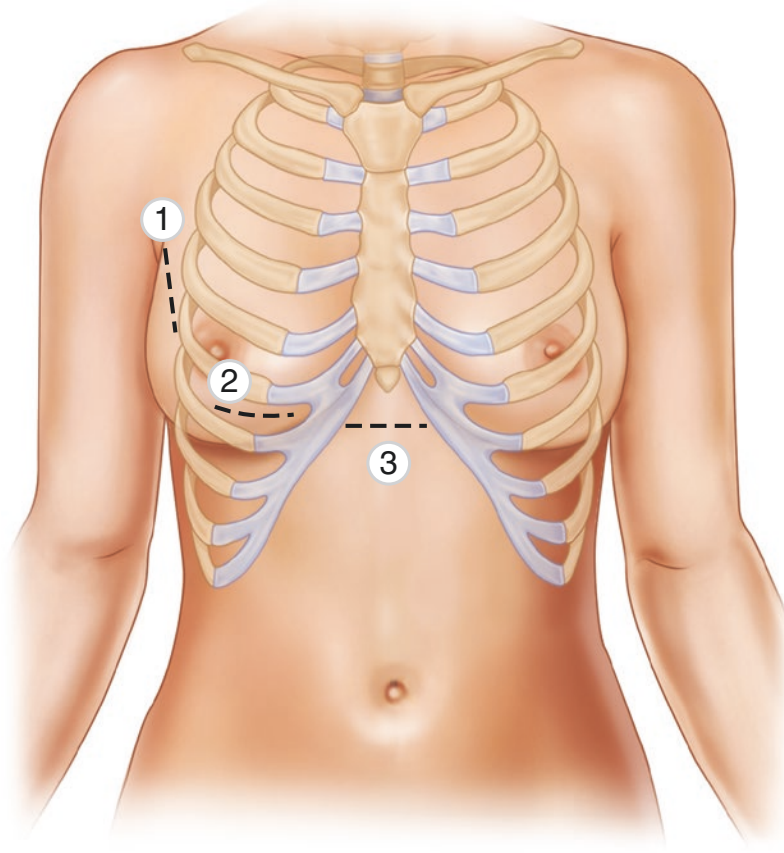

Fig. 43.3 Options in placement of incision for implantation of silicon prosthesis. (1) Axillary incision, (2) Sub-mammary incision and (3) Epigastric incision

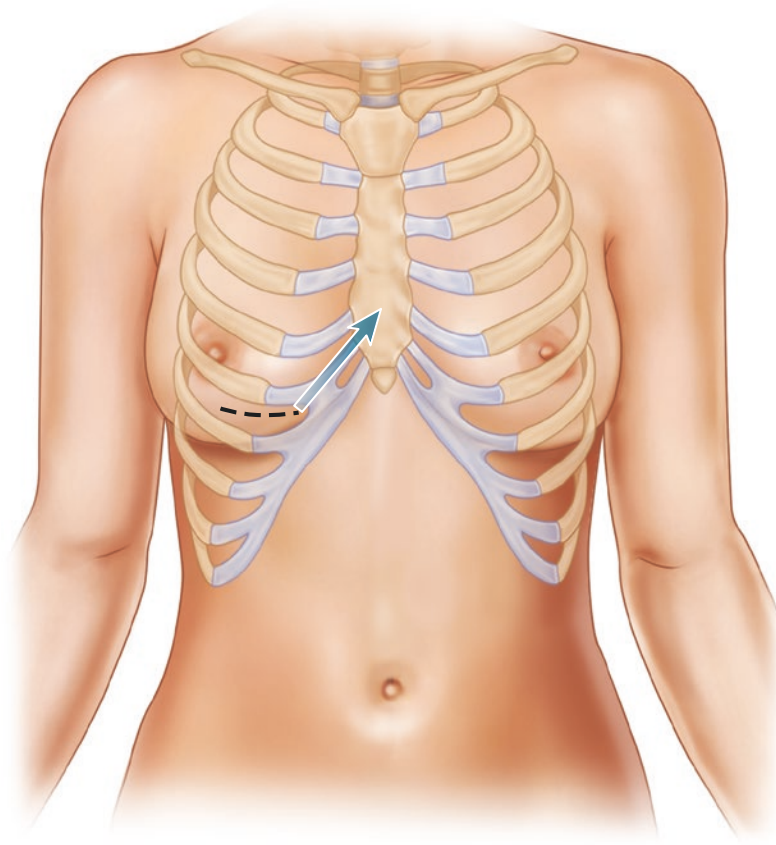

Fig. 43.4 The pocket created is washed with betadine first and then with saline. Meticulous hemostatis is crucial. The silicone implant is then inserted and positioned into the defect

patient is fully mobilised with an output of <30 ml in a 24 h period. Once the drain has been removed the patient is discharged home with strict instructions to use the pressure garment for 6 weeks, 24 h a day only removing it to shower and to avoid contact sports for at least 6 weeks. The patients are followed up in the Plastic Surgery dressing clinic at 1 week following discharge and Plastic Surgery outpatients department at 6 weeks and 3 month. Figure 43.6a, shows a 25 year old female patient with pectus excavatum, who underwent reconstruction of her chest wall deformity using a customised silicone implant produced by 3D CT scan reconstruction. Figure 43.6b shows visible improvement post insertion of the customised silicone implant via a right inframammary incision.

Potential Complications

- Bleeding
- Infection
- Seroma
- Implant rotation/malposition/displacement
- Capsular formation

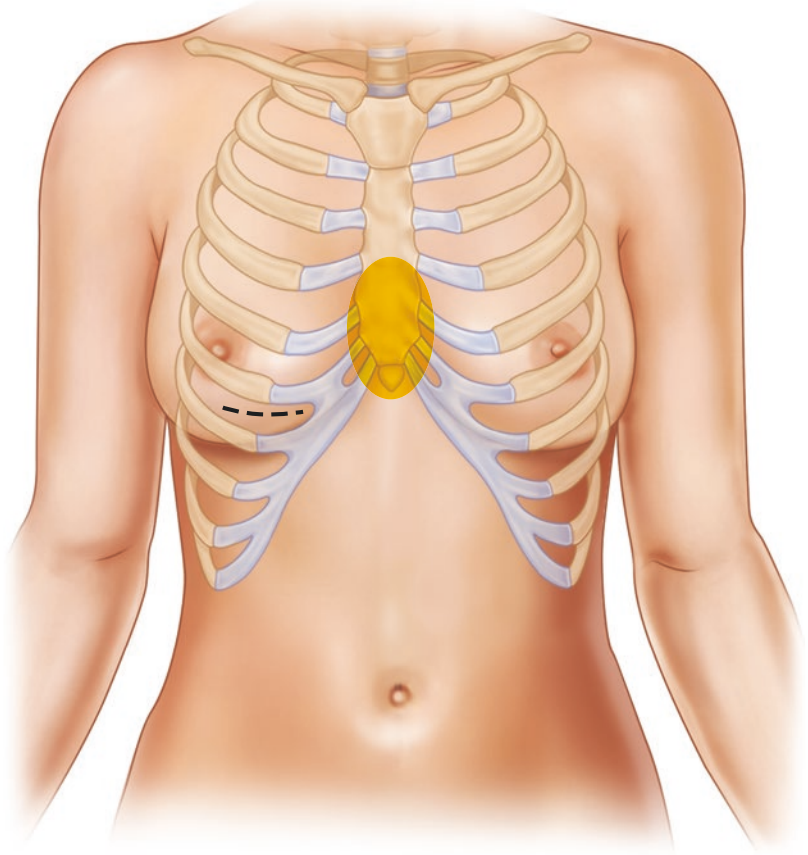

Fig. 43.5 In a female patient with pectus excavatum a submammary incision is preferred

Summary

An array of procedures have been described to address chest wall deformities including the use of fillers, pedicled muscle flaps and procedures involving alteration of the skeletal structure such as the Ravitch operation. These more complex procedures are associated with a higher risk of complications.

The technique of customised silicone implants for the reconstruction of chest wall defects such as Pectus Excavatum, Poland's syndrome and acquired chest wall defects is a less complex method of reconstruction with fewer associated complications.

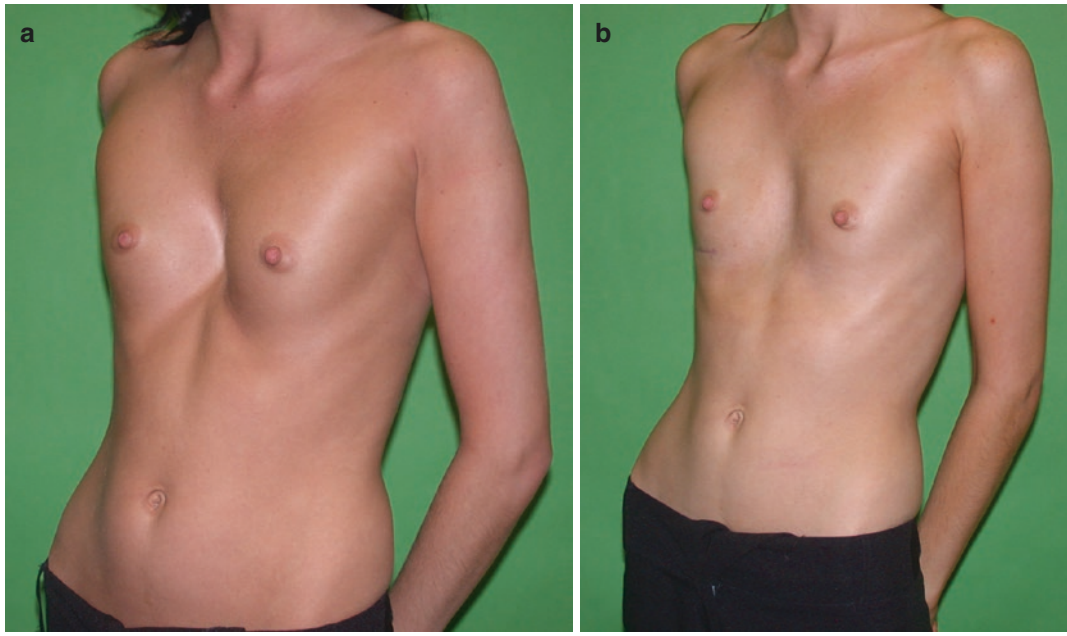

Fig. 43.6 (**a**) Obvious pectus excavatum defect of the chest wall. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. Journal of Plastic, Reconstructive & Aesthetic Surgery. October 2008. Vol 61, Issue 10. With permission from Elsevier. (**b**) Visible improvement post insertion of the customised silicone implant. Note the right inframammary incision. From Samer Saour, Hassan Shaaban, Jane McPhail, Paul McArthur. Customised silicone prostheses for the reconstruction of chest wall defects: technique of manufacture and final outcome. *Journal of Plastic, Reconstructive & Aesthetic Surgery*. October 2008. Vol 61, Issue 10. With permission from Elsevier

Procedure Related Images
(Figs. 43.6a, b)

Suggested Reading

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Part V

Pectus Carinatum: General Aspects

Overview of Repair of Pectus Carinatum Type of Deformities

44

Amulya K. Saxena

Introduction

Pectus carinatum, or protrusion deformity of the chest, occurs less commonly than pectus excavatum. It has been estimated that pectus carinatum comprises about 15 % of patients with chest wall deformities [1]. The protrusion may be in the manubrium and the superior costal cartilages [1], which is termed a chondromanubrial deformity or pigeon breast. However, chondrogladiolar is the most common variety which is the symmetric or asymmetric protrusion of the gladiolus and inferior costal cartilages. With asymmetric chondrogladiolar type, unilateral overgrowth of the costal cartilages results in a rotational deformity of the sternum and a keel-like appearance of that side of the chest. Protrusion in pectus carinatum patients may be presenting unilateral, bilateral, or mixed [2]. There is a male preponderance of 4:1. Although the etiology of pectus carinatum is still unclear [3], a genetic component has been confirmed in close to 25 % of patients with a family history of chest wall defect [1]. Pectus carinatum continues to progress over a variable length of time, often over a period of years into adolescence (Fig. 44.1). Pectus carinatum can occur in patients following surgery for pectus excavatum [4] (Fig. 44.2).

The natural course of this deformity differs from pectus excavatum. There is generally a delay in clinical presentation of pectus carinatum when compared to pectus excavatum, this may be due to the fact that the protruding infantile abdomen accentuates pectus excavatum in early childhood and recession of the abdominal contour which occurs in the later years emphasizes the protrusion defect in early adolescence. Most patients are asymptomatic, and when symptoms occur, they are confined to tenderness at the site of the protrusion [3, 5]. Associated mitral valve disease has also been reported [6, 7]. In patients without congenital heart disease, cardiopulmonary symptoms resulting from the condition has not been reported. Other associations include Marfan's syndrome and scoliosis [1]. Connective tissue disorders involving structural abnormalities of the major vasculature and heart valves have also been associated with pectus carinatum which have been described in a separate Chapter on Syndromes.

Apart from the possible physiologic consequences, pectus carinatum can have a significant psychological impact. Patients with milder cases are less affected and often do not seek medical attention than those with moderate or severe forms. Patients with moderate or severe pectus carinatum are affected by the distorted shape of the chest that harms their self-image and confidence, which

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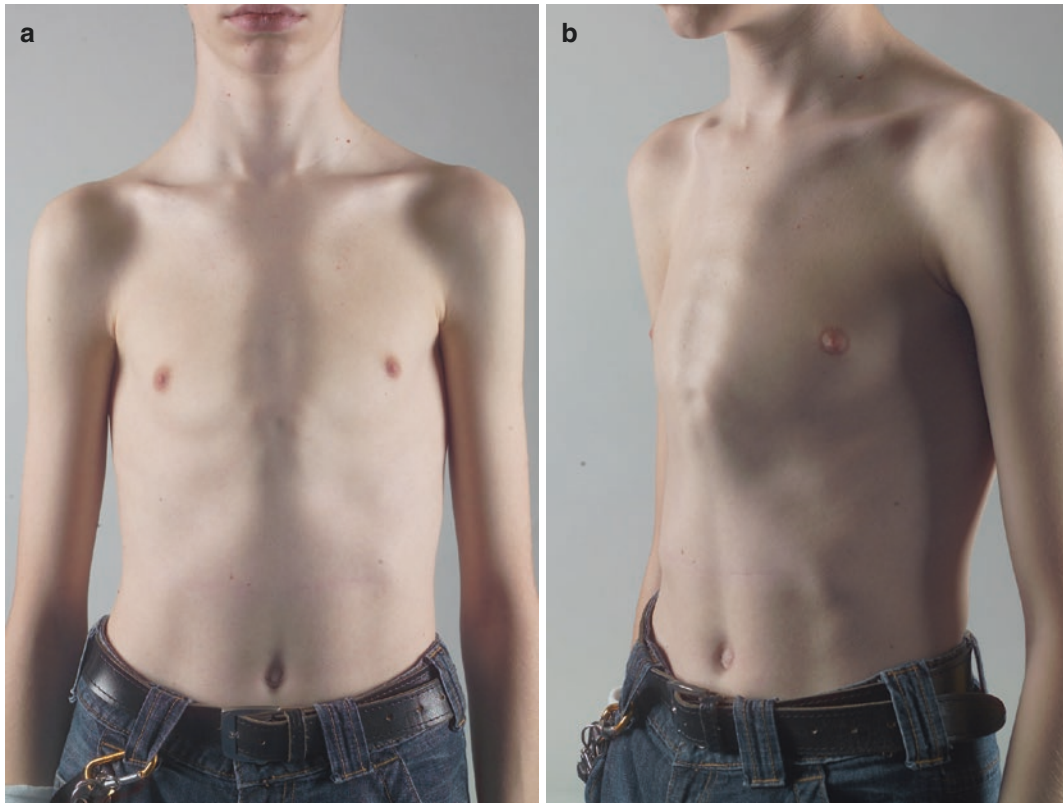


Fig. 44.1 Patient with a pectus carinatum exhibiting the protruding chest wall deformity (**a**- frontal view and **b**- lateral view)

could be responsible for disruption of social connections and a cause for them to feel uncomfortable throughout adolescence and adulthood.

Overview of Procedures

Pectus carinatum (pigeon chest), a protrusion deformity is the second most common malformation of the anterior chest wall. Ravitch in 1952 was the first to suggest that surgical correction was the only effective method for the treatment of this deformity and corrected the chondromanubrial prominence by resecting the multiple deformed costal cartilages and performing a double osteotomy (Fig. 44.3) [8]. His technique involved the dettachment and reflecting the pectoral as well as rectus abdominis muscles. The prominence of the sternum was exposed. The double osteotomy did not involve the deformed part of the sternum which was left intact, but involved a cuneiform osteotomy at the manubriosternal junction permitting the corp body of the sternum to be elevated forward followed by a second osteotomy in the reverse direction in the mid-sternum permitting the distal portion of the sternum to be flexed posteriorly, in order to give a better surgical result. The deformed cartilages closed to the sternum were shaved away without any manipulations on the costo-sternal junctions. Deformed central portions of the cartilages were excised after incising the perichondrum, after which the perichondrium was reefed with mattress sutures to form a taut support for the pleura in its new expanded position. Ravitch later abandoned the approach of sternal osteotomy when he presented his modified approach 8 years later in 1960.

Lester in 1953 performed a repair involving resection of the anterior part of the sternum, but abandoned this approach due to unsatisfactory results. He then reported a second less radical technique which involved subperiosteal resection

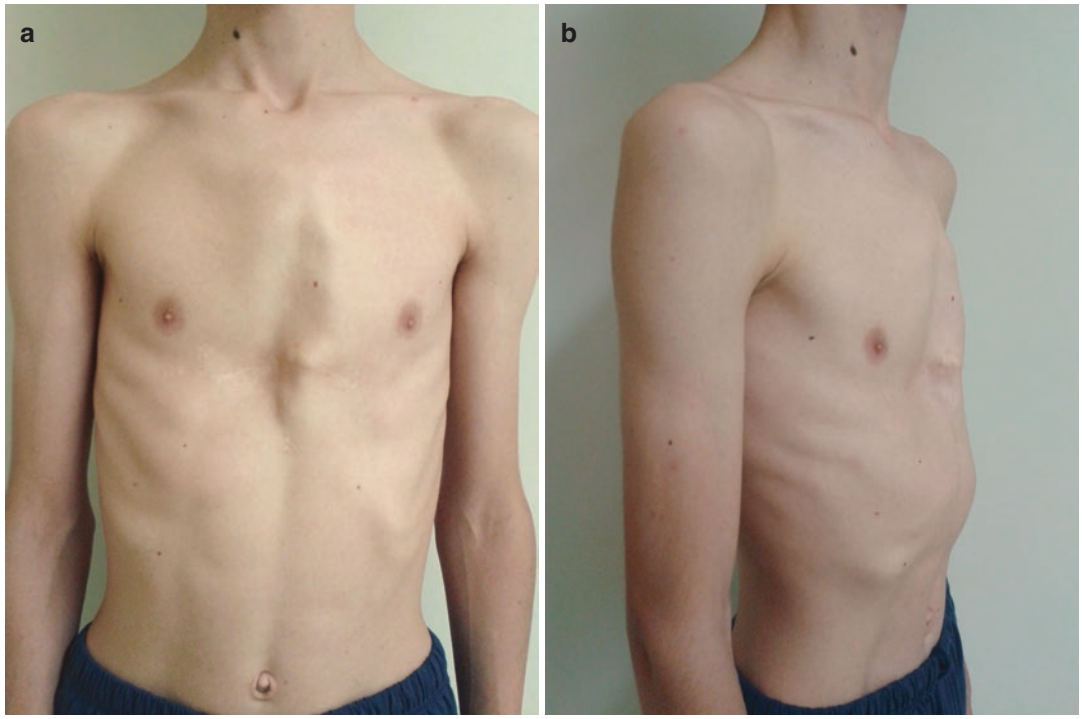
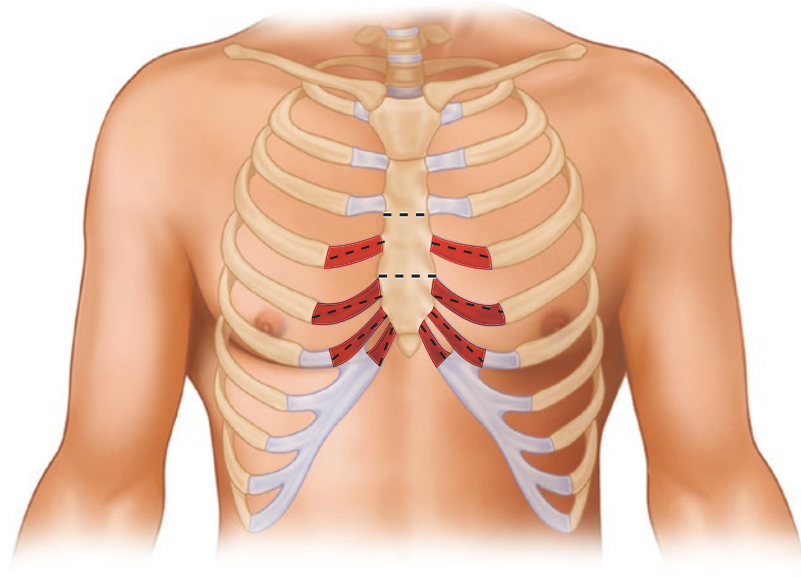


Fig. 44.2 Patient with reactive pectus carinatum after open repair for pectus excavatum (**a**- frontal view and **b**- lateral view)

Fig. 44.3 Schematic view of pectus carinatum repair as performed by Ravitch



of the lower body of the sternum and sternal ends of the costal cartilages [9]. In his technique for sternal protrusions performed a longitudinal incision over the midline of the sternum and the skin along with subcutaneous tissue was reflected (Fig. 44.4). Both pectoralis major muscle were detached from their sternal insertions and also reflected. This was followed by subperichondrial

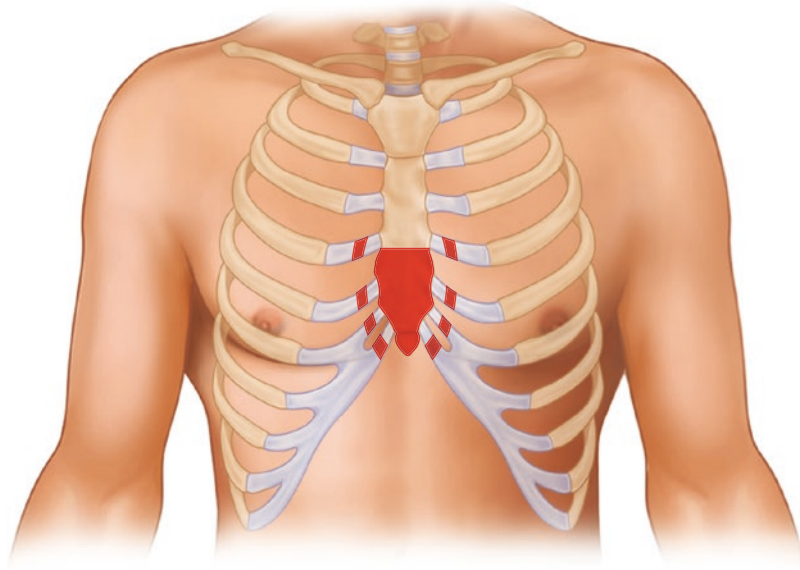


Fig. 44.4 Schematic view of pectus carinatum repair as performed by Lester

resection of about 2 cm of the sternal end of each deformed cartilage attached to the sternum. The sternal periosteum was then incised longitudinally and the periosteum stripped laterally from the outer surface of the lower four segments of the sternum. The xiphoid was then carefully detached by a transverse incision going down to the substernal ligament, and this, with the periosteum of the under surface of the sternum, was stripped off, leaving the denuded sternum free. The denuded sternum was removed by transecting it in the second segment. The pectoralis major muscles are sutured together snugly across the midline and the wound closed. A pressure dressing may be applied to maintain the depression. On the other hand Lester applied a different technique similar to a thoracoplasty. In the less severe deformities he resected only the involved rib and cartilage through with an incision over the deformity. If there is a bilateral involvement of the costal arch only, both sides were operated upon at the same time, using similar incisions for the sake of uniformity.

If the deformity was extensive, Lester avoided complete removal of the deformed ribs at one operation to avoid the risk dangerous paradoxical respiration [9]. For that reason he performed the operation in two stages. The first stage was done through an incision placed parallel to the protruding ridge. Subperichondrial resection of the involved cartilages was then performed after gaining access to the cartilages by splitting the fibers of the pectoral muscles, so as to avoid injury to the muscle attachments to the sternum. The effect of this stage was to leave the deformity practically unchanged. The second stage was done a week or 10 days later, through a longitudinal incision in the axilla. The involved ribs were resected subperiosteally from the costochondral junctions to the anterior axillary line or farther if the deformity requires it. The pectoral muscles were retracted for exposure. This stage corrected the deformity. The objection to the two-stage operation was that there were two scars, one of which, the anterior, being noticeable.

Chin in 1957 and Brodkin in 1958 employed an operative procedure which used the traction effect of the rectus muscles to relocate and maintain the sternum in a corrected position (Fig. 44.5) [2, 10]. Chin, suggested that the anterior portion of the diaphragm, in patients with pectus carinatum, was attached to the rectus abdominis and not to the xyphoid and to the lower cartilages. He pointed out that the Harrison's grooves are accentuated on inspiration, the lower sternum moves forward on inspiration, but that the rectus sheath dimples near the tip of the xyphoid on inspiration. All of this was presumed by him to be

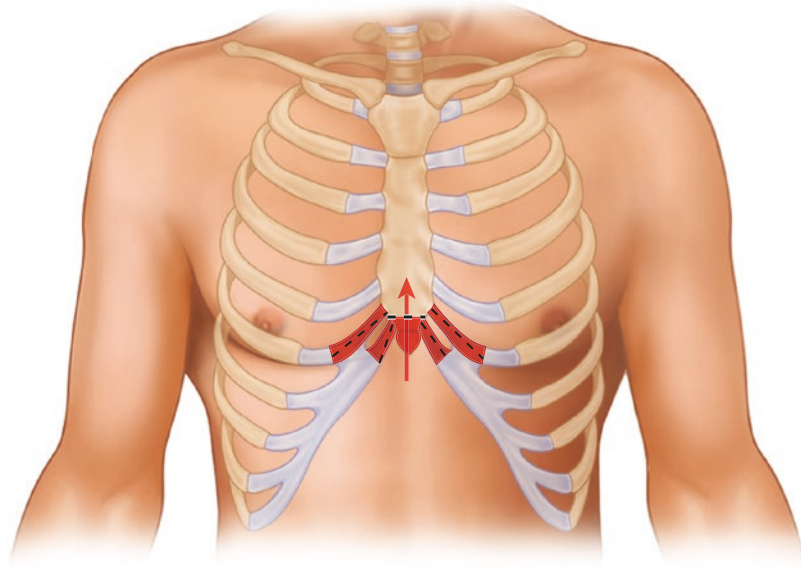


Fig. 44.5 Schematic view of pectus carinatum repair as performed by Chin

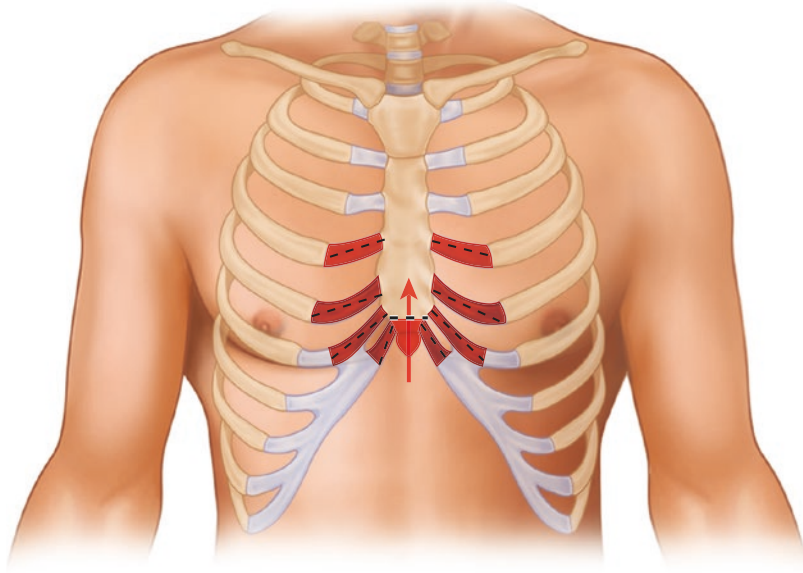


Fig. 44.6 Schematic view of pectus carinatum repair as performed by Howard

associated with over-development of the anterior-lateral portion of the diaphragm. His operation, consisted of bilateral resection of the medial portions of the sixth and seventh costal cartilages sub-perichondrially, detaching the xyphoid and reinserting it into a slot made for it in the sternum, at the level of the fourth costal cartilage. In younger children it was presumed to pull the sternum back into proper position with the passage of time.

Howard from Melbourne Australia, in 1958 further modified this method by radical resection of cartilages and a sternal osteotomy (Fig. 44.6) [11]. He employed the same maneuver as reported by Chin but added to it the subperichondrial resection of all of the deformed costal cartilages.

Ravitch, in 1960, reported a surgical procedure that left the sternum alone, but involved the resection of the affected costal cartilages along with the shortening of perichondrial strips with reefing

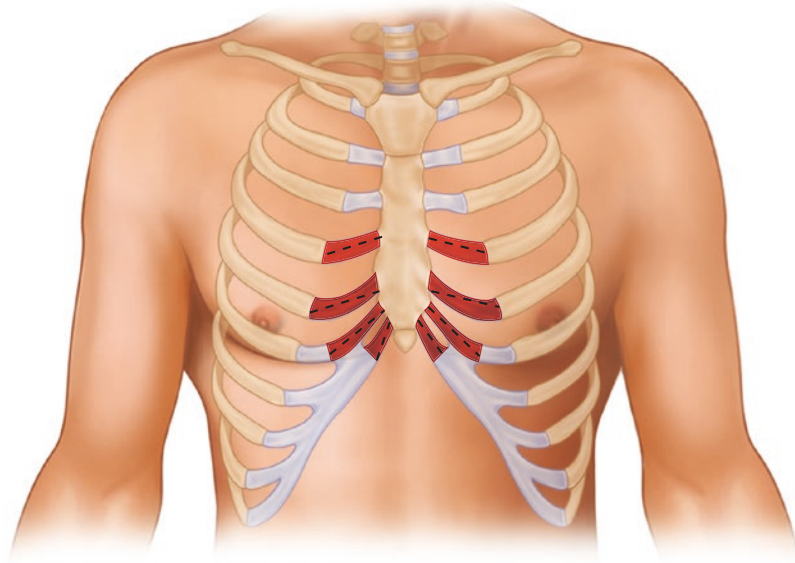


Fig. 44.7 Schematic view of pectus carinatum repair as performed by Ravitch

sutures (Fig. 44.7) [12]. The first operations performed by Ravitch were a two-stage procedure as he feared that the patient would not tolerate a bilateral procedure. However, subsequent procedures were performed with bilateral corrections in the single staged procedure. Procedures were performed by dissecting and reflecting the pectoral as well as rectus abdominis muscles. The prominence of the sternum that was due to the deformed cartilages was exposed. The deformed cartilages closed to the sternum were sliced away, leaving the costo-sternal junctions intact. Involved central portions of the cartilages were excised after opening of the perichondrum, after which the redundant perichondrium was reefed with mattress sutures to form a taut support for the pleura. The procedure was completed with the reattachment of the pectoralis and rectus adominis muscles.

In 1963, Ramsay utilized a rectal muscle flap to fill the lateral defects resulting from the protrusion without altering the position of the sternum or resecting the deformed costal cartilages (Fig. 44.8) [13].

Also in 1963, Robicsek reported a technique that involved subperichondrial resection of the deformed lateral asymmetric costal cartilages, transverse sternal displacement, and resection of the protruding lower portion of the sternum along with the reattachment of the xiphoid and rectus muscles to the new lower margin of the sternum (Fig. 44.9) [14]. Welch in 1973 and Pickard in 1979 reported techniques which were similar and involved costal cartilage resection and sternal osteotomy (Fig. 44.10) [15, 16].

In Münster, Germany, thoracic reconstruction operation for pigeon chest type deformity was performed on 111 patients during a 13 year period (1984–1997) using the Willital-Hegemann technique (Fig. 44.11) [17]. A vertical midline incision was made in boys; otherwise in girls, a submammary incision that was curved upward at the midpoint (over the deepest point of the sternal defect) was preferred. Skin, fat and pectoral muscles are then reflected in a single flap and the entire impression of the deformity generally formed by 5–8 pairs of ribs is exposed (third to tenth rib). The subperichondrial dissection is carried out and the deformed costal cartilages were resected parasternally from their junction with the rib to within 1 cm of the sternum as well as at the level of transition to the normal ribs. The attachment of the rectus muscle to the sternum was severed in order to elevate the sternum and dissected it free from its anterior mediastinal tissue attachments. A partial transverse sternal wedge osteotomy was performed at the Angle of

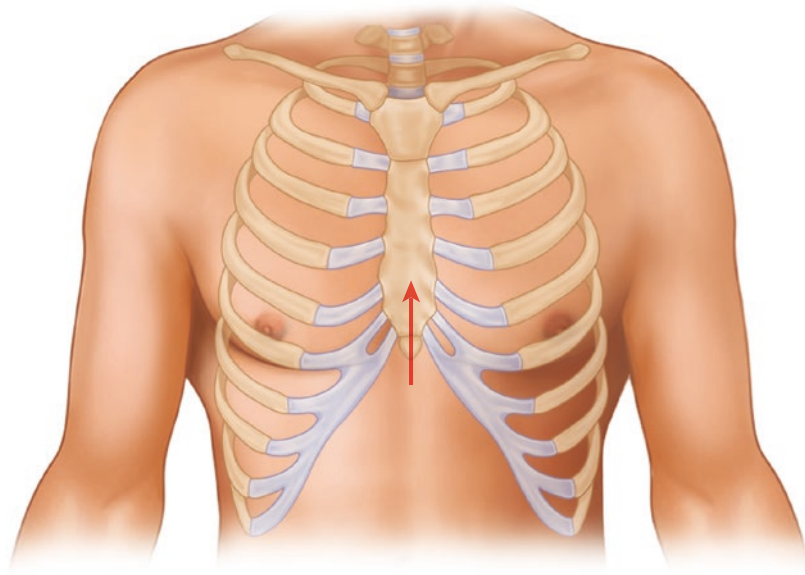


Fig. 44.8 Schematic view of pectus carinatum repair as performed by Ramsay

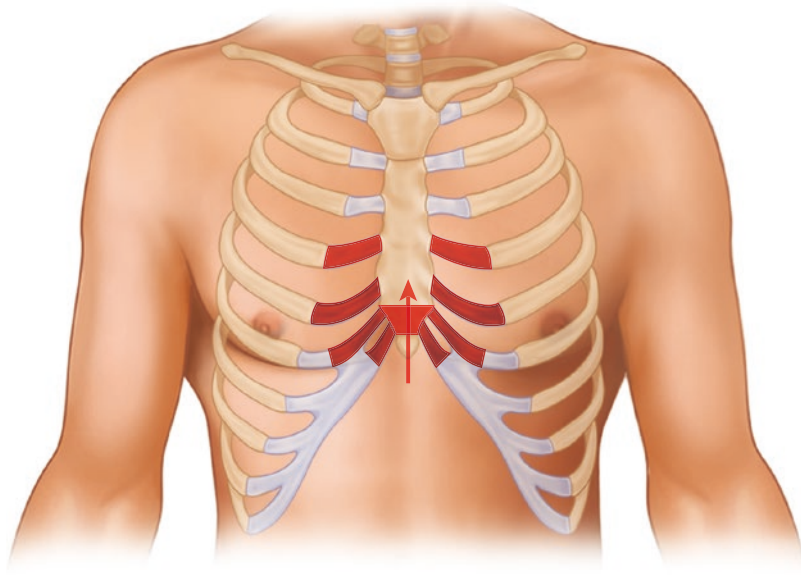


Fig. 44.9 Schematic view of pectus carinatum repair as performed by Robicsek

Ludovici and the sternum is modeled to conform to anatomic contours. A perforated stainless steel strut was then passed trans-sternally, and the sternum was repositioned to its corrected position. Two parasternal metal struts were also employed, with the points of fixation being the second rib and the lowest end of the rib cage. The trans-sternal strut is fixed to the two parasternal struts with stainless steel wires for additional support. The pectoral muscle flaps and the severed rectus muscles are then sutured and fixed to the sternum.

Minimal Access Repair of Pectus Carinatum

In 2009, Abrahmson reported a 5-year experience with minimally access surgical technique for treatment of pectus carinatum on 40 patients

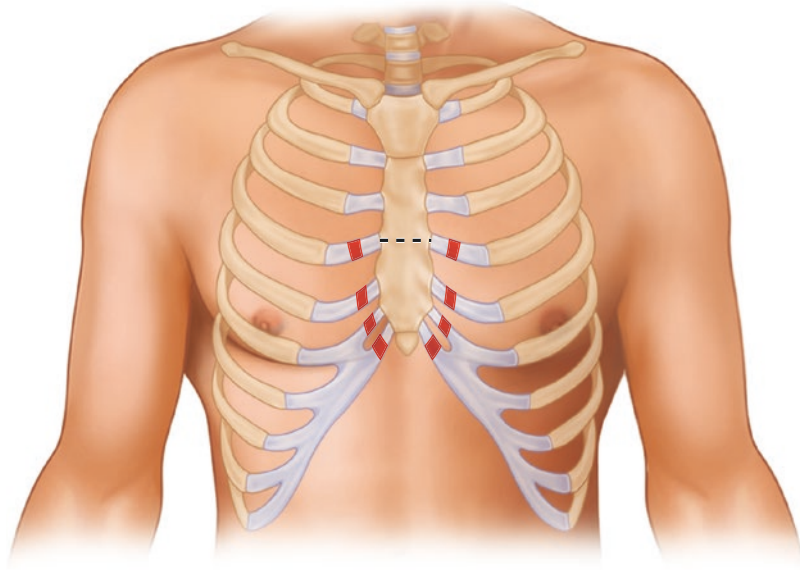


Fig. 44.10 Schematic view of pectus carinatum repair as performed by Welch

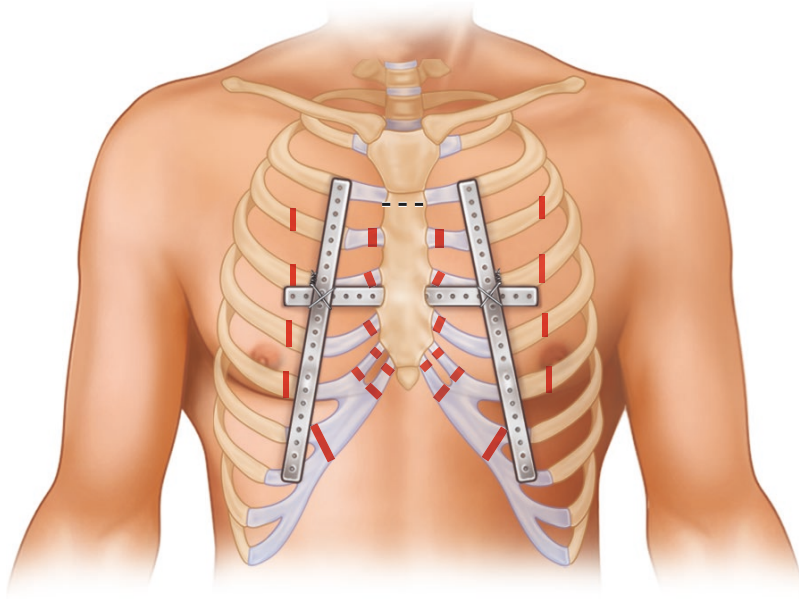


Fig. 44.11 Schematic view of pectus carinatum repair as performed by Willital

(Fig. 44.12) [18]. To achieve this, a bent pectus bar was placed subcutaneously on top of the sternum via lateral thoracic incisions. The bar was inserted through a polyvinyl chloride tube with the convexity facing posteriorly. The polyvinyl chloride tube was positioned pre-sternal through a port. Subperiosteal wires were employed to attach the small fixation plates to the ribs laterally, and the bent bar was secured to the small fixation plates with screws applying manual pressure to the anterior chest wall until the desired configuration is achieved. The compressive elongated bar was attached to the fixation plate with screws. Both symmetric and asymmetric protrusions were treated with this method. Patients, whose chest is not malleable, do not qualify for this technique. Following the report of Abramson, minimal access techniques for pectus carinatum

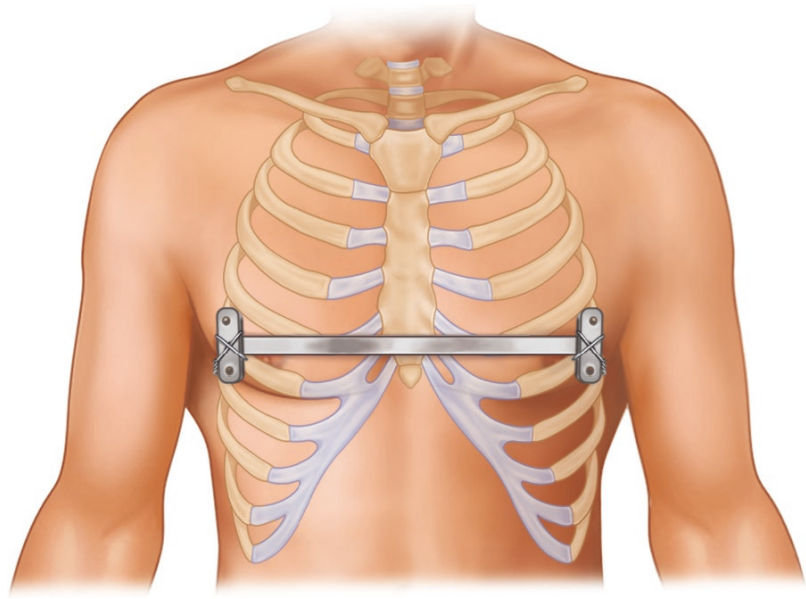


Fig. 44.12 Schematic view of pectus carinatum repair as performed by Abramson

with modifications were reported by Hock and Yüksel [19, 20]. Since these initial reports further surgical procedures with modifications of these techniques have been emerged in the literature.

Outcomes

As in pectus excavtum, there are numerous surgical procedures that have been designed with modifications for the repair of pectus carinatum, comparison between the procedures here is not possible. It is important that comparisons be performed with series that were performed in similar time frames and technology and techniques undergo marked changes after a decade. Future comparisons with open repairs in pectus carinatum will not be feasible since minimal access repairs will prevail if surgical correction is required, and non-operative treatments now predominate in the conservative management of these deformities.

Outcomes of Open Repairs

Since the results of open repairs have varying end points and sample size, the results of the 111 patients operated by the Willital- Hegemann technique are presented. Most of the patients at the time of surgical correction were children (n = 58) in the 11–15 years age group. The postoperative course of all patients was generally uneventful and the repairs were completed with complication rate of 4.5 % (pneumothorax 3, superficial wound infection 2). Blood transfusion was required only in one patient. Furthermore postoperative recovery in children <12 years was 2–4 days; which was significantly faster than 4–7 days in teenagers and adults. Major recurrence was found in 2 patients (1.8 %) and both the patients were re-operated. Minor recurrences we found in 3 patients (2.7 %). Satisfactory long-term results were obtained in the rest of the patients. Scars were revised in all the patients at the time of strut removal.

The follow-up period ranged from 4 weeks to 12 years, mean 7.2 years and the documentation of the preoperative deformity as well as the status of surgical repair included (a) measurement of the sagittal thorax diameter using a pelvimeter (b) electrocardiogram (c) pulmonary function tests in children >8 years of age (d) thoracic ultrasonography, (e) clinical photograph and videostereoraster graphs; which are routinely obtained to demonstrate the preoperative deformity and the result of surgical repair.

Outcomes of Minimal Access Repair

Since minimal access repair is still evolving, the results of the initial experiences have been presented in this chapter. In Abramson series, of the 40 patients treated, 20 had undergone bar removal at the time of his report with the following results: ten excellent, four good, four fair, and two poor. Average blood loss was 15 ml with a mean length of hospital stay during repair being 3.8 days and during removal 1.4 days. Complications in this series included pneumothorax (n = 1) which required a chest tube suction, skin adherence (n = 8), seroma (n = 6), wire breakage (n = 3), persistence of pain (n = 1) and infection (n = 1).

In the series reported by Hock, the operation time ranged from 75 to 110 min [19]. The chest tube that was placed during surgery was removed on the second postoperative day and the patients were discharged between the fifth and seventh postoperative day. There was one dislocation in which the bar was removed, and in three patients a dislocation of the end of the strut through intercostal space that necessitated re-fixation using wires.

In the series reported by Yüksel, excellent esthetic results with 94.4 % patients satisfied with the operation [20]. Two of the bars have been removed during the 18-month follow-up. Three fixating steel wire breakages requiring re-fixing of the stabilizers and two local skin adhesions over the bar were seen as postoperative complications.

Standardization of Results

The radical change in management of pectus carinatum from open repairs and minimal access repairs to non-surgical conservative treatment makes it difficult to compare results as the indications for each of these techniques varies on multiple factors. Comparison of results will have to consider the following factors prior to and after management before data interpretation based on evidence is possible:

1. Type of pectus deformity (symmetric/asymmetric)
2. Shape of the thorax (presence/absence of platytorax)
3. Presence of combined deformities
4. Age of the patient at the time of management
5. Imaging (Computed tomography/Videosteroraster/Caliper/Flexible ruler)
6. Evaluation of result by patient/parent
7. Evaluation of the result by surgeon
8. Psychological evaluation
9. Use of a standardized management (open/minimal access/non-surgical)
10. Complications during and immediately after the procedure
11. Late complications
12. Complications during bar removal (if a strut or bar was used)
13. Long-term follow up

Standardizing the spectrum of presentation of pectus carinatum and the age at which management is commenced along with the opted technique will be the three important denominators that will determine the basis for comparison of results.

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Nonoperative Management of Chest Wall Deformities

45

Richard G. Azizkhan and Aliza P. Cohen

Overview

Pectus carinatum (PC) comprises a spectrum of anterior chest wall deformities characterized by convex protrusion of the sternum and adjacent costal cartilages. Although the incidence of these deformities has not been recently validated, historical reports indicate that it ranges from one in 1500–1 in 1700 live births [1, 2]. PC occurs more frequently in boys (4:1) [3, 4]. It may occur as a solitary abnormality, but is often associated with scoliosis (15–30 %) and other musculoskeletal abnormalities [4, 5]. PC also occurs in association with trisomy 18 and a number of connective tissue disorders and syndromes, including homocystinuria, Marfan syndrome, Ehlers-Danlos syndrome, and Noonan syndrome [4–6].

The etiology of this anomaly is unclear; however, the increased incidence of a positive family history suggests a genetic linkage in some (25 %) patients [1, 4]. In a small number of patients who have undergone thoracic or mediastinal surgery, damage to cartilage growth contributes to the development of PC at an early age.

Three predominant types of PC have been identified. Type 1, also referred to as chondrogladiolar deformity, is the most commonly seen type; it involves the inferior costal cartilages and the gladiolus, resulting in a protrusion of the middle and lower sternum (Fig. 45.1); Type 2 (<1 %), also referred to as chondromanubrial deformity, involves the superior costal cartilages and the manubrium, resulting in manubrial and upper sternal protrusion (Fig. 45.2). Both type 1 and type 2 can occur either symmetrically or asymmetrically. Type 3, a mixed PC variant, appears as an asymmetric lateral protrusion of the costal cartilage with a contralateral depression (Fig. 45.3).

What manifests as a mild deformity during the neonatal period or early in childhood typically becomes more severe and thus more noticeable as the child grows, particularly during the pubertal growth spurt. As the deformity worsens, teenagers are often faced with significant cosmetic and psychosocial concerns that prompt their families to seek clinical advice.

Historically, surgeons have advocated reconstructive techniques employing resection of the deformed cartilages together with sternal osteotomy [7, 8]. Despite the relative success of these operative approaches, the risks associated with any major operation remain. In prepubertal children, there are also concerns that the most

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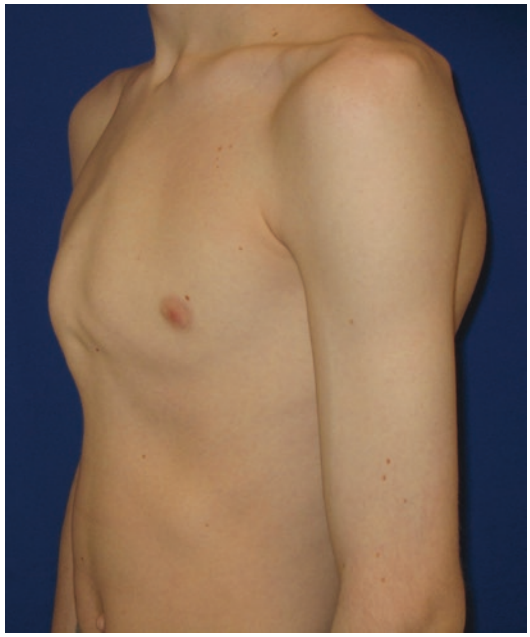

Fig. 45.1 Illustrates protrusion of the middle and lower sternum seen in patients with chondrogladiolar deformity (type 1 PC)

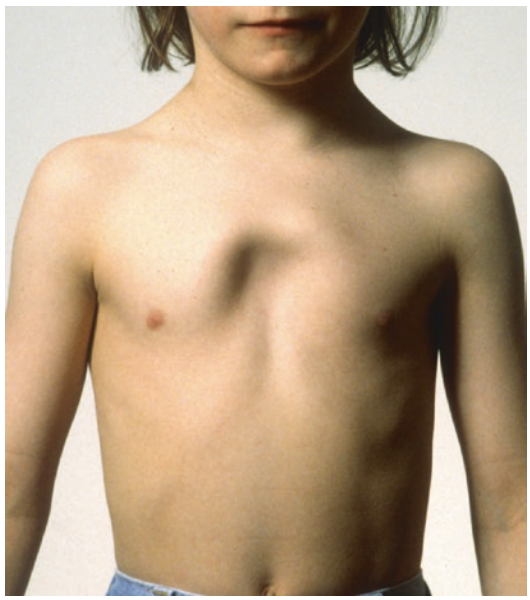

Fig. 45.2 Illustrates manubrial and upper sternal protrusion seen in patients with chondromanubrial deformity (type 2 PC)

common procedure used (Ravitch procedure) may interfere with chest wall growth because of damage to the costal cartilage growth plates. In an effort to eliminate these risks and avert the need for major surgery when possible, a number of authors over the past several decades have suggested nonoperative management approaches for selected patients. These noninvasive alternatives are based on the fact that the anterior chest wall remains compliant during puberty and is amenable to remodeling by applying external compression [5, 6, 9, 10].

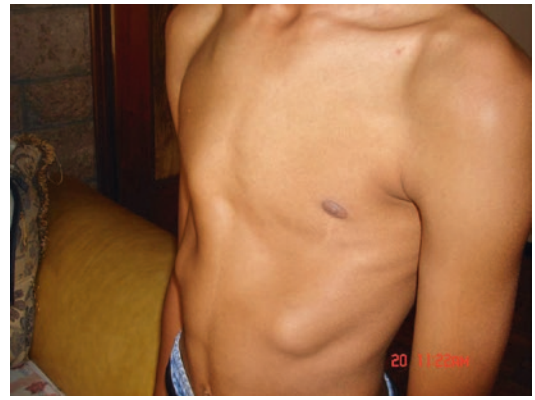

Fig. 45.3 Illustrates asymmetric lateral protrusion of the costal cartilage with a contralateral depression seen in patients with mixed PC variant (type 3 PC)

In recent years, nonoperative management has gained widespread popularity. In many pediatric centers, the use of an orthotic compression brace for managing patients with chondrogladiolar PC has become the first line of treatment, with corrective surgery reserved for patients who do not adhere to this management protocol or in whom bracing has been unsuccessful.

This chapter provides a contemporary overview of the nonoperative approach. We will briefly describe the methods and efficacy of bracing reported to date, with particular emphasis on the senior author's (RGA) personal experience over a 17-year period.

Evolution of Nonoperative Approaches

Several early case reports documenting success with casting and bracing techniques challenged the prevailing belief that nonoperative treatment was universally ineffective. In 1964, Jaubert de Beaujeu et al. [11] described serial casting of a

3-year-old boy with PC. Their approach involved applying corrective forces with a strap across the sternal prominence. Correction was subsequently maintained with a body cast. This approach reportedly achieved a good cosmetic result over 12 months, though lateral rib depressions were only partially corrected.

Several years later (1968), Bianchi [12] described limited success using a modified windowed body cast to attain correction in 20 patients. A wooden bar was placed in the cast over the area of sternal prominence. The bar was attached by screws to a pressure platform outside of the cast. By tightening the screws, a posteriorly directed vector of force could be transmitted to the sternum via the bar in the cast. The screws were turned daily for 30–40 days. Photographs and radiographs demonstrated correction, though the author noted that lateral sternocostal depressions did not disappear.

Drawing on accepted orthopaedic tenets of molding growing bone and cartilage with orthotic devices, Haje and Raymundo (1979) [13] published the first of several Spanish-language reports on the successful treatment of PC with an orthosis referred to as a dynamic chest compressor (DCC). Almost a decade later, Haje et al. (1988) [14] reported a classification for PC and an improvement in the DCC. In 1992, Haje and an American colleague, Richard Bowen [15], developed a custom-fitted compressive orthotic that placed greatest external forces on the point of the most prominent sternal protrusion of chondrogladiolar deformity. The orthotic was constructed with two U-shaped metal rods (one anterior and one posterior) placed around the patient's chest, with an anterior pressure pad and two posterior counter-pressure pads. The two metal rods were fixed to one another laterally by screws, which could be used to adjust the pressure applied by the pressure pad. Improvement in the deformity was rated on a subjective scale according to appearance. According to this scale, all patents with inferior and lateral types of PC who underwent treatment showed improvement (minimal 1-year followup). The authors concluded that when the orthotic was applied under medical supervision, it could likely avoid the need for operative treatment in most patients.

1 year later (1993), Mielke and Winter [2] published their experience managing a 14-year-old girl with a symmetrical elevation of the chest wall involving the lower third of the sternum and no associated anomalies. The child was placed in a windowed underarm body cast with both anterior and posterior molding. After the cast was removed (6 weeks later), a brace constructed with anterior and posterior shells was placed and worn. It was periodically tightened over 7 months, during which time it was worn fulltime. The brace was worn at night only for an additional 10 months. After 2 years, the midline prominence was corrected, leaving a slight right-sided sternocostal prominence. The outcome remained unchanged at 5-year followup, once again illustrating that nonoperative management of PC could be effective. The authors emphasized that patients must be motivated and have growth potential remaining.

By the year 2000, nonoperative management of PC had gained more widespread acceptance as an alternative to surgical reconstruction, though this approach was limited to only a few centers.

Egan et al. (2000) [9] developed a compressive orthosis similar to that of Haje and Bowen [15] and sought to better quantify treatment outcomes with objective radiographic criteria in five patients, reporting positive preliminary outcomes in those who were compliant. In 2006, both Banever and colleagues [10] in the United States and Kravarusic and colleagues [6] in Canada reported positive outcomes with lightweight, custom-fitted chest braces. During the same year, a nonoperative management study conducted by Azizkhan and colleagues [5] yielded positive outcomes in all compliant patients (N = 26), leading the authors to conclude that compression bracing was a safe and effective treatment for children with chondrogladiolar (type 1) PC. In the largest study to date, Martinez-Ferro et al. [16] in Argentina (2008) reported that 99 of 112 (88.4 %) patients treated with a specially designed dynamic compression system using a custom-made aluminum brace (Fig. 45.4) achieved good to excellent results. This device has an adjustable lateral tension component that allows for adjustment during the duration of treatment. A specially designed pressure monitoring device is applied over the

Fig. 45.4 Dynamic compression system designed by Martinez-Ferro et al. in Argentina

deformity until the desired thoracic configuration is obtained. The authors showed that the initial measurement in pounds per square inch could be used to predict outcomes and required duration of the therapy.

Lastly, in a relatively recent (2009) case report, Nehra et al. [17] provided imaging results that clearly demonstrated remarkable improvement after 2–3 months of bracing in 2 highly motivated teenage boys with moderate and severe disfigurement respectively. Their findings corroborated the previously cited outcomes, leading the authors to conclude that bracing is a potentially effective nonoperative option in compliant, well-motivated, skeletally immature children. Moreover, the authors noted that they had achieved excellent outcomes with this option in ten additional cases prior to completing their report.

Cincinnati Children's Hospital Protocol

Initial Management

All patients initially undergo a thorough physical examination. At this time, the clinician determines whether the patient should be observed for a variable period of time or if definitive treatment should be initiated. Patients with chondrogladiolar deformity who appear to be well-motivated and committed to complying with the requirements of bracing are considered appropriate candidates for this approach.

Orthotic Brace

Through the collaborative efforts of the clinician and a certified orthotist, a custom-fitted, easily concealable chest compression brace is designed. The brace is constructed with separate anterior and posterior compression plates that are anchored to aluminum struts. The struts of each compression plate are bound together by an adjustable strap on each side. The construction allows for the application of continuous anterior-posterior chest compression, pushing the sternum with the midback serving as a stable base (Fig. 45.5a, b).

Treatment Regimen

Patients are instructed to wear the brace over a body hugging undergarment such as a T-shirt for 16–18 h a day and to symmetrically tighten the straps as tolerated. Initially, the straps are tightened every 1–2 weeks. This is followed by adjustment tightening at less frequent intervals.

After the initial brace fitting, patients are seen at 6 weeks to 3 months. Subsequently, they are seen at 6–12-month intervals. Pressure on the anteroposterior projection of the chest is easily monitored; when pressure is sufficient, a red mark over the area of sternal protrusion persists for several hours after brace removal.

Patients and families are informed that the brace is to be worn until linear growth is complete, which is generally 2–2.5 years. Because prepubertal patients experience growth spurts, they may require a longer period of bracing.

Bracing Outcomes

Over a 17-year period, the senior author (RGA) has nonoperatively managed 200 children ranging from age 3 to age 18. Compliance with a prescribed bracing protocol has resulted in successful outcomes in 196 (98 %) of these patients. The four patients who failed to achieve successful outcomes did not comply with the full bracing protocol. All patients were monitored for 1–2

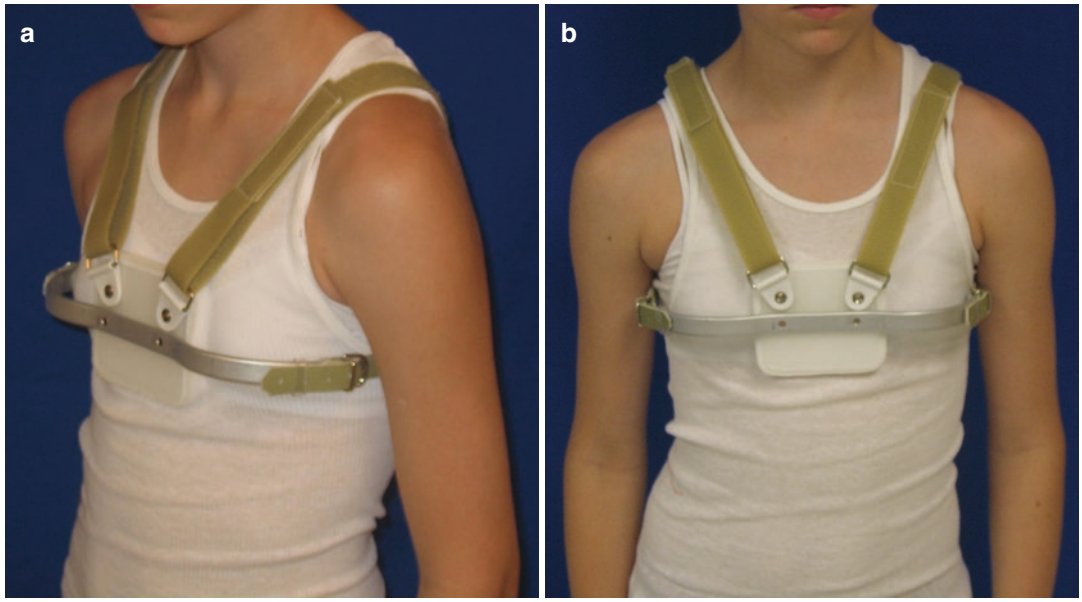

Fig. 45.5 (**a**, **b**) Orthotic compression brace worn by patients (oblique and front views)

years after completion of bracing and usually after linear growth was complete. Two prepubertal patients who underwent treatment redeveloped PC after undergoing additional growth spurts 1 year after bracing. These children required re-bracing, which was successful.

In summary, we believe that the positive outcomes discussed in this chapter, together with close to two decades of personal experience, indicate that bracing is indeed a safe and efficacious option for the management of highly motivated pediatric patients with chondrogladiolar PC. Furthermore, at a time when cost containment in healthcare is imperative, it should be noted that the total cost of surgical correction vs. bracing at our institution in 2012 was close to $30,000 vs. less than $1,000, respectively.

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Dynamic Compressor System for Pectus Carinatum Repair

46

Marcelo Martinez-Ferro and Carlos Fraire

Introduction

Pectus carinatum (PC) is a common pediatric condition characterized by an abnormal overgrowth of the costal cartilages, which results in protrusion of the sternum and adjacent costal cartilages. It is more frequent in males than in females (4:1 ratio) and may be classified into typical (90 %) or atypical (10 %) forms. Typical PC can be symmetric (Fig. 46.1a) or asymmetric (Fig. 46.1b) whereas atypical forms might be associated to Currarino-Silverman (Fig. 46.2), combined pectus carinatum/excavatum (Fig. 46.3), and Poland, Marfan or Von Recklinghausen syndromes, among other connective tissue disorders (Fig. 46.4). Even though its etiology is unknown, PC may be genetically linked considering its frequent occurrence in families [1].

Apart from the external appearance which most commonly concerns patients and families, the majority of children present with relatively mild symptoms; the most frequently reported are tenderness, bone pain or mild exercise intolerance. Even though psychosocial issues secondary to body image need to be promptly addressed in all cases, since pectus deformities and their associated effects as scoliosis, tend to become more severe during adolescent growth years and may worsen throughout adult life, the physiological concerns must take precedence without exception.

Despite the early work of Jaubert de Beaujeu and Bianchi and coworkers [2, 3] – the pioneers in non-operative treatments for PC – open surgery has been the treatment of choice over the last five decades [4–6]. Most of the existing surgical procedures consist of modifications of the Ravitch technique that employ resection of the deformed costal cartilages along with sternal osteotomy [3, 7]. Our experience before 2001 using a modified Ravitch technique is summarized in (Table 46.1) [8–10]. In these 94 surgical patients, the overall complication rate was 22 %. The most frequent complications observed, were wound infection in 12 patients (12.7 %), pleural effusion in 6 (6.3 %), pneumothorax in 5 (5.3 %), recurrence in 12 (12.7 %), and hypertrophic scarring in 16 (17.0 %). Even though patients were generally pleased with the improvement of their chest's shape, surgery could not address the usual problem of the flaring of ribs and a visible scar was always left. On top of that, we know now that surgery does not result in complete thorax remodeling in comparison to the non-operative treatments.

Recently, there have been many publications that propose less radical resection but still remain

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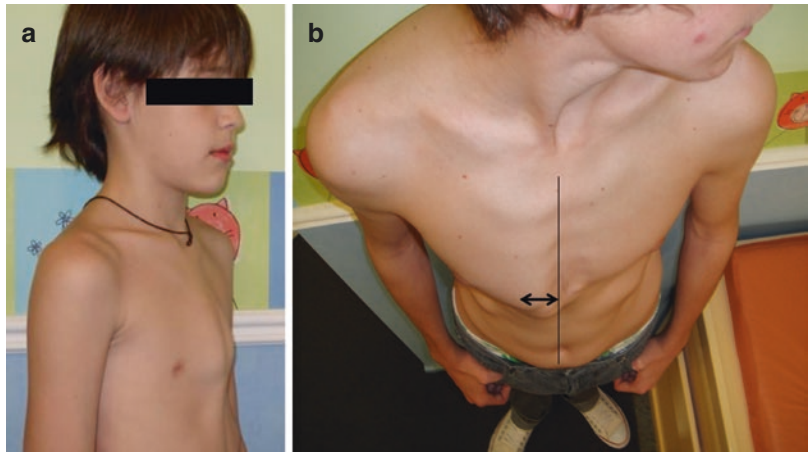


Fig. 46.1 Typical PC. (**a**) Symmetric. (**b**) Assymetric (offsite from the center)

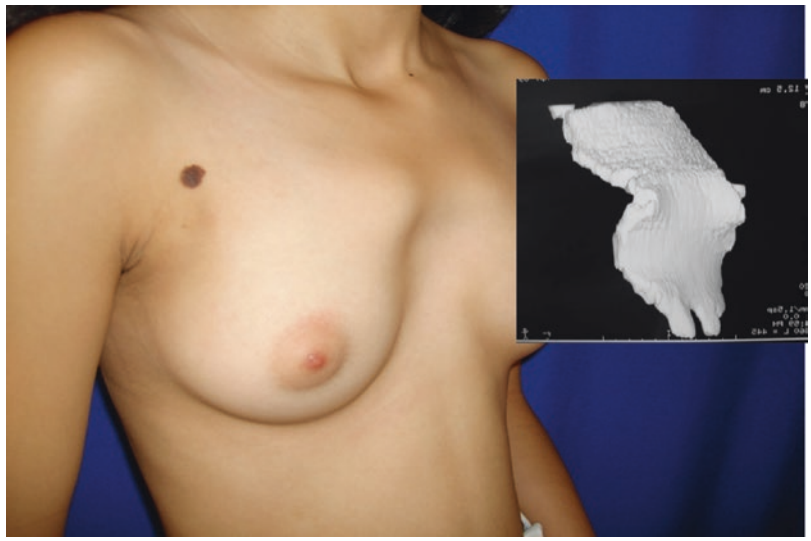


Fig. 46.2 Atypical PC associated to Currarino-Silverman syndrome

major surgical procedures [11–13]; consequently, because of the inherent risks of major operations, surgical treatment has been reserved for the most severe cases, thus leaving many of the patients untreated [14].

More lately, several authors have suggested a variety of alternative non-operative approaches for these undertreated patients, based on the fact that the anterior chest wall is still compliant during puberty and that it permits remodeling by applying external compression [15–21]. Drs. Haje DP, Haje SA and coworkers have shared their extensive experience in treating patients with PC using a Dynamic Chest Compressor [22–25].

The Nuss procedure for pectus excavatum introduced a paradigm shift by demonstrating that the thoracic wall is a very elastic and malleable structure in children [26]. Inspired by this concept, early in the year 1999, we began assessing chest wall compliance in patients with mild to moderate forms of PC by applying manual

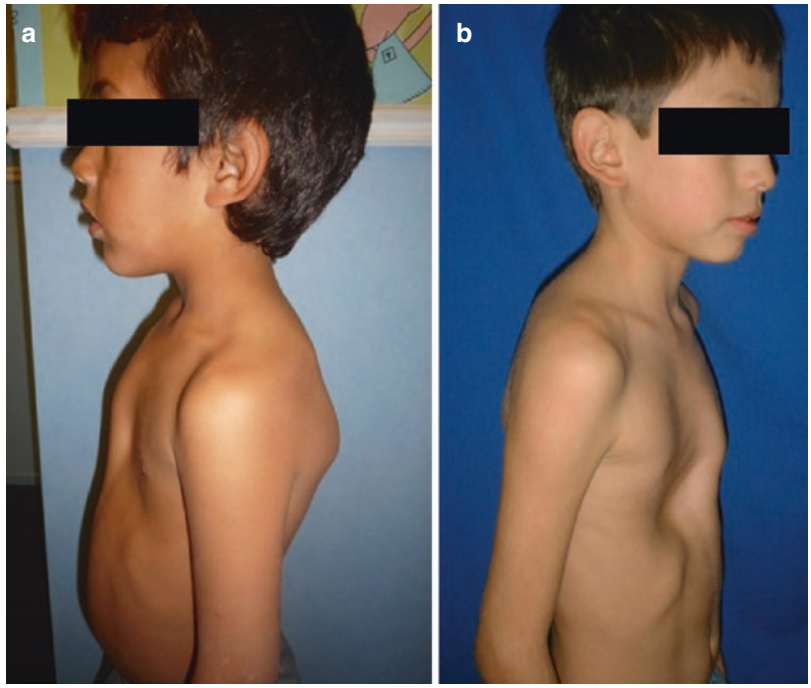


Fig. 46.3 Combined pectus carinatum/excavatum. (**a**) Carinatum to the right and excavatum to the left. (**b**) Excavatum with flare to the right and carinatum to the left

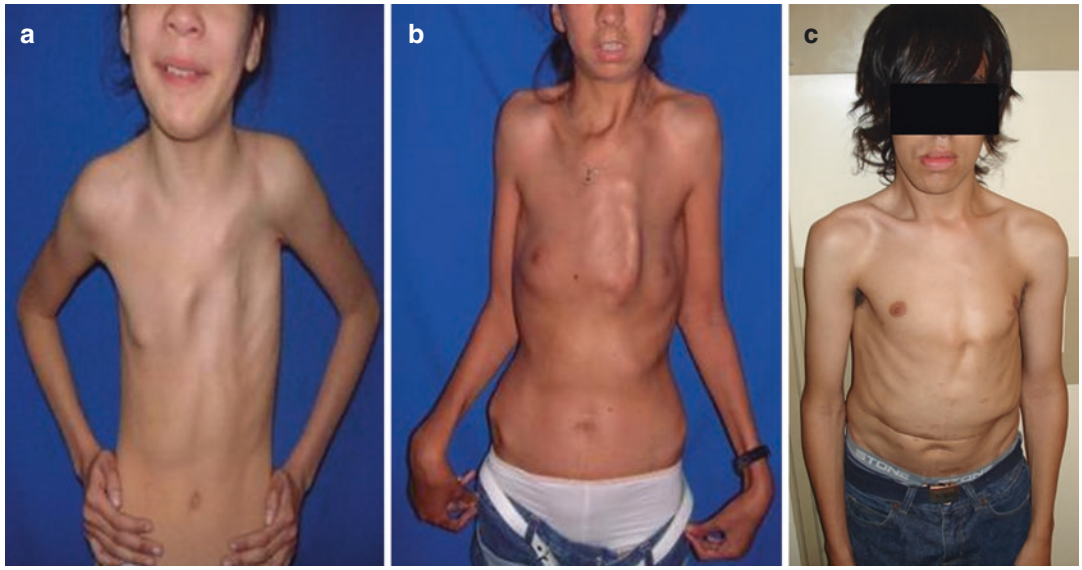


Fig. 46.4 Atypical PC associated to (**a**) Poland, (**b**) Marfan and (**c**) Von Recklinghausen syndromes

compression to the defect (Fig. 46.5). Since we observed it could be corrected without pain, a non-operative prospective study was designed and implemented at our chest wall deformities outpatient clinic with the advantage that, in comparison to the treatment for pectus excavatum, there was no need for an implant as the protrusion could be compressed externally. A Dynamic

Compressor System (DCS) was developed and utilized for this purpose.

Additionally, since by that time, there were no reports about the record and analysis of pressure measurements to compare series of patients, we decided to investigate on that particular topic.

Two quantifiable variables were defined to statistically compare objective data collected at every consultation:

- Pressure for Initial Correction (PIC), that is, the pressure applied to the patient at the time of his first consultation until the proper shape of the thorax is achieved. It is a measure of the patient's chest wall flexibility.
- Pressure of Treatment (POT), that is, the pressure required to treat the patient. It must be measured regularly at every consultation to adjust the DCS.
- By the beginning of 2001, the DCS design was finished and its protocol approved by our IRB.

Table 46.1 Surgical approach versus non-surgical approach for pectus carinatum repair

	Surgical (modified ravitch; *n* _ 94)	Non-surgical (DCS; *n* _ 388)
Operative time (hours)	3.5	0
Hospital stay (days)	5.2	0
Deaths	0	0
Overall complications	22 %	4.6 %
Excellent to good results	89 %	96.7 %
Fair to poor results	11 %	3.3 %

First Projects

At first, the authors developed different kinds of plastic and then metallic orthotic devices that actually did not work. They noticed that when the patient's thorax was compressed, it expanded laterally and concluded the reason for their failure was that thoracic lateral expansion occurring naturally during inspiration had not been taken into consideration.

With the aid of mechanical and electronic engineers, the authors designed an external DCS as exposed previously, but loaded with an electronic Pressure Measuring Device (PMD) to measure the PIC (Fig. 46.6) and POT. The DCS included a brace, which was an expandable and regulable belt which aside from being inflexible, it was of low-profile and light weight. In order to avoid referals, the brace was designed in such a way that it could be ordered, assembled and implemented at different, distant locations with ease (Figs. 46.7 and 46.8). The PMD was developed to convert the mechanical energy exerted to the patient (pressure) into electrical energy visible as numbers

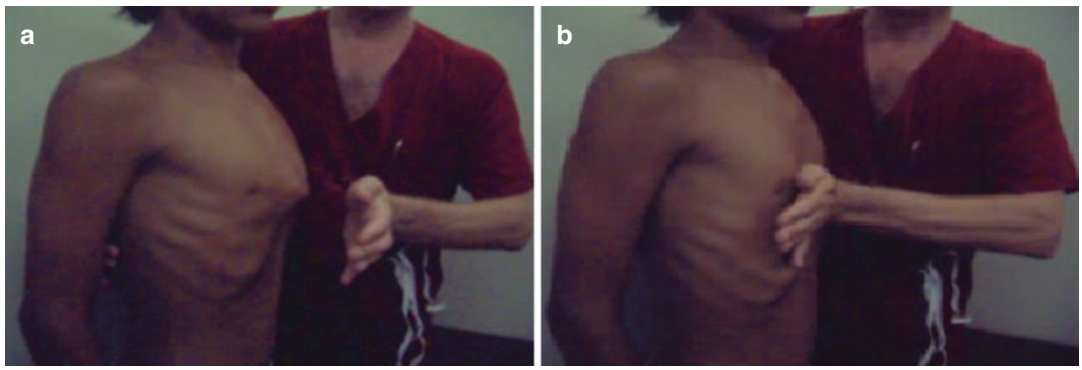


Fig. 46.5 (**a**) Patient with typical, symmetric PC in whom (**b**) thoracic elasticity is checked by applying manual compression to the defect. This is corrected without pain and led Dr. Fraire C and Dr. Martinez-Ferro M to the design of a Dynamic Compressor System to exert pressure on the protrusion just as in the picture

in a screen (measurement of pressure). The PMD was needed at every consultation due to the differences in chest wall flexibility among patients and because chest remodeling gradually reduces the pressure of correction needed by each patient throughout the treatment. The variables PIC and POT quantified changes in the pressure required to compress the chest wall deformity during the follow up and until proper defect correction. In addition they proved useful to predict duration of treatment and prognosis, and thereby to aid physicians assess the patient and his family.

After an intensive clinical research, it was concluded that the most appropriate unit of measurement was pounds per square inch (PSI). In

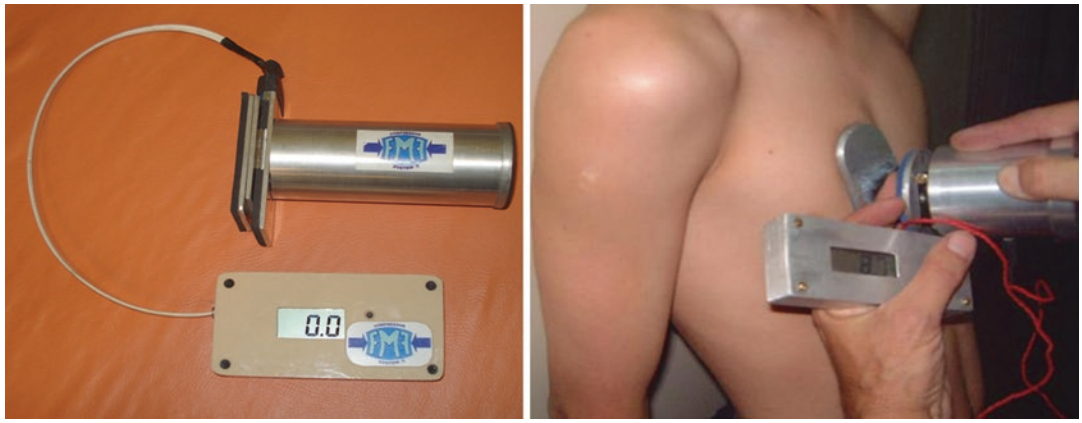


Fig. 46.6 *Left*: Early version of the electronic PMD used to measure the PIC in PSI units. *Right*: The same device compressing the defect of a patient with PC during inspiration

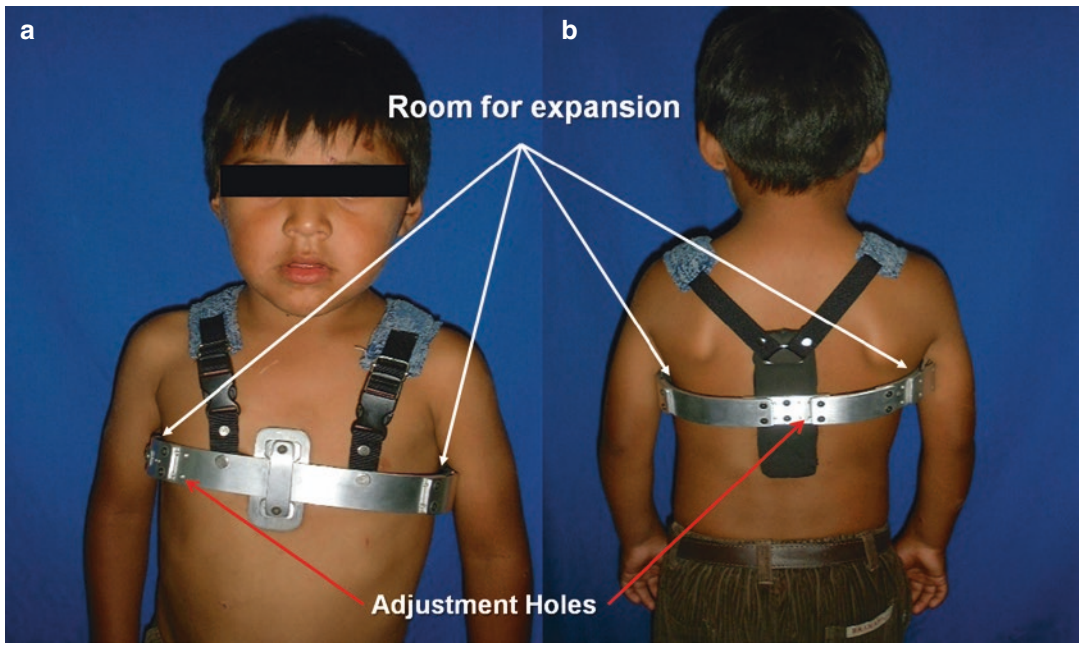


Fig. 46.7 Six-year old patient standing up in (**a**) ventral and (**b**) dorsal position, wearing the 1° prototype of the FMF® Dynamic compressor system. Note the brace has adjustment holes to attach the PMD and enough room to enable lateral expansion

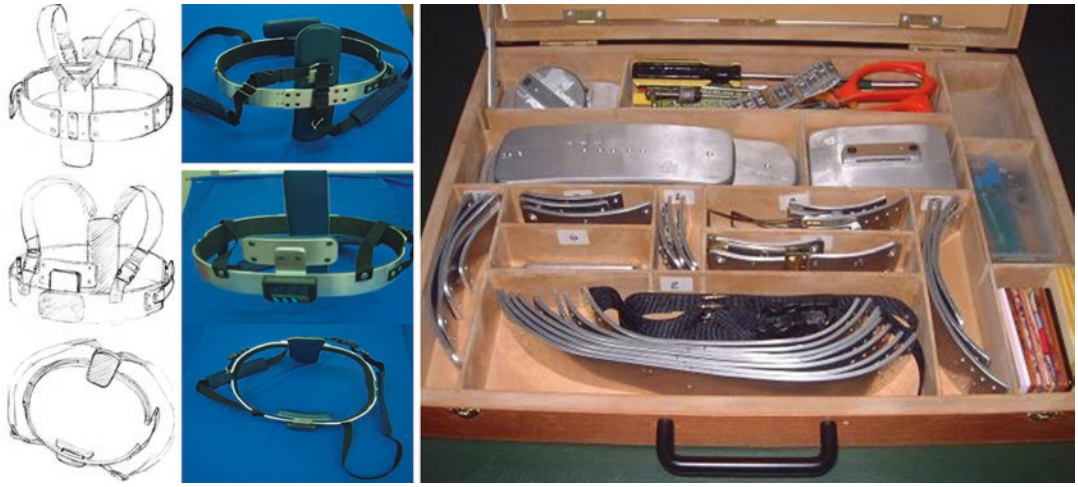


Fig. 46.8 Drawings and their corresponding pictures of the 1° prototype of the FMF® Dynamic compressor system

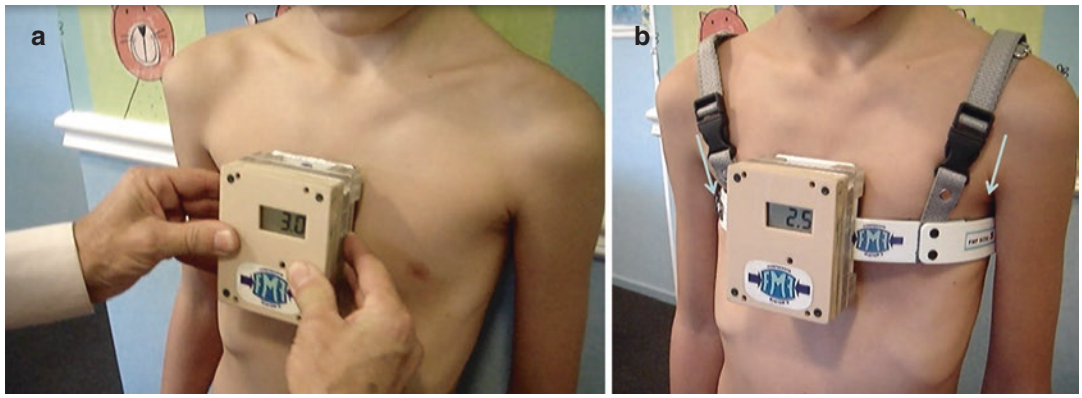


Fig. 46.9 The currently employed PMD is used in (**a**) to measure the PIC (3.0 PSI) and (**b**) the POT which is set to 2.5 PSI during maximal inspiration to avoid skin ulceration and eventual non-compliance with the treatment. Note that, at maximal inspiration, there is no lateral space left (*arrows*)

effect, this is the unit of pressure that is currently still employed (Fig 46.9).

Throughout the following years, several modifications were introduced to the DCS. For instance, the posterior compression pad was removed as it was not useful and caused skin lesions upon the spine and dorsal tissue (Fig. 46.10). Because skin erosion could be prevented, there was better tolerance from patients to finish the treatment. A docking station was designed for attaching the PMD to the brace for regulation of POT (Fig. 46.11). A locking system to avoid patient manipulation and a portable plate bender to model lightweight aluminum curved segments during consultation (Fig. 46.12) have also been developed.

We published our final product in 2008, called the FMF® Dynamic Compressor System (FMF stands for Fraire/Martinez-Ferro) [8] and standardized a series of measurements to collect data for scientific evaluation [9, 10].

The following chapter summarizes our last 12-year experience (2001–2013) using a nonsurgical approach by means of a DCS, the facts observed as we analyzed the pressure data recorded and the proposed algorithm of treatment for pediatric patients with PC deformity.

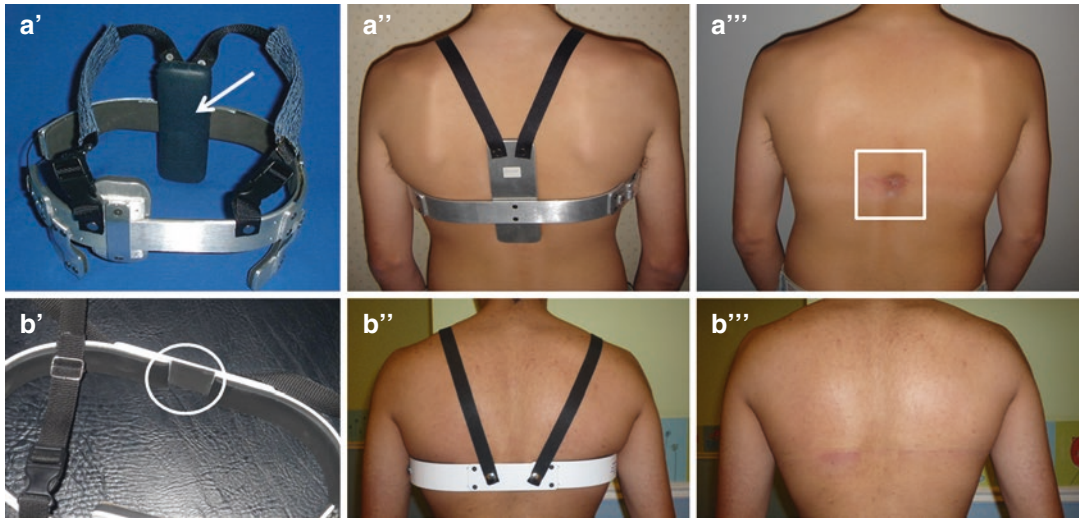


Fig. 46.10 (**a**) Patient wearing the 1 prototype of the FMF® Dynamic compressor system with a posterior compression pad (*arrow*). (**b**) Patient wearing a newer version of the FMF® Dynamic compressor system in which the compressor pad has been removed (*circle*) to avoid skin erosion as observed in (*A''*)

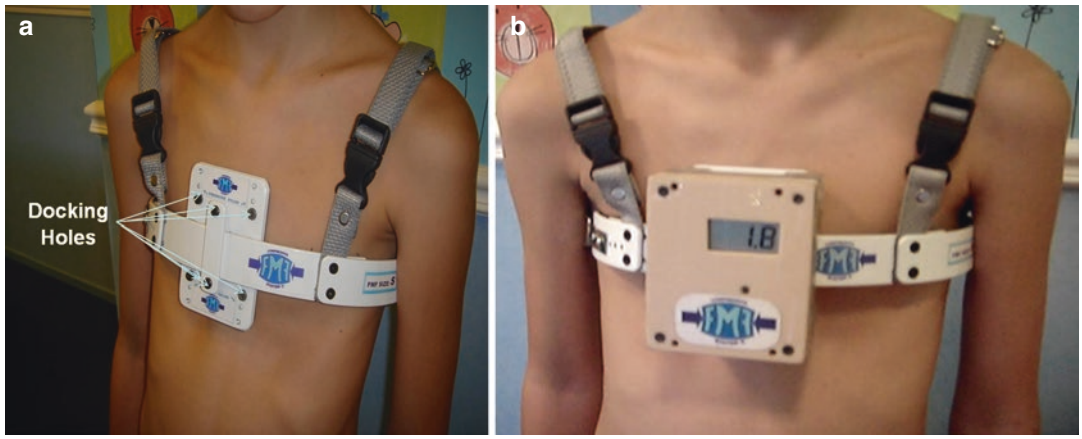


Fig. 46.11 (**a**) FMF® Dynamic compressor system in place for the attachment of the PMD for pressure determination. Take notice of the docking holes in the brace's compression plate. (**b**) The docking station is fixed to the brace and measures the pressure exerted to the defect. In these patient, POT = 1.8 PSI. On the upper left, a picture of the docking station with a removable and interchangeable compression pad

Materials

Dynamic Compressor System

The FMF® Dynamic Compressor System is not just a brace, it is a system. This system is comprised of various elements:

1. A custom-fitted, low-profile aluminum brace that is adjustable to any thoracic shape or size;
2. Various kinds of shoulder straps each time better designed for the patients comfort.
3. Different sizes and shapes of cushioned compression plates adaptable to distinctive sternal protrusions, independently of their locations, sizes and shapes;

Fig. 46.12 Portable plate bender to model lightweight aluminum curved segments during consultation

4. Different compression pads that can be adhered to the compression plate to cushion the defect, prevent skin lesions and avoid non-compliance;
5. A Pressure Measuring Device, which consists of a docking station attached to a removable compression plate;
6. Standardized measuring instruments (chest measuring ruler and metric tape) to record the data needed to assemble each brace;
7. A portable plate bender to bend the lightweight aluminum curved segments according to the patient's continuous re-shaping thoracic anatomy;
8. Specific tools as screwdrivers and screws.

The custom made brace is assembled for each patient using multiple lightweight aluminum curved segments to obtain a rigid belt that surrounds the thoracic wall at the level of the defect. One unique cushioned compression plate is attached to its anterior segment and placed at the level of the protrusion (Fig. 46.13). The locking system is situated on the side where the prominence is most evident to enhance compression.

The FMF® Dynamic Compressor System corrects the defect by pushing the sternum backwards: the continuous anterior–posterior compression progressively re-shapes the chest, which is widened as the defect is pushed in, into a normal thoracic shape and position. Thoracic widening additionally occurs because of patient's growth. The multiple lightweight aluminum segments can be adjusted, bent and eventually replaced at every consultation to permit proper lateral thoracic expansion (Fig. 46.9b). An excessively tight brace causes non-compliance and treatment interruption.

Pressure Measurement and Regulation

Initial FMF® Dynamic Compressor System Fitting Phase

This is the screening period since patients first visit the chest wall deformities outpatient clinic until they begin using the brace.

PIC (in PSI) is obtained at the time of the first consultation with the PMD. Measurement of PIC is available for the last 375 patients.

The patient is asked to stand up against a wall facing the physician. The PMD is applied directly against the thoracic wall at the point where the chest wall deformity is most pronounced. The thorax is gradually compressed until a "normal shape" is observed. The process is repeated three times, and the average pressure required to compress the protrusion and re-shape the thorax is considered as the PIC (Fig. 46.9a). Because of the variation of pressure during breathing, the latest versions of the PMDs calculate the average pressure of multiple measurements taken in 5 s automatically, by pressing a single button (the average button).

Other standardized measurements are always taken at the time of first consultation to order and assemble the FMF® Dynamic Compressor System. Specific instruments were designed for

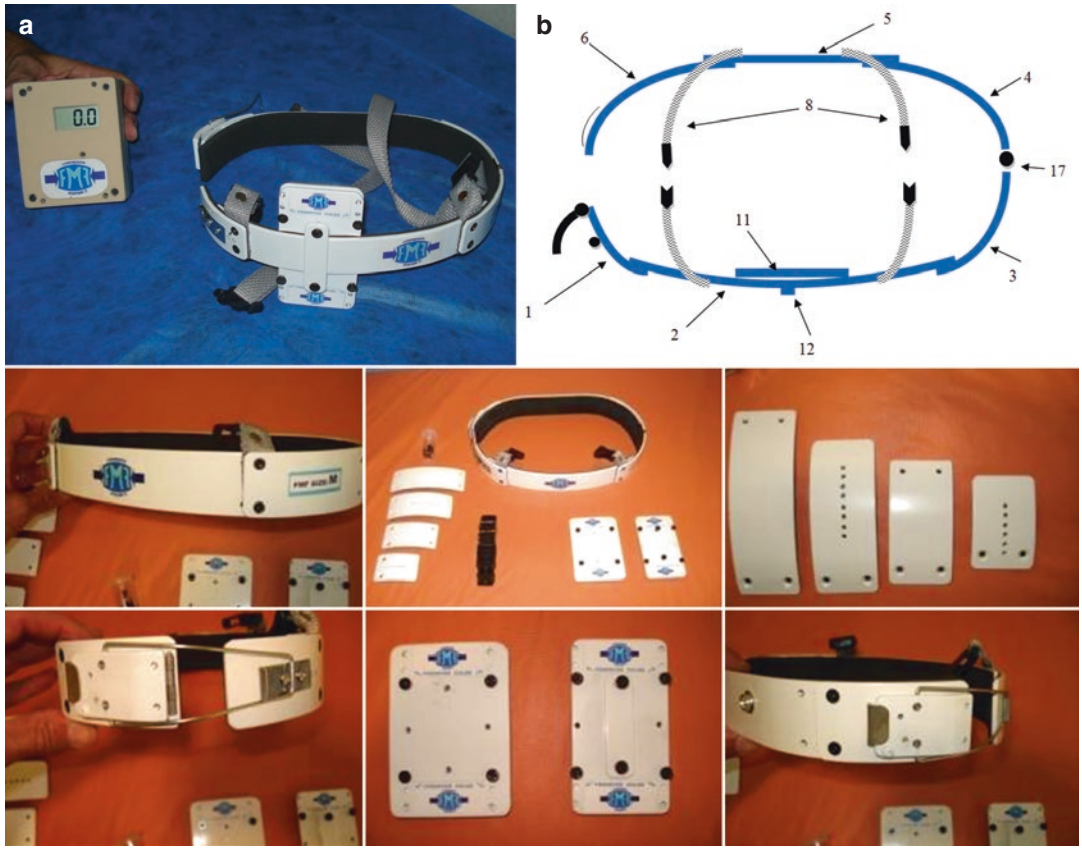


Fig. 46.13 Later version of FMF® Dynamic compressor system. (**a**) Brace and PMD. (**b**) Schematic drawing of the system constituted by 17 pieces and of the corresponding pictures denoting the FMF® Dynamic compressor system's complexity and the multiple combinations that can be employed to adjust the brace to different thoracic shapes and sizes

this purpose, including a chest measuring ruler to quantify the antero-posterior length, before and after manual compression of the protrusion (carinatum height), the transverse length and, in asymmetric patients, the off-site distance from the center. A metric tape is used to measure the thoracic perimeter at the level where the defect is most prominent (Figs. 46.14 and 46.15). All these data (in cm) are entered into the FMF® Dynamic Compressor System Order Form (Fig. 46.16).

Upon consent to follow the treatment, a custom-fitted brace with a compression pad adhered to a compression plate is assembled for each patient.

Correction Phase

This is the period between initial FMF® Dynamic Compressor System fitting and the reversion of the defect (including complete thoracic remodeling).

POT (in PSI) is measured when the patient wears the FMF® Dynamic Compressor System for the first time, and every time he comes to the chest wall deformities outpatient clinic for adjustment. To measure POT, the PMD is attached to the docking station placed in the anterior segment of the brace and over the defect (Fig. 46.11). POT is available for the last 251 patients. It is set at 2.5 PSI or less, independently of the PIC measurement, and during maximal inspiration to avoid skin ulceration.

Patients are checked monthly during the first 3–5 months, and less frequently then, until complete correction is achieved. Regulation of POT enables modifying the pressure of correction to the desired level. It prevents setting it too high, as

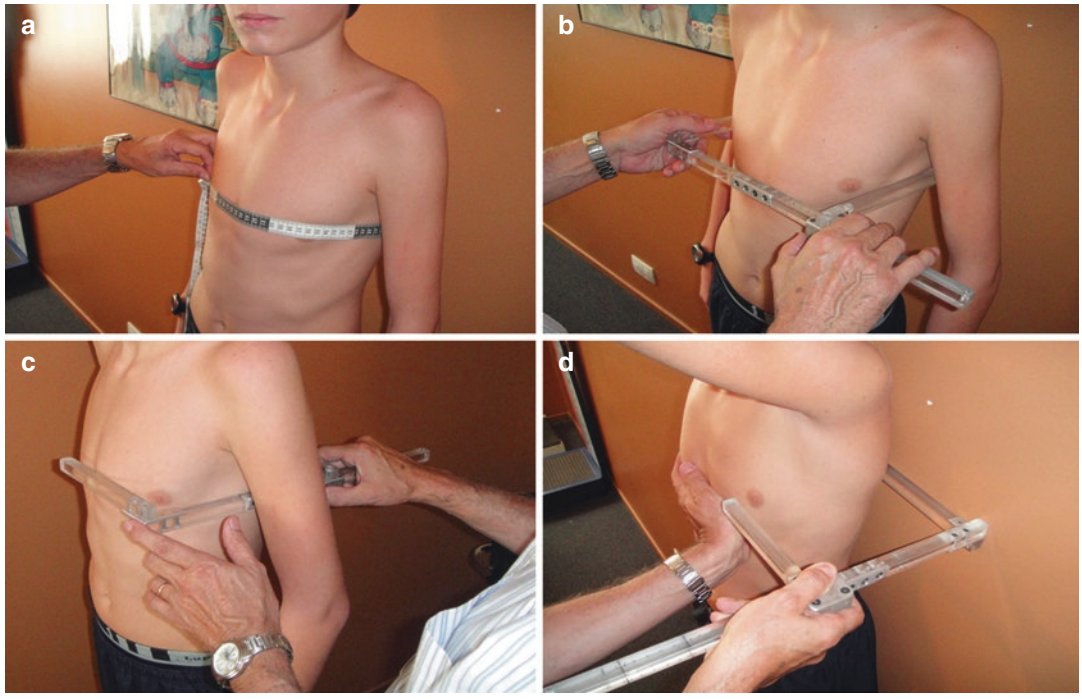


Fig. 46.14 Standard measurements taken to assemble the brace before initial brace fitting. (**a**) The patient's perimeter is obtained with the metric tape where the protrusion is more evident. A chest measuring ruler is used to quantify the (**b**) lateral and antero-posterior measurements (**c**) before and (**d**) after manual compression of the defect. The difference between the antero-lateral measurements in (**c**, **d**) permit calculating the height of the protrusion

Fig. 46.15 Briefcase with the instruments and materials required to obtain the standardized measurements to order the brace. Ref: (*1*) chest measuring ruler, (*2*) metric tape, (*3*) PMD. It includes a screwdriver and screws, too

this results in pressure necrosis and/or noncompliance (Fig 46.5).

The correction phase ends when the surgeon and patient (or parents) agree that the deformity has been fully repaired.

Patients

Patients referred to the chest wall deformities outpatient clinic were evaluated and those who met the inclusion criteria (typical PC, PIC < 9 PSI, consent to follow the treatment) were asked to join an institutional approved prospective study.

Patients with PICs < 7.5 PSI were instructed to wear the FMF® Dynamic Compressor System every day, overnight and as much as possible during the day, depending on their activities; they

FMF DYNAMIC COMPRESSOR SYSTEM® ORDER

Date Ordered: ____/____/____

Patient's Name: _____ Patient's Age: _____

Address: _____ City: _____

State: _____ Country: _____ Phone: _____

Patient Gender: ☐ Male ☐ Female

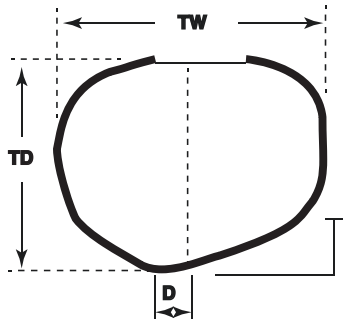
PIC: Pressure of Initial Correction ☐ PSI

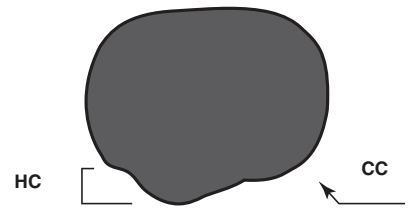
CC: Chest Circumference: ☐ cm. HC: Height of Carintum: ☐ cm.

Breast Development: ☐ Non-Existent ☐ Minimal ☐ Considerable

Position of Deformity: ☐ Left ☐ Central ☐ Right

Pad Length: ☐ Standard (10x7) ☐ Special: _____ ☐ Small ☐ Carved out





TW: ☐ cm: TD: ☐ cm: D: ☐ cm:

Notes: _____

Referring physician: _____ Signature of Physician: _____

Fig. 46.16 FMF dynamic compressor system order form

were only allowed to remove it during sports and while having a shower. On the other hand, adolescents and young adults with PICs between 7.5 and 9 PSI, commenced wearing the FMF® Dynamic Compressor System 1 or 2 h per day, in order to enhance tolerance and treatment compliance. The hours of usage were gradually increased to as much as possible during the day and overnight.

Setting of POT during the correction phase was always 2.5 PSI, as already mentioned, except in patients with sensitive skin, age lower than 4 years old, stiff chests or reporting pain. In these cases, POT was set to pressures lower than 2.5 PSI (range 2.4–1 PSI).

Measurement of the time of usage was difficult to calculate objectively because it depended on what the patient and family transmitted to the physician (which might not always be accurate). Hence, for the moment, the author's results were based on a very subjective scale. A double-blinded patient-physician or family-physician scale from 1 to 10 was designed and applied. At the end of the treatment, at the time of the last examination, patients and/or parents (depending on age) were asked to judge the final outcome from 1 to 10. Each treating physician on the team also submitted an undisclosed judgment. The lowest two numbers were used for the final result. Historical pictures of each patient were always available at any time for consultation at the physician's desk.

During the correction phase the patient was indicated a series of physical therapy daily exercises and had to attend regular reviews at the chest wall deformities outpatient clinic to constantly monitor the FMF® Dynamic Compressor System in order to ensure the correct correctional forces were being applied and to reinforce brace compliance and physical activity at home.

Weaning Phase

Once the defect was reverted, the FMF® Dynamic Compressor System was gradually withdrawn to avoid eventual recurrences. During this weaning phase, patients were allowed to wear it during the day or overnight (they generally preferred overnight), every day for the first month, every 2 days for the second month and every 3–4 days until in the third month. The weaning period, that is the time between the correction treatment termination and FMF® Dynamic Compressor System withdrawal, was not contemplated in the calculation of the duration of treatment. The FMF® Dynamic Compressor System was basically used as a "retainer". POT remained invariable in this post-correcting period.

Results

Between April 2001 and February 2013, 420 patients were prescribed the staged, non-operative treatment, with the FMF® Dynamic Compressor System and counseled to follow the protocol.

Three hundred and thirty-six were males (80 %) and the mean age was 12.5 ± 1.7 years (range: 1–32 years). 32 patients (7.6 %) abandoned treatment and could not be evaluated for final results. Of these, 2 declared pain and 4 reported skin intolerance as the cause of noncompliance; the other 23 claimed social discomfort, and 3 patients were lost to follow-up. Of the remaining 388 patients, 348 completed the treatment (89.7 %), and 40 (10.3 %) are still actively using the FMF® Dynamic Compressor System. The mean time of use per patient was 16 ± 3 h/day (range 7–21 h) for a mean period of 10 ± 5 months (range 3–24 months).

Applying the previously described scale, 375 (96.7 %) achieved a 7- to 10-point correction (excellent, very good, and good results) and 13 (3.3 %) patients achieved only 1- to 6 -point corrections (poor and bad results). The mean PIC value was 4.9 ± 0.6 PSI (range 1–9 PSI).

The following complications were observed in 18 of the 388 patients (4.6 %): back pain ($n = 8$), hematoma ($n = 1$), and skin ulceration ($n = 9$). No other complications were seen or reported. Even though they caused a delay in completion of treatment, they were not the cause of treatment termination. Skin ulceration was mild in all cases and was treated by withdrawal of the FMF® Dynamic Compressor System and topical skin lotions until the skin healed completely. The

other complications were treated with temporary loosening of the FMF® Dynamic Compressor System to lower the POT.

Mean follow-up time was 8 years (11.8 years to 1 month). During the follow-up, 35 patients (9 %) presented with a partial recurrence. The latter was mostly observed during periods of rapid growth and typically 7.3 ± 2.1 months after treatment discontinuance. All partial recurrences were mild, and were successfully treated with the FMF® Dynamic Compressor System, by modifying its shape and size to suit the patient's larger and widened thorax. All patients responded well and were promptly cured.

Analysis of Results

The collected pressure data denoted several interesting facts.

PIC could be correlated with age, final cosmetic results, and treatment duration.

Patients were divided into four groups: less than 2.5 PSI (Group I), from 2.5 to 5 PSI (Group II), from 5 to 7.5 PSI (Group III) and from 7.5 to 9 PSI (Group IV) (Table 46.2).

Younger patients had lower PICs than older patients. In other words the lower the age, the greater the thoracic flexibility. Even though better cosmesis was associated with lower PICs, what attracted the authors attention most, was that the duration of treatment could be predicted at the time of the first consultation, as lower PICs were associated to lower durations of treatment. That is, patients which more elastic and malleable thoraces cured faster.

Table 46.2 Correlation between the four groups versus final results and treatment duration

	Group I (<2.5 PSI)	Group II (2.5–5 PSI)	Group III (5–7.5 PSI)	Group IV (7.5–9 PSI)
Patients (n = 388)	144	189	45	10
Age (years)	7 (1–16)	12 (6–17)	15 (10–18)	21 (17–32)
Final Results (1–10)	9.5 (7–10)	8 (5–10)	7.5 (2–9)	6.5 (4–8)
Duration of Treatment (months)	3 (1–8)	5 (3–17)	11 (7–20)	17 (15–24)

The pressure data revealed that patients with PICs lower than 2.5 PSI were cured in approximately 3 months (Fig. 46.17) whereas those belonging to Group II, got corrected in 3–4 months, and cured in 6 months (Fig. 46.18). Group III patients were generally cured in 1 year (Fig. 46.19). Those in Group IV needed between 15 and 24 months to revert their PC.

Correlation between PIC and the duration of treatment resulted very useful in helping patients understand what was going to happen to them throughout the treatment (Fig. 46.20).

Less than 2 % of the patients (in particular Group I patients) showed a tendency to overcorrection to pectus excavatum. The treatment was immediately stopped until spontaneous correction of the defect took place. That is the reason why patients require monitoring with a monthly frequency for the first months.

Radiologic (Fig. 46.21) and computed tomographic scan (Fig. 46.22) changes were found, too. These changes consisted in flattening of the sternum and complete thoracic re-shaping from an abnormal to a normal chest shape. These studies are not routinely indicated, unless the patient presents with an uncommon PC which demands further investigation.

Data review from past experience pointed out that patients with PICs higher than 9 PSI should not be treated non-operatively, because they show intolerance and have a greater incidence of failure. POTs higher than 2.5 PSI should be avoided because they produce skin erosion and lesions, hence POT must always be equal or smaller than 2.5 PSI throughout the entire treatment. In general the higher the PIC, the lower the POT employed, that is, patients with stiff PCs will commence with a POT of 1 whereas those with a PIC of 3 will be treated with a POT of 2.5. Physicians are encouraged to refer not only adolescents but also small children. The earlier patients are treated, better results are obtained and the faster they are cured. Even though younger patients might

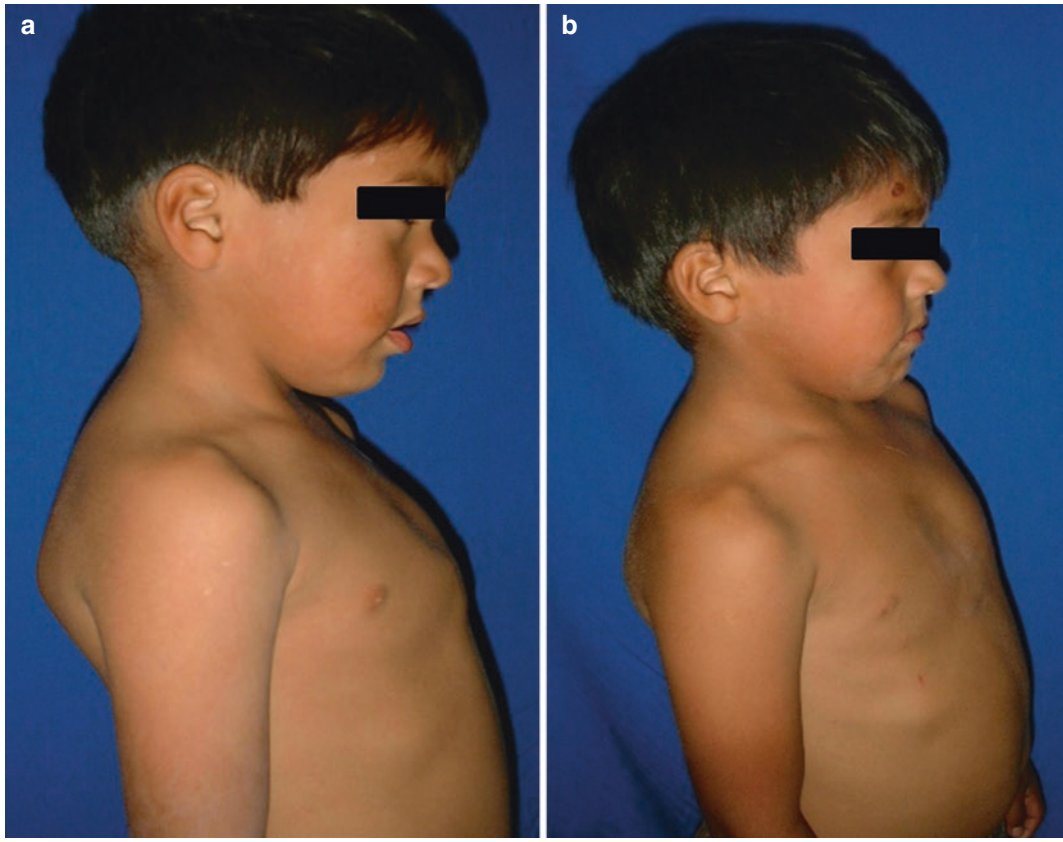


Fig. 46.17 Group I patient (PIC < 2.5 PSI): (**a**) At commencement of treatment. (**b**) After 3 months

present with partial recurrences related to growth in the future years, these are mild and easily corrected by using the FMF® Dynamic Compressor System for a short period of time. However older patients with stiffer PCs will require longer periods of time for final rectification of the defect.

Continuous Improvement Process

New projects are currently being developed to improve the FMF® Dynamic Compressor System and to reduce data recollection bias. Authors are currently working on the development of a time sensor, activated with body temperature, in order to measure the real "using time". An FMF® software is currently being developed to process the measurable and applicable data, with implications for prognosis and treatment of PC.

Proposed Algorithm of Treatment

An algorithm of treatment is proposed in Fig. 46.23.

For patients with PICs lower than 9 PSI, treatment with the FMF® Dynamic Compressor System is prescribed. Upon non-operative treatment tolerance, surgery is commonly avoided. However, upon treatment failure or

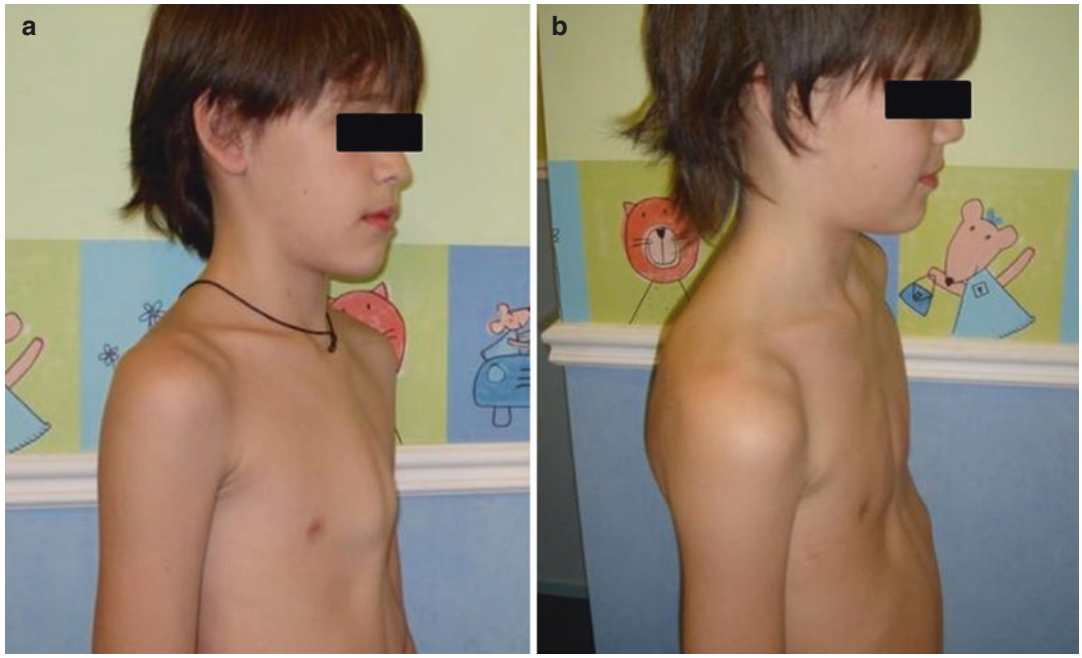


Fig. 46.18 Group II patient (2.5–5 PSI): (**a**) At commencement of treatment. (**b**) After 6 months

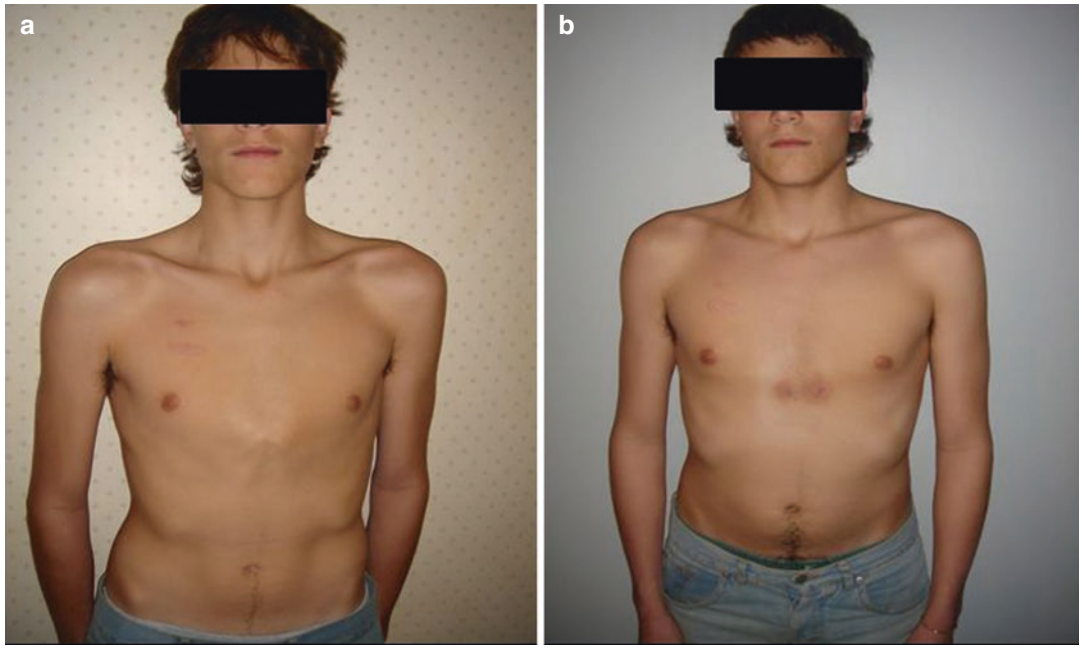


Fig. 46.19 Group III patient (PIC > 5 PSI): (**a**) At commencement of treatment. (**b**) After 12 months

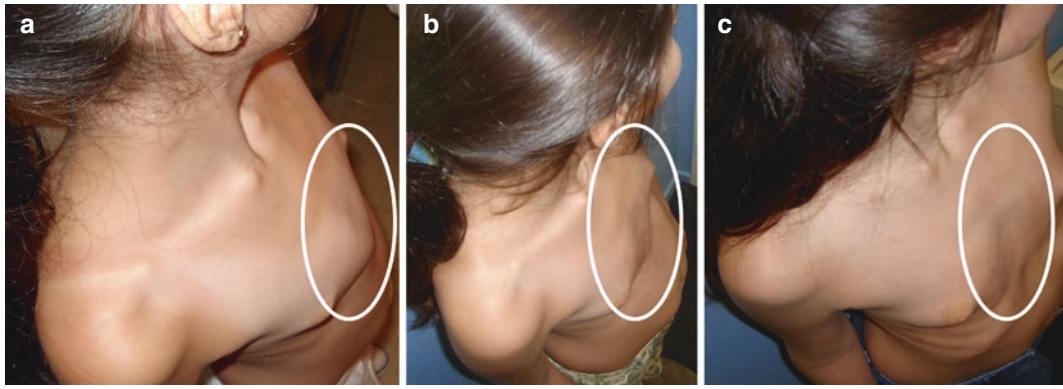


Fig. 46.20 Group I patient (PIC < 2.5 PSI): (**a**) At commencement of treatment. (**b**) After 1 month. (**c**) After 3 months

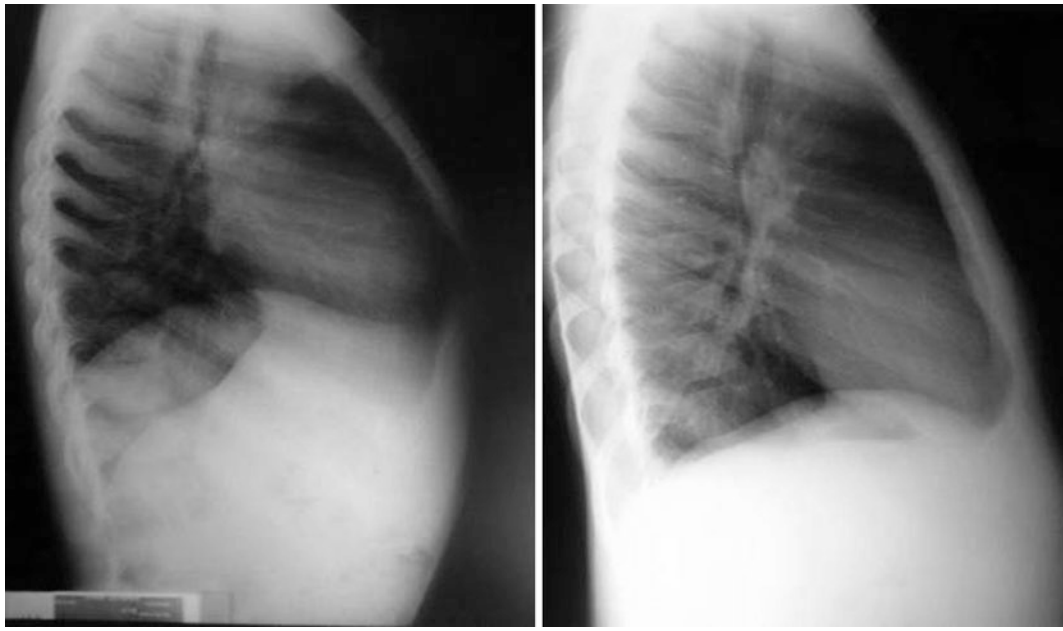


Fig. 46.21 Lateral chest X -ray of a group II patient (2.5–5 PSI) before (*Left*) and (*Right*) after treatment. Note the sternum protruding forwardly before treatment

for those patients who are unlikely to be compliant with bracing, an MIS is indicated. This approach may be extrathoracic [12, 13] or intrathoracic [27–30]. Our current technique is the Abramson technique (Reversed Nuss) [12]. No thoracoscopy is needed as the operation consists of the insertion of subcutaneous and submuscular Nuss/Biomed® modified bars and stabilizers.

If the patient requires a pressure of correction higher than 9 PSI, surgery is indicated. Generally, if the patient's chest is symmetrical, an MIS is the first choice. In most of the asymmetrical cases, even though an MIS can be tried, a classic thoracoplasty (using the modified Ravitch technique) is indicated. The same is the case for those patients who additionally have a very stiff chest or a failed previous MIS.

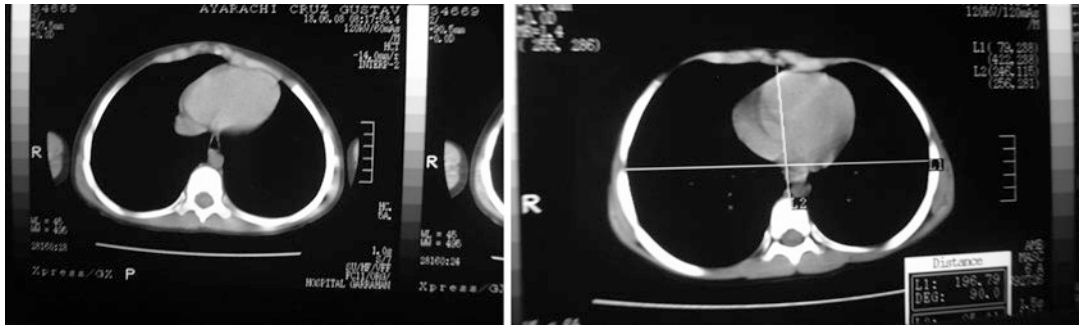


Fig. 46.22 TAC of a group II patient (2.5–5 PSI) before (*Left*) and (*Right*) after treatment. Notice complete thoracic re-shaping to a normal contour after treatment

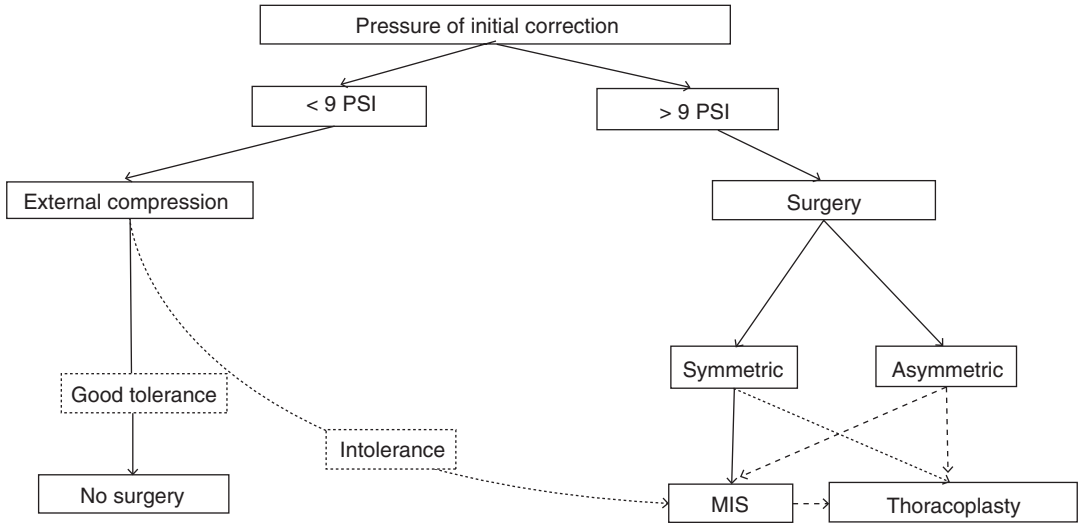


Fig. 46.23 Proposed algorithm for PC

Discussion

The Nuss procedure for pectus excavatum introduced a paradigm shift by demonstrating that the thoracic wall is an elastic and malleable structure, especially in children [26]. Following this idea, early in the year 1999, we started a prospective study with the objective of treating PC patients using the same concept but with the advantage that there was no need for an implant as the protrusion could be compressed externally. We agree with Lee RT and coworkers, who expressed in their recent paper, that a non-operative approach for PC essentially mirrors the effects of the internal bar in pectus excavatum patients, remodeling the growth pattern of the deformed chest wall cartilages [31].

By the year 1999, except for the pioneer papers of Haje and coworkers [22–24], no other authors supported a non-operative approach for the treatment of mild to moderate cases of PC. By the beginning of 2001, the DCS design was finished and its protocol approved by our IRB. Starting almost simultaneously with us, several other authors suggested a variety of non-operative approaches based on the same concept: that the anterior chest wall is still compliant during puberty and permits remodeling by applying external

compression [14–21]. Our results and conclusions are very similar to the above-mentioned papers.

Dr. Kelly RE Jr and his teamwork is currently treating patients amenable to bracing with the FMF® Dynamic Compressor System at a subsequent chest wall deformities outpatients clinic in Norfolk, USA with very good results [32]. They have observed that one of its advantages over other orthotics, is that the FMF® Dynamic Compressor System objectively measures the pressure of correction to guide treatment decisions. Because the position of the compression plate is early adjusted on the aluminum frame, flattening of the sternum in asymmetric cases is enhanced. Even for patients who fail bracing, treatment may assist in softening the chest wall in preparations for MIS or open surgery.

Comparing our own historical open surgery results with those of our new non-operative treatment, the benefits of the latter are superlative. To begin with, the FMF® Dynamic Compressor System results not only in permanent remodeling of the sternum and cartilaginous ribs but also in complete thoracic re-shaping. Secondly, it totally eliminates the risks of anesthesia and of major surgeries, decreases the complication rate, leaves no visible scar, avoids hospital admission, avoids activity restrictions associated to implant placing and dramatically reduces the cost of treatment. When considering all of these factors and reviewing our own and other authors experience, there should be little doubt that no patient should be selected as a candidate for surgery before trying a non-operative approach.

On the other hand, when analyzing our experience, at least two differences were established with the rest of the literature. In the first place, from the beginning we understood that, as anterior–posterior compression was applied to the thorax, a considerable lateral thoracic widening could be observed. To achieve a good and effective thoracic re-shaping, the FMF® Dynamic Compressor System was designed to permit lateral expansion. Secondly, we noticed that, depending on the age and other factors, the pressure needed for complete thoracic re-shaping showed a big range between patients, because of differences in chest wall elasticity.

At that time, we decided that measuring the pressure exerted to the protrusion would be crucial for understanding and treating patients with PC. When analyzing the recorded pressure data, we concluded that PIC increases with age and can be used to predict treatment duration, as patients with pressures lower than 2.5 PSI ended treatment almost four times faster than those requiring pressures higher than 5 PSI (3 versus 11 months). Although with less precision, PIC might predict the final cosmetic outcome; Group I patients showed better esthetic results than those of Groups II–IV. POT measurement enabled the additional observation that pressures greater than 2.5 PSI were lesser tolerated, mostly because of skin ulceration. Consequently, in the last 251 patients in whom the device was available, POT was set up to be equal or less than 2.5 PSI, and as a result, the FMF® Dynamic Compressor System resulted in satisfactory treatment tolerance and compliance.

Conclusion

We conclude that the DCS is an effective tool for the treatment of patients with PC and that PIC and POT seem promising complementary resources.

The FMF® Dynamic Compressor System (1) permits thoracic lateral expansion (re-shaping), (2) permits pressure measurement and control, (3) permits prediction of treatment duration and prognosis, (4) permits in-situ outpatient clinic adjustments, (5) avoids patient manipulation, (6) avoids spine and dorsal injury, thereby (7) providing increased tolerance. It can be (8) indicated, placed and controlled by any physician, who can additionally (9) collect objective data to enable him adjust the FMF® Dynamic Compressor System and perform further scientific evaluations.

The implementation of a staged treatment, consisting of three distinctive phases: (1) Initial FMF® Dynamic Compressor System Fitting Phase, (2) Correction Phase and (3) Weaning Phase, enables patients to be treated non-operatively with optimum reversion of their PC and complete thoracic remodeling. Upon failure, open or video-surgery is always a viable alternative.

Supplemental experience will aid to define which patients are better suited for non-operative dynamic compression with pressure measurement.

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Part VI

Pectus Carinatum: Surgical Techniques

Minimally Access Repair of Pectus Carinatum

47

Horacio Abramson and Leonardo Abramson

Abbreviations

P.C. Pectus Carinatum
PVC polyvinyl-chloride

Technical Highlights

The Minimal Access Repair of Pectus Carinatum MARPC does not require violation of the intra-thoracic cavity, and thus can avoid inflammatory, infectious or traumatic complications that involve the pleura, mediastinum, great vessels, heart and pericardium.

The technical highlights of this novel method are:

(a) Two 2-cm diameter horizontal incisions in both hemi-thoraxes along the middle axillar line in males and sub-mammary incisions in females.
(b) Dissection of the skin and muscle until the costal level.
(c) Sub-periosteal dissection of two costal arches on both sides.
(d) Sub-periosteal passage of little size polyvinyl-chloride (PVC) tubes around the ribs.
(e) Sub-periosteal passage throughout the PVC tubes of the steel wires around the ribs.
(f) Application of the fixation plates on the ribs by means of those wires.
(g) Sub-pectoral tunnel in both hemi-thoraxes.
(h) Insertion of a 1-cm. PVC tube in sub-pectoral location.
(i) Bending the bar in an appropriate design helped by template.
(j) Bar passage throughout the PVC tube.
(k) Linkage of the bar by means of screws with the fixation plates placing it along the fixation plate grooves.
(l) Progressive adjustment of the pre-sternal bar until achievement of the desired chest contour.
(m) Checking the integrity of the pleural cavity and closure of the muscle and skin layers.

Indications

1. Pectus carinatum in patients older than 10 years-old.

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2. Symmetric and caudal located protrusions (Chondrogladiolar type) give the best results

Preoperative Considerations

1. Routine lab and complete coagulation tests are required.
2. Lung function tests are required for assessment of the pulmonary function.
3. Cardiac evaluation using echocardiogram.
4. Front and lateral chest films.
5. Thoracic Computed Tomography is not mandatory unless Pectus Excavatum is also present.
6. Presence of Scoliosis must be assessed which is frequent in the asymmetric forms.
7. Psychological assessment of patient and her/his relatives.
8. Patients with Chondromanubrial type (Pouter Pigeon Breast or Currarino & Silverman Syndrome) are not suitable candidates for this procedure.
9. Detailed history of allergies to metal. Cutaneous tests for nickel and chrome if necessary.
10. Photographs of the chest wall documented in different angles.
11. In patients with limited flexibility of the chest wall, pre-surgical treatment begins with kinetic therapy which includes compressive massages, muscle and breathing exercises. Compressive orthotic treatment in the protruded region for a period between 30–60 days could also be considered.
12. Patients with a highly rigid chest wall or those that would present technical complexity due to extended deformity and cranial localization of the deformity are not suitable candidates for this procedure. These patients are best suited for open repairs.
13. Chondrotomies and osteotomies by Video Assisted Thoracic Surgery (VATS) both for unilateral or bilateral forms facilitate compression of the pectus bar.
14. Antibiotic therapy is initiated during surgery with Cefazolin.

Special Instruments

This technique requires special instruments:

1. Tools for costal dissection: periosteal elevators of several types.
2. Curved instruments to circumscribe the rib.
3. PVC tubes of small caliber to surround the rib and allow passage of the pericostal wires.
4. Pectus bars of multiple sizes.
5. Chest wall templates to configure the appropriate design of the bars.
6. Fixation plates with threaded holes and non-threaded holes.
7. Screws of several sizes and shapes to link the bar to the plate.
8. Wires of different calibers.
9. Long scissors.
10. Metal hook.
11. Benders of different types: manual benders, table benders and metal-bar benders to allow adequate contour of the bar and its fine tuning once implanted, without having to take it out to correct details in the bar design.

Surgical Technique

Small horizontal incisions of 2-cm are made in both axillar regions on the middle axillary line. The incisions are located over a horizontal line that goes through the highest level of protrusion in the symmetric forms. In the asymmetric forms this horizontal line rule is not followed with the intention of locating the bar in an oblique direction. The location of the incisions must be equidistant between two costal arches to allow an accurate fixation to the thoracic wall. Initial anesthetic blockage is recommended.

Skin, subcutaneous and muscular layer are dissected until the ribs are reached. Two costal arches are dissected in such a way as to prevent opening the parietal pleura, thus avoiding pneumothorax. Two small PVC tubes are introduced around the ribs. The integrity of the pleural cavity must be verified.

Then, a sub-pectoral bilateral tunnel is created. This is performed creating a tunnel in one hemi-thorax using a large scissors. Through this tunnel a trocar covered with a 1 cm PVC tube is inserted. The trocar is removed and the PVC tube is retained in its sub-pectoral location.

The sub-pectoral tunneling maneuver is repeated in the contralateral hemi-thorax where another trocar is introduced. The end of the trocar is introduced into the PVC tube and pushed until it appears in the contralateral incision. Care is required to prevent the tip of the trocar from entering the thoracic cavity.

The PVC tube is removed and replaced with a larger one. Through this tube a pectus bar that has been bent to the desired thoracic contour is introduced.

Stainless steel wires (316 L) are inserted in both pericostal PVC tubes. They are slipped around the ribs inside the tubes avoiding lung damage. The quantity and adequate wire size are chosen considering the age of the patient and chest wall flexibility.

A sub-muscular pocket is performed bilaterally in order to lodge the fixation plates. These plates are located and secured to the ribs using the periosteal wires through the peripheral holes of the fixation plate. This fixation must be initially loose to facilitate the union between the bar and the plate.

The bending of the bar is performed aided by a template. The passage of the bar is commenced with the concavity facing anteriorly, and is rotated while it is inserted, eventually to achieve the concavity facing the chest wall downwards.

Compression and fixation is performed in a progressive manner. The ends of the compressive bar are secured with screws to the threaded holes located in the fixation plates. The procedure begins with the ends of the bar installed in the first holes of the fixation plate.

Once fixed in this initial position at both ends, a process of changing the location of the screws in progressive, paused and alternating manner starts at both ends. Then a gradual compression is applied until the adequate chest contour is obtained. Finally, the pericostal wires are firmly adjusted and the muscular and skin layers are closed. The final result of the P.C. repair is achieved through this method.

Procedure Overview (Figs. 47.1, 47.2, 47.3, 47.4, 47.5, and 47.6)

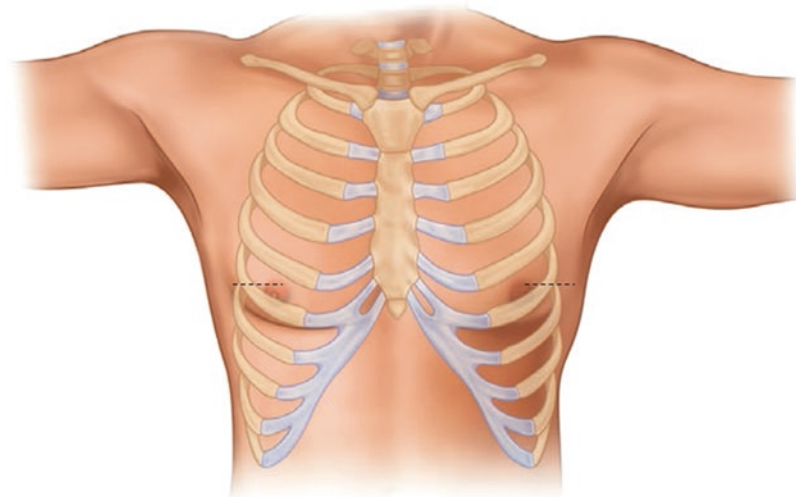

Fig. 47.1 Small horizontal incisions of 2-cm are made in both axillar regions on the middle axillar line. Skin, subcutaneous and muscular layer are dissected until the ribs is reached

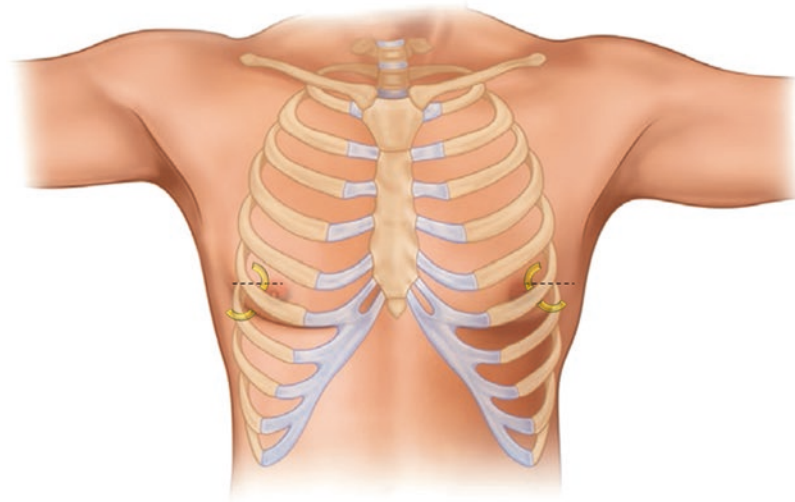

Fig. 47.2 Two small PVC tubes are introduced around the ribs

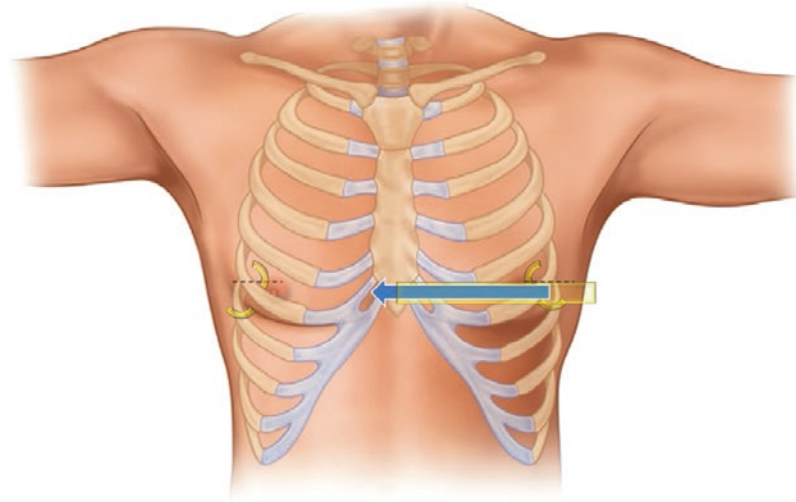

Fig. 47.3 A sub-pectoral bilateral tunnel is created. This is performed creating a tunnel in one hemi-thorax using a large scissors. Through this tunnel a trocar covered with a 1 cm PVC tube is inserted. The trocar is removed and the PVC tube is retained in its sub-pectoral location

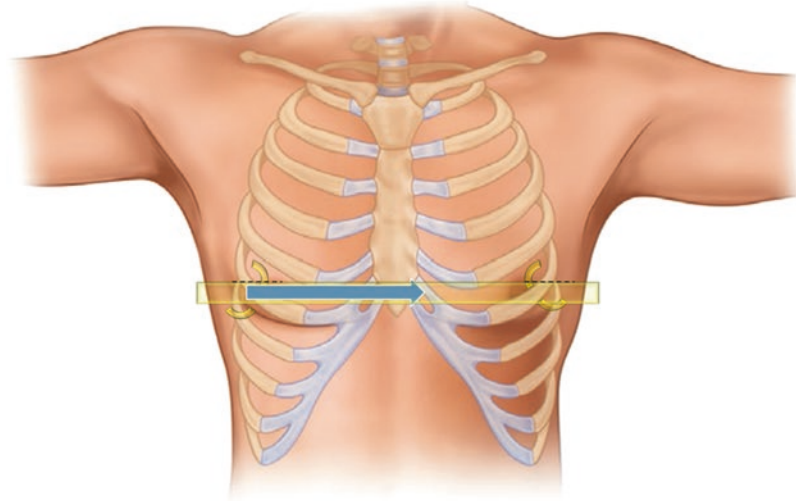

Fig. 47.4 The sub-pectoral tunneling maneuver is repeated in the contralateral hemi-thorax where another trocar is introduced. The end of the trocar is introduced into the PVC tube and pushed until it appears in the contralateral incision

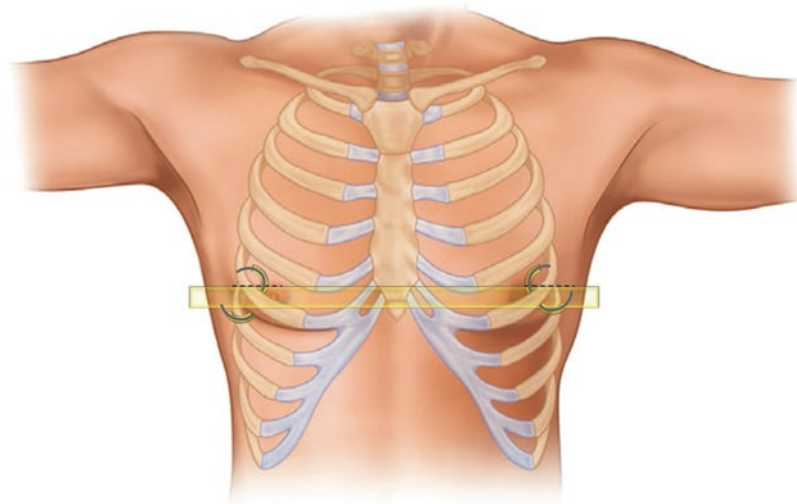

Fig. 47.5 Stainless steel wires (316 L) are inserted in both pericostal PVC tubes. The passage of the pectus bar is commenced through the PVC tube with the concavity facing anteriorly, and is rotated while it is inserted, eventually to achieve the concavity facing the chest wall downwards

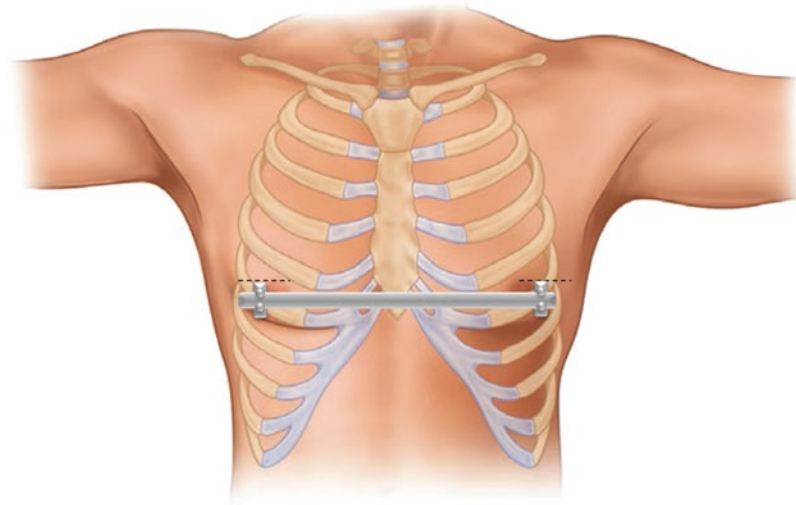

Fig. 47.6 The ends of the pectus bar are secured with screws to the threaded holes located in the fixation plates. First the ends of the bar are installed in the first holes of the fixation plate. Once fixed in this initial position in both ends, a process of changing the location of the screws in progressive, paused and alternating manner starts at both ends. Then a gradual compression is applied until the adequate chest contour is obtained, after which the pericostal wires are firmly adjusted

Postoperative Management

Postoperative care includes pain treatment, antibiotic therapy during surgery (cefazolin) which continuous until discharge and parenteral hydration for 24 hrs.

Patients with elastic chest wall tolerate pain reasonably well. Older patients or with low flexibility may experience pain within the first week after surgery. Chest compression and comprehensive correction of the thoracic bone structure are the reasons for frequent spine pain.

Early mobilization is recommended and the patient is discharged from the hospital after 2–4 days.

The steady fixation of the bar by means of four pericostal wires offers stability to resist the body movements. Both the flexion and thoracic torsion do not present a risk in this method of P.C. repair technique.

Massages in the pre-sternal region as well as in both axillary regions are important in the initial post-operative period. This is to avoid adherence of the bar to the skin. Due to the limited

fatty tissue that these patients usually present, it is important to take precautions to avoid this complication.

Patients are controlled weekly in the period after surgery, and then every 3 months until the pectus bar is removed, the time of which is variable. The completion of body growth is the most appropriate time to plan the pectus bar removal. Usually the bar is retained for 2 years after which it is removed. Patients are requested to avoid extreme activities and refrain from participating in contact sports.

Results

Evaluations have been performed on a group of 40 patients that have been treated with this procedure between 2002 and 2009, in whom 20 have had pectus bar removals. Mean follow-up since the time of surgery was 2.49 years and mean follow-up from the time of pectus bar removal was 1.53 years. The average duration of hospital stay during pectus implantation was 3.8 days, whereas it was 1.4 days after pectus bar removal. Patients resumed routine activities after 14 days.

The criteria for results evaluations we applied are:

1. Excellent: harmonic thoracic perimeter, lack of dermatological complications.
2. Good: improvement of the chest contour with some asymmetries.
3. Fair: reasonable improvement of the chest and/or presence of dermal problems such as adherence and hyperpigmentation.
4. Poor: slight improvement as a result of premature removal because of pain, infection or dermal adherence with hyperpigmentation, ampoules, or ulcers.

Results of Patients after bar removal were Excellent: n = 10; Good: n = 4; Fair: n = 4; Poor: n = 2; Total: n = 20

All patients retained the desired thoracic contours after bar removal, and there were no relapses of the malformations. No intraoperative complications were observed. With an average blood loss of 14.7 ml, no transfusions were necessary.

Complications

- The skin-bar adherence is the most common complication observed in these 40 cases. Reasons that explain this complication are: allergic cutaneous reaction, infection and also the low density of fat tissue that is present in this area.

  The dermatological adherence appeared in 8 of the first 24 cases. Later we modified the technique to achieve a sub-pectoral placement of the pectus bar, hence reducing the frequency of this reaction.
- Seroma is also a frequent finding and it appears due to compression and rubbing of the bar surface adjacent to the sternum and cartilages. Seroma is treated with local ice appliance, aspiration of fluid by puncture, anti-inflammatory medication and cutaneous massages. If a seroma recurs and the punctured secretion obtained is turbid, removal of the pectus bar is planned.
- Wire breakage occurred in three patients as a consequence of growth and physical activity. The 0.3-mm wire 316 L was replaced by a thicker one of 0.9 mm, which did not break. When breakage or loosening of the wires happens in a late period as consequence of excessive growth, breakage is not considered as a complication. Close to the completion of

first year after implantation, the bar is covered by a fibrotic envelope that compensates for broken wires that were used for fixation and therefore the retained bar continues to be effective.

- In five cases, we noticed excessive growth of the xiphoid region below the zone where the bar is located. This growth can turn the bar toward the caudal region. In 3 of these 5 patients, the first bar was removed and replaced by a second bar in a lower localization to resolve the residual protrusion.

  Two other patients previously treated by modified Ravitch technique were re-do successfully treated by this P.C. method.
- One patient with a rigid chest wall required bar removal due to persistent pain and skin adherence.
- One patient developed infection in the one of the scars in the axillary region. The infection was managed with antibiotics and appropriate wound care. In spite of this treatment, the patient developed skin adherence and eventually the bar was removed after 3 months post-surgery.
- In two patients there were incomplete correction observed and the desired result was not achieved.
- One patient with pneumothorax required a pleural drain.

Procedure Related Images:

(Figs. 47.7, 47.8, 47.9, 47.10, 47.11, 47.12, 47.13, 47.14, 47.15, 47.16, 47.17, 47.18, 47.19, 47.20, 47.21, 47.22, 47.23, 47.24, 47.25, 47.26, 47.27, 47.28, 47.29, 47.30, 47.31, 47.32, 47.33, 47.34, 47.35, 47.36, 47.37, 47.38, 47.39, 47.40, 47.41, 47.42, 47.43, 47.44, 47.45, 47.46, 47.47, 47.48, 47.49, 47.50, 47.51, 47.52, 47.53, 47.54, 47.55, 47.56, 47.57, 47.58, 47.59, 47.60, 47.61, 47.62, 47.63, 47.64, 47.65, 47.66, 47.67, 47.68, 47.69, 47.70, 47.71, 47.72, 47.73, 47.74, 47.75, 47.76, 47.77, 47.78, 47.79, 47.80, 47.81, 47.82, 47.83, 47.84, 47.85, 47.86, 47.87, 47.88, 47.89, 47.90, 47.91, 47.92, and 47.93)

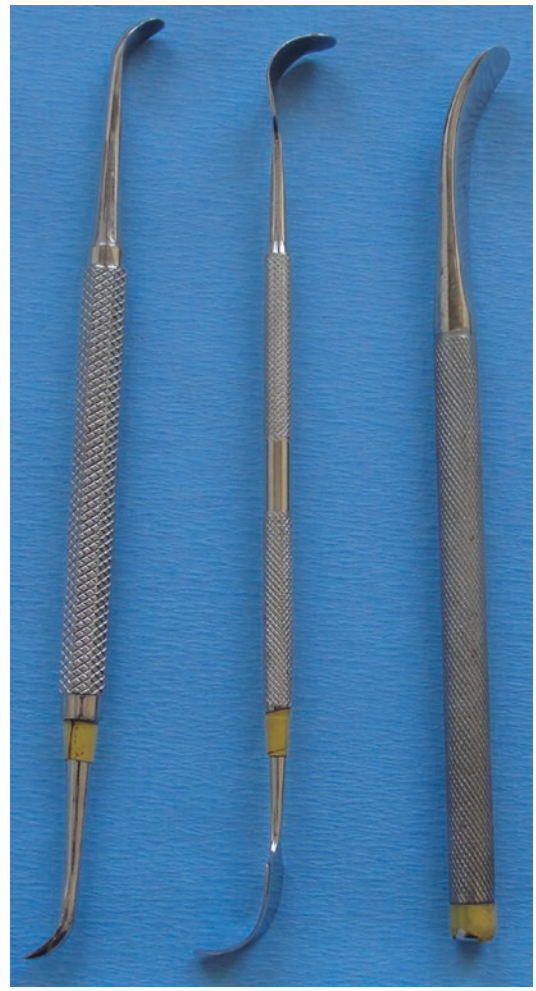

Fig. 47.7 Tools for costal dissection: periosteal elevators of several types

Fig. 47.8 Curved instruments to circumscribe the rib

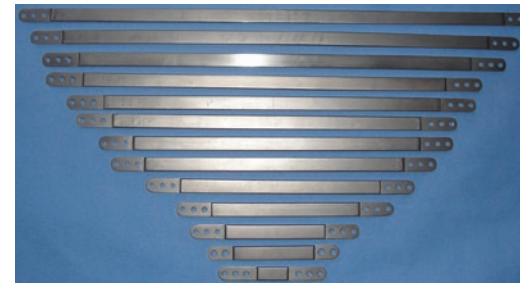

Fig. 47.9 Pectus bars of various sizes

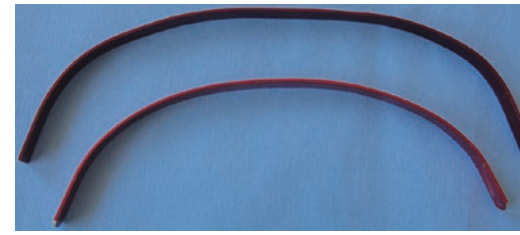

Fig. 47.10 Chest wall templates to configure the design of the pectus bars

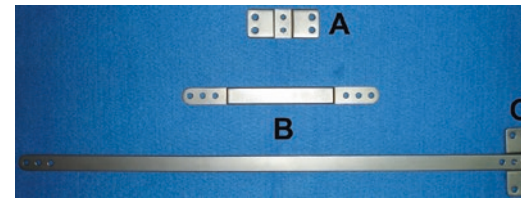

Fig. 47.11 Fixation plates with threaded holes and non-threaded holes

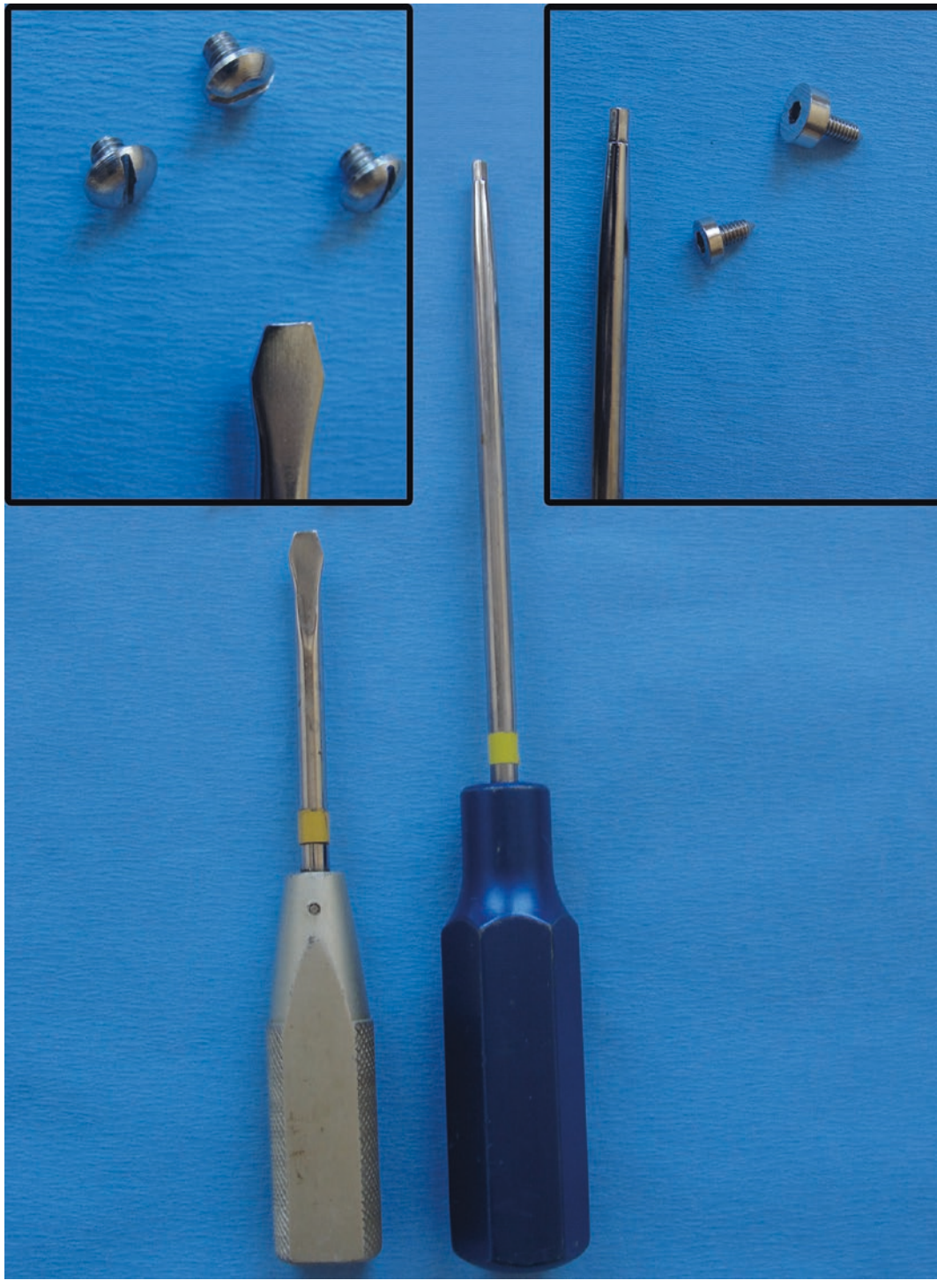

Fig. 47.12 Screws of several sizes and shapes to attach the bar to the plate

Fig. 47.13 Long scissors for preparation of the tunnel

Fig. 47.14 Metal hook

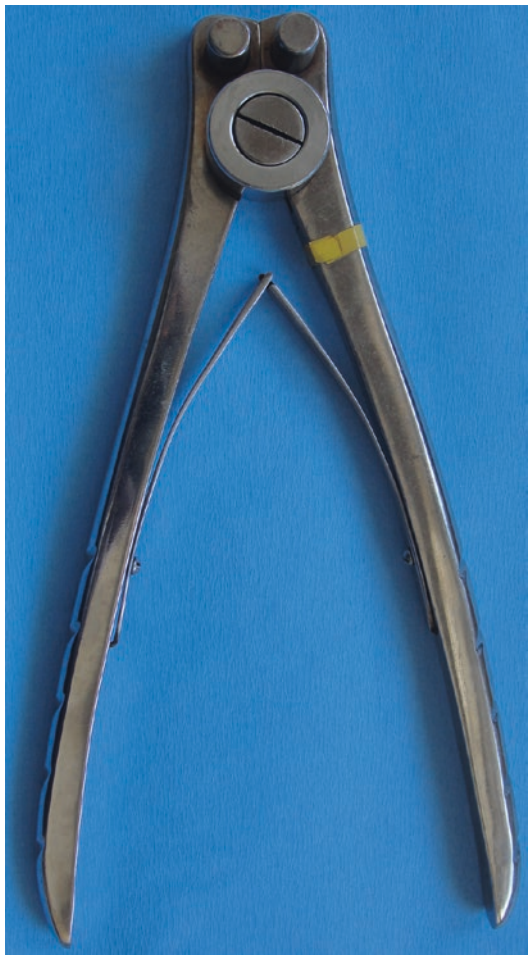

Fig. 47.15 Manual hand held benders

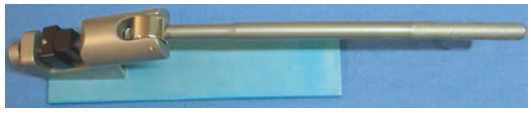

Fig. 47.16 Table benders

Fig. 47.17 Metal-bar benders to allow the contouring of the bar

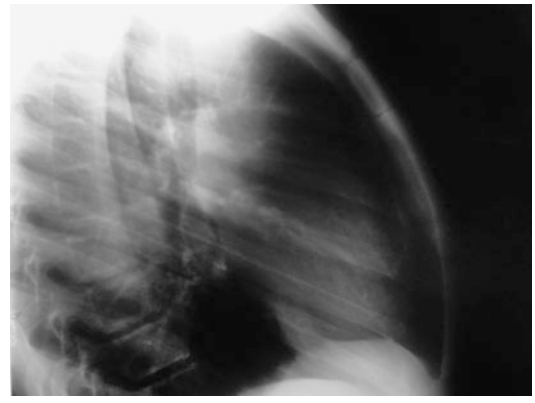

Fig. 47.18 Chest films lateral view

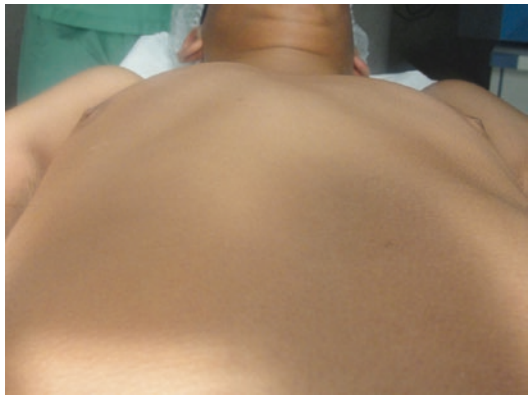

Fig. 47.19 Incision in asymmetric forms, intention is to locate the bar in an oblique direction

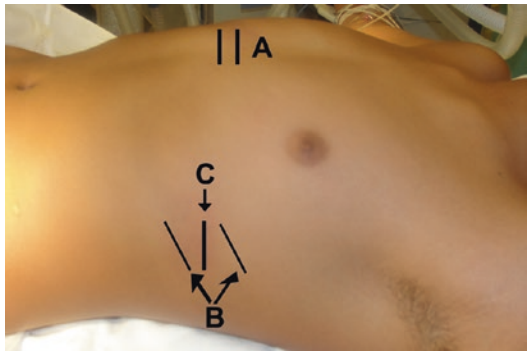

Fig. 47.20 Skin markings (*A*: pre-sternal tunnel, *B*: costal arches, *C*: incision sites)

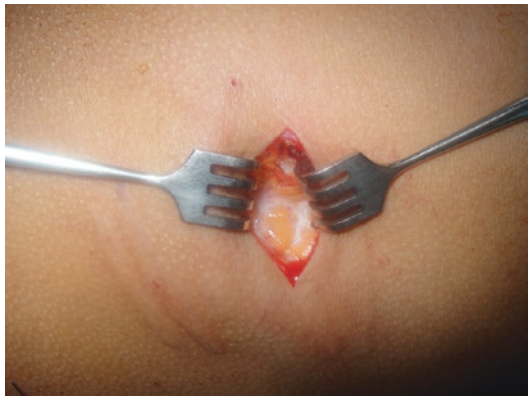

Fig. 47.21 Incision of the skin and subcutaneous layers

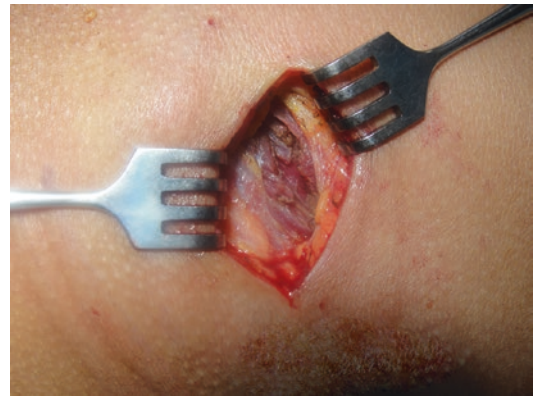

Fig. 47.22 Preparation of the muscle layer

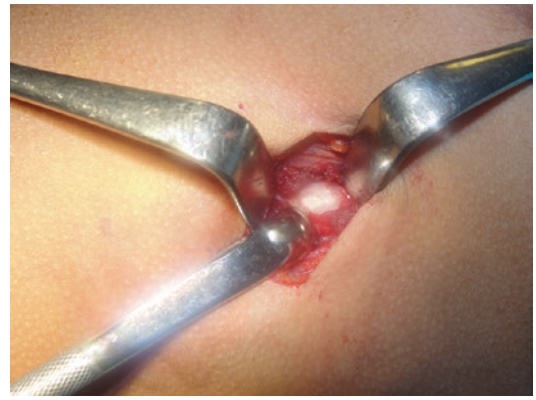

Fig. 47.23 Shaving the rib with a periosteal elevators

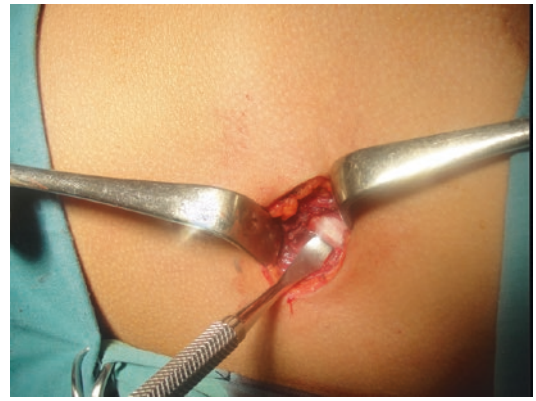

Fig. 47.24 Scrapping the posterior surface of the rib

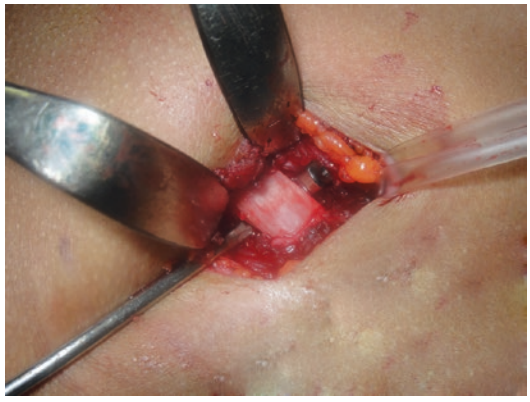

Fig. 47.25 Circumscribing the rib

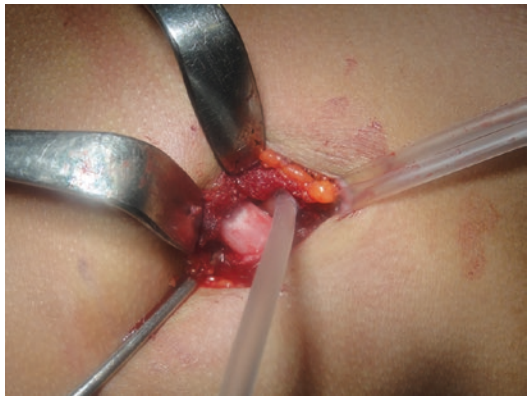

Fig. 47.26 Two small PVC tubes are introduced around the ribs

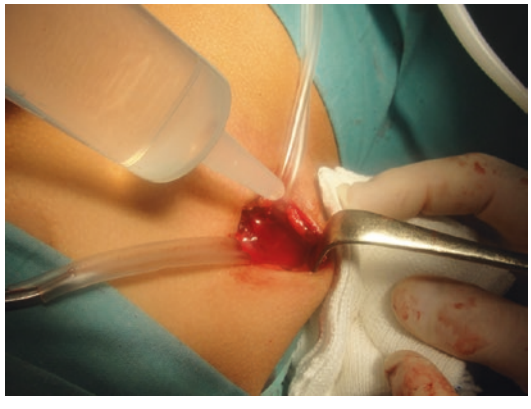

Fig. 47.27 The integrity of the pleural cavity is verified

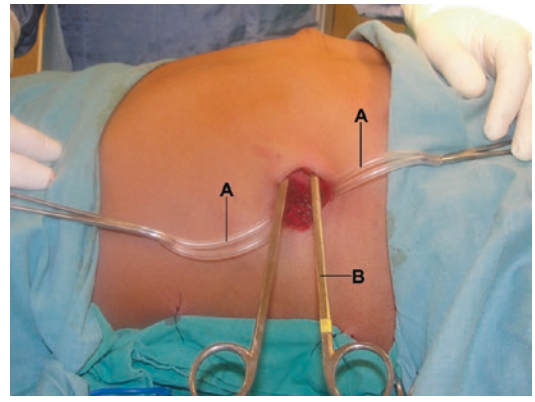

Fig. 47.28 Creation of a bilateral sub-pectoral bilateral tunnel using a large scissors

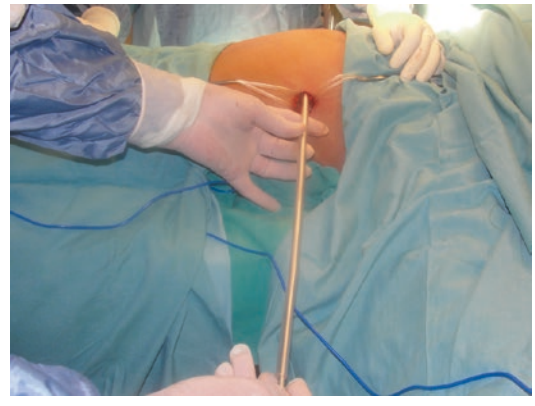

Fig. 47.29 Placement of the PVC tube with a trocar

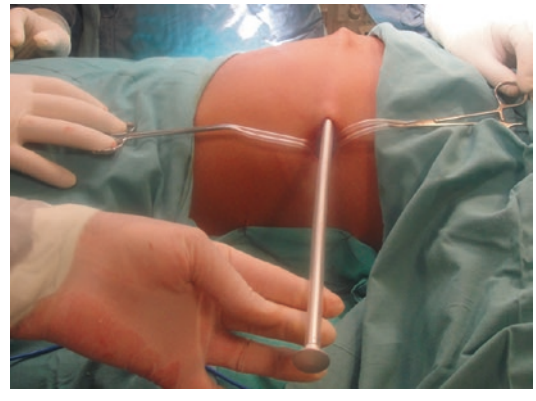

Fig. 47.30 Trocar and PVC tube in sub-pectoral location on the left side

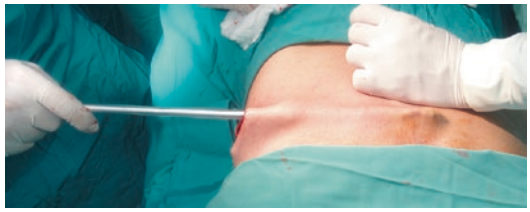

Fig. 47.31 Trocar and PVC tube in sub-pectoral location on the left side

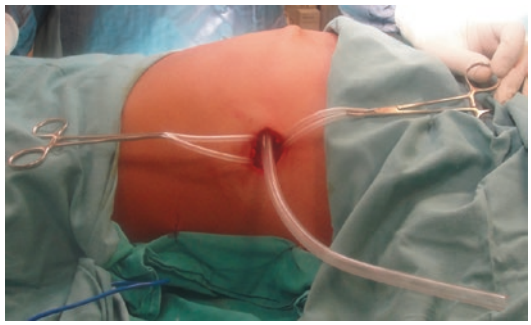

Fig. 47.32 The trocar is removed and the PVC tube is retained in its sub-pectoral location

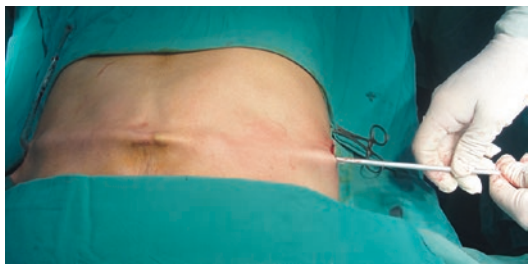

Fig. 47.33 The sub-pectoral tunneling maneuver is repeated in the contralateral hemi-thorax where another trocar is introduced

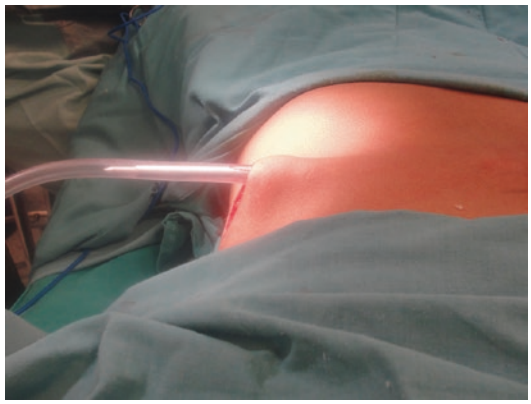

Fig. 47.34 The end of the trocar is introduced into the PVC tube and pushed until it appears in the contralateral incision

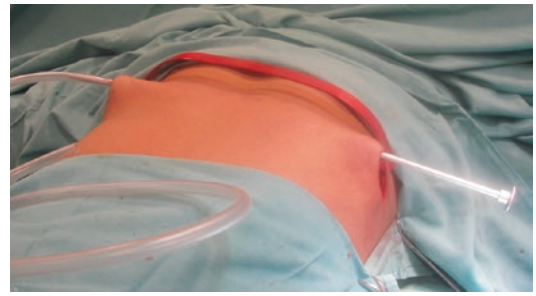

Fig. 47.35 The PVC tube is removed and replaced with a larger PVC tube

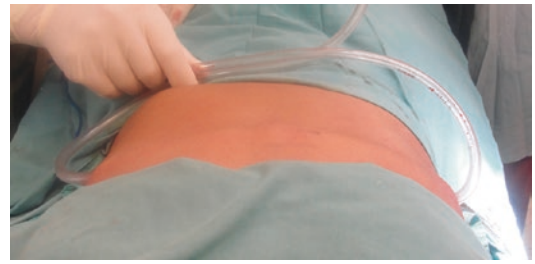

Fig. 47.36 The longer PVC tube without the trocar

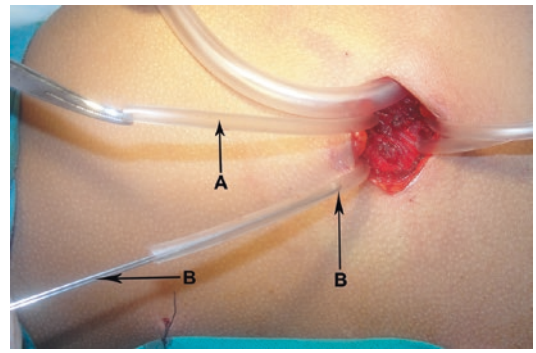

Fig. 47.37 Pericostal wire progression (*A*: wire inside the PVC tube)

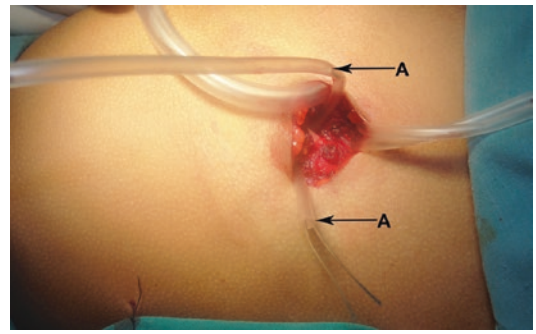

Fig. 47.38 Pericostal wire progression (*A*: wire inside the PVC tube)

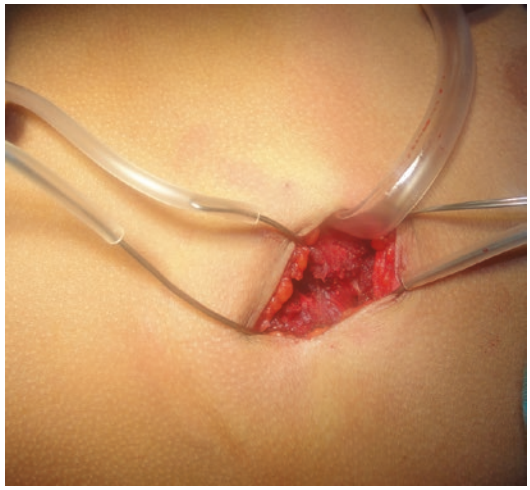

Fig. 47.39 Pericostal wires

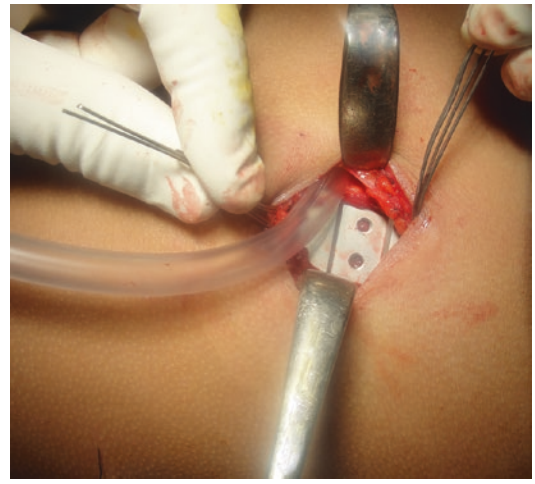

Fig. 47.42 Submuscular fixation plate location

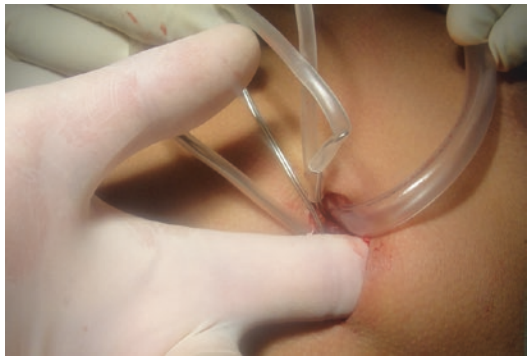

Fig. 47.40 A sub-muscular pocket is performed bilaterally in order to lodge the fixation plates

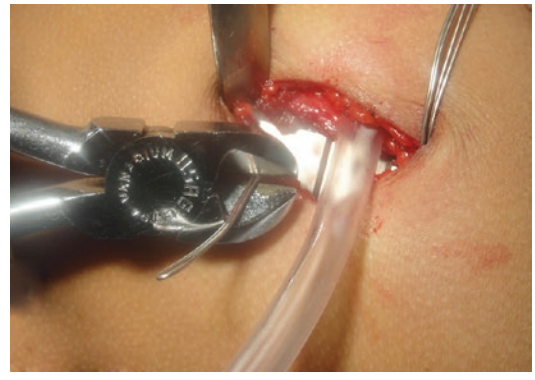

Fig. 47.43 The ends of the wires are cut

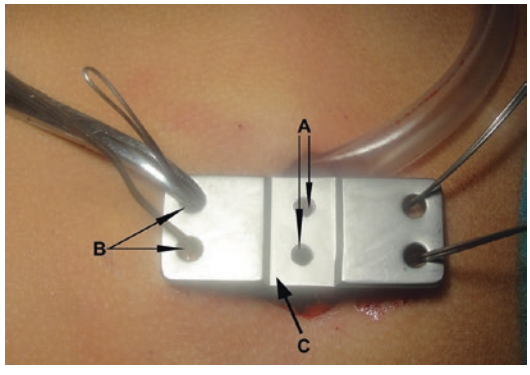

Fig. 47.41 Fixation plate insertion (*A*: threaded holes, *B*: non-threaded holes, *C*: grooves)

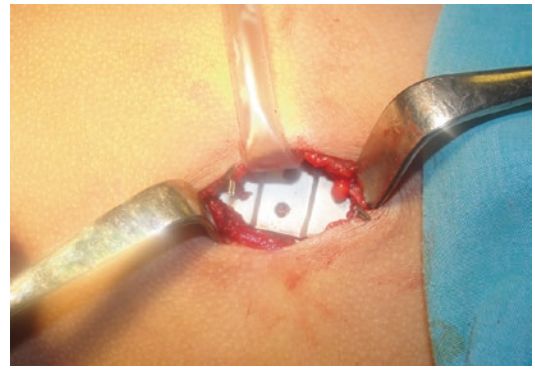

Fig. 47.44 The fixation must be initially loose to facilitate the union between the bar and the plate

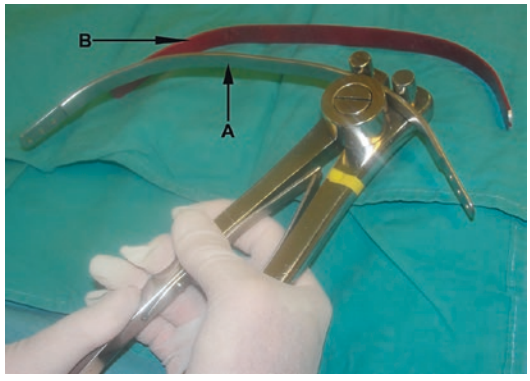

Fig. 47.45 The manual bending of the bar is aided by a template (*A*: pre-sternal bar, *B*: template)

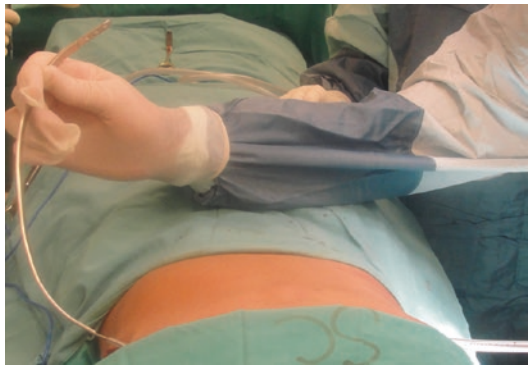

Fig. 47.46 Insertion of the pre-sternal bar guided by the longer tube

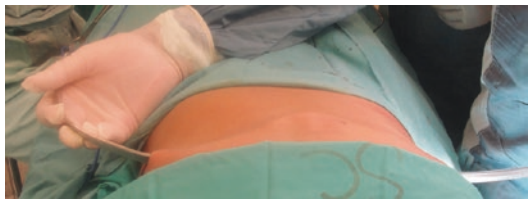

Fig. 47.47 Advancing and turning the bar

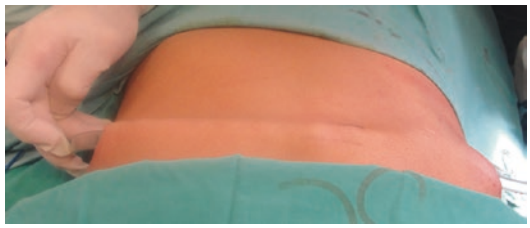

Fig. 47.48 The bar reaching the fixation plate

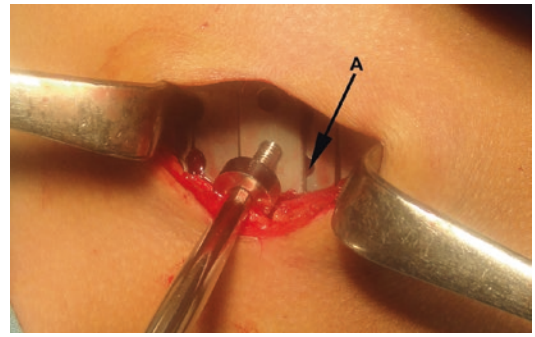

Fig. 47.49 Initial adjustment approximating the bar and fixation plate (*A*; threaded hole)

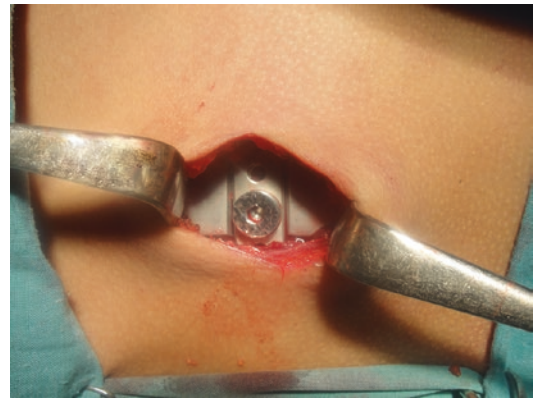

Fig. 47.50 Final adjustment on one side

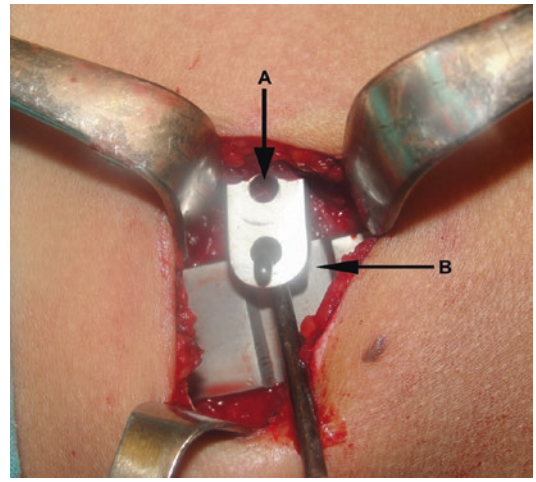

Fig. 47.51 Approximating the bar with the fixation plate on the other side aided by the hook (*A*: non-threaded hole, *B*: groove of the fixation plate)

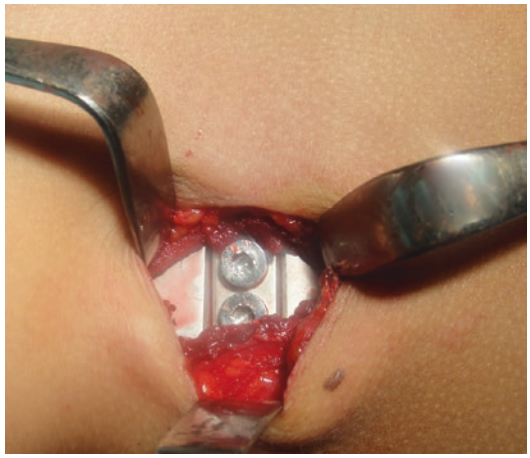

Fig. 47.52 Second screw insertion when desired compression is reached

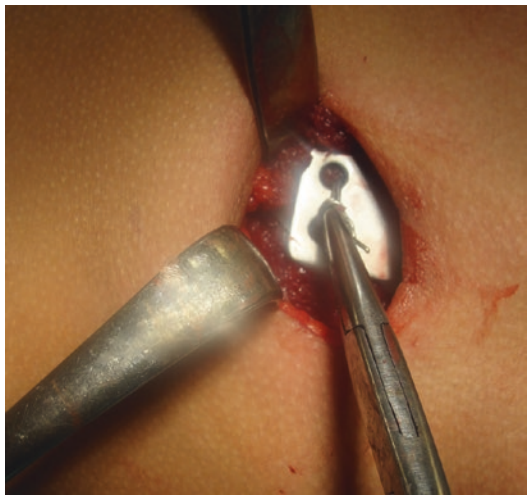

Fig. 47.53 The pericostal wires are firmly adjusted

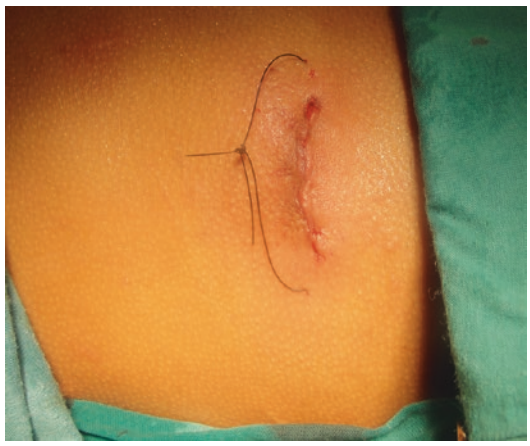

Fig. 47.54 Closure of the skin layer

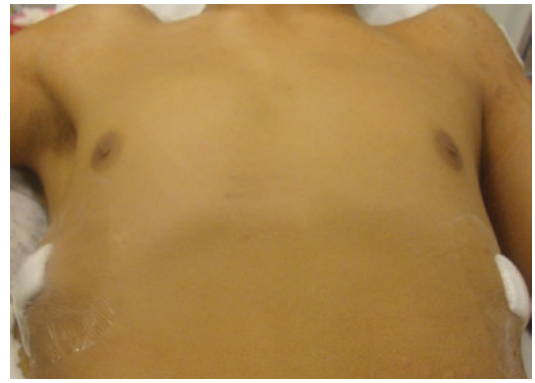

Fig. 47.55 The final result of *Pectus Carinatum* repair

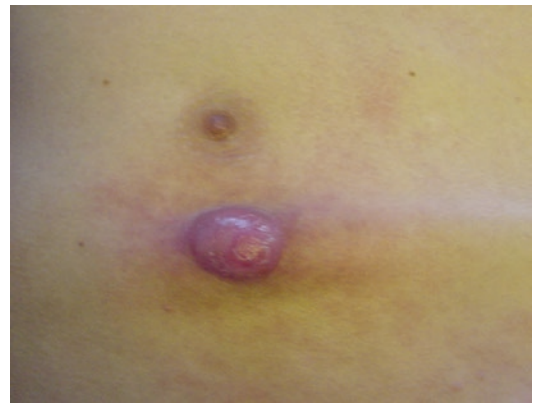

Fig. 47.56 Skin-bar adherence (ampulla) complication of the procedure

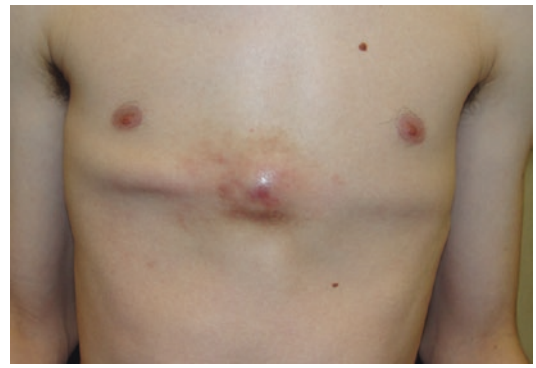

Fig. 47.57 Skin-bar adherence (seroma) complication of the procedure

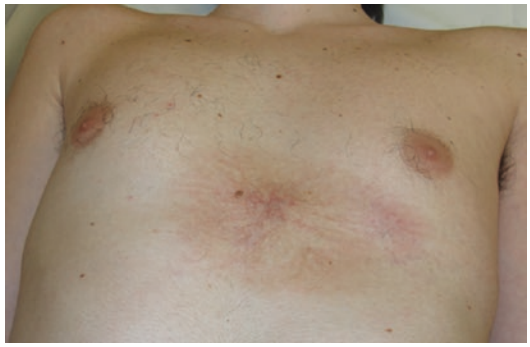

Fig. 47.58 Skin-bar adherence (seroma and dermal irritation) complication of the procedure

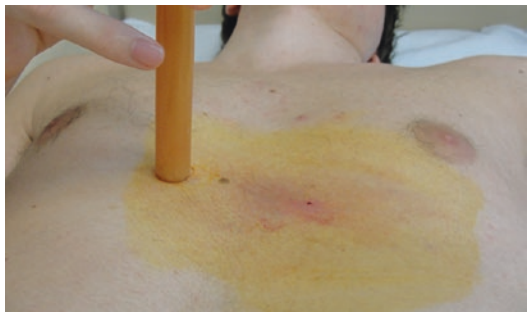

Fig. 47.59 Skin-bar adherence (seroma and dermal irritation post puncture) complication of the procedure

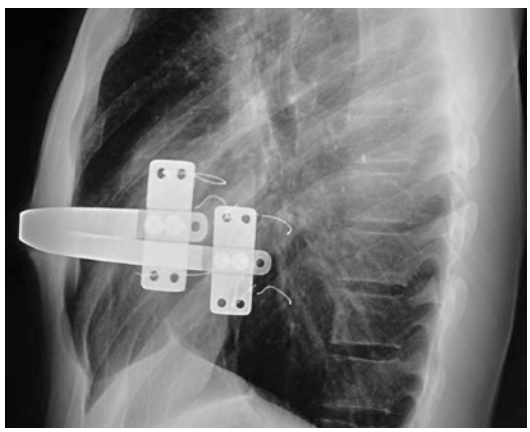

Fig. 47.60 Wire breakage: chest film lateral view 3 years post-surgery (Patient Case Nr. 1- Image 59)

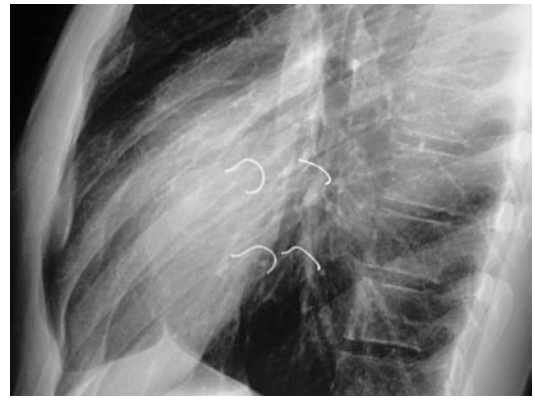

Fig. 47.61 Wire breakage chest film lateral view after pectus bar removal (Patient Case Nr. 1- Image 59)

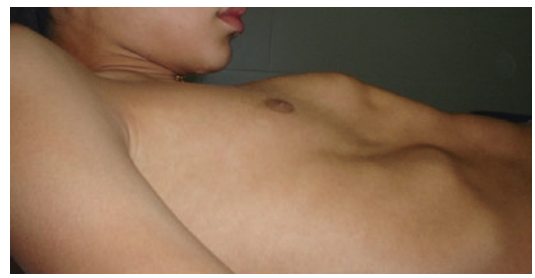

Fig. 47.62 Case Nr 1: Severe *Pectus Carinatum* lateral view

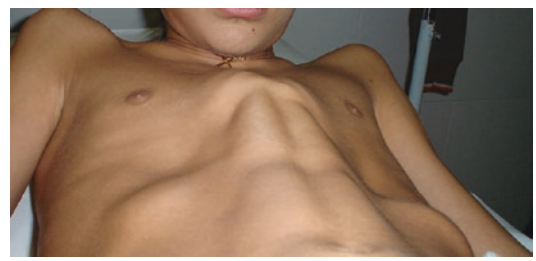

Fig. 47.63 Case Nr. 1: Severe *Pectus Carinatum* raising the head, anterior view

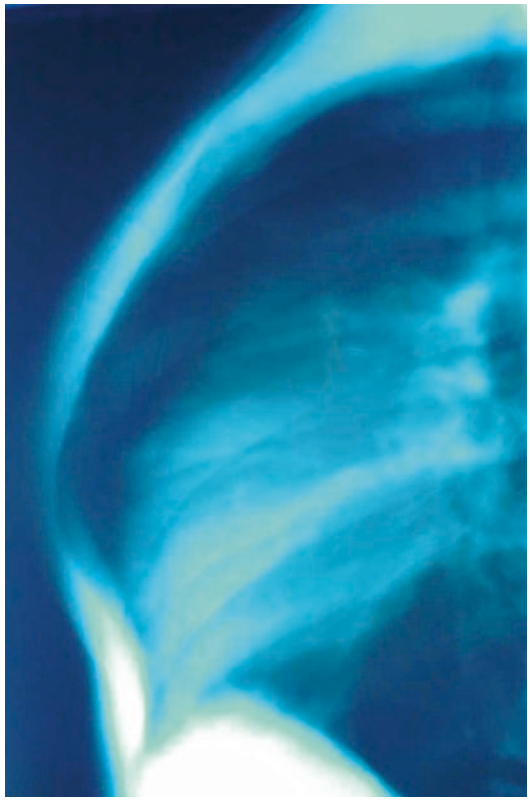

Fig. 47.64 Case Nr. 1: Severe *Pectus Carinatum* X-ray lateral view

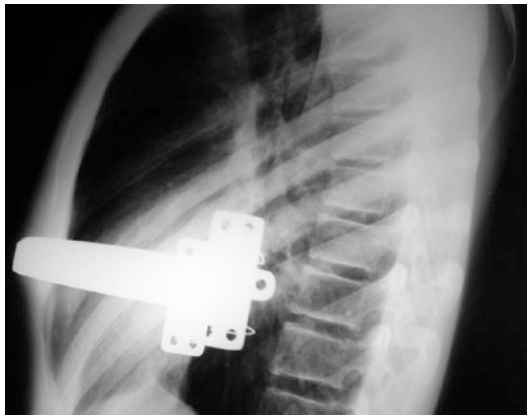

Fig. 47.65 Case Nr. 1: Severe *Pectus Carinatum* X-ray lateral view after surgery

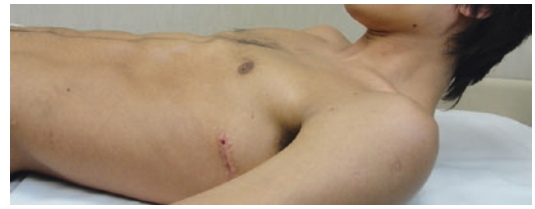

Fig. 47.66 Case Nr. 1: Severe *Pectus Carinatum* after bar removal with head raised in lateral view

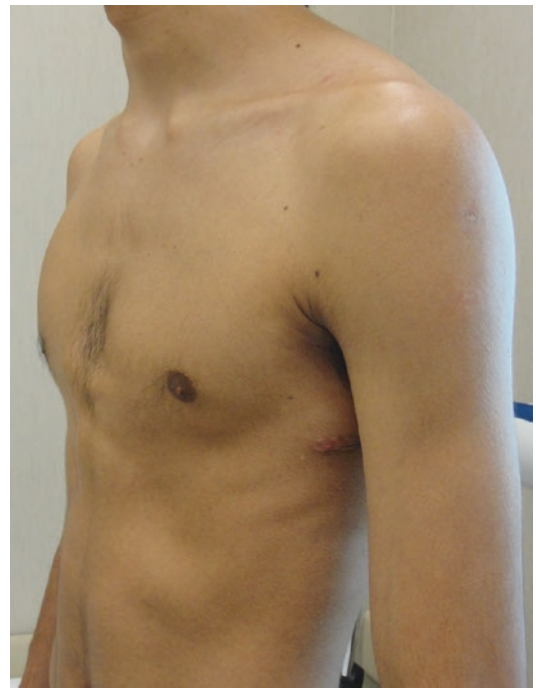

Fig. 47.67 Case Nr. 1: Severe *Pectus Carinatum* after bar removal, lateral view

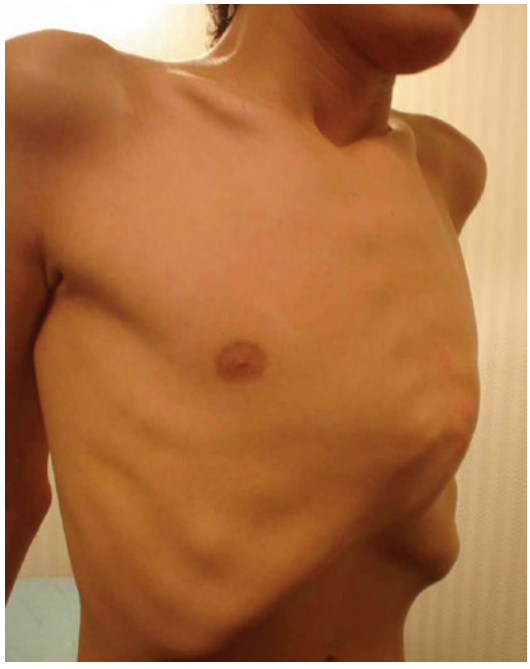

Fig. 47.68 Case Nr. 2: Severe asymmetric *Pectus Carinatum*

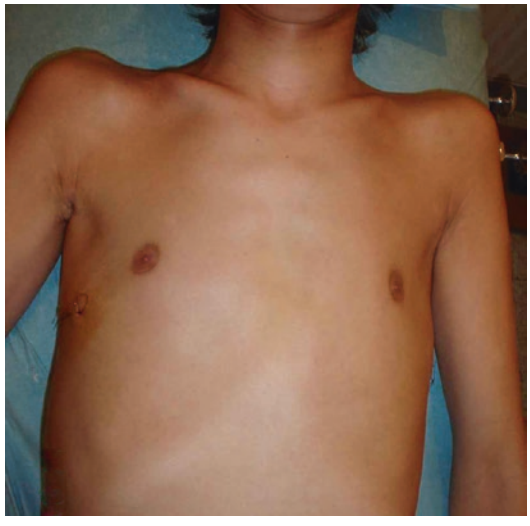

Fig. 47.69 Case Nr. 2: Severe asymmetric *Pectus Carinatum* after surgical correction

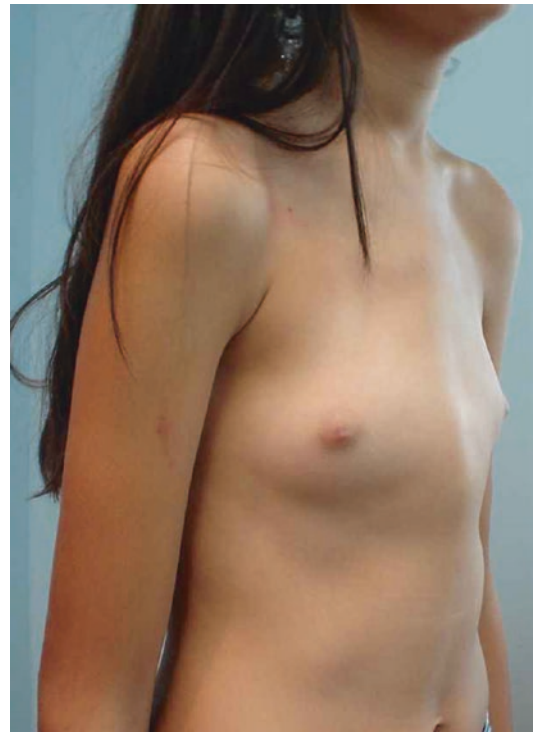

Fig. 47.70 Case Nr. 3: Female Chondrogladiolar *Pectus Carinatum*

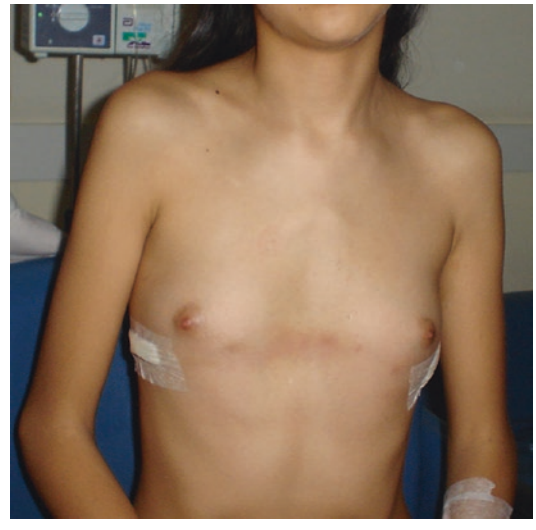

Fig. 47.71 Case Nr 3: Female Chondrogladiolar *Pectus Carinatum* post-surgery

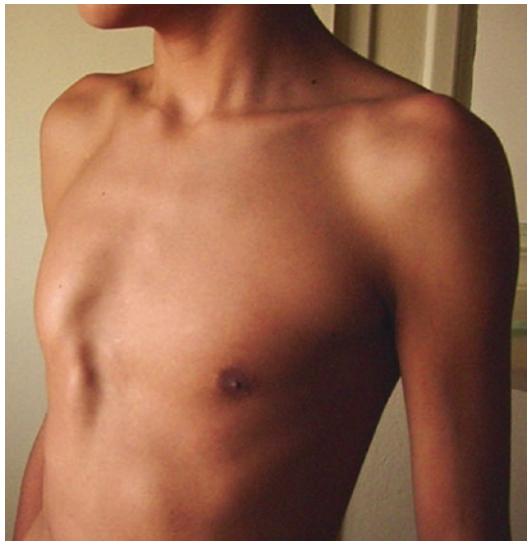

Fig. 47.72 Case Nr. 4 (elder sibling of Case Nr 5): asymmetric *Pectus Carinatum* with lateral view

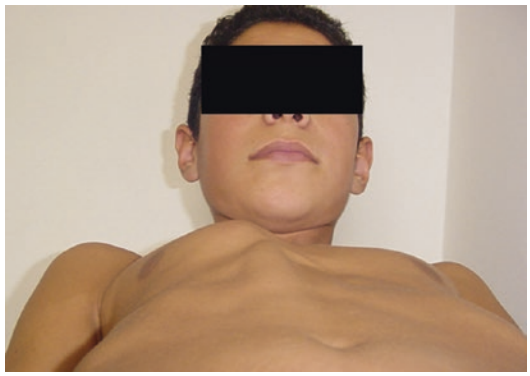

Fig. 47.73 Case Nr. 4 (elder sibling of Case Nr 5): asymmetric *Pectus Carinatum* with raised the head and caudal view

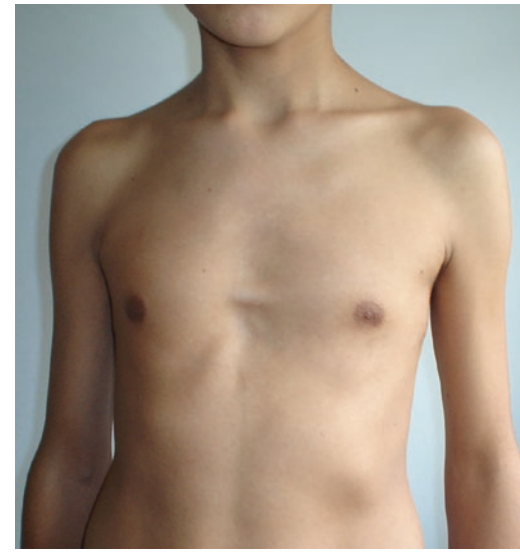

Fig. 47.74 Case Nr. 4 (elder sibling of Case Nr 5): asymmetric *Pectus Carinatum* after surgery

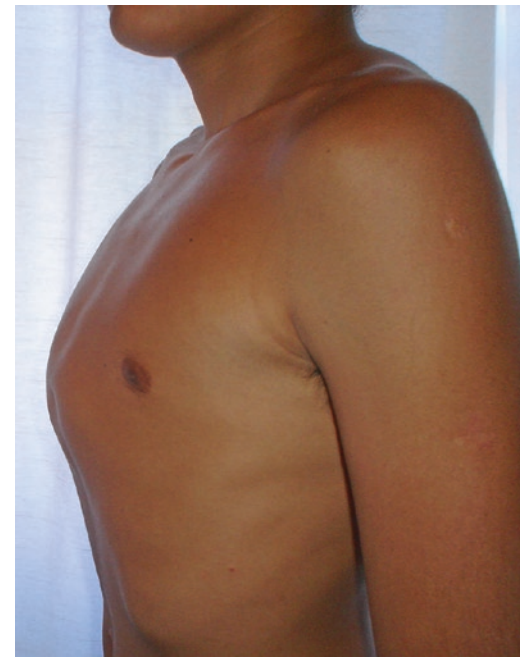

Fig. 47.75 Case Nr. 5 (younger sibling of Case Nr 4): Symmetric Chondrogladiolar *Pectus Carinatum* with lateral view

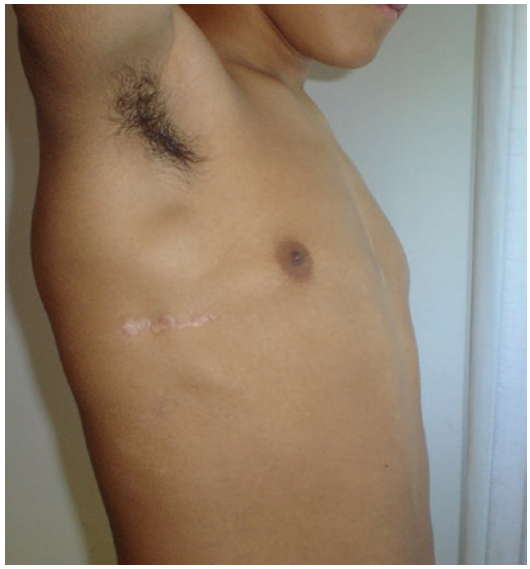

Fig. 47.76 Case Nr. 5 (younger sibling of Case Nr 4): Symmetric Chondrogladiolar *Pectus Carinatum* lateral view after surgery

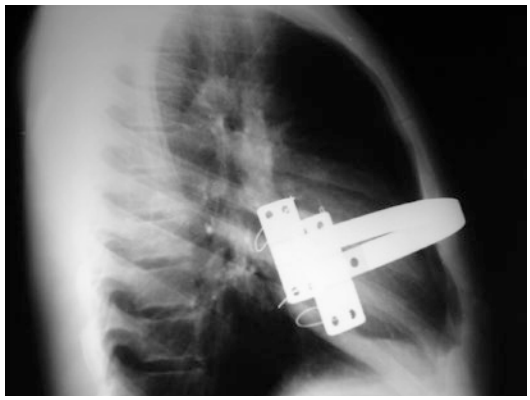

Fig. 47.77 Case Nr. 5 (younger sibling of Case Nr 4): Symmetric Chondrogladiolar *Pectus Carinatum* chest film after surgery

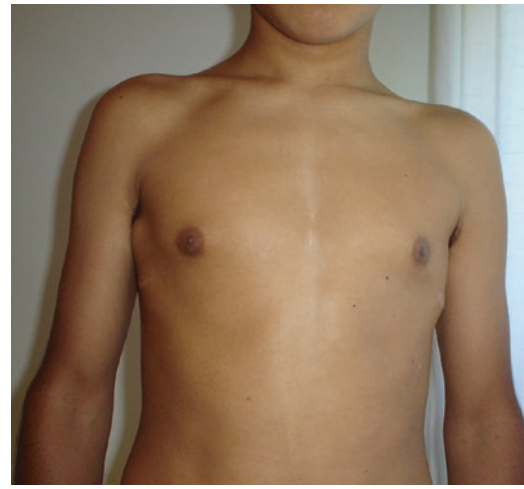

Fig. 47.78 Case Nr. 5 (younger sibling of Case Nr 4): Symmetric Chondrogladiolar *Pectus Carinatum* in anterior view

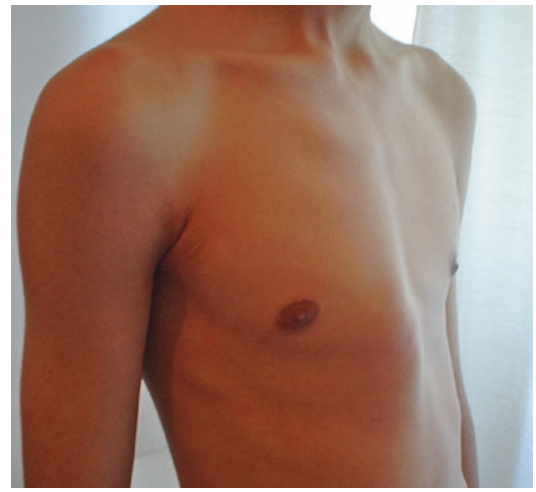

Fig. 47.79 Case Nr. 6: asymmetric *Pectus Carinatum* in lateral view

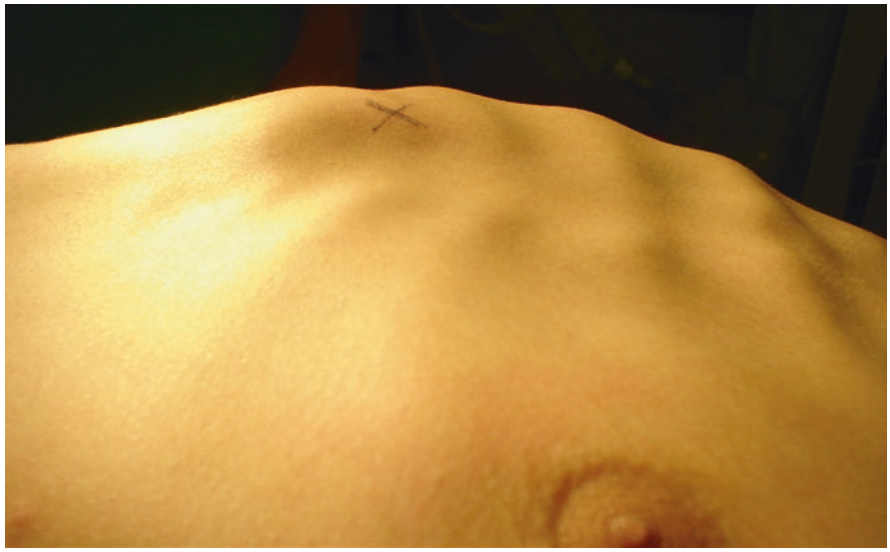

Fig. 47.80 Case Nr. 6: asymmetric *Pectus Carinatum* in lateral view

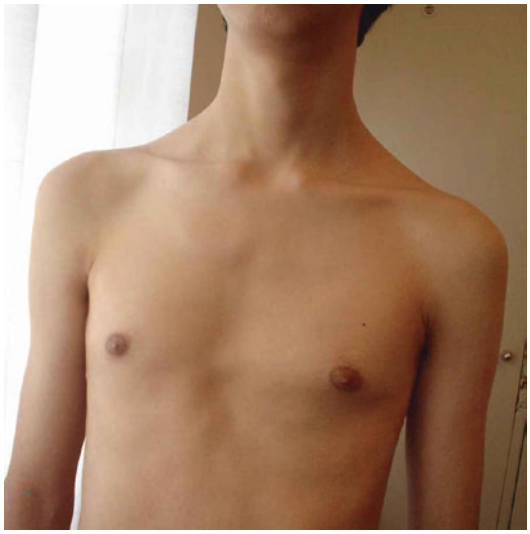

Fig. 47.81 Case Nr. 6: asymmetric *Pectus Carinatum* frontal view after surgery

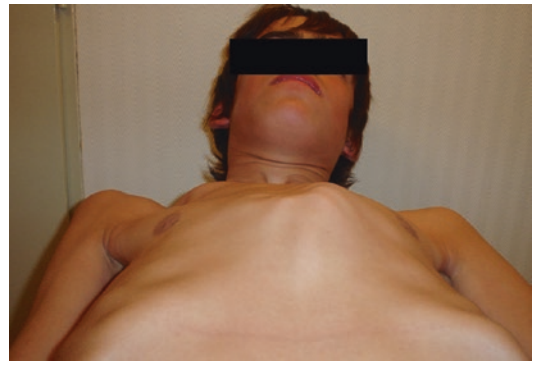

Fig. 47.82 Case Nr. 7: Severe asymmetric *Pectus Carinatum* with raised head

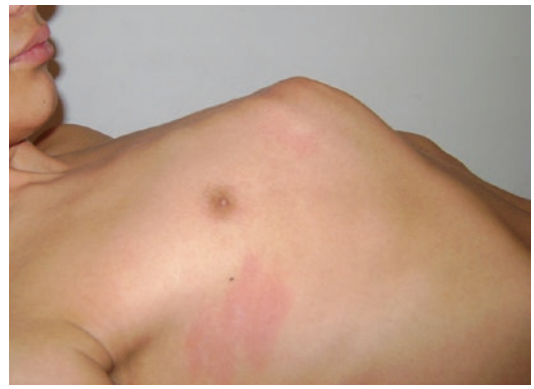

Fig. 47.83 Case Nr. 7: Severe Symmetric *Pectus Carinatum* with raised head in lateral view

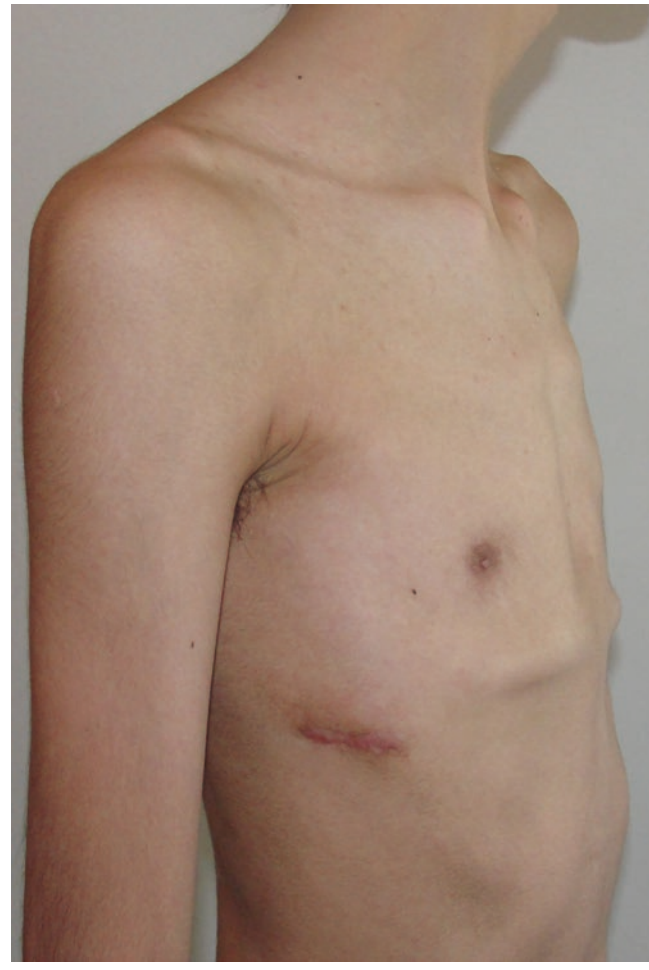

Fig. 47.84 Case Nr. 7: Severe Symmetric *Pectus Carinatum* lateral view after surgery

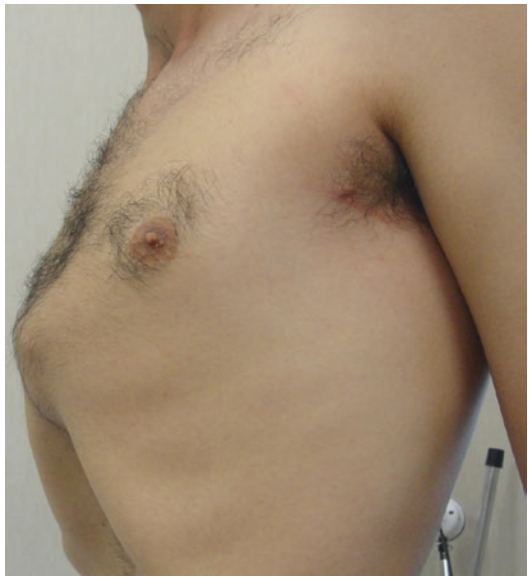

Fig. 47.85 Case Nr. 8: Severe Symmetric *Pectus Carinatum* in lateral view

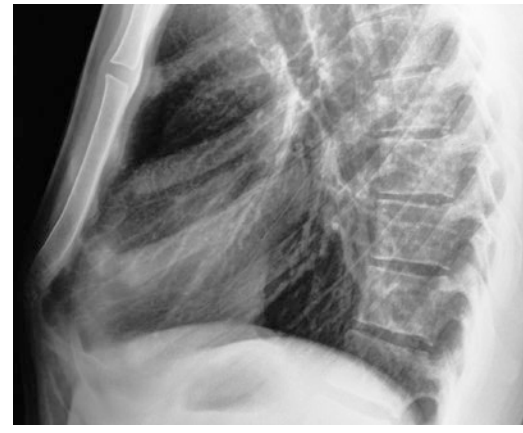

Fig. 47.86 Case Nr. 8: Severe Symmetric *Pectus Carinatum* in chest film in lateral view

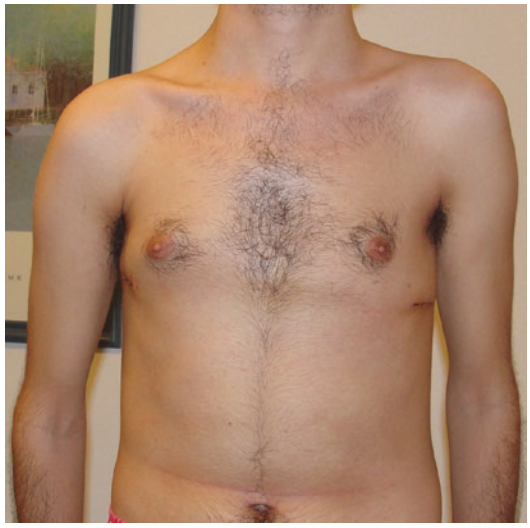

Fig. 47.87 Case Nr. 8: Severe Symmetric *Pectus Carinatum* after surgery on anterior view

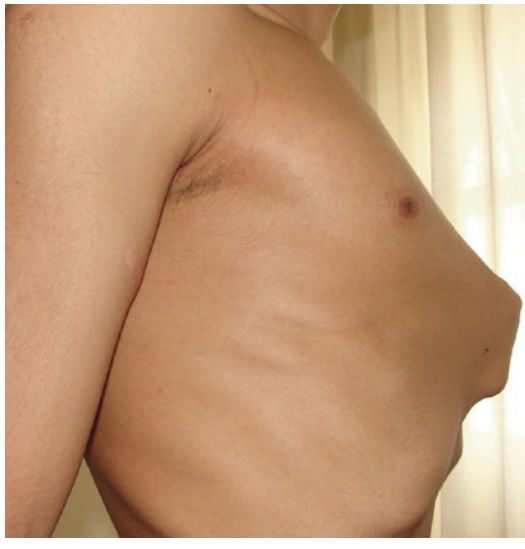

Fig. 47.88 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* of the right side

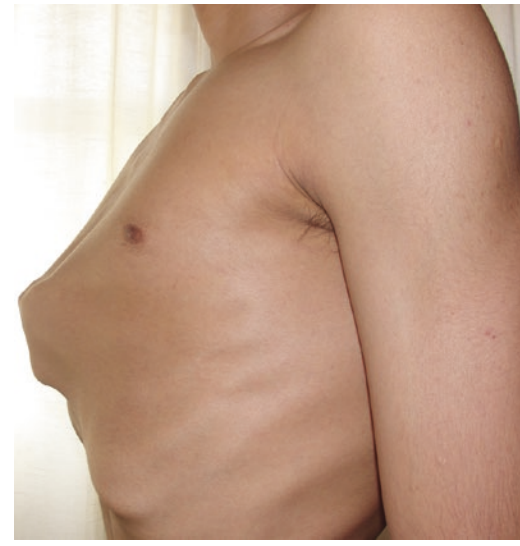

Fig. 47.89 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* of the left side

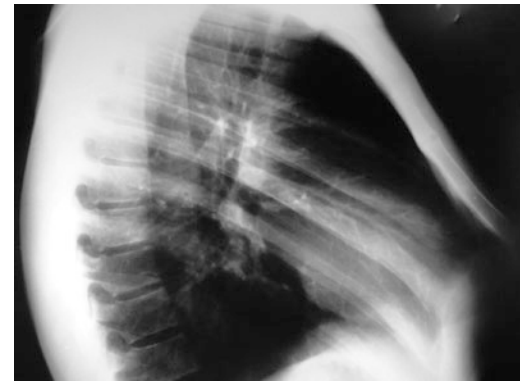

Fig. 47.90 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* on chest film in lateral view

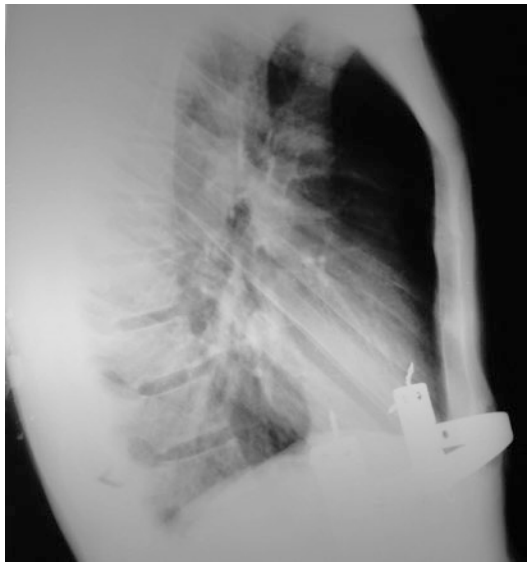

Fig. 47.91 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* after surgery on chest film in lateral view

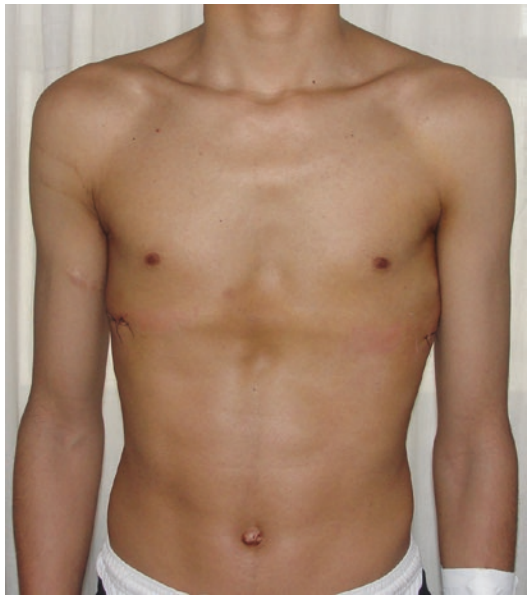

Fig. 47.92 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* after surgery on anterior view

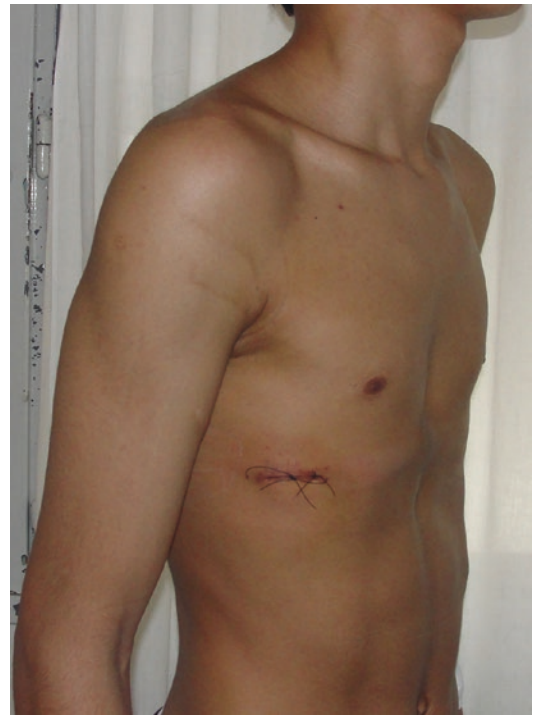

Fig. 47.93 Case Nr. 9: Severe Chondrogladiolar *Pectus Carinatum* after surgery on lateral view

References

1. Abramson H. A minimally invasive technique to repair pectus carinatum. Preliminary report Arch Bronconeumol. 2005;41:349–51.
2. Abramson H, D'Agostino J, Wuscovi S. A 5-year experience with a minimally invasive technique for pectus carinatum repair. J Pediatr Surg. 2009; 44(1):118–23.
3. Abramson H, Pisoni S, Rodriguez M, Segura G. Pectus carinatum repair by minimally invasive procedure. New trends about pectus carinatum repair. J Clin Anal Med. 2010;1(1):61–4.

Minimal Access Repair of Pectus Carinatum

48

Andras Hock

Technical Highlights

Minimal access repair of Pectus Carinatum or the Hock techniques is suitable for repair of varying degrees of protruding deformities of the sternum.

The main technical highlights of this procedure are:

(a) Selection and preforming of pectus bar prior to thoracoplasty
(b) Transverse left lateral chest wall incision is made traversing the mid-axillary line
(c) Blunt preparation of the intercostal region where the bar will be positioned
(d) Creation of a transverse subcutaneous tunnel in front of the sternum using clamps
(e) The tunnel should be up to the level of the right sternocostal margin
(f) Insertion of the pectus introducer in the presternal tunnel
(g) Blunt dissection and opening of the right intercostal space with the pectus introducer
(h) The left thorax is now opened and the curved clamp is moved up to the sternal edge
(i) Creation of a space with the clamp from within the thorax at the left sternal edge
(j) Removal of the clamp and introduction of the pectus bar in the left thorax
(k) Bar is then passed through the divided intercostal muscle close to the sternal edge
(l) The bar is flipped 180° and moved over the sternum to enter the right thorax
(m) Positioning of both the bar ends within the thoracic cavity
(n) End of the bar is secured to the ribs using PDS cords
(o) Subcutaneous and skin sutures

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Indications

Pectus carinatum that belong to one of the following categories:

- Pectus Carinatum affecting the mid part of the sternum
- Pectus Carinatum affecting the lower part of the sternum
- Symmetric protruding deformities
- Deformities with mild form of asymmetry

Preoperative Considerations

1. The procedure should be offered patients >14 years of age as a general rule.
2. Adult patients with stiff chest walls may not suited for this procedure.
3. Chest radiographs are taken to access the extent of the deformity.

A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_48

4. Lung function tests are performed to evaluate the pulmonary function.
5. Electrocardiograms to diagnose or rule out cardiac pathologies.
6. Video-stereo-raster investigations.
7. Chest perimeter plots to quantify the deformity at three different points: manubrium, sternum and xiphoid.
8. Photographic documentation of the deformity
9. Coagulation tests are performed to rule out disorders in bleeding disorders
10. Careful patient history is taken to rule out nickel allergy as stainless steel struts are used. If in doubt or an unclear history, a nickel allergy test should be performed.
11. Antibiotics are administered on the day of the procedure and are continued for a period of 5 days after the procedure. Cefuroxime is the choice of antibiotic.

Special Instruments

Besides the instruments used for general thoracic surgical procedures, the following instruments are additionally required for this procedure:

1. Pectus bar
2. Pectus bar bender (to shape the bar contours)
3. Pectus introducer
4. Pectus flipper

Surgical Technique

The patient is positioned supine with both arms abducted. Selection and bending of the *pectus bar* is also identical to that of the minimal access procedure of PE and is performed in the operating room prior to the thoracoplasty. A transverse left lateral chest wall skin incision is made traversing the mid-axillary line. With blunt preparation that intercostal region where the bar will enter the thorax is exposed. After that from the incision site a transverse subcutaneous tunnel running in front of the sternum is created with a long, slightly curved clamp reaching to the right sterno-costal margin. It is essential to create this pre-sternal tunnel because the pectus-bar will into the right thorax be placed through it. After that the *pectus introducer* is inserted into the pre-sternal tunnel through the skin incision. It will be then pushed forward to the level of the right sternocostal margin. At the right edge of the sternum the intercostal space is dissected bluntly with the tip of the *pectus introducer* thus preparing it for the insertion of the *pectus bar* into the right thoracic cavity.

The *pectus introducer* will be removed and the left thorax is opened at the incision site with the curved clamp, which is forwarded inside the thorax to the left sternal edge with the tip sliding on the inner surface of the thoracic wall. After reaching the left parasternal intercostal space, it will be divided with the clamp from inside thus preparing it for the *pectus bar* placement. The clamp is removed and the bent *pectus bar* with its convexity facing posteriorly is inserted into the left thoracic cavity through the left lateral intercostal space. The tip of the bar is going to be further pushed forward carefully with the tip sliding on the inner surface of the chest wall. The bar then is passed through the thoracic wall at the previously divided left intercostal space into the pre-sternal subcutaneous tunnel. At this point, the *pectus bar* will be flipped over 180° and further pushed into the right thoracic cavity through the previously prepared intercostal opening at the right sternocostal border. Placing the bar into the right thoracic cavity can be technically challenging. The *pectus bar* is brought into his final position and the left end of the strut is placed into the chest.

Prior to closure of the thorax a drain will be placed into the chest through a separate intercostal space under visual control. The end of the bar is fixed to the chest wall with 1/0 non-absorbable sutures tied around the neighbouring ribs. The thoracotomy is closed in the conventional way and air is aspirated from chest. The subcutaneous and the skin will be closed separately.

Procedure Overview (Figs. 48.1, 48.2, 48.3, 48.4, 48.5, and 48.6)

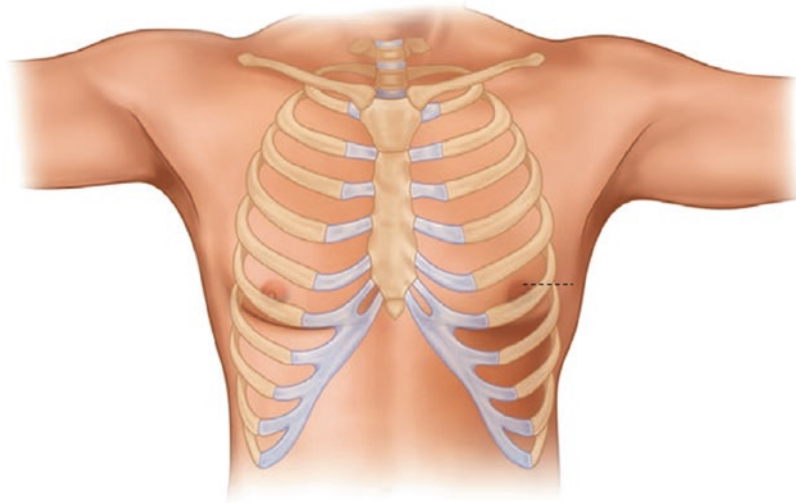

Fig. 48.1 A transverse left lateral chest wall skin incision is made traversing the mid-axillary line

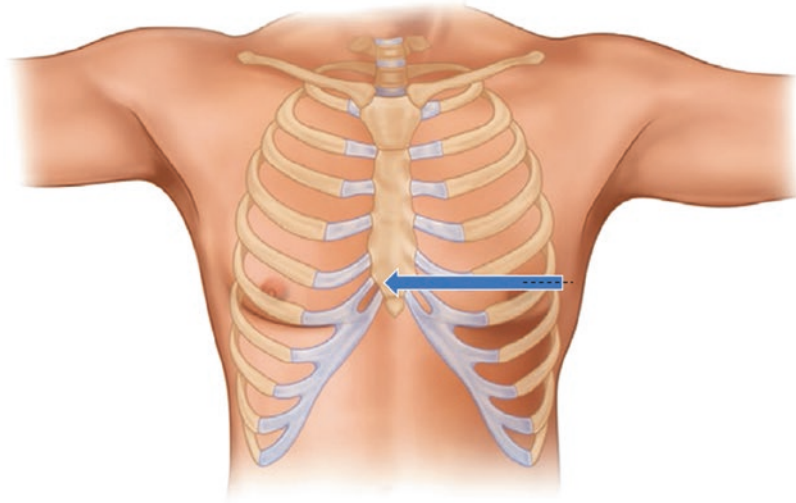

Fig. 48.2 A transverse subcutaneous tunnel running in front of the sternum is created with a long, slightly curved clamp

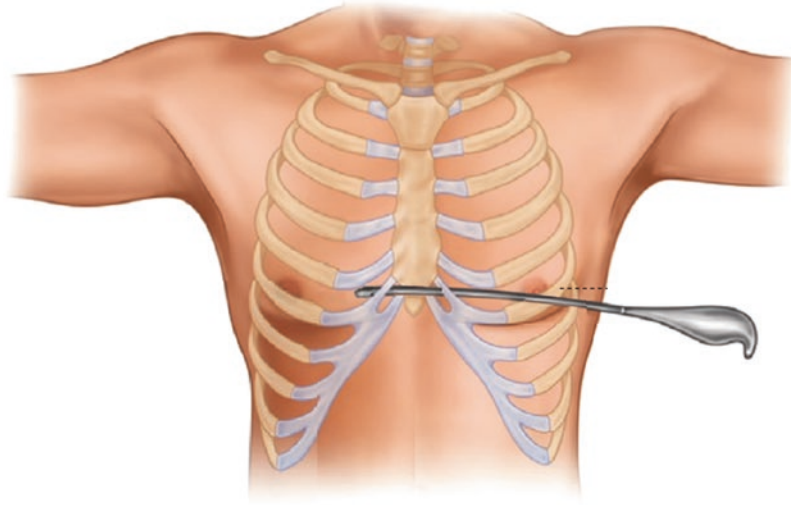

Fig. 48.3 The *pectus introducer* is inserted into the pre-sternal tunnel and pushed forward to the level of the right sternocostal margin. At the right edge of the sternum the intercostal space is dissected bluntly with the tip of the *pectus introducer*

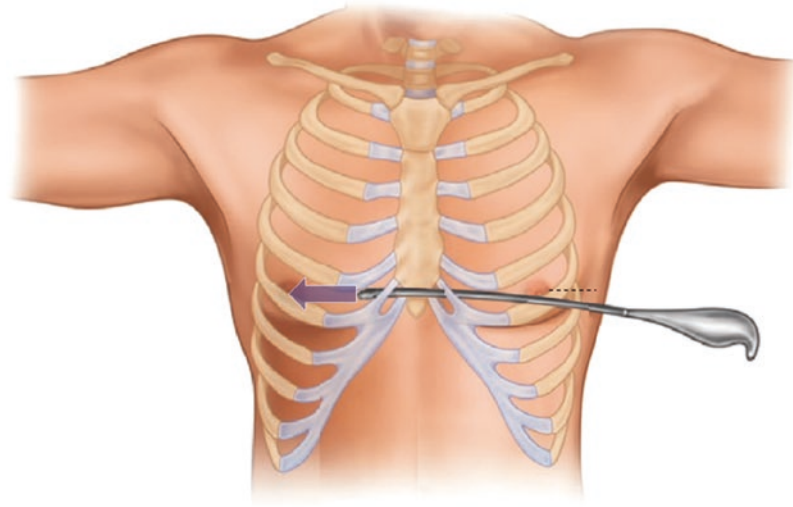

Fig. 48.4 The left thorax is opened at the incision site with the introducer, which is forwarded inside the thorax to the left sternal edge with the tip sliding on the inner surface of the thoracic wall to create an opening in the left parasternal intercostal space (*dark arrows- bar bending points*)

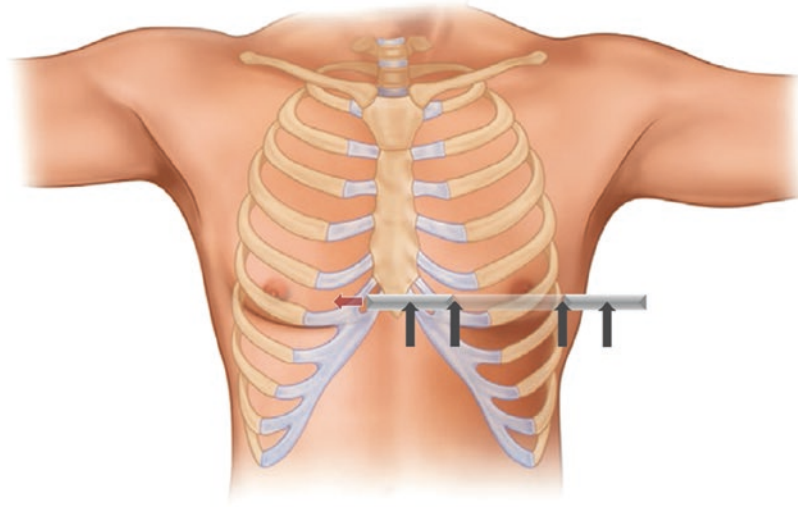

Fig. 48.5 The bent pectus bar with its convexity facing posteriorly is inserted into the left thoracic cavity and pushed forward with the tip sliding on the inner surface of the chest wall. The bar then is passed through the thoracic wall at the previously divided left intercostal space into the pre-sternal subcutaneous tunnel and then flipped 180°

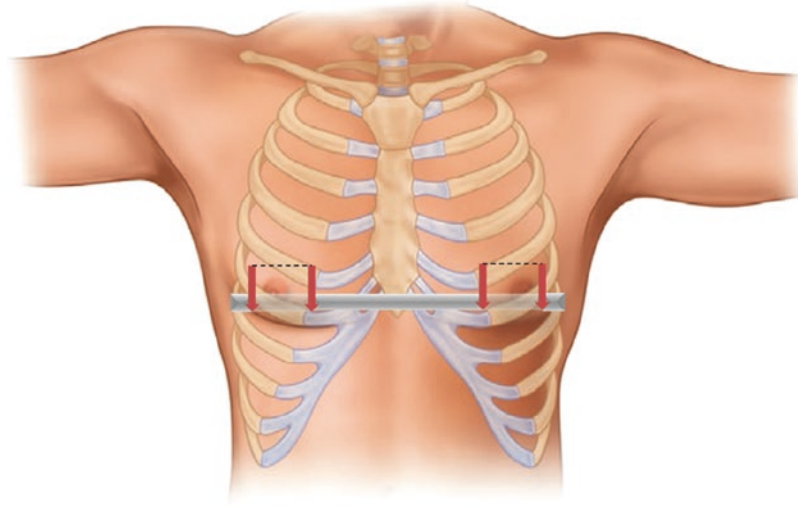

Fig. 48.6 Placing the bar into the right thoracic cavity can be challenging. The pectus bar is brought into his final position and the left end of the strut is placed outside the chest (*intrathoracic segment between 2 red arrows*)

Postoperative Management

Perioperative Prophylactic antibiotics were given preoperatively and administered for 5 days postoperatively. The patients were observed in the intensive care unit. In four patients there was no need for peridural anesthesia. The postoperative pain was managed with intravenous nalbuphin and was adequate and effective. There was no need for ventilatory support in any patient. During the first 2 postoperative days, suction of 8–10 cm H_2O was applied on the chest tubes. The patients were followed-up at regular intervals (3, 6, 12 and 15 months) after the procedure. The struts are removed after a period of 3 years.

Results

In the first patient operated with this procedure, insertion of the *pectus bar* was carried out by creating an additional anterior chest wall skin incision which aided in the exploration of those parasternal spaces where the end of the *pectus bar* was brought out from and pushed into the thoracic cavity. Thereafter, no anterior incisions were performed and only the bilateral thoracic exploration was done in the next set of two patients. Postulating that a bilateral fixation of the bar might be unnecessary, in the last two patients only a single-sided, left exploration was performed, which resulted in a reduction of the operating time.

There was one major and three moderate complications. In one patient an early *pectus bar* dislocation was encountered, which required *pectus bar* repositioning 3 weeks after the initial procedure. In the same patient, re-dislocation of the *pectus bar* occurred 2 weeks following the refixation. This patient opted for *pectus bar* removal and no further correction. The *pectus bar* in this patient was removed 6 weeks after the primary operation. In three patients, the tip of the bar perforated the intercostal space at the site of the bar insertion. In these patients, the tip of the *pectus bar* was repositioned and fixed of with wire sutures. After that no recurrence of this complication occurred. All patients were discharged on the 7–8th postoperative day. No limitation of the spontaneous physical activity was advised.

With regards to the aesthetic outcome, all ten patients were satisfied with the early postoperative result of the operation. In all patients a normalization of the Haller-index could be achieved (based on the postoperative chest x-ray measurements). In the four patients who were successfully treated, the *pectus bar* was removed between 2 and 3 years after surgery. During the follow-up after the removal of the *pectus bar*, neither clinical nor radiological signs of a recurrence of PC were observed.

Procedure Images (Figs. 48.7, 48.8, 48.9, 48.10, 48.11, 48.12, 48.13, 48.14, 48.15, 48.16, 48.17, 48.18, 48.19, 48.20, 48.21, 48.22, and 48.23)

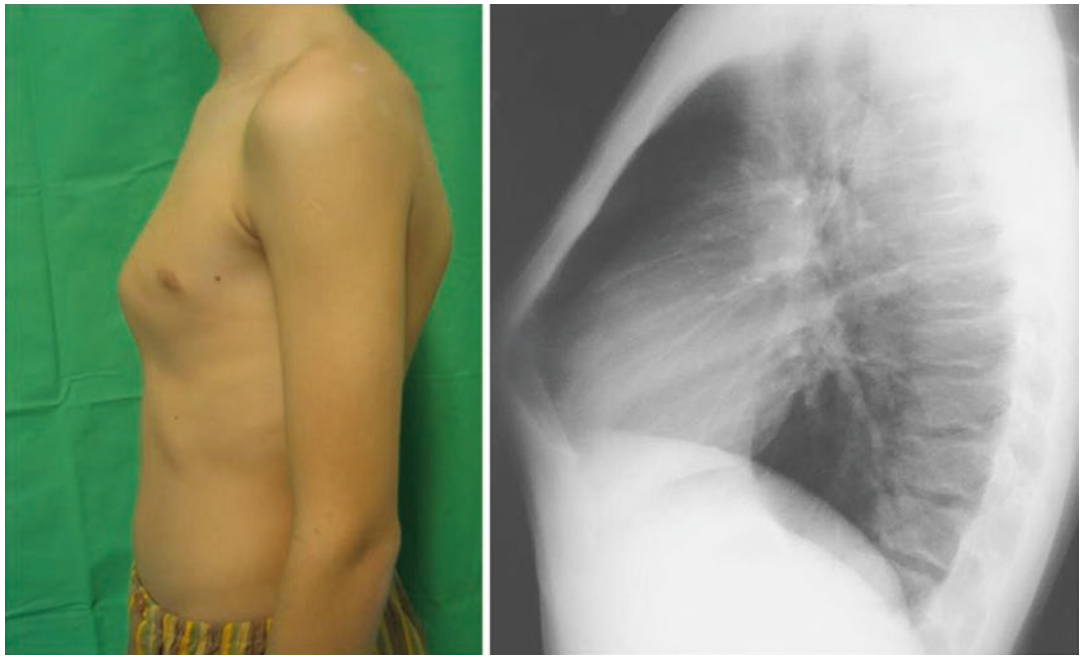

Fig. 48.7 Lateral view of a 14-year old patient with pectus carinatum (*left*) correlated with lateral chest films (*right*)

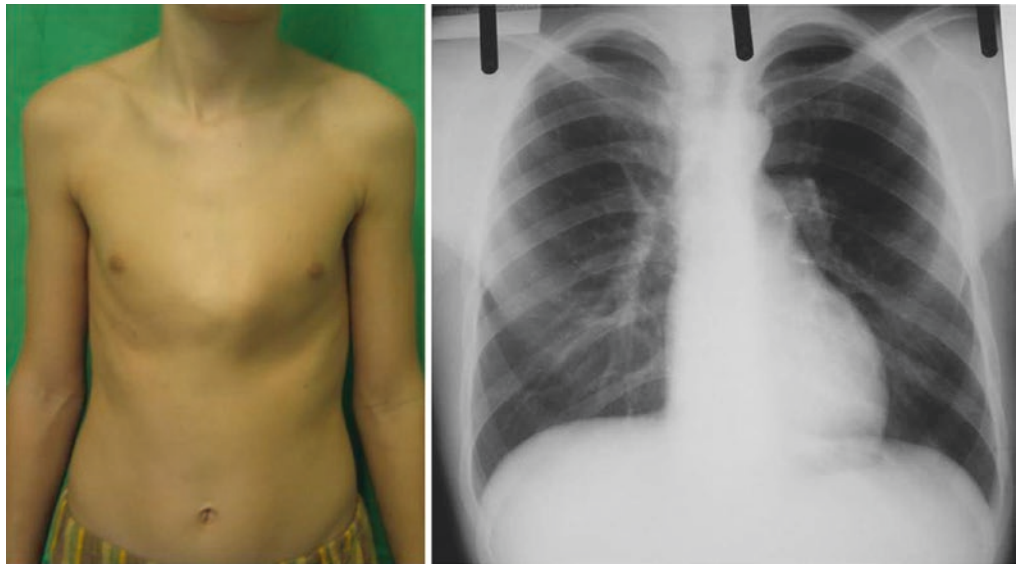

Fig. 48.8 Frontal view of a 14-year old patient with pectus carinatum (*left*) correlated with anterio-posterior chest films (*right*)

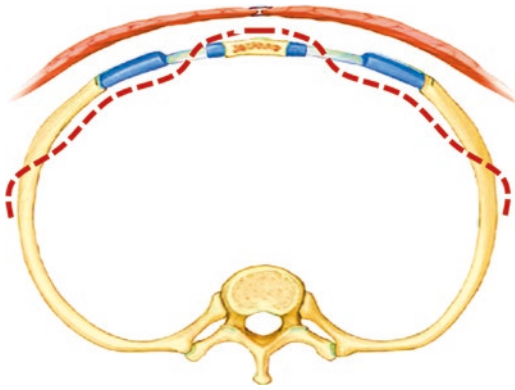

Fig. 48.9 Schematic overview showing the trajectory of the bar (*red line*) with relation to the sternum and ribs

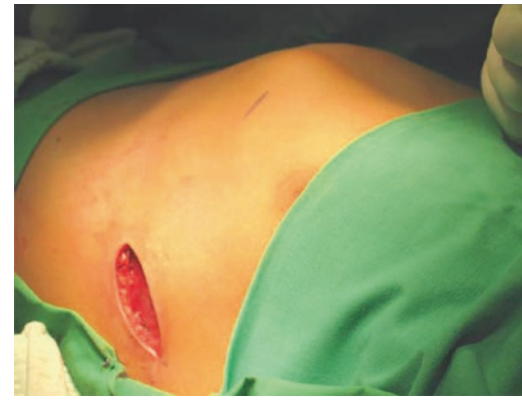

Fig. 48.12 The skin is incised and the subcutaneous tissue around the incision is mobilized

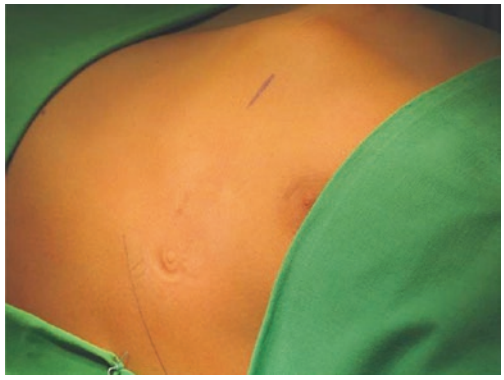

Fig. 48.10 Operative view of the incision site marked on the left side of the chest wall (*right*). left lateral chest wall skin incision is made traversing the mid-axillary line

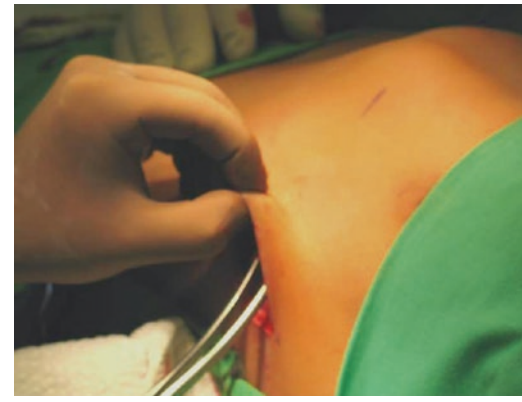

Fig. 48.13 A long curved clamp is used to dissect the subcutaneous tissue with the preparation directed towards the sternum (aided by a Pectus Introducer so as to reach the right sternocostal area and to create an opening in the intercostal muscles)

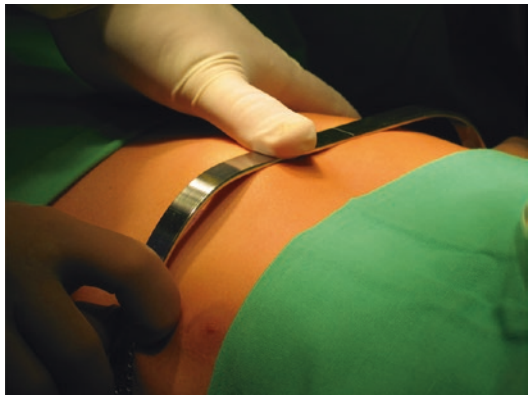

Fig. 48.11 A suitable Pectus bar is chosen depending on the dimensions of the thorax and is bent to size using a Bar Bender

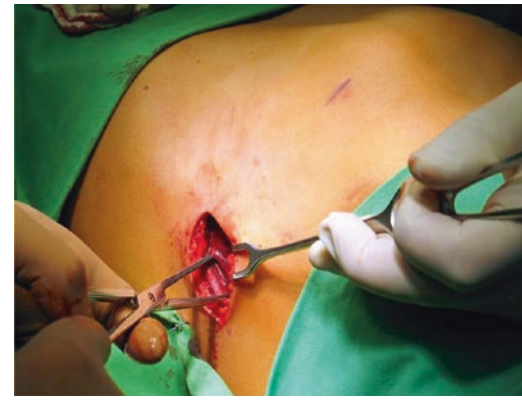

Fig. 48.14 Access is now gained into the thorax after splitting the intercostal muscles at the site of the incision

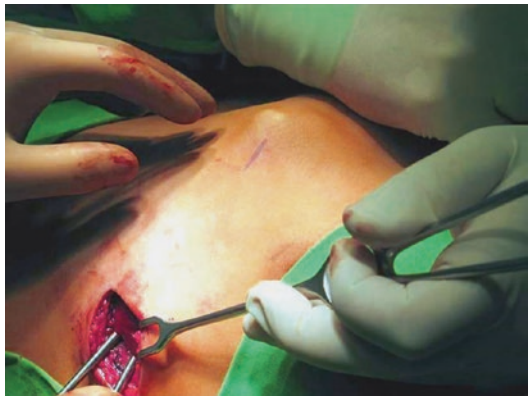

Fig. 48.15 A long curved clamp is introduced into the thorax and is moved along the chest wall until the left sternocostal area is reached. The intercostal muscle is dissected until the tips of the clamps are visible

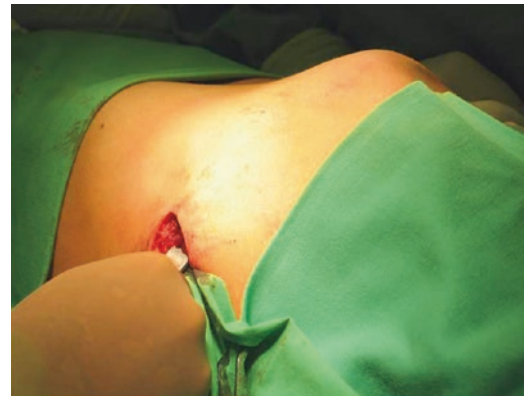

Fig. 48.17 Over the sternum the bar is flipped 180° with the tip pointing towards the right intercostal muscle gap created before

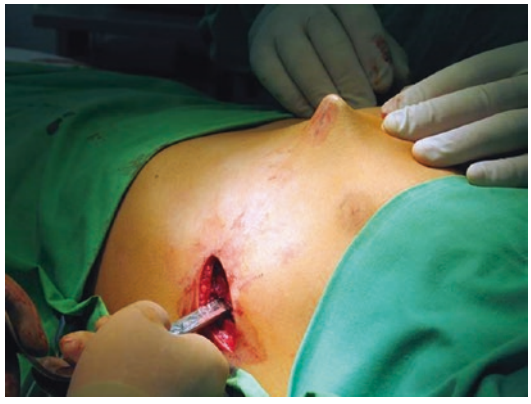

Fig. 48.16 The Pectus bar is then inserted through the thoracic cavity and pushed out through the intercostal muscle gap

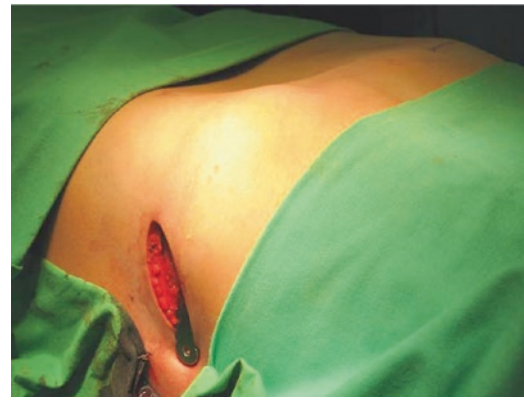

Fig. 48.18 Operative view of the Pectus bar after correct positioning on deformity

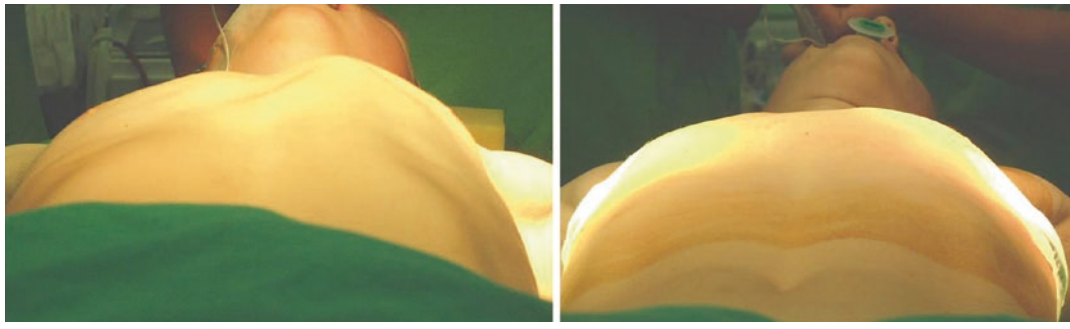

Fig. 48.19 View of the chest wall contours before (*top*) and after (*below*) completion of the procedure

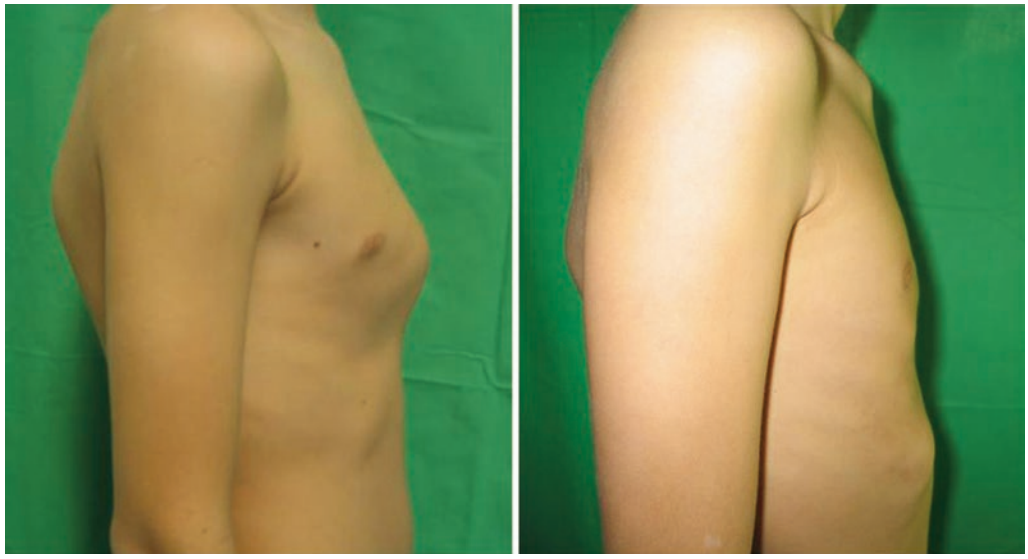

Fig. 48.20 Clinical image (lateral view) of a 14 year-old patients before (*left*) and after (*right*) correction

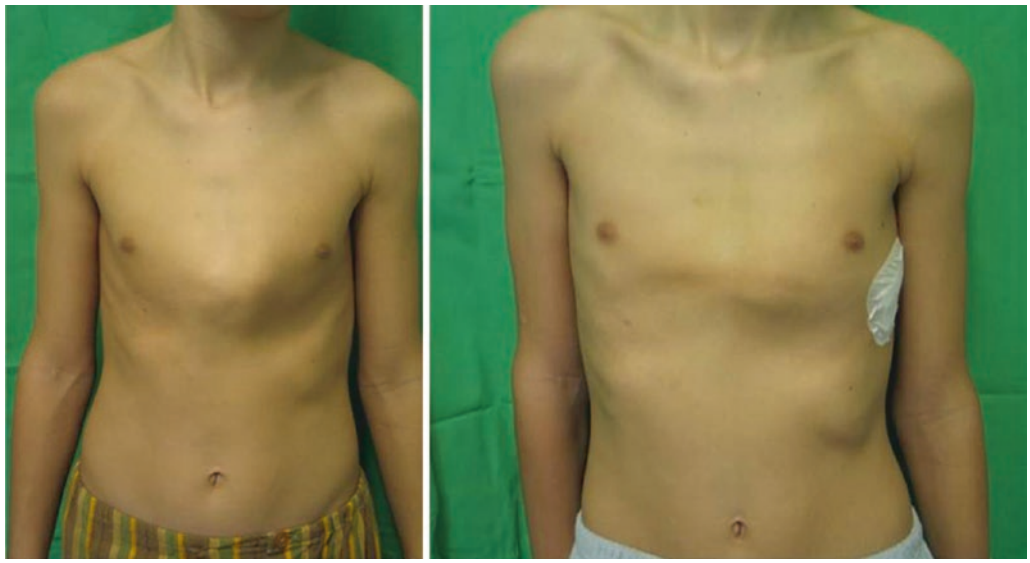

Fig. 48.21 Clinical image (frontal view) of a 14 year-old patients before (*left*) and after (*right*) correction

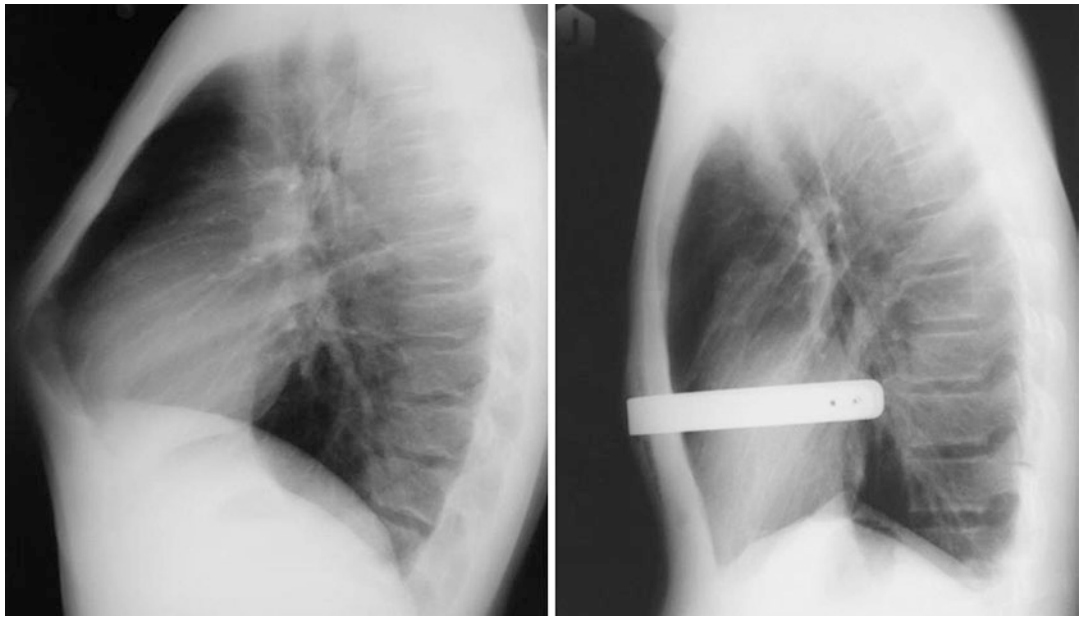

Fig. 48.22 Chest film (lateral view) of a 14 year-old patients before (*left*) and after (*right*) correction

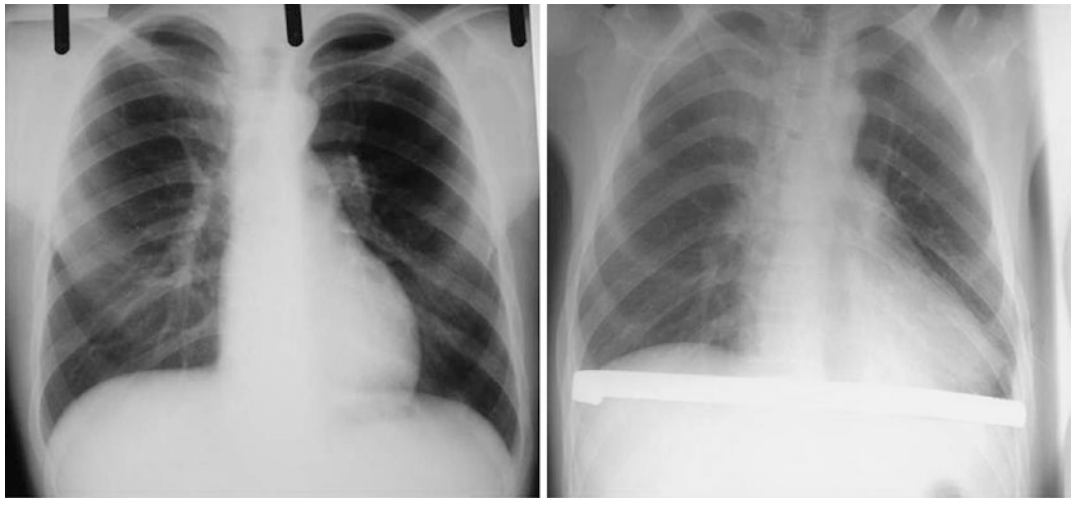

Fig. 48.23 Chest film (anterioposterior view) of a 14 year-old patients before (*left*) and after (*right*) correction

Suggested Reading

1. Abramson H, D'Agostino J, Wuscovi S. A 5-year experience with a minimally invasive technique for pectus carinatum repair. J Pediatr Surg. 2009;44: 118–23.
2. Hock A. Minimal access treatment of pectus carinatum: a preliminary report. Pediatr Surg Int. 2009;25: 337–42.
3. Kálmán A. Initial results with minimally invasive repair of pectus carinatum. J Thorac Cardiovasc Surg. 2009;138:434–8.

49 Thoracoscopic Approach for Surgical Repair of Pectus Carinatum

David Pérez, José Ramón Cano, and Luis López

Technical Highlights

Surgery for correction of Pectus Carinatum according to our technique is a fully endoscopic procedure that involved placement of major part of the pectus bar within the thorax with only the medial segment is positioned to compress the sternum and correct the deformity.

The main technical highlights of this procedure are:

(a) Bilateral perimammilar incisions are preferred in young male patients with deformity of the middle sternum and sub-mammary incisions in female patients. Protusions of the lower third of the sternum are better approached through incisions at anterior axillary lines.
(b) A transverse sternal osteotomy is performed in patients with a rigid deformity.
(c) Special surgical dissectors and optical tool are inserted paralell into the thoracic cavity through the right and left single port.
(d) Dissectors cross the thoracic midline over the sternum, thus creating a presternal tunnel.
(e) A short pectus bar is selected and be bent with a mild curvature only at the ends.
(f) The bar is inserted in its final position with the help of the guide-thread.
(g) Both lateral ends of the pectus bar are fixed to the ribs, the left pectus bar end in located within the thorax and the right end rests on the rib, so as to facilitate pectus bar removal.

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Indications

Symmetric Pectus Carinatum in non-rigid thorax (standard technique).
Symmetric Pectus Carinatum in rigid thorax (sternal osteostomy required).
Relatively asymmetric Pectus Carinatum (utilization of longer pectus bar)

General Preoperative Considerations

1. Chest films in anterior-posterior and lateral views
2. Electrocardiogram is also necessary for preoperative assessment
3. General laboratory tests including basic coagulation test.

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4. Since Pectus Carinatum is generally not associated with dysfunction of the intra-thoracic organs, respiratory or cardiologic functional studies are not required unless conditions affecting such systems have been detected during history taking, the physical examination or the basic preoperative investigations.
5. Complex quantitative estimations of deformity degree, such as volumetric or planimetric studies, are not considered necessary or efficient, since the results of such studies do not influence the technique choice.
6. Photographic documentation of the deformity before and after correction.
7. Procedures are conducted under general anesthesia, preferentially with selective intubation with double lumen tube. Orotracheal intubation can be used; however, such a procedure requires low ventilation volume or repeated apnea, which may interfere with the surgery.
8. Postoperative epidural analgesia is recommended for a minimum of 4 days postoperatively. Please note that however, when sternal osteotomy is performed, this notably reduces postoperative pain.
9. Wide-spectrum antibiotic prophylaxis, with preference for Cephalosporin. Antibiotics should be administered intravenously 20 min before intervention and administered until the 5th postoperative day.
10. Antithrombotic prophylaxis with subcutaneous low-molecular-weight-heparin should be applied.

Special Instruments

The specific tools necessary for this procedure include instruments for Minimal Access Repair of Pectus Excavatum (MARPE) and instruments for general thoracic surgery.

Specific instruments needed for this technique:

1. Surgical tool set for MARPE, including pectus bar benders and pectus introducers (Biomet Microfixation Pectus Bar, Jacksonville, Florida).
2. Scopes of 30 and 5 or 10 mm diameter.
3. Flexible endoscopic ports (Flexipath; Ethicon Endo-Surgery, Blue Ash, Ohio).
4. Osteotome and/or Lebsche knife (Codman and Shurtleff, Inc, Bridgewater, MA, USA).
5. Multifilament steel cable system and Dall-Milles cable grip (Stryker Orthopaedics, Mahwah, NJ, USA).

Surgical Technique

The patient is placed in the supine position with a small interscapular pillow and with the arms in abduction. The first important step is to estimate thorax rigidity by manual depression of the in order to determine if a sternal osteotomy is indicated; this is recommended in patients whose sternum cannot be easily depressed. The distance between both nipples is measured and a pectus bar is selected that is 4 cm longer than that distance. The pectus bar is then bent with a mild curvature only at the ends.

Incisions of 2–3 cm length are performed on the lateral margins of both nipples in male patients or on the lateral portion of the submammary creases in female patients.

The mammary fat and pectoral muscles are then displaced and the flexible ports are inserted. The flexible ports allow for parallel insertion of the scope and the instruments and cause less postoperative pain than rigid ports. Both pleural cavities are then explored with the scope.

In case sternal osteotomy is needed, it has to be done before placement of the pectus bar. The sternal osteotomy is performed under endoscopic monitoring of the right mammary vessels by placing the scope in the right thorax. After initial breaching of the periosteum, the osteotome is withdrawn and the procedure continued with a Lebsche knife and a hammer until bone resistance decreases.

The pectus introducer is then inserted parallel to the optical scope, through the right areolar or

submammary incision and moved forward towards the mediastinum until the right internal thoracic vessels are visible. The end of the pectus introducer is directed upwards, from the margin of the internal thoracic vessels, and the pectus introducer is pushed out into the pre-sternal space. In this way, the bar will not compress the mammary vessels. The pectus introducer is then rotated and with its end oriented towards the left hemi-thorax. It is then moved forwards in a plane that is closely adjacent to the outer sternal surface until the opposite intercostal space is reached, thus completing the pre-sternal tunnel.

Before entering the left pleural space, the anaesthetist has to facilitate to collapse the left lung with ventilation continuing with the right lung. This is necessary to allow the scope to be moved into the right hemi-thorax to monitor the maneuver. Manual depression of the sternum remarkably facilitates inserting the pectus introducer into the left thorax at a point lateral to the left internal thoracic vessels. Once the pectus introducer enters the left thorax, it is moved forward towards the left port; the end is rotated towards the port and the movement continued until the dissector leaves the thorax.

A cotton tape is tied to the end of the pectus introducer and retrieved through the incision. A second larger pectus introducer is then inserted and moved forward, with the help of the cotton tape, with the aim of widening the path. Finally, the bent pectus bar is inserted. The Pectus bar is secured at both ends with to the rib using the multifilament Dall-Milles steel cable system. The wire tensor of this system allows application of controlled gradual tension until the desired point (rib notching).

The surgical site is closed in layers by using 4–0 absorbable suture in the muscle and subcutaneous layers and 4–0 non-absorbable monofilament single stitches in the skin layer. During the closure, the anaesthetist will utilize high ventilation volume to reduce residual pneumothorax.

Procedure Overview (Figs. 49.1, 49.2, 49.3, 49.4, 49.5, and 49.6)

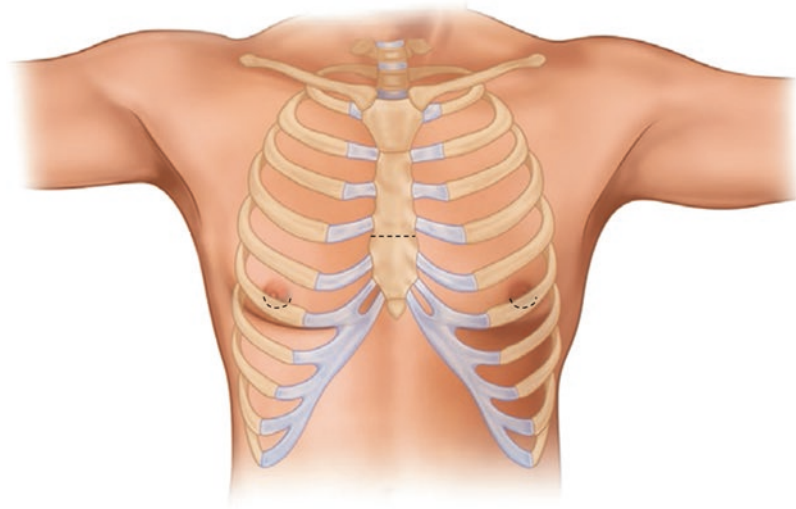

Fig. 49.1 Peri-mammilary incisions in male patients or in the submammary creases in female patients and an additional 2 cm incision to perform the sternotomy are made

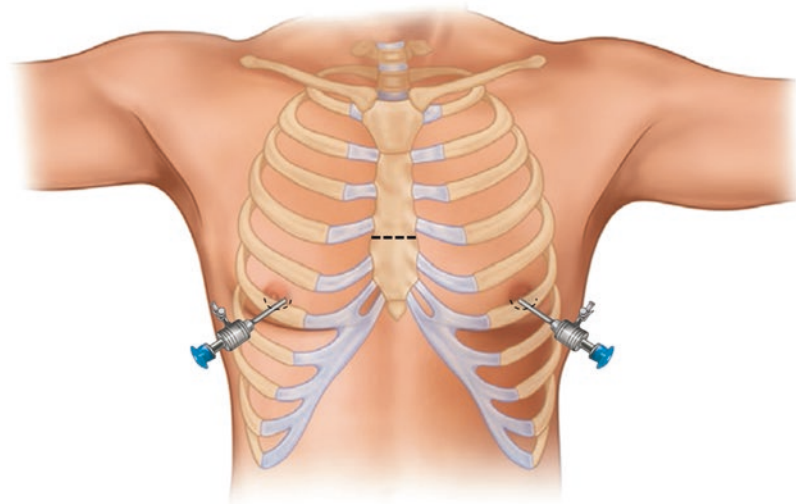

Fig. 49.2 Bilateral insertion of ports after which a sternotomy is performed under endoscopic vision of the right mammary vessels after which a dissector is used to create a working space

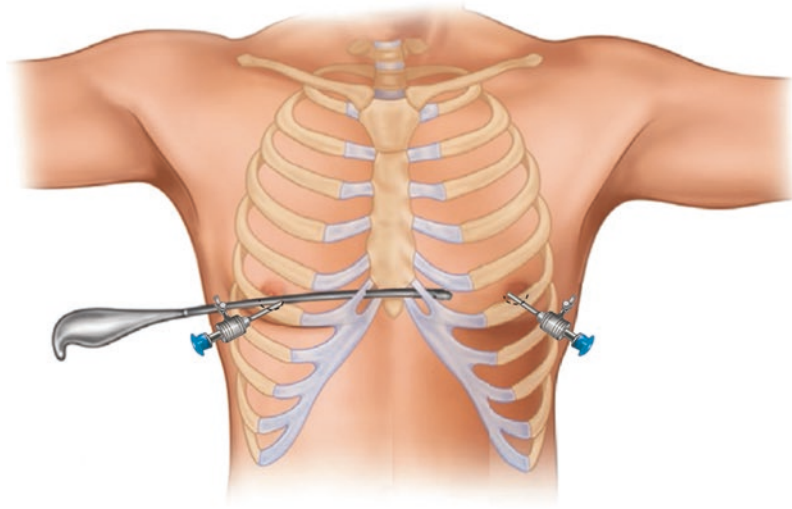

Fig. 49.3 The pectus introducer is then inserted parallel to the scope, through the right areolar or submammary incision and moved forward towards the mediastinum until the right internal thoracic vessels are visible. The end of the pectus introducer is directed upwards, from the margin of the internal thoracic vessels, and the pectus introducer is pushed out into the pre-sternal space. In this way, the bar will not compress the mammary vessels. The pectus introducer is then rotated and with its end oriented towards the left hemi-thorax

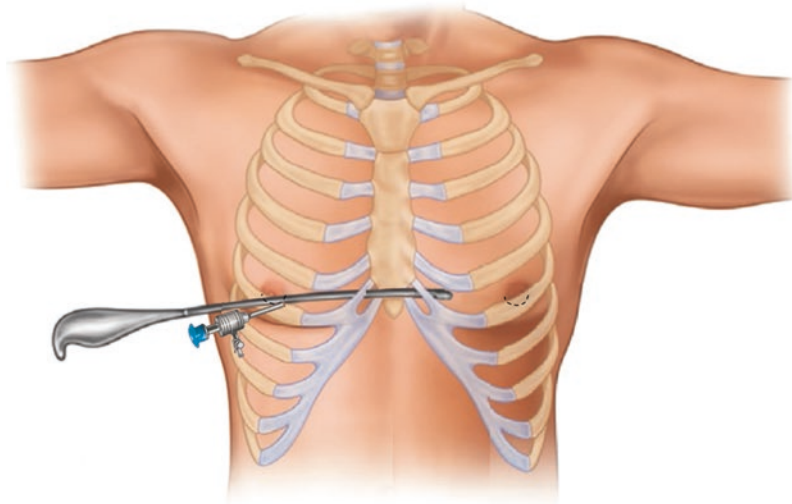

Fig. 49.4 A larger introducer is used to extend the tunnel after which a cotton band is passed through the introducer tip to help guide the bar into its final position

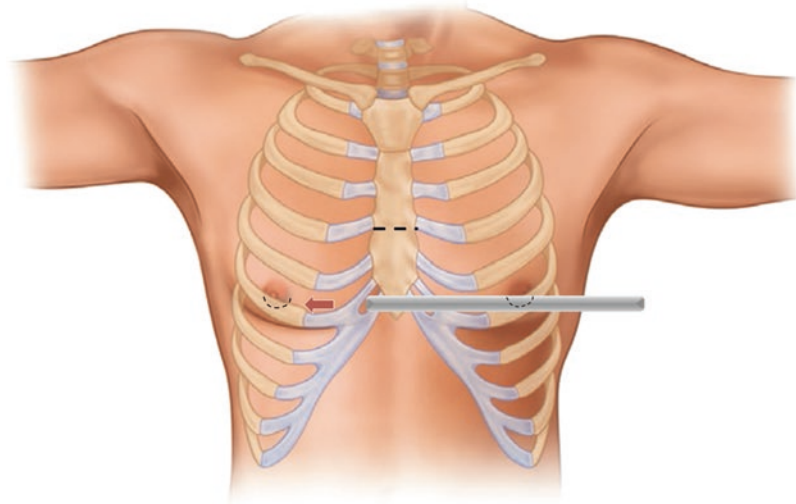

Fig. 49.5 The pectus bar is set into the desired position

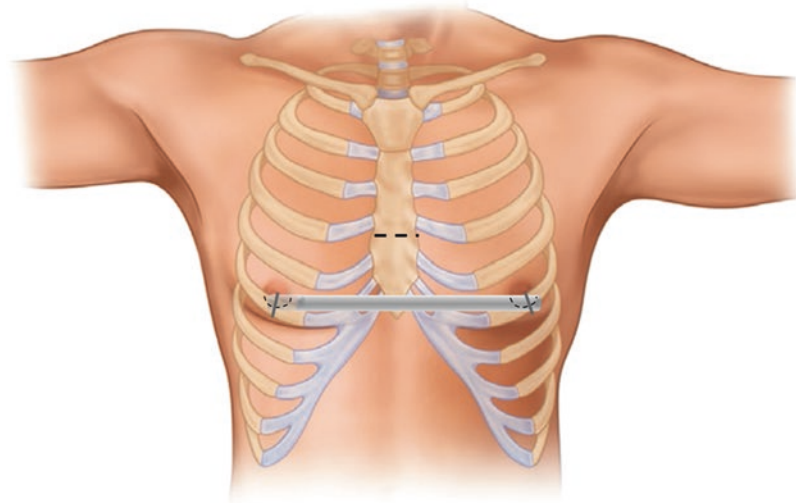

Fig. 49.6 Both ends of the pectus bar are attached to the ribs with multifilament wires using a Dall-Milles steel cable system tied around the rib

Postoperative Management

Patients are extubated in the operation room and transferred to the recovery room for the first few hours, where they are monitored, the analgesic treatment is adjusted and an immediate post-intervention radiological control is carried out. The epidural and intravenous analgesic treatment will be maintained for 72 h and then substituted by an oral medications according to the analgesia management protocol (WHO analgesic scale), generally alternating between two different non-steroidal anti-inflammatory medications. Please note that however, when sternal osteotomy is performed, this notably reduces postoperative pain.

Wide-spectrum antibiotic are administered, with preference for Cephalosporin. Antibiotics should be administered intravenously 20 min before intervention and administered until the 5th postoperative day. Respiratory physiotherapy are commenced 24 h after the procedure. Mobilization is encouraged 48 h after surgery. Patients may be discharged on the fifth day after surgery.

Relative rest is recommended for 2 weeks. Patients should avoid sport activities in general for 6 months and competitive or contact sports for 1 year. Follow-up visits should take place 3, 6, 12 and 24 months after surgery.

In general, the pectus bar removal is recommended after 3 years; however, it can be removed earlier in patients who underwent transverse sternal osteotomy since, once the sternal fracture has consolidated in the new position, the pectus bar is no longer needed.

Procedure Related Images

(Figs. 49.7, 49.8, 49.9, 49.10, 49.11, 49.12, 49.13, 49.14, 49.15, 49.16, 49.17, 49.18, 49.19, and 49.20)

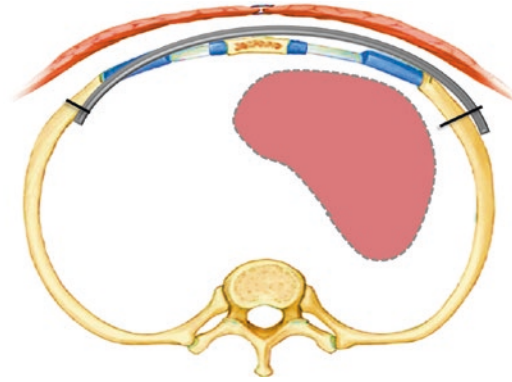

Fig. 49.7 Schematic diagram demonstrating the positioning of the pectus bar

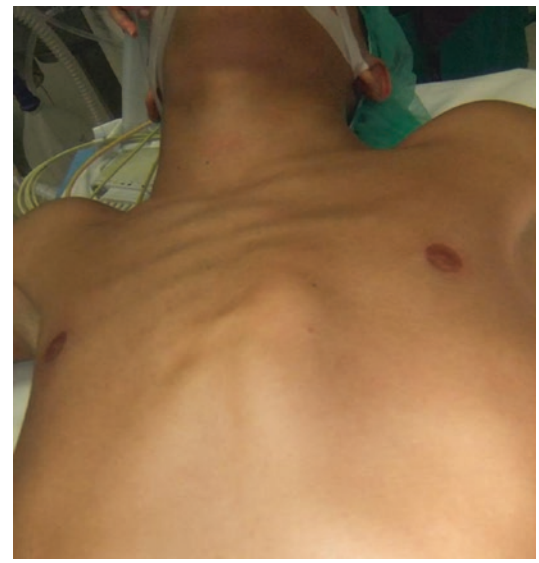

Fig. 49.8 Incisions are performed on the lateral margins of both nipple demarcation in male patients or on the lateral portion of the submammary creases in female patients

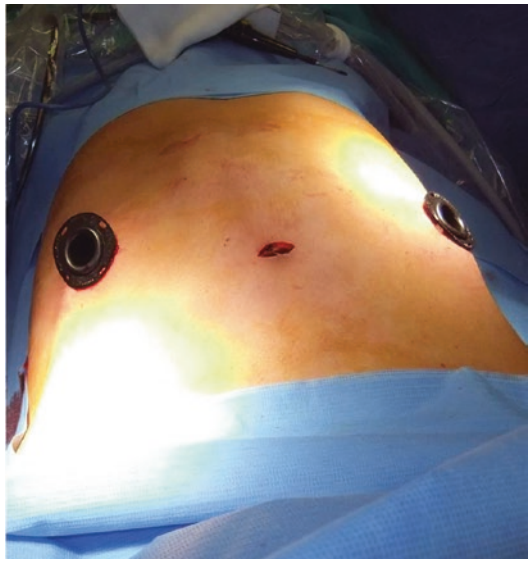

Fig. 49.9 In case sternal osteotomy is necessary, an additional 2 cm incision is made at a two-finger-width distance above the planned path of the Pectus bar

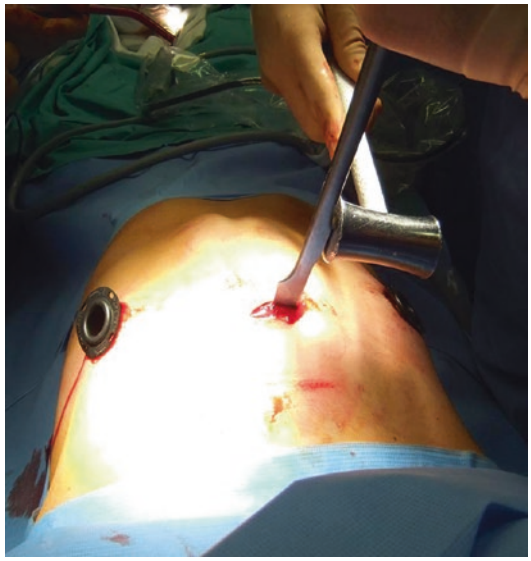

Fig. 49.10 The sternal osteotomy is performed under endoscopic vision of the right mammary vessels

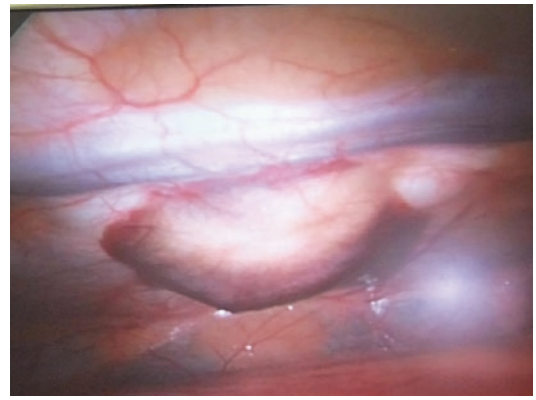

Fig. 49.11 A small mediastinal hematoma can be visualized on endoscopic view after breach of the posterior sternal wall during osteotomy

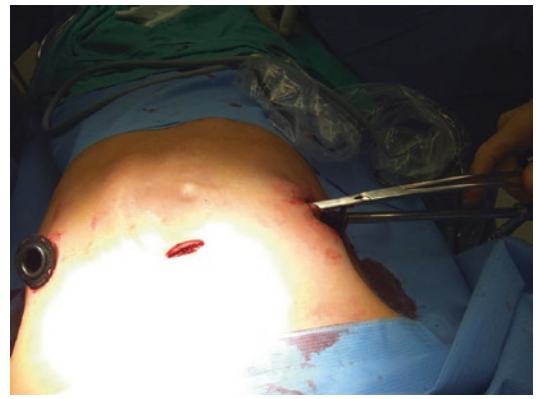

Fig. 49.12 The pectus introducer inserted parallel with the scope, is moved forward until the right internal thoracic vessels are reached and is forced out into the pre-sternal space in order to create the pre-sternal tunnel

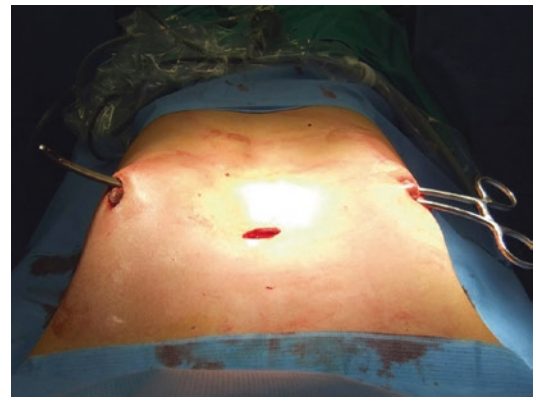

Fig. 49.13 The dissector is moved forward and exits the chest cavity at the left working port

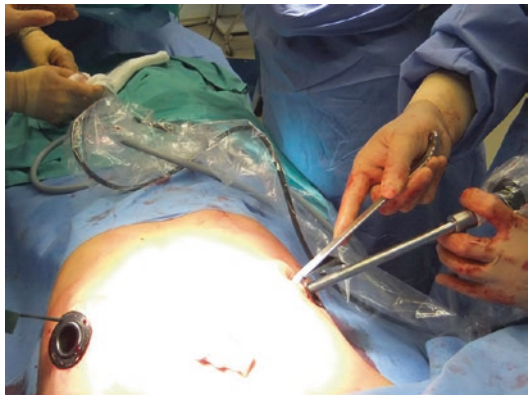

Fig. 49.14 A cotton tape tied to the end of the dissector preserves the created trajectory and facilitates the passage of a larger pectus introducer and Pectus bar

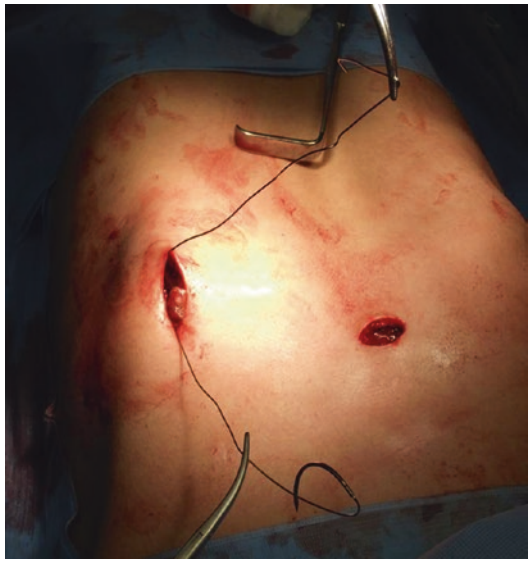

Fig. 49.15 Both ends of the bar are fixed with metal wires

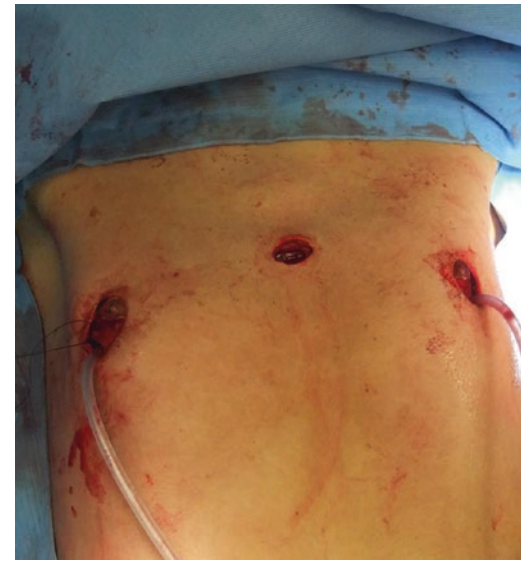

Fig. 49.16 During the closure tubes are used in order to avoid residual pneumothorax

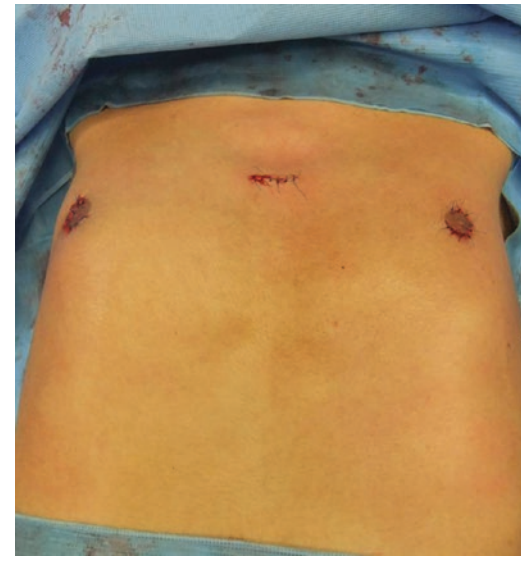

Fig. 49.17 The surgical site is closed in layers; remodelling of the chest wall is adequate and scars will become barely noticeable

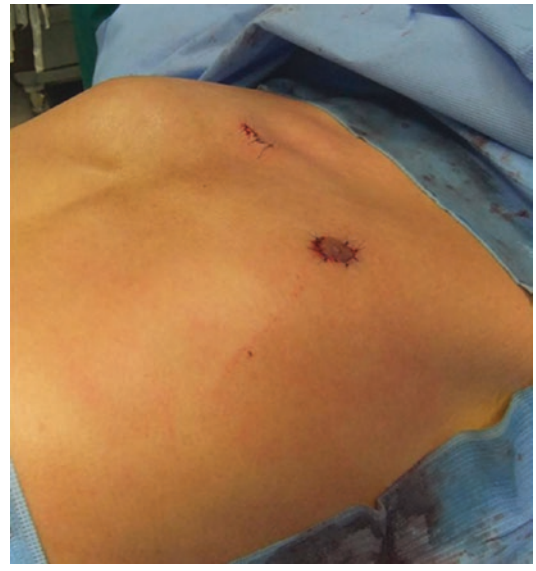

Fig. 49.18 Another view of closure of the incisions

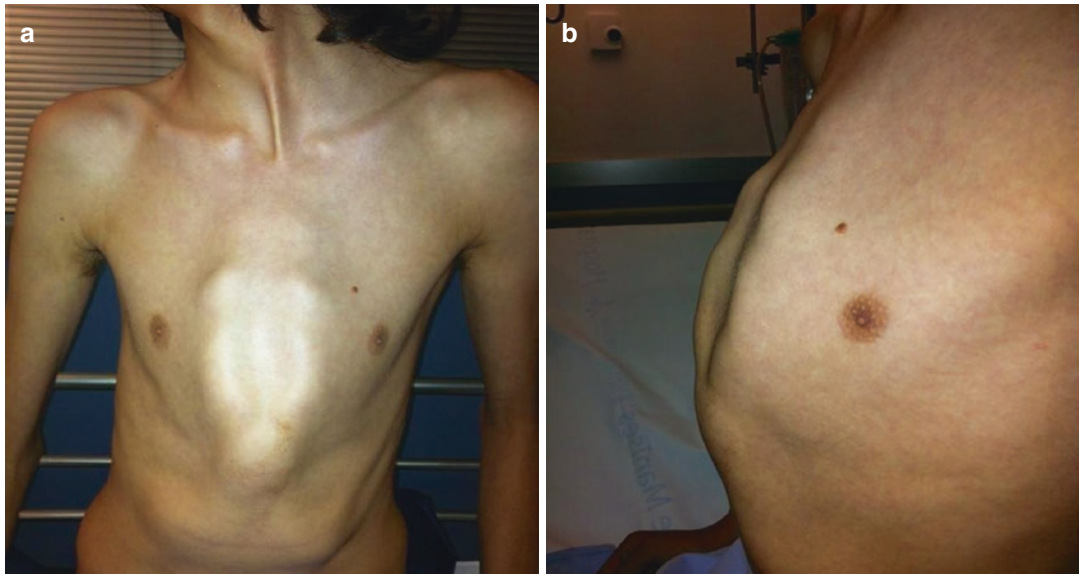

Fig. 49.19 Case presentation of patient with severe form of pectus carinatum (**a**: front view) and (**b**: lateral view)

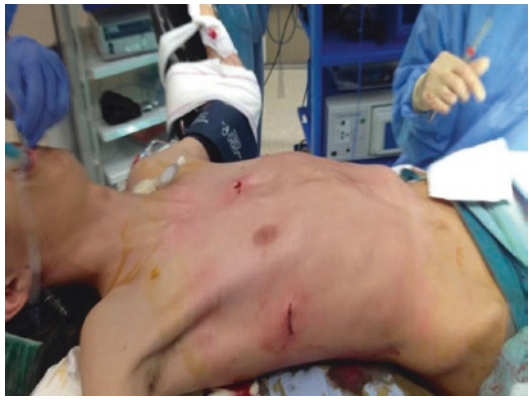

Fig. 49.20 This endoscopic surgical procedure is offered to patients with large Pectus Carinatum deformities as demonstrated in Image 13. A longer bar has to be used for which incisions are located in the axillary lines. The procedure provides of excellent results; besides correcting the anterior deformity, this bar will also expand the ribcage by re-shaping the lateral thoracic walls

Suggested Reading

1. Abramson H. A minimally invasive technique to repair pectus carinatum. Preliminary report. Arch Bronconeumol. 2005;41:349–51.
2. Kalman A. Initial results with minimally invasive repair of pectus carinatum. J Thorac Cardiovasc Surg. 2009;138:434–8.
3. Perez D, Cano JR, Quevedo S, Lopez L. New minimally invasive technique for correction of pectus carinatum. Eur J Cardiothorac Surg. 2011;39:271–3.
4. Pérez D, Cano JR, Quevedo S, López L. Videothoracoscopic repair of pectus excavatum with sternal transection for adult patients with non-elastic deformity. J Thorac Cardiovasc Surg. 2011; 142:942–3.

Minimal Access Repair of Pectus Carinatum: Istanbul Technique

50

Mustafa Yuksel

Technical Highlights

The technical highlights of the Minimal Access Repair of Pectus Carinatum (MARPC) using the Istanbul technique are as follows:

- A horizontal line is drawn through the region with the highest degree of protrusion on the anterior chest wall.
- Two bilateral transverse incisions are are made at the midaxillary line.
- The periosteum of two adjacent ribs on both sides for the placement of the stabilizers is incised.
- The ribs are encircled a Doyen rib periosteal elevator or a hook and then with a suction catheter to serve as a sheath for the steel wires.
- The stabilizers for the pectus bar are placed perpendicularly on the ribs and secured with the wires on both sides.
- The pectus bar of appropriate size is bent into a convex configuration.
- Using clamps, bilateral subcutaneous tunnels are created toward the sternum
- A polyvinyl chloride tube with a trocar is passed from one incision to the other, posterior to the pectoralis major, then through the pectoralis major close to its origin near the sternum.
- The trocar is removed from the lumen of the tube, and the bar is inserted into it with the concavity facing posteriorly
- Compressing the bar over the sternum, both edges are placed into the stabilizers and secured with metal screws on both sides.
- The wounds are closed in layers with absorbable sutures.

Surgical Indication

- Pectus carinatum

The best age range to perform MARPC is between the age of 12 and 18 years, as this is the rapid growth phase of puberty when the deformity becomes more prominent and the chest wall is still flexible.

Preoperative Considerations

- Chest films with postero-anterior and lateral view
- Pulmonary function tests

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- Electrocardiography and echocardiography examinations
- Routine laboratory blood tests and coagulation tests
- Computed tomography is done for all patients
- A compression test to estimate the rigidity of the chest wall is performed. If the sternum poses high rigidy to compression, open surgery is preferred for the correction of PC. Recently, our group has developed a device to measure sternal compression pressures to standardize our treatments of choice: open surgery, MARPC or orthotic bracing. If the measured value is less than 10 kg orthotic bracing is preferred, between 10 and 25 kg MARPC is offered and if more than 25 kg the open surgical option is indicated.
- The pectus bar and stabilizing system for MARPC is a nickel alloy, hence patients for nickel allergy should be cleared otherwise dermatological allergey tests are performed.
- Patient satisfaction is done through questionnaires preoperatively and in the postoperative 6th month [1].

Surgical Technique

The patient is placed in the supine position with the arms abducted at 90°. General anesthesia with single-lumen tube is preferred. A horizontal line is drawn through the region with the highest degree of protrusion on the anterior chest wall to determine the site for the incisions. Two 2.5 cm transverse incisions are made on both sides at the mid-axillary line. After preparation of the muscle fibers, 1 cm incisions are made into the periosteum of two adjacent ribs for the placement of the stabilizers. While the momentary cesation of ventilation line, the ribs are circumscribed subperiostally with a Doyen rib periosteal elevator or a hook and then with a suction catheter to serve as a sheath for the placement of multi-ply steel wires. The cesation of ventillation is to prevent the breach of the pleura during this maneuver. Following the placement of the multi-ply steel wires, the suction-catheter sheath is removed and the ventilation is commenced. Specially designed stabilizers plates with grooves and screws for the pectus bar are placed perpendicularly on the ribs and secured with the wires on both sides. The bar of appropriate size is then bent into the desired convex configuration. Using clamps, subcutaneous tunnels toward the sternum are created on both sides and a polyvinyl chloride tube with a trocar is passed from one incision to the other, posterior to the pectoralis major, then through the pectoralis major close to its origin near the sternum. The trocar is removed from the lumen of the tube, and the bar is inserted into it with the concavity facing the sternum after which the tube is removed. The bar is then pressed over the sternum and to push the edges into the stabilizers at appropriate levels. These are then secured with metal screws on both sides. The wounds are closed in layers with absorbable sutures.

Postoperative Overview (Figs. 50.1, 50.2, 50.3, 50.4, and 50.5)

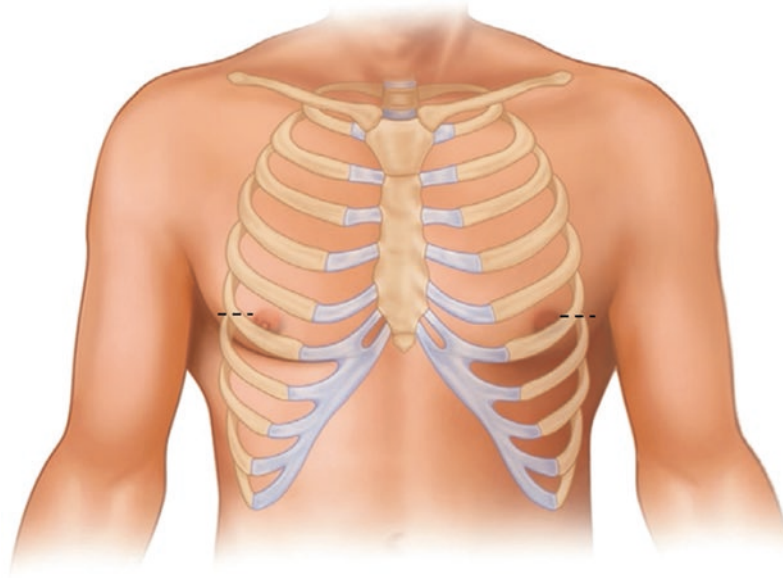

Fig. 50.1 A horizontal line is drawn through the region with the highest degree of protrusion on the anterior chest wall. Two bilateral transverse incisions are are made at the midaxillary line

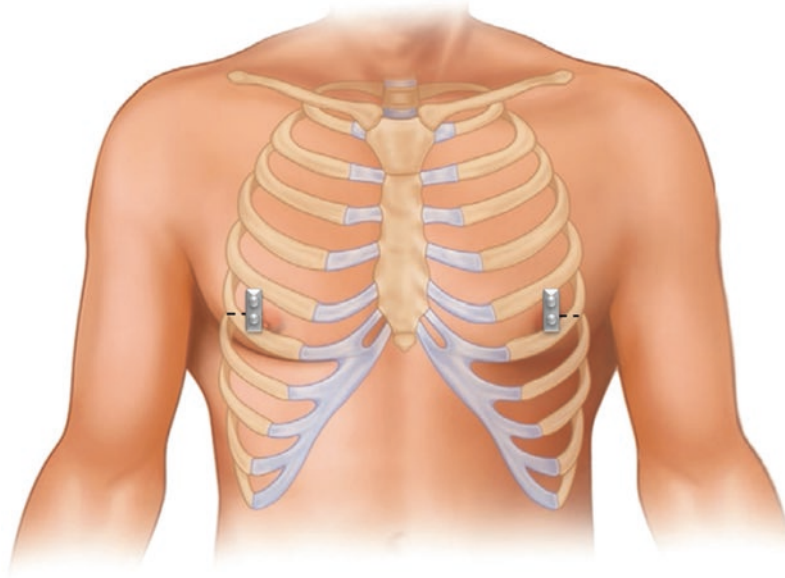

Fig. 50.2 The periosteum of two adjacent ribs on both sides for the placement of the stabilizers is incised. The ribs are encircled by a periosteal elevator or a hook and a suction catheter is used to serve as a sheath for the steel wires. The stabilizers plates are placed perpendicular on the ribs and secured with the wires

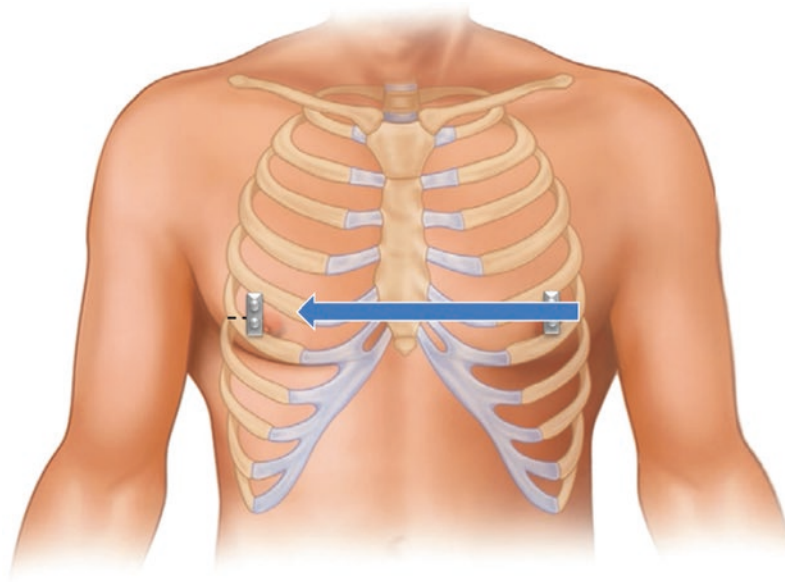

Fig. 50.3 Using clamps, bilateral submuscular tunnels are created toward the sternum

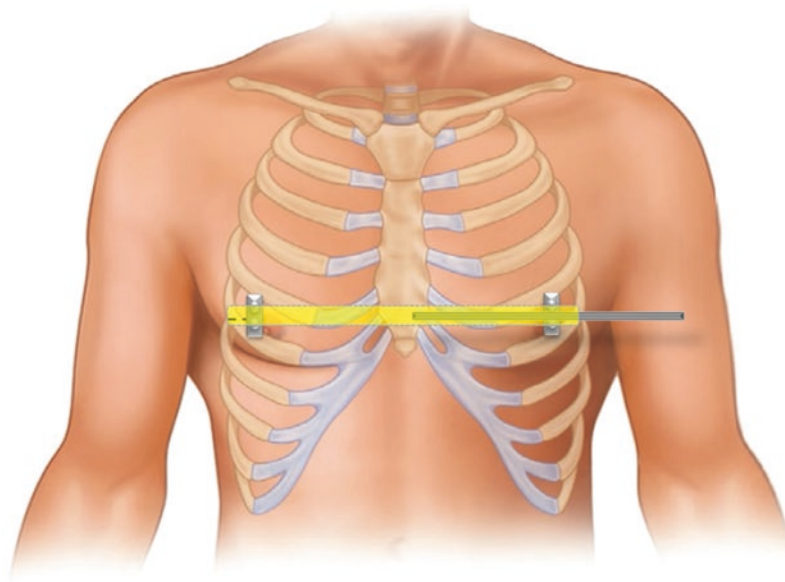

Fig. 50.4 A polyvinyl chloride tube with a trocar is passed from one incision to the other, posterior to the pectoralis major, then through the pectoralis major close to its origin near the sternum

Postoperative Management

A postoperative chest film is obtained routinely, to view the position of the pectus bar in place and to access the thorax for pneumothorax. No intervention is done for residual pneumothorax of less than 10 % of the lung. If necessary patients are followed up with chest films for an increase in the pneumothorax rate to access the need for a chest tube. Pain control is maintained with patient-controlled analgesia (PCA) using epidural fentanyl and bupivacaine during the postoperative 48 h, followed by oral tramadol and etodolac. Patients are mobilized as early as possible and are sent home within the first 5 days in general. Patients are also advised to return

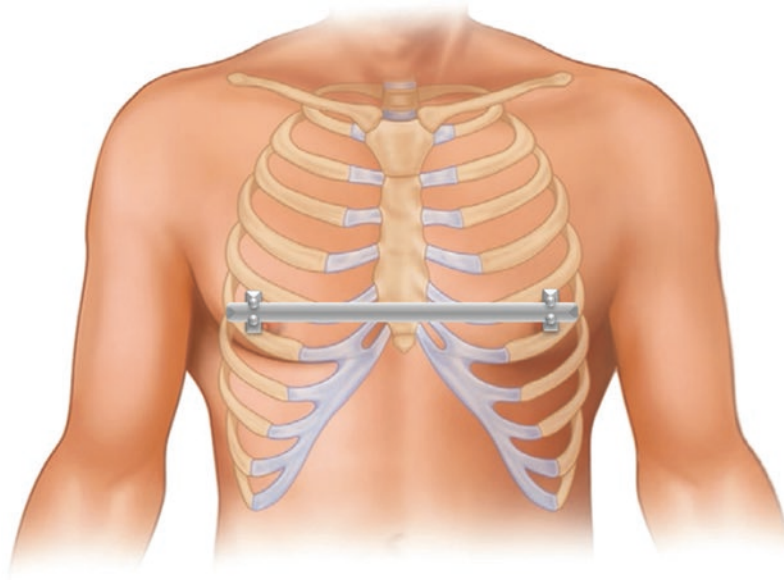

Fig. 50.5 The bar is inserted into it with the concavity facing posteriorly. Compressing the bar over the sternum, both edges are placed into the stabilizers and secured with metal screws on both sides

early to regular activities, however advised not to commence with active sports before the 1 month. Return to all forms of sports is allowed after the 3rd month. All pectus bars are left for maximum 2 years after which they are removed under general anesthesia.

Results

As MARPC is not an intrathoracic procedure less severe complications then with minimal access repair of pectus excavatum (MARPE) are expected. The most common complications encountered in the perioperative and postoperative period are pneumothorax, wound infection, seroma, cutaneous adherence on the bar, breakage of the steel wires, metal allergy, overcorrection and insufficient correction [2].

Long term results with MARPC are as satisfying for repair of chondrogladiolar type of pectus carinatum. Asymmetric deformities can also benefit from this technique, however as chondromanubrial type of pectus carinatum is commonly rigid and not well suited for the placement of a presternal pectus bar, open repairs are preferred for this type of deformity [3]. Earlier removal of pectus bars even before the end of the second year may cause recurrence of the deformity. On the other hand leaving the pectus bar for an extended period of time may lead to the formation of an iatrogenic pectus excvatum, or rigid thorax especially during puberty.

At our series on 126 MARPC, there were 122 males with a median age was 16.5 years (range 10–33). The deformity was symmetric in 88 patients whereas it was asymmetric in 38. Following the first three patients who were operated using conventional MARPE pectus bars, another bar and stabilizing system was developed by our group and modified subsequently. These were then used in the rest of the paitents. One bar and two stabilizers were used in all patients for the correction of the deformity The median operation duration was 60 min (range: 45–110) and the median duration of hospital stay was 5 days (range: 2–18). The bars have been removed in 38 patients as planned without any recurrence. According to the evaluation of the quality-of-life questionnaires 95.2 % of the patients are satisfied with surgical outcome.

Comments

Minimal access repair for pectus carinatum is initiated by Abramson and has been gaining support in recent years [4, 5]. MARPC mainly consists of

a pre-sternal placed pectus bar compressing the sternum, fixed on both sides of the chest wall on metal plates. Our carinatum bar and stabilizing system enables extra grip on the lateral ends for the screws, making the bar almost impossible to disengage. In addition, it can be adjusted for the patient very precisely with the use of its notches. The problem of steel wire breakage was addressed by using the wire in multifilament construction, or by using steel cords. Together with the stabilizers plates, it is a safe and easy-to-use prosthesis for MARPC.

Procedure Related Images

(Figs. 50.6, 50.7, 50.8, 50.9, and 50.10)

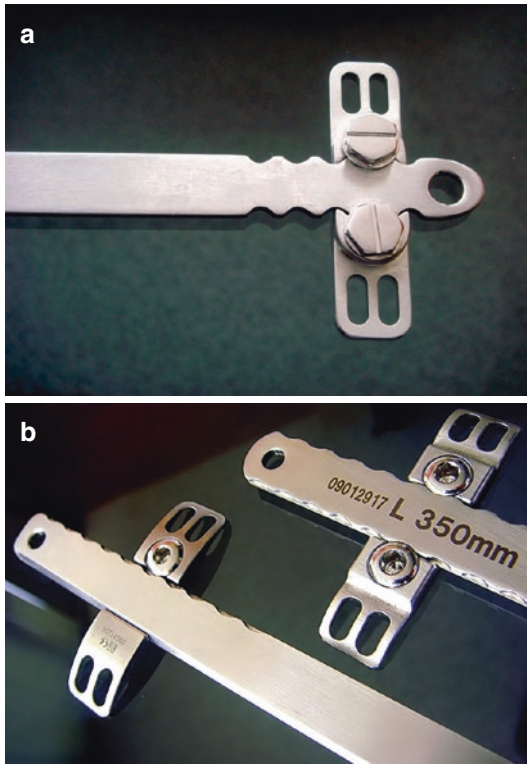

Fig. 50.6 (**a**) First generation carinatum bar and stabilizer. (**b**) Third generation carinatum bar and stabilizers

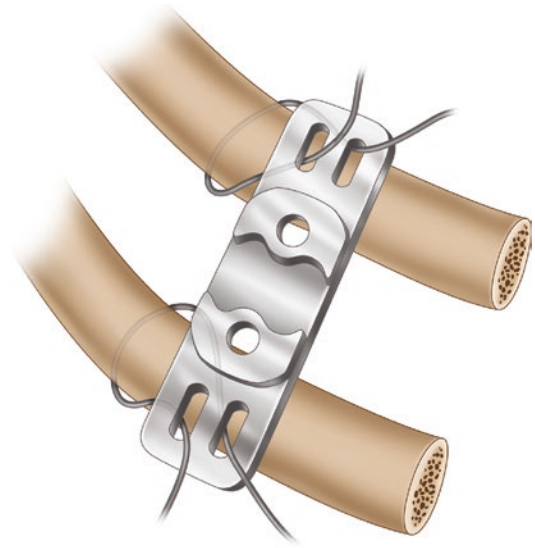

Fig. 50.7 Stabilizer plate being secured to the ribs

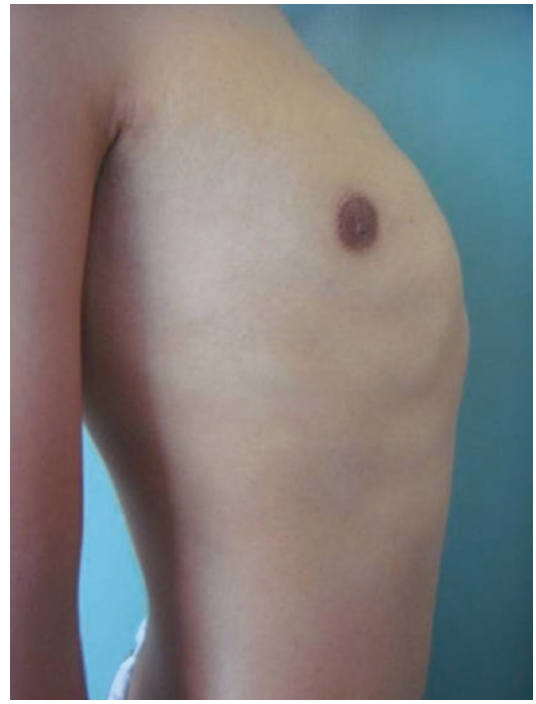

Fig. 50.8 Fourteen year-old male patient with a symmetric pectus carinatum deformity

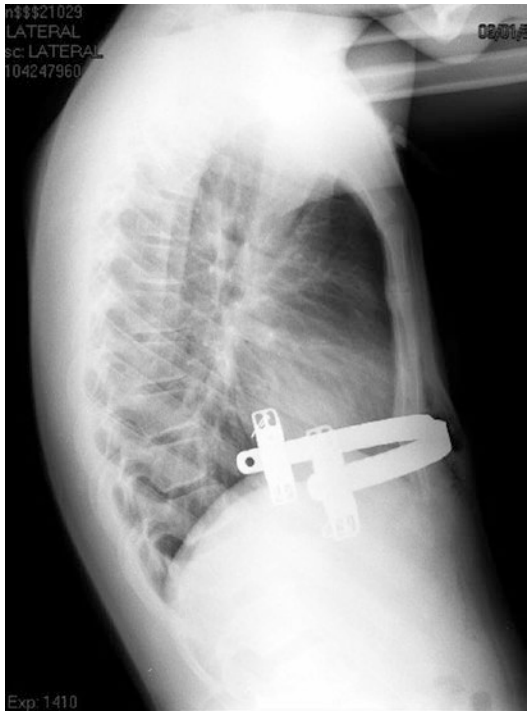

Fig. 50.9 Postoperative lateral x-ray of the patients following MIRPC

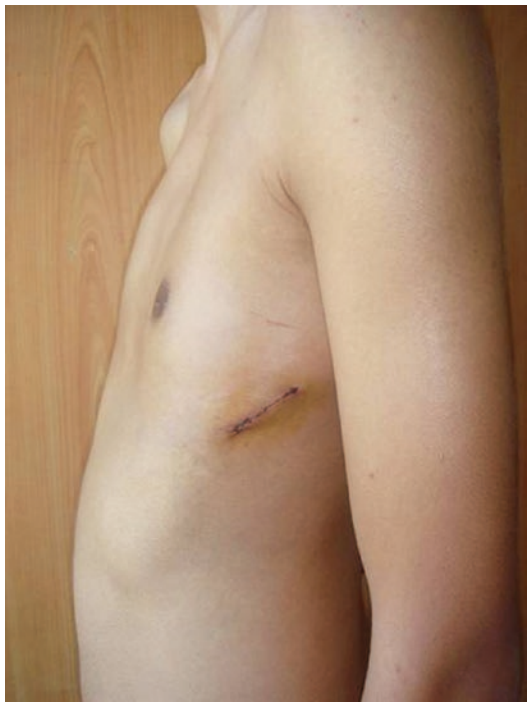

Fig. 50.10 The same patient as in image 4 in the third postoperative month

Acknowledgment I would like to thank Dr Korkut Bostanci for translation of the manuscript and for correction of the English grammar.

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51 Thoracoscopic Repair of Pectus Carinatum: Complete Cartilage Resection with Perichondrial Preservation Technique

Michele Torre, Marcello Carlucci, Luca Pio, and Patricio Varela

Technical Details

This technique for the thoracopscopic repair of pectus carinatum involves complete cartilage resection with perichondrial preservation (CCRPP). The technical details are as follows:

- Patient must be intubated as single-lung ventilation is essential for this procedure
- All anomalous cartilages are identified and the overlying skin is marked.
- The surgical team is positioned at the back of the patient.
- Three 5 mm ports are placed lateral to the posterior axillary line
- The camera port should be positioned few centimetres below the level of the nipple.
- The parietal pleura and posterior perichondrium is incised using a cautery hook.
- Intercostal vessels and nerves are preserved on both margins as the cartilages are progressively exposed.
- The cartilages are exposed lateral up to the chondro-costal junction and medial limit to the chondro-sternal junction
- The internal thoracic vessels are coagulated and dissected.
- Cartilages are excised using a thoracoscopic rongeur, leaving the anterior perichondrium intact.
- Multiple bites of the cartilage are necessary until anterior perichondrium is reached.
- Excision is facilitated by the compression on the anomalous cartilages on the anterior chest wall.
- A continuous suction from the third port is necessary during the surgery to guarantee good vision.
- On completion of the procedure a chest tube is positioned under thoracoscopic vision.

Indications

This thoracoscopic technique is not directed towards surgery on the sternum. For this reason, this approach is reserved for:

- Pectus carinatum limited to cartilage deformations
- Patients affected with unilateral pectus carinatum without rotation of sternum.

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A.K. Saxena (ed.), *Chest Wall Deformities*, DOI 10.1007/978-3-662-53088-7_51

Preoperative Considerations

1. Chest films in anterior-posterior and lateral views
2. General laboratory tests including basic coagulation test.
3. Respiratory or cardiologic assessment if associated conditions are suspected during history taking, the physical examination or the basic preoperative investigations.
4. Photographic documentation of the deformity before and after correction.
5. Patient must be intubated and single-lung ventilation may be helpful to the surgeon to better expose the surgical area.
6. The patient is positioned in the lateral decubitus position, with the unilateral pectus carinatum side raised up.
7. All anomalous cartilages are identified and the overlying skin is marked before commencing the procedure.

Special Instruments

General instruments for Video Assisted Thoracic Surgery (VATS). For this procedure, additional special instrument required is:

- Thoracoscopic rongeur

Surgical Technique

The surgical team is positioned towards the back of the patient. Three 5 mm ports are placed lateral to the posterior axillary line. One port is for the 30° 5 mm scope, second for the thoracoscopic rongeur and third for additional instruments required for manipulation. The optic port is positioned few centimetres below the level of the nipple. Carbondioxide is insufflated and maintained at maximal intra-thoracic pressure of 6 mmHg. With the help of cutaneous markers, local compression of the anterior thoracic wall on the anomalous cartilages allows the thoracoscopic identification of the anomalous cartilages. The parietal pleura and posterior perichondrium is incised using a hook cautery. Intercostal vessels and nerves are preserved on both superior and inferior margins as the cartilages are progressively exposed. The cartilages are exposed medially up to the level of internal thoracic vessels with the medial limit being the chondro-sternal junction. The internal thoracic vessels are coagulated and dissected. The cartilages are laterally exposed up to the chondro-costal junction that can be identified by change of colour glossy white to matt yellow. After complete exposure, cartilages are excised using a thoracoscopic rongeur, leaving intact the anterior perichondrium. Multiple bites of the cartilage are required until the anterior perichondrium is reached. Excision is facilitated by the compression on the anomalous cartilages from the anterior chest wall. A continuous suction through the third port is needed to achieve good visualization during the procedure. At the end of the procedure a chest tube is positioned under endoscopic vision. Surgical wounds are closed with absorbable sutures.

Procedure Overview (Figs. 51.1 and 51.2)

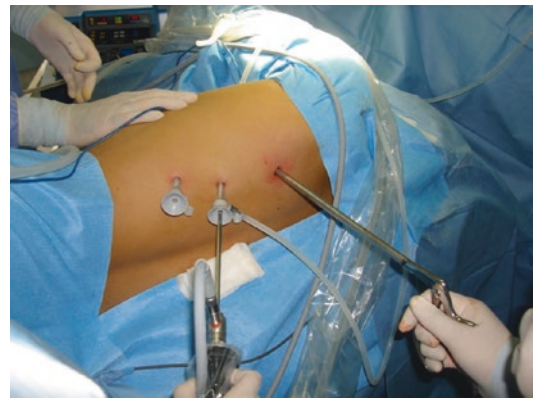

Fig. 51.1 The positioning of the patient for the procedure and the placement of ports

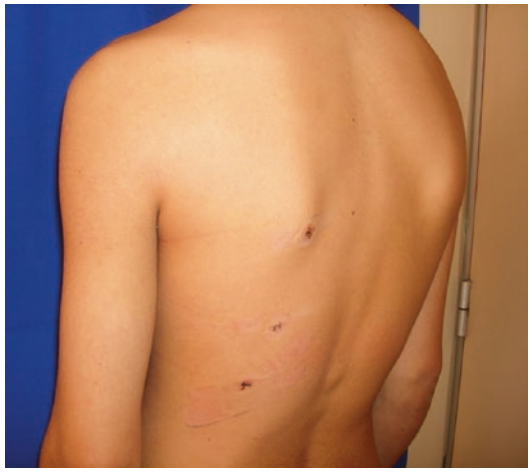

Fig. 51.2 Postoperative view of the port site incisions

Postoperative Management

Intravenous antibiotics are administered for 24 h. Analgesic treatment including morphine, Ketorolac, Paracetamol is continued for all the hospitalization. Epidural catheter is not necessary. Chest drain is left in place for 3–4 days, until the secretions reduce to below 60–70 ml in 24 h. The patient is mobilized as soon as thoracic drain is removed and discharged on the 4th or 5th day.

An elastic bandage with mild compression is applied around the thorax to maintain the sternum in the right position thus optimizing the result, and should be maintained from the day of surgery for 3 months. Sports have to be avoided for at least 3–6 months, corresponding to the period in during which consolidation of the cartilages is usually completed.

Results

No major complications have been observed with this technique. Bleeding may occur during surgery due to intercostals or internal thoracic vessel injury. It must be borne in mind, that there are two intercostal bundles are present, the one along the inferior margin of the rib being larger than the one along the superior edge.

In our experience, results are good if the cartilages are removed entirely and the sternum is not rotated. When dissection and cartilage exposure are not extended medially to the internal thoracic bundle, the removal of the cartilage is not complete which could lead suboptimal. No recurrences have been observed in our series, however insufficient correction has been observed in very severe anomaly, sternal rotation or when cartilage resection was incomplete.

Comments

Classical treatment of pectus carinatum has been based on two main principles: costal cartilage excision and sternal osteotomy [1]. In order to reduce OR to minimize morbidity of open procedures options such as orthotic brace system [2] and Abramson technique [3] have been proposed, both of which are based on the longer duration of sternum compression, however without cartilage excision. Kim and Idowu [4] were the first to describe the thoracoscopic treatment for unilateral PC in 2009. They applied the VATS principles of the minimal cartilage resection method proposed by Fonkalsrud in 2006 [5]. Our group [6] in 2010 proposed a modification of this technique applying the principles of the open surgical techniques that provide the complete cartilage resection of the abnormal ribs, preserving the anterior perichondrium (CCRPP). In fact, a complete resection of the abnormal costal cartilages reduces the chest circumference, facilitating the sternal depression and achieving optimal results. The best age for thoracoscopic approach has not been established, however we believe that the thoracoscopic approach is best suited for young teenagers, in whom the cartilages are not ossified. Recently Bell, Idowu and Kim [7] reported a combined technique in the minimally access repair of symmetric PC based on bilateral thoracoscopic chondrotomies and suprasternal compression bar placement.

The main advantages of thoracoscopic approach are: (1) Accesses for thoracoscopic approach are almost invisible and placed on the

posterior chest wall, (2) less pain, due to the absence of muscle dissection and (3) no muscle related complications that may be present in other techniques.

Procedure Related Images
(Figs. 51.3, and 51.4)

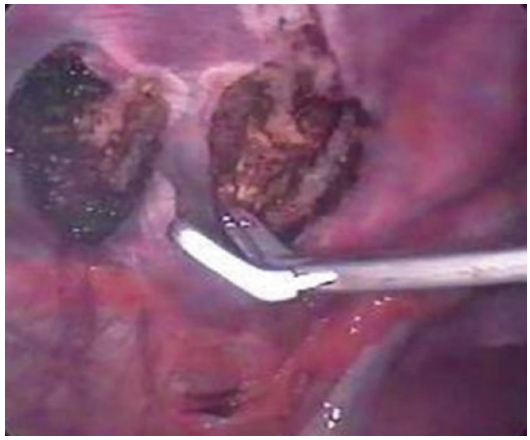

Fig. 51.3 Endscopic view of the cartilage resection using the thoracoscopic rongeur. Multiple bites of cartilages are taken while preserving the anterior perichondrium

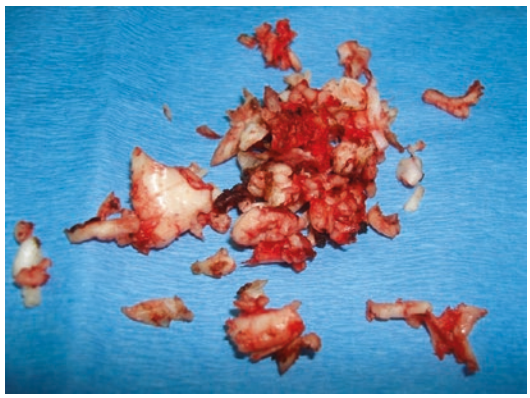

Fig. 51.4 The cartilage sections resected using VATS

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Part VII

Surgical Techniques Common for Pectus Excavatum, Pectus Carinatum, and Complex Chest Wall Deformities

Non-prosthetic Surgical Repair for Pectus Deformities

52

Ida Giurin and Ciro Esposito

Technical Highlights

The highlights of the prosthesis-free technique as reported by Marakawo et al. [1] are as follows:

- A 6–10-cm transverse skin incision was performed over the mid-portion of the defect.
- Subcutaneous, pectoral and rectus muscle flaps were elevated to the extent of the deformity
- The muscle flaps were elevated until they can oppose to the midline without excessive tension.
- Bilateral subperichondrial resection of deformed cartilage
- Transverse sternal osteotomy and xiphisternum excision to reposition sternum
- Wedge excision osteotomy is used to elevate the distal sternum for pectus excavatum
- Wedge osteotomy with bone fragments harvested from rib ends allowed depression of the sternum for pectus carinatum.
- The sternum is released to a neutral position and stabilized to the overlying muscle raphe closure

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Indications

This procedure can be employed for depression and protrusion type of chest wall deformities:

- Pectus excavatum
- Pectus carinatum

Preoperative Considerations

1. The procedure should be offered and performed in patients >12 years of age
2. Adult patients with rigid chest wall are well suited for this procedure.
3. Chest films are taken to access the extent of the deformity.
4. Lung function tests are performed to evaluate the pulmonary function.
5. Echocardiography and electrocardiograms to rule out cardiac pathologies.
6. Computed Tomography scan in severe thoracic asymmetry
7. Photographic documentation of the deformity
8. Coagulation tests are performed to rule out disorders in bleeding disorders
9. Blood reservation for auto-transfusion.

Special Instruments

Instruments for general thoracic surgical procedures.

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Surgical Technique

A 6–10-cm transverse skin incision is positioned over the center of the defect. Subcutaneous, pectoralis and rectus muscle flaps are elevated to the proximity of the deformity. The muscle flaps are elevated until it is possible to oppose them to the midline without excessive tension. Bilateral subperichondrial resection of deformed cartilage, transverse sternal osteotomy and xiphoid excision are performed to allow the sternum to position the sternum to normal morphological contours.

Wedge excision osteotomy is used to elevate the distal sternum for pectus excavatum. However for patients with pectus carinatum a wedge is placed in the osteotomy with bone fragments harvested from rib ends to allow depression of the sternum. Two heavy absorbable sutures are placed across the osteotomy to stabilize the sternum. An additional heavy suture is placed through the distal body of the neutrally placed sternum and later secured to the muscle raphe for additional stability. The perichondrium is reapproximated, and the pectoralis and rectus muscles are approximated. At this stage, the body of the sternum is fixed to this raphe using the previously placed suture. Submuscular and subcutaneous drains were placed.

Procedure Overview (Figs. 52.1, 52.2, 52.3, 52.4, and 52.5)

Postoperative Management

After surgery patients were managed on the ward with patient-controlled intravenous analgesia. This is switched to regular oral analgesia within 48 h. Drains are removed only after the total daily drainage is less than 50 ml. Patients are mobilized on the first day. An important aspect in achieving a good result is to subject the patient to a posture-maintaining exercise regimen, which should be continued for at least 6 months.

Comments

The key point of this procedure is that the sternum achieves a neutral position without any support. Its position is supported by the attachment to the overlying muscle raphe that hold it in the desired position, assisted by maintenance of good

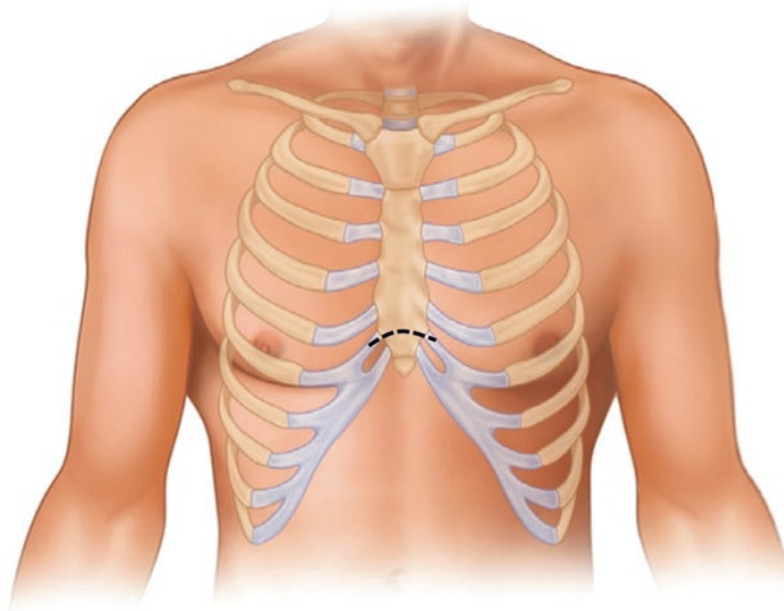

Fig. 52.1 A transverse skin incision is made over the center of the defect

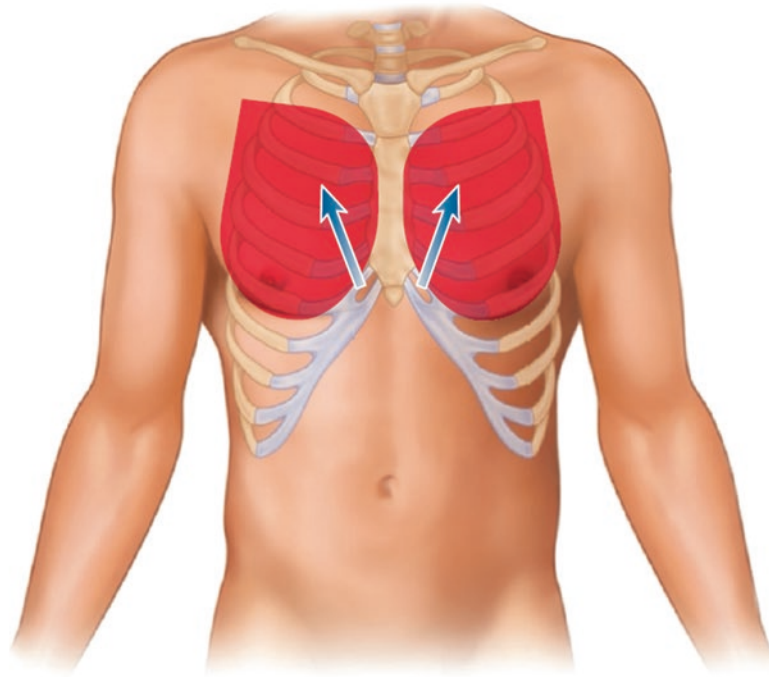

Fig. 52.2 Subcutaneous, pectoral and rectus muscle flaps were elevated to the extent of the deformity

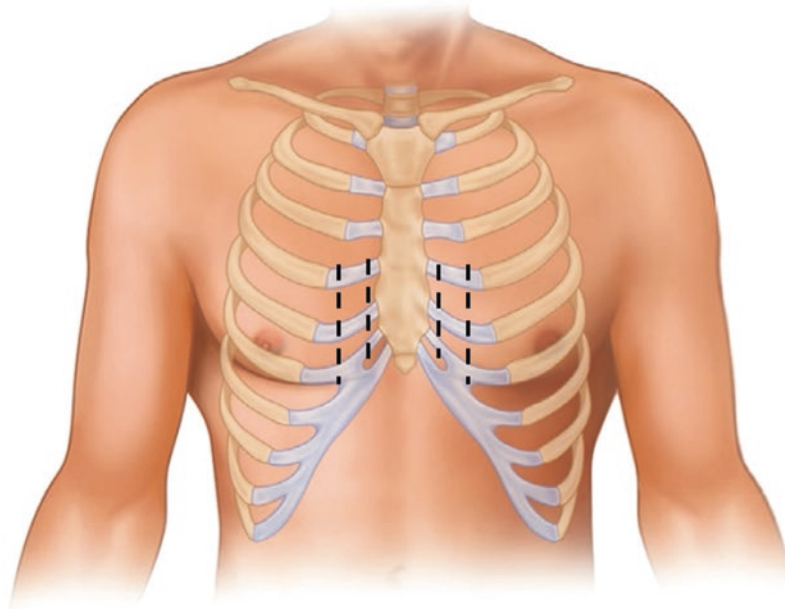

Fig. 52.3 Bilateral subperichondrial resection of deformed cartilage

posture after surgery. The natural role of the pectoralis muscle complex is to aid inspiration by pulling the ribs forwards and outwards, making it a suitable brace on which to attach the corrected sternum. This produces a stable repair with no evidence of a postoperative flail chest. Thourani and co-workers described other metal-free techniques whose sternal stabilization is dependent on relocation of the xiphoid–rectus complex to beneath the sternum as a buttress to prevent

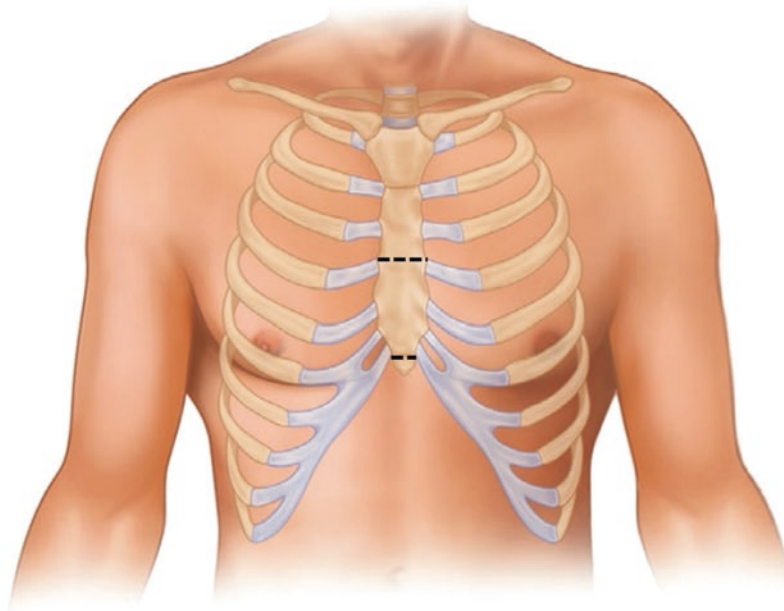

Fig. 52.4 Transverse sternal osteotomy and xiphioid excision to reposition sternum. Wedge osteotomy to elevate the distal sternum for pectus excavatum and with bone fragments interposition to allow depression of the sternum for pectus carinatum

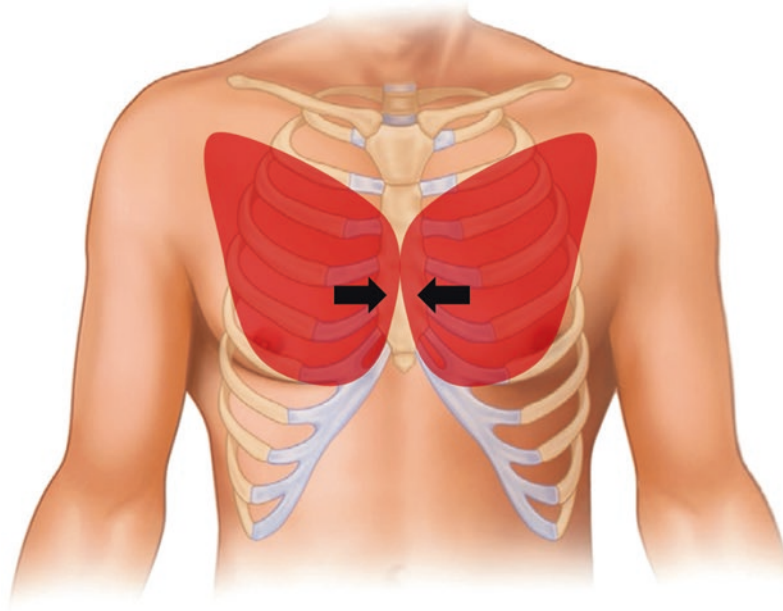

Fig. 52.5 The sternum is released to a neutral position and stabilized to the overlying muscle raphe closure

posterior collapse [2]. The initial practice of Fonkalsrud and colleagues varied because sometimes they do not use the muscle raphe to augment the repair [3].

Although correction of pectus carinatum without a metal bar is not novel, Makarawo et al. described this prosthesis-free technique for the surgical treatment of pectus carinatum also, using the muscle raphe closure to stabilize the repair [1]. The key advantage of the present technique lies in the avoidance of the metal prosthesis. The only limitation of this technique is the short follow-up. Long-term follow-up will determine the outcomes of the techniques.

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53 Minimally Access Open Repair of Pectus Excavatum and Carinatum (Robicsek Procedure)

Francis Robicsek and Benjamin B. Peeler

Repair of Pectus Excavatum

Technical Highlights

The procedure that is coined as "minimal access open repair" is performed as follows:

- A transverse, pre-sternal, midline, slightly upward convex 4–6 cm skin incision is made
- The depth of the incision is to reach the sternum and the pectoralis fascia and the broad, full-thickness skin-flap is widely mobilized
- Using electrocautery, the pectoralis major muscles are detached from the sternum, retracted to expose the costal cartilages
- Using the upper limit of the sternal depression as a guide, the deformed cartilages are removed one-by-one, using sharp and blunt dissection.
- The length of the chondral resection is proportional with the degree of depression
- It is seldom necessary to resect portions of the third, and almost never of the second rib.
- The resection is done sub-perichondrally to promote the regeneration of the cartilages. For the same reason, a minimum a 1-cm length of cartilage containing its growth center is left in continuity with the bony ribs.
- Using a wide osteotome and a small caliber gage, a wedge-osteotomy is performed in line with the upper level of the sternal depression.
- The xiphoid process is detached from the sternum and is allowed to withdraw caudally.
- The lower tip of the sternum is grabbed with a towel-clip and, using blunt dissection, the sternum is freed of tissue connections with the pericardium and the pleura.
- Using electrocautery, the sternum is disconnected from the costal and intercostal tissue.
- The right pleural cavity is entered and a wide connection is established between the retrosternal and right pleural space. Using a catheter inserted through a separate stab wound, underwater drainage of the right pleural space is established.
- The sternum is now forcefully bent forward into a corrected position.
- To maintain the sternum elevated, a Prolene® (Ethicon, Inc) mesh is placed under the entire portion of the mobilized sternum and using 0–0 non-absorbable filaments, sutured under moderate tension bilaterally to the stumps of the ribs.

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- The xiphoid process is mobilized and used as the lower anchor to the mesh.
- The pectoralis muscles are united in front of the sternum and the wound is closed.
- In asymmetric pectus excavatum the transverse sternotomy is carried deeper. The repair also requires uneven bilateral cartilageous resection. The new, twisted position of sternum is secured with a reversed "Z" heavy steel-wire suture.

Surgical Indications

- Symmetric pectus excavtum
- Asymetric pectus excavatum

Preoperative Considerations

1. Both pediatric and adult patients are well suited for this procedure.
2. Chest radiographs are taken to access the extent of the deformity.
3. If necessary electrocardiograms or echocardiography to rule out cardiac pathologies.
4. Photographic documentation of the deformity

Special Instruments

General instrument set for thoracic surgical procedures. Special material requirements:

- Prolene® (Ethicon, Inc) mesh

Surgical Technique

The patient is placed in the supine position with the spinal curvature exaggerated, using a posteriorly placed sandbag or rolled-up sheet. The anterior chest is disinfected and draped. On the right side, the exposed area is extended to allow the insertion of an intercostal pleural catheter. A transverse, pre-sternal, midline, slightly upward convex skin incision is made in accordance with Langer's lines in the length of not more than 4–6 cm. The depth of the incision is to reach the sternum and the pectoralis fascia. The skin and the subcutaneous tissues are not separated. The broad, full-thickness skin-flap is widely mobilized in a way to allow to be moved by retraction freely over the entire operative field. Using electrocautery, the pectoralis major muscles are detached from the sternum, retracted laterally exposing the costal cartilages to the costochondral junctions, cranially to the upper limit of the depression. Using the upper limit of the sternal depression as a guide, the deformed cartilages are removed one-by-one, using sharp and blunt dissection.

The length of the chondral resection is proportional with the degree of depression, but in general, the shortest segments are resected from the uppermost deformed cartilage and progressively longer portions are removed as the procedure progresses downward. It is seldom necessary to resect portions of the third, and almost never of the second rib. The resection is done subperichondrally to promote the regeneration of the cartilages. For the same reason, a minimum a 1-cm length of cartilage containing its growth center is left in continuity with the bony ribs. These precautionary measures are especially important in the growing child to assure the continued normal development of the thoracic cage. In general, the resection involves the depressed portion of the cartilages, but it should not include removal of any part of the ribs which are at level or above the most anterior portion of the chest wall. To extend the resection into the bony portion of the ribs is seldom necessary, and if it is done, it should not involve more than 1–2 ribs.

Using a wide osteotome and a small caliber gage, a wedge-osteotomy is performed in line with the upper level of the sternal depression. The osteotomy should not extend into the posterior lamina and it is made in line with the respective intercostal space, rather than with the chondrosternal junction.

The xiphoid process is detached from the sternum and is allowed to withdraw caudally. The lower tip of the sternum is grabbed with a towel-clip and, using blunt dissection, the sternum is freed of tissue connections with the peri-

cardium and the pleura. Using electrocautery, the sternum is disconnected from the costal and intercostal tissue. In the course of this last maneuver, care is taken to preserve the integrity of the mammary vessels.

The right pleural cavity is entered and a wide connection is established between the retrosternal and right pleural space. Using a catheter inserted through a separate stab wound, underwater drainage of the right pleural space is established. This seemingly unnecessary maneuver assures drainage of the retrosternal space, thus proper wound healing.

The sternum is now forcefully bent forward into a corrected position. To maintain the sternum elevated, a large-pore (to allow drainage), cut-to-size Prolene® (Ethicon, Inc) mesh is placed under the entire portion of the mobilized sternum and using 00 non-absorbable filaments, sutured under moderate tension bilaterally to the stumps of the ribs. If in effort to preserve the growth center, a small portion of the deformed ribs is left *in loco*, sutures are placed through the most prominent portion of the ribs overlapping the amputated tips. By incising the anterior rectal fascia, the xiphoid process is mobilized and used as the lower anchor to the mesh. At the end of the suturing, the mesh supporting the sternum should be "tout as a drum", but not under undue tension.

After appropriate hemostasis, the pectoralis muscles are united in front of the sternum and the wound is closed without additional drainage. Any pressure-dressing should be avoided.

Procedure Overview (Figs. 53.1, 53.2, 53.3, 53.4, 53.5, and 53.6)

Comments

Pectus excavatum and carinatum are the two most commonly occurring anterior chest deformities. They are characterized by a funnel shaped depression and a keel-like protuberance of the sternum and adjacent cartilages, respectively. They occur in about 1 % of the general population as isolated anomalies, but may also be part of complex musculoskeletal syndromes. Patients with pectus excavatum outnumber those with carinatum 10:1. Males are affected more frequently than females, as to a proportion 3:1. Positive family history of chest deformity has been reported as high as 37 % [1]. In our surgical experience, different varieties of pectus excavatum and carinatum constituted about 9:1 of more than 1200 patients operated upon for anterior chest deformities [2–31].

Our recommended interventions to correct both pectus excavatum and carinatum contain several elements of the original operations of Sweet [32] and Ravitch [33–35]; however, they differ in several respects, such as avoiding wide exposure, utilizing a posterior mesh support in patients with pectus excavatum and shortening of the sternum and applying the traction of the rectus abdominis muscles in pectus carinatum. The avoidance of rods and plates in any form and position is strongly emphasized as being unnecessary, dangerous and cost-contributory.

All of our interventions [2–31] are based upon the concept that in these conditions the sternum is but an "innocent bystander", and the real culprits are the overgrown and elongated costal cartilages. This concept has been first emphasized by Richard Sweet of the Massachusetts General Hospital:

> Unless appreciable length of lower cartilages are resected, it is impossible to correct the deformity even after releasing the pull of the substernal, diaphragmatic and fascial attachments and the removal of a wedge of bone at the upper limits of the gladiolus. One gets the impression that the deformity may be primarily skeletal. [3]

Whenever the elongated cartilages push the sternum in the retrograde direction, pectus excavatum develops. If the abnormally long cartilages force the sternum forward, the process results in pectus carinatum Uneven, unilateral overgrowth of the cartilages leads to asymmetrical pectus deformities, while abnormality of a single or a few cartilages may create localized, asymmetrical anomaly. Other proposed causes include anomalous development of the diaphragm and premature fusion of the sternal growth centers [34].

According to these principles, Sweet in 1944, in the course of the operation of two patients with

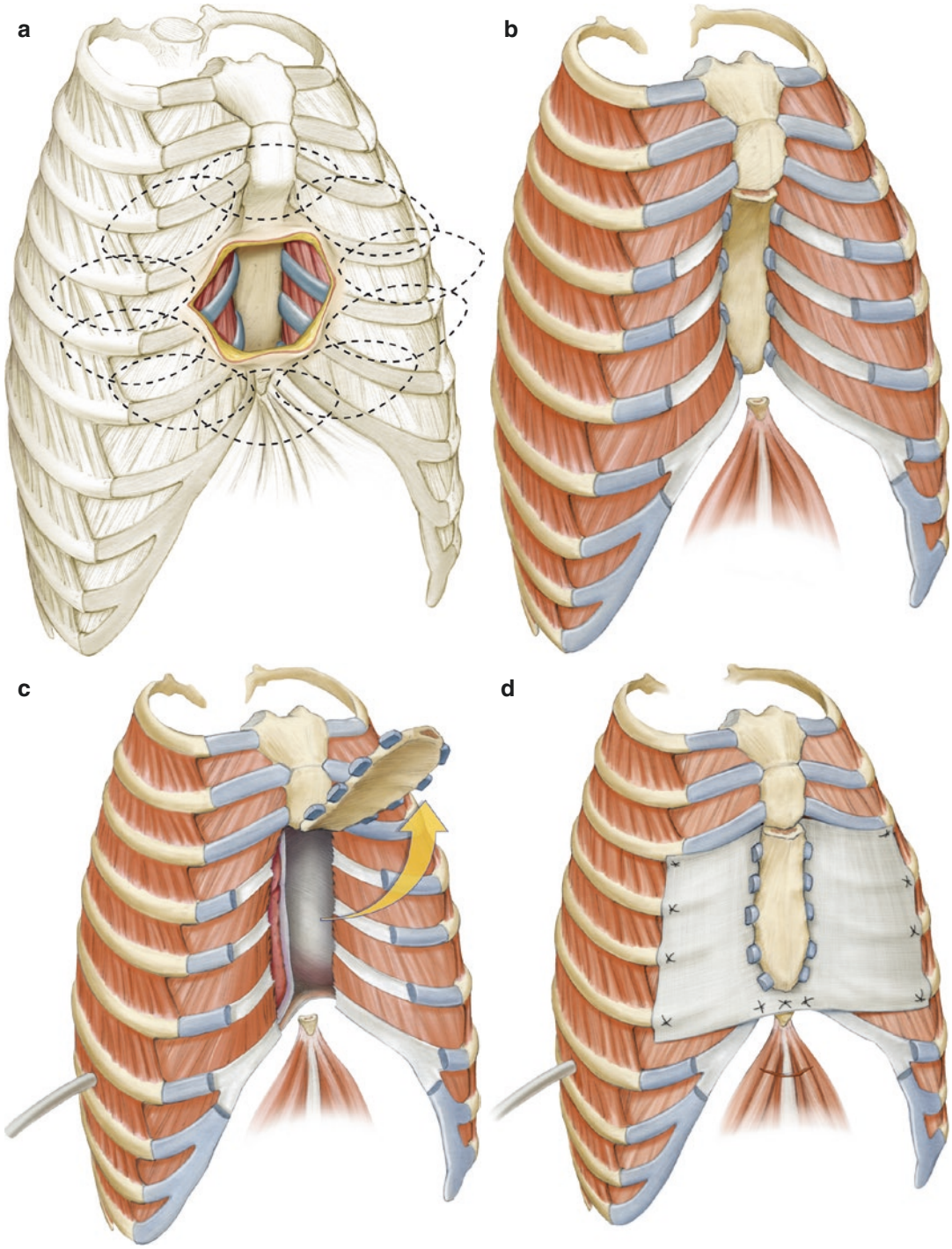

Fig. 53.1 (**a**) A small 4–6 cm long, transverse, midline, slightly upward convex incision is made over the sternum. A full-thickness skin flap developed and mobilized to provide necessary exposure to the different stages and areas of the intervention. (**b**) Using electrocautery, the pectoralis muscles are detached from the sternum, retracted and the costal cartilages are exposed. Using the upper limit of the deformity as the guide, the deformed cartilages are sub-perichondrally resected, progressively long segments as the procedure progresses downward. Using a wide osteotome a transverse sternal wedge osteotomy is performed in-line with the respective intercostal space. The xiphoid process is detached from the sternum. (**c**) The sternum is freed of its posterior and interior connections, forcefully bent forward (**d**) and supported by a synthetic large pore-mesh sutured to the costal stumps of the rib cartilage

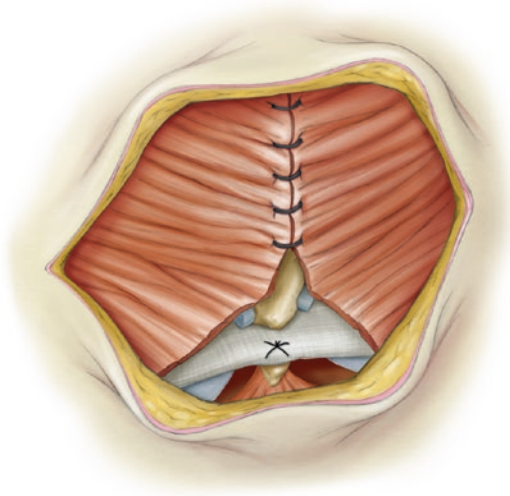

Fig. 53.2 After the procedure is completed, the pectoralis muscles are united pre-sternally

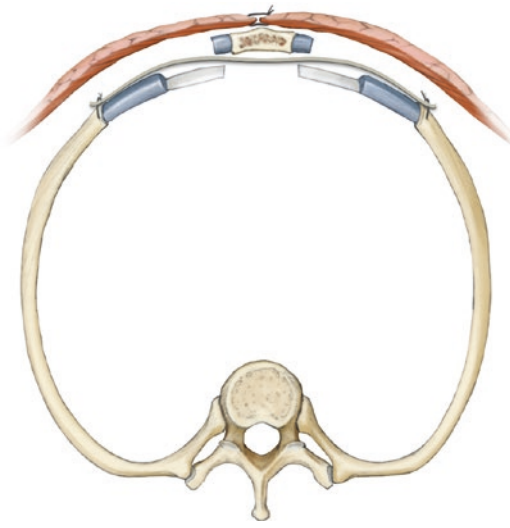

Fig. 53.3 Cross-section of the bony thorax after minimally invasive open repair of pectus excavatum

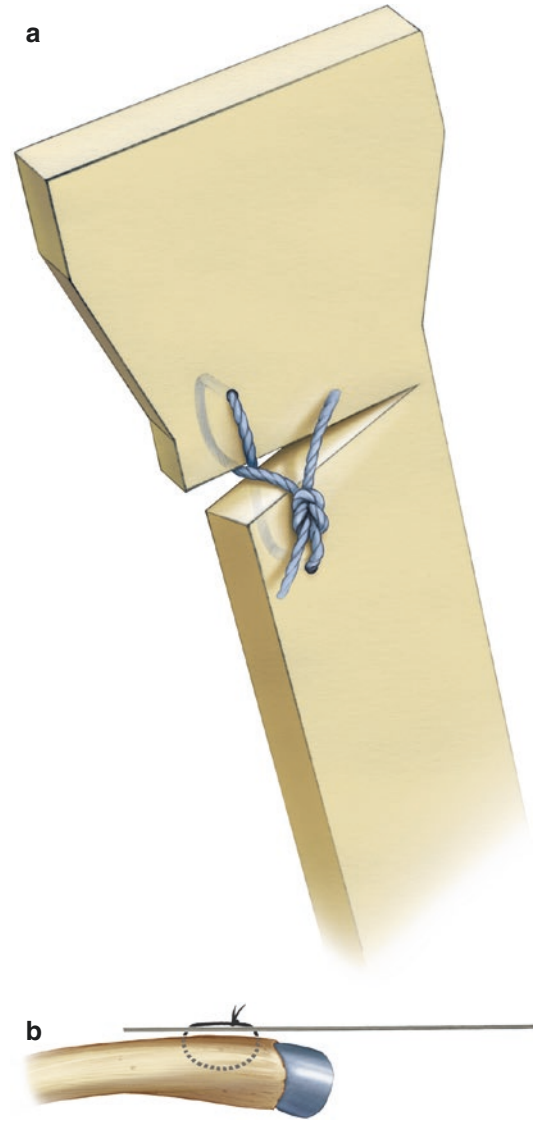

Fig. 53.4 The rotation of the sternum is corrected by twisting the sternum toward the protruding side and supporting it with a reverse "Z" heavy steel wire suture. (**a**) It is mandatory that in the growing child, minimum, 1-cm length of the cartilage containing the growth center should be left in continuity with the bony ribs to assure regeneration of the cartilages. If this segment is bending inward, the mesh should be anchored more laterally at the most prominent portion of the rib cage (**b**)

pectus excavatum, excised the xiphoid process, performed segmental resection of the costal cartilages from the III down, and did a transverse wedge osteotomy of the sternum just below the second cartilage. He also closed the chondral defects with sutures and maintained the sternum in an overcorrected position with wire-sutures placed across the osteotomy [33]. The "basic" operation of Ravitch, performed in 1949, was very similar to that of Sweet, with the exception that he detached the intercostal bundles of the sternum and did not remove the xiphoid process. This operation, which became generally known as

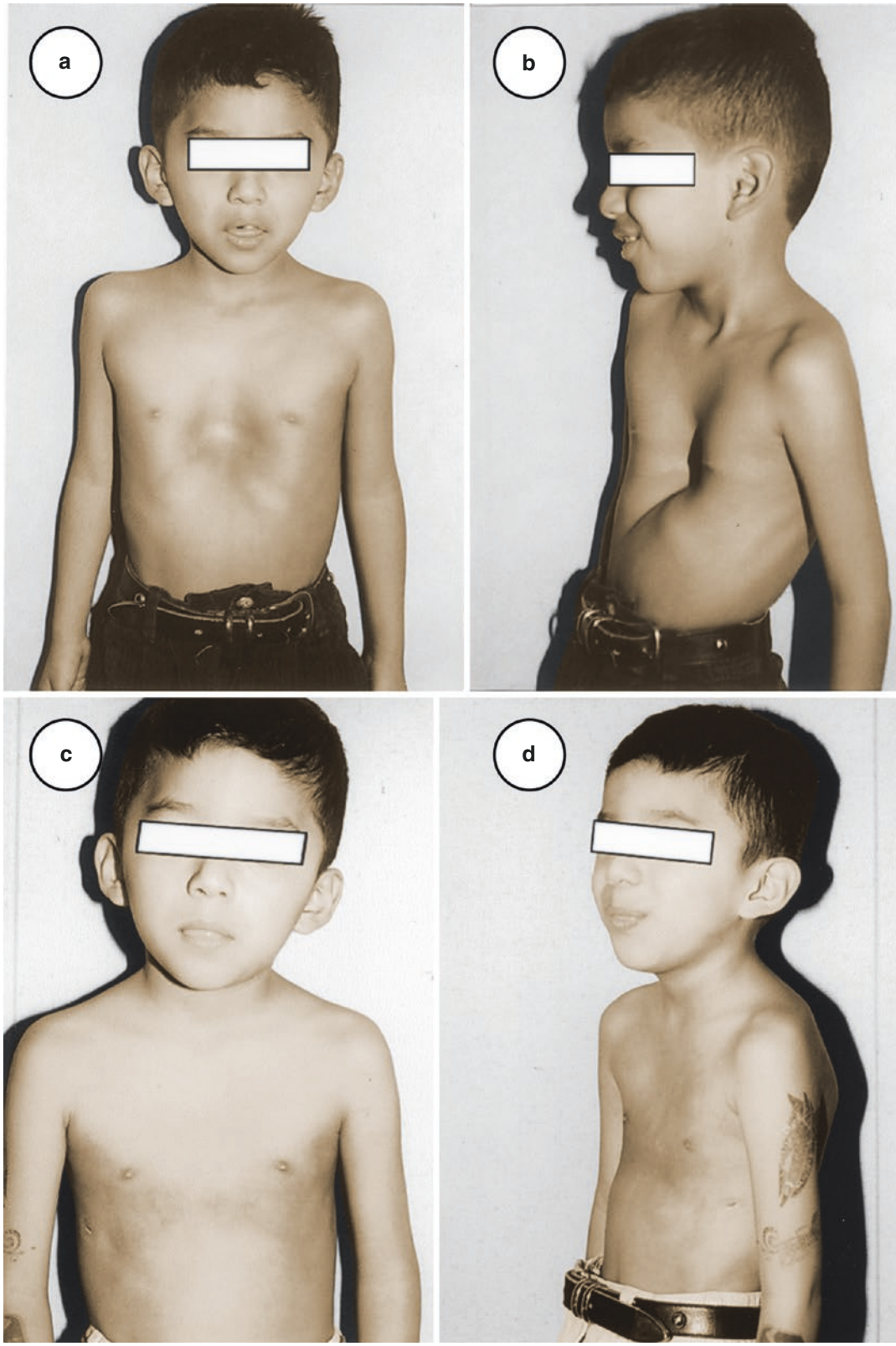

Fig. 53.5 A 6 y.o. boy before (**a**, **b**) and after (**c**, **d**) minimally invasive open correction of pectus excavatum

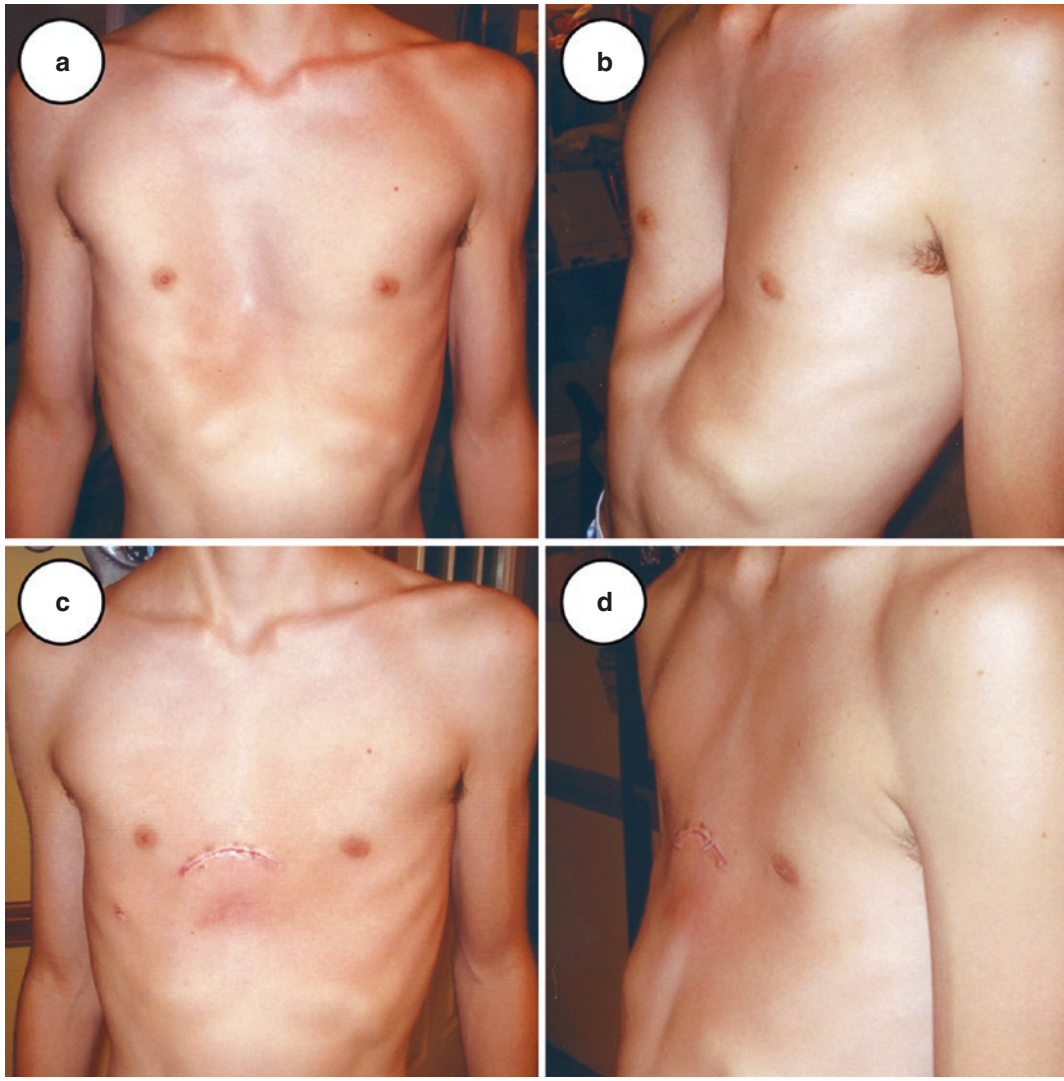

Fig. 53.6 A 17 y.o. patient before (**a**, **b**) and after (**c**, **d**) minimally invasive open correction of pectus excvatum

the "Ravitch-procedure" [34, 35], was applied to thousands of patients by three generations of surgeons. It yielded fairly good results, however, recurrences were common. The reason for this was that while the procedure satisfactorily addressed some of the pathogenic factors, such as the elongated cartilages and the displaced sternum, it failed to reliably insure that the position of the sternum indeed remained corrected. (Note: In the ongoing comparative evaluation of different forms of modern pectus excavatum surgery, the authors suggest to eliminate the term "Ravitch operation" altogether, because now it is improperly applied by many to all forms of "open" repairs. Nowadays, there are many procedures that may include some of the features of the classic Ravitch operation, but their general approach is widely different).

The high recurrence rate was already evident to Ravitch [34, 35], as well as to other surgeons [36–38], who recommended various retro and intraosseous "supports" of different materials, primarily metal rods, to assure that the posture of the sternum remains corrected indeed. While substernal rigid supports significantly improved the postoperative results, they also inherently increase the dan-

gers of infection and other complications relevant to dislodgement of supportive rods, and made reoperation to remove the implanted material a necessity. This induced us to apply retrosternal autogenous [2, 3] and later synthetic mesh support [8, 10, 11, 13–15, 19, 21, 22, 24, 28, 30] to which none of these features were inherent.

Procedure Related Images (Figs. 53.7, 53.8, 53.9, 53.10, and 53.11)

Repair of Pectus Carinatum

Technical Highlights

The "minimal access open repair" is performed as follows:

- A transverse, pre-sternal, midline, slightly upward convex 4–6 cm skin incision is made
- The depth of the incision is to reach the sternum and the pectoralis fascia and the broad, full-thickness skin-flap is widely mobilized
- Using electrocautery, the pectoralis major muscles are detached from the sternum, retracted to expose the costal cartilages
- The deformed cartilages are removed one-by-one, using sharp and blunt dissection.
- The resection is done sub-perichondrally to promote the regeneration of the cartilages. For the same reason, a minimum a 1-cm length of cartilage containing its growth center is left in continuity with the bony ribs.
- Using a wide osteotome and a small caliber gage, a wedge-osteotomy is performed in line with the upper level of the sternal depression.
- Using electrocautery, the sternum is disconnected from the costal and intercostal tissue.

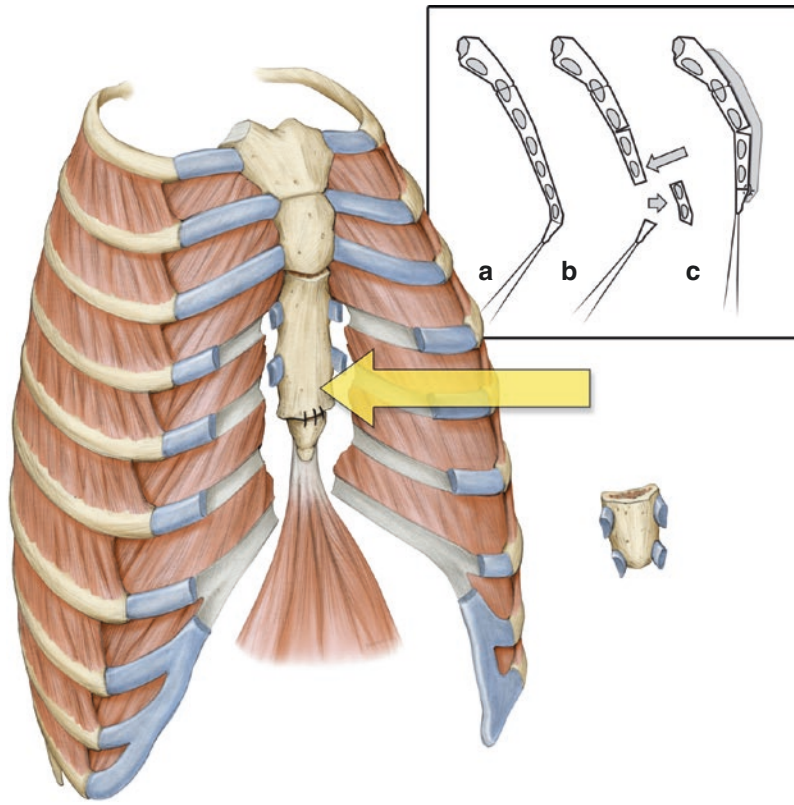

Fig. 53.7 In Type A pectus carinatum, the lower position of the sternum forms a 90° angle with the xiphoid process. After performing a transverse sternotomy, bilateral chondral resection (*a*), and correction of the sternal axis a short portion is resected from the lower sternum (*b*), and the xiphoid process is reattached to the sternal stump. Pre-sternal closure of the pectoralis muscles assures the corrected position of the sternum and a smooth anterior chest wall (*c*). In Type B anomaly, the sternal resection may be omitted

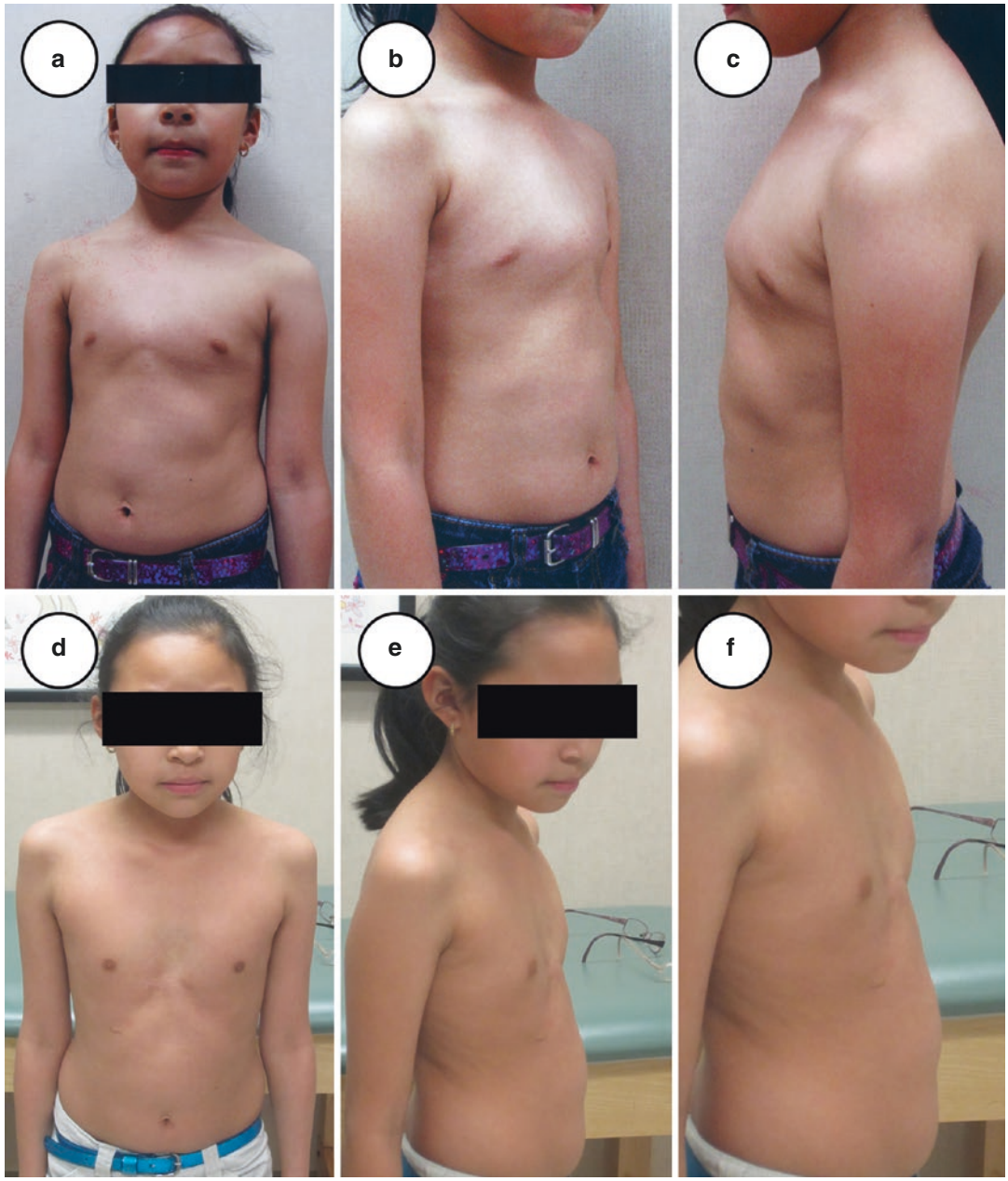

Fig. 53.8 A 12 y.o. girl before (**a–c**) and after (**d–f**) minimally invasive open correction of Type A pectus carinatum

- In Type A, short portion is resected from the lower tip of the sternum, and then the sternum is bent to achieve a normal straight axis. The xiphoid process is re-attached to the resected stump of the sternum with a pair of stainless steel wire sutures. Sternum is further secured by closing the previously detached pectoralis muscles pre-sternally.
- In Type B, no sternum shortening and no detachment of the xiphoid process is necessary.
- In Type C pectus carinatum, bilateral parasternal costal resection usually suffices.
- The pectoralis muscles are united in front of the sternum and the wound is closed.

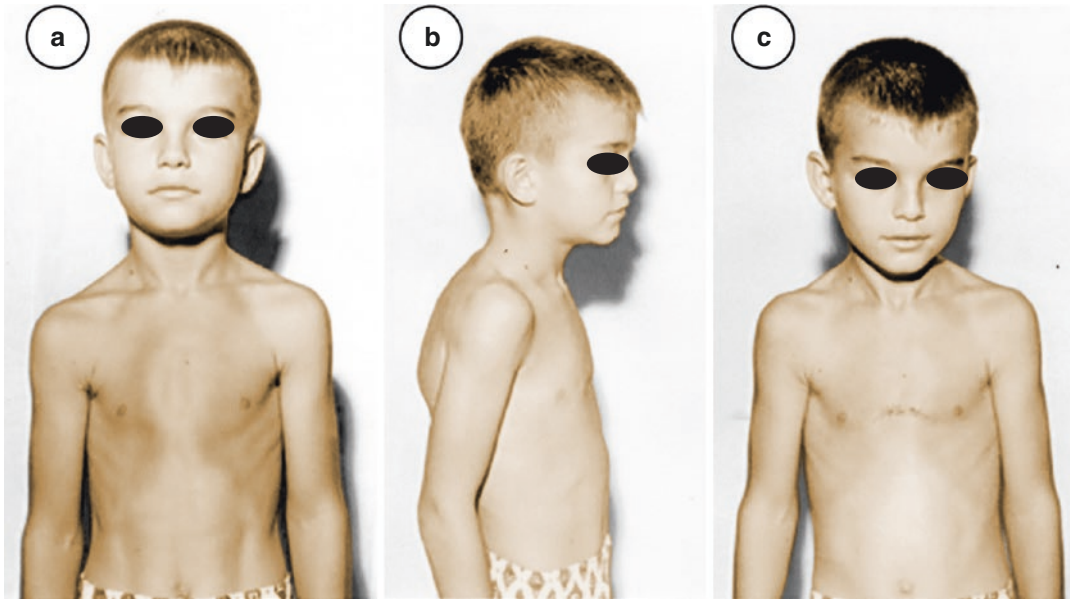

Fig. 53.9 A 14 y.o. boy before (**a**) and after (**b**, **c**) minimally invasive open correction of pectus carinatum

Indications

- Type A Pectus carinatum (protrusion of lower sternum with bent xiphoid)
- Type B Pectus carinatum (protrusion of lower sternum along with xiphoid)
- Type C Pectus carinatum (bilateral protruding ridge of costal cartilages)

Preoperative Considerations

Please refer to the section on preoperative consideration for pectus excavatum in this chapter

Surgical Technique

The initial stages of surgical correction, i.e., exposure of the sternum, subperichondrial resection of the involved cartilages, transverse sternal wedge osteotomy of both types of pectus carinatum, are identical with those described in Paragraph one for the repair of pectus excavatum. However, the right pleural space is not entered and the intercostal and chondral strips are left in continuity with the sternum. In Type A, short 4–6 cm portion is resected from the lower tip of the sternum, and then the sternum is forcefully bent backward to achieve a normal straight axis. The xiphoid process is re-attached to the resected stump of the sternum with a pair of stainless steel wire sutures. This way the pulling effect of the rectus abdominis muscles will preserve the sternum in its corrected position, which is further secured by closing the previously detached pectoralis muscles pre-sternally. In *Type B*, where the xiphoid process is "in line" with the sternal axis, sternum shortening and detachment of the xiphoid process is usually deemed unnecessary. The wound is drained with a small caliber suction device, closed, and a moderately compressive dressing applied.

In a less frequently seen symmetrical carinatum anomaly, *Type C pectus carinatum*, the sternal prominence is moderate or absent, but there is a bilateral protruding ridge of costal cartilages. In such cases, bilateral parasternal costal resection usually suffices.

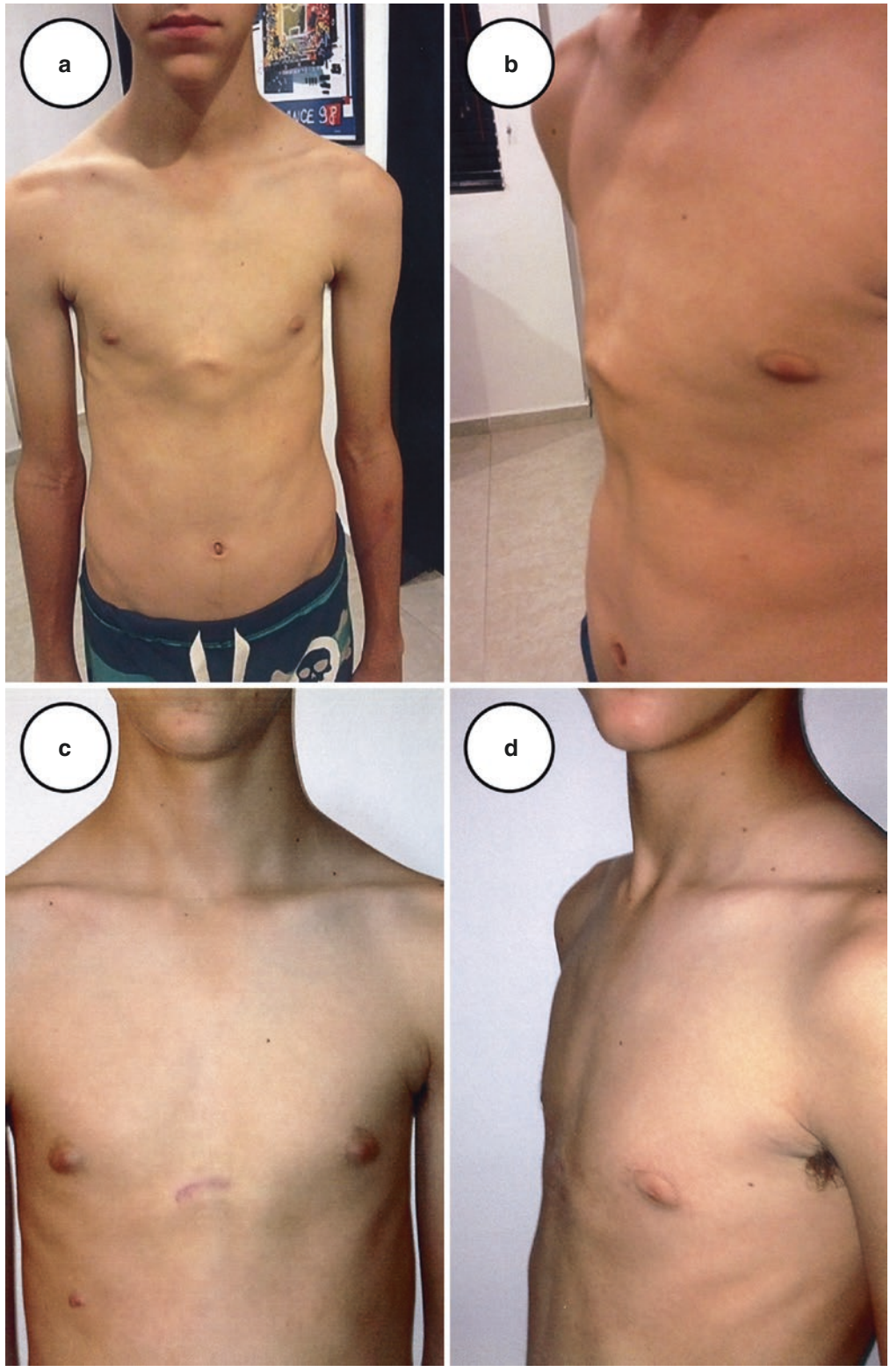

Fig. 53.10 A 17 y.o. patient before (**a**, **b**) and after (**c**, **d**) minimally invasive open correction of asymmetrical pectus carinatum

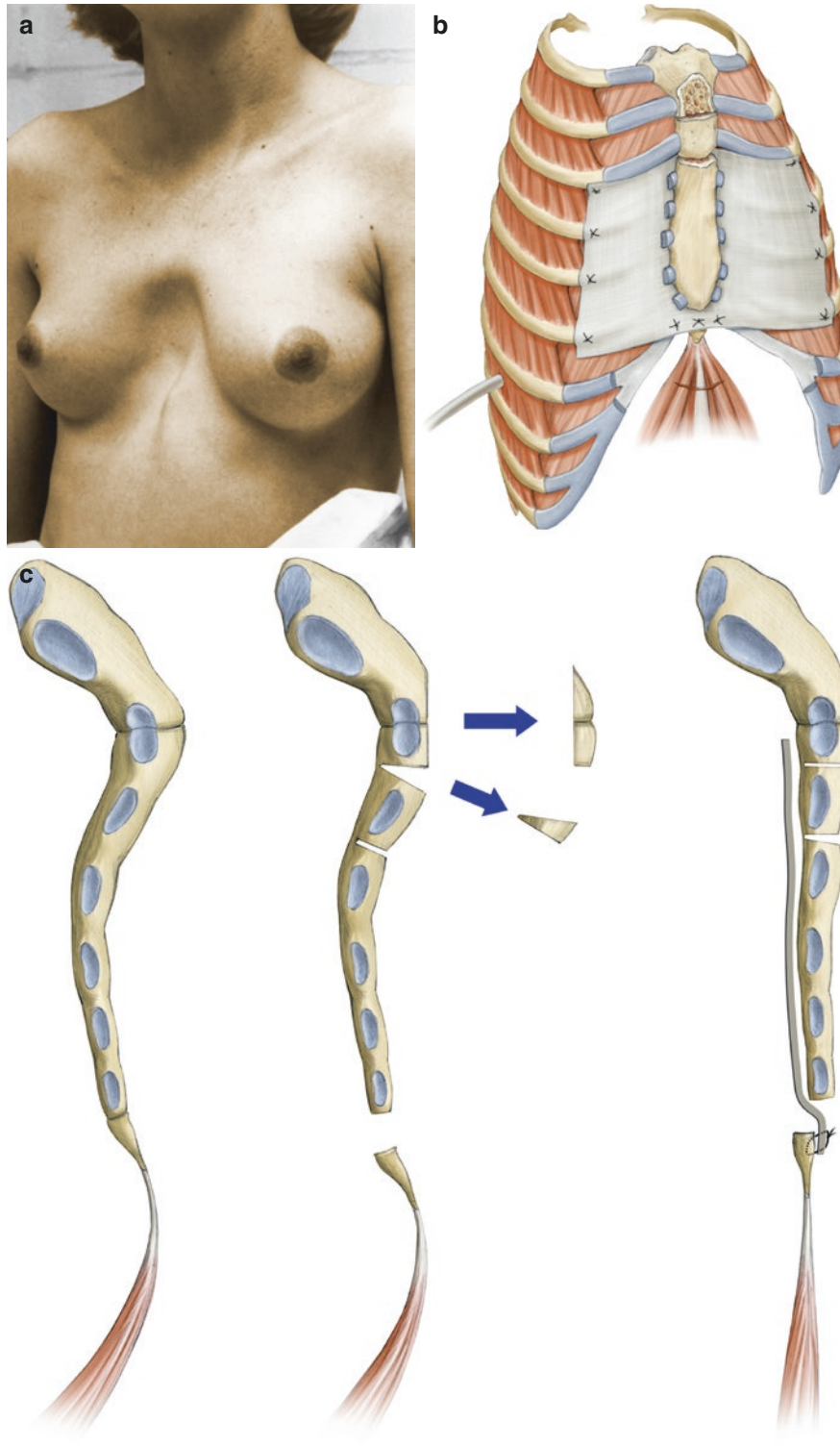

Fig. 53.11 While in Type A, pouter pigeon breast is limited to removal of the manubrial protrusion, the correction of Type B pouter pigeon breast (**a**) also requires, besides the removal of the manubrial protrusion, a second transverse sternotomy, bilateral resection of the deformed cartilages, correction of the sternal axis and substernal mesh-support (**b**, **c**)

Procedure Overview

Comments

Pectus carinatum is a spectrum of progressive inborn anomalies of the anterior chest wall named after the keel (carina) of the ancient Roman ship characterized by protrusion of the sternum and the adjacent costal cartilages. Pectus carinatum is often associated with various other conditions, notably Marfan disease, homocystinuria, prune belly, osteogenesis imperfecta, Noonan syndrome and mitral valve prolapse [1, 39]. All of these may complicate the formation of the plan for correction. The suggestion that there is a strong link with congenital heart disease [1] may be misleading, because a variety of congenital heart conditions result in right ventricular hypertrophy, which in turn may cause bulging of the precordium and give the appearance of keel chest (pseudo pectus carinatum). Overzealous pectus excavatum repair may also result in pectus carinatum [39], especially after the usage of forward-convex rigid bars, like those used in the Nuss-procedure. A heretofore unknown form of iatrogenic pectus carinatum has been recently described and attributed to fibroelastic changes of the sternum and the costal cartilages stimulated by implanted metal bars [40].

The initial description of pectus carinatum stems from Hippocrates:

> The chest becomes sharply pointed and not broad and becomes affected with difficulty in breathing and hoarseness. [41]

The first repair of pectus carinatum was carried out by Lester in 1943 [42]. Later on, Ravitch performed a sternal osteotomy and cartilageous resection to correct pectus carinatum. Subsequently he modified his technique by adding perichondrial "reefing" sutures to secure the corrected sternal posture [36].

I. *Symmetrical pectus carinatum*
Symmetrical pectus carinatum is the condition that fits best with the term of "keel chest". The sternum broadly arches forward often with mild, relative to the sternum, depressed costal cartilages which may accentuate the sternal prominence [36, 42]. Symmetrical pectus carinatum has three common varieties:
- *Type A pectus carinatum* is characterized by protrusion of the lower portion of the sternum and formation of 90° backward open angle with the xiphoid process.
- In *Type B pectus carinatum*, the lower portion of the sternum is protruding but the longitudinal axis of the xiphoid process remains "in line" with it.
- In *Type C pectus carinatum*, the sternal prominence is moderate or absent, but there is a bilateral protruding ridge of costal cartilages.

II. *Asymmetrical Pectus Carinatum*
Localized protrusions of the chest wall, not involving the sternum, are treated according to their individual morphological features, and are limited to resection of the protruding portion of the sternum and/or costal cartilages. Larger defects may require mesh-closure.

Procedure Related Images

Pouter Pigeon Breast

(Currarino-Silverman syndrome, costogladiolar prominence with costosternal depression).

Pouter pigeon breast is a relatively infrequently seen "mixed" anomaly, which may be classified either in the "excavatum" or in the "carinatum" group. *Type A Pouter pigeon breast* is limited to the protrusion of the abnormally thick sternal manubrium and the adjacent one to three costal cartilages. In the more frequently seen *Type B Pouter Pigeon Breast*, besides the proximal sternal protrusion, the C shaped sternal body is depressed, backward arching and the xiphoid process points slightly anteriorly. This further exaggerates the sunken appearance of the meso-sternum. Pouter pigeon breast is attributed not only to the abnormal elongation of the costal cartilages, but also to premature fusion or absence of the sternal ossification centers [43].

In the authors' technique of pouter pigeon breast repair, the skin incision is made somewhat higher than in carinatum repair. Depending on the individual morphology, the sternum is remodeled in the following way:

In Type A pouter pigeon breast, where the depression of the midsternum is moderate or absent and the abnormal anatomy is dominated by protrusion of the manumbrium and adjacent cartilages, the repair is limited to that area. First, a transverse groove is created in the sternum at the level of the 2nd intercostal space in about two-thirds of the sternal thickness. Then a wide osteotome is inserted in the groove, aimed in the cranial direction and all the protruding tissues are "shaved off".

In Type B anomaly, where the sternal body is depressed, in addition to the above, the depressed sternal body is treated similar to a primary pectus excavatum repair with an additional transverse sternotomy performed at the level of the third or fourth intercostal space [2–5, 10, 11, 14–17, 20, 21, 24, 25].

General Remarks to Repair of Pectus Deformities

I. *Timing of the procedures*
We prefer to correct anterior chest deformities between the ages of 8–12, when the chest wall is still easily adaptable and the subperichondreal resection is easier to perform. However, we would not hesitate to operate well below or above this age. The authors do not share the opinion of those [44, 45] who suggest that the repair of pectus deformities should be delayed until puberty or later because of the danger of restricting the development of the juvenile thorax. Nevertheless, at any age care should be taken not to destroy the cartilaginous growth plates at the costochondral junctions, and to preserve the perichondrium thus, assuring the regeneration of the cartilages. Neglecting these guidelines may have grave consequences indeed, i.e., development of acquired thoracic dystrophy or arrested growth of the chest [46]. We believe that if the operation is performed properly, it could be done at any age and, in fact, have found the age limit imposed on small children especially unnecessary. However, we advise caution when operating on individuals past teenage unless psychologically or physiologically severely restricting anomaly exists. In adult patients, the operation is more challenging, but good cosmesis and functional improvement may still be achieved even in adulthood [18, 22].

II. Drainage of the wound
We recommend that in the course of pectus excavatum repair, the operative field should be drained by connecting the newly created substernal space with the right pleural cavity, which in turn is drained with a chest tube inserted through a separate stab-wound. We found that this maneuver virtually eliminated the vexing complication of subcutaneous seromas, often occurring despite subcutaneous drains. To adequately drain the substernal space, as well as the area under the subcutaneous flap, a large-pore mesh which allows the free passage of fluid should be used to uphold the sternum. If the pores on the mesh are not large enough, several additional large holes should be created.

In pectus carinatum, where the repair does not require substernal dissection, subcutaneous drains suffice.

Summary

The proper requirements for the modern surgical treatment of anterior chest deformities include the following:

Should be simple and swift.
Should not leave rigid foreign material in the body.
Should not create a potential for serious complications. Mortality is unacceptable.
Should provide uniformly excellent cosmetic and functional results.

Should not necessitate extended and frequent follow-up.
Should not require re-intervention.

Nothing short of these requirements should be accepted.

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Pectus Less Invasive Extrapleural Repair (PLIER): Saxena Technique

54

Amulya K. Saxena

Technical Highlights

Pectus Less Invasive Extrapleural Repair (PLIER) or the Saxena technique is an open procedure for the correction of the various forms of Chest Wall Deformities (CWD).

The main technical highlights of this procedure are:

(a) Sagittal incision in males and sub-mammary incision in females
(b) Dissection of the rectus muscle and exposure of the Xiphoid process
(c) Dissection of the pectoral muscles to expose the sternum and ribs
(d) Digital separation of the sternum from the pericardium and pleura
(e) Bilateral parasternal chondrotomy to release the sternum from the ribs
(f) Bilateral parasternal rib fractures in severe deformities
(g) Sternal wedge osteotomy and stabilization of the sternum using sutures
(h) Using of one trans-sternal metal strut (additional parasternal struts in severe asymmetry)
(i) Re-anastomosis of sternum to the rib cartilages
(j) Stabilization of the fractured ribs using figure-of-eight sutures
(k) Suture fixation of the rectus muscle to the sternum
(l) Suture fixation of the pectoral muscles

Indications

All types of Chest Wall Deformities can be corrected using this technique. However special indications for this technique are:

- Severely asymmetric Pectus Excavatum
- Severely asymmetric Pectus Carinatum
- Combined forms of Pectus Excavatum and Pectus Carinatum
- Chest Wall Deformities associated with Poland's Syndrome

Preoperative Considerations

1. The procedure should be offered and performed in patients >12 years of age as a general rule. Due to variations in body growth among teenagers, surgery should be reserved for teenagers whose body sizes have reached adolescent or adult proportions.

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2. The procedure should be performed in girls only after demarcation of the breast contours. This is mainly to avoid potential breast deformation that may result from submammary skin incisions in females operated before reaching pubertal changes.
3. Adult patients of any age with relatively stiff chest wall are well suited for this procedure.
4. Chest radiographs are taken to access the extent of the deformity.
5. Lung function tests are performed to evaluate the pulmonary function.
6. Electrocardiograms to diagnose or rule out cardiac pathologies.
7. Video-stereo-raster investigations.
8. Chest perimeter plots to quantify the deformity at three different points: manubrium, sternum and xiphoid.
9. Measurement of thorax diameter using a pelvimeter
10. Photographic documentation of the deformity
11. Coagulation tests are performed to rule out disorders in bleeding disorders
12. Since nickel is present in stainless steel struts, careful patient history is taken to rule out nickel allergy. Alternatively a nickel allergy test should be performed.
13. Antibiotics are administered on the day of the procedure and are continued for a period of 5 days after the procedure. Cefuroxime is the choice of antibiotic.

Special Instruments

Besides the instruments used for general thoracic surgical procedures, the following instruments are additionally required for this procedure (see Chapter on Instruments):

1. Stainless steel metal struts (Karl Lettenbauer GmbH, Erlangen, Germany)
2. Surgical toothed Pliers (Karl Lettenbauer GmbH, Erlangen, Germany)
3. Surgical Spanners (Karl Lettenbauer GmbH, Erlangen, Germany)

Surgical Technique

A vertical midline incision 7–10 cm long is preferred in males, while a submammary incision that is curved upward at the midpoint is preferred in females, in order to avoid the complications of breast deformity and impaired breast development. In the incision, skin, fat and pectoral muscles are reflected in a single flap and the entire dissection is performed with a needle-tipped electrocautery. The insertion of the rectus muscle to the xiphoid is exposed and the rectus muscle is severed to free the xiphoid. The attachment of rectus muscle to the lower costal cartilages is also dissected free. Precaution should be taken to avoid injury to the large subcostal vessels present between the rectus muscle and the caudal costal cartilage.

The xiphoid is then grasped using a Kocher clamp and retracted. Under traction, the sternum is freed from its anterior mediastinal attachments using careful digital preparation. The costal cartilages are then exposed and parasternal wedges of the deformed cartilages are resected by placing a perichondral elevator around the cartilage and incising it with a scalpel. Successive bilateral cartilage blocks are resected as the dissection proceeds cranially gradually freeing the sternum from its caudal to cranial attachments. The pectus deformity generally involves 5–8 pairs of ribs (third to tenth rib) and dissection is stopped once sufficient sternum has been freed from the distal costal cartilages, still remaining attached cranially. Precaution should also be taken to avoid injury to the internal mammary vessels. During the entire preparation, the integrity of the pleura is maintained. In case the pleura space integrity is compromised, a 5–0 vicryl suture is used to close

the opening. The deformed costal cartilages are additionally resected at the level of transition to the normal ribs to form obtain acceptable thoracic contours.

A partial transverse sternal wedge osteotomy is performed, if required, at the Angle of Ludovici. Once the sternum was dissected free as described, a perforated Hegemann steel strut (Lettenbauer, Erlangen, Germany) is passed trans-sternally. The distal part of the sternum is everted out of the thorax to pass the strut through it. This method is preferred to avoid injury to any intra-thoracic structure while passing the strut through the sternum using a hammer. The strut is then bent with metal clamps so that it fits the thorax wall perfectly at the edge of the impression. On achieving the desired shape of the strut, the strut and the sternum are returned back into the thoracic cavity. Two parasternal metal struts are also employed, with the points of fixation being the second rib and the lowest end of the rib cage. Heavy sutures are used to suture the cartilages as well as the trans-sternal strut to the ribs. Two parasternal struts are employed to provide anchorage to the mobile chest segments, which are formed as a result of double bilateral chondrotomy. The trans-sternal strut is secured to the two parasternal struts with PDS (polydiaxone) cords The pectoral muscle flaps and the severed rectus muscles were then sutured and fixed to the sternum. The overlying subcutaneous and cutaneous structures are finally united in the conventional manner to restore the normal chest wall anatomy.

Procedure Overview (Figs. 54.1, 54.2, 54.3, 54.4, 54.5, 54.6, 54.7, 54.8, 54.9, and 54.10)

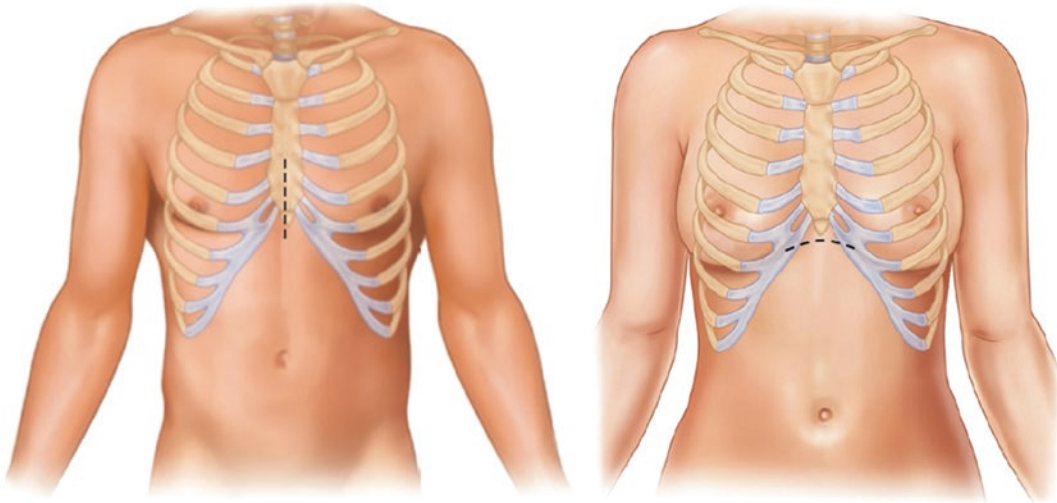

Fig. 54.1 Vertical midline incision are preferred in males (*left*) and submammary incisions in females (*right*)

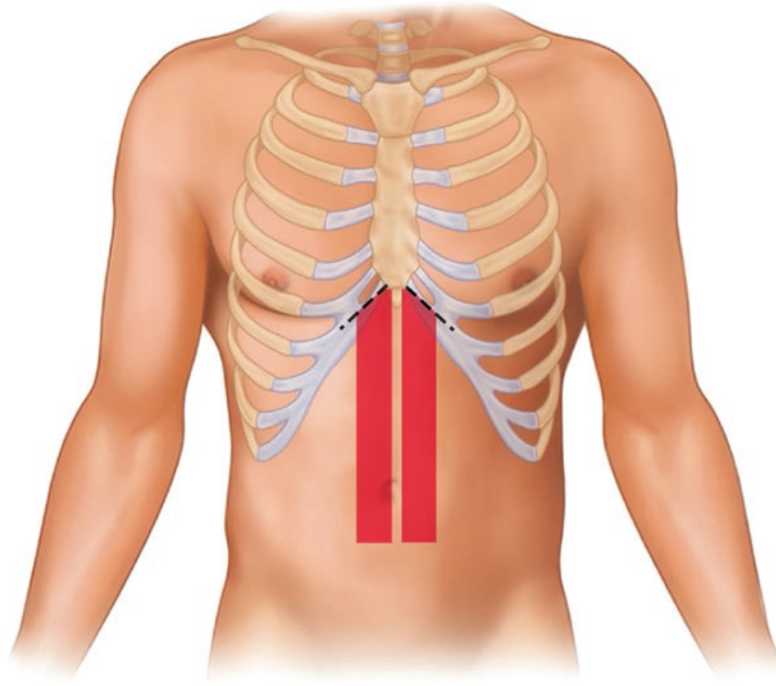

Fig. 54.2 The rectus muscle is severed from its attachments to the sternum to expose the xiphoid. The dissection is enlarged along the lower rib. Care should be taken to prevent injury to the lower subcostal vessels

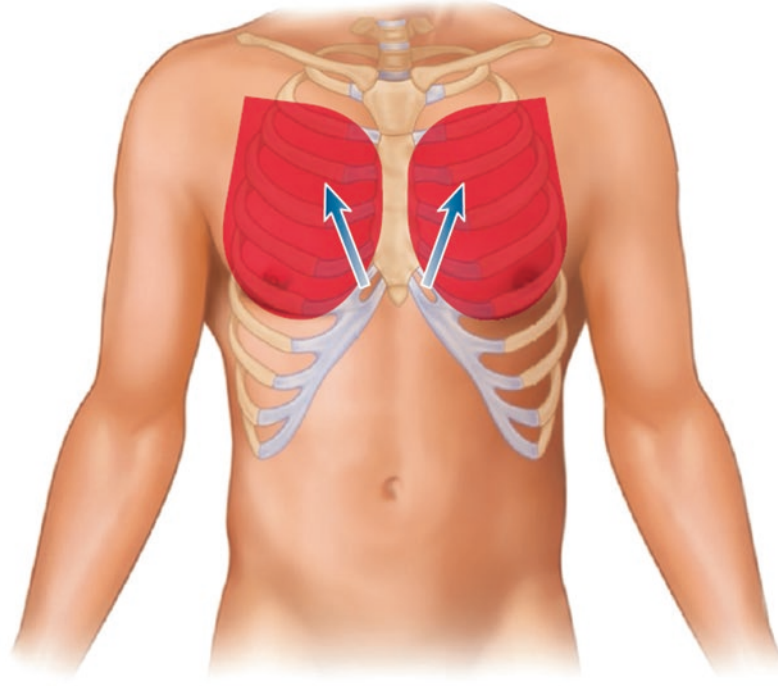

Fig. 54.3 En-bloc dissection of the pectoralis muscles is then performed to obtain maximum lateral exposure of the sternum and the ribs. The wound edges are secured with towels hitched to subcutaneous tissue

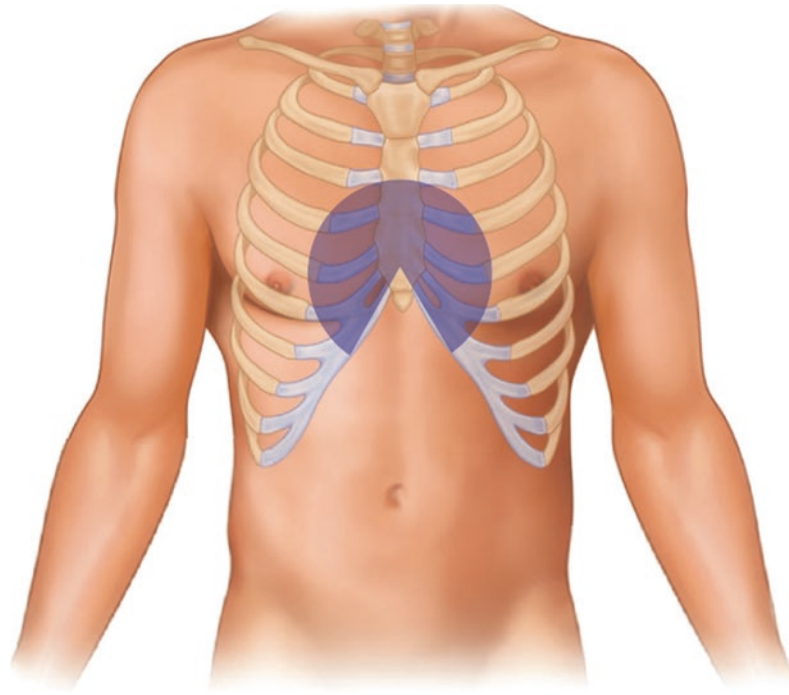

Fig. 54.4 The retrosternal tissue preparation is then performed. The xiphoid is secured with a clamp to provide retraction and facilitate digital tissue preparation. Precaution is taken to keep the pleura closed

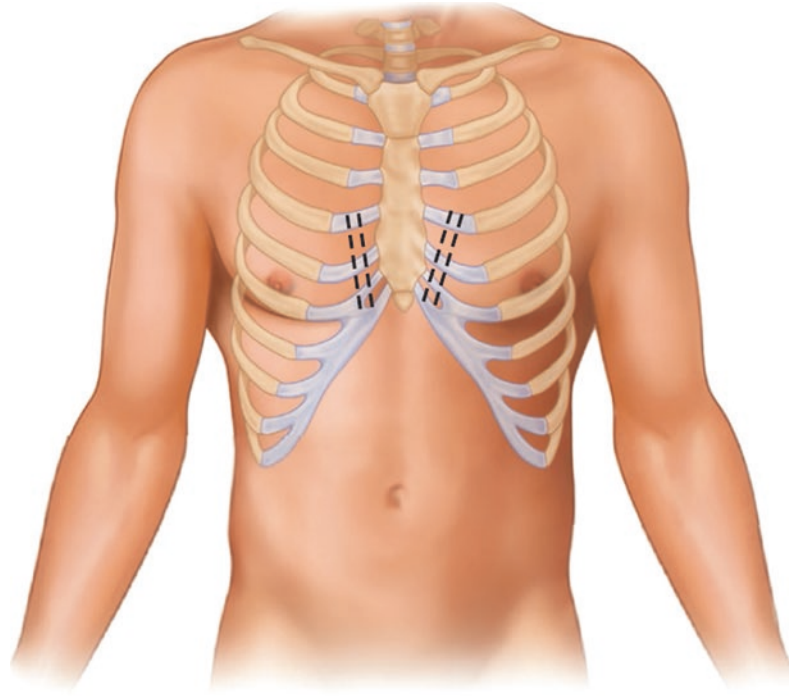

Fig. 54.5 Bilateral parasternal chondrotomy is then performed. The cautery is used to incise the perichondrium and 1 cm segments of cartilage are removed. The dissection proceeds from caudal to cranial with precaution taken to avoid injury to the Internal Thoracic Vessels

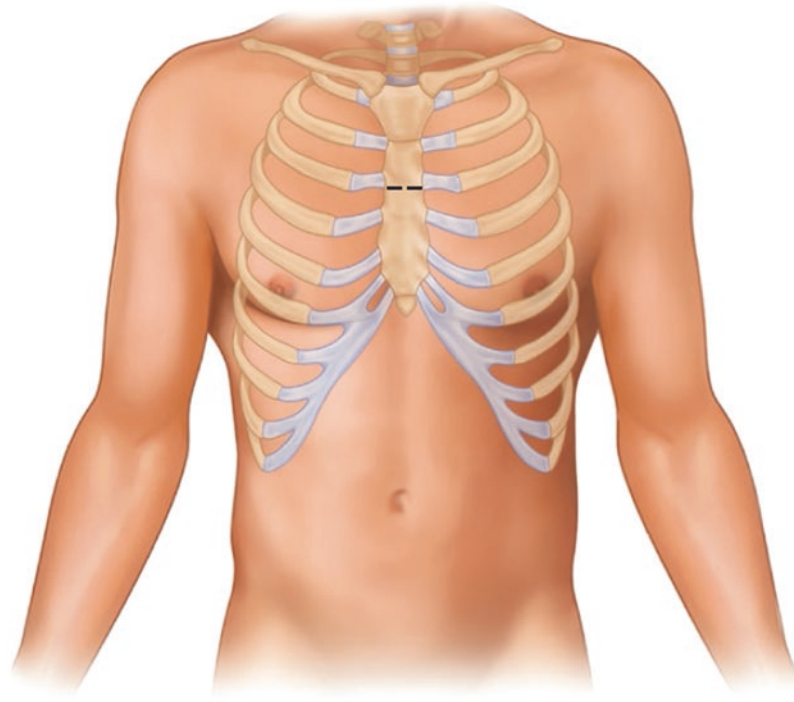

Fig. 54.6 Partial wedge sternal osteostomy is performed if necessary. The edges of the sternum after partial osteotomy are secured using braided non-absorbable suture. The sharp edges of the sternum are shaved using a scalpel or a chisel

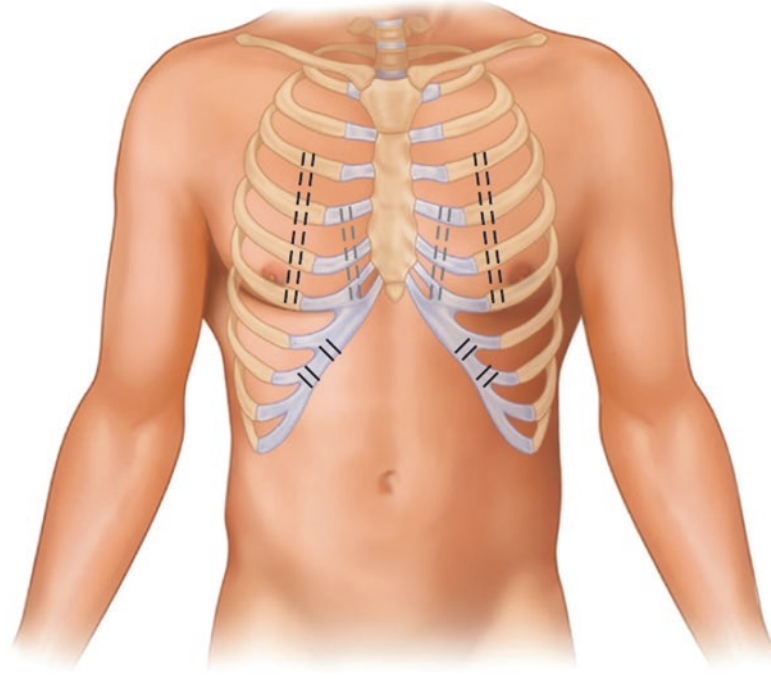

Fig. 54.7 Ribs are fractured when platythorax or lower rib eversions are present. However, lower rib eversions are corrected by chondrotomy and re-anastomosis of the cartilages

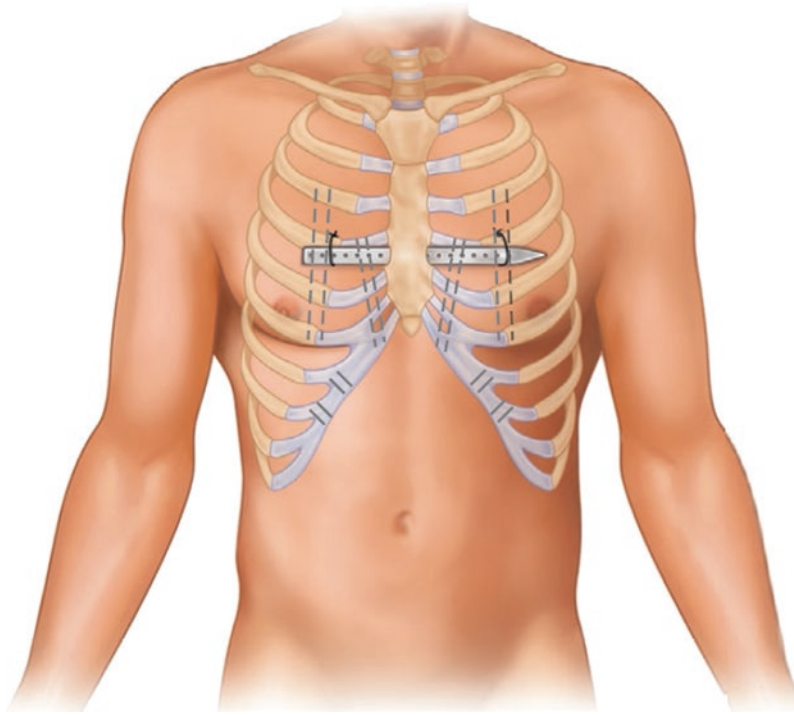

Fig. 54.8 A trans-sternal metal strut with a sharp-pointed edge is passed through the sternum using a hammer. A retractor is used on the other end to prevent injury during the passage. After placement the sharp edge of the strut is cut using pliers and blunted with a hammer

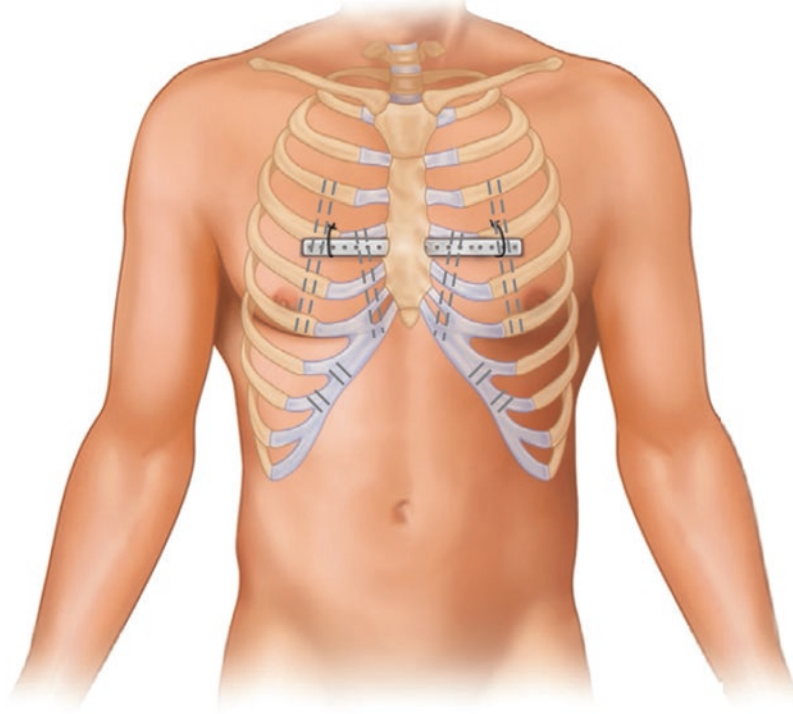

Fig. 54.9 The trans-sternal strut is secured to the underlying rib using PDS cords. The cartilages are sutured to the sternum from cranial to caudal direction using absorbable heavy sutures

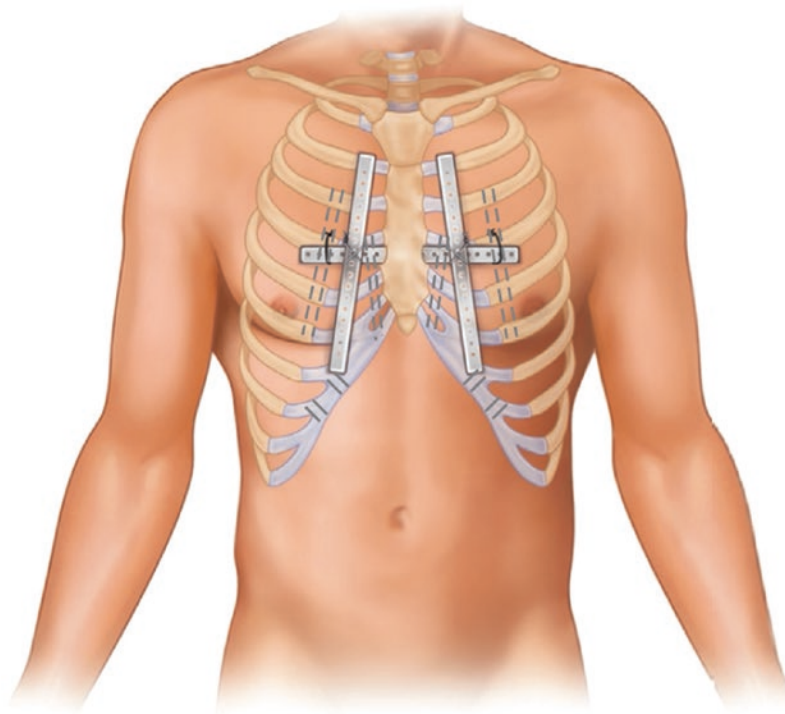

Fig. 54.10 Additional longitudinal para-sternal struts may be necessary in combined deformities or in platythorax or to stabilize the ribs if extensive correction is necessary. Longitudinal struts are placed on the ribs to which they are secured with heavy absorbable sutures. On the culmination points to the trans-sternal struts PDS cords are used to secure the struts

Postoperative Management

Perioperative antibiotic therapy was administered; with Ceftriaxone being the drug of choice. All patients were also administered strong analgesics for first 24 h. The patients are mobilized on the second day after surgery. On the fourth day pulmonary exercises are commenced. Since there was no compulsion to discharge the patients on the third day, patients were kept in hospital to follow pain and recovery after this procedure for 1 week. Patients who preferred to be discharged earlier were permitted to leave the hospital. At the time of discharge patients were advised to avoid body contact sports until the struts had been removed. Regular swimming was encouraged along with light athletic exercises. The patients were followed-up at regular intervals (3, 6, 12 and 15 months) after the procedure. The struts are removed after a period of 12–15 month.

Results

The PLIER procedure was carried out on 25 PE and 18 PC patients with a median age of 15.7 years (range 13–19 years). The procedure was carried out on 29 males and 6 females who presented with Type-1 (13 patients), Type-2 (12 patients), Type-3 (4 patients) and Type-4 (14 patients) pectus deformity according to Willital's classification. The mean size of the incision was 7.5 cm (7–10.5 cm). The mean operation time was 85 min (75–115 min). The procedures were completed with three struts and two metal wires in all the patients. Wound drains were placed in 31 patients and were removed after 48 h. There were no intraoperative complications and no blood transfusions were required. In 37 patients, the pleura integrity was preserved, however in 6 patients the pleura was extremely thin and accidentally opened during preparation and had to be sutured. Post-operative x-ray films demonstrated a minimal pneumothorax in these patients, but no drainage was required.

None of the patients required chest tubes during or after the procedure. Strong analgesics were administered for 2 days and the patients were mobilized on the second day. On the third post-operative day a chest ultrasound was carried out on all the patients. Pleural effusions were not observed in any of the patients. Pain medication (acetaminophen) was not required in most of the patients after 3 days, however 3 patients required

pain medication for 5 days. Antibiotics were administered for 1 week and no wound infections were observed.

The follow up period ranged from 6 months to 2 years. The struts were removed after a period of 15 months and no recurrences after strut removal were observed. Hypertrophic scars were seen in 8 patients and were corrected at the time of strut removal. There were no recurrences in the series and all patients were satisfied with the esthetic results of the procedure and were followed up 3, 6 and 12 months after the procedure. The 43 patients graded their operation from very good to excellent. The follow up investigations included (a) measurement of thorax diameter using a pelvimeter (b) electrocardiogram (c) pulmonary function tests, (d) chest ultrasound, (e) clinical photograph and video-stereo-raster examinations in 35 patients.

Procedure Related Images

(Figs. 54.11, 54.12, 54.13, 54.14, 54.15, 54.16, 54.17, 54.18, 54.19, 54.20, 54.21, 54.22, 54.23, 54.24, and 54.25)

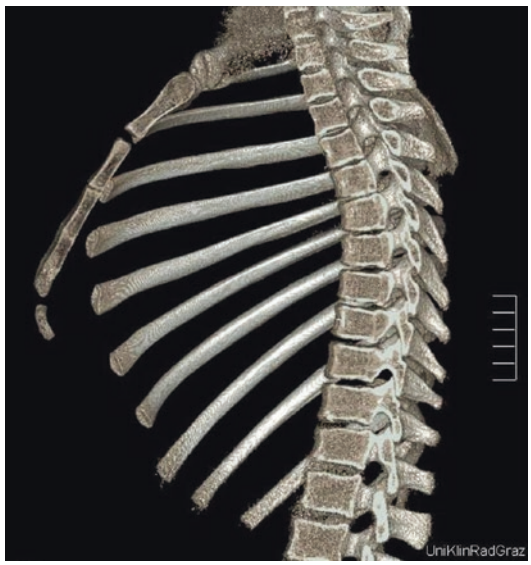

Fig. 54.11 Preoperative 3-D computed tomography image of a 14-year old male patient with a severe pectus carinatum deformity who underwent a PLIER correction

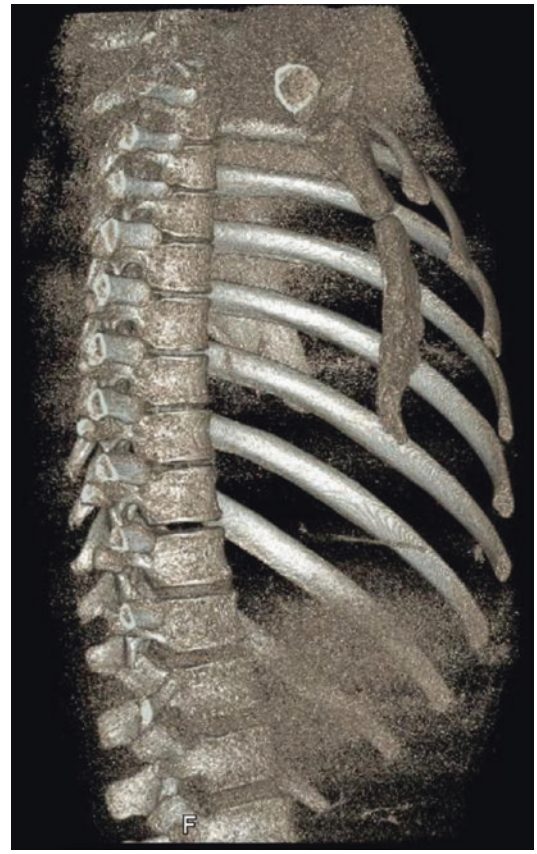

Fig. 54.12 Preoperative 3-D computed tomography image of a 15-year old female patient with a severe asymmetric pectus excavatum deformity corrected with the PLIER technique

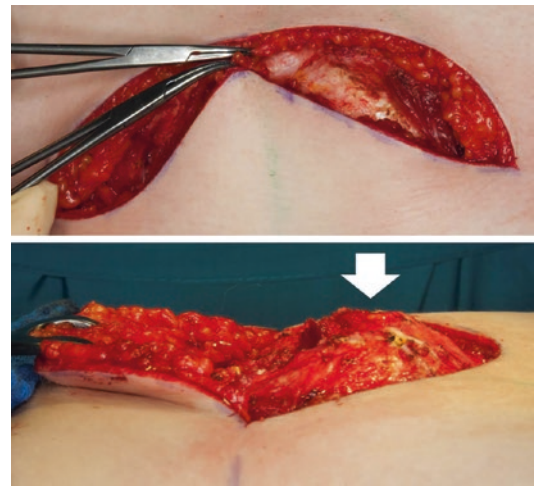

Fig. 54.13 A submammary incision (*top*) in a 25-year old patient with a combined pectus carinatum-excavatum deformity. Lateral view during surgery (*below*) showing the elevated sternum (*arrow*) positioned above the level of the ribs

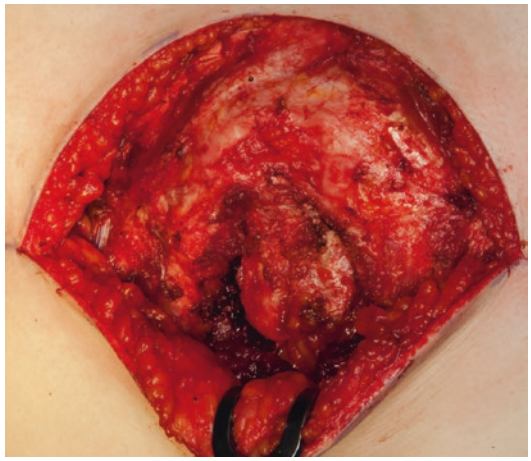

Fig. 54.14 Exposure of the Xiphoid which is depressed by the sternal evelevation

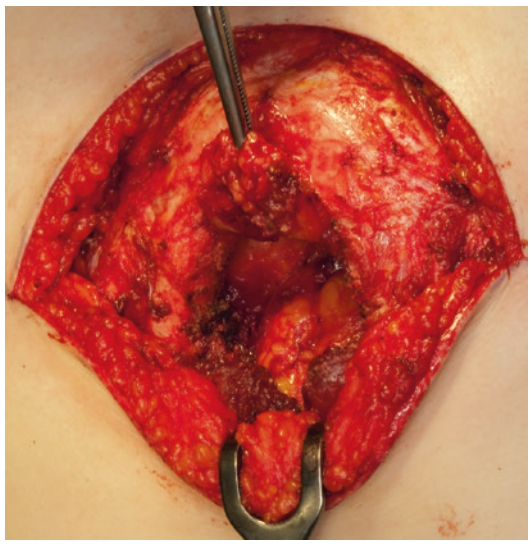

Fig. 54.15 The rectus muscle attachments are then dissected to free the Xiphoid and the edges of the lower ribs to permit blunt dissection of the retrosternal extrapleural space

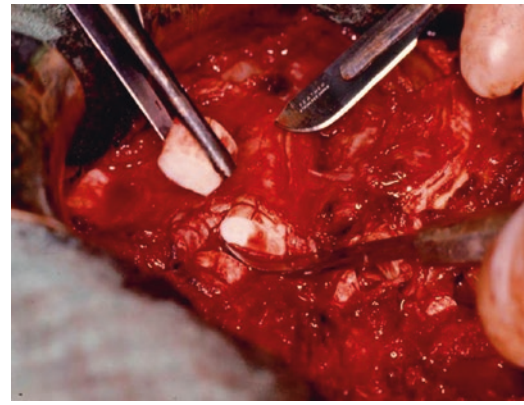

Fig. 54.16 En-bloc bilateral resection of costal cartilages is performed using a scalpel with removal of cartilages from the caudal to cranial direction

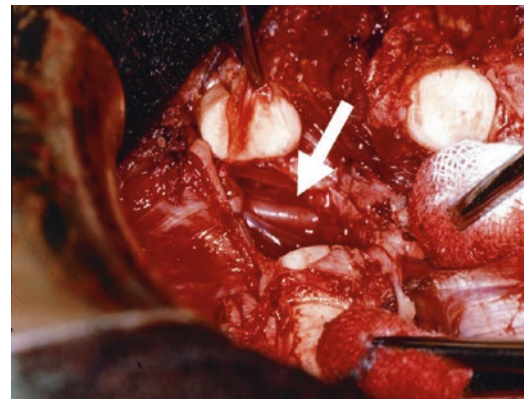

Fig. 54.17 Precaution should be taken to identify the internal thoracic vessels (*white arrows*) which run just below the line of dissection so as to avoid injury to them during cartilage resection

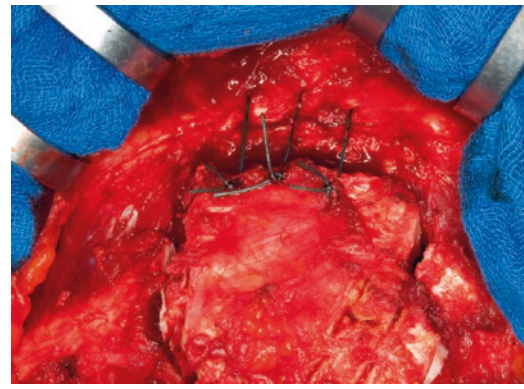

Fig. 54.18 The elevated sternal deformity is corrected by a wedge osteostomy, after which the partial sternotomy edges are secured using interrupted non absorbable suture material

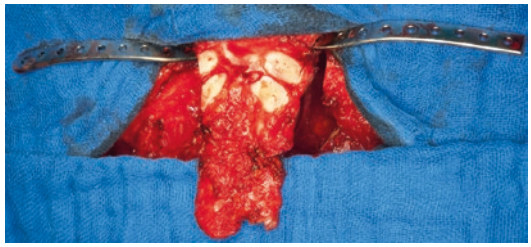

Fig. 54.19 The edges of the costal cartilages approximated to the sternum are shaved to achieve desired contours and a stainless steel strut is passed through the sternum and bent appropriately according to the chest wall countours

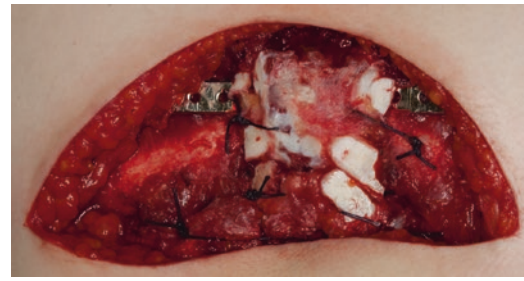

Fig. 54.20 The trans-sternal strut is secured to the ribs with absorbable and nonabsorbable suture. The edges of the cartilage after resection are readapted using absorbable sutures

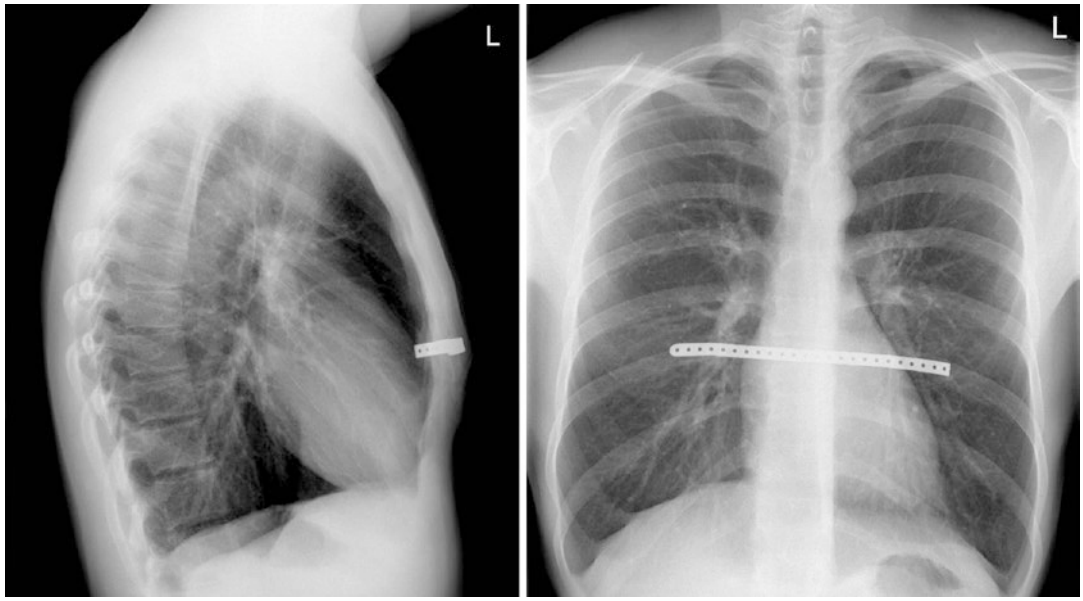

Fig. 54.21 Postoperative films demonstrating the position of the strut in a patient with severe pectus carinatum corrected using the PLIER technique (*left image*-lateral view, *right image*-anteroposterior view)

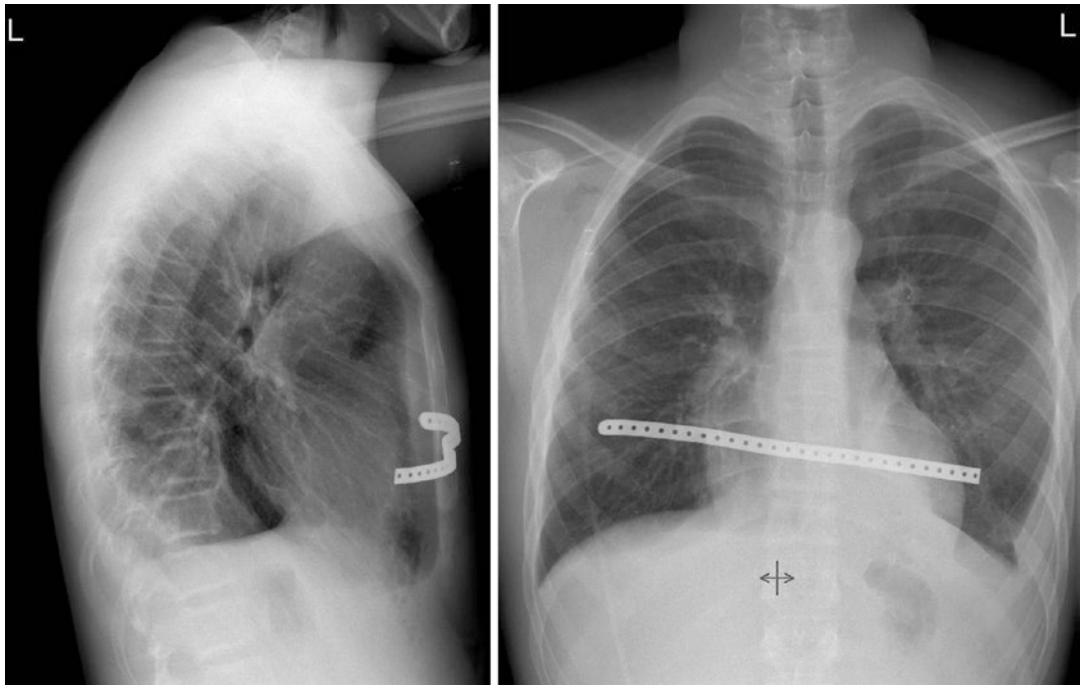

Fig. 54.22 Postoperative films demonstrating the position of the strut in a patient with severe asymmetric pectus excavtum corrected using the PLIER technique (*left image*-lateral view, *right image*-anteroposterior view)

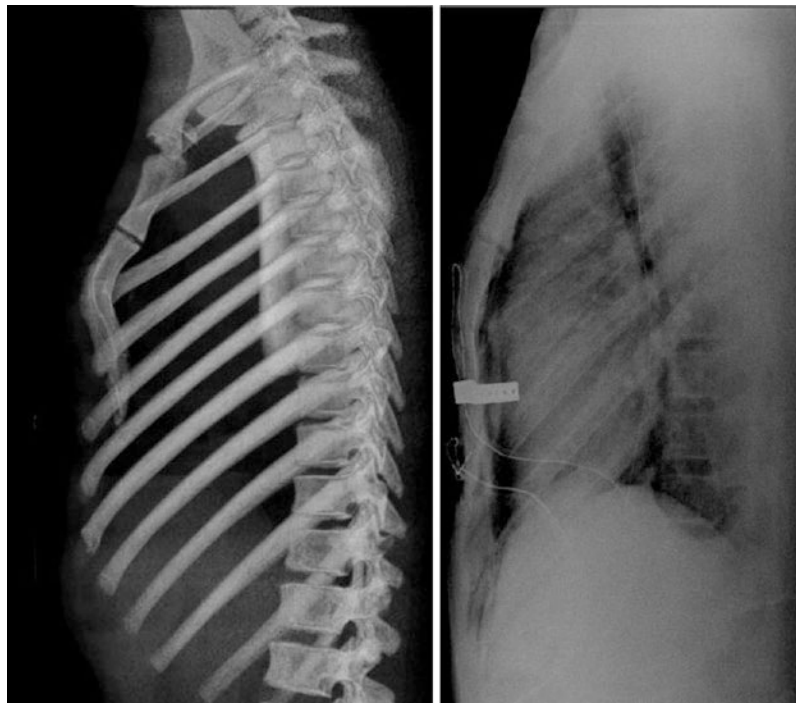

Fig. 54.23 Preoperative view (*left image*) of a combined pectus carninatum (cranial) excavatum (caudal) deformity. Immediate postoperative film (*right image*) demonstrating the results of PLIER correction and the position of the strut. Subcutaneous drains were placed (seen in *right image*) were removed after 24 h

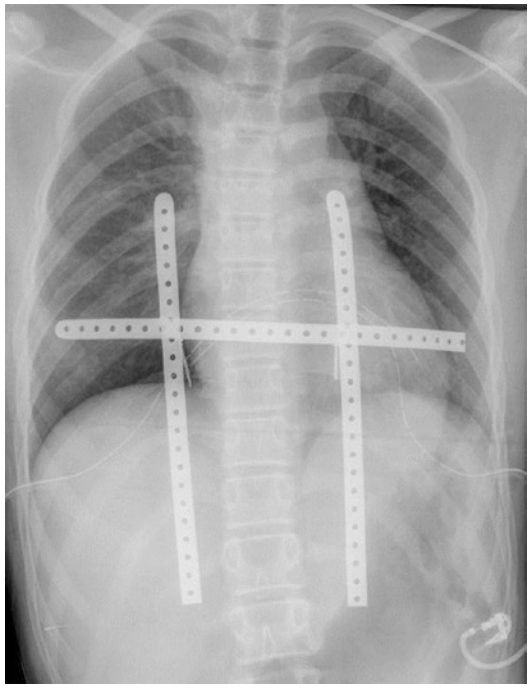

Fig. 54.24 Postoperative film (anteroposterior view) demonstrating the position of the struts in a patient with severe combined asymmetric pectus excavtum- pectus carinatum with platythorax corrected using the PLIER technique. Additional struts are required to stabilize the flail segment of the thorax which are created as a result of rib osteotomies to achieve desired chest wall contours

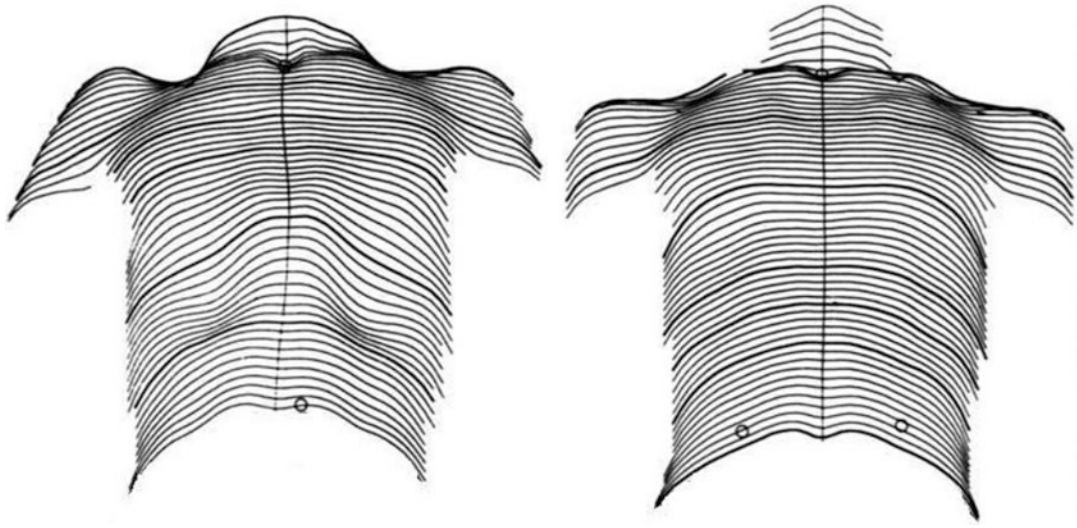

Fig. 54.25 Video-stero-raster results of PLIER procedure in pectus carinatum. Preoperative (*left*) and postoperative (*right*) findings

Suggested Reading

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Part VIII

Other Chest Wall Deformities: Rare Chest Wall Deformities

Poland's Syndrome

55

Amulya K. Saxena

Introduction

Poland's syndrome is a rare congenital anomaly characterized by hypoplasia of the breast and nipple, scarcity of subcutaneous tissue, absence of the costosternal portion of the pectoralis major muscle, lack of the pectoralis minor muscle, aplasia or deformity of the costal cartilages or ribs II to IV or III to V, alopecia of the axillary and mammary region, and unilateral brachysyndactyly [1] (Figs. 55.1 and 55.2). The presence of these manifestations may vary between patients and it is extremely rare for all the above individual manifestations to be present together [2, 3] (Fig. 55.3). Poland's syndrome almost exclusively occurs unilaterally, although one bilateral case has been reported in the literature [4]. Besides this, there are no sorts of correlation between the extent of chest wall, breast and hand deformities involvement [5, 6].

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Incidence

Poland's syndrome has an incidence that ranges from 1:7,000–100,000, with predominance in males 2–3:1 [7, 8]. In almost 60–75 % of the patients, the right side of the body is involved. Poland' syndrome can be sporadic or have a familial incidence. Sporadic cases have a male predominance with the right side affected more than twice in males. Whereas in sporadic cases that involve females, there is almost an equal affection with regards to the side involved. On the other hand, in familial cases, the incidence of Poland's syndrome in males and females is equal and without a right-side predominance [8–10].

Etiology

Poland's syndrome is a not a genetic congenital disorder and has an extremely low risk of familial reoccurrence [11, 12]. The vertical transmission from parents to children or between affected siblings with normal parents is related to a delayed mutation of an autosomal dominant gene which may have a higher chance of occurrence surprisingly with increased paternal age [13–16].

The etiology of Poland's syndrome is best explained by the theory that at the end of the sixth week of gestation, when the upper limb bud adjacent to the chest wall is still in a stage of development, there is an interruption of the embryonic

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blood supply which causes hypoplasia of the ipsilateral subclavian artery or one of its branches. The site and degree of this impairment in blood flow may determine the extent and the severity of the anomaly. Also, in case of hypoplasia of the internal thoracic artery occurs, this could lead to the absence of the sternocostal portion of the pectoralis major muscle; on the other hand such a hypoplasia of the brachial artery may lead to hand abnormalities [10, 17].

Fig. 55.1 Spectrum of tissue involvement in patients with Poland's syndrome

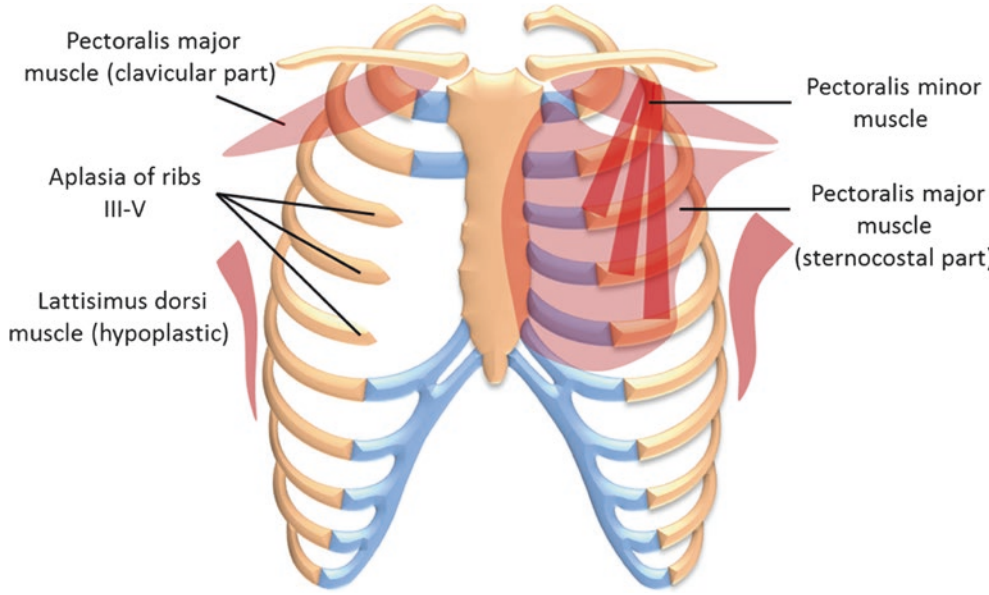

Fig. 55.2 Schematic representation of Poland's syndrome manifestations in the thorax

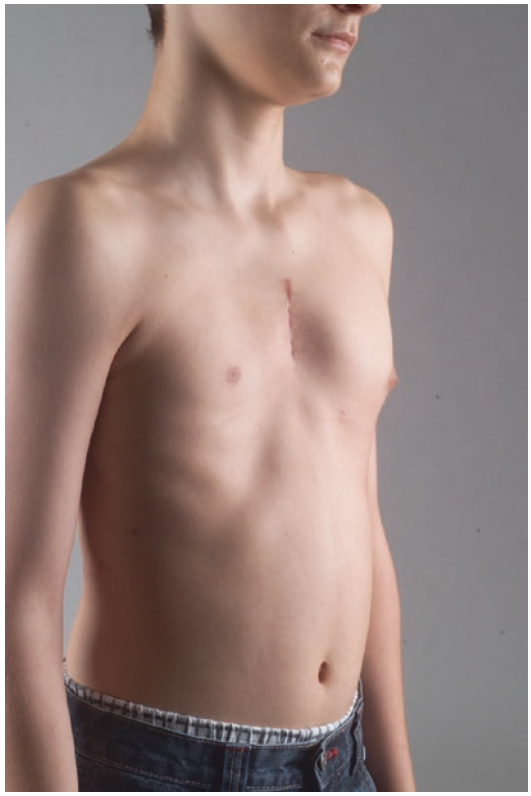

Fig. 55.3 Poland's syndrome in a 16-year old male patient with right side chest wall affection

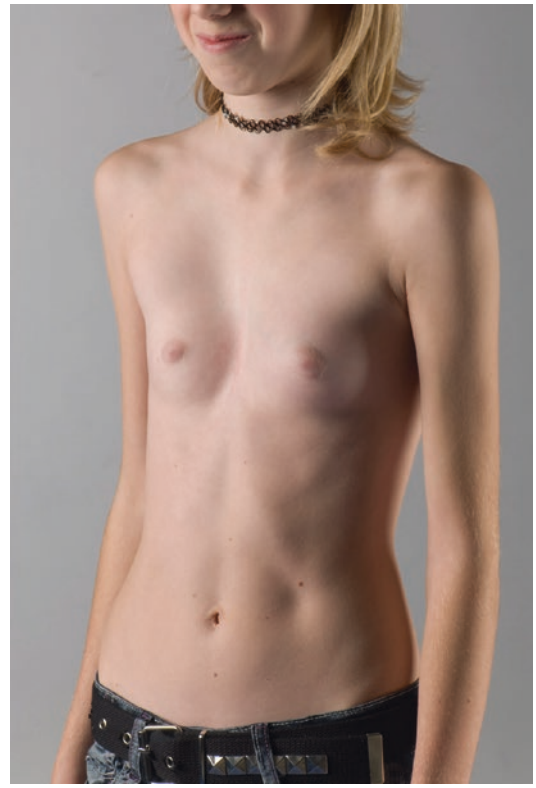

Fig. 55.4 Poland's syndrome in a 14-year old female patient with chest wall and breast involvement on the right side

Another theory points to the disruption of the lateral plate mesoderm from which the pectoralis muscle develops, an event which occurs between 16 and 28 days after fertilization may be responsible for the defects [9]. Besides these theories, a variety of other factors, such as autosomal dominant inheritance, gene defects, trauma, viral infection, intrauterine insult following attempted abortion, the teratogenic effects of environmental substances, or maternal smoking during pregnancy have also been implicated in the etiology of Poland's syndrome [18–20].

Clinical Features

Poland's syndrome is characterized by the absence of the sternocostal head of the pectoralis major muscle. The pectoralis minor muscle may also be absent in number of patients. The absence of these muscles does not necessarily translate into functional impairment of the affected side in the patients [21–23]. Some patients may also present with the affection of other muscles such as the latissimus dorsi, external oblique, and serratus anterior. There are in fact large variations in the presence or absence of these structures, and three-dimensional computed tomography has been found to be an ideal investigating method to reveal the details of individual patient [24].

Breast involvement is present in more than a third of female patients and could range from mild hypoplasia to amastia, which is the complete absence of breast tissue [25] (Fig. 55.4). The nipple and areola are usually hypoplastic or even absent, a condition which is known as athelia. Another feature of Poland's syndrome is the presence of supernumerary nipples. A recent report confirmed that the hyoplastic breast can lactate normally and can offer normal breast feeding function [26].

Most patients clinically present with a unilateral depression of the chest wall which results

from hypoplasia of the ribs and cartilages. Ribs II to IV, or III to V are most frequently involved, but the second rib is less affected (Fig. 55.5). In almost one-fourth of the patients, a severe chest wall depression occurs with aplasia of the anterior portions of between one and three ribs (Fig. 55.6) [25, 27]. Rib defects are also evident in 15 % of patients who present with pectoralis major defects on the right side [28]. In some patients, the sternal ends of aplastic ribs may be fused together which leads to rotation of the sternum towards the aplastic ribs and clinically manifests as an asymmetrical pectus carinatum type of deformity.

Isolated pectoral hypoplasia is a variety of Poland's syndrome [29]. Hence, women seeking mammary augmentation for breast asymmetry should be evaluated for the isolated absence of the pectoralis major muscle with breast hypoplasia, which is not an uncommon in Poland's syndrome. Patients without hand involvement are more common than Poland's syndromes patients which exhibit most of the manifestations and are referred to as "partial Poland's sequence"[21].

With regards to pulmonary involvement, chest films in Poland's syndrome patients demonstrate a hyperlucent lung on the involved side. Lung herniation is observed in small subset of patients, in whom paradoxical motion of the thoracic cage may be of clinical significance during stress or severe pulmonary infections. In patients with Poland's syndrome the vital capacity is reduced and is found to be half of what is observed in normal patients; improvement of which has been reported after surgical correction [30].

With regards to cardiac manifestations, dextrocardia has been reported in patients with Poland's syndrome, which in fact appears to be a dextroposition without inversion associated with rib defects on the left-side [28, 31]. Unlike isolated dextrocardia, dextrocardia in Poland's sequence is not associated with other cardiovascular anomalies. It has been estimated that around 5.6 % patients with Poland's syndrome have dextrocardia, which may be as high as 9.6 % in patients in patient with left-side defects [10]. There is an increased occurrence of dextrocardia among the children of consanguineous parents [28, 32].

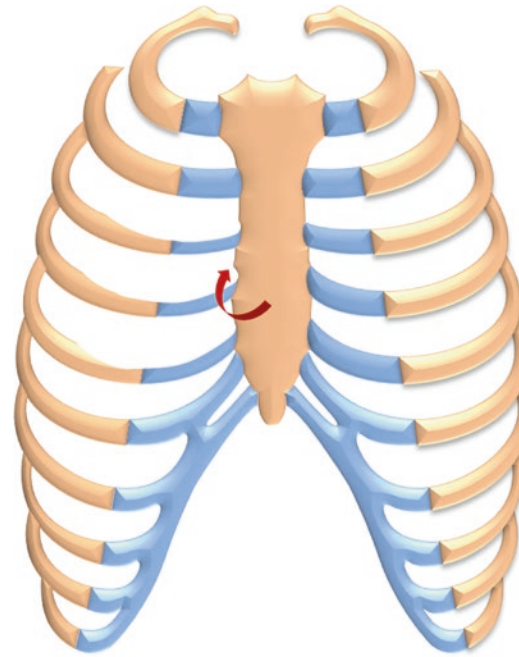

Fig. 55.5 Schematic view of chest involvement in moderate form of Poland's syndrome showing chest wall depression with hypoplasia of ribs III to V and mild torsion of the sternum

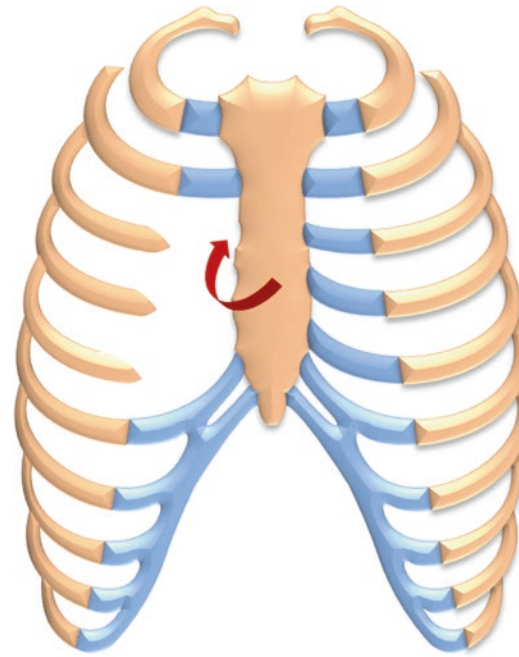

Fig. 55.6 Schematic view of chest involvement in severe form of Poland's syndrome showing chest wall defect with aplasia of ribs III to V and rotation of the sternum

Hand abnormalities in Poland's syndrome are unilateral and present on the same side of chest involvement. Hand involvement contralateral to the chest deformity is rare [19, 33]. Hand deformities vary from shortness of the middle phalanges (hypoplasia or aplasia) with cutaneous webbing (syndactyly), to complete absence of the hand (ectrodactyly), and their occurrence in Poland' syndrome patients is variable [21, 29]. Aplasia of the middle phalanges of the hand is more common on the ulnar side than on the radial side. The thumb may appear disproportionately small and malrotated. Involvement of the lower extremity in Poland's syndrome is very rare [34].

Poland's syndrome is associated to Möbius syndrome, which is a bilateral congenital facial nerve palsy with paralysis of the ocular abductor muscles [35]. Patients with facial nerve palsy, paralysis of ocular abductor muscles, absence of pectoral muscles and ipsilateral hand deformities constitutes the Poland's–Möbius syndrome [36, 37]. Poland's–Möbius syndrome can have a familial etiology [38]. Rarely, atrial septal defects can be also associated with Poland's–Möbius syndrome [39].

Few patients with Poland's syndrome may present with elevation and winging of the scapula which is described as Sprengel's deformity or scapula alata [10]. This is due to the absence of the upper portion of the serratus anterior, which probably is a result of decreased of blood flow in the suprascapular arteries.

Poland's syndrome may also be associated with Klippel-Feil syndrome, in which patients exhibit a characteristic short neck due to fusion of the cervical vertebrae and to abnormalities of the brainstem and cerebellum [10]. Klippel-Feil syndrome could be attributed to a delay in the development of the vertebral arteries.

An association of aplasia of the pectoralis major muscle with renal anomalies has been identified and termed as an acro-pectoral-renal field defect [40]. Renal anomalies such as unilateral renal agenesis or duplication of the urinary collecting system occur as part of this defect and may be responsible for renal hypertension.

Associations between tumors and Poland's syndrome has been established which can be best explained by the abnormal homeobox and tumor suppressor genes present in the patients affected [41]. Poland's syndrome patients have shown associations with non-Hodgkin's lymphoma, leukemia, leiosarcoma, cervical cancer, and lung cancer [42]. Invasive ductal carcinoma has also been reported in the hypoplastic breasts of patients with Poland's syndrome [43].

Surgical Management

There are no absolute indications for surgical management of the unilateral chest wall deformity in patients with Poland's syndrome. Relative indications to offer surgery to these patients include:

1. Possibility of its progression of the unilateral depression of the chest wall
2. Inadequate shielding of the thoracic organs
3. Paradoxical chest wall movement
4. Plastic surgical reconstruction of the affected female breast
5. Esthetic defect due to absence of pectoralis major muscle and axillary fold in males

The plan for surgical strategy for the treatment of Poland's syndrome should be based on the severity of the deformity, the age of the patient and the sex of the patient. Surgical procedures may be a single procedure or may be offered as two-stage procedures.

In children with mild forms of Poland's syndrome, which is limited to absence of the pectoralis muscles and breast hypoplasia, the single surgical management should be performed after the patient has reached puberty. In females patients this could be combined with the reconstruction of the breast. If severe aplasia of the ribs and pronounced depression of the chest wall is present, surgical management should involve a two-stage procedure in children. In the first stage, the rib defect should be reconstructed, this should be followed by the second procedure of muscle flap transposition which should be performed should after puberty. In female patients the second procedure may be combined again with the reconstruction of the breast.

In adults, a single-stage procedure which involves reconstruction of the chest wall along with muscle transposition is preferred. In female

patients, augmentation mammoplasty may be performed as part of this single-stage procedure [25].

Surgical Procedures

Ribs and Chest Wall Reconstruction

In the presence of unilateral rib deformities, correction procedures such as the Willital-Hegemann procedure or the PLIER technique could be beneficial. These procedures in Poland's patients involve the minimal resection of the sternochondral cartilage, partial rib wedge osteotomy, sternal detorsion with or without partial sternal wedge osteotomy and re-adaptation with heavy non-absorbable sutures and the stabilization of the reconstructed thorax with struts [44, 45]. Besides the above procedures, costal cartilage-sparing modified Ravitch procedure can also be employed in these patients [46]. At the time of strut removal which is approximately 15 months after the procedure, muscle augmentation or breast reconstruction can be performed in the same procedure. Minimal access repair of the chest wall could be another option in these patients [47].

In the presence of large rib defects that are associated with lung herniation, the chest wall can be reconstructed using subperiostial split rib grafts from the unaffected contralateral side, bone grafts or polypropylene mesh-patch (Fig. 55.7). If bone grafts are utilized, the medial ends of the grafts are to be sutured into caverns made into sternum edge. The grafts are attached on the lateral ends to the freshened surface of the ends of the ribs. The reconstruction of the ribs and cartilages has three advantages, in that enables the anatomic correction of the defect, prevents paradoxical respiration and provides a base for a muscle flap or for a prosthetic implant. If mesh is used alone, it should be stretched taut and sutured to the edges of the defect. If a combination of bone grafts and mesh is used, the mesh should be sutured to the rib grafts as well. If a sternal rotation is present, this should be corrected by performing a transverse sternotomy and readaptation of the sternum with non-absorbable sutures (Fig. 55.7b) [48]. Transfer of the latissimus dorsi muscle and the implantation of breast prosthesis may be performed at the same time.

If the defect is limited to two ribs, the normal ribs may be split above and below the defect and attached to the ends of the aplastic ribs (Fig. 55.8). If the ribs are fused at their sternal ends, they are to be detached and then reattached to the sternum [49].

In patients with hypoplastic ribs and chest wall depression, the procedural choice is to per-

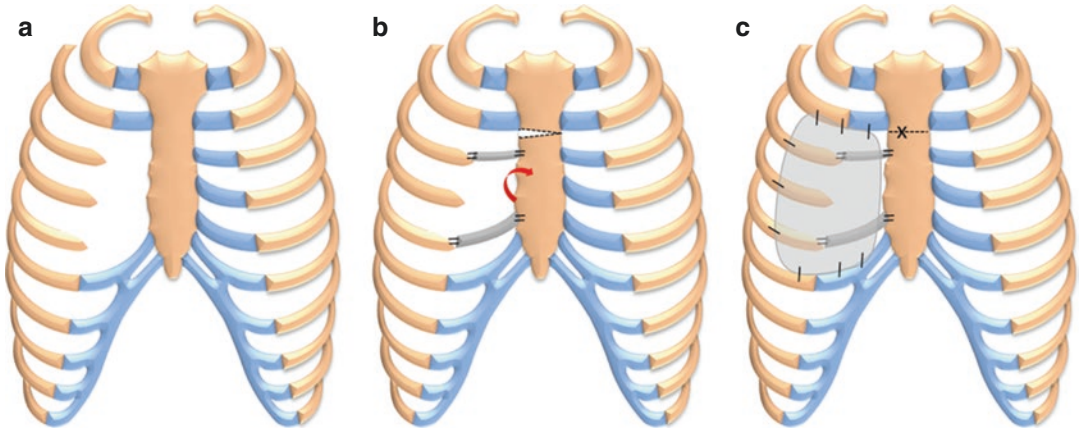

Fig. 55.7 Schematic view of operative repair of a severe form of Poland's syndrome with aplasia of ribs III to V and sternal rotation. (**a**) Split rib grafts are harvested from the contralateral side. (**b**) These grafts are sutured into previously created sternal notches on the medial side, and to the ends of the aplastic ribs on the lateral side. Sternal rotation is corrected by osteotomy and secured with a figure-eight suture. (**c**) If a prosthetic mesh is used it is sutured on top of the rib grafts and to the edges of the defect

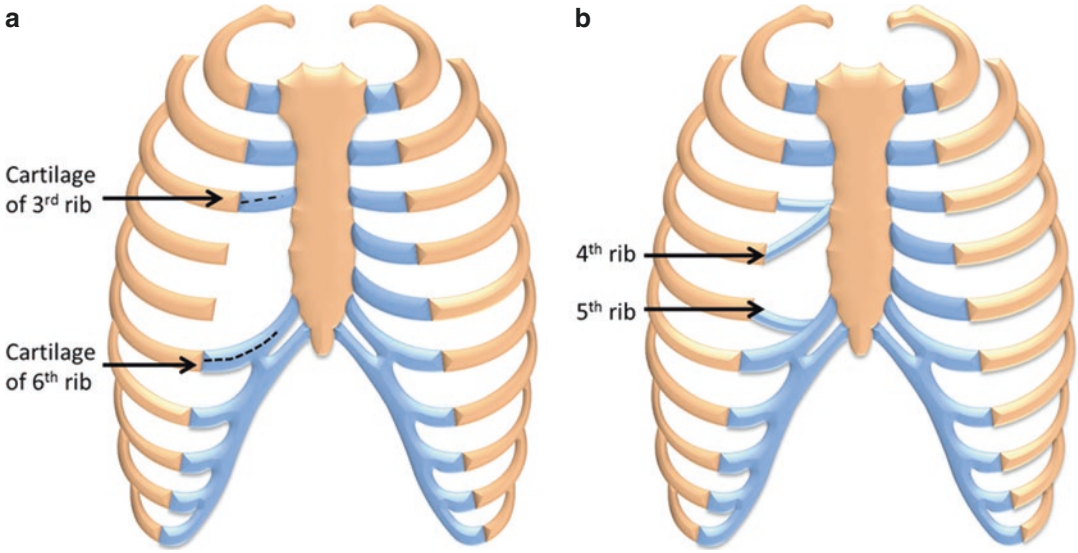

Fig. 55.8 Schematic view of surgical correction of Poland's syndrome with aplasia of the anterior portion of two ribs. (**a**) Anatomic situ and proposed lines of incisions. (**b**) Normal ribs cranial and caudal to the defect are split and sutured to the ends of the aplastic ribs

form a subperichondrial resection and elevation of the involved costal cartilages. If necessary, this could be done with mesh reinforcement.

Muscle Flaps

In female patients, prosthetic implants or musculocutaneous flaps could be employed to augment hypoplastic breast [50]. These procedures should be done only after puberty so that the reconstruction of the size of the affected breast is proportional to the size of the contralateral normal fully developed breast. A musculocutaneous flap is preferred over a simple muscular flap because the skin and subcutaneous tissues are generally extremely thin and not strong enough to accommodate a considerable volume of transferred muscle and prosthesis beneath them (Fig. 55.9) [51]. Early attempts using free latissimus dorsi flaps were largely unsuccessful because of atrophy occurring due to omission of a neurovascular pedicle [49].

Another option here could be the transplantation of a latissimus dorsi musculocutaneous flaps from the contralateral side to correct the loss of the anterior axillary fold and muscle mass. However, such muscle The auto-transplantations have the disadvantage of creating an additional thoracic scar, as well as taking away one of the major muscles of the shoulder and arm on the unaffected side [6]. An endoscopcially assisted minimally access reconstruction technique could be a better option to reduce donor site morbidity [52].

The state of the latissimus dorsi muscle is important in determining the success of the surgical approach. In patients in which the hypoplastic latissimus dorsi was replanted, partial skin necrosis was also reported [53]. Latissimus dorsi can be present in a hypoplastic form as an avascular fibrous sheet in the presence of a normal posterior axillary fold [54], hence in order to avoid surgical pitfalls the latissimus dorsi must be carefully evaluated by computed tomography or magnetic resonance imaging scans [55]. If the latissimus dorsi muscle is hypoplastic or absent, microsurgical transfer of the contralateral latissimus dorsi would be a feasible option. Upper gluteal flaps, microvascular-free transverse rectus abdominis muscle (TRAM) flaps nourished by the internal thoracic vessels as recipient vessels or transverse musculocutaneous gracilis (TMG) flap could be other options [56–58].

Lipomodeling has recently been proposed as an option to augment breast in Poland's syndrome

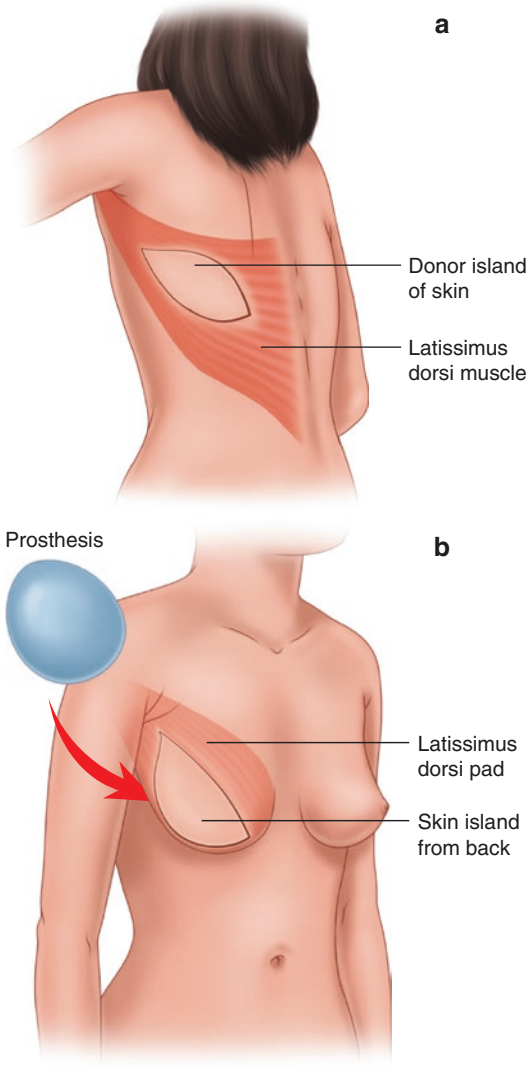

Fig. 55.9 Schematic view of single stage reconstruction of the chest in adult patients with Poland's syndrome. (**a**) Dorsal view of myocutaneous flap with preserved thoracodorsal neurovascular pedicle, translocation of the latissimus dorsi flap through an axillary tunnel, and suture of the anterior chest wall skin to the transplant. (**b**) Frontal view of myocutaneous flap reconstruction with preservation of the thoracodorsal neurovascular pedicle, translocation of the latissimus dorsi flap through an axillary tunnel, and suture of the anterior chest wall skin to the transplant

using surgical procedures as well autologous injections of fat [59, 60]. Another promising option with lipomodeling that has recently been reported is the omental flap technique which is performed using minimal access surgery and offers superior amelioration of the deformities caused by Poland's syndrome when compared with other reconstructive options [61]. There were initial concerns with lipomodeling, that implanted fat tissue may comprise breast cancer detection in female patients, however there has been no evidence to confirm this hypothesis [62].

In some patients with large rib defects and muscle hypoplasia, a lone transferred muscle flap could be inadequate to achieve the desired chest wall contours [50, 63]. For such patients, a prefabricated chest wall implant could be an feasible option [63].

Hand Reconstruction

Hand reconstruction depends on the severity of the hand deformity. The surgical procedures must be based depending on the type of deformity based on the possible option available. Parents of these children should be counseled with regards to their expectation that could be associated with hand reconstruction.

Hand reconstruction, especially the separation of the fingers should be performed in the early years of infancy, preferably in the first year of life. The main reason for this is to achieve correction before the development of abnormal function patterns of the fingers and before progression of the deformity. Restoration of some hand function in patients with ectrodactylia is possible either by toe-transfer or by implanting prosthesis.

Summary

Poland's syndrome is a rare congenital disorder characterized by unilateral chest wall hypoplasia, ipsilateral hand abnormalities, and a variety of associated anomalies. Hypoplasia of the subclavian artery or one of its branches is considered to be the responsible etiologic factor for the development of the deformities. When considering and planning the surgical options, the extent of the chest wall deformity and the degree of muscular involvement must be carefully taken into account

based on a case by case basis. Surgical corrections could be offered as a single or two stage procedures. Reconstruction or stabilization of the hypoplastic ribs can be achieved by open repair or minimal access surgical repairs, however in case of aplastic ribs techniques using bone grafts or a prosthetic mesh patch are advocated. Muscle flaps, breast prosthesis and lipomodeling are possible options to offer reconstructive for muscle deficiency and breast hypoplasia and to accomplish an acceptable esthetic result. Due to the established association between Poland's syndrome and tumors patients with Poland's syndrome should be screened and not be lost to follow-up as they make the transition between medical practitioners from the pediatric age to the adult.

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Asphyxiating Thoracic Dystrophy (Jeune's Syndrome): Congenital and Acquired

56

J. Duncan Phillips

Jeune's Syndrome (Congenital)

Overview

Familial asphyxiating thoracic dystrophy (ATD), also called asphyxiating thoracic dysplasia and thoracic-pelvic-phalangeal dystrophy (TPPD), is a rare multisystem autosomal recessive disorder originally described in the literature by Jeune in 1954 [1]. In this initial report, the authors encountered two siblings with broad, foreshortened, horizontal ribs and short upper extremities, who died from severe progressive respiratory insufficiency at the ages of 8 and 18 months.

Subsequent investigations have mapped the affected portion of abnormal genetic material to chromosome 15q13 [2]. Frequency of this condition is estimated to be between 1 in 100,000 and 1 in 130,000 live births [3].

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Additional Characteristics

Jeune's syndrome is one of at least seven short-rib syndromes that have been described (see Table 56.1). Most affected children with Jeune's syndrome have short ribs and short limbs, with dwarfism (see Fig. 56.1). The most common manifestation of the disorder is alveolar hypoventilation due to limitation of chest wall expansion. Affected children can present with variable severity, ranging from mild respiratory impairment to profound restrictive disease and death in infancy [4]. Most reported children have died within the first year of life from respiratory failure.

Organs affected in Jeune's syndrome, other than the chest wall, are variable and may include renal, hepatic, pancreatic, and retinal abnormalities. The characteristic chest wall configuration has been detected by in-utero ultrasonography as early as 14 weeks gestation.

Initial Evaluation

A thorough investigation to determine the extent of affected organ systems must be carried out early in the evaluation of these children. As a minimum, evaluation should include:

1. A thorough physical examination, including ophthalmoscopy, chest auscultation, and a detailed musculoskeletal assessment of the extremities.

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Table 56.1 Short rib syndromes

Syndrome	Common manifestations	Distinguishing features
Asphyxiating thoracic dystrophy (Jeune's syndrome)	Thoracic constriction	
	Respiratory distress (often severe)	
	Polydactyly	
	Limb shortening (dwarfism)	
	Renal disease	
	Liver disease	
Chondroectodermal dysplasia (Ellis-van Creveld dysplasia)	Thoracic hypoplasia and postaxial polydactyly are less pronounced	Ectodermal dysplasia
	Cleft lip/palate	
	Epispadius or cryptorchidism	
	Tooth abnormalities	
	Sparse/absent/fine hair	
	Heart defects common	
Short rib – polydactyly syndrome (SRPS), which includes:		
I-Saldino-Noonan type	Severe pulmonary hypoplasia	Usually fatal in neonatal period
	Genitourinary and gastrointestinal anomalies	Pointed femoral ends
II-Majewski type	Short/narrow thorax	Short ovoid tibia
	Micromelia, polysyndactyly/brachydactyly	
	Hydropic appearance at birth	
	Prominent forehead, low-set ears	
	Lobulated tongue, micrognathia	
	Cleft lip/palate, short/flat nose	
III-Verma-Naumoff type	Thoracic constriction	
	Renal abnormalities	
IV-Beemer-Langer type	Severe pulmonary hypoplasia	Usually fatal in neonatal period
	Cleft lip/palate	
	Genitourinary and Gastrointestinal anomalies	
	Omphalocele	
Thanatophoric dwarfism		
Chondrodysplasia punctata		
Osteogenesis imperfect		
Camptomelic dysplasia	Thoracic constriction	
	Femur/tibia bowing	
	Hydronephrosis/hydroureter	

2. Chest and extremity radiographs, to include the pelvis.
3. Ultrasound examination of the kidneys, liver, and (if possible) pancreas.
4. Infant pulmonary function tests, if available.

Formal surgical planning, if necessary (see below), will typically require:

1. Computerized tomography (CT) of the chest, to include 3-dimensional reconstructions, if possible. The typical cross-sectional images are those of a "clover leaf" deformity (See Fig. 56.2).
2. Laryngoscopy and bronchoscopy, since some children with Jeune's syndrome have tracheomalacia or bronchomalacia.

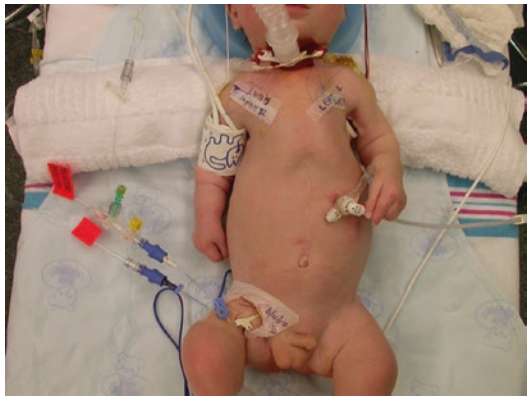

Fig. 56.1 2 month old boy with Jeune's syndrome. Note short ribs and limbs, with dwarfism. Child has already undergone tracheostomy and gastrostomy tube placement and is hospitalized, ventilator-dependent, with severe respiratory distress

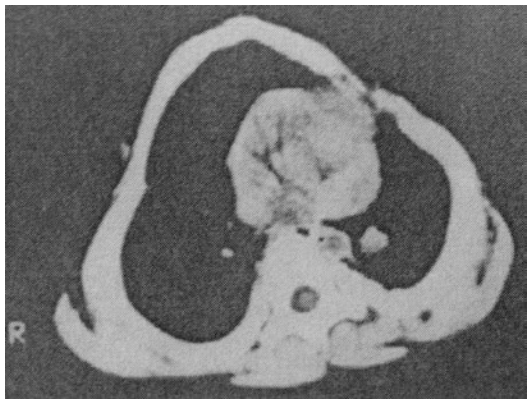

Fig. 56.2 Computed tomography (CT) of child with Jeune's syndrome, demonstrating the classic "clover leaf" deformity on cross-sectional imaging (From Takada et al. [8]. With kind permission of Springer Science+Business Media)

Involvement of multiple pediatric subspecialty physicians may be necessary and is usually advisable. In addition to the child's primary care physician, most of these children require involvement of a pediatric pulmonologist and a pediatric otorhinolaryngologist (or general pediatric surgeon, with extensive experience in management of airway issues, including infant tracheostomy). Depending on the results of the evaluation, other specialty physicians may include gastroenterologists, nephrologists, and ophthalmologists. Of course, involvement of a pediatric genetics specialist is advisable also, in order to exclude other short rib syndromes (see Table 56.1) and counsel parents on prognosis and potential for recurrence of the disorder in future-born siblings.

Most severely-affected children will require both tracheostomy and gastrostomy placement.

Surgical Treatment

Several different surgical approaches have been described for the treatment of Jeune's syndrome. Unfortunately, reports are limited by: (1) small numbers (most reports involve single cases or, at most, several patients); (2) brief follow-up (since the mortality rates are so high); and (3) bias toward certain approaches (with no reports able to compare/contrast different techniques from the same institution/authors). It is important to inform parents and other care-givers during the preoperative period that: (1) none of the surgical treatments for this disorder have been shown to "cure" the respiratory deficiency of the condition, (2) surgical treatments are probably best for use only in severe life-threatening cases, and (3) surgery is intended as an adjunct to medical management rather than a replacement.

Rib growth seems to improve in survivors following infancy and the respiratory difficulties tend to decrease [3, 4]. Therefore, some authors argue that aggressive surgical approaches to Jeune's syndrome in children during the first 2 years of life are reasonable, if designed to temporarily alleviate severe respiratory distress [4]. However, the high frequency of progressive renal failure in many of these children has led some authors to question the wisdom of embarking on major surgical reconstruction of the chest [3].

Sternal Split with Interposition

This is probably the most popular technique published [5–13]. Most reports describe a median sternotomy with interposition of various prosthetic materials to keep the two sternal halves separated (see Fig. 56.3). In theory, this increases chest wall circumference by the width of the

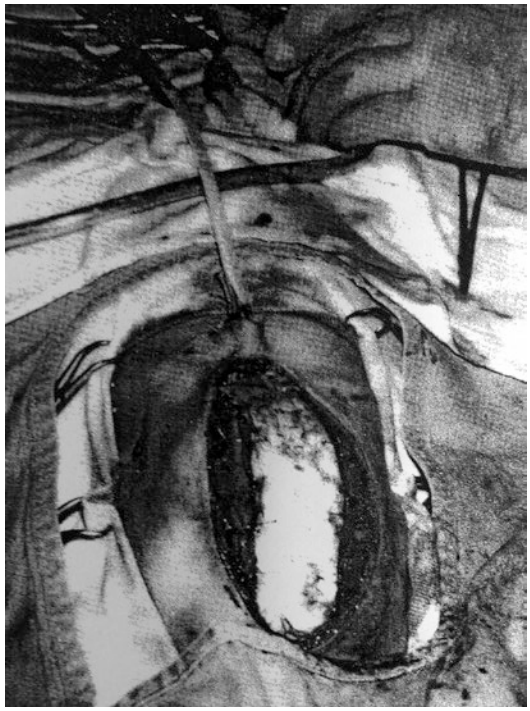

Fig. 56.3 Sternal split with interposition of methyl methacrylate prosthesis to keep the longitudinally-divided sternal halves separated (From Todd et al. [7]. With permission from Elsevier)

prosthetic material, allowing improved lung expansion and ventilation, as well as more room for lung growth during subsequent months and years. Table 56.2 summarizes reports of this technique, to date.

As noted by Aronson [11], the interposed material does not grow/expand with the patient and therefore, even if temporary improvement in respiratory function occurs, it seems inevitable that, with time, respiratory insufficiency will recur. In order to overcome this problem, Kaddoura and colleagues [12] developed a modified sternal spreader, with an adjustable screw, which they placed, following sternal split, between the two sternal halves (see Fig. 56.4). Pectoralis myocutaneous flaps were developed and mobilized medially, to cover the device, which was implanted, with the adjustment screw exteriorized through a stab wound incision. This apparently allowed 2 mm expansion procedures to be performed on a weekly basis until, at 8 months follow-up, the distance between sternal halves had been increased from 3 to 6 cm. Duration of follow-up on this case is unknown.

Conroy et al. [13] reported one case where distraction osteogenesis was used to achieve an

Table 56.2 Published reports of sternal split with interposition for surgical treatment of Jeune's syndrome (congenital variant)

Author/year	Age at surgery	Interposition material	Followup duration
Barnes et al. (1971) [5]	7, 20, and 30 months (3 operations)	1. Rib grafts, sterilized bone grafts	4 weeks (after third operation) (survived)
		2. Additional rib grafts and Dacron patch	
		3. Lower grafts secured to patch	
Karjoo et al. (1973) [6]	10 and 15 months (2 operations)	1. Stainless steel bar and Marlex mesh	15 months (after second operation) (survived)
		2. Bar removal	
Todd et al. (1986) [7]	9 months	Methyl methacrylate	Until hospital discharge (survived)
Takada et al. (1994) [8]	15 months	Methylmethacrylate	2.5 years (survived)
Sarimurat et al. (1998) [9]	13 months	Methylmethacrylate	2 months (death)
Sharoni et al. (1998) [10]	3 months (2 operations)	Methylmethacrylate then bilateral pectoralis muscle flaps	15 months (survived)
Aronson et al. (1999) [11]	4 months	Homologous bone graft (adult tibia from bone bank)	18 months (death)
Kaddoura et al. (2001) [12]	4 and 11 months (2 operations)	Methylmethacrylate then implanted expandable modified sternal spreader (see text)	8 months (survived)
Conroy et al. (2010) [13]	3 months	Distractor then Leibinger plates and mesh	27 months (survived)

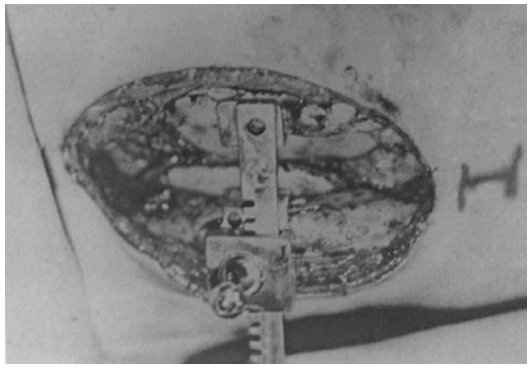

Fig. 56.4 Dynamic thoracoplasty technique of Kaddoura. A modified sternal spreader has been placed between the two sternal halves, to allow repeated gradual expansion procedures (From Kaddoura et al. [12]. With permission from Elsevier)

improved outcome. Following sternal split, the authors placed a mid-face distractor to slowly expand the separation, allowing bone to form between the two sternal edges. Following subsequent operations, the sternal distraction was repeated, in order to obtain more distance between the sternal halves.

Vertical Expandable Prosthetic Titanium Rib (VEPTR)

This device, developed in San Antonio, Texas by pediatric orthopaedist Robert Campbell and pediatric surgeon Melvin Smith, and first used in 1989 in humans, was initially developed to treat thoracic insufficiency syndrome caused primarily by early-onset progressive thoracic scoliosis [14]. Modification of the device (see Fig. 56.5) allowed the placement of multiple devices in a single patient for possible treatment of Jeune's syndrome [15–17].

The VEPTR device is a curved titanium rod, placed subcutaneously along the chest wall, and anchored, via "cradles" to a superior rib (such as the second or third) and an inferior rib on the same side of the chest. Multiple ribs are transected laterally to create, essentially, "flail segments" of chest wall which are then pulled laterally by thick wires attached to the VEPTR device. Progressive expansion of the VEPTR device requires reoperation every 4–6 months, to allow progressive expansion of the chest wall.

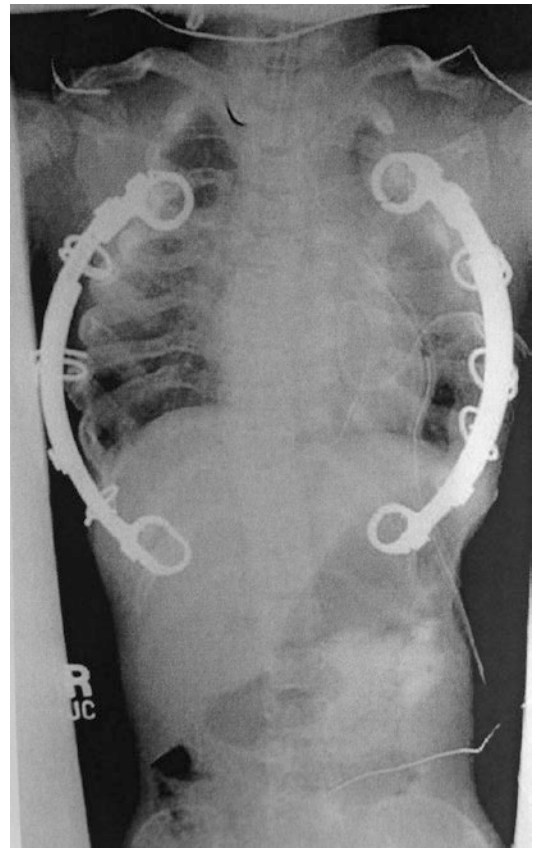

Fig. 56.5 Chest radiograph of child with Jeune's syndrome with bilateral vertical expandable titanium rib (VEPTR) devices in place. Multiple wires have been placed around ribs in order to allow lateral distraction of the chest wall by the VEPTR devices (From Waldhausen et al. [15]. With permission from Elsevier)

Although several authors have reported successful VEPTR placement in patients with Jeune's syndrome, the largest series reported from a single institution is that of Campbell's group [16], with 13 patients treated, with a minimum follow-up of 2 years and mean follow-up of 9.2 years. Transverse chest diameters increased an average of 8.6 % per year and thorax lengths increased an average of 7.3 % per year. Maximum benefit appeared to be achieved during the first five expansions, in most children. Radiographic studies suggested that the implanted "cradles" gradually migrated medially, making lateral chest wall expansion difficult and suggesting that the rib osteotomies might be fusing. The authors have suggested that, perhaps, more frequent expansions and

possibly earlier repeat rib osteotomies might improve the efficacy of the device.

Reports from a multicenter registry of VEPTR patients [17] noted that in 19 patients with Jeune's syndrome treated with the VEPTR device, four died within 1 year of surgery and others suffered from multiple life-threatening events, emphasizing the delicate nature of these children.

Lateral Thoracic Expansion (LTE) Technique

This novel approach was originally described by Davis in 1995 and has now been reported by several authors [18–23]. In this technique, ribs four through nine are dissected free from surrounding tissue laterally, including the underlying parietal pleura, and then divided in a staggered fashion (see Fig. 56.6). Underlying periosteum and chest wall and pleura is then divided in a staggered fashion in the opposite direction of the rib divisions. Divided ribs are then plated to each other (long ends to long ends) in order to expand the chest circumference laterally (see Fig. 56.7).

This operation is typically not performed until age 6–9 months, in order to allow the ribs to be of sufficient size and strength to support the expansion technique. Typically, one side is operated on and children are then maintained on mechanical ventilation for 4–6 weeks, to allow rib regeneration from the remaining periosteum. After 4–6 months, the contralateral side can undergo an expansion operation.

Davis initially reported his results on eight patients with Jeune's syndrome, using this technique [18]. Of the three patients with severe symptoms, operated on within the first year of life, two died within 2.5 months of surgery. However, of the five patients with milder forms of Jeune's syndrome, operated on between ages 3 and 9 years, symptomatic improvement in pulmonary function occurred. Davis was later able to show evidence of rib healing and new bone formation with this technique.

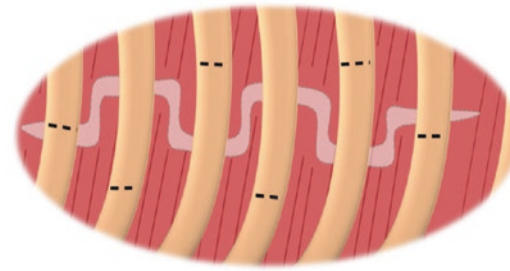

Fig. 56.6 Lateral thoracic expansion technique of Davis. Multiple staggered rib osteotomies have been performed (From Davis et al. [18]. Redrawn with permission from Elsevier)

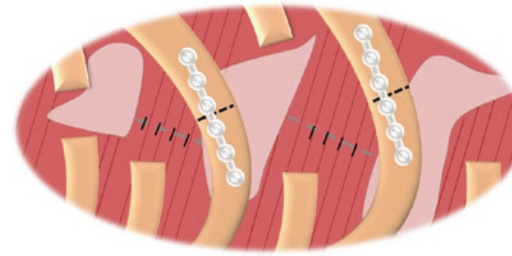

Fig. 56.7 Lateral thoracic expansion technique of Davis. Multiple titanium plates have been placed to approximate ribs in an alternating fashion (long ends to long ends) to expand chest circumference laterally (From Davis et al. [18]. Redrawn with permission from Elsevier)

A later report summarized his experience with 27 LTE procedures in 18 patients [21]. (It is unclear how many of these 18 had Jeune's syndrome and how many suffered from different, but similar, disorders). Although 4 of the 18 patients died, most of the 14 survivors demonstrated "clinical improvement." Serial pulmonary function tests, obtained postoperatively in 5 of the 18 patients, showed an increase in total lung capacity. After accounting for somatic growth, it was felt that LTE resulted in approximately a 7–15 % increase in lung capacity. Unfortunately, this improvement in lung function does not seem to persist since continued somatic growth appears to be greater than the increase in lung capacity and,

as a result, with time these patients actually end up with lung capacity that is a smaller percentage of what is predicted, based on height and weight.

Andrade et al. recently reported the use of this technique following a previous sternal split procedure with cartilage graft interposition [22]. Although Davis has published promising early results with LTE, other authors have been unsuccessful in achieving similar outcomes with this operation [23]. Indeed, Davis himself admits that "although the LTE procedure results in clinical improvement in the short run, the fundamental problem of Jeune syndrome, that growth of the thorax does not keep up with that of the body is not altered" and that there is not sufficient long-term follow-up to know "if the benefits of the procedure will last, and, if they don't, if reoperation is effective or even possible" [21].

"Nuss Technique"

Kiruchi et al. have reported the use of a modified "Nuss technique" for the treatment of two siblings, ages 14 and 9 years, with less-severe forms of Jeune's syndrome [24].

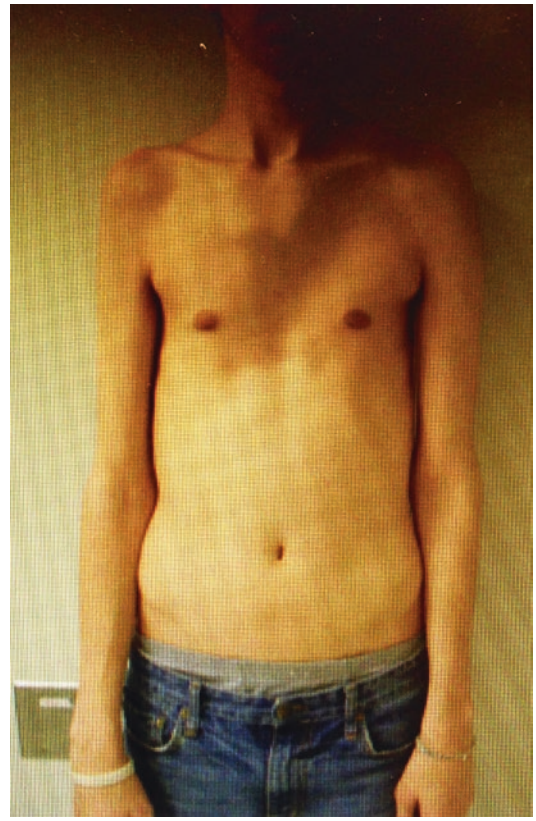

Fig. 56.8 14 year old boy with Marfan's syndrome and acquired asphyxiating thoracic dystrophy (AATD) 9 years following "open" (Ravitch-type) repair of congenital pectus excavatum. Chest circumference is markedly diminished

Acquired Asphyxiating Thoracic Dystrophy (AATD)

Acquired asphyxiating thoracic dystrophy (AATD), also called acquired restrictive thoracic dystrophy (ARTD) is an unusual condition, initially recognized by Azizkhan and Haller in the early 1990s in a group of children who had previously undergone repair of pectus excavatum or carinatum using "open" techniques, such as that described by Ravitch, with subperichondrial resection of deformed cartilages and sternal osteotomy [25]. This condition is typically called "acquired Jeune's syndrome." Twelve patients were described in detail in the original report in 1996.

Children with AATD may present many years following their original surgical procedures with progressive dyspnea and/or recurrent respiratory infections. Physical examination typically reveals a small, narrowed thoracic cavity with decreased circumference (Fig. 56.8). Previously untouched ribs (especially numbers one and two) may have undergone dramatic compensatory hypertrophy (Fig. 56.9). Pulmonary function tests (typically done at rest) usually reveal fairly marked reductions in FEV1 (forced expiratory volume in 1 s) and FVC (forced vital capacity) to only 30–50 % of predicted values, based on height and weight and age tables. Chest radiographs usually reveal a classic "tubular" shape to the upper chest (extending inferiorly to about the level of the 7th rib) and then may show "flaring" of the lowest ribs.

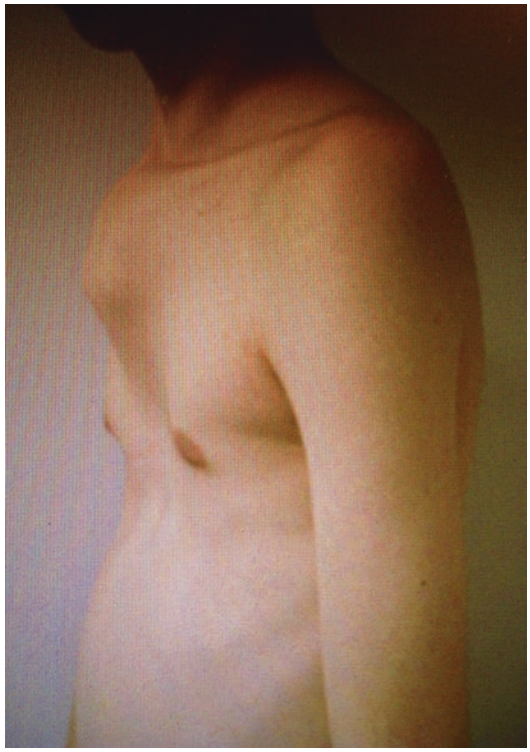

Fig. 56.9 Compensatory growth of ribs 1 and 2, with anterior displacement of the manubrium, in same child from Fig. 56.8, 9 years following "open" (Ravitch-type) repair of pectus excavatum

According to Haller, this condition appears to be a result of early, overly-aggressive resections of the deformed cartilages, with damage to the normal "growth plates" of affected ribs, impairing subsequent rib growth during later childhood and adolescence. Almost all children who have been described with AATD underwent their initial surgical procedures at age 5 years or younger. More recently, Robicsek and Fokin have attributed the condition to possible "overzealous" resection of involved cartilages (including the second costal cartilage) and/or suturing the remaining perichondrial strips together behind the sternum [26].

During the 1970s and 1980s, many authors (including Haller) advocated early "open" pectus excavatum repairs to prevent the long-term problems with respiratory impairment that can occur in some of these children [27–30]. Many pediatric surgeons were taught, in their training, to recommend these procedures to parents of children as young as 2 years of age. Unfortunately, the exact incidence of AATD remains unknown and therefore many as-yet unrecognized patients with AATD may yet become apparent and present themselves in the future with symptoms, requesting treatment. Surprisingly, some authors appear to still be advocating early pectus repairs. AATD has not been described following the minimally-invasive (Nuss-type) procedure nor following the pectus less invasive extrapleural repair (PLIER).

Evaluation

In addition to a thorough physical examination, children and adults with AATD typically require:

1. Pulmonary function testing, with spirometry. It is also helpful to get estimates of total lung capacity, if possible.
2. Echocardiography, since chronically-affected patients may develop right ventricular hypertrophy and even right heart failure due to pulmonary hypertension.
3. Computed tomography (CT), with 3-dimensional image reconstructions [31]. Shaded surface display (SSD) has been found to be extremely helpful in defining the specific anatomical issues in these patients and assists in surgical planning [32] (see Fig. 56.10).

Preoperative Preparation

AATD patients should be counseled extensively, prior to any attempts at surgical repair, regarding expected benefits (and risks) of the surgical procedure (see below). Medical treatment of any "reversible" components of respiratory insufficiency, such as reactive airways disease, should be maximized with a combination of bronchodilators and inhaled corticosteroids, if possible.

It is clear that surgical approaches to AATD are not indicated to improve the cosmetic appearance of the chest. Indeed, these operations often result in a chest wall configuration that some

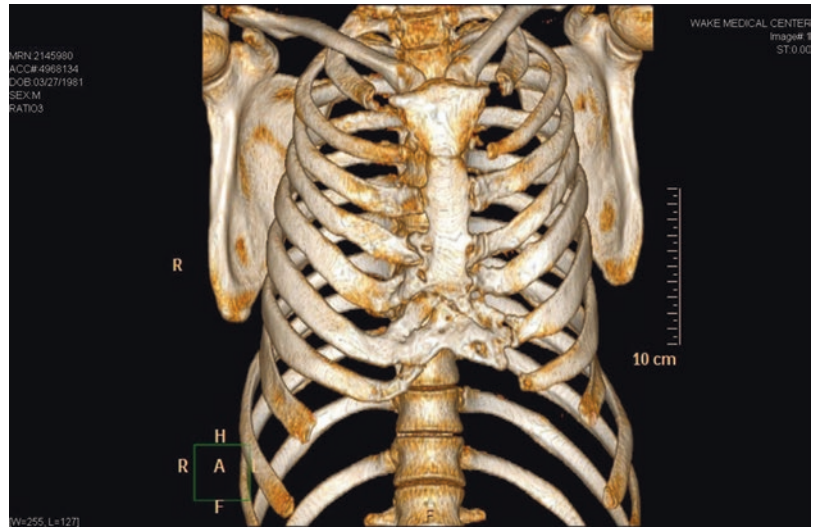

Fig. 56.10 Three-dimensional reconstruction of chest computed tomography (CT) of 30 year old man, 26 years after "open" (Ravitch-type) repair of pectus excavatum. Shaded surface display (*SSD*) technique aids in clarification of regrown calcified cartilages, with "fusion" of ribs to sternum, as well as markedly diminished volume of chest cavity

might deem unsightly and somewhat unusual [33]. It is important that patients and their family members be warned of this.

It is also clear that these extensive, complex, lengthy revisional operations can be anticipated to provide only a small objective improvement in measured lung function (see below). It is important that patients and their family members not have unrealistic expectations regarding expected improvements in quality of life.

Surgical Technique

In Haller's initial report of this condition, he advocated what is essentially a modified "re-do Ravitch" approach, with removal of the regenerated cartilages (and calcifications) and then placement of modified Rehbein splints anterior to the sternum [25]. The splints were intentionally curved anteriorly, into a gentle arc, so that the sternum could be elevated anteriorly and anchored to the splints with wire. This was to allow increased room in the chest for lung expansion. Unfortunately, early postoperative studies in the first six patients who underwent this repair showed no change in pulmonary function tests but preliminary exercise pulmonary function tests done 3–6 months later did show mild to moderate improvement. A total of 11 patients were reported with this procedure and, unfortunately 5 of the 11 developed complications from the Rehbein splints, including breakage, erosion, and detachment from the sternum.

In 1998, Weber described an alternative approach to AATD [34], utilizing modifications of techniques originally reported for treatment of Jeune's syndrome (see above). In this technique, a midline sternotomy is performed and the pleura is opened widely to enable both lungs to herniate anteriorly into the mediastinum. Subperiosteal lateral osteotomies of all ribs attached to the sternum allows the two sternal halves to be separated 4–8 cm. Three rib segments are then harvested and wedged/wired, in a transverse orientation, to keep the sternal halves apart (Fig. 56.11).

Weber reported followup on ten patients [35]. Pulmonary function tests showed gradual improvement in measured parameters over a 24 month period following surgery with typical values (such as FVC, or forced vital capacity or FEV1, or forced expiratory volume in 1 s) increasing from roughly 50% of predicted (based on height and weight) to roughly 2/3 of predicted. All patients reported subjective improvement in symptoms. (Interestingly, this mild degree of measured "objective" improvement in lung function, as well as subjective clinical improvement, is quite similar to that reported by Davis for LTE for Jeune's syndrome—see above).

We have utilized a modification of Weber's technique for the treatment of children and adults with acquired Jeune's syndrome. Some patients have benefited from implantation of

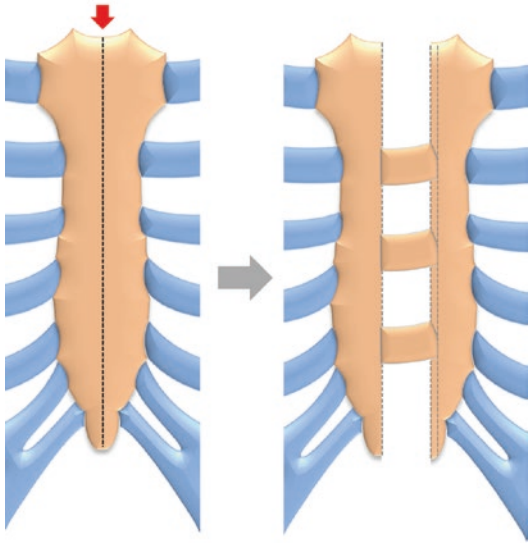

Fig. 56.11 Sternal split technique, with rib graft interposition, for the treatment of acquired asphyxiating thoracic dystrophy (*AATD*), as originally described by Weber [34]

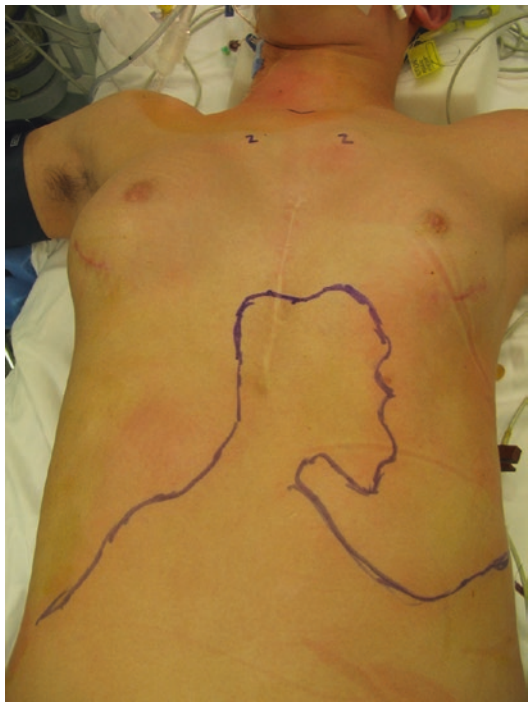

Fig. 56.12 14 year old boy with acquired asphyxiating thoracic dystrophy syndrome (*AATD*), 9 years following "open" Ravitch-type repair of pectus excavatum. Bilateral subpectoral tissue expanders have been placed and gradually expanded with serial injections of saline. The previous healed midline scar can be seen. The costal margin has been outlined in purple. Medial heads of the second ribs have been marked superiorly

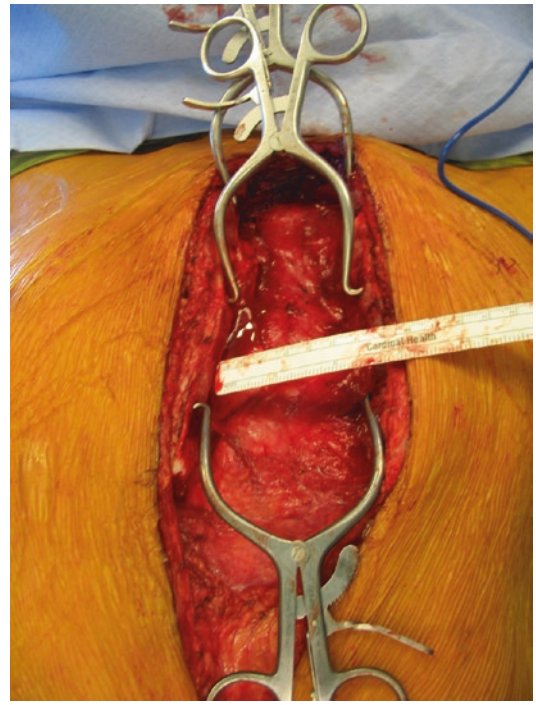

Fig. 56.13 Same patient from Fig. 56.12, undergoing sternal split procedure. Retractors hold sternal halves approximately 8 cm apart, prior to rib graft placement

tissue expander devices beneath their pectoralis major muscles, prior to sternal split procedures (Fig. 56.12). This can aid in progressively stretching the overlying muscle, soft tissue, and skin for easier approximation in the midline and better coverage of the rib grafts. Sternotomy is performed via midline incision (Fig. 56.13). We have found that expansion of up to 8 cm can be achieved. Multiple rib grafts may then be placed and held in place with sternal wires (Fig. 56.14) or metal plates.

In some patients, we have utilized unilateral pectoralis major muscle flaps to cover the rib grafts anteriorly (Fig. 56.15). In others, such as the patient seen in Figs. 56.12, 56.13, and 56.14 who had tissue expanders implanted prior to sternotomy, pectoralis muscles can be approximated in the midline anteriorly (Fig. 56.16).

Followup 3-dimensional CT scans have documented complete healing of the interposed rib grafts. The mechanism of respiratory improvement is assumed to be the ability of the lungs to herniate anteriorly, in front of the pericardium,

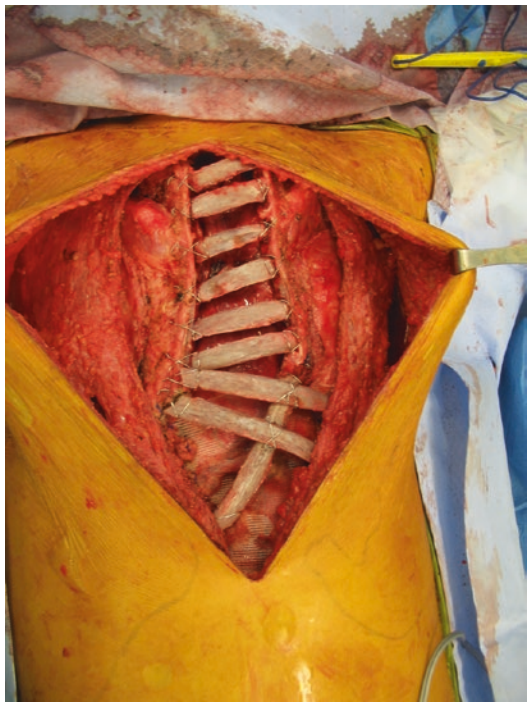

Fig. 56.14 Same patient from Figs. 56.12 and 56.13, with interposition of 9 rib grafts, secured to sterna halves with stainless steel wire

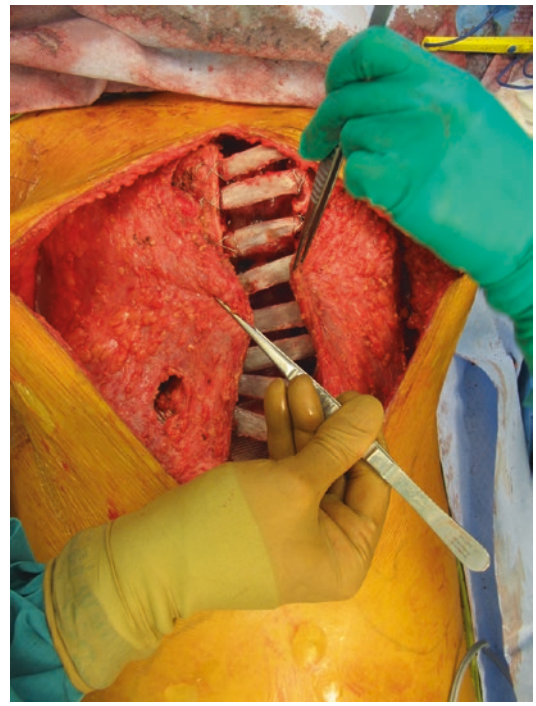

Fig. 56.16 Same patient from Figs. 56.12, 56.13, and 56.14, with pectoralis major muscles being approximated anteriorly, in front of the rib grafts

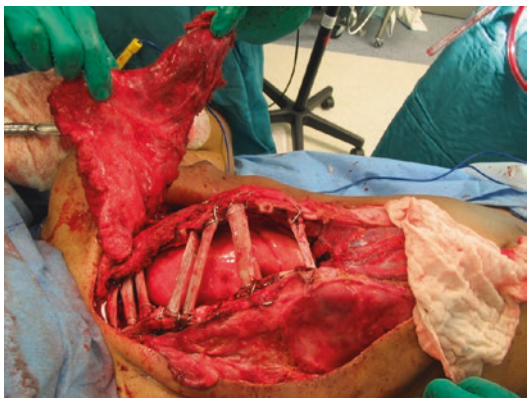

Fig. 56.15 Pectoralis major flap used, in some patients, to cover the rib grafts. Note parietal pleura has been opened and lungs have been allowed to "herniate" anteriorly

and essentially expand and contract in a "new space" created by the surgeon, medial to the two pleural cavities. Interestingly, this is similar to the mechanism of Haller's approach with modified Rehbein splints (see above) and the sternal split procedures described by various authors for the treatment of Jeune's syndrome.

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Post-traumatic and Post-surgical Chest Wall Deformities (Acquired Chest Wall Deformities)

57

Helmut Wegmann and Amulya K. Saxena

Chest wall deformities may be congenital or acquired secondary to conditions that affect the patient at any stage of life. Although, emphasis has been placed in the diagnosis and treatment of the congenital types of chest wall deformities, acquired chest wall deformities present a completely different aspect depending on the underlying cause that led to the deformity. Acquired chest wall deformities constitute less than 1 % of all thoracic deformities and can be classified in four major groups as presented in Table 57.1 [1].

Introduction

In order to prevent the development of acquired chest wall deformities in young patients resulting from iatrogenic damage which in turn results in impaired chest wall growth after thoracic surgical procedures, it is crucial to have knowledge on the mechanisms of chest wall growth and pathophysiology of the chest wall growth plates.

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Approximately, 75 % of the longitudinal growth of a rib takes place at the sternal end and occurs predominantly as a result of endochondral bone formation [3]. Experimental data has demonstrated that chest wall development is severely affected if growth centers of the ribs in the costochondral junction are not preserved during surgical procedures or more than four ribs are resected during a surgical intervention in the thorax; in this case the antero-posterior growth of the chest wall may be affected and significantly retarded [4]. Not only the costochondral, but also the removal of

Table 57.1 Types of acquired chest wall deformities based on the etiological factors

1. Intrinsic pathological processes within the thoracic
Congenital cardiac anomalies with enlargement of the heart
Thymic tumors associated with excessive growth
2. Pathologies involving the ribs and muscles in the chest wall
Tumors of anterior chest wall
Infections of the rib(s)
3. Post-surgical interventions
Operative procedure associated with deformities include
Repair of various types of chest wall deformities
Harvesting of rib grafts
Cardiothoracic procedures [2]
4. Post-traumatic
5. Secondary to alterations in the vertebral column
Scoliosis
Kyphosis

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sub-perichondrial rib cartilages has been reported to result in the formation of a chest wall deformity within 1 year after surgery in up to 50% of the patients [5]. Another important point to be considered here is the damage to the sternal growth center during complete upper transverse ostotomy which is associated with accompanying blood supply disruption, that might play a role in the development of these deformities [6]. Hence, the localization and knowledge of sternal and costochondral growth centers is important, to avoid damage of these centers during surgical procedures in order to decrease the possibility of an iatrogenic related development of a chest wall deformity [1].

Development of Human Sternum and Ribs and Ossification

The embryological development of the sternum is important to understand its formation and to understand the surgical considerations with regards to its embryology. The sternum is of mesodermal origin and is a structure that is formed by fusion of embryonic tissue in the midline of the developing thorax [7]. Cells from two separated bands of mesoderm on either side of the anterior chest wall - *the sternal bars* - migrate toward the midline, and fuse around the 10th week of gestation to form the sternum (Fig. 57.1) [8, 9].

The sites of ossification appear from the cranial to the caudal direction in the manubrium and mesosternum prior to birth, but are not present in the xiphod process until the 6th year of life. The ossification sites are formed in the midline within the intervals between the articular depressions for the costal cartilages in the intercostal spaces. While variations are possible, generally a single site in the manubrium and three major sites in the mesosternum represent most sternal patterns that have been observed at the time of birth [10]. Postnatal sternal ossification is inherent, but three basic patterns have been distinguished and are presented in Table 57.2 (Fig. 57.2) [11]:

Fusion of adjacent mesosternebrae proceeds in a caudal-to-cranial direction, opposite to the initial appearance of the ossification centers. The sternal maturity is characterized by the coalescence of expanding ossification centers, which may be influenced by biological forces to the sternum [12]. The manubrium and mesosternum are completely ossified by the 21st year of life. Ossification of the xiphoid process is usually completed by the 35th year of life but in rare instances may remain cartilaginous throughout life. The ossification centers of the sternum develop from the posterior aspect of the perichondrium first to the lateral part. The anterior domain and the costosternal joints are the last part to ossify [10].

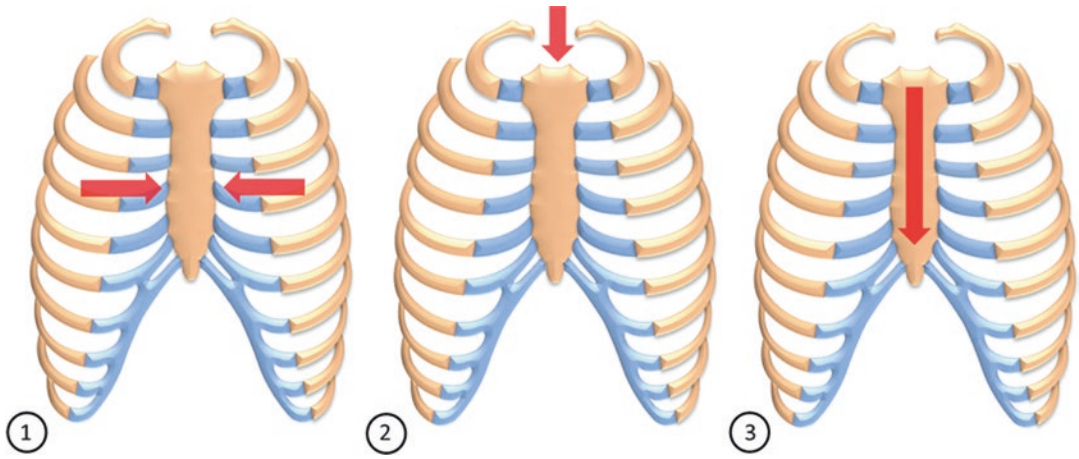

Fig. 57.1 Embryological development of the sternum showing (*1*) the migration of the sternal bars towards the midline. (*2*) The sternal bar fuse towards the cranial part initially to form the jugulum and (*3*) later the caudal progression of fusion leads to the complete formation progressing from the manubrium towards the formation of the xiphoid process

Table 57.2 The three basic patterns of poststernal ossification observed in sternum

Type I	*Ossification is localized in a single midline center in the manubrium and within each of the three mesosternebra which is the square area between the sternal ends of the costal cartilages*
Type II	*Ossification is localized in a single midline center in both the manubrium and first mesosternebra, and also in two bilateral centers of the more caudal mesosternebra*
Type III	*Ossification is localized in a single midline center in the manubrium along with bilateral centers in each of the mesosternebra*

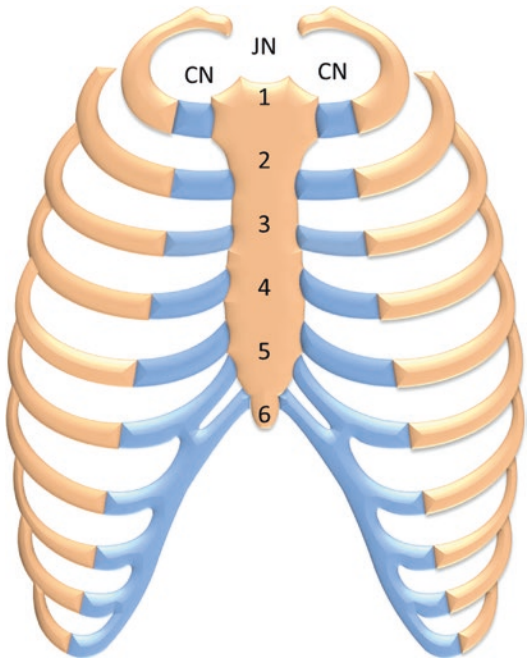

Fig. 57.2 Human sternal anatomy with ossification centers. Clavicles have their insertion in the clavicular notches (*CN*) separated by the jugular notch (*JN*) [9]

Costal cartilage ossification seems to be inhomogenic. The ossification process of the first rib proceeds from the costal toward the sternal end of the cartilage in an anteromedial direction [13]. Approximately 75 % of the longitudinal growth of a rib takes place at the cartilaginous portion of the costochondral junction [3].

There is no satisfactory explanation to understand the etiology of pectus deformities, but there might be evidence from rodent model that damage to the growth plates of the sternum result in predictable patterns of aberrant growth. If sternebrae are seen analogous to epiphyseal centers, surrounded by regions of hyaline cartilage with growth plate potential, then physiological damage might translate in lost potential similar in magnitude and direction to that described for long bones [10].

Iatrogenic Pectus

Etiology of Deformities After Pectus Surgery

Deformities of the thorax have been reported in patients that have undergone correction of severe forms of congenital pectus deformities [6, 14]. Due to the clinical findings in these patients who present with a narrow torso, an immobile anterior chest wall and various degrees of dysnpea with decreased vital capacity and forced expiratory volume, this conditioned was termed as "Acquired Restrictive Thoracic Dystrophy" (ARTD) [15]. ARTD was observed in very young children undergoing open pectus excavatum repair with unusual abundant rib resection – that involved removal of three to eight ribs on each side or five to six total rib cartilages bilaterally.

Hence, the normal recommendation for a basic operative procedure would be to resect a 2.5 cm segment of three to four cartilages on each side with preservation of the perichondrium. This observation has further led to the recommendation of delaying open repair in pediatric patients until the 6th–8th year of life [6, 16], with the expectation that the impact on growth failure would be more moderate in older patients if growth plates were to be damaged during a surgical procedure.

It has also been demonstrated that removal of the costal cartilages leads to reduction of the antero-posterior diameter of the thorax, while preservation of 1/4th of the cartilage length at the costochondral junction protects the growth centers of the ribs (Fig. 57.3) [17]. Furthermore damage to the sternal growth center by complete upper transverse osteotomy with corresponding blood supply disruption might cause deformities [6].

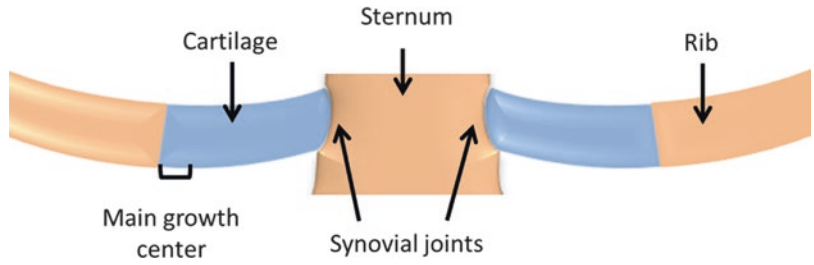

Fig. 57.3 Preservation of 1/4th of the cartilage at the costochondral junction has found to protect the growth centers of the ribs

Chest Wall Deformities After Cartilage Graft Harvesting

Costal cartilage harvesting for autologous cartilage tissue transfer is used in many occasions in plastic and rhino-facial surgery, when rigid support for soft tissue reconstruction is needed. These include surgical procedures for reconstruction in microtia which are often performed in young patients [18, 19]. In this procedure to reconstruct the auricular framework, two or more whole costal cartilage grafts are often necessary, with some patients even requiring additional grafts [5]. Donor site complications after such procedure have been rarely reported in the literature; but, increased amount of harvested cartilage is associated with chest wall deformities.

Ohara et al. controlled 18 patients after cartilage harvesting of the ribs for microtia reconstruction. Each cartilage was harvested subperichondrial from the costochondral junction to its free end or attachment to the sternum. The perichondrium, muscle, fascia and subcutaneous tissue were closed in layers. All grafts were taken from the sixth to ninth costal cartilages. It was observed that 16 of 32 ribs from which costal cartilage had been harvested showed increased inward bowing on radiographs. The rate of deformation correlated with age and was more frequent in patients younger than 10 years of age. The deformities were more conspicuous and severe when two or more costal cartilages had been harvested. The rib deformities could usually be observed on radiographs within 1 year of surgery. Interestingly, harvest of the sixth costal cartilage induced over 100 % rib deformities [5].

In 1976, Radford et al. observed in a 6–13 years follow up depression of the chest wall in 16 % of the patients after cartilage harvesting for microtia reconstruction. The technique for harvesting involved the removal of the sixth, seventh and eighth rib cartilages together with the perichondrium, while leaving the synchondrosis between the two larger ribs intact [19].

Thomson et al. also harvested the 6th to 8th rib. Their approach involved the re-adaptation of the intercostals muscle and remaining cartilages with interrupted non-absorbable suture material (Vicryl®, Ethicon Inc, polyglactin 910 sutures), and positioning of the 9th rib cartilage up to the level of the 6th without tension to obliterate the potential margin deformity. The serratus anterior muscle was then approximated with a running suture of 2–0 Vicryl®. Using this approach, a total of 25 % chest wall deformities were observed, with 8 % of them seen in the 6–12 year old children and 33 % in 2–3 year old children [18].

Due to the well-known long term occurrence of chest wall deformity after costal cartilage graft harvesting, a more conservative way of harvesting costal cartilage was developed for rhinoplasty [20]. In this approach, only a central portion of the rib is resected with preservation of intact costal cartilage on three sides (Fig. 57.4).

Due to the better outcomes with this procedure, a brief description of the steps is provided. A 3 cm incision inferior to the infra-mammary crease in the female patients and below the inferior border of the pectoralis major muscle in the male patient is created. The dissection is focused then towards the cartilage of the 7th rib. The rib is then exposed from the lateral osseocartilaginous border to the medial junction of the rib and the sternum. A 27-gauge needle is used to evaluate the cartilage and minimize the harvesting of calcified tissue. The perichondrium is incised with a scalpel in the shape of a "T" and periosteal elevator is then used to lift the perichondrium carefully. A 15-blade scalpel is then used to make incisions both at the medial and at

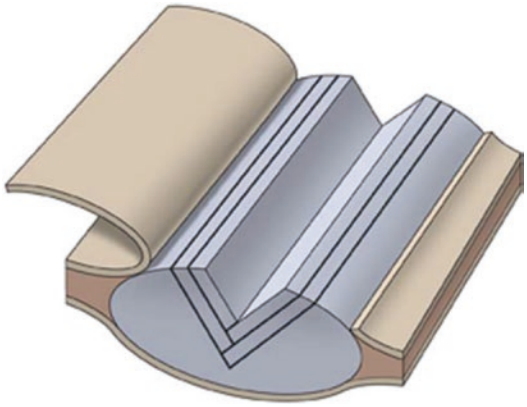

Fig. 57.4 Illustration demonstrating the technique for costal cartilage harvests which decreases donor site morbidity and minimizes warping or displacement (Lee et al. [20]. With permission from John Wiley and Sons)

the lateral aspect of the proposed graft. The incisions should extend through the cartilage but not violate the underlying perichondriuum. At this time an incision is made directly perpendicular to the rib at the mid portion of the rib. The incision is angled at 45° and moves to remove the graft. The initial harvest is usually the largest, shaped like a triangle. Further grafts can be harvested as needed leaving the rib borders (three sides) and underlying perichondrium intact. Only the amount of cartilage that is needed is harvested in order to minimize the amount of tissue that may be discarded later. The overlying perichondrium is now approximated with 4–0 Vicryl suture. The wound is then closed in multiple layers with 3–0 Vicryl used to approximate the muscle. The deep dermis is closed with 4–0 Vicryl, and finally a 4–0 Monocryl is used for the epidermis. Drains are not typically placed in the wound bed after the completion of the procedure. This technique allows for sufficient graft tissue for rhinoplasty, while decreasing donor site morbidity and minimizing warping or displacement.

Iatrogenic Pectus Carinatum After Chest Wall Surgery

Post-surgical pectus carinatum deformities are rare. Swanson et al. reported three patients with "reactive pectus carinatum" after correction of pectus excavatum. Two of these patients were observed to demonstrate this deformity after pectus excatum correction with the Minimal Access Repair of Pectus Excavatum (MARPE) technique while it was observed in one patient after Ravitch procedure [21]. All three patients developed a ventral displacement of the sternum within 1 year after primary intervention. In both patients who underwent MARPE, the bars were removed after 5 months. While in one patient the defect resolved after his pubertal growth spurt, the other patients required two subsequent surgical interventions with costal excision to return the sternum to a neutral position to correct the recurrent pectus carinatum deformity. After the second revision, the sternum was stable in neutral position. In the patient with Ravitch procedure metal removal was done and after 3 years of expectant management, at the age of 17, with computed tomography (CT) scans demonstrating persistent pectus carinatum with bony and fibrous hyperplasia in the anterior chest wall. This patient underwent operative repair involving perichondrial excision and resection of the involved cartilage, and a sternal osteotomy was performed with wedge excision and cartilage graft to deflect the sternum. There was no recurrence of pectus deformity in this patient (Fig. 57.5).

Thompson et al. reported acquired chest wall deformity in 3 month old boy with congenital heart defect that underwent a cardiac surgical procedure [2]. Sternotomy was done twice – once at age of 3 months and repeated at age of 10 months. Approximately, 8 months after the second surgery pectus carinatum was observed. In this case, a revision was performed, and all but one banding wires and excess tissue were removed. During surgery, it was found that the costal cartilage did not appear elongated, but the lower pole of the sternum was prominent. This was shaved down to match the level of the rest of the sternum. Recurrence of pectus carinatum in this patient was not observed.

Post-traumatic Chest Wall Deformities

Reports of post-traumatic pectus excavatum deformities have been rarely reported and documented in the historical literature. In 1931,

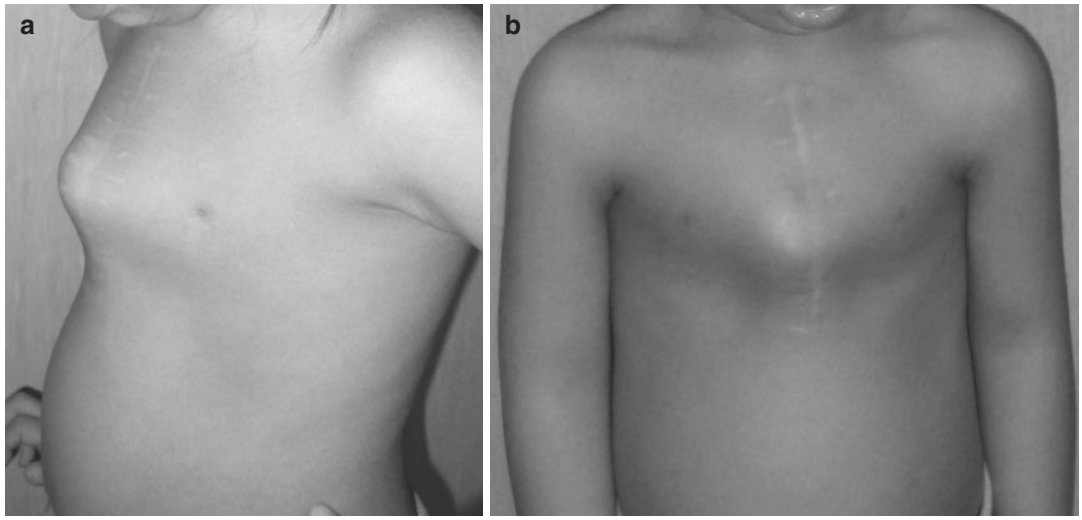

Fig. 57.5 (**a**, **b**) The formation of a pectus carinatum in a 4-year old boy after sternotomy for a cardiac procedure 1 year ago

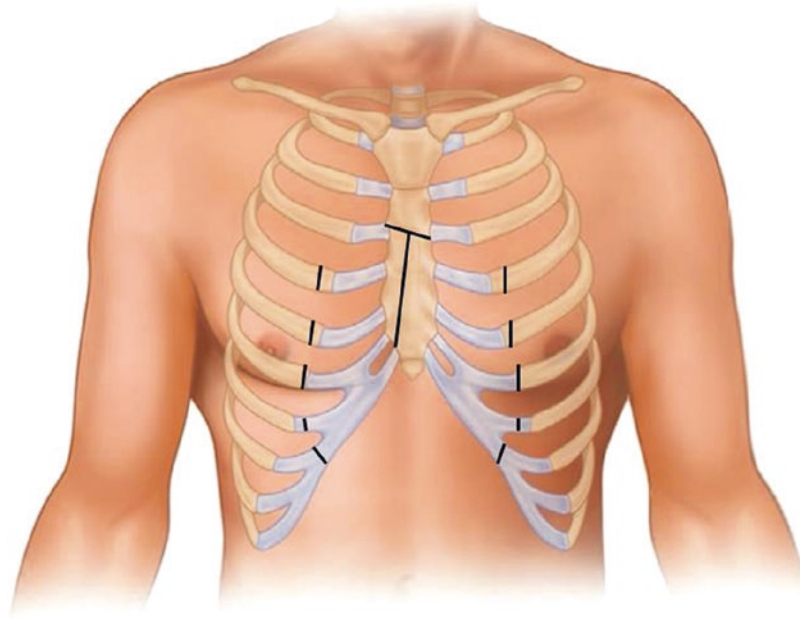

Fig. 57.6 Heavy lines indicating the division of the sternum, ribs and cartilages in order to repair the deformity in the patient [22]

Alexander reported two adolescent patients with post-traumatic pectus excavatum. In one patient who presented at 16 year of age, blunt trauma to the chest wall occurred 4 years ago during a bout of wrestling. The second patient who was a 20 year old woman, was reported to have a blunt chest wall trauma in a car accident 2 years ago. Due to severe pectus excavatum deformity both developed symptoms of dyspnoea and palpitations. The author presumed that multiple fractures of the thoracic cage lead to an inward bending of the frontal thoracic cage due to negative pleural pressure. Both were operated and respiration was improved. In the latter patient excessive sternal callus formation was reported at the time of surgical correction (Figs. 57.6, 57.7 and 57.8) [22].

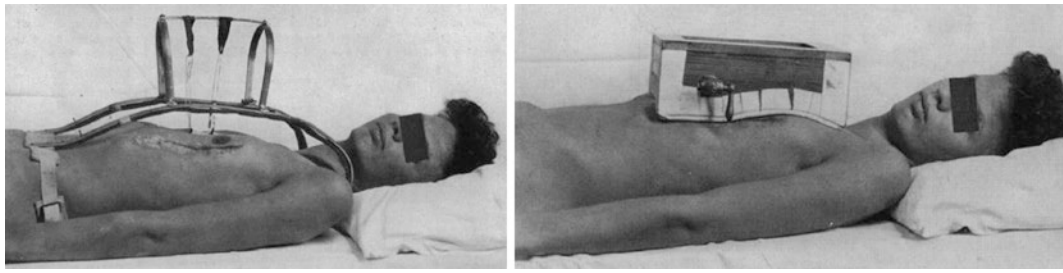

Fig. 57.7 Historical image: device by which reduction of the sternal deformity was maintained for 5 weeks after operation. The band and wire surrounding the sternum are suspended to a bar by rubber bands. *Right*: Box that was air-tight with skin when negative pressure created in it by connecting the pipe with the hospital air suction system. This box was used after removal of the device pictured above in order to maintain reduction of the sternum deformity until firm bony union of the divided sternum and ribs occurred (Alexander [22]. With permission from Wolters Kluwer)

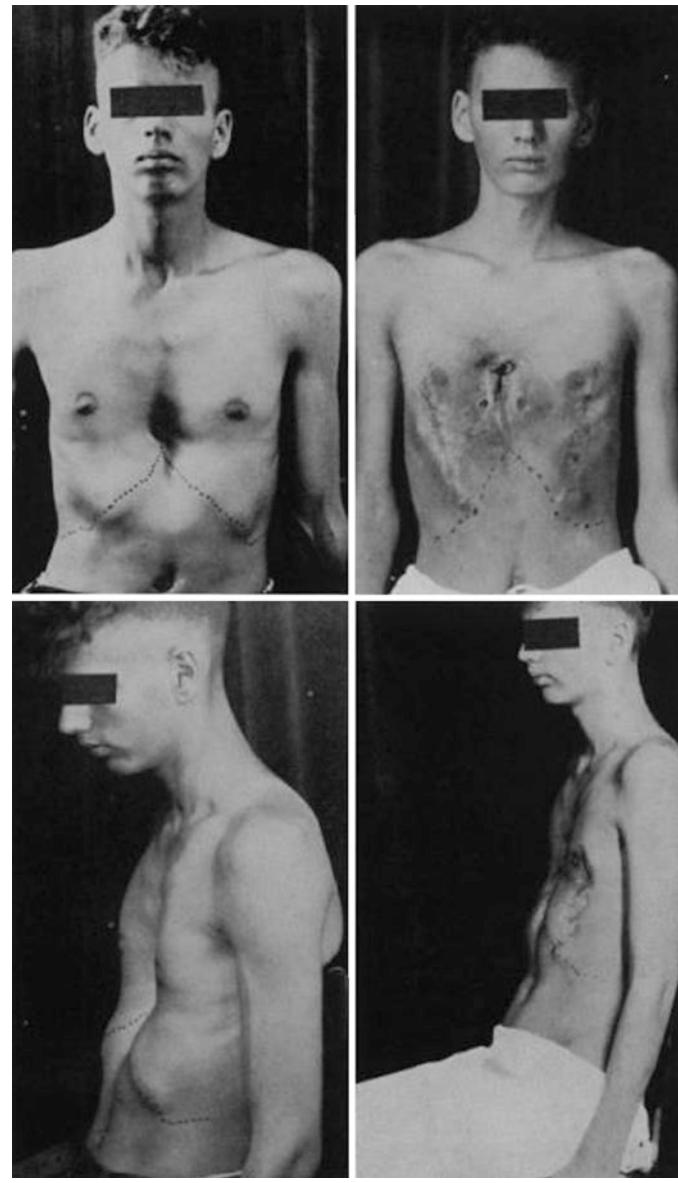

Fig. 57.8 Historical image. The patient who developed a traumatic pectus excavtum- before (*left*) and after (*right*) correction (Alexander [22]. With permission from Wolters Kluwer)

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Thoracic Reconstruction in Chest Wall Tumors

58

Alireza Basharkhah and Amulya K. Saxena

Introduction

The chest wall protects vital organs like the heart, lung and liver as a cage and at the same time provides a flexible skeletal framework to stabilize the movements of the shoulder and arm, and enabling respiratory movement with more than 20,000 breaths a day [1].

Chest wall reconstruction involves both bony and soft tissue elements with the general rule that all full-thickness skeletal defects that have the potential for paradoxical chest movement should be reconstructed [2]. Meanwhile although large tumors of the chest wall can be treated by resection and soft tissue converge, several studies have demonstrated that stabilization of the chest wall in addition to soft tissue coverage reduces ventilator dependence and over-all hospital stay [3–5].

In case of large chest wall defects that result from tumor resection (primary, recurrent, metastatic lesions, or locally invasive), irradiation necrosis, trauma or infection, there is a disruption of the chest wall integrity, which may result in significant morbidity, rapid life-threatening conditions and paradoxical chest movement, known as flail chest. In a flail segment larger than 5 cm ventilation becomes progressively inefficient. Therefore, adequate reconstruction of large chest wall defects is mandatory to avoid flail chest resulting from resection larger than 5 cm or loss of more than four or five ribs [6].

The major goals of chest wall resection and reconstruction in oncologic patients include complete removal of the local extent of the tumor, restoration of adequate protection to the thoracic viscera chest wall stability, elimination of dead space, preservation of respiratory function, restoration of physiologic function, providing for adequate chest wall growth and an acceptable chest wall appearance and avoiding infection [6, 7].

Chest Wall Tumors

Chest wall tumors (CWT) are rarely seen in childhood with incidence rate less than 1 in 1,000,000 [8]. Just 1.8 % of solid tumors are located in the chest wall in the pediatric population [9] whereas about 22 % of the thoracic tumors in childhood are located in the chest cavity [10].

CWT consist of a wide range of benign and malignant diseases. In childhood, the most predominant malignancies are Ewing tumors; whereas in adults chondrosarcomas are most often encoun-

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tered [11, 12]. Shamberger et al. reported Ewing tumors to be 56 % of chest wall malignancies, followed by rhabdomyosarcomas (23 %), lymphomas (15 %), fibrosarcomas (5 %), osteosarcoma (3 %) and chondrosarcomas (3 %) [12]. Benign tumors like eosinophilic granuloma, aneurysmal bone cyst (ABC), hamartoma, osteoma, osteochondroma, chondroma and metastatic tumors such as osteogenic sarcoma, neuroblastoma and Wilms' tumor are rarer than primary malignant tumors [13]. Meanwhile the most common location of CWT are ribs, but clavicle sternum, scapula and soft tissue may also be involved [7].

Ewing Sarcoma Tumor Family (ESTF)

ESTF is a group of tumors with single neoplastic entity, sharing common phenotypic and molecular features. This group consists of Ewing sarcoma (ES), primitive neuroectodermal tumor (PNET) and malignant small cell tumor of throacoppulmonary region, called Askin tumor (Fig. 58.1). In the intergroup study, 13 % of patients with Ewing patients had rib tumors [14]. ES-PNET most commonly develops in the second decade of life (64 %), although it is not uncommon for a diagnosis to be made before puberty or even into adulthood [15]. CWT lesions comprise 6.5 % of primary ES and when grouped with PNETs, these are the most common chest wall tumors in children followed by rhabdomyosarcoma [12].

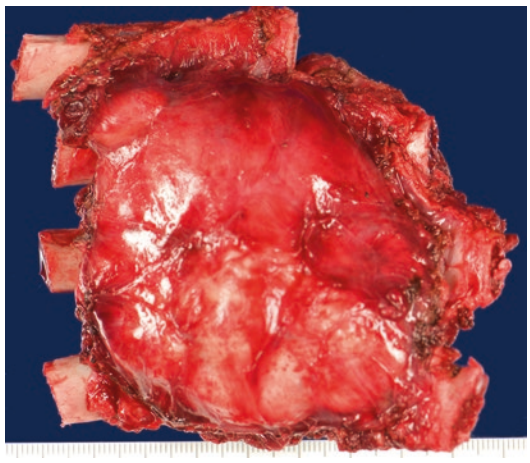

Fig. 58.1 Malignant small cell tumor of throacopulmonary region also known as Askin tumor demonstrated after surgical resection

Askin Tumor

Epidemiology and Symptoms

Askin tumor is very rare but highly malignant tumor and comprises the most common malignancy among CWT. It was first described in 1979 by Askin [16]. This report described a unique clinicopathologic entity characterized as a malignant small cell tumor of the thoracopulmonary region in 20 children and adolescents (mean age 14.5 years) with a female predilection (75 %).

The diagnosis is based on the histological presence of small cells with neural differentiation. The blue small round cells are presumed to arise from embryonic migrating cells of the neural crest [17]. In addition to this histological changes, chromosomal translocation t(11;22)(q24;q12) in the tumor cells [18, 19], other translocations like t(21;22), t(7;22) and t(17;22) have been observed [20]. Southern blotting has revealed that the position of the breakpoints on chromosome 22 which have been found to be tightly clustered within a 7-kilobase (kb) fragment of the genomic DNA, within a gene designated EWS [21, 22]. A tri/tetrasomy of chromosome 8 is to be found in 50 % of patients with ESFT [23]. This seems to be a proliferative advantage of the tetrasomy 8 cells based on the enrichment seen in experiments with culturing of bone marrow cells in ES [24]. Beside these chromosomal aberrations, other diagnostic features like positive PAS (Periodic acid Schiff) reaction, CD99 and NSE (Neuron specific enolase) can be demonstrated in tumor cells.

Immunohistochemically, MIC-2/HBA-71 is found to be positive in many patients. The MIC-2 gene is a pseudoautosomal gene located on the short arms of the X and Y- chromosomes in Ewing's sarcoma cells and peripheral PNET (Primitive neuroectodermal tumor) cells. The gene product, a cell membrane protein, is recognized by the monoclonal antibody (MoAb) HBA-71 [25].

The symptoms of Askin are usually detected after the tumor has achieved substantial size, therefore the tumor is generally detected at an advanced stage [26]. The associated symptoms are often very unspecific. These may include non-specific pain and swelling or mass at the affected throacopulmonary region, collapse, night sweat, orthostatic dysregulation, dyspnea, cough, decreased condition, weight loss, fever and Horner's syndrome.

Diagnosis

At the time of presentation, imaging studies show typical bone destruction in the affected rib and rather large extension of the tumor mass to the adjacent areas. Usually, the first examination that is performed is a chest film. These may reveal unilateral chest wall mass, pleural fluid and thickness and bone involvement (Fig. 58.2).

In order to detect any invasion to the adjacent lung parenchyma, pulmonary nodules and sometimes lymphadenopathy computed tomography is performed [27]. Computed tomography can provide more detailed information about vascular supply of the tumor, its extent, location and its composition. Additionally, a bone scan should be performed to exclude any osseous metastasis [13].

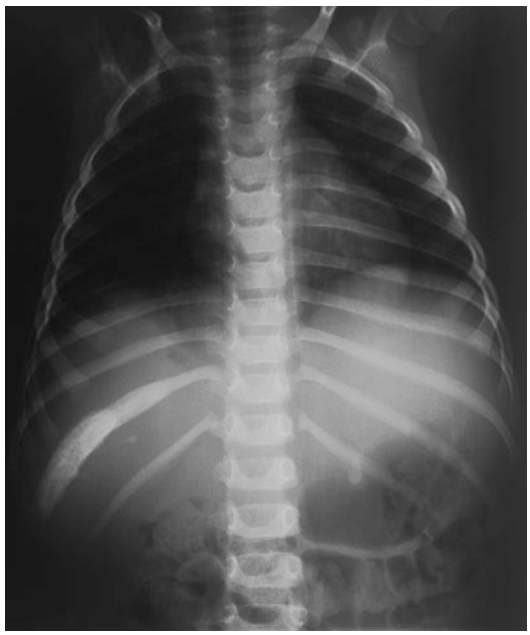

Fig. 58.2 Chest film of Askin tumor presenting as unilateral chest wall mass with bone thickening (lower rib on the right side) and pleural effusions on the same side

The diagnosis of Askin tumor rests on histopathological investigations and immunohistochemical examinations; therefore a biopsy is generally performed. The diagnosis is based on the presence of small cells with neural differentiation. The diagnosis is then confirmed by cytogenic examinations with the expression of CD99 in all cases and no expression of cytokeratins or desmin [28]. Contesso et al. reported invasion in regional ribs, destruction and tumor infiltration in soft tissue [29]. In order to detect any bone marrow involvement a bone aspiration or biopsy should be performed.

Therapy

As mentioned, Ewing sarcoma is the most common malignant CWT in children and adolescents. Askin is extremely aggressive and requires an intensive local therapy with extensive resection, radiotherapy occasionally, neoadjuvant and adjuvant therapy. It is recommended to apply an aggressive multimodal approach after primary biopsy [14, 30–32].

In Europe, chemotherapy is admitted according to the EUROpean Ewing tumor Working Initiative of National Groups 1999 (EURO-E.W.I.N.G. 99) protocol, which describes six courses of vincristine, ifosfamide, doxorubicin, and etoposide (VIDE) as intensive induction chemotherapy for Ewing tumors [33]. Patients are initially stratified into risk groups depending on prognostic factors such as site of metastases and size of tumor. Following VIDE induction and local therapy (surgery or if tumor is not amenable to surgery then radiotherapy), further stratification into standard and high-risk patients takes place depending on the histological response to VIDE. Standard risk patients are randomized for consolidation therapy with either vincristine, actinomycin D, and cyclophosphamide (VAC) or vincristine, actinomycin D, and ifosfamide (VAI); high risk patients for either VAI or high-dose chemotherapy (busulphan and melphalan in combination) followed by stem cell rescue [33]. By applying neoadjuvant chemotherapy a reduction in tumor size can be

achieved. In a retrospective study at our center, a significant reduction of the tumor volume was observed. The mean initial tumor volume prior to chemotherapy was 391.1 ml and the amount after chemotherapy was 49.7 ml (Fig. 58.3) [34].

By this modality complete resection with negative margins can be achieved. Thus, compared to postoperative chemotherapy, preoperative chemotherapy allows a more conservative but radical surgical resection and reduces the risk of intra-operative tumor rupture and tumor cell dissemination [35]. Additionally, it has been suggested, that initial chemotherapy led more limited chest wall resection [36].

In ES/PNET distant micrometastasis must be assumed to be present, therefore all patients receive adjuvant chemotherapy to control the distant disease [30]. There is no survival benefit caused by primary resection of the tumor, however, patients undergoing definitive surgery after initial chemotherapy had a higher frequency of successful resection of the tumor, eventually avoiding radiation therapy to the chest [30].

Surgical resection of CWT should be performed after the neoadjuvant chemotherapy. Grosfeld described that more than 50 % of initially unresectable patients achieved disease-free survival after late resection [37]. Generally, a wide excision of the tumor and involved structures should be performed. In case of the involvement of a rib, the affected rib and the two adjacent ribs and surrounding soft tissue should be resected (Fig. 58.4). Complete resection is the goal, and if this is accomplished with adequate margins, it allows the patient to avoid radiation therapy with its potential long-term complications. Shamberger et al. showed that complete resection of the primary tumor of the chest wall was accomplished more frequently in resections performed after initial adjuvant chemotherapy compared to primary resection (77 % vs 50 %) [30]. In this intergroup study, there was a significant decrease in the proportion of patients requiring radiation therapy to chest in patients with delayed resection [30].

Radiation therapy is recommended just in limited cases only if there is positive margin at a delayed resection or in case of malignant effusion, paravertebral involvement, and apical location of the tumor. It is known, that radiation therapy in doses recommended for the treatment of Ewing sarcoma/PNET has been associated with a significant incidence of secondary tumors in these patients [38]. Furthermore, radiation therapy may be associated with other complications such as pulmonary fibrosis due to damage of large volume of lung [39]. Additional to cardiomyopathy caused by chemotherapeutic agents, like doxorubicin, radiation therapy induces damages to heart and influences coronary artery disease [40].

Prognosis

Appelbaum et al. described 5-year survival of patients with localized extra-skeletal Ewing sarcomas to be about 70 % [41]. In another study, the overall survival rate after a median follow-up

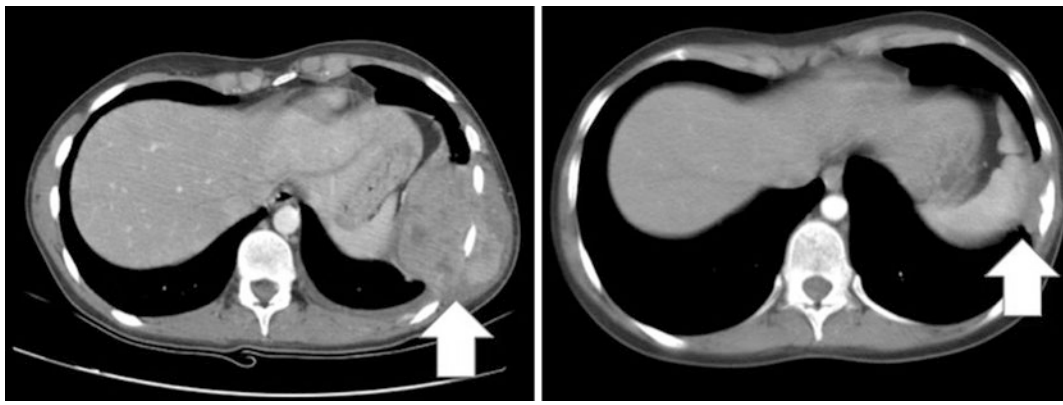

Fig. 58.3 Computed tomography images demonstrating the initial tumor (*white arrow*) prior to chemotherapy (*left*) with a volume of 390 ml and post neoadjuvant chemotherapy (*right*) with a volume of 50 ml (*white arrow*)

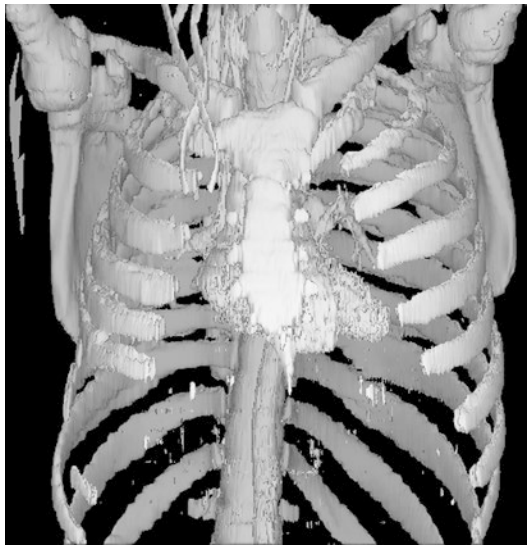

Fig. 58.4 Wide excision of the tumor and involved structures should be performed with the resection of the affected rib and the two adjacent ribs and surrounding soft tissue. Computed tomography demonstrating the wide area of resection in the right lower thoracic cage

of 28 months was 45 % [42]. The overall survival in our cohort is 71 % and 5-year survival rate counts 86 % [34]. By an aggressive multimodal approach a high survival rate can be achieved. The age of the patients has been demonstrated to be an important factor in the determination of prognosis. Younger patients have an improved survival compared to older patients [43].

The presence of a positive margin after surgery in patients with localized disease does not affect the overall survival or EFS [44]. Additionally, recurrences seem to be an unfavorable prognostic factor. Patients who suffer from a relapse have a poor survival rate.

Principles of Surgery

Reconstruction of chest wall defect is most commonly required following resection of chest wall neoplasm but it may also be necessary after infection, radiation injury and trauma. Incidence of local tumor recurrences and positive resection margin has led to performing of extensive surgical resections of the tumor and surrounding chest wall tissue. Small defects can be closed with only soft tissue. However, if structural stability is required, autogenous tissue (such as fascia lata or rib) or prosthetic material (various meshes, metals or soft tissue patches) may be used [2]. In case of Askin tumors, wide excision should include parts of adjacent ribs, muscles and underlying pleura.

For high-grade malignancies, resection involving 3–4 cm margins have been found to be adequate [34, 45]. On the other hand, it is recommended to resect anything that resembles residual or scarred tumor, because in very fibrotic tissue, resulting from chemotherapy, microscopic island of residual tumor have been found to be present [30]. The size and location of the defect influences the decision whether a reconstruction should be performed or not. A thoracic reconstruction is necessary in defects over 10 cm in its greatest diameter in the posterior parts of the chest and over 5 cm elsewhere [2].

Chest Wall Reconstruction

Although chest wall reconstruction remains challenging, the primary goal is to provide rigid fixation without compromising respiration and ideally with minimal foreign body deposits [46]. The first chest wall resection was performed in eighteenth century under difficult conditions, but in present times extensive chest wall resections can be performed due to improved surgical technique, peri- and postoperative care, antibiotics, and refinements in reconstruction techniques [47].

Prior to the surgery, an exact radiographic study with regards to the extent and localization of tumor should be performed. In addition to this, the patient's medical history should be taken in consideration, as any previous courses of radiotherapy, history of other operations, and even smoking may play an important role in planning the reconstruction.

A defect following the excision of one rib does not usually require reconstruction. In these cases, covering of defect by overlying muscles and subcutaneous tissue is sufficient with or without approximation of ribs [13]. Using muscle flaps in soft-tissue reconstruction seems to be sufficient for wound stabilization, whereas in case of large

defects of anterior chest wall a skeletal reconstruction is indicated. Deschamps et al. reported that the decision not to reconstruct the chest wall skeleton depends on the size and location of the defect. Defects less than 5 cm in diameter or posterior defects less than 10 cm underlying scapula are usually not reconstructed unless they are located at the tip of the scapula where entrapment of the scapula can occur during movement of the arm [2].

Even though the necessity of reconstruction remains controversial, it is generally accepted that *en-bloc* resections resulting in a defect larger than 5 cm or four or more ribs will benefit from reconstruction/stabilization. It is recommended to perform a chest wall reconstruction to avoid flail segment induced by "*Pendelluft* principle", in which air flow back and forth between the lungs, resulting in increased dead space ventilation [48].

There are three tenets of surgical resection, which should be borne in mind when performing these procedures [49]. First, a sufficient amount of tissue must be resected. However, there is always a dilemma between having sufficient safety margins and causing a large chest wall defect requiring an adequate reconstruction. Second, in large chest wall defects a rigid reconstruction and replacement of the resulted defect should be applied to avoid a flail chest. Third, to seal the visceral organs, great vessels and pleura, a soft-tissue coverage is necessary. As a wide resection of the chest wall often threatens stability, and reconstruction plays a crucial role in determining postoperative morbidity and mortality. The main principles of thoracic reconstruction are stabilization of the skeletal defect to avoid any paradoxical chest wall movement, obliteration of intrathoracic dead space in order to prevent sepsis, protection of intrathoracic viscera, and soft tissue coverage of the chest wall defect [50]. There are different groups of materials, which can be used to reconstruct the chest wall deformity. In following part, there is a description of those groups.

Autologous Reconstruction

For long time, autologous reconstruction was the method of choice, due to lack of prosthetic materials.

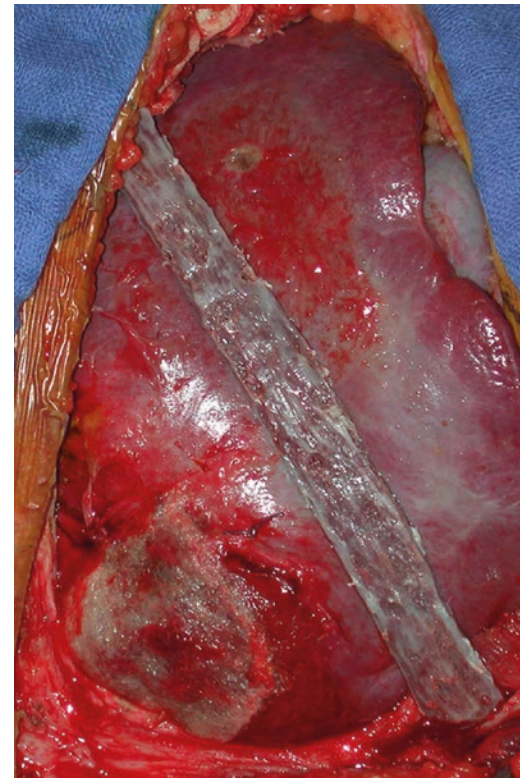

Fig. 58.5 Operative view of a donor rib being secured to the defect created after en-bloc tumor resection. Pain or instability of the donor side should be considered when these procedures are performed in children

The main advantage of autologous reconstruction is the avoidance of alloplastic tissue and a lower risk of infection that may occur due to (re)-vascularization. The main restriction to this method is the limited amount of the available tissue, the resulted instability and pain in the donor site.

Autologous Rigid Reconstruction

Numerous autologous bone grafts have been used for achieving the skeletal stability of chest cage. The most common donor sites for chest wall reconstructions are contralateral ribs or the anterior part of the lateral defect. A rib can be taken entirely, as a segment or split longitudinally. The possible pain or instability on the donor side should be taken in consideration when performing these procedures in children. The donor rib can then be secured to the defect as seen in (Fig. 58.5). Another possible autologous bone for chest wall reconstruction is fibula.

The autologous transplant may be vascularized or should be placed in a well-vascularized bed to facilitate graft survival and osteoconduction. If the graft fails to integrate on the host and the bone is not re-vascularized, the graft will be absorbed over time, leaving behind a firm capsule, which may occasionally be rigid enough to provide the sufficient stabilization and protection [48].

Dast et al. described a new option for autologous anterior chest wall resection performed with a composite thoracodorsal artery perforator free flap with sixth and seventh ribs vascularized on serratus muscle [51]. This option was performed in a patient who underwent subtotal sternectomy with partial resection of the clavicle and anterior arches of the upper three ribs due to sternoclavicular joint chondrosarcoma. In 2010, Chang reported of harvesting of a combined free vascularized iliac osteocutaneous flap and pedicled pectoralis major myocutaneous flap in a patient with a composite defect of sternum and ribs as well as huge skin defect, due to an ablative surgical procedure [52]. In follow-up, a well-incorporated iliac bone (into the recipient bone) and a stable chest can be observed. Good incorporation into the recipient bone, low rate of infection, better vascular supply to a poorly healing wound and avoidance of usage of foreign bodies are advantages of a vascularized bone graft [52]. One advantage of the iliac bone graft is its curved shape, which can simulate the curvature of the chest wall. Based on this fact osteotomy may be considered unnecessary.

Autologous Non-rigid Reconstruction

A variety of techniques are described to provide adequate wound coverage and allow quick healing, rehabilitation, and acceptable cosmesis. For defects not requiring stabilization or for additional coverage of alloplastic tissue and skeletal reconstruction pedicled muscle or myocutaneous flaps are commonly used. Fasciocutaneous, perforator, and omental flaps can also be utilized for reconstruction. Muscular flaps may be deployed as pedicled or as free implant if the area of defect is difficult to be reached with pedicled flaps, or in case of lack of the local muscle, and when a single regional flap is inadequate for covering the chest wall defects.

Muscular Flaps

The coverage with either pedicled or free muscle should be performed to provide well-vascularized tissue to cover the thoracic wound or the used alloplastic materials, to avoid or control infection, to obliterate dead space.

A muscular flap can be deployed as pedicled or as free implant. A free flap is indicated if the area of defect is difficult to be reached with pedicled flaps, in case of lack of the local muscle due to irradiation or previous operations, and when a single regional flap is inadequate for covering the chest wall defects. Arnold et al. emphasized the importance of utilizing non-radiated tissue, because the possibility of developing necrosis is very high, when irradiated muscles are transposed [53].

In this part, we describe the three workhorse muscle flaps in chest wall reconstruction. For this purpose, pectoralis major, latissimus dorsi, and rectus abdominis have been found to be robust and reliable, have consistent vascular supply and anatomy, and have versatility to cover small or large defects and the potential to include overlaying skin paddles [50]. Beside these workhorse muscle flaps, external oblique and thoracoepigastric flaps are uncommonly transposed to reconstruct the chest wall defect.

Pectoralis Major

Pectoralis major is well-suited muscle in reconstruction of anterior central thoracic wall, especially in the upper part of the thorax, but it may also be used to cover supraclavicular, axillary, or lateral chest wall defects. It can be transposed through the intrathoracal cavity for obliteration of dead space [50]. It originates partly from medial half of clavicle as clavicular head and partly from anterior manubrium, sternum, and cartilages of first to sixth rib, as sternocostal head, as well as aponeurosis of the abdominal external oblique muscle, considered as abdominal head. Both heads together insert as flat tendon laterally in intertubercular groove of the humerus.

The pectoralis major receives its innervation by the medial pectoral nerve and the lateral pectoral nerve to facilitate medially rotation, adduction, drawing scapula anteriorly and inferiorly, and functioning as auxiliary respiratory muscle.

The blood supply is provided by the thoracoacromial artery, arising from axillary artery, which divides in calvicular, pectoral, acromial, and humeral branches. The pectoral branch gives rise cutaneous perforators, which supply the skin. Additionally, the three perforating branches rising from the internal thoracic artery enter the pectoralis major via first to third intercostal space. By preserving the perforators, a medially turnover flap can be used. The pectoralis major may be used as rotation-advancement flap to cover dehisced or debrided sternotomy wounds. This flap is based on the thoracoacromial pedicle; therefore the intercostal arteries and internal thoracic artery perforators are ligated carefully. This technique has the benefit of allowing a second sternotomy, if required [53].

The alternative flap is pectoralis major turnover flaps based on the internal thoracic artery perforators. This flap is used to cover the inferior part of sternal wound. To minimize the morbidity to the donor site during the pectoralis major turnover flap, a modification of this method was introduced in which the thoracoacromial pedicle is identified and divided. The medial part of the pectoralis major can then be used as a turnover flap supplied from the internal thoracic artery perforators.

Latissimus Dorsi Flap

Anterior or anterolateral wounds and chest wall defects can be reconstructed with latissimus dorsi, either as muscular or as musculocutaneous flap. The latissimus dorsi, as the largest muscle in the human body with robust vessels, can be used with versatility for coverage of not only extrathoracal but also intrathoracal defects to obliterate dead space [50].

The latissimus dorsi has multiple origins. It arises, from the spinous processes of T7-T12, as the vertebral part, from the thoracolumbar fascia and the posterior third of the iliac crest, as the iliac part, from the 10th–12th ribs, as the costal part, and often additionally from the inferior angle of the scapula, as the scapular part. Latissimus dorsi inserts together with teres minor into intertubercular groove of the humerus and is innervated by the thoracodorsal nerve to facilitate adduct, rotate internally the arm, and extend transversally of the shoulder. It receives its blood supply from the thoracodorsal artery, a terminal branch of the subscapular artery, which arises from the axillary artery. The main pedicle divides into two branches, one running as upper horizontal and the other as descending oblique branch. Based on this vascular anatomy, a partial harvesting of the muscle for coverage, may minimize donor morbidity [54]. A branch of the lumbar artery and a branch of the intercostal artery arise dorsally and perfuse the distal part of the latissimus dorsi. The 8th to 11th intercostal branches are the largest vessels among the secondary pedicles.

The latissimus dorsi can be used as muscle flap for intrathoracic and as musculocutaneous flap for chest wall defects. With a high degree of reliability and an excellent arc of rotation, due to its vascular anatomy, this muscle can be utilized to reconstruct large anterior, anteriolateral, and posterior chest wall defects. In addition, latissimus dorsi remains a workhorse flap when a previously used muscle flap has failed or any further debridement is required. In such cases, infected or ischemic tissue must be debrided completely prior to the coverage in order to achieve a successful and satisfactory reconstruction. Retrospectively, four patients were found whose chest wall defects resulted from Askin tumor were reconstructed with a latissimus dorsi flap in single stage. All four cases showed successful outcome regarding the chest wall reconstruction [34].

Rectus Abdominis Flap

The rectus abdominis muscle (RAM) was first described for the reconstruction of the abdominal wall defects, but soon it was also used to reconstruct anterior chest wall defects either as muscle or musculocutaneous flap. The rectus abdominis, arising from the outer surface of the fifth to seventh costal cartilages, xiphoid process, and the expanding ligaments between xiphoid process and costal cartilages, inserts into the pubic crest. Three tendinous intersections, consisting of aponeurotic extensions of the external, internal obliques, and the transversus abdominis muscles can be found between xiphoid process and umbilicus. The left

and the right rectus abdomins meet each other in the midline at the linea alba. The intercostal nerves (T5-T12) innervate the rectus abdominis.

The two dominant vascular pedicles of the rectus abdominis are superior epigastric artery, as the terminal branch of the internal mammary artery, and deep inferior epigastric artery, arising from the external iliac artery, which gives a medial and lateral braches. The latter two anastomose to the superior epigastric artery.

The rotational flap of the rectus abdominis is especially used in case of large centrally located chest wall defect due to large sternal debridement, central soft tissue defect inferior to xiphoid process and/or when ipsilateral is not sufficient or available due to previous operation [50]. By ligation of the inferior epigastric artery and mobilization of the rectus abdominis, this muscle can be divided from the pubic crest and used as a rotational muscular flap to cover the central chest wall defects.

The blood supply of the abdominal skin and subcutaneous tissue come from several perforators, coming from the epigastric arteries. Therefore, the rectus abdominis can be harvested as musculocutaneous flap. The skin component can be oriented transversally (TRAM) or vertically (VRAM), depending on the geometry and location of the chest wall defect. In our group of patients who underwent chest wall reconstruction due to Askin tumor, there was one patient who needed a TRAM, because of lack of the latissimus dorsi, which was used in previous operations (Fig. 58.6). In this patient, the course according to TRAM was successful and uneventful [34].

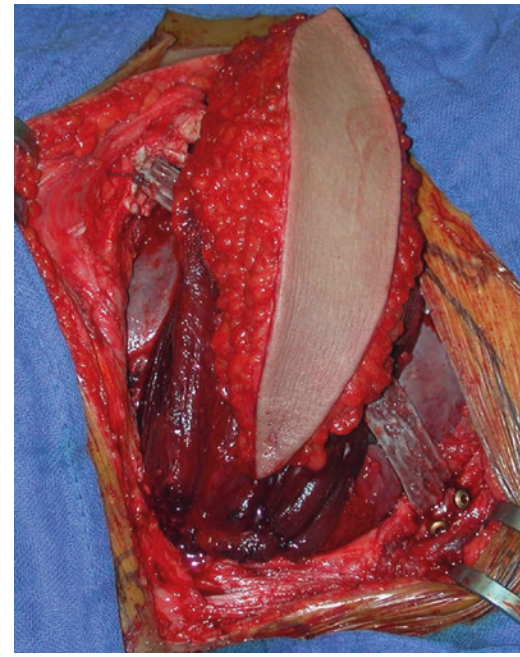

Fig. 58.6 Operative view of chest wall reconstruction after Askin tumor resection using transverse rectus abdominal muscle (TRAM) flap

Other Autologous Materials

Greater Omentum Flap

The greater omentum flap may be used as a pedicled or free flap. Arnold et al. considered the greater omentum as "security blanket" for chest wall reconstruction because of its reliable blood supply of undamaged tissue for patients who showed osteoradionecrosis of the anterior chest wall after irradiation, when the pectoralis major, the latissimus dorsi, and the superior epigastric artery are damaged due to previous surgery or irradiation. They described that greater omentum flap was applied in 10 % of all patients who underwent a chest wall reconstruction and the repair was in reliable fashion [53, 55]. The main advantages of the greater omentum are the double dominant blood supply (based on the right and the left gastroepiploic arteries), the large size and arc of rotation. In addition, the greater omentum is an ideal donor tissue, because there is no postoperative loss of function and low morbidity to donor [56].

The greater omentum can be harvested through a laparotomy or by laparoscopy. In addition, laparoscopy may be advantageous, because the size of the omentum is unpredictable. One major disadvantage of the pedicled omentum flap is the creation of a hernia either to the abdominal fascia or diaphragm.

Tensor Fascia Lata and Dura

Other materials employed as autologous transplants are tensor fascia lata and dura. The thickness of the fascia lata makes it strong biomaterial

for this purpose. It is easily harvested and can be used with or without a skin paddle. Matloub et al. reported that there was no loss of tensile strength in autologous fascia over the 1-year study period and an adequate abdominal wall reconstruction can be achieved. Furthermore, in contrast to Marlex (CR Bard, Murray Hill, NJ) no intraperitoneal adhesions to fascia lata were seen [57]. A successful placement of initially "devascularized" fascia lata in an infected setting has been reported to result in no graft loss [58]. This might be due to the revascularization from the surrounding tissue. The fact that viable fascia seems to be more resistant to bacterial contamination, is important in immunosuppressed hosts and in cases that present major contamination [59].

Microsurgery for Muscular Flaps

Microsurgery can augment blood flow to a pedicled flap and allow the surgeon to transfer free tissue from a distant site. This technique is indicated in case of large defect (with radiation fibrosis), salvage procedure after (partial) loss of regional flaps, intrathoracic dead space, and auxiliary anastomosis for "supercharged" TRAM [60].

Microsurgery provides a safe, successful and reliable method of single-stage chest wall reconstruction with low morbidity for the patient.

Alloplastic Reconstruction

A wide range of possible techniques, prosthetic, and reconstructive options are available today; therefore, it is mandatory to find an individual and appropriate procedure for each patient. Alloplastic tissues are widely available, inherently flexible, and may be fitted to the shape and size of the chest wall defect. Using new methods and synthetic meshes in combination with muscle flaps can achieve a safe, and effective one-stage reconstruction of major chest wall defects.

In 1983, LeRoux and Shama described the ideal characteristics of a prosthetic tissue as follows [61]: *rigidity* to abolish paradoxical chest motion, *inertness* to allow in-growth of fibrous tissue and decrease the likelihood of infection, *malleability* so that it can be fashioned to the appropriate shape at the time of operation, and *radiolucency* to allow radiographic follow-up of the underlying problem. Until now, there is no material, which fulfils all the needed criteria.

By using the alloplastic materials no donor site morbidity is caused. Even if alloplastic tissues are expensive, they are available in large quantities [48] and provide more material than that can often be obtained from patient, also diminishing operation time necessary for harvesting the autologous grafts. Prior to the operation, the vascularity of the recipient site and sufficiency of soft tissue coverage of alloplastics should be explored, in order to avoid any exposure or extrusion of the implant. The soft tissue coverage should be as thick as possible. An appropriate size of implant should be chosen to avoid any tension of overlying soft tissue coverage. The alloplastic tissue has to be secured to a stable adjacent structure, otherwise migration might occur. Other factors, which should be considered preoperatively, are mobility of surrounding tissues, adequacy of pocket dissection and the possible excessive mechanical loading.

Alloplastic Non-rigid Reconstruction

There are variety of reasonably functioning synthetic meshes with different thickness and properties but the optimal choice of prosthetic material is very confusing. Therefore the decisions to use a specific material has been based on a surgeon's individual preference.

Polypropylene Knitted Meshes

Polypropylene knitted meshes are non-resorbable synthetic meshes, which are available as single and double–knit fabrics. Marlex (CR Bard, Murray Hill, NJ) is a single knit fabric which is unidirectional stretchable with tendency towards fraying [49]. Prolene (Ethicon Inc., Somerville, NJ) is another mesh that is double-knitted by a process which interlinks each fiber junction and provides for bidirectional elasticity. This

construction permits the mesh to be cut into any desired shape or size without unraveling.

Both meshes should be sutured under tension and induce intense fibrovascular infiltration, therefore this result in the incorporation into the surrounding tissue [48]. Mansour et al. favor these two meshes for their in-growth and their pliability and achieved satisfactory results when no rigid reconstruction was necessary in a not too large chest wall defect and after all contaminated is resected [47]. A conservative management can be occasionally performed with precaution if polypropylene meshes are exposed or infected.

Polyester

Polyethylene terephthalate (Dacron; Invista, Wichita, KS) is a biocompatible, flexible, nonabsorbable polymer that is used as a mesh (Mersilene; Ethicon Inc.) in abdominal and chest wall reconstruction. Mersilene polyester fiber mesh affords good strength, durability and surgical adaptability, along with maximal porosity for necessary tissue ingrowth and provides for elasticity in both directions.

Biodegradable polyester polyglycolic acid (Dexon; Covidien, Mansfield, MA) is a lightweight and partially absorbable mesh. Polylactic Acid polymers are degraded in the body at physiologic pH over the course of weeks to months. These resorbable polymers are available as mesh sheets for body wall reconstruction. Polyglactin (Vicryl, Ethicon Inc, Somerville, NJ) is a synthetic absorbable undyed knitted copolymer of glycolide and lactide, derived respectively from glycolic and lactic acids. It has been shown to be inert, nonantigenic, nonpyrogenic and to elicit only a mild tissue reaction during absorption.

Polytetraflourethylene

PTFE (Teflon; Gore Inc., Flagstaff, AZ) is an inert and highly biocompatible polymer that is extremely useful in soft tissue augmentation and was the first PTFE graft to be used in plastic surgery. Expanded Polytetraflourethylene (e-PTFE/Gore-Tex; Gore, Flagstaff, AZ) is a pliable, durable, inert, and biocompatible prosthetic material combined with two antimicrobial preservative agents (chlorhexidine diacetate and silver carbonate, both active against Gram-positive and Gram- negative organisms) and also provided of a smooth visceral surface to minimize adhesions and a textured external surface for tissue ingrowth [62]. It has some tissue ingrowth, little inflammatory reaction and almost no encapsulation.

In literature, it is described that PTFE, in contrast to prolene mesh, is much easier to suture, stretch, and mold into the wound and provides a water- and airtight barrier between the pleural and subcutaneous space. Secured 2 mm thick patch with heavy interrupted non-absorbable sutures is recommended. Although there was no reduced rate of seromas, wound infections, and other postoperative complications by using PTFE [2]. Also Gapany et al. favored the usage of Gore-Tex combined with latissimus dorsi mucle flap for large thoracic defects [63], but due to a negligible fibrovascular incorporation into the sorrounding tissue, ePTFE seems to be less resistant to infection, and in such a case a surgical removal is usually required [48].

Proplast (Vitek, Houston, TX) is a polymer of polytetrafluoroethylene and carbon filaments with a porous structure. It was first thought to be favorable but biomechanical failure, infection, inflammation, capsule formation, and extrusion were seen.

Silicone

Since 1950, silicone was used in many reconstructive surgeries due to its inert character, resistance to deformation and degradation, and being non-allergic. It is available as gel, rubber, and solid implants. Meanwhile silicone rubber is very weak and tends to tear, for chest augmentation solid silicone implants are used.

Stainless Steel

Stainless steel mesh, which shows a gird structure, has slight elasticity with appropriate hardness and softness but the metal fibers may cause injuries to the adjacent soft tissue, like lung, heart and large vessels. Therefore, it is described in literature to fold the edges of stainless steel mesh, to secure it to the ribs, and to be covered by Marlex mesh [64]. Yoshino et al. favors the

utilizing of stainless steel mesh in combination with Marlex and muscular flap [64]. Another usage of steel mesh was, when it was added to the methylmethacrylate-Marlex composite to prevent fragmentation of the hardened methyl methacrylate [65]. This technique is not utilized further and no steel meshes are imbedded in Methylmethacrylate (MMM) [66].

Alloplastic Rigid Reconstruction

A rigid reconstruction of the chest wall defect is mandatory to maintain the rigidity of thorax and to avoid the paradoxical ventilation especially in large anterolateral or sternal chest wall defects.

Polylactic Acid Bar/Plate

Miller et al. reported of 25 patients who underwent a chest wall reconstruction, using biomaterials with a total of preoperative infected sites in 10 patients. Fifteen of these had polylactic acid bar (BioBridge, Acute Innovations, Hillsboro, OR). In addition, in 11 of 15 patients bovine pericardium patch (Veritas; Synovis Life Technologies Inc, St Paul, MN) was implanted. In one patient, a removal of only one doublet polylactic acid bar was required, due to inflammatory reaction. Therefore, the author recommends using polylactic acid bar in combination with bovine pericardium patch in infected sites, lateral chest wall and small to moderate sternal defects. Further PLA bar can be applied for single- or double-rib defects with sufficient overlying soft tissue [67].

Lactosorb copolymer plate (Lorentz; Jacksonville, FL) is a bioresorbable plate, consisting of 82 % L-lactic acid and 18 % glycolic acid, which is moldable in warm water and can be cut with a hot knife or scissor. It is available, as solid or perforated plate in different sizes and thicknesses. It shows no late inflammatory reaction, due to its amorphous structure and uniform degradation, due to missing crystallinity. This material provides stability and rigidity, because it keeps its tensile strength for months postoperatively. It has been previously used in craniofacial and maxillofacial reconstruction in children. Tuggle et al. described their 2-year experience with Lactosorb in chest wall reconstruction and considered it as an excellent addition to the available reconstructive materials [68]. Although short-term results seem to be very good, long-term follow-up is missing.

Marlex Mesh- Methylmethacrylate (MMM) Sandwich

In 1981, McCormack et al. described the MMM (Simplx P; Stryker Howmedica Osteonics, Mahwah, NJ) technique for rigid reconstruction [65]. This prosthesis can be adapted to any size, shape or contour of chest wall defect. First two pieces of Marlex are cut slightly larger than the chest wall defect, then some methylmethacrylate is applied on one sheet of Marlex and a stainless steel mesh is added. After pressing the second piece of Marlex into the paste, the "sandwich" can then be molded to conform to body contours. Attention should be paid, because the exothermic process of hardening may develop a temperature up to 140 ° F. In addition intraoperative hypotension and metabolic acidosis secondary to anion replacement with methacrylate may occur [69]. A modification of this technique was published in 2005 by Weyant et al. in which they reported that the stability of the prosthesis was adequate even without the usage of the additional layer of wire mesh [66].

In order to allow some tissue ingrowth and better fluid drainage from the thoracic wall soft tissue into the pleural cavity, the usage of cross-hatching strips of methylmethacrylate instead of entirely solid plate, was introduced [66]. However, a higher rate of wound complication, fracture, tilting or extrusion of the implant has been reported by their use at different Centers [2, 47, 53, 62, 66]. Despite aforementioned points, the long intraoperative time required for MMM to be prepared and shaped to the chest wall defect, and difficult handling, MMM sandwich (with Prolene or Marlex mesh) is recommended to achieve a excellent rigid stability, physiologic and aesthetic success, and low respiratory complications.

Although a variety of materials can be used in the reconstruction of the chest wall defect, there

is no consensus on the most physiologic or efficacious material. Each mesh or composite has its advantages or disadvantages; the decision about the choice of optimal mesh or composite depends on surgeon's preferences and of course on the type, size, and characteristics of the chest wall defect. There is a limitation in usage of alloplastic meshes in case of existing contamination or in patients who were subjected to irradiation previously. Therefore, many investigations are carried out to find an appropriate bioprosthetic material.

Bioprosthetic Materials

A main characteristic of bioprosthetic materials is to support matrix remodeling, cell ingrowth, and new collagen deposition [70]. Therefore, various processing technique have been developed by manufactures. One of the important methods seems to be collagen cross-linking, which prolongs the lifespan of the mesh by adding to the three-dimensional structure of the collagen to mechanically strengthen the matrix and impede degradation by collagenase [71]. When the graft is positively recognized by the host, the process of ingrowth begins with penetration of macrophages and mast cells into a mesh scaffold (neocellularization). Simultaneously, new blood vessels proliferate within the graft (neovascularization). Then fibroblasts are attracted by cytokines and other signaling factors secreted by mononuclear cells. Therefore new collagen synthesis and deposition take place. Importantly, this process has to occur not only at the mesh/host interface but also within the graft itself. By this process ingrowth, incorporation, and new collagen deposition within the mesh are achieved [71]. Bioprosthetic materials are classified based on their origin of harvesting in allograft or xenograft.

By using the bioprosthetic implants, large defects can be reconstructed successfully without any site morbidity and a relatively unlimited material supply. Additionally, allografts are described to be more favorable for radiated or contaminated wounds with potential for cutaneous exposure [48]. The xenografts do not need any rehydration and can be placed in clean or clean-contaminated wounds.

Allografts

Allografts are of human origin and are usually derived from decellularized cadaveric dermis. Decellularization is required to remove the antigenic material of the tissue.

AlloDerm (LifeCell Coropration, Branchburg, NJ)

AlloDerm is a non-cross linked and freeze-dried cadaveric human dermis, which is chemically and physically processed to achieve an acellular tissue with preserved extracellular matrix and basement membrane. The material should be rehydrated 20–40 min prior to implantation. AlloDerm is the most investigated bioprosthetic material.

Holton et al. showed in a study that there was neither incidence of flail thorax nor respiratory distress among operated rabbits. Additionally, No lung herniation or graft dehiscence was found. They reported that 33 % of the AlloDerm group developed seroma compared to 25 % of the ePTFE group without any infection of seromas. Histologically, AlloDerm grafts demonstrated cellular infiltration with significant vascular growth [46]. The material seems to have a low infection rate in contaminated wounds [72, 73] and tolerance of cutaneous exposure without the need for surgical removal [74]. If an infection occurs, it may be treated medically or with local wound care.

Flex HD (Ethicon Inc, NJ) and AlloMax (Tutogen Medical Inc., Alachua, FL)

AlloMax is a non-cross-linked acellular dermal collagen that retains its constituent elastin fibers. It is prepared using Tutoplast (Tutogen Medical Inc., Alachua, FL) process. Flex HD is an acellular hydrated dermis through an alliance with the Musculoskeletal Transplant Foundation (MTF). The tissue is prehydrated and ready to use. On both Flex HD and AlloMax, there are

currently no published human data for chest wall reconstruction.

Xenografts

Xenografts are derived from either porcine or bovine. Although xenografts are available in greater supply and larger sizes, but the risk of cross-contamination with bovine spongioform encephalopathy or porcine endogenous retrovirus even in acellularized xenograft may remain. Many studies have shown that allografts and xenograft differ in pattern and degree of fibroblast penetration, remodeling and the immunogenic response. Armour et al. showed that fibroblast infiltration, necessary for effective graft healing, was supported in a significantly greater number of samples of human acellularized dermal matrix than porcine dermal matrix [75]. On the other hand, immunogenic responses to biologic scaffolds may occur despite thorough cleaning, lysis to achieve decellularization [76].

Porcine Biomaterials

Surgisis (Cook Biomedical, Bloomigton, IN)

Surgisis is derived from porcine small intestine submucosa and was approved by the United States Food and Drug Administration (FDA) in 1999. Helton et al. reported that Surgisis performs well in clean and minimally contaminated area and it had satisfactory short-term outcomes. For dirty and grossly contaminated wounds. However, complications and recurrent hernia were described to be high in critically ill patients with dirty wounds undergoing attempted repair with Surgisis [77]. The graft was modified by the manufacturer and is now marketed as Biodesign. Whether the modifications improve the outcome is subject of further investigations.

Permacol (Covidien, Norwalk, CT)

Permacol surgical implant is a porcine dermal collagen implant, which is decellularized by a gentle process without damaging to the 3D collagen matrix. It is then cross-linked for enhanced durability. Shawn et al. reported of five patients undergoing a chest wall resection due to a primary malignant and following reconstruction with using Permacol. In this group, there was no postoperative implant-related complication [78]. Permacol shows prolonged resorption (incomplete at 6 months), maintained tensile strength, and low rates of infection [79].

Strattice (Life Cell Inc., Branchburg, NJ)

Strattice is a non cross-linked porcine dermis matrix, processed in order to eliminate all cellular elements and to keep the intact extracellular matrix. The matrix provides a scaffold, promoting integration of the connective tissue and thus being revascularized and repopulated by the patient's own cells. It should be soaked around 2 min prior to usage. Itani et al. presented a multicenter single-arm study with patients undergoing a repair of clean-contaminated, contaminated, or dirty ventral incisional hernias. Not unexpectedly, 66 % of patients experienced wound events but no removal of Strattice was required. There were 28 % hernia recurrences, without any correlation to wound events. In this group, a successful, single stage reconstruction could be achieved in over 70 % of contaminated ventral incisional hernias [80].

XenMatrix (Davol Inc., Warwick, RI)

XenMatrix is a Non-cross-linked, regenerative porcine collagen matrix for hernia and abdominal wall repair. In repair of high-risk incisional hernias and traumatic abdominal wall defects with XenMatrix, Byrnes et al. showed that the defects could be repaired with a low recurrence rate of 7.2 % [81].

CollaMend (Davol Inc., Warwick, RI)

CollaMend is a porcine, heavily cross-linked dermal matrix, approved for usage in 2006. Chavarriaga et al. presented the data of 18 patients with complex abdominal wall defect undergoing a reconstruction with CollaMend. They reported that the recurrence rate and postoperative wound complications were 44 % and 38.9 %, including infection in 22.2 %. The removal of the matrix was required in all of the

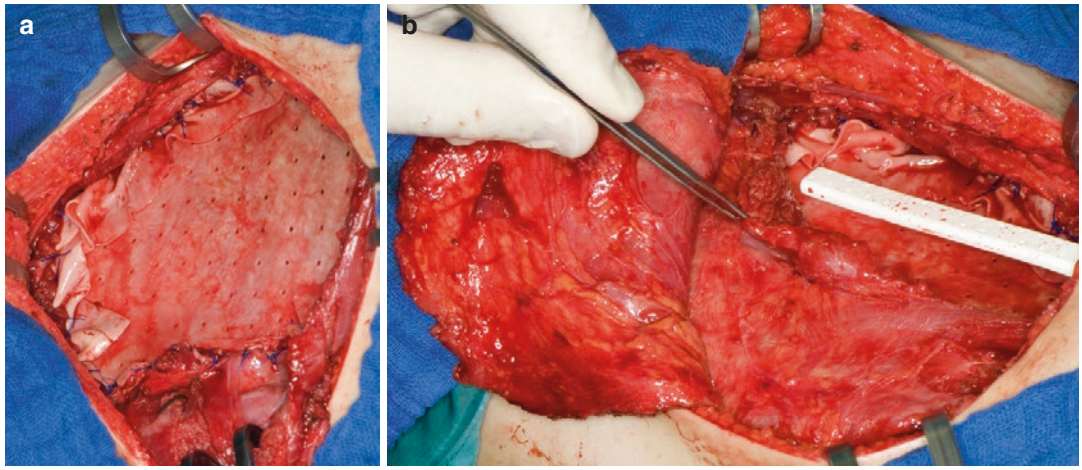

Fig. 58.7 Operative views of (**a**) reconstruction using bovine pericardium collagen (Tutopatch®) and (**b**) placement of a Jacksons drain to minimize seroma formation

patients with infection as a result of encapsulation rather than incorporation of the biologic prosthesis [82]. On the other hand, Federico et al. described in a case report, that good result could be achieved in a patient with traumatic infected chest wall defect [83].

XCM Biologic Tissue Matrix (KenseyNash, West Chester, PA)

XCM Biologic Tissue Matrix is a sterile non cross-linked prehydrated 3D matrix derived from porcine dermis. It undergoes Kensey Nash's proprietary Optrix process, which is strong enough to inactivate viruses and decellularize the tissue, and gentle enough to maintain the bulk of the natural extracellular matrix. It was approved for usage in 2011, and there are no human published series available for review.

Bovine Biomaterials

Tutopatch (Tutogen, Alanchua, FL)

Tutopatch is made of bovine pericardium. The bovine pericardium is subjected to a proprietary, multistep Tutoplast process, which comprises meticulous cleaning of the tissue and gentle solvent dehydration, monitored by continuous quality controls. Saxena et al. reported of 29 children who underwent abdominal wall reconstruction with Tutopatch due to gastrochisis or omphalocele. Twenty-eight patients were treated successfully [84]. In our department, we had a single case of chest wall reconstruction after Askin, using Tutopatch. Although the course of patient was lethal due to multiple metastases, there was no infection, wound event or removal of the graft (Fig. 58.7) [34].

Veritas/Perigurad (Synovis Surgical Inc., St. Paul, MN)

Veritas and Periuard are both bovine pericardial products. Veritas is remodelable and incorporates into the surrounding tissue over a 3- to 6-month period. In 2013, Miller et al. described 21 cases of usage of Veritas. In 3 patients, a removal of Veritas was required and a seconde bovin pericardial patch was placed within 7 days. In two patients, the removal was done prophylactically during debridement of the partially necrotic overlaying muscle flap [67].

Surgimend (TEI Bioscience Inc., Boston, MA)

Surgimend is processed from fetal bovine dermis. It was approved for use in 2002.

In literature it is demonstrated that in the short term, Veritas can be used in complex high-risk hernia repairs, many with contaminated wounds at the time of initial repair, with no mortality and low morbidity [85].

Surgical Techniques

After the extensive tumor resection, rib edges are blunted to avoid any injury to the graft or adjacent viscera. The cavity is then irrigated by antibiotic solution and an adequate rehydrated graft in appropriate size and thickness is applied to the defect with ideal tension. A tense, drum-like feel of the graft is considered as ideal in literature [86]. The graft should be secured with non-absorbable sutures, by either quilting or single interrupted sutures to the surrounding soft tissue or ribs in onlay technique. To allow better reepitheliazation into the graft, the basement membrane surface should face to outer-laying soft tissue [87]. The coverage of the graft with well-vascularized healthy soft tissue is necessary to allow for cellular infiltration and revascularization of the graft. To prevent seroma formation a Jackson-Pratt drain, which remains till the output is less than 25–30 cc over 24 h, is placed between the mesh and the soft tissue.

If there is a contaminated wound, clinical infection may appear; therefore the usage of bioprosthetic material is advisable. In case of infection, it may be beneficial to stage the definitive chest wall reconstruction after several debridement and dressing changes [48]. In this time a negative-pressure wound therapy (NPWT) may be administered. Two other studies recommended single-stage management with immediate debridement and reconstruction in patients with deep and complicated sternal wounds, in order to decrease morbidity, mortality and length of hospital stay [88, 89].

Osteosynthesis

Mandibular Titanium Plate (Stryker-Leibinger, Freiburg, Germany)

In 2006, Sunil et al. reported of using a combination of Gore-Tex and mandibular titanium plates with locking screws (Stryker-Leibinger, Freiburg, Germany) to reconstruct and stabilize the chest wall defect in a 16-year-old boy after a wide PNET resection. The patient underwent a subtotal sternectomy, which involved removal of the body of the sternum and adjacent bilateral 2–3 cm of costal cartilage [69]. In the aforementioned case, stability of the reconstructed chest wall defect and avoidance of the postoperative ventilation could be achieved. In addition, it should be considered that titanium is both osteoconductive and osteoinductive, and when used, it induces the deposition of hydroxyapatite on the surface of the titanium bar intracorprally [90].

Titanium Bar and Rib Clips (Thoracic Osteosynteses System- Stratos; MedXpert GmbH, Heiterheim, Germany)

The Stratos titanium bar, which is available in three different lengths (15, 19, and 23 cm) and with differently angled titanium rib clips (straight 45° or 22.5° left and right), is moldable, feasible, quick to use, and simple to attach to the resected edges of the ribs; therefore, it can be shaped and fitted individually to achieve a rigid reconstruction of the chest wall defect. Gonfiotti favored the Stratos titan bar an option in reconstruction in wide chest wall deformities and applied it in patients who underwent anterolateral resection or complete sternectomy. He combined the Stratos titanium bar with a multiperforated titanium plate for longitudinal reconstruction and stated that an excellent final cosmesis and minimizing of the risk of any wound complication can be achieved [62].

Vertical Expandable Prosthetic Titanium Rib

Vertical Expandable Prosthetic Titanium Rib (VEPTR; Synthes GmbH, Freiburg, Germany) was first introduced for dysplastic thorax deformities, scoliosis, and flail chest. However, it has been also applied to achieve a rigid chest wall reconstruction for large defects after resection of malignant tumors [63, 91]. VEPTR is an implanted expandable device, which can be either affixed to healthy ribs above and below the defect by using the cradles or adapted can be adapted as a hybrid device with pedicle screws for inferior fixation, providing support of the thorax. Campbell emphasize not to use the first rib since migration of the device cephalad may endanger the brachial plexus [92].

The system is amenable up to 12 cm lengthening and can therefore provide stabilization in pediatric population throughout the growth phase. In contrast, MMM and other rigid constructs without a mechanism for growth may minimize the risk of convexity due to the resection and lack of structural support in the short term but they will fix the chest wall and lead to long-term chest wall asymmetry, growth failure, and scoliosis. Stephenson et al. favor utilizing of VEPTR chest wall reconstruction after tumor resection or for primary chest wall deformities. They showed that the reconstruction could be effectively accomplished and satisfactory result could be achieved with no intraoperative complications, no postoperative mechanical ventilation, and no wound complications [91].

Long-term complications reported by Stephenson et al. were breakage of the intramedullary wires, which were used for additional fixation in two patients, with requiring a removal of the broken wire in one patient.

Chest Wall Reconstruction for Sternal Defects

Although pediatric primary tumors of the sternum count less than 1 % of all bone tumors, they are almost always malignant [93]. Also here the complete removal of the affected parts of sternum with 2–3 cm of the adjacent bilateral ribs and consecutive reconstruction of the skeletal defect is the major goal to avoid flail chest, paradoxical respiration and to protect the visceral organs. Beside the usage of methylmethacrylate-Marlex composite and homologous, many other systems are introduced and described in the literature.

Using Stratos titanium bar for transverse and multiperforated titanium plate for longitudinal reconstruction combined with ePTFE in cases after complete sternectomy was favored by Gonfiotti et al. [62]. To their opinion, a rigid reconstruction is not necessary in case of subtotal strenectomy if a small part of manubrium including sterno-clavicular joint or a caudal part of sternum are conserved. A non rigid reconstruction was carried out by usage of ePTFE and muscular flap but also Prolene or Marlex mesh can be used. The workhorse flap for sternal reconstruction is the pectoralis major due to its proximity and versatility. It can be used unilaterally or bilaterlally.

Sunil et al. reported in 2006, the reconstruction by using 2-mm Gore-Tex Dualmesh and locking mandibular multiperforated titanium plates, secured with 2.8 mm screws to the adjacent ribs after subtotal sternectomy which involved removal of the body of the sternum with the adjacent bilateral 2–3 cm of costal cartilages. A mediastinal drainage was left for 48 h. Soft tissue coverage was performed by mobilization of the pectoralis major muscle flaps from either sides [69].

Complications After Chest Wall Reconstruction

Deschamps et al. described a rate of complication up to 46 % [2]. One of the main complications after chest wall reconstruction is persisting infection which might be due to the unfilled dead space, exposed prosthetic materials, remaining necrotic tissue or infected bone or cartilage and visceral fistulas. By performing wide debridement and second stage a reduced mortality to less than 3 % and re-infection less than 5 % could be achieved and therefore there is no the need of serial debridement [94].

Even late complication like bowel obstruction and peritoneal infection may occur, usage of omentum is favored by some authors to control sternotomy infection. Anyhow coverage via muscle or omental flap may resolve persistent infection.

It is mandatory to perform a thorough debridement of necrotic soft tissue and infected bone and cartilage. Furthermore the usage of a rigid plate and the pectoralis major to separate the sternal wound is favored in literature to prevent infected mediastinitis [95, 96]. Formation of seroma was the most common complication in patients who were treated by using human acellular dermal matrixes, as described by Mahabir [48]. Additionally, bulges or material laxity was seen in cases with infection or prolonged cutaneous

exposure and when it was used to bridge fascial gaps.

Another possible complication after chest wall reconstruction might be reduction of the shoulder girdle function due to loss of chest wall skeletal stability, therefore a rigid fixation and restoration of midline support for sternal wounds are promoted recently [95, 97].

Respiratory complications are reported to be present in 20–24 % of patients [2, 47]. In patients with a large chest wall defect paradoxical respiration may occur if an adequate reconstruction is not perform. In this case air flow back and forth between the lungs, resulting in increased dead space ventilation because in a flail segment larger than 5 cm ventilation becomes progressively inefficient and the pulmonary toilette is very poor. Restrictive pulmonary function due to surgery or irradiation, hypoplasia of the soft tissue e.g. mamma and secondary tumors may occur. These late complication can have a tremendous effect on psychological well being of the patient [8].

Atrial fibrillation, flap hematoma, mesenteric ischemia, acute renal failure, pancreatitis and donor site hernia are other mentioned complication after chest wall resection and reconstruction [47].

Painful sternal nonunion is considered as sternal clicking or instability for longer than 3 months in absence of infection or medistitnitis. This late but rare complication requires stable fixation of the chest wall after the thorough debridement of all non viable tissue [98].

It is known that surgery or irradiation may lead to scoliosis due to disarticulation of the costovertebral joint, resection of the posterior segment of the chest wall or lower ribs or irradiation of the heimthorax or hemivertebra. The resulting scoliosis can range from slight to severe deformity leading requiring a surgical treatment.

Summary

In patients with Askin an aggressive multimodal approach after primary biopsy should be applied. This consists of a radical extent tumor resection with a tumor free margin of 3–4 cm and chemotherapy. This fact allows the patient to avoid radiation therapy with its potential long-term complications. The necessity of irradiation should be considered case by case.

A chest wall reconstruction should be performed depending on the location an the size of the resulted defect. Defects smaller than 5 cm anywhere on the thorax do not usually need reconstruction. If not located at the tip of the scapula, no reconstruction is required in posterior defects less than 10 cm. Smaller defect can usually be reconstructed with autologous tissue alone.

In large chest wall defects a rigid reconstruction and replacement of the resulted defect should be applied to avoid a flail chest. A shorter overall hospital stay and reduced ventilation dependence can be achieved in these patients.

The choice of prosthetic material is often based on surgeon's preference because there is no substance fulfilling all criteria. There is a general agreement in literature, that contaminated wounds are not reconstructed with synthetic meshes but rather with bioprosthetics.

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Operative Options in the Management of Cleft Sternum

59

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Introduction

Since the 1850's, fascination with cleft of the manubrium has been evident because the heart and great vessels visibly beat under the skin. This produces an alternate protrusion and retraction movement upon respiration, specifically during coughing or the Valsalva maneuver. In this challenging anomaly, the underlying mediastinal structures (heart and great vessels) may easily be injured by external trauma. In addition, it is an unpleasant cosmetic deformity and quite alarming to the young patient's family [1].

Sternal cleft (SC) is a rare congenital anomaly of the sternum. It consists of a defect, of variable degree, arising from the incomplete fusion of the sternal bars in embryonic life. Immediate correction in neonates or in infancy is beneficial in terms of providing skeletal cover with little or no need for any prosthetic material for reconstruction. In the absence of internal midline defects, one should aim to provide complete correction when the chest wall is still compliant, and cardiorespiratory compromise is not expected after correction.

The first surgical correction was proposed in 1888 [2], but was successfully performed only in 1947 [3]. Despite its long-term description, articles usually report only small series. Acastello et al. [4] found that SC accounted for 0.15 % of all chest wall malformations. Other defects, such as vascular dysplasia, PHACES Syndrome (which go for posterior fossa abnormalities, hemangiomas, arterial lesions, cardiac abnormalities/aortic coarctation, abnormalities of the eye, and sternum defects), midline fusion defects including the abdominal wall, and Cantrell's pentalogy are possibly associated with SC [5].

An antenatal diagnosis with ultrasound or with magnetic nuclear resonance is likely, particularly when the clefts are extensive and associated with other conditions such as diaphragmatic hernia, abdominal wall defects or ectopiacordis are nowadays possible and evaluable. Quite often, the diagnosis is obvious at the time of delivery, in both isolated clefts and in extensive variants [6].

Although primary repair in the neonatal period, according different authors, is the best management [6, 7], in our series it was not always possible because some patients were referred to us as a child or as adolescents. Our initial experience were reported in 1998 [1], and 2009 [7], but in this chapter contemporary surgical technique that are preferred have been elaborated.

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Embryology

The sternum is of mesodermal origin and has a common origin with the pectoral musculature. During the 6th week, it arises from paired longitudinal concentrations of mesenchymal tissue located on each side of the anterior chest wall and called "sternal bars". In sequence, these sternal bars migrate towards the midline and fuse to form the sternal plate. This fusion begins at the cephalic end of the sternal bars and progresses in a cranio-caudal direction and is completed by the 10th week [8].

After maturation into cartilage, the sternum undergoes several transverse divisions into a series of six cartilaginous segments called "sternebrae". There is one segment for the manubrium, four for the body and one for the xiphoid process. The sternum ossifies gradually from these cartilaginous precursors. Union of adjacent "sternebrae", initiated through a central osseous bridge, progresses through anterior, lateral, cephalocaudal, and posterior domains to achieve bone formation. A similar early disturbance of the midline mesodermal structures at this stage along with the cutaneous tissue has also been thought to be the cause of the association of sternal malformations with vascular dysplasias such as haemangiomas [9].

Additionally, the Hoxb gene might be involved in the development of SC [10] and the "sternebrae" do not always ossify to the same degree so there is a wide variability in the number and configuration of the ossification centers of the developing sternum. Furthermore, the ossification may be asynchronous. The failure of this physiologic epiphysiodesis of the sternal bars leads to congenital anomalies. The most frequent defect is the upper cleft. Inferior clefts are usually associated with other abnormalities of midline fusion. Complete cleft sternum has only been reported on a few occasions as well as sternal fenestration.

SC can be classified as complete or incomplete [11]. A complete cleft is the rarest form and fewer than ten cases have been reported in the medical literature [12, 13]. Incomplete clefts are subdivided into superior or inferior ones. The latter may occur as an isolated entity, although it is usually associated with other developmental defects of the anterior chest wall such as ectopia cordis, or a combination of defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart in Cantrell's pentalogy. In the incomplete superior type, a bridge of bony tissue exists at the lower ends of the sternal bars, joining the two edges of the cleft in a U shape (proximal to the fourth cartilage) or V shape (reaching the xiphoide process). These last two types are frequently presented as an isolated entity [14].

Surgical Techniques

Diagnostic work-ups include computed tomography scans or MRI evaluation of the chest and abdomen, focusing on those rare but complex associated anomalies. They are quite helpful in prognosticating the extent of the lesion, and directing the appropriate surgical therapy.

There is a tendency to write in the literature to recommend performing the surgical correction of sternal clefts in the first days or weeks of life. At this age structures are less ossified, more flexible, the size of the defect is small and easier to close. The only reason to postpone the procedure is when there are major cardiac malformations or clinic contraindication. However, in the reality of our country this never happens, we almost always receive all the patients or were ask to a second opinion after months or years after the neonatal period.

Since the patients have no critical disorder they are referred later in life. In these situations, in adolescents or young adults, foreign material can be or usually utilized as titanium bars, prolene mesh, stainless steel mesh, acrylic, silicone and Teflon associated to autologus costal cartilages or tibial periosteum. Our preference is to close the defect primarily utilizing only or specifically autologus tissues [15].

In our preference the sternum is accessed through a midline skin incision, exposing the defect and the fused sternum caudally. The pectoralis major muscle is freed from its insertions in the sternum. With the defect completely exposed

the periosteum is incised and raised anteromedially all along the "U" or "V" defect and sutured in the midline if possible.

The bars of the sternum are approximated with three or four unabsorbable stitches close to the midline. If there is extreme tension in the suture line, two to four chondrotomies can be made bilaterally until the base of the "U" or "V" defect, bending the upper double sternum bars together.

When the sternum cannot be approximated without tension a cartilage graft, harvested from the near rib, is embedded in the middle of the defect. It can be stabilized with a sheet of Marlex or absorbable mesh. The pectoralis muscle is ressutured over the defect. Several techniques were developed to repair the cleft, but we believe that periosteal flaps, muscle mobilization and cartilage or bone graft stabilized with sutures, Marlex or absorbable mesh can be used with this objective.

The periosteal flaps bridge offers an anatomic subtract for osteogenesis allowing a neosternum remodeling. Our proposal is based on two previous concepts. One of them is the use of autogenous tissue without the use of any synthetic material. Another concept is the primary approximation of the sternal bars. Surgery was done through a vertical midline incision, as mentioned before and the sternal edges were well defined. In some patients, the inferior fusion between the sternal bars is divided and further mobilization is carried out to isolate the sternal edges from both sides [1].

The periosteum is incised on the opposing edge of the sternum on both sides to create both raw surfaces for union, and a posterior sternal wall (PSW) with the periosteal flap. A sliding flap is created between the second until the fourth or fifth costal cartilages, and the sternal bars from both sides are advanced to meet in the midline. In order to create a PSW the periosteum of each bar is incised on its lateral border and elevated from its anterior and medial surfaces. Both flaps are then turned upside down and sutured together in the midline with 3–0 absorbable sutures. This provides in the majority of the patients a thick and stable PSW [7].

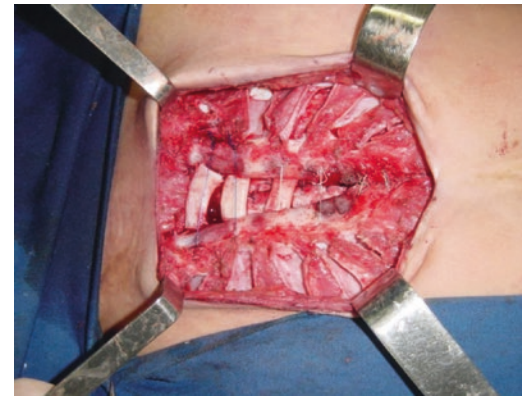

Fig. 59.1 Surgical view of the PSW technique

Each gap that still remain between the two sternal bars, is then filled with two or three chondral grafts that were removed subperichondrially with attempts to preserve the perichondrial sheet's integrity on both sides and at the level of the cleft. After that, the distal end of the cleft (an existing bridge of cartilaginous tissue at the xiphoid area) is resected to facilitate the approximation of the sternal bars. The latter are approximated with heavy stainless steel wire (Fig. 59.1). The chondral grafts are kept in position by these sternal wires. Finally the perichondrial sheets are closed bilaterally.

Internal Organs Compression

Cardiac compression is one of the most threatening complications of SC reconstruction. It is more frequent in wide defects. For this reason one advantage of using periosteal flaps is the enlargement of the thoracic perimeter/diameter. Autologus repair with rib grafts can be accomplished safely without the use of prosthetic material.

This technique allows normal growth of the chest wall and provides sufficient strength and rigidity in the neosternum. In three patients, however, Marlex mesh was required, not as a substitute for the chest wall, but as a bridge for muscle (pectoralis major) approximation. Absorbable meshes that are commercially available today can be used for this purpose.

Cervical Muscle Sparring

In our series it was rarely necessary to mobilize cervical muscles to stabilize the superior thorax inlet (Fig. 59.2). It was necessary in only one of our patients to suture the sternohyoid and sternothyroid muscles or to slide the origins of the sternocleidomastoid medially to "stabilize the thorax aperture and elude the tendency toward herniation of the lungs and pulsation in the neck," as proposed by Asp and Sulamaa in 1961, modified by Daum and Hecker in 1964 [2], and also used by Greenberg and colleagues in 1991 [16].

On the other hand, it can be agreed with the Greenberg and colleagues and with Salley and Stewart [17], regarding the advantages of contact between the cartilaginous tissue of the two sternal bars, when the periosteal flaps are raised to facilitate primary union. We also concur that mobilization of the pectoralis muscles may further protect the repair site and reducing site by reducing lateral tension of the advanced rib segments due to approximation of the sternal bars.

Cartilage Mobilization

Regarding cartilage mobilization, it is assumed that contact between the cartilaginous tissues with sternal bars can facilitate sternal fusion. One of the earliest successful procedures for correction was devised by Sabiston in 1958 [11]. In this procedure, by utilizing multiple bilateral sliding chondrotomies the sternal halves were brought together while simultaneously increasing chest wall dimensions and flexibility. Another advantage, using the chondral grafts to fill all the gaps between the sternal bars with a solid and resistant tissue, a real protection can be provided from the first post-operative day. With the passage of time there will be remodeling and transformation into a thick layer and calcified tissue to generate a new sternum.

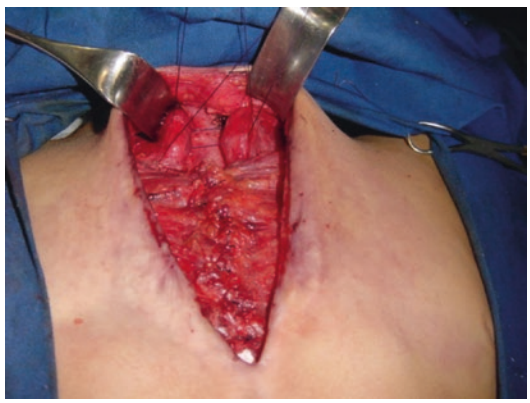

Fig. 59.2 Surgical technique to mobilize cervical muscles

Pectoralis Major Mobilization

For this the muscles are approximated in the median plane. This simple and easy maneuver seems to protect the repaired thoracic wall and to reduce lateral tension secondary to the approximation of the sternal bars. The detached pectoralis major muscle are sutured back on the area of the costal slide to close the dead space on the cartilages on both sides. A single mediastinal drain is preferably left *in situ*, and a further suction drain is left in the subcutaneous plane for further wound closure.

Knox and colleagues [18], in 1994 stated that "autologous repair with rib grafts can be accomplished safely without the use of prosthetic material which reduces the risk of infectious complications while allowing normal growth of the chest wall and providing sufficient strength and rigidity in the neosternum, with excellent cosmetic results." In few (03) patients of our series, however, smalls strips of Marlex mesh (2x3x3 to 2x3x4 cm) were used, not as a substitute for the chest wall, or at the level of the bones and intercostal muscles, but as a bridge for the major pectoralis muscles that could not be sutured in the middle line, despite wide mobilization.

Prosthetic Material

It is our preference not to use prosthetic materials in sternal cleft reconstruction, considering the risks of infection and the inability of these inert materials to remodel themselves according to the

patient growth [15]. Hence the emphasis is laid towards the feasibility of repair without using prosthetic material regardless of patient age. The confirmation that a new sternum can be formed over a biological substrate as can be demonstrated on late postoperative computed tomographic scans (Fig. 59.3).

Primary surgery is the treatment of choice, and should be performed usually soon after diagnosis is made and if possible without the use of prosthetic material. If a simple advancement of lateral sternal bars is ineffective or impossible, in case a loss of tidal volume or restriction of thoracic contents is noticed, a lateral release is usually necessary and our technique of the PSW associated with sliding chondrotomy or remove parts of the cartilages and using as a chondrol graft, is effective in achieving the release of sternal bars to be approximated well enough to avoid the associated restrictive effect.

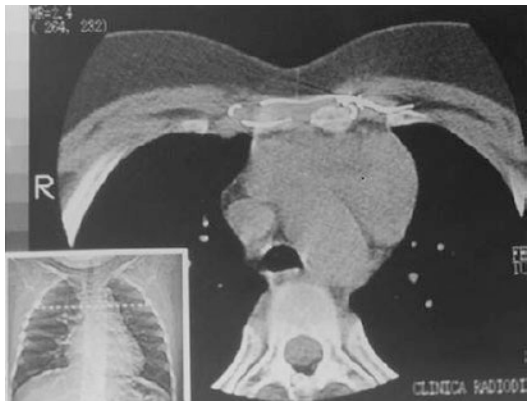

Fig. 59.3 CT Scan of one patient 8 years after the procedure (From Campos JRM, Filomeno LTB, Fernandez A, Ruiz RL, et al. Repair of Congenital Sternal Cleft in Infants and Adolescents. *Ann Thorac Surg*, 1998;66:1151–4. With permission from Elsevier)

Personal Experience

Our series consists of 22 patients (16 females and 6 males), with ages between 1.5 to 19 years. Eighteen of 22 patients had only SC, two concomitant pectus excavatum, one subaortic obstruction and one had Cantrell's pentalogy (Table 59.1).

Since 1999 [7], our preference has shifted to a single technique. Using this technique, 14 patients have been managed (Table 59.2) in which the posterior sternal flaps are rotated to the median plane (PSW), followed by chondral grafts. Chondral grafts and stainless steel wire are used to fill the gap between sternal bars. One patient is being observed as we await for the parents' consent for surgery. Another patient who presented at a later age, two techniques were used, the Nuss technique to correct the severe pectus excavatum and the PSW to manage the SC. It is recommend to close the perichondrial sheets (Fig. 59.4) with interrupted suture using absorbable material and the placement of suction drains posterior to the pectoralis major muscles. Skin is closed always with intradermal sutures.

Table 59.1 Initial experience, first 8 cases

1	Male	11	79	U-superior + PEx	Chondrotomies + resection
2	Female	2	85	Total	Sliding chondrotomies
3	Female	1.5	86	Total + sub. obstr.	Sliding chondrotomies + aortic
4	Male	1.9	91	U-superior	Sliding chondrotomies
5	Female	12	92	V-superior	PSW + chondral grafts
6	Female	4	93	V-superior	PSW + chondral grafts
7	Female	6	97	U-superior	PSW + chondral grafts
8	Female	19	97	Inferior + Caltrell	PSW + chondral grafts + Cantrell
n	Sex	Age	Year	Diagnosis	Technique

From Campos JRM, Filomeno LTB, Fernandez A, Ruiz RL, et al. Repair of Congenital Sternal Cleft in Infants and Adolescents. *Ann Thorac Surg*, 1998;66:1151–4. With permission from Elsevier

Table 59.2 Experience, during the last few years, until the number 15, all the patients were published at the European Journal of Cardio-Thoracic Surgery

9	Female	5	99	V-superior	PSW + chondral grafts
10	Female	14	2000	V-superior	PSW + chondral grafts
11	Male	7	01	U-superior	PSW + chondral grafts
12	Female	9	03	V-superior	PSW + chondral grafts
13	Female	6	05	U-superior	PSW + chondral grafts
14	Male	12	06	V-superior	PSW + chondral grafts
15	Female	4	07	V-superior	PSW + chondral grafts
16	Female	4	09	U-superior	PSW + chondral grafts
17	Female	5	11	V-superior	PSW + chondral grafts
18	Female	6	12	U-superior	PSW + chondral grafts
19	Male	1.5	12	V-superior	Observation
20	Female	6	13	U-superior	PSW + chondral grafts
21	Male	12	13	Inferior	PSW + chondral grafts
22	Female	19	13	U-superior + PEx	PSW + chondral grafts + Nuss
N	Sex	Age	Year	Diagnosis	Technique

Campos JRM, Das-Neves-Pereira JC, Velhote MC, Jatene FB. Twenty seven-year Experience with Sternal cleft Repair. How-to-do-it. *Eur J Cardio Thorac Surg*, 2009;35:539–41. With permission of Oxford University Press [7]

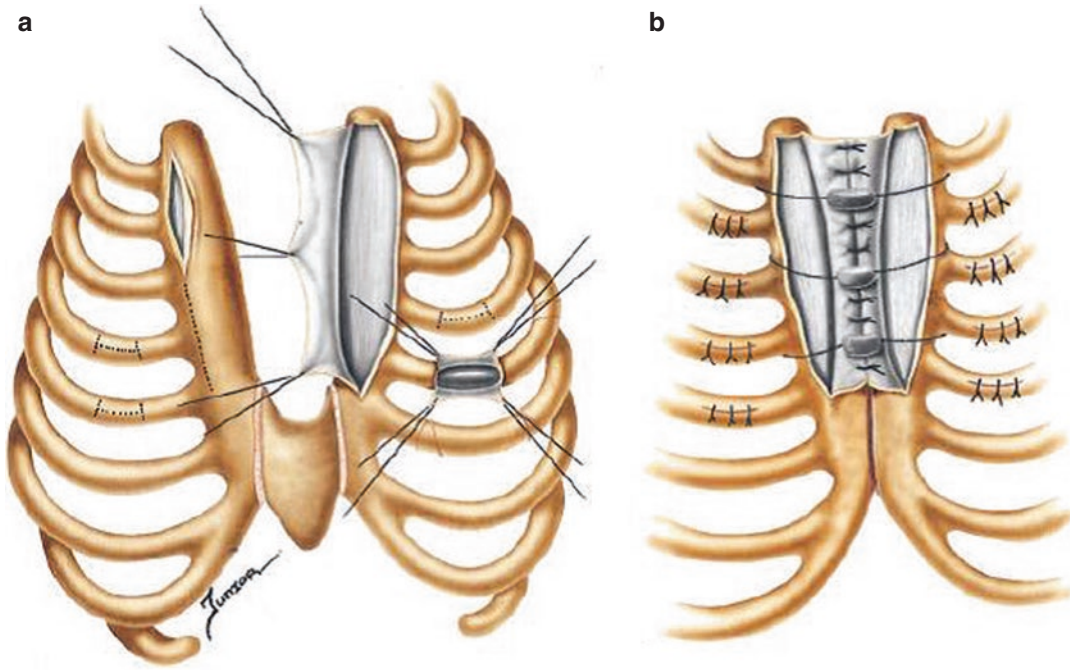

Fig. 59.4 (**a**) initial techniques (**b**) final aspect of the surgery. Posterior Sternal Wall (PSW) technique (From Campos JRM, Filomeno LTB, Fernandez A, Ruiz RL, et al. Repair of Congenital Sternal Cleft in Infants and Adolescents. *Ann Thorac Surg*, 1998;66:1151–4. With permission from Elsevier)

There is no explanation that we know for the marked female predilection in our series with 06 (30 %) boys and 16 (70 %) girls (Tables 59.1 and 59.2). However, it is similar to that of Gorlin and colleagues [19] and Heron and colleagues [20], who found the number of female patients was

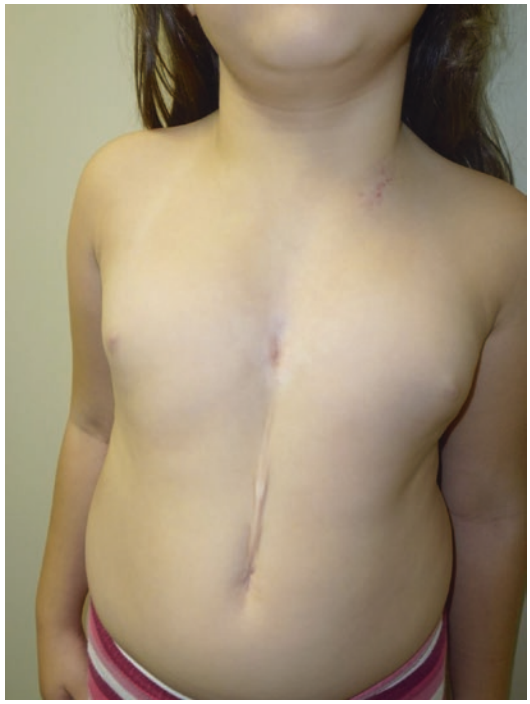

Fig. 59.5 Female patient with a sternal defect accompanied by a supraumbilical raphe

significantly greater when a sternal defect was accompanied by a supraumbilical raphe but not by facial hemangiomas (Fig. 59.5).

In addition to being effective, our procedure of building a PSW with periosteal flaps raised from the sternal bars, together with the chondral grafts, prepares a natural bed for new bone formation which, despite the reabsorption of the grafts, will provide a genuine bone sternum to protect the mediastinal structures.

Results

Postoperative follow-up ranged from 4 months to 30 years. There was no mortality. The mean hospital stay was 6 – 9 days. Early postoperative complication was subcutaneous fluid collected in two patients, but without infection. In long term follow-up up to 6 months one partial piece of Marlex mesh had to be removed because it becomes exposed (Fig. 59.6). All patients have a good mediastinal protection and

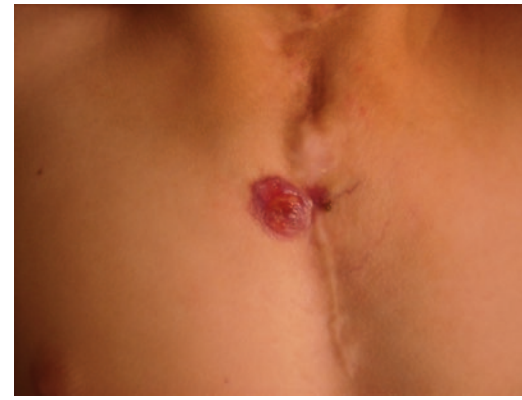

Fig. 59.6 Long term follow-up up to 6 months a partial piece of Marlex mesh becomes exposed

are satisfied with the aesthetical aspects of the surgery.

In the late postoperative period, it has been observed in CT scans in some of our patients, that the posterior wall becomes like a structural base. With the use of the chondral graft over it, rigidity is immediately obtained and after which the development of fibrous and osseous tissue commences to build a new sternum. Some of these patients have more than 25 or 30 years follow up demonstrating the presence of an almost perfect sternum. There are some concerns as certain degrees of pectus excavatum may be associated with repairs, as reported by Acastelo et al. [4]. However if the sternal bars are placed close to the middle line and 3–4 sutures are used to secure them with the chondral grafts, the development a postoperative pectus excavatum can be minimized.

Conclusions

Whether dealing with older children or young adults, the rigidity of the chest wall and the lack of new space for accommodation of the intrathoracic organs must be considered, especially when the defect is too large to be closed directly. The heart is the least tolerant organ to reduced space inside the thoracic cage. Enlargement of the thoracic cage diameter is the main reason for our choice to use the PSW, apart from the fact that in older groups approximation of the sternal bars is almost physically and anatomically impossible.

The procedure of building a PSW with periosteal laps raised from the sternal bars, together with the chondral grafts, prepares a natural bed for new bone formation which, despite their absorption of the grafts and will provide a genuine bone to build a new sternum to protect the mediastinal structures. Also according to the reported literature [14, 16, 18, 20, 21], it is always better to avoid the use of prosthetic materials, considering the risks of infection and mainly the inability of these inert materials to grow with the patient.

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Part IX

Other Chest Wall Deformities: Surgical Techniques- Rare Chest Wall Deformities

Repair of Cleft Sternum

60

Laura Jackson and Dakshesh Parikh

Technical Highlights

Complete and partial cleft sternum requires either primary repair, or repair by bridging the gap using native musculoskeletal tissue or synthetic material. When performed early in the first few months of life primary closure is usually achievable. Late reconstruction, when the chest wall is less compliant, requires additional manoeuvres in order to accommodate and protect the vital organs within the thoracic cavity.

The main technical highlights of cleft sternum repair are:

(a) Ventral midline incision for complete and superior clefts, or transverse incision for isolated inferior clefts
(b) Dissect the skin and subcutaneous tissues
(c) Dissect the medial attachments of the pectoralis major. Dissect rectus abdominis from the sternal bars and costal insertions for inferior and complete clefts only
(d) Blunt dissection to separate the pericardium and pleura from the sternal bars as far as the intercostal spaces
(e) Incise sternal bars along their medial edge and raise periosteum anteriorly and posteriorly
(f) For children with superior sternal clefts either excise a wedge from the sternal bridge or perform complete excision in the sagittal plane
(g) Multiple heavy monofilament absorbable sutures are passed around the sternal bars
(h) Retrosternal suction drain is placed
(i) Sutures are tied sequentially from inferior to superior while monitoring for respiratory or cardiovascular compromise
(j) Pectoralis major and rectus abdominis muscles are approximated over the sternal repair using absorbable suture
(k) Subcutaneous and cutaneous layers are repaired with absorbable suture
(l) In the event primary closure cannot be achieved a number of alternatives can be employed to reconstruct the anterior chest wall

Indications

(a) **Protection**: Reconstruction of the anterior chest wall provides protection to the mediastinal vessels and viscera which are otherwise vulnerable to injury.

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(b) **Management of paradoxical respiration**: Paradoxical movement is often asymptomatic but may be associated with recurrent chest infections, dyspnoea and intermittent cyanotic episodes.
(c) **Cosmesis**: The pulsation of the heart is seen through the overlying skin, which may become thinned and ulcerated. In some children pectus excavatum may be an accompanying feature.

Pre-operative Considerations

The following assessments and investigations should be considered pre-operatively:

1. Clinical pre-operative photograph
2. Pre-operative radiological imaging to delineate the chest wall malformation and assess the defect: Plain X-rays (AP and Lateral); Chest CT scan with intravenous contrast is essential to assess the defect and thoracic anatomy. Occasionally MR angiogram is advisable to demonstrate vascular anatomy.
3. Pre-operative anaesthetic assessment
4. All children should have cardiology consultation, an electrocardiogram and echocardiogram to diagnose or rule out associated congenital cardiac malformation
5. In the presence of a cardiac defect correction can be carried out simultaneously in certain instances
6. In older children lung function tests are performed to evaluate pulmonary status
7. Neuroradiologic imaging and/or opthalmological examination should be undertaken in children with a high suspicion of associated anomalies
8. In children with cutaneous haemangiomas a laryngotracheobronchoscopy should be performed to exclude sub glottic haemangiomas

Special Instruments

A standard thoracic set of instruments will be required. The availability of necessary synthetic material should be established prior to the procedure.

Surgical Technique

The patient is placed in the supine position. The same incision is used in both males and females. Female patients with sternal cleft have normal breast development. Care should be taken during dissection to avoid injury to the mammary glands. A midline skin incision extending the length of the defect is performed. Skin with subcutaneous tissue is dissected from underlying mediastinal structures/pericardium and flaps are raised on either side with sharp dissection and/or by electrocautery to expose the sternal bars. For superior clefts the sternoclavicular insertions of pectoralis major are raised on either side, while for inferior and complete sternal clefts the insertions of the rectus abdominis muscles are also mobilised and reflected to expose the sternal bars and costal cartilages. Blunt dissection is used to free the pericardium and pleura from the posterior aspect of the sternal bars up to the intercostal spaces so that sutures can be placed without injuring these structures.

The chest wall is first assessed to determine whether primary closure is achievable without cardiovascular compromise. If this is deemed possible the sternal bars are incised medially along their length and the periosteum raised anteriorly and posteriorly for a short distance.

In a superior sternal cleft it may be necessary to either excise a wedge from the inferior sternal bridge or alternatively completely excise the inferior sternal bridge.

Multiple heavy polydioxanone (PDS) sutures should be placed around the sternal bars to approximate them and close the defect. The sutures are tied sequentially from inferior to superior in close consultation with the anaesthetist ensuring there is no evidence of cardio-respiratory compromise.

A retrosternal drain is placed, the pectoralis major and rectus abdominis muscles are approximated over the sternal repair. The subcutaneous and cutaneous layers are repaired using absorbable sutures.

Procedure Overview (Figs. 60.1, 60.2, 60.3, 60.4, 60.5, 60.6, and 60.7)

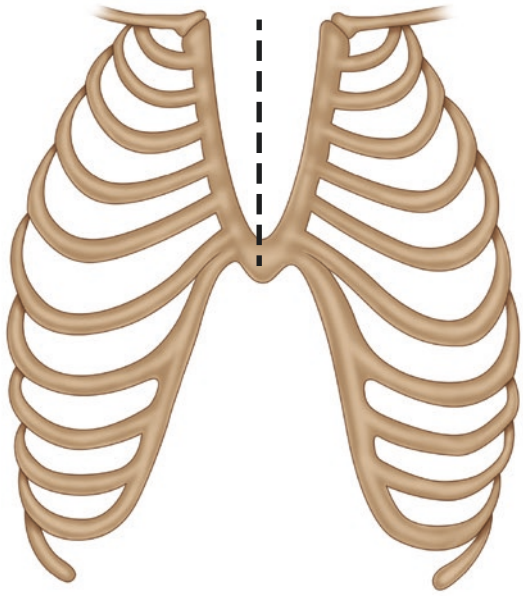

Fig. 60.1 Ventral midline incision for complete and superior clefts to expose the length of the defect. A transverse incision may be used for isolated inferior clefts

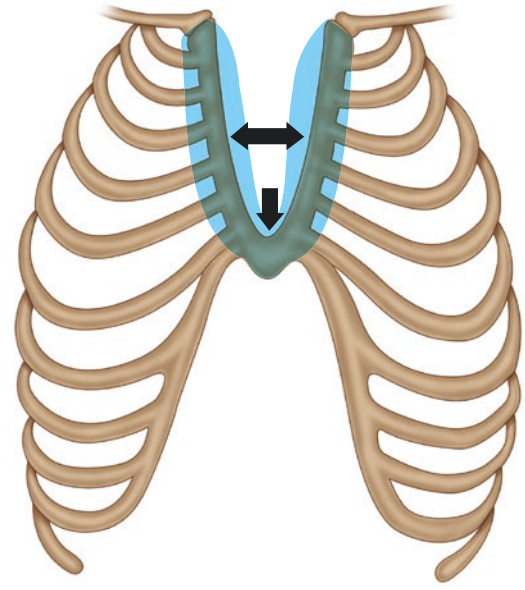

Fig. 60.3 Blunt digital dissection is used to separate the pleura and pericardium from sternal bars thus creating a space to pass sutures posterior to the sternal bars. Care should be taken to avoid pleural or pericardial tears

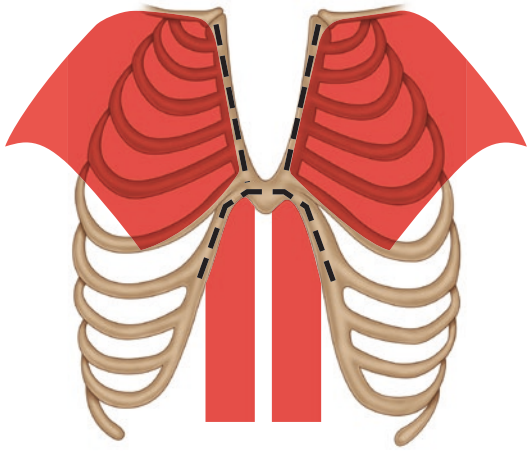

Fig. 60.2 Dissect the sternocostal head of the pectoralis major for all types of sternal cleft. Dissect rectus abdominis from the sternal bars and costal insertions for inferior and complete clefts only

Postoperative Management

1. Monitor vital signs and fluid balance.
2. Cardiac monitoring is required.
3. Broad spectrum intravenous antibiotics: prophylactic for 48 h for primary repair and up to 5 days if grafts are employed for reconstruction.
4. When recovered from the anaesthesia patients are allowed to feed and increase to full feeds as tolerated.
5. Analgesia: Analgesia is required postoperatively routinely and continued as judged by the pain score. Initial morphine infusion with nurse led top up is administered. Subsequently, either oral/rectal/IV paracetamol, with anti-inflammatory medications or oral/rectal codeine phosphate is prescribed.
6. The medistinal drain is removed 48–72 h following surgery.
7. Mobilisation is stimulated as soon as feasible in children.
8. Chest x-ray is advisable to rule out pneumothorax the next day.
9. Discharge is generally feasible in most cases within 3–5 days in uncomplicated cases.
10. Follow-up of these patients is essential with a view to judging the effectiveness of the reconstruction. Anatomical and cosmetic outcome can be investigated by AP and lateral chest x-ray, and in the case of reconstructive grafts

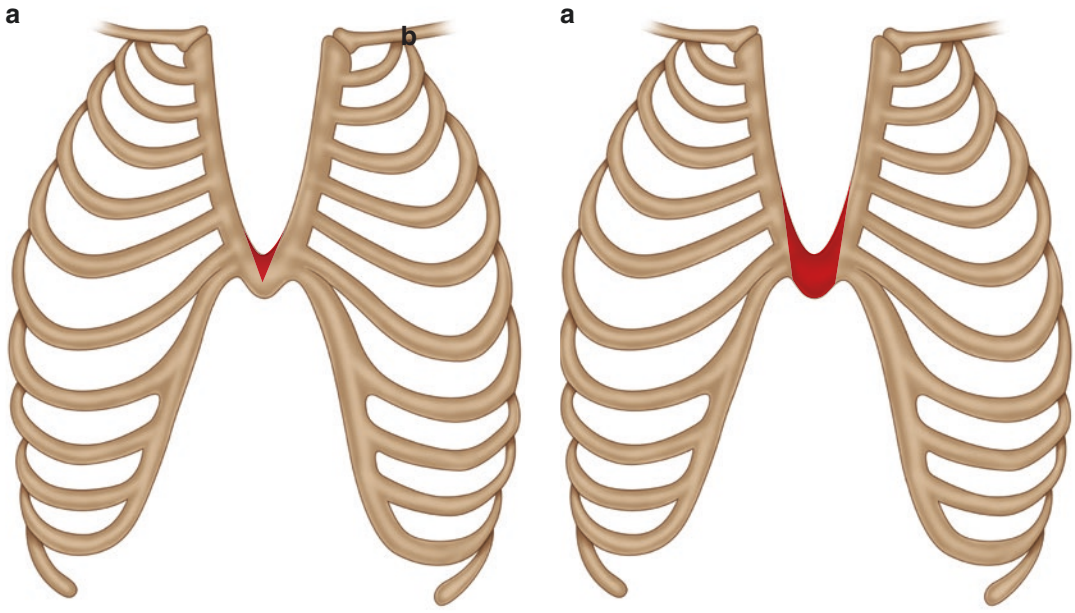

Fig. 60.4 (**a**) For a U-shaped superior sternal defect a wedge can be excised from the inferior sternal bridge to aid closure. (**b**) Alternatively the bridge can be completely excised in the sagittal plane thus converting it to a complete cleft

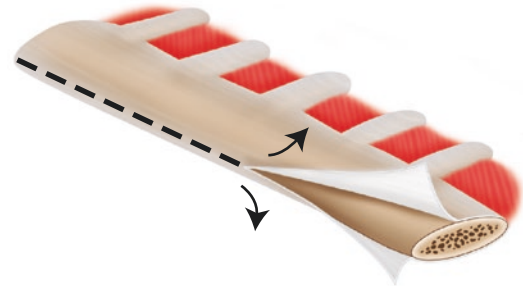

Fig. 60.5 Using electrocautery incise the sternal bars along their medial edge and raise periosteum anteriorly and posteriorly to expose underlying bone

3D CT scans should be considered. In older children lung function test will demonstrate the physiological outcome.

Results

There are only a few retrospective case series reporting the outcome of children treated for sternal cleft. The most comprehensive review is by Torre et al. who reported an institutional series of seven patients and a literature review involving 70 patients who had undergone surgical repair.

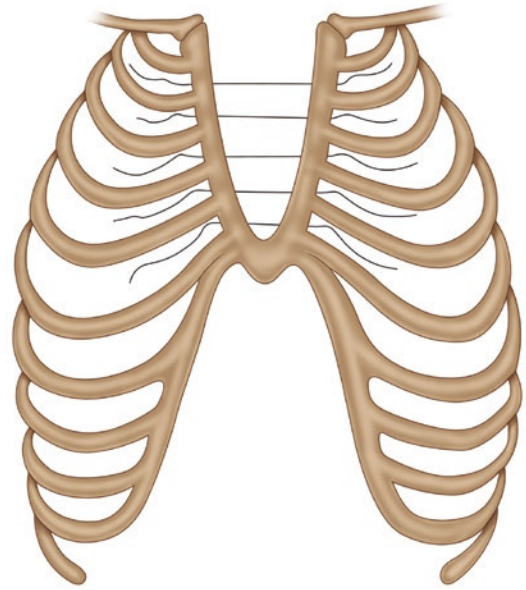

Fig. 60.6 Multiple heavy absorbable monofilament sutures are passed around the sternal bars

Primary repair was the most common procedure performed in 31/77 (40 %) cases. The periosteal flap was used in 14/77 (18 %), sliding chondrotomies in 10/77 (13 %) and cartilage resection in

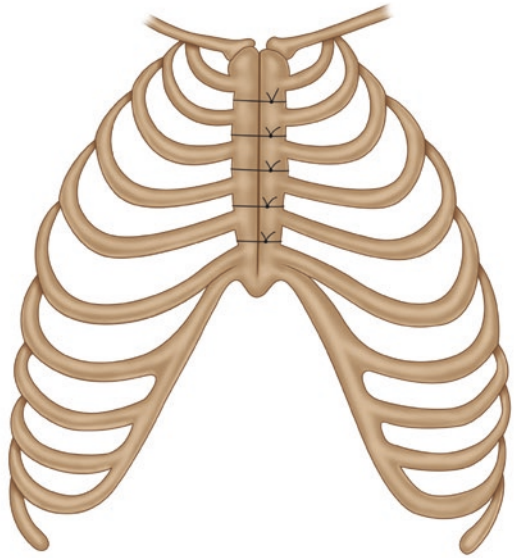

Fig. 60.7 The sutures are tied sequentially from inferior to superior while monitoring the patients' cardiovascular and respiratory status

4/77 (5 %). Placement of a prosthesis was the most common alternative procedure used in 8/77 (10 %). Bone graft placement was used in 7/77 (9 %) cases, muscle flaps in 2/77 (2.6 %) and two stage primary closure in 1/77 (1.3 %) [1].

Intraoperative complications were reported in 3/77 (4 %) patients. These were during sternal bar dissection causing pericardial or pleural tears. Post-operative complications were seen in 13/77 (17 %). These were mostly retrosternal seromas and pneumothorax. Two post-operative deaths were reported related to underlying cardiac anomalies. No recurrences have been reported in the literature [1].

A good cosmetic outcome and sternal resistance to palpation was reported in all 7 patients in the institutional series. This was unaffected by the type of prosthesis used. One out of 7 patients subsequently developed pectus excavatum, which was successfully repaired by an open technique [1].

A large series reporting mostly isolated superior sternal cleft repairs produced a good cosmetic outcome for all of their 15 patients operated over a 27 year period. Complications included seroma formation in two patients [2]. In another series reporting 8 sternal cleft patients, only one child had a poor outcome. This child was initially repaired using rib homograft and a prosthetic mesh and had excessive mobility of the repair. This was successfully corrected by re-operation [3].

The surgical results are therefore encouraging and although primary repair within the first few months of life is desirable, a good cosmetic and functional outcome can be achieved in the majority of patients regardless of the method of closure. Ultimately survival is determined by associated anomalies rather than the sternal cleft.

Comments

Sternal cleft is a rare congenital chest wall malformation resulting from defective embryological development and fusion of the two mesodermally derived sternal halves during the sixth to tenth week gestation [3, 4]. The Hoxb gene has been implicated in the development of sternal cleft [5] although no direct causative factors, teratogens or a familial basis for the abnormality has been identified [4, 6, 7].

The reported incidence is 0.15 % of all chest wall malformations and there is a female predominance [1, 3]. The defect is classified as either complete or partial. Partial sternal clefts are further subdivided into superior and inferior defects [8].

The superior sternal cleft is the most common, affecting the manubrium and upper sternum for a variable distance. It is usually an isolated anomaly, and is characterised by either a broad u-shaped or a narrow v-shaped defect [8] (Fig. 60.8). An inferior sternal cleft results from failure of fusion of the distal sternum and although it may occur as an isolated abnormality it is usually associated with other developmental abnormalities of the anterior chest wall such as ectopia cordis or combinations of defects as occur in the Pentalogy of Cantrell; omphalocoele, intracardiac defects, anterior diaphragmatic hernia and ectopia cordis [6, 7] (Fig. 60.9). A complete sternal cleft is very rare and characterised by the presence of two unconnected parallel sternal bars with diastasis of the rectus abdominis [8] (Fig. 60.10). Isolated sternal clefts, and particularly superior defects, usually have a

good outcome as they have an orthotopic heart without significant intracardiac abnormalities. The pericardium is intact and skin coverage normal [3]. In the presence of thoracic or thoracoabdominal ectopia cordis the mortality is much higher. This is due to the complex associated cardiac defects and available thoracic cavity to accommodate thoracic organs enabling them to function effectively [4].

Preoperatively neonates and children presenting late with sternal clefts should be thoroughly investigated for the purposes of the surgical repair, and for identification of associated anomalies. Various specialists should be consulted for the feasibilities and availability of appropriate materials at the time of surgery. Associated abnormalities have been reported in up to 72 % of children with sternal clefts [1]. Some of these are evident on physical examination; exomphalos, pectus excavatum, cleft lip/palate, precordial skin tags, supraumbilical raphe, maxillo-facial haemangiomas and gastroschisis [1, 8, 9]. Other abnormalities are demonstrated by further investigation and include cardiac defects, aortic coarctation, noncutaneous haemangiomas, ocular abnormalities, posterior fossa abnormalities and vascular dysplasias, particularly of the vertebrobasilar system [1, 8]. Associated defects may be seen in combination in the neurocutaneous disorder PHACE syndrome (posterior fossa anomalies, haemangiomas, arterial anomalies, cardiac abnormalities/aortic coarctation and eye abnormalities) and the Pentalogy of Cantrell [1, 10].

Surgical treatment and timing of surgery depends on the age of the patient, the type of defect and the presence of associated abnormalities. Primary closure is feasible and best performed in the neonatal period or early infancy when the thoracic wall is most compliant [3, 4, 11]. Late presenters require various innovative cleft repair techniques to protect and restore the thoracic viscera. With regards to the technique, if a superior sternal cleft is present it may be necessary to either excise a wedge from the inferior sternal bridge [3, 12, 13] or alternatively completely excise the inferior sternal bridge [11, 14]. Furthermore, if the sternal bars are too widely separated or the thoracic cage is too stiff, primary closure may result in respiratory and cardiovascular compromise secondary to reduced intrathoracic volume and impairment of venous return. This scenario is more common in patients beyond the neonatal period [1]. Alternative techniques to effect closure are available in such circumstances and include:

- Partial or total thymectomy [1]
- Sliding/rotating chondrotomies [15]
- Clavicle dislocation [3, 7]
- Periosteal flaps [2, 6]
- Pectoralis muscle flap advancement [16]
- Autogenous tissue/cancellous bone graft [2, 4, 6]
- Insertion of synthetic or biological prosthetic grafts [1, 3]

Bilateral sliding chondrotomies involve oblique transections of the ribs allowing approximation of the sternal bars in the midline [15]. Rotating chondrotomies can be employed to achieve the same purpose. In certain instances concomitant clavicular dislocation may be required [3, 7].

Insertion of autogenous bone graft or a synthetic prosthesis avoids significant chest wall dissection but carries an increased risk of infection. Autologous tissue grafts including; iliac crest [4] (Fig. 60.4), costal cartilage [2, 6], rib [3], parietal skull and tibial periosteum [4, 6] have been used successfully.

Autologous tissue grafts are generally preferred to synthetic grafts, which do not grow with the child and are associated with tissue reactions and an increased risk of infection [4]. Synthetic grafts have however been used successfully, either in isolation or as a composite graft, with good functional and cosmetic outcome. Materials used include non-absorbable grafts such as; stainless steel mesh, Teflon, polypropylene, acrylic, silicone elastomer, polyester, and calcium phosphate cement [1, 4, 17]. Absorbable prostheses such as Lactosorb® (Biomet microfixation, Jacksonville, FL, USA) have also been used in combination with non-absorbable prostheses [1].

Procedure Related Images
(Figs. 60.8, 60.9, 60.10, and 60.11)

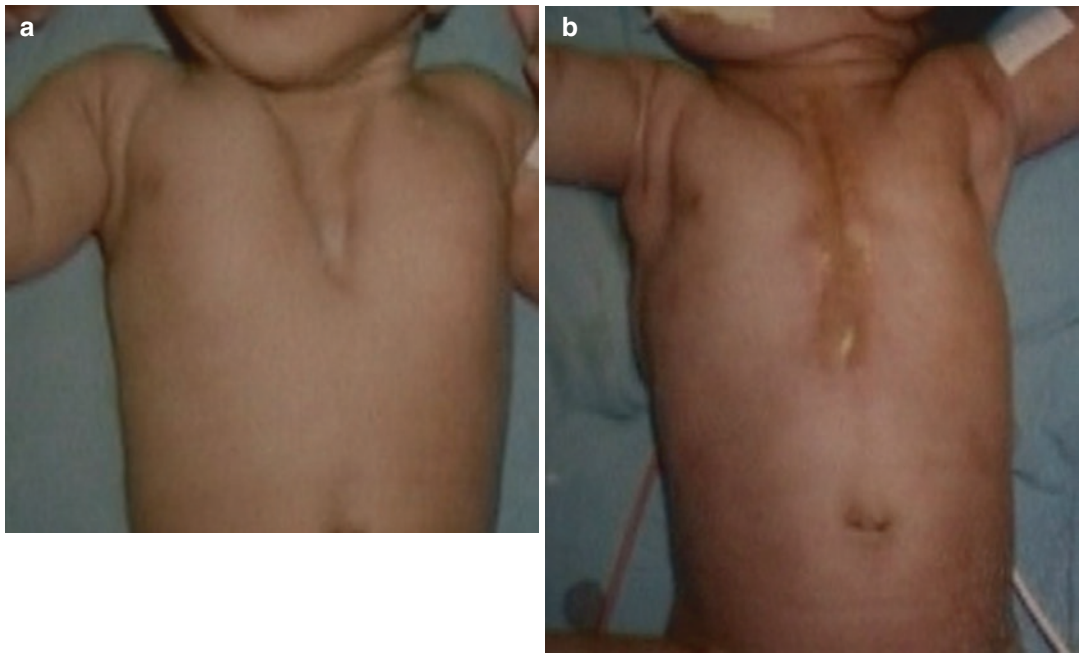

Fig. 60.8 (**a**) Superior sternal cleft in an infant. (**b**) Appearance following primary repair in the same infant

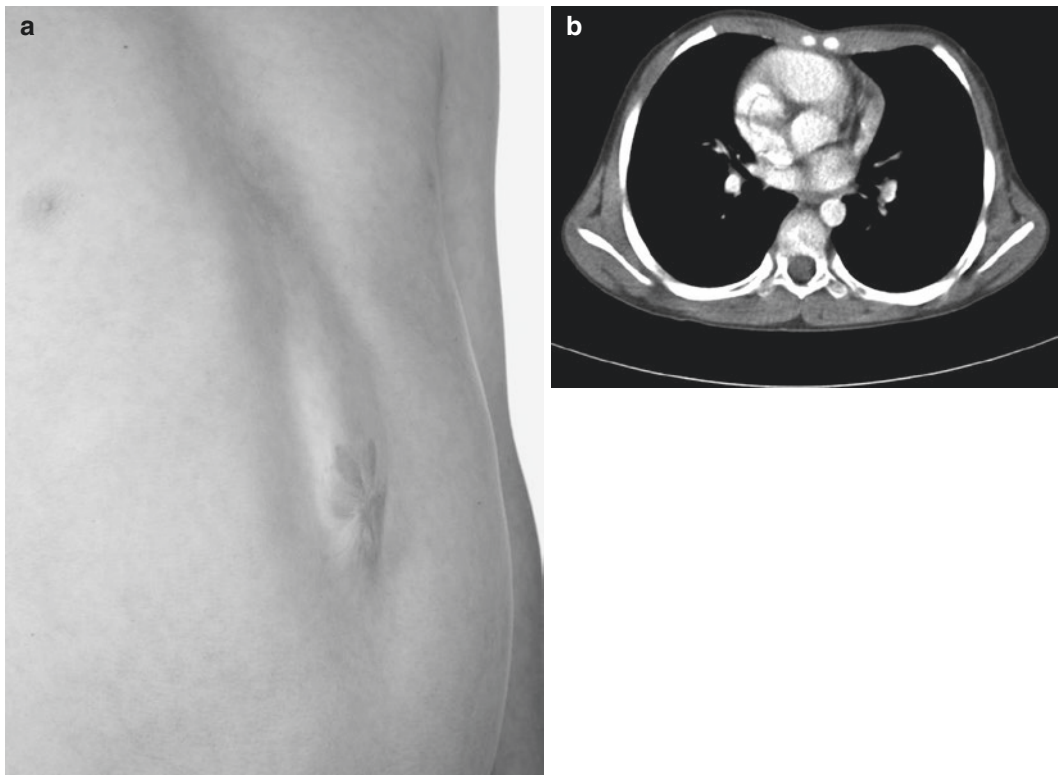

Fig. 60.9 (**a**) Inferior sternal cleft in a child with a previously repaired exomphalos. (**b**) Axial CT scan in the same child demonstrating separation of the inferior sternal bars. (**c**) Sagittal CT scan in the same child demonstrating the inferior sternal bony defect. (**d**) Inferior sternal cleft associated with exomphalos major in a neonate. (**e**) Appearance following primary repair of inferior sternal cleft and exomphalos in the same neonate

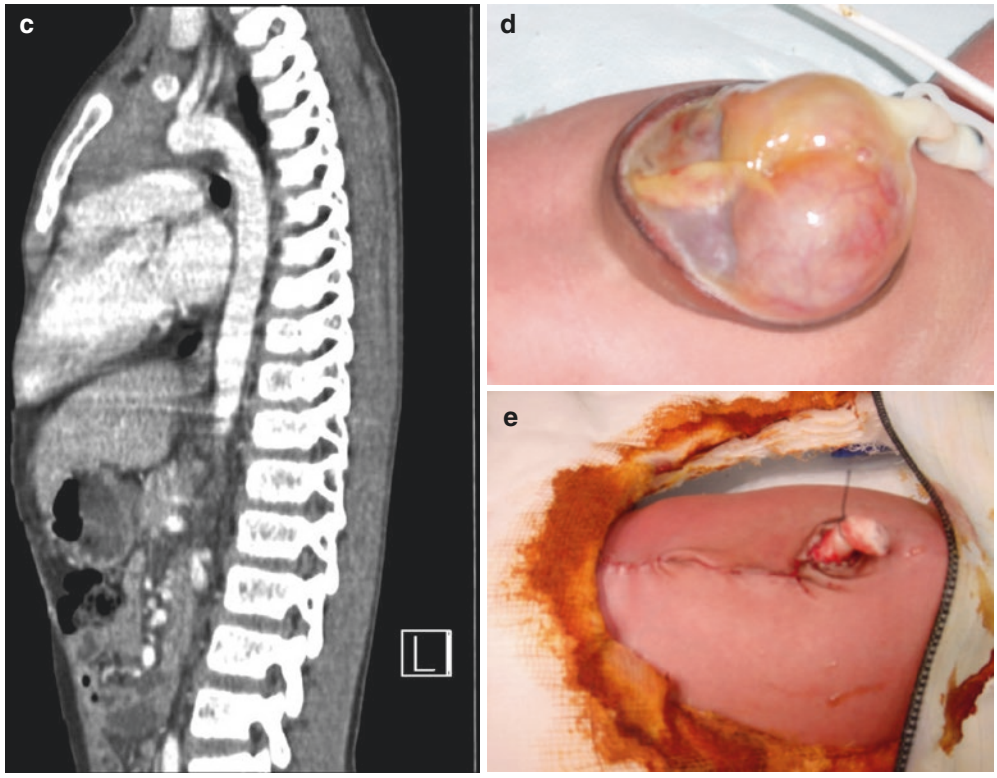

Fig. 60.9 (continued)

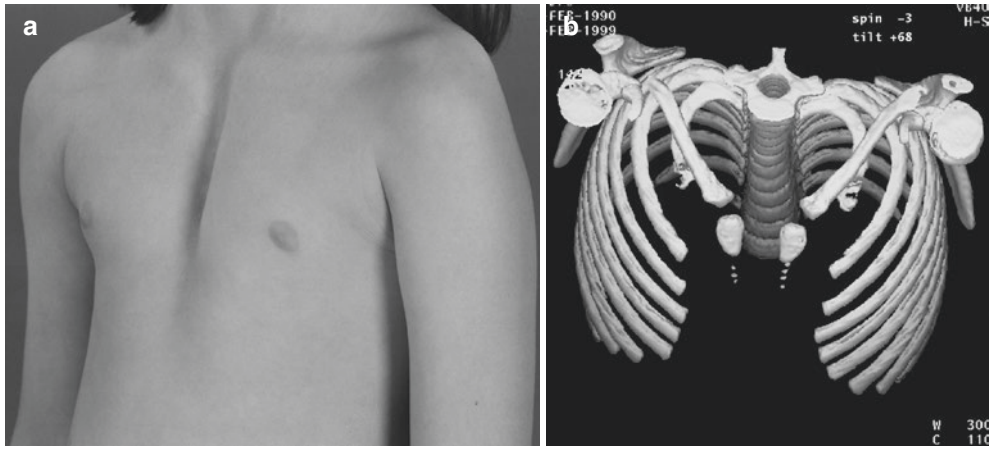

Fig. 60.10 (**a**) Complete sternal cleft in a child. (**b**) Pre-operative three dimensional CT scan demonstrating the bony defect in the same child

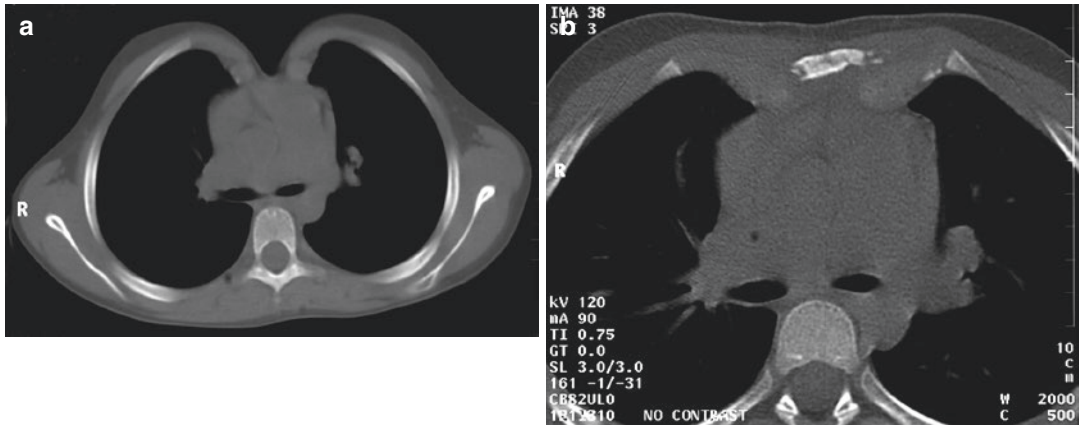

Fig. 60.11 (**a**) Preoperative axial CT scan in a child with a complete sternal cleft. The bony defect and underlying mediastinal structures are demonstrated. (**b**) Postoperative axial CT scan at 6 weeks in the same child demonstrating incorporation of a cancellous free graft from the iliac crest

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Poly-Lactic Acid Plate for Chest Wall Repair in Pentalogy of Cantrell

61

Hui-Ling Chia and Vincent Kok-Leng Yeow

Technical Highlights

This technique is suitable for the reconstruction of inferior sternal defects, a feature of pentalogy of Cantrell [1]. This rare syndrome also causes defects involving the heart, pericardium, diaphragm and anterior abdominal wall.

The main technical highlights of this procedure are:

1. Sagittal incision
2. Dissection of pectoral muscles to expose sternum
3. Median sternotomy and correction of congenital intra-cardiac defects by cardiothoracic surgeons, if indicated
4. Repair of pericardial defect using expanded polytetrafluoroethylene (ePTFE) membrane
5. Sternotomy closure with wire fixation
6. Suture approximation of divaricated rectus muscle
7. Cut and mold poly-L-lactic-polyglycolic (PLLA-PGA) plate to fit defect
8. Inlaying of plate and suture fixation of plate to sternal bar and rectus muscle
9. Elevation of bilateral skin and pectoralis major flaps
10. Advancement and suture fixation of bilateral pectoralis major flaps
11. Skin closure

Indications

- Chest wall defects in pentalogy of Cantrell
- Inferior sternal defects causing ectopia cordis

Preoperative Considerations

1. Echocardiogram to evaluate cardiac defects
2. Collaboration with cardiothoracic and pediatric surgeons as cardiac, pericardial, diaphragmatic and abdominal wall defects may be repaired in the same setting
3. Chest and abdominal computed tomography (CT) scan to assess extent of deformity
4. Photographic documentation of the deformity
5. Referral to geneticist if other associated anomalies are present.

Special Instruments (Implant)

Poly-L-lactic-polyglycolic (PLLA-PGA) plate (Lactosorb®, Walter Lorenz Surgical, Jacksonville, FL)

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Surgical Technique

The reconstruction may be performed during the same surgical setting as the repair of intracardiac, pericardial, diaphragmatic and abdominal wall defects. In these cases, a median sternotomy is performed. After corrective cardiac surgery, the pericardial defect is repaired using Gore-tex® (W.L. Gore & Associates, Inc.; Flagstaff, Arizona, USA), an expanded polytetrafluoroethylene (ePTFE) surgical membrane. This is positioned without folds and secured using interrupted Prolene sutures.

In inferior sternal clefts, the upper rectus muscles are widely separated and are attached laterally onto the lower ribs. This V-shaped defect is reduced by approximating the superior rectus muscles using polyglactin sutures. The superior sternotomy is closed using interrupted stainless steel wires (Fig. 61.3).

Using a paper template, the sternal defect is outlined before transferring onto a PLLA-PGA plate, allowing it to be cut precisely to shape. The plate is made pliable, by placing it into a warm sterile water bath, and shaped into the desired contour. The molded plate is inlaid into the sternal defect and fixed superiorly to the sternal bar and inferiorly to the rectus muscle using polypropylene sutures (Fig. 61.4). One drain is placed.

Using needle-tipped cautery, bilateral pectoralis major flaps are dissected. The muscle flaps are then advanced and sutured together to cover the plate before skin closure.

Procedure Overview
(Figs. 61.1 and 61.2)

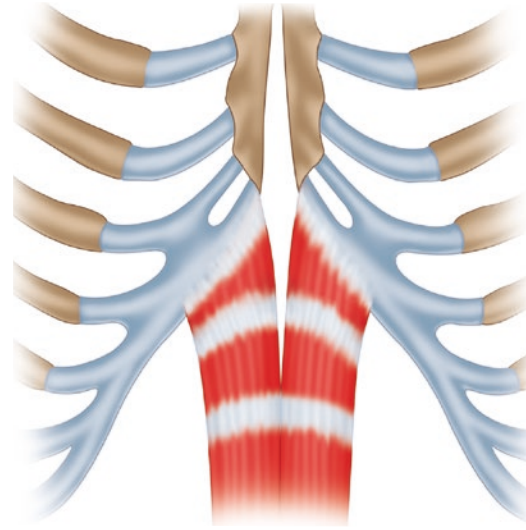

Fig. 61.1 Diagram showing inferior sternal defect. Median sternotomy of the superior sternum was performed. Apart from a large lower sternal defect, the upper rectus muscles were also widely separated, leaving a deep V-shaped defect

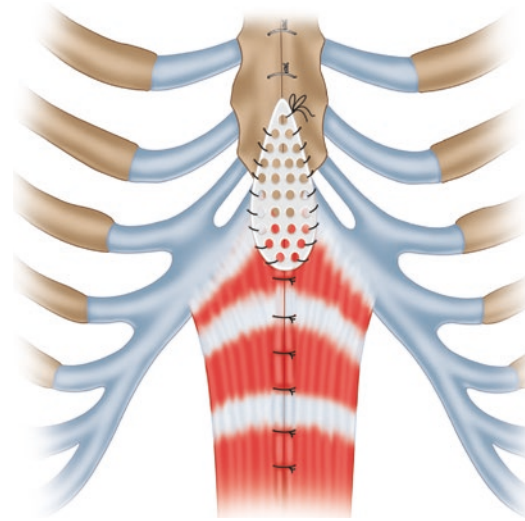

Fig. 61.2 The upper rectus muscles are approximated. The PLLA-PGA plate is placed in the lower sternal defect and sutured to the superior sternum and superior rectus muscles. Pectoralis muscles are approximated in order to cover the plate before closure

Postoperative Management

Perioperative intravenous antibiotic (Cefazolin) is administered. Patients are admitted to the cardiothoracic intensive care unit for 24 h postoperatively. After about 3 days, the drain is removed when the drainage is less than 20 ml over 24 h. Patients are discharged after about 1 week and followed up regularly in the outpatient clinic.

Results

In infants, the paucity of reconstructive options due to hypoplastic or anomalous ribs and pectoral muscles may preclude a robust and

reliable autologous reconstruction. We propose a staged procedure to repair the sternal defect using a PLLA-PGA plate in the first stage. This acts as a temporary shield and can be performed concurrently with the repair of other defects.

This technique was performed in a 10-month old infant with Pentalogy of Cantrell (Image 3). Although the time to complete resorption of the polymer was reported to be between 9 and 15 months [2], the reconstructed anterior chest wall remains firm to palpation at 3 years (Image 4). This could be explained by the combination of ossification from the freshened edges of the sternum, fibrosis secondary to retention of the plate in this period of time and reinforcement by the bilateral pectoralis major advancement flaps [3]. Autologous reconstruction is planned in the second stage when the child is older and when more substantial autologous tissue is available for a more reliable reconstruction (chapter on bilateral sternal turnover flaps, Chap. 63).

Comments

This is a simple technique for sternal reconstruction in patients who are less than 2 years of age [3]. For patients with pentalogy of Cantrell or inferior sternal defects causing ectopia cordis, corrective cardiac surgery and sternal reconstruction is usually performed after 2 years of age if there are minimal intracardiac defects (when the child gains more cardiovascular reserve and to allow growth of the thorax).

Autologous reconstruction is recommended due to a lower risk of infection and its ability to remodel according to patient growth, as opposed to alloplastic materials. However, autogenous tissue may not be adequate in patients who require surgery (eg. early corrective cardiac surgery) before 2 years of age. The PLLA-PGA plate is resorbable and can be placed during this setting. It acts as a temporary shield, obviating the use of permanent alloplastic materials. When the child is older, definitive sternal reconstruction can be performed when more reliable autologous reconstruction options are available. For definitive reconstruction of inferior sternal clefts, the use of bilateral sternal bar turnover flaps is preferred (chapter on bilateral sternal turnover flaps, Chap. 63).

PLLA is resistant to degradation and PGA produces a rapidly resorbing implant. Combining these properties, the copolymer creates better strength and resorption profiles than exist in the homopolymers alone. Large studies have shown that these implants have similar resistance to deformation and fracture as one's native skull [4]. Its moldable property will facilitate plate conformation to the contour of the anterior chest wall. The macromolecules used in this copolymer are in a non-inflammatory pathway for dissolution and are less prone to inflammatory complications than metallic systems.

Procedure Related Images (Figs. 61.3, 61.4, 61.5, and 61.6)

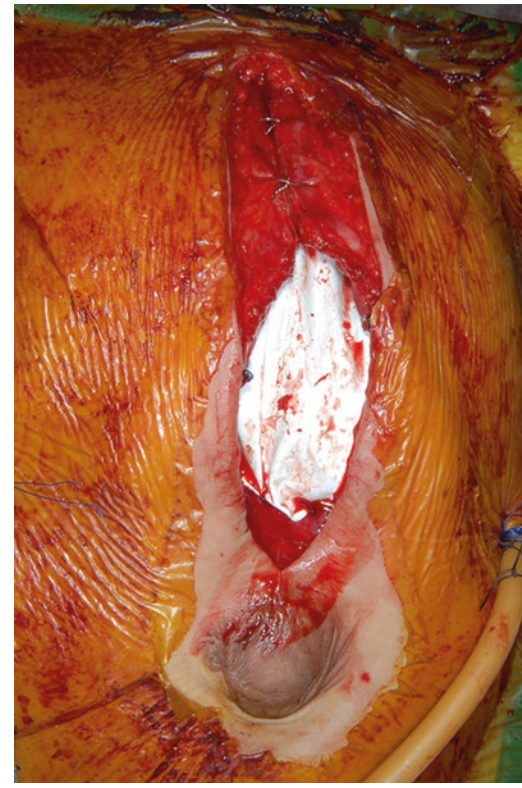

Fig. 61.3 The pericardium is closed with an expanded polytetrafluoroethylene (ePTFE) membrane, followed by sternotomy closure with wire fixation

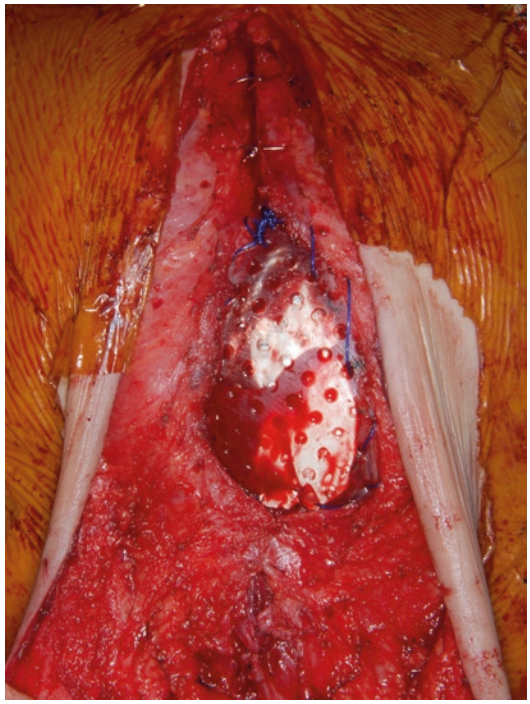

Fig. 61.4 The upper rectus muscles are approximated. The PLLA-PGA plate is placed in the lower sternal defect and sutured to the superior sternum and superior rectus muscles

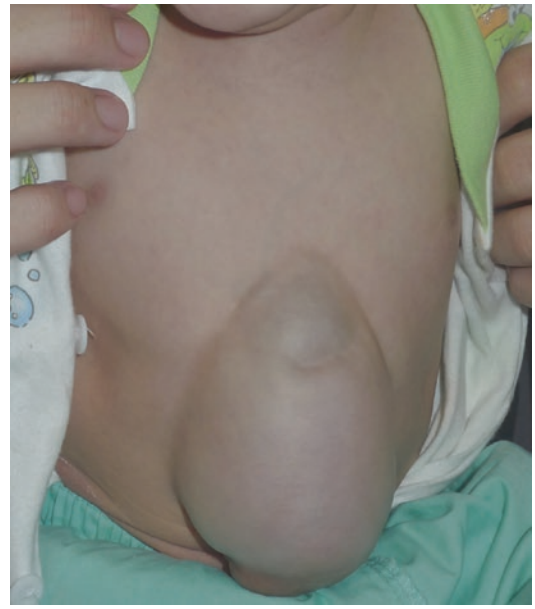

Fig. 61.5 Preoperative photo of a 10-month-old patient with pentalogy of Cantrell, presenting with abdomino-thoracic ectopia cordis. There is herniation of the heart through a lower sternal defect, which was covered by very thin skin alone, allowing the cardiac impulse to be seen. An epigastric hernia was also present

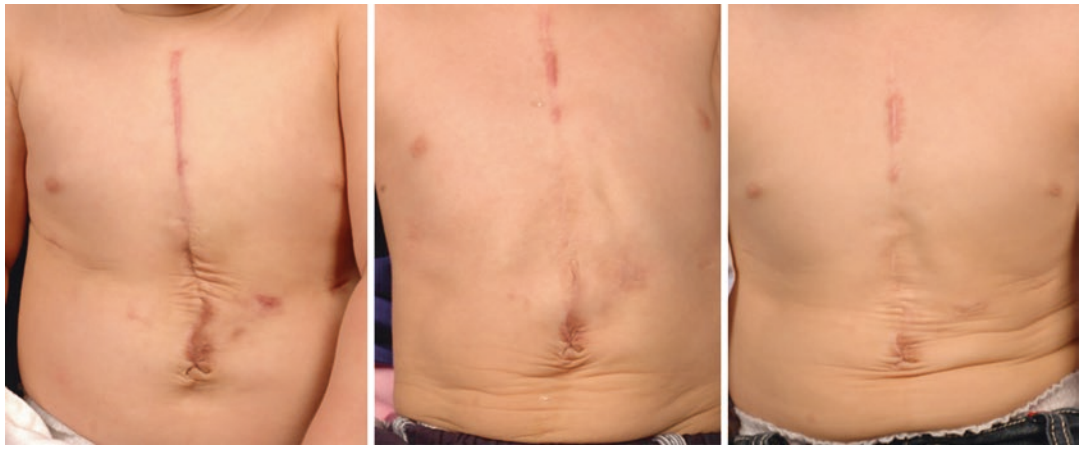

Fig. 61.6 Postoperative photos of the patient at 3 months (*left*), 9 months (*middle*) and 3 years (*right*)

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2. Eppley BL, Morales L, Wood R, et al. Resorbable PLLA-PGA plate and screw fixation in pediatric craniofacial surgery: clinical experience in 1883 patients. Plast Reconstr Surg. 2004;114:850–6; discussion 857.
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Poland's Syndrome Treatment with Omentum Flap

62

Sirlei dos Santos Costa and Rosa Maria Blotta

Introduction

Physical characteristics that affect patients with Poland's syndrome are (a) the presence of a transverse skin fold in the anterior axillary pillar caused by the absence or hypoplasia of the pectoral muscles, (b) the infraclavicular depression and (c) the anomalous breast contour.

The main technical highlights of the laparoscopic harvested omental flap procedure are:

(a) Laparoscopy using 4 ports and elevation of the gastric wall.
(b) Localization of the right gastro-epiploic artery.
(c) Ligations of the short gastric arteries.
(d) Liberation of the transverse colon segment.
(e) Ligation of the left gastro-epiploic artery.
(f) Incision in the peritoneum.
(g) Digital maneuvering to pull it from the abdominal cavity to the breast region.
(h) Securing the omentum in the defect.

Indications

- Chest wall augmentation in patients with Poland's syndrome

Preoperative Considerations

- Almost all patients are suitable candidates for laparoscopic omentum transposition but previous states such as prior surgery, radiotherapy or infection could compromise the viability of the omentum.

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- Chest films with postero-anterior and lateral view
- Electrocardiography and echocardiography examinations
- Routine laboratory blood tests and coagulation tests

Special Instruments

General instruments and equipment to perform laparoscopic procedures

Surgical Technique

Harvesting of the omentum flap is performed using standard laparoscopic surgical techniques. Four ports are usually placed and CO_2 pneumoperitoneum of 8–10 mmHg is maintained during the procedure. Dissection of the flap is commenced by grasping and elevation of the gastric wall. The right gastro-epiploic artery (RGEA) is isolated and preserved; ligations of the short gastric arteries along the greater curvature are then performed until the left gastro-epiploic artery (LGEA) is reached. The omentum is disconnected from the transverse colon by a careful dissection in order to preserve the mesocolon vascularity. The flap is liberated when ligations of the LGEA are completed adjacent to the left colic flexure.

Through a small incision in the inframammary fold a subcutaneous tunnel is created until the costal border to open the aponeurosis in the medial line, in the direction to the abdominal cavity. With a digital maneuver the omentum is extracted from the abdominal cavity to the breast region for placement of the flap over the thoracic wall. This tunnel is placed to the left or right side of the round ligament depending on the side that needs reconstruction. The area of the deformity is then dissected and filled in with the omentum flap, which is fixed into place. Both procedures are performed under endoscopic visualization.

Procedure Overview (Figs. 62.1, 62.2, 62.3, and 62.4)

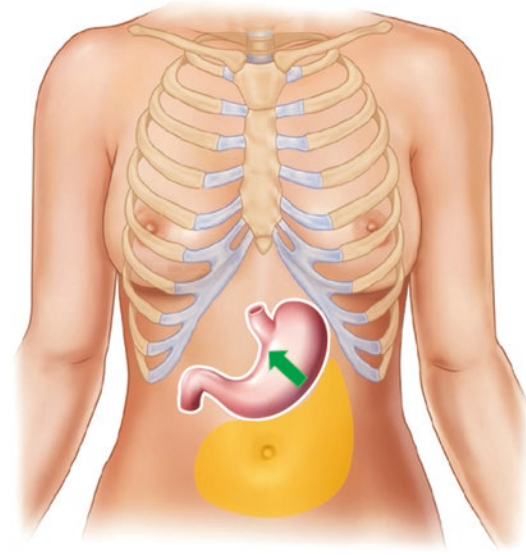

Fig. 62.1 Laparoscopic dissection of the omentum flap using a 4 port technique is commenced by grasping and elevating the stomach wall to expose the greater curvature

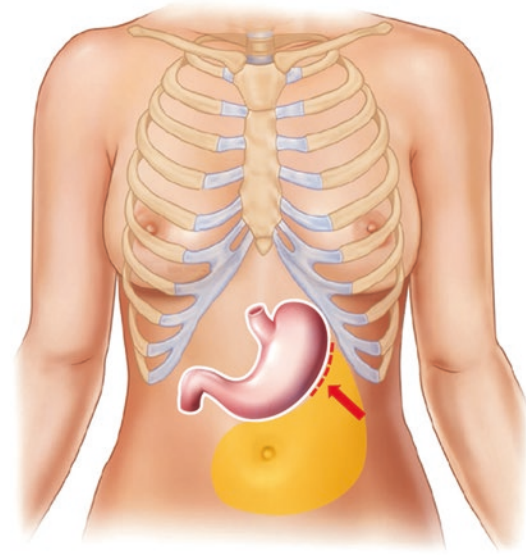

Fig. 62.2 Ligations of the short gastric arteries along then performed along greater gastric curvature

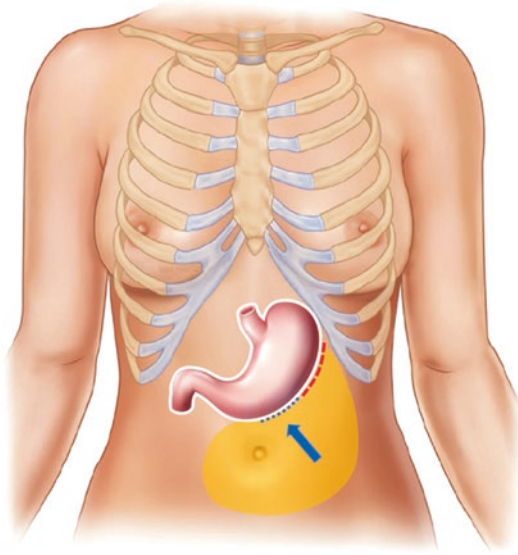

Fig. 62.3 The omentum is separated from the transverse colon by a careful dissection

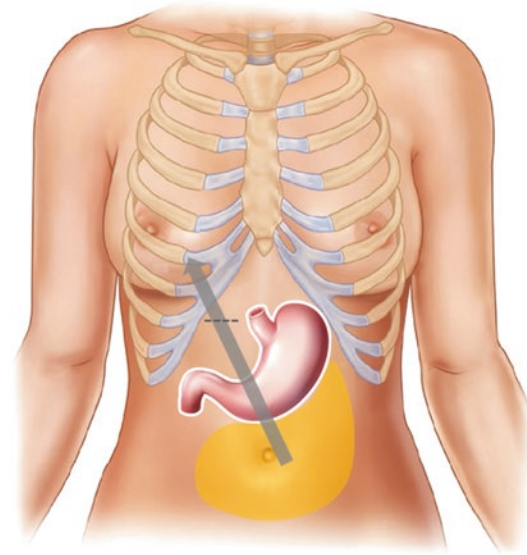

Fig. 62.4 Through a small incision in the infra-mammary fold a subcutaneous tunnel is created to access abdominal cavity. With a digital maneuver the omentum is extracted from the abdominal cavity to the breast region for placement of the flap over the thoracic wall

Postoperative Management

The mean hospital stay is short and ranges between 1 and 5 days. Patients resume normal activities depending on individual treatment progress. Some patients have post-prandial discomfort and moderate gastric distension with spontaneous regression for 48 h after the procedure.

Results

When laparoscopically harvested omentum flap is used to treat breast deformities, a significant volume increase of the omentum is noticed in the first months following its transposition in all the patients. At the time of the transposition of the omentum flap to the breast region, the adipose tissue is subjected to a transitory hypoxia, which might have stimulated VEGF and, consequently, neoangiogenesis, leading to the growth of the volume of the flap.

The increase in the volume of the flap is caused, probably, by the hypertrophy of the adipocyte resulting from neoangiogenesis, caused by the stimulus of VEGF induced by temporary hypoxia caused by dissection and mobilization of the flap from the abdominal cavity to the breast region.

Patients with Poland's syndrome treated with omentum flap achieved a final aesthetic result better than those patients who were treated with other techniques. With the omentum flap, it is possible to correct specific details of breast contour due to flap tissue malleability, an outcome impossible to achieve with all other techniques. The palpation of the flap in the new site is similar in consistency to a normal breast. All other flaps do not achieve similar consistency of the breast like omentum flap.

Comments

Patients with Poland's syndrome may present with numerous ailments such as absence of the sternal-costal portion of the *pectoralis* major muscle, upper extremity hypoplasia, brachysyndactyly, and syndactyly. Various other muscles may also be affected: *pectoralis* minor, latissimus dorsi, serratus anterior, external oblique, and deltoid. Skeletal deformities such as partial agenesis of the ribs, *sternum*, and spine (sometimes with *scoliosis*) may occur. Breast hypoplasia or aplasia, nipple abnormalities, skin atrophy, and absence of the sweat glands and surrounding structures are other features.

In Poland's syndrome, thoracic wall deformities are not as obvious at birth as hand deformities. However, when female patients reach adolescence, the thoracic deformity seems to become more evident as absence or asymmetry of the developing breasts occurs. To minimize this, a tissue expander may be placed in the developing breast to accompany contra-lateral breast growth. Unfortunately, however, surgical treatment of the breast deformities cannot be accomplished before 17–19 years of age, when development of the body is complete.

Numerous alternatives of treatment have been proposed to correct the most important deformities caused by this syndrome such as handmade expanders and prosthesis, latissimus dorsi flap and rectus abdominals muscle flap, but all have offered only a partial correction of the deformities, with poor aesthetic results.

The utilization of the omentum flap, in our experience, offers the possibility to treat these deformities with excellent cosmetic results. The omentum has been utilized in reconstructive surgery for more than a 100 years. In 1888, Senn employed it to protect an intestinal anastomosis; and in 1963, Kirikuta described the use of the great omentum as a flap in cases of breast cancer surgery.

In 1972, McLean and Buncke described the omentum free flap and in 1976, Arnold and Jurkiewicz used the pedicled omentum in the reconstruction of the chest wall, including two patients with Halsted mastectomy by a one-stage reconstruction using transposed omentum to cover a silastic gel prosthesis and to support an overlying skin graft. Laparoscopic harvesting of the omentum was carried out for the first time by Saltz in 1993 in order to repair soft tissue defects in the knee. In 1998, our group utilized the omentum flap to treat Poland's syndrome deformities.

The employment of the omentum flap provides reparation of the thoracic wall and breast deformities caused by Poland's syndrome, with superior outcomes when compared to other reconstructive options. The advantages of the omentum flap are numerous and significant: it is extremely malleable, adapts easily to irregular surfaces, and has a long and reliable vascular pedicle. The flap measures approximately 25×35 cm and its volume varies according to patient size. In the first six months after the procedure the omentum flap presents a variable

growth that needs to be considered when planning repair of the deformity.

Other important advantage include the flap's large absorption capacity, which reduces the postoperative time period during which drains are needed, since the flap helps absorb the lymphatic fluid resulting from lymph node dissection. The use of laparoscopy to harvest the flap offers minimal insult to the abdominal wall, ensuring a short and comfortable postoperative recovery period. Finally, the resulting consistency is very similar to the contra-lateral breast, enabling more satisfactory reparation of the anterior axillary pillar than any other reconstructive option.

A difficulty of this technique is that it is not possible to precisely define the final omentum volume available in order to plan the reconstruction. The complementation of the volume and equalization with the contra-lateral breast may be done during the same surgical time or, even better, at another time, 4–6 months later; when it is possible to take into consideration the spontaneous growth of the flap that always occurs after its transposition. The skeletal deformities that come with this syndrome may be minimized but, when very severe, may still distort the aesthetic appearance of the thoracic wall and breasts, in some cases obliging for another complementary procedure.

Procedure Related Images

(Figs. 62.5, 62.6, 62.7, 62.8, 62.9, 62.10, and 62.11)

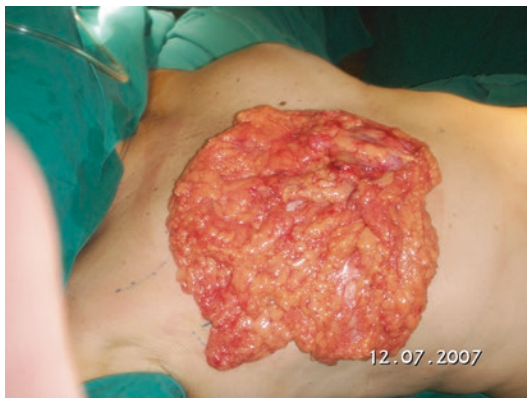

Fig. 62.5 View of the omentum flap after laparoscopic preparation and exteriorization over the incision

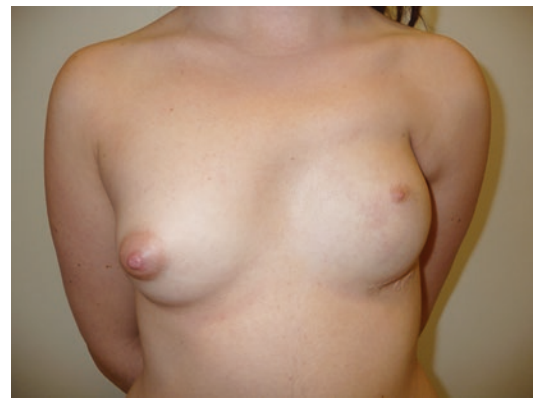

Fig. 62.6 Patient with Poland's syndrome demonstrating the extent of the deformity on the LEFT side

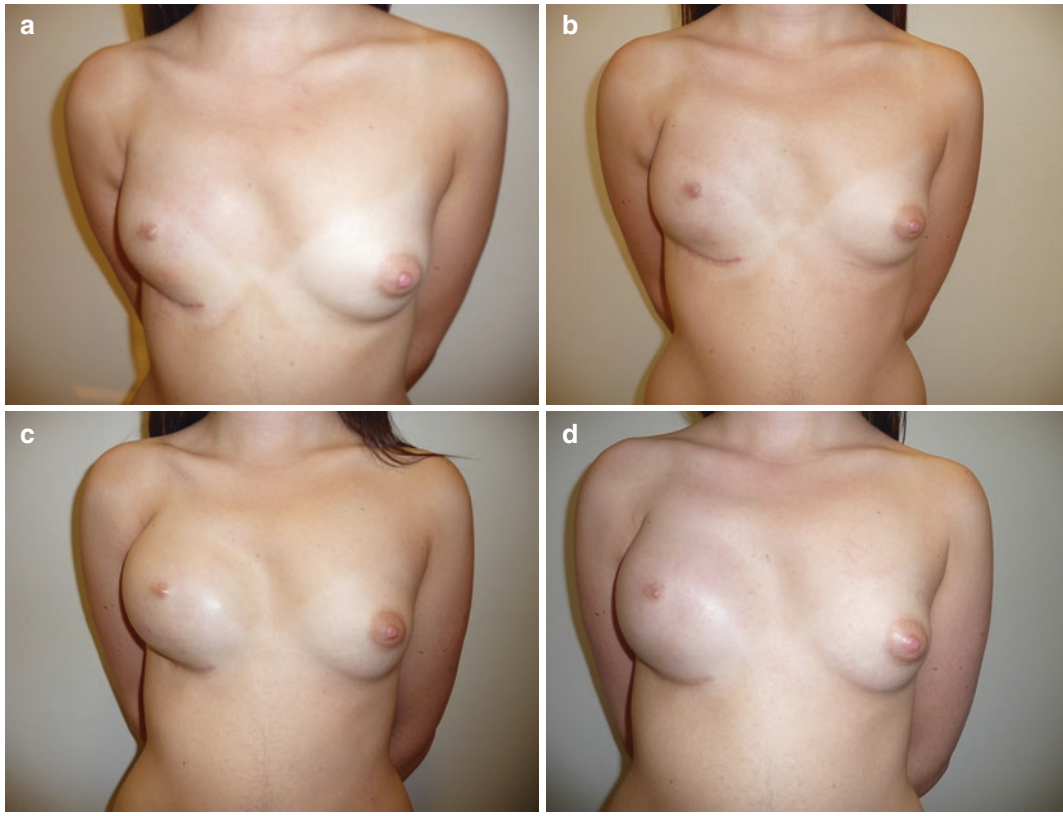

Fig. 62.7 Volume increase of the omentum flap after transposition to the breast region. (**a, b**) preoperative and (**c**, **d**) postoperative images

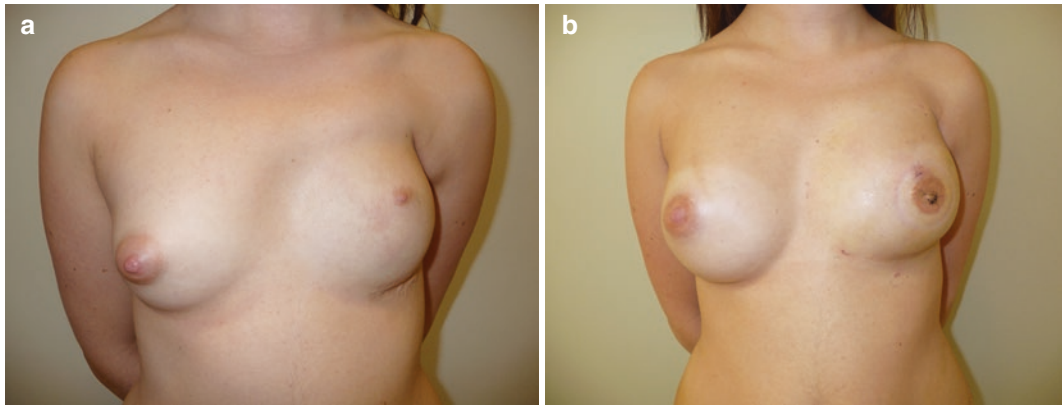

Fig. 62.8 Pre (**a**) and post operative (**b**) view of omentum flap transposition to the left breast and rigth breast augmentation with implants

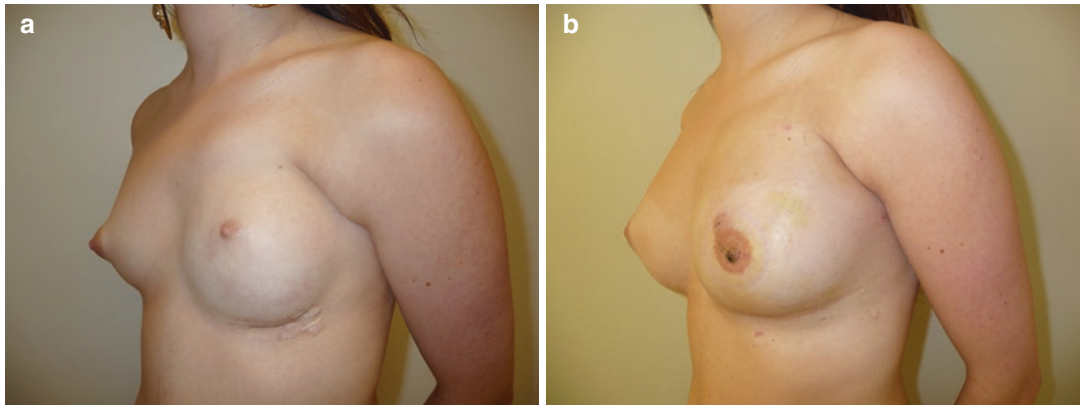

Fig. 62.9 Oblique view after reconstruction with omentum flap transposition

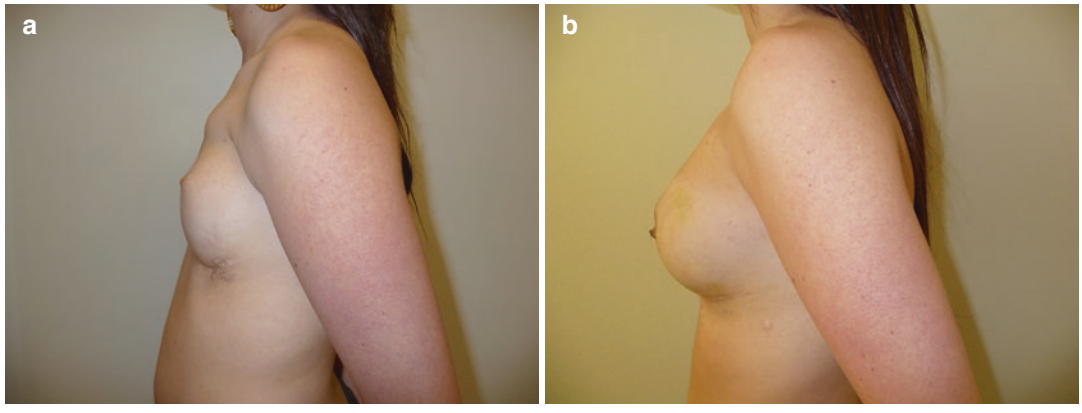

Fig. 62.10 Lateral view after reconstruction with omentum flap transposition

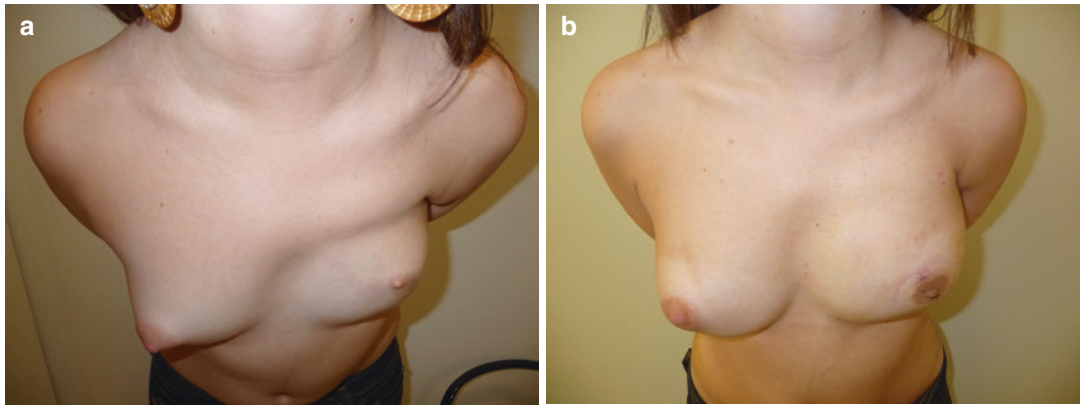

Fig. 62.11 Results after reconstruction with omentum flap transposition, patient view of her breasts

Suggested Reading

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Bilateral Sternal Bar Turnover Flaps for Reconstruction of Inferior Sternal Cleft

63

Hui-Ling Chia and Vincent Kok-Leng Yeow

Technical Highlights

Bilateral sternal bar turnover flaps is a technique for reconstruction of inferior sternal defects.

The main technical highlights of this procedure are:

1. Sagittal incision in the area of the deformity
2. Elevation of skin and pectoralis major muscle flaps
3. Dissection of superior rectus attachments from the lower sternal bar and ribs
4. Suture apposition of the divaricated superior rectus muscles
5. Bisection of cartilaginous sternal bars
6. Medial turnover of anterior half of the bisected sternal bars
7. Suture apposition of sternal bar flaps in the midline
8. Re-attachment of superior rectus to the sternal bar flaps
9. Placement of bilateral bupivacaine epidural catheters *under* the sternal bars
10. Advancement and suture fixation of bilateral pectoralis major flaps in the midline

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Indications

- Children above 2 years of age with inferior sternal cleft deformities, as part of Cantrell's pentology.

Preoperative Considerations

1. Echocardiogram to evaluate for cardiac defects
2. Collaborate with cardiothoracic and pediatric surgeons in patients with pentalogy of Cantrell, as cardiac, pericardial, diaphragmatic and abdominal wall defects may be repaired in the same setting
3. Chest and abdominal computed tomography (CT) scan to assess extent of deformity
4. Photographic documentation of the deformity
5. Referral to geneticist if other associated anomalies are present.

Special Instruments

Instruments for general thoracic surgical procedures

Surgical Technique

The patient is placed in supine position and a lower vertical midline incision is made. Skin and pectoralis major muscle flaps are raised using needle

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diathermy, exposing the sternal bar and proximal costal cartilages. The superior rectus attachments are dissected from the lower border of the sternal bar and ribs. Using a scalpel, the cartilaginous sternal bars are bisected, along its length, from lateral to medial, leaving the medial perichondrium intact. This allows the anterior half of the bisected sternal bars to be turned over medially to cover the inferior sternal defect. The cartilaginous flaps are sutured together in the midline using polypropylene sutures, constituting a complete thoracic skeletal ring. After suture apposition of the divaricated superior rectus muscles with polyglactin sutures, this is re-attached to the reconstructed lower sternum using polypropylene sutures. For sustained postoperative pain control, 0.025 % bupivacaine is administered using elastomeric infusors via two epidural catheters. These are placed *under* the sternal bars, one on each side. The bupivacaine will infuse at 2 ml per hour over the next 48 h. We place the drainage catheter *over* the sternal bars to avoid draining out the infused bupivacaine solution. Bilateral pectoralis major flaps are advanced and sutured in the midline to cover the reconstructed sternum before skin closure.

Procedure Overview (Figs. 63.1, 63.2, and 63.3)

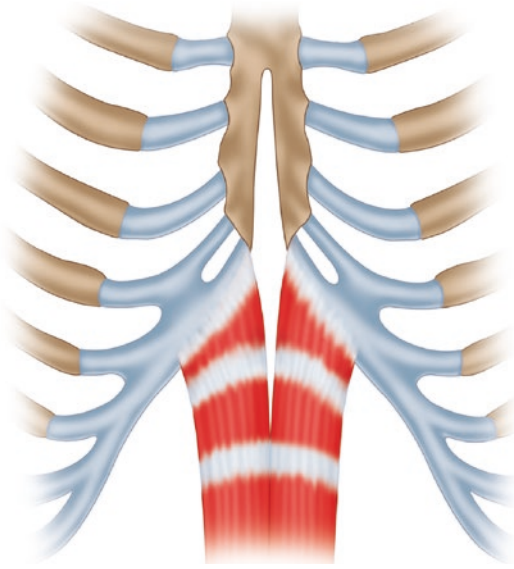

Fig. 63.1 View of the inferior sternal cleft and separated upper rectus abdominis, forming a diamond-shaped defect (Corresponds to Fig. 63.5)

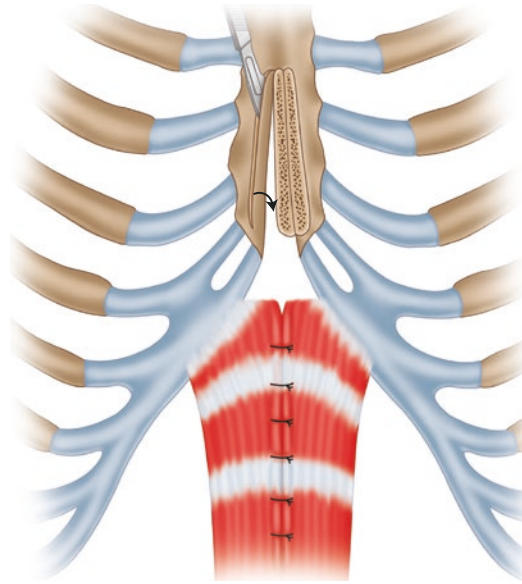

Fig. 63.2 The cartilaginous sternal bars are bisected, along its length, from lateral to medial, leaving the medial perichondrium intact. The anterior half of the bisected sternal bars can be turned over medially to cover the inferior sternal defect (Corresponds to Fig. 63.6)

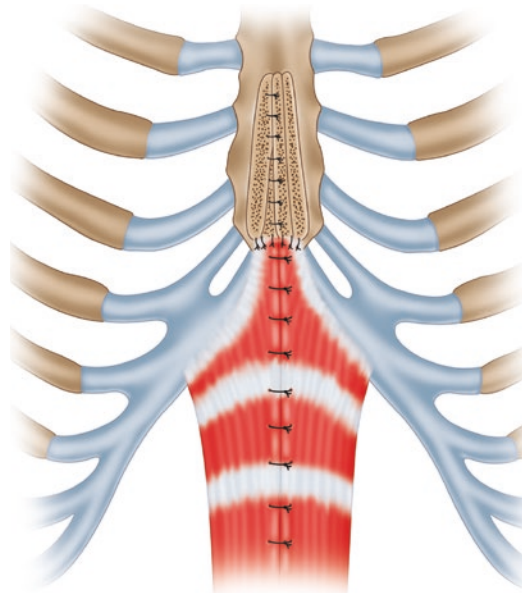

Fig. 63.3 The cartilaginous flaps are sutured together in the midline using polypropylene sutures. The divaricated superior rectus muscles are apposed with polyglactin sutures and re-attached to the inferior sternal bar flaps using polypropylene sutures (Corresponds to Fig. 63.7)

Postoperative Management

Patients are admitted to a high dependency ward for 24 h. The epidural infusion catheters are removed after 48 h and the drainage catheter after 3 days.

Results

Bilateral sternal bar flaps have the advantage of providing a rigid reconstruction without significant donor site morbidity. Both contour and strength of the anterior chest wall can be achieved using this technique. The continuous infusion of bupivacaine via epidural catheters provides effective sustained analgesia. Patients remained relatively pain-free postoperatively and are discharged by postoperative day 5. No complications or recurrences were encountered.

Comments

This technique is indicated for children above 2 years of age with inferior sternal cleft deformities, usually occurs as part of Cantrell's pentad [1].

Chest wall reconstruction and repositioning of the heart is preferably delayed until the child gains more cardiovascular reserve, and to allow for growth of the thoracic cavity. Frequently, this operative intervention is also associated with cardiopulmonary compromise. Children less than 2 years of age have small sternal bars and these may not provide a reliable reconstruction. When mild intracardiac defects are present, major reconstruction should be delayed until the child is 2 years of age [2].

In cases where surgical correction is indicated in children less than 2 years of age, the reconstruction is performed in two stages. In the first stage, a resorbable plate is insetted to act as a temporary shield [3]. Definitive rigid reconstruction of the defect with bilateral sternal bar turnover flaps is performed in the second stage, when the child is older and the thoracic skeleton is more developed.

Procedure Related Images
(Figs. 63.4, 63.5, 63.6, and 63.7)

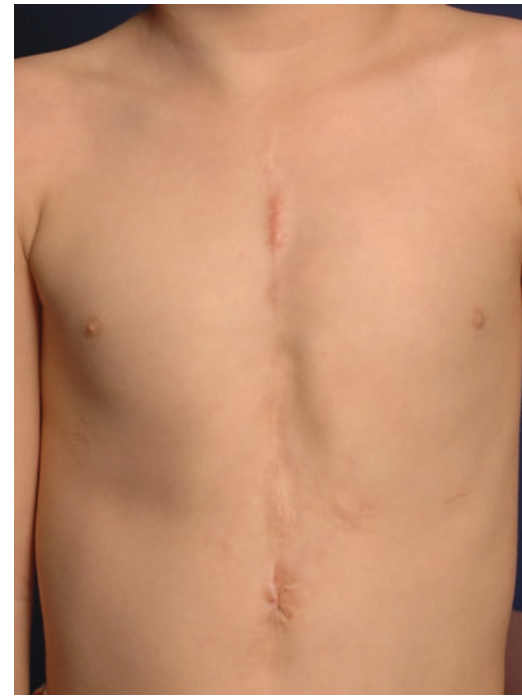

Fig. 63.4 Pre-operative photo. Image of a 4-year-old boy with inferior sternal cleft, as part of Cantrell's pentad who underwent corrective cardiac surgery and a resorbable plate was used to reconstruct lower sternal defect at 10 months of age. Definitive reconstruction with bilateral sternal bar turnover flaps was planned

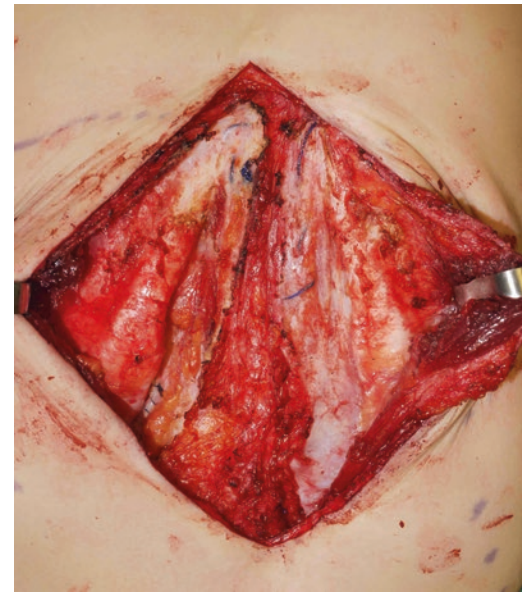

Fig. 63.5 Inferior sternal cleft. Intraoperative photo showing the inferior sternal cleft and separated upper rectus abdominis, forming a diamond-shaped defect (Corresponds to Fig. 63.1)

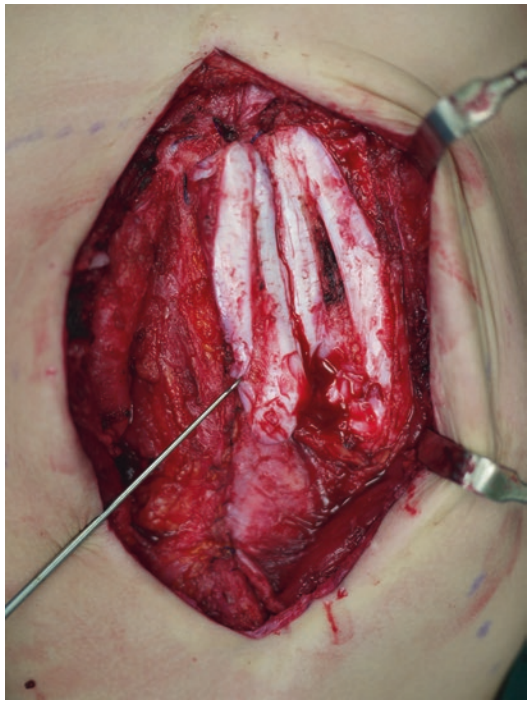

Fig. 63.6 Bisection of sternal bars. The cartilaginous sternal bars are bisected, along its length, from lateral to medial, leaving the medial perichondrium intact. This allows the anterior half of the bisected sternal bars to be turned over medially to cover the inferior sternal defect (Corresponds to Fig. 63.2)

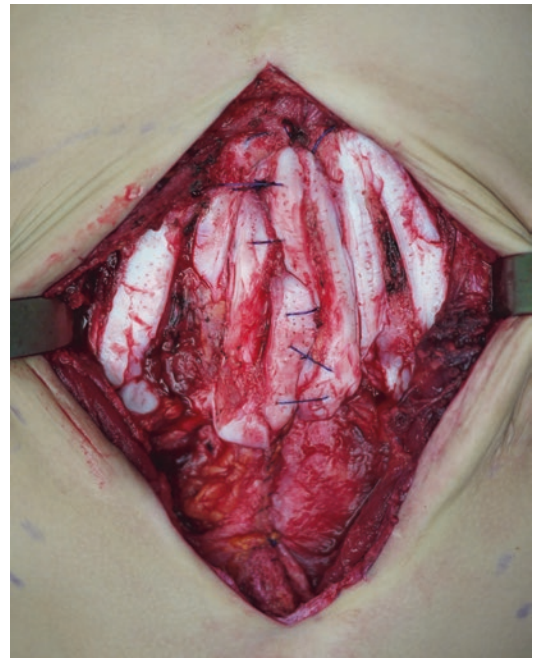

Fig. 63.7 Apposition of flaps. The cartilaginous flaps are sutured together in the midline using polypropylene sutures. The divaricated superior rectus muscles are apposed with polyglactin sutures and re-attached to the inferior sternal bar flaps using polypropylene sutures (Corresponds to Fig. 63.3)

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Part X

Comments on Pectus Repairs

Pros and Cons of the Nuss-Procedure

64

Amulya K. Saxena and Francis Robicsek

The Pros

Surgery is always invasive; it is only the route that is minimal access

Successful pectus repairs with conventional open surgical techniques were offered at some Centers throughout the world that were able to further develop these techniques and expertise due to high volume of patients being treated. Centers will low volumes of patients struggled to replicate these results, with the consequence that either these procedures were completely abandoned from their surgical armament or indications were not given to operate these patients. Hence, during the 1980's and 1990's Centers that were presenting data on larger series of pectus repairs were often criticized for their indications for surgery rather than the good results obtained after correction. With this scenario, there was lack of enthusiasm in low volume Centers and the majority of global Centers to develop their pectus services and justify reasons for surgical indications.

With the report on the Minimal Access Repair of Pectus Excavatum (MARPE) by Donald Nuss, a sudden global surge in the interest to manage patients with pectus deformities was observed. Indications for surgery were then being offered throughout the world, especially for those patients who have been suffering for psycho-social reasons due to their deformities. Hence, the Nuss procedure firstly highlighted the plight of these patients with pectus deformities as well as gave a global impetus towards the management using a minimal access technique. Secondly, the popularity of this technique gave the surgical experts who were involved with the treatment of pectus deformities, a common basis to compare and discuss their results. None of past techniques to date were able to generate such a response from the surgical community in the treatment of pectus deformities in former times. Thirdly, the expansion of the internet at the time the Nuss procedure was being introduced was instrumental in the technique gaining popularity and the dissemination of results obtained to a global audience of surgeons willing to treat their patients with a technique that avoided open surgical manipulations around the parasternal area. Fourthly, the introduction of endoscopic visualization that offered a safer possibility to complete the procedure and the

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impetus in pediatric endoscopic surgery, were two factors that further won enthusiastic surgeons to adopt this technique. Finally, the standardization of instruments through creation of pectus sets and their global sales by Walter Lorenz Surgical Inc. assisted in the dissemination of the technique which was further aided by globalization. The ability of surgeons to attend Courses and visit Centers after relaxation of travel restrictions over the past 2 decades added to the list of factors that added in worldwide acceptance.

Nuss procedure has been successful mainly because of 2 reasons when seen from the patients' perspective, namely their ability to conceal their scars after surgery and their ability to understand the "concept" of this procedure through the consenting surgeons and/or the internet. As with every technique, the limitations of this technique must be well understood before successful application to achieve desired results. Since the majority of patients present with a symmetric pectus excavatum to which Nuss procedure is aptly suited, results have been reported also to be excellent. However, complex and combined forms of pectus deformities, as well as patients presenting with severe asymmetries should not be subjected to this technique to achieve sub-standard results. It should be clear that the "*Surgeon should not use a favorite technique*, *but choose the most appropriate technique for the patient- as one size does not fit all*". Surgeons should be aware of the limitations of Nuss procedure, and should be ready to offer the appropriate technique as chest wall deformities present a "wide spectrum" of structural anterior chest wall malformations.

Nuss procedure and the refinement of this technique has added an excellent surgical option for the surgical repair of pectus excavatum especially in view of the changing trends in global fashions where exposure of the body image with a midline scar is now acceptable even to a lesser extent by teenagers and adolescents that would qualify to have repairs. Nuss procedure has a series of steps and technical considerations, which when respected can offer excellent results along with other advantages of short hospital stay and early recovery. The judicial use of an accessory pectus bar has been found to offer advantages in patients with rigid chest walls or those presenting with a platythorax in offering excellent results as well as to aid in minimizing of postoperative pain or discomfort.

Nuss procedure in the suitable sub-set of patients offers a single technique with global acceptance and for comparison of results based on a common denominator. The technique has laid the basis for the principle that the chest wall is not rigid and that it there is possibility to mold it into a desired shape and contours. This principle of "*chest wall correction through molding*" by Nuss procedure has triggered the development of the minimal access technique for the repair of pectus carinatum. This principle of "*chest wall correction through molding*" has had a further domino effect which has led to the revolutionary evolution of conservative modes of treatment for patients with pectus deformities with the use of (a) Vacuum bell for pectus excavatum and (b) orthotic braces for pectus carinatum.

The Cons

> *Damocles was enjoying himself immensely until he saw the sharp sword suspended over his head on a horse-hair … Cicero in Tusculian Disputes*

During the last decade, those who are involved in the treatment of pectus excavatum, and lately also pectus carinatum, are facing the choice to apply either the conventional "open" approach or the more recent Nuss-operation, a procedure which applies inlaying steel bars, to remodel the thoracic cage. In this debate, advocates of the Nuss-procedure are using the now outmoded open Ravitch procedure as a "straw-man" against which the merits of the Nuss-operation is argued. As far as the Ravitch-operation is concerned, one has to realize that while the elements of the "classic" Ravitch-operation, performed through a long vertical incision and leaves the sternum unsupported, are indeed the basis of most open pectus repairs, the procedure as such is but history. The

Ravitch-operation has largely been abandoned because of the poor long-term results, and replaced with its modern modifications, performed through a 5–6 cm exposure and using permanent support. These procedures provide excellent results and re-operation unnecessary.

When we compare these methods to the Nuss-operation, it needs to be emphasized that the Nuss-operation is not only "minimally invasive" as touted by its advocates, but in reality it is "maximally intrusive". Depending upon the number of rods inserted, it needs 2–6, one to one-and-half cm long lateral incisons, plus an additional hole for the videoscope. This certainly "adds up" to the single 5–6 cm long (albeit, anterior) incision needed to perform an up-to-date limited exposure "open" repair. Also, how can anybody call an operation "minimally invasive" where 2–3 foot-long metal rods are driven through both pleural cavities, passed by the width of a hair between the heart and the sternum, turned around to pressure-fracture all the costal cartilages and left there for extended periods of 3–5 years, then the incisions are to be opened up and the same procedure is performed in a "reverse" fashion?

Even if one accepts some of the aspects of the Nuss-operation as an advantage over open procedures, it has to be considered that the metal bars remain a threat for health and life, and like the sword of Damocles, endanger the patient as long as they are left *in loco*. This has been proven by the unprecedented number of publications during the last decade which we have had the opportunity to review. The articles published not only reveal what appears to be a significant increase in the total number of pectus repairs reported, about 20,000 in 10 years and four-fifths were Nuss-procedures, but also an astonishing number of complications (31 % Nuss-procedures *versus* 8.55 % open operations). We have also noted the emergence of a number of complications with the Nuss-procedure, which were never seen with the open approach: Cardiac arrest, laceration of the heart, the lung, the mammary artery, the inferior vena cava or the diaphragm, brachial plexus injury, Horner's and thoracic outlet syndromes. Late complications such as fracture, rotation and displacement of the bar, metallic allergy, hemorrhage and persistent pain occurring months and even years after the operation. There were also a disproportionately large number of reports of perioperative pneumonia, pericardial and pleural effusion, pneumothorax, atelectasis, recurrence of the deformity, reactive carinatum, and so on.

The argument that open pectus excavatum repair may induce development of acquired thoracic dystrophy, does not hold any water either. We strongly believe that restrictive thoracic dystrophy should never occur following a properly performed open pectus excavatum repair. Furthermore, contrary to the Nuss-procedure, open repair may be performed at even the very early age, but it should be performed correctly. In contrast, because of the specific concern as how the Nuss-bars may alter the growth of the costal cartilages, the procedure is not recommended in the growing child.

After all the above is said, one may wonder how the Nuss operation became so popular and that it is now considered by some the "gold standard" of therapy for pectus excavatum and even carinatum? The answer is not as complicated as it sounds.

First of all, the proponents of the procedure injected into the debate the magic words of contemporary medical marketing: "non-surgical" and "minimally invasive", despite that the Nuss-procedure requires two operations, up to six incisions thus, it is more invasive than any other pectus operation, save the sternal turnover of Nissen and Wada. The second reason for its popularity is that by its nature the Nuss-procedure, because of the lack of necessity of chest wall resection, allows specialists not formally trained in thoracic surgery to enter the new area uncharted by thoracic surgeons. As usually happens, the thoracic surgeons readily followed, especially because the news that "pectus excavatum now may be corrected without surgery", induced patients and pediatricians alike who may have been reluctant to face the scalpel, are not readily willing to accept the rods. (Figs. 64.1, 64.2, and 64.3).

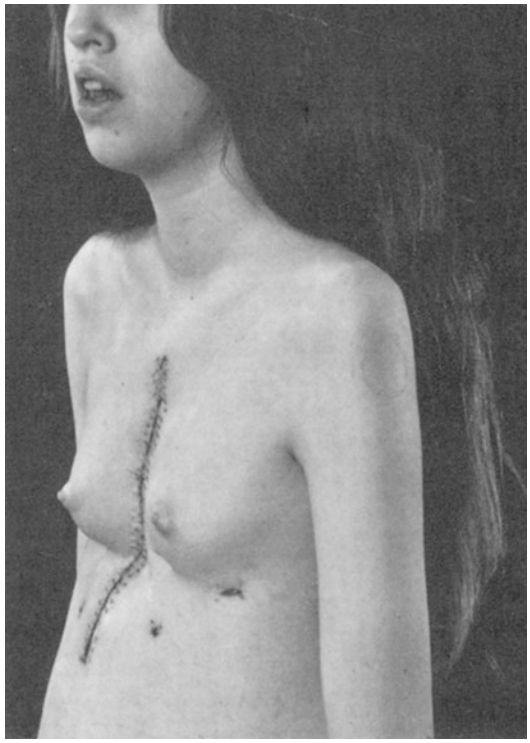

Fig. 64.1 Incision used to correct pectus excavatum in the 1970's (From: Ravitch MM: *Congenital deformities of the chest wall and their operative correction*. WB Saunders Co. Philadelphia, London, Toronto, 1977. With permission from Elsevier)

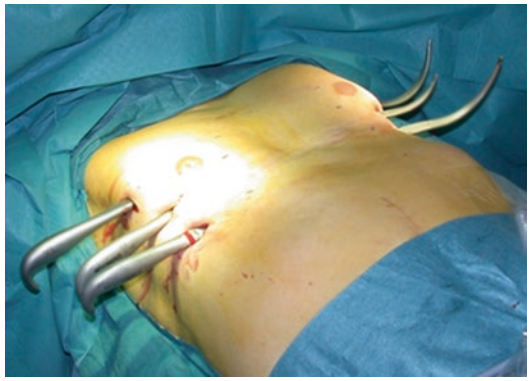

Fig. 64.2 Insertion of "guide-rods" in the course of the "minimally invasive" Nuss-procedure (From: Robicsek F, Hebra A: To Nuss or not to Nuss? Two opposing views. *Thorac Cardiovasc Surg*. 2009;21: 85–88 Courtesy of Dr. Hans Pilegaard. With permission from Elsevier)

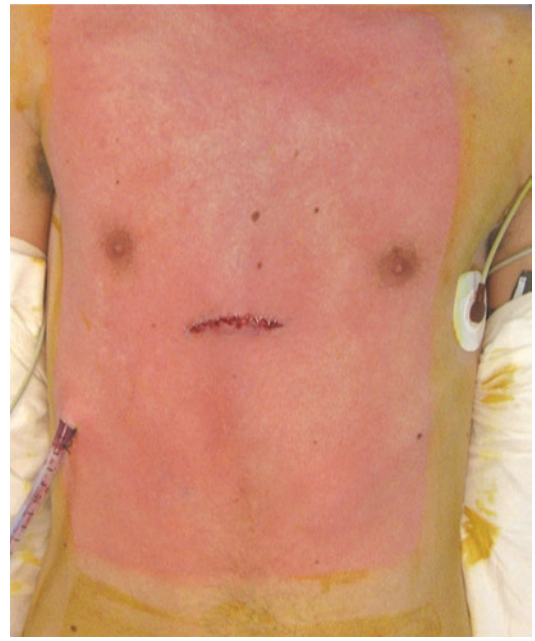

Fig. 64.3 Incision applied in the course of minimally invasive open repair of pectus excavatum (From: Robicsek F, Hebra A: To Nuss or not to Nuss? Two opposing views. *Thorac Cardiovasc Surg* 2009;21:85–88. With permission from Elsevier)

Suggested Reading

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