Stuart A. Green
Mark T. Dahl

Intramedullary Limb Lengthening

Principles and Practice

Springer

Intramedullary Limb Lengthening

Stuart A. Green • Mark T. Dahl

Intramedullary Limb Lengthening

Principles and Practice

Springer

Stuart A. Green, MD
Department of Orthopedic Surgery
University of California - Irvine
Orange, CA, USA

Mark T. Dahl, MD
Director, Limb Length and Deformity
Correction Clinics
Gillette Children's Specialty Healthcare
and University of Minnesota
St. Paul / Minneapolis, MN, USA

ISBN 978-3-319-60296-7 ISBN 978-3-319-60297-4 (eBook)
DOI 10.1007/978-3-319-60297-4

Library of Congress Control Number: 2017950662

Printed on acid-free paper

This Springer imprint is published by Springer Nature
The registered company is Springer International Publishing AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

Preface

This book evolved from a series of scientific exhibits for the American Academy of Orthopaedic Surgeons' annual meetings, created on behalf of The Limb Lengthening and Reconstruction Society–North America (LLRS–NA). Our 2013 exhibit, "Complications of Limb Lengthening and Deformity Correction," emphasized that switching the method of limb elongation from external fixation to intramedullary lengthening nails, although resulting in a far more comfortable experience for our patients, had not eliminated any of the potentially serious complications associated with limb lengthening, save for the eradication of pin and wire site pain, inflammation, and sepsis.

The LLRS–NA 2013 AAOS scientific exhibit, "Assessment for Deformity Correction: Identifying Hidden Deformities," created by the senior author with the assistance of Janet Conway, MD of Baltimore, focused attention on the dangers of not fully defining both the obvious and hidden deformities before embarking on a limb realignment program.

In 2016, with the help of Dror Paley, MD of Miami, the senior author reviewed "The History of Limb Lengthening," in an AAOS scientific exhibit, taking the viewer from the earliest attempts at limb elongation to the modern, fully implantable intramedullary lengthening nails.

Around that time, Ed Roschak, then CEO of Ellipse Technologies (now NuVasive Specialized Orthopaedics), manufacturer of the PRECICE® intramedullary nail, suggested that the senior author create a monograph summarizing the features of the aforementioned exhibits emphasizing, how knowledge of limb lengthening with external fixation systems teaches us to properly and safely use intramedullary lengthening devices.

The project expanded to encompass all such implants then available in North America. Fortunately, Mark Dahl, the junior author, was one of only two American orthopedic surgeons who had extensive clinical experience with all three intramedullary lengthening nails being used in North America. Thus, we were able to provide the reader with a comprehensive overview of the current status of the monograph's subject matter.

We also solicited clinical contributions from colleagues who helped produce the LLRS exhibits for the AAOS annual meetings, including Drs. Dror Paley, Rob Rozbruch, Austin Fragomen, John Herzenberg, and Janet Conway. These surgeons, along with Drs. Rainer Baumgart, Matthew Gardner, and Matt Dawson, generously

and unstintingly shared their experience and clinical cases with us for inclusion in this volume.

NuVasive also provided a grant to hire a talented artist and medical illustrator, Matthew Brownstein, to grace the pages of this monograph with outstanding images. We are most grateful for his contribution.

We have also included in this monograph an updated version of the cross-section atlas from the senior author's 1981 book, *Complications of External Skeletal Fixation: Causes, Prevention and Treatment*. We reasoned that the use of an intramedullary lengthening nail shares with the application of external skeletal fixation many transcutaneous protocols—percutaneous osteotomy, transverse locking screws, blocking screws, and guidance screws—inserted through minimally invasive incisions. Thus, an updated cross-section atlas seemed in order. In fact, we included a body part, the forearm, for which no foreseeable intramedullary lengthening implant is on the horizon, due to the narrow diameter of the involved bones. Nevertheless, engineering creativity often finds a way to solve such problems.

The reader should be advised that some of the applications of intramedullary lengthening nails illustrated in this monograph are "off-label"—not yet approved by the FDA for marketing in the USA. We have indicated the off-label usage when appropriate. The pioneering orthopedic surgeons who have contributed such cases to this book have sought to use existing implants in ingenious ways to solve serious clinical problems, always acting in the best interests of their patients. The results they have thus achieved bear witness to the resourcefulness of such practitioners.

Finally, by way of disclosure, we both serve as consultants for NuVasive and have royalty arrangements for elements of the PRECICE® nail and its derivatives. The senior author also receives royalties for Smith & Nephew's rancho cubes, used with their Ilizarov external fixation system and the Taylor Spatial Frame®.

Orange, CA, USA — Stuart A. Green, MD
Minneapolis, MN, USA — Mark T. Dahl, MD

Contents

1 Historical Background 1
Introduction 1
Early Pioneers 3
Parkhill 3
Codivilla 4
Putti 4
Abbott 4
Bost and Larsen 6
G. A. Ilizarov 6
First Patient 6
Distraction Osteogenesis 7
Ilizarov Apparatus 8
Ilizarov Institute, Kurgan, Siberia 9
Skepticism About Ilizarov's Method 9
Dissemination of the Ilizarov Method 10
The Cold War 10
Italian Connection 10
Worldwide Dissemination 12
ASAMI and LLRS 12
References 13
Further Reading 14

2 The Regenerate Bone 17
Histologic Features of the Regenerate 17
Histology, Early Phases 17
Histology, Vascularization 17
The Fibrous Interzone 19
Maturation Phases 19
Ossification 20
The Ring of Ranvier 21
Factors Influencing Regenerate Quality 21
Stability 21
Osteotomy/Corticotomy 22
Rate and Rhythm of Distraction 22

Ilizarov Terminology . . . 22
Introduction . . . 22
Location . . . 22
Sequence . . . 23
Action . . . 23
Objective . . . 23
Summary . . . 23
Conclusion . . . 24
Applications of the Ilizarov Method . . . 24
References . . . 25

3 Modifications of the Ilizarov Apparatus . . . 27
Half-Pin Mounting . . . 27
Computer-Controlled Hexapod Fixators . . . 27
Monolateral External Fixation . . . 29
Strategies to Shorten Time in Fixation . . . 30
Lengthening Over a Nail . . . 30
Lengthening and Then Nailing . . . 30
Lengthening and Then Plating . . . 31
References . . . 31

4 Intramedullary Lengthening Devices . . . 33
Background . . . 33
Elimination of the External Fixator . . . 33
Ratcheting Devices . . . 34
Bliskunov Intramedullary Nail . . . 34
Albizzia Intramedullary Nail . . . 35
Internal Skeletal Kinetic Distractor . . . 36
Rotating Spindle Devices . . . 37
Fitbone® Intramedullary Nail . . . 37
Phenix® Intramedullary Nail . . . 39
PRECICE® Intramedullary Nail . . . 39
Transferring Energy Through Soft Tissues . . . 39
Correcting Deformities with Intramedullary Lengthening Nails . . . 43
Achondroplasia and Related Conditions . . . 43
Achondroplasia . . . 43
Hypochondroplasia . . . 44
Turner's Syndrome . . . 45
Constitutional Short Stature . . . 45
Stature Surgery in Dwarfing Conditions . . . 45
Clinical Illustration: Four-Segment Intramedullary Lengthening . . . 46
Technical Considerations . . . 46
Uses of Intramedullary Lengthening Nails . . . 47
References . . . 48
Further Reading . . . 49

5 Surgical Principles of Intramedullary Lengthening 51
General Principles 51
Similarity to Intramedullary Trauma Nails 51
Differences from Intramedullary Trauma Nails 51
Guide Wire Issues 52
Short or Long Nails 52
Preoperative Planning 52
Assessing Deformities 53
Hidden Deformities 56
Full-Length X-ray Studies 57
Clinical Illustration: Failure to Identify Hidden Deformity 58
Level of the Osteotomy 59
The Osteotomy Formula 59
Fixator-Assisted Nailing 60
Periosteal Elevation 61
Venting 62
Depth Plunging 63
Compartment Syndrome 63
Thermal Injury While Drilling Vent Holes 64
Osteotomy Technique 64
Transverse Locking Screws: Proximal Guide 65
Testing the Implant 66
Preliminary Osteotomy Gap 66
A Spare Implant 66
Correcting Deformities with Intramedullary Lengthening Nails 66
Considerations for Tibiofibula Lengthening 67
The Fibula 67
Osteotomy of the Fibula 67
Fibular Fixation to the Tibia 69
Blocking Equinus 69
Tibial Insertion Point 71
Transverse Locking Screws: Distal Targeting 71
Considerations for Femoral Lengthening 75
Antegrade and Retrograde Nailing 75
Trochanteric or Piriformis Femoral Entry Portal 76
Retrograde Femoral Nailing 77
Bone Curvature 80
Clinical Illustration: Cortical Penetration 80
Considerations for Humeral Lengthening 82
Antegrade and Retrograde Nailing 82
Nail Size 83
The Proximal Humeral Portal 83
Clinical Illustration: Humeral Lengthening via Proximal Portal 85
The Distal Humeral Portal 85
Clinical Illustration: Humeral Lengthening via Distal Portal 86

Forearm . . . 88
Reaming the Canal . . . 88
References . . . 90
Further Reading . . . 91

6 Preventing Complications During Limb Lengthening . . . 93
Background . . . 93
Operative Strategies to Prevent Bone Deformities During Lengthening . . . 94
Bone Deformities During Lengthening . . . 94
Tibial Deformation . . . 94
Femoral Deformation . . . 94
Forearm Deformation . . . 95
Preventing Deformities (External Fixator Principles) . . . 95
Ring Prophylaxis . . . 95
Strategic Pin Placement . . . 96
The Waypoint Method . . . 96
Biomechanical Axis Considerations . . . 97
Correction with External Fixators . . . 99
Valgus Prophylaxis with Intramedullary Lengthening Nails . . . 100
Planning the Correction . . . 101
Preventing Deformities During Lengthening (Intramedullary Lengthening Nail Principles) . . . 101
Background . . . 101
Blocking Screws . . . 103
Clinical Illustration: No Blocking Screws . . . 105
Clinical Illustration: Secondary Blocking Screws . . . 107
Clinical Illustration: One Blocking Screw . . . 108
Clinical Illustration: Many Blocking Screws . . . 110
Blocking Screws as Guidance Screws . . . 110
Drill Bits for Guidance . . . 111
Regenerate Enhancement with Blocking Screws . . . 112
Excessive Correction . . . 113
The Osteotomy Site . . . 113
Angulation . . . 114
Translation and Rotation . . . 114
Angulation, Translation, and Rotation . . . 115
Latency, Rate, and Rhythm Modifications . . . 117
Rationale . . . 117
Latency . . . 117
Rate . . . 117
Rhythm . . . 118
Surgical Soft Tissue Release . . . 118
Rationale . . . 118
Subluxations and Dislocations . . . 120
References . . . 123
Further Reading . . . 124

7 Postoperative Management Considerations 125
Basic Principles 125
Initial Management 125
Postoperative Neurological Problems 125
Postoperative Vascular and Circulatory Problems 126
Compartment Syndrome 127
Contractures, Subluxations, and Dislocations 127
Avoiding Complications 128
Physiotherapy 128
Stretching 129
Splinting 130
Night Positioning 131
Functional Use of the Limb 134
Failure to Control Progressive Contractures 135
Reference 135
Further Reading 135

8 Postoperative Care, Day by Day 137
In Operating Room 137
After Surgery 137
Day-by-Day Management 137
Postoperative Day 1 137
Postoperative Days 2–4 138
Postoperative Days 5, 6, and 7 or Later 138
Latency 138
Distraction 139
Visualizing the Regenerate New Bone 139
After One Week of Distraction 139
Two Weeks of Distraction 141
Three Weeks of Distraction 141
Four Weeks of Distraction 141
Visits During Distraction 141
Judging the Regenerate 142
Assessment 142
Classification of Regenerate 143
Bone Mineral Density 143
Pixel Value Ratio 143
Ultrasound 144
The Regenerate Around a Lengthening Nail 144
Patterns of Regenerate 145
Biology of the Regenerate 145
Gardening 145
Biological Factors Retarding Regenerate 146
Chemical Factors Retarding Regenerate 146
Mechanical Factors Retarding Regenerate 147

Maturation (Consolidation) . . . 147
Visits During Maturation . . . 147
Tardy Regenerate Ossification . . . 147
Stimulating Regenerate Ossification . . . 148
Mechanical Enhancement . . . 149
Pulsed Electromagnetic Field . . . 149
Pulsed Ultrasound . . . 149
Chemical Enhancement . . . 150
Biological Enhancement . . . 150
Bone Grafting . . . 151
Hyperplastic Regenerate . . . 151
When Length Is Achieved . . . 152
Planning Implant Removal . . . 152
Post-Removal Management . . . 153
Conclusion . . . 153
References . . . 153
Further Reading . . . 155

9 Alternatives to Limb Lengthening . . . 159
Pelvic Tilting . . . 159
Shoe Lifts . . . 159
Surgery on the Normal Side . . . 160
Growth Arrest . . . 160
Shortening the Normal Limb . . . 161
Intramedullary Segmental Shortening . . . 161
Amputation . . . 162
Major Limb Deficiencies . . . 162
Assessing the Medical Literature . . . 163
The Limb Lengthening and Reconstruction Society-North America . . . 163
Problems with Publications . . . 163
The LLRS-AIM Complexity Score . . . 164
Judging Patient Wishes . . . 165
Patient-Reported Outcomes . . . 165
The Limb Deformity Modified SRS (LD-SRS) Score . . . 165
Section I: All patients . . . 166
Section 2: After Completion of Treatment . . . 168
References . . . 169
Further Reading . . . 170

10 The Future of Intramedullary Limb Lengthening . . . 171
Device Modifications . . . 171
Lengthening Plates . . . 171
Trauma Nail . . . 171
High Tibial Osteotomy . . . 172
Residual Limb Elongation . . . 174
Smart Remote Controllers . . . 175

Bone Transport 176
Bifocal Treatment 179
Expanded Indications 180
Cosmetic Stature Surgery 180
Conclusion 181
References 182
Further Reading 183

11 Cross-Section Atlas 185
Development of the Atlas 185
Background 185
Derivation of the Atlas 186
The Need for a Cross-Section Atlas for Intramedullary Lengthening . 186
Plunge Depth 186
The Atlas 186
Principles of the Atlas 186
References 203

Index 205

Historical Background

1

Introduction

Hind limb walking has been a defining feature of humans and their ancestors for at least 2 million years. Bipedalism confers clear advantages: the ability to see prey and predators while standing in tall savannah grasses and deep streams, the capacity to reach higher fruit in trees, and the aptitude to carry food and tools with the forelimbs.

Numerous subtle, but critical, adaptations evolved to make walking upright more energy efficient. The inward-sloping human femurs, for instance, reduce side-to-side displacement of the body's center of mass, saving calories during walking (chimpanzees can walk short distances on their hind limbs but waddle while doing so—a fatiguing gait pattern) (Fig. 1.1).

Moreover, an orthograde posture allows for an anteriorly positioned foramen magnum—the hole at the base of the skull for the spinal cord—permitting larger cranial capacity for the brain (Fig. 1.2).

Likewise, the slight posterior downslope of the knee's tibial weight-bearing surface permits a 10° knee flexion angle at midstance, thereby reducing up-and-down displacement of the center of mass—another energy saver. (A person with a straight stiff knee walks with a bouncing gait, like a car on a rutted road.)

Such anatomical modifications allow us to walk so efficiently that our center of mass displaces no more than 1 inch in any direction as it spirals through space-time, while we transition from right to left—and then back again—during the normal gait cycle.

Bipedalism comes, however, with a price. Unlike a three-legged dog, happily chasing tennis balls in the park, the loss of a lower limb in a human makes walking impossible without an ambulatory aid of some kind. Equally important, dysfunction of one or both lower limbs causes limping, a tiring way to get from place to place.

Although joint pain is the predominant cause of altered gait, unequal limb length disturbs gait as well. Ordinarily, humans can mask up to 1 inch of limb length difference by a combination of pelvic tilt and spinal curvature. Any discrepancy

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_1

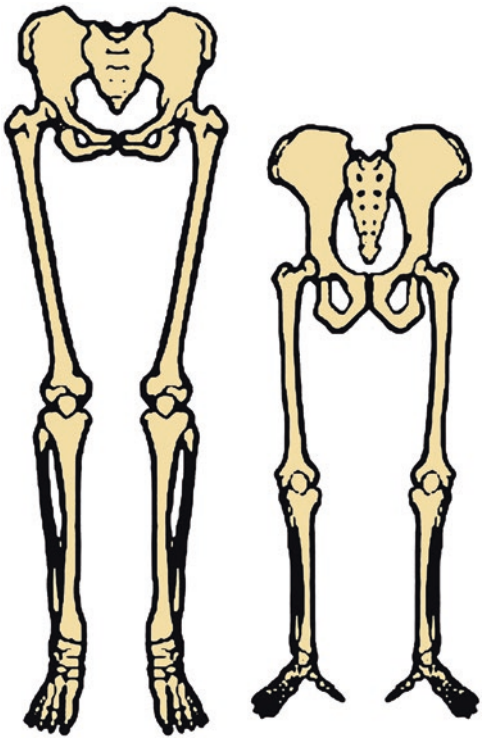


Fig. 1.1 Inward-sloping human femora (*left*), by placing the knees closer to the midline, reduce side-to-side waddling, compared to the chimpanzee (*right*). Copyright 2016 Zeeca Publishing Co

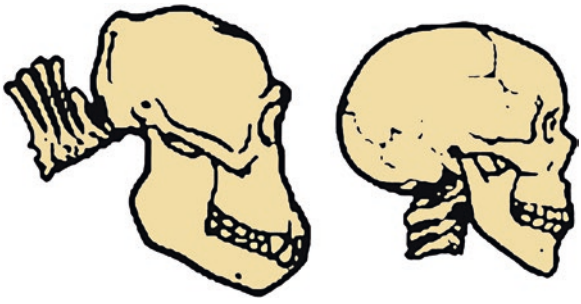


Fig. 1.2 Human upright posture moves the spinal column forward, allowing greater brain development. Copyright 2016 Zeeca Publishing Co

beyond that becomes an obvious impediment to efficient movement. Moreover, the incidence of osteoarthritis of the knee increases if limb lengths are unequal [1].

Until the middle of the last century, a shoe lift, sometimes many inches high, proved the only safe way to equalize lower limb length. With the advent of the aseptic operating room, pioneering surgeons searched for ways to elongate a short limb—or shorten the longer one—without causing a life- or limb-threatening infection while doing so.

There exists a distinct difference between elongating limbs that were originally of normal length in an adult, but were shorted by trauma, and those limbs whose

childhood growth was stunted by a birth defect or by a traumatic, infectious, or developmental process, inhibiting natural function of the growth cartilage. In the former situation, the limb's soft tissues were of normal length before the injury, so elongation becomes a matter of *restitutio ad integrum*. With inhibited childhood growth, the bone and soft tissues were never of normal size, so the periosseous soft tissues resist elongation of the limb.

Early Pioneers

Parkhill

Considering the above thoughts, it is no surprise that the first attempts at limb lengthening were to overcome post-trauma shortening. Clayton Parkhill, a surgeon working in Denver, Colorado, at the turn of the nineteenth century, devised the first practical external fixator that could be used in a variety of clinical situations [2]. Among the illustrations in his published report, Parkhill corrected a femoral malunion with side-to-side healing and substantial fragment overlap. In general, his results with such operations proved favorable (Fig. 1.3).

Fig. 1.3 Clayton Parkhill, of Denver, shown with his Parkhill bone clamp being used to treat a femoral malunion (1895). Copyright 2016 Zeeca Publishing Co

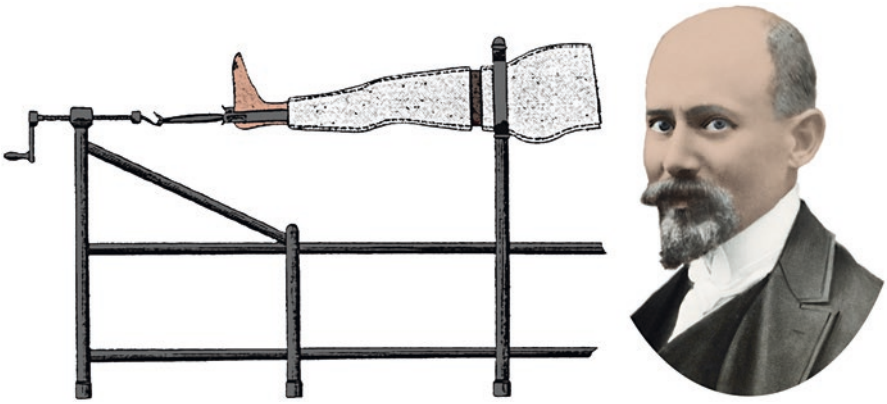


Fig. 1.4 Alessandro Codivilla, with his traction device for gaining limb length after osteotomy of the bone (1905). Copyright 2016 Zeeca Publishing Co

Codivilla

Alessandro Codivilla, a prominent Italian surgeon, used strong traction to restore length to shortened limbs [3]. Unfortunately, his patients remained hospitalized throughout the elongation process (Fig. 1.4).

Putti

Vittorio Putti, a student of Codivilla, first used an external skeletal fixator to slowly elongate a bone after a step cut osteotomy, thereby allowing some overlap and side-to-side contact between fragments when the full length was achieved [4]. The lengthening mechanism contained a spring within the elongating tube to maintain tension throughout the process, which no longer required continuous hospitalization. However, bone grafting was often necessary at the end of the elongation process (Fig. 1.5).

Abbott

San Francisco became a center of limb lengthening for children when LeRoy Abbott created a more stable external fixator in 1939 than the one used by Putti a generation earlier [5]. Abbott's device used two transosseous pins in each fragment, thereby enhancing the stability of the bone-fixator construct (Fig. 1.6). His innovative mind inspired generations of San Francisco orthopedic surgery residents. For this reason, the orthopedic alumni association of the University of California, San Francisco, is, to this day, named "The LeRoy Abbott Society."

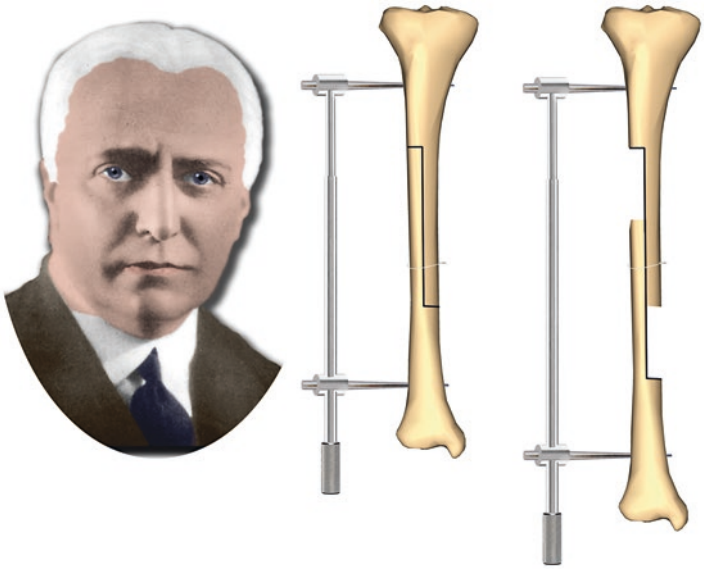


Fig. 1.5 Vittorio Putti and his spring-containing external fixator for limb lengthening (1921). Copyright 2016 Zeeca Publishing Co

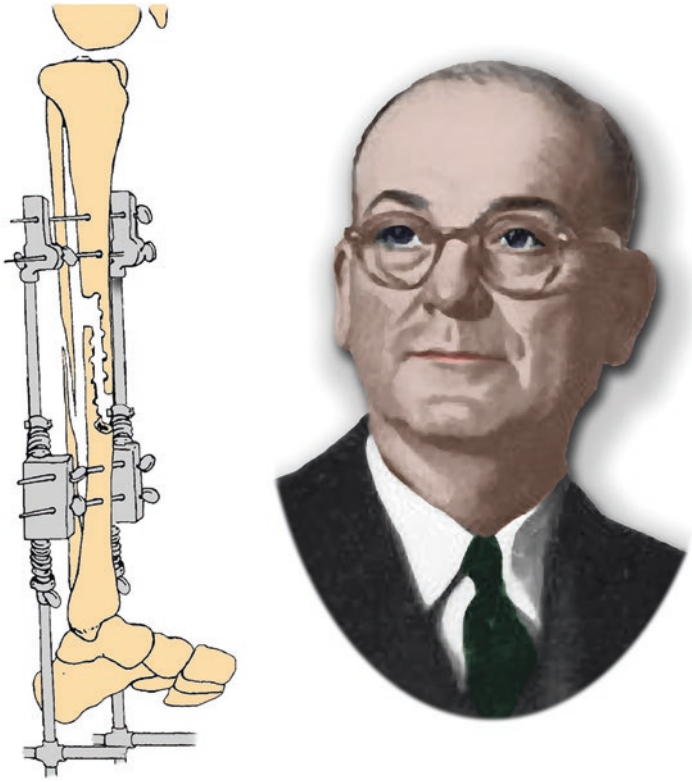


Fig. 1.6 LeRoy Abbott and his lengthening fixator (1939) containing two transosseous pins in each fragment. Copyright 2016 Zeeca Publishing Co

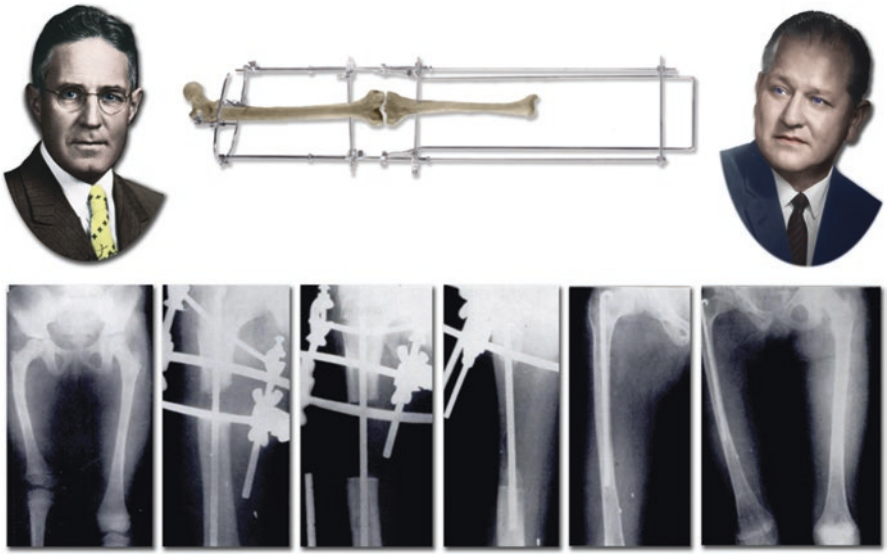


Fig. 1.7 Fredrick Bost (*left*) and Loren Larsen (*right*) of San Francisco, with their distraction system (*top*). In about half their cases, new bone formed in the distraction zone, as shown. Copyright 2016 NuVasive

Bost and Larsen

Fredrick Bost and Loren Larsen, both working at the San Francisco Unit of the Shriners Children's Hospital System, reported on their experience with a new external fixator for limb lengthening in 1956 [6]. It incorporated features of a Thomas splint and transfixion pins to slowly distract an osteotomized bone (Fig. 1.7). In about half of their patients, newly formed bone filled the widening distraction zone, a harbinger of Ilizarov's discoveries. However, the phenomenon was unpredictable, and half of their patients required bone grafting to fill the distraction gap.

G. A. Ilizarov

In 1951, Soviet surgeon G. A. Ilizarov (Fig. 1.8) unlocked a previously hidden capacity of bone to form new osseous tissue reliably in a widening distraction gap under the appropriate conditions of stability and elongation [7].

First Patient

While serving as a physician at a veterans' clinic in Kurgan, Siberia, USSR, Doctor Ilizarov cared for a patient who had sustained a traumatic below-knee amputation

Fig. 1.8 Gavriil Abramovich Ilizarov, 1921–1992. Copyright 2016 Zeeca Publishing Co

during the Second World War. As sometimes happens in such cases, a 90° flexion deformity of the knee evolved, making it impossible to fit the retired soldier with an artificial limb.

Moreover, the knee joint underwent spontaneous bony ankylosis, such that a solid mass of bone filled the entire region that had formerly been the man's knee.

Ilizarov's treatment plan started with an oblique cut through the bone mass, followed by application of a simple external fixation device, with one tensioned wire through the femur, and another through the tibial stump. These wires were secured to half rings, which, in turn, were connected by threaded distraction rods.

The patient was instructed to turn the threaded rods to a small extent each day, thereby separating the half rings, a maneuver that would gradually straighten the knee. Ilizarov had planned to fill in the resulting triangle-shaped bone defect with a bone graft once the distraction had been completed, eventually allowing proper fit of an artificial limb. The grafting operation, however, was delayed as Dr. Ilizarov went on a Crimean peninsula summer vacation.

To his surprise, upon returning, Ilizarov discovered that newly formed bone had filled the entire distraction gap, eliminating the need for a bone graft.

Distraction Osteogenesis

Dr. Ilizarov gradually came to realize that he had made a unique discovery, one that he called *distraction osteogenesis*—formation of new bone tissue in a widening distraction gap [8].

Fig. 1.9 Distraction osteogenesis with the Ilizarov apparatus. Copyright 2016 NuVasive

Over time, Dr. Ilizarov and his Russian co-workers developed strategies to lengthen not only the long tubular bones of the upper and lower extremities but also the tarsal and carpal bones, as well as flat bones of the cranium, mandible, and maxilla [9–19]. Ilizarov's external skeletal fixator, as it evolved over time, consisted of stainless steel rings that surrounded the limb and were secured to the involved bone with tensioned transcutaneous, transosseous wires (Fig. 1.9).

Ilizarov Apparatus

By incorporating hinges in the external fixator configuration, Ilizarov realized that he could utilize distraction osteogenesis to correct virtually any bone deformity, whether congenital or acquired. Likewise, Ilizarov developed a strategy to overcome substantial defects in long bones by osteotomizing one of the remaining bone fragments and dragging the intercalary piece of bone slowly through the limb with wires connected to the apparatus [20]. This technique, now called "bone transport" in orthopedic literature worldwide, was one of the most revolutionary of Ilizarov's ideas [21] (Fig. 1.10).

Fig. 1.10 Bone transport with the Ilizarov apparatus. The intercalary bone fragment is moved downward to eliminate a bone defect. Regenerate bone fills in the distraction zone during the process. Copyright 2016 NuVasive

Ilizarov Institute, Kurgan, Siberia

What started as a log cabin, one-room veterans' clinic eventually grew into the largest orthopedic hospital in the world (Fig. 1.11), staffed by more than 350 surgeons, who perform surgeries based exclusively on the Ilizarov method of distraction osteogenesis. A child with a congenital (Fig. 1.12) or acquired (Fig. 1.13) limb length deficiency may be on a list for several years awaiting surgery at the institute.

Skepticism About Ilizarov's Method

Most doctors observing such a phenomenon would have dismissed the formation of new bone tissue in a widening distraction gap as an oddity unique to the patient being treated [22]. This is because in the 1950s (an era before the standard use of internal fixation—plates, screws, and intramedullary rods), long bone fractures of the lower extremity were typically treated with prolonged bed rest with a skeletal traction system, created by a combination of ropes and pulleys and weights, that aligned the fracture fragments, while they slowly united over a period of several months.

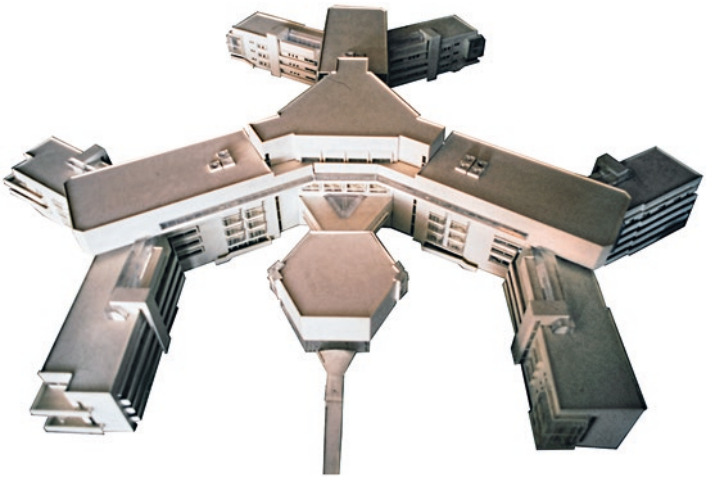


Fig. 1.11 The Russian Ilizarov Scientific Center for Restorative Traumatology and Orthopaedics, in Kurgan, Siberia, Russia, is the world's largest orthopedic hospital

With such longitudinal traction treatment strategies, excessive weight on the alignment ropes led to distraction of the bone ends, commonly resulting in non-union of the fracture. Therefore, distraction was considered the ultimate antithesis of fracture healing.

Dissemination of the Ilizarov Method

The Cold War

Between 1951 and 1981, Ilizarov's methods gradually became utilized throughout the Communist world. However, as the Iron Curtain limited discourse between the East and West, virtually nothing was known of these techniques outside the Soviet sphere of influence. Thus, by 1983, Cuban orthopedic surgeons had produced a Spanish language textbook on the Ilizarov method [23], while 90 miles away, in the United States, surgeons knew nothing of Ilizarov's discoveries. In fact, the Red Army wanted it that way: techniques employing distraction osteogenesis could return a fully trained but seriously injured soldier to his unit to fight another day.

Italian Connection

In the late 1970s, Italian orthopedic surgeons learned about the Ilizarov method from patients who returned to Italy after being treated for serious motor scooter injuries sustained while vacationing in nearby Communist Yugoslavia. Likewise, Carlo

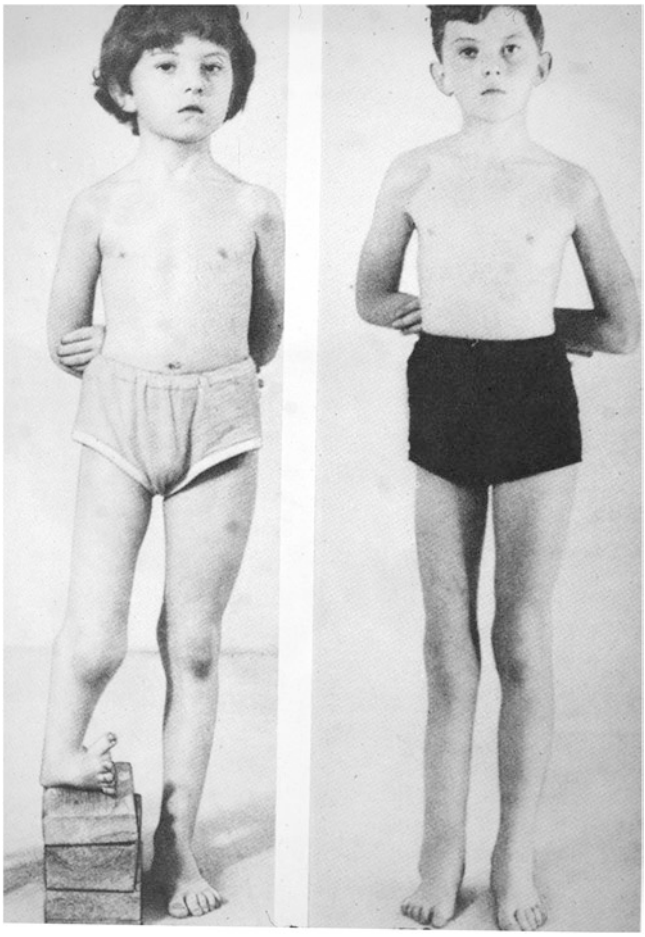


Fig. 1.12 Ilizarov patient: Simultaneous deformity correction and limb lengthening for congenital defect of tibia and fibula. (*Left*) Before treatment. (*Right*) After treatment

Fig. 1.13 Ilizarov patient: Simultaneous deformity correction and limb lengthening for postinfection growth arrest. Reprinted from *Transosseous Osteosynthesis*, "Correction of Deformities of Long Tubular Bones with Simultaneous Limb Lengthening," 1992, pp. 329–368, G. A. Ilizarov, Copyright Springer-Verlag Berlin Heidelberg. With permission of Springer

Mauri, a famous Italian explorer/writer, went to Siberia to be personally treated by Dr. Ilizarov for a recalcitrant infected non-union of his tibia. Mauri's return to his native land after a successful cure stimulated interest among Italian doctors.

In 1981, the city of Milan, Italy, had a Communist party mayor who arranged for a group of Italian surgeons to travel into the heartland of the Soviet Union to visit and learn from Professor Ilizarov. Shortly thereafter, these surgeons began applying distraction osteogenesis principles in their own clinics. Five years later, the Italians presented their preliminary results at international conferences, stimulating the worldwide interest in the Ilizarov method.

Worldwide Dissemination

In 1986, Canadian orthopedist Dror Paley joined a group of Italians visiting Ilizarov in Kurgan, becoming the first North American to visit the institute in Siberia. A year later Dr. Stuart Green of California, a Professor of Orthopedic Surgery at UC Irvine, became the first American to do so.

In 1987, the first English-speaking workshop was held in Kurgan, USSR. Many others have followed.

ASAMI and LLRS

In nations far and wide, surgeons interested in limb lengthening and deformity correction soon formed societies to provide a forum for the dissemination and interchange of ideas about Ilizarov's methods. The first of these groups formed, naturally enough, in Italy. The associations were typically named the Association for the Study and Application of the Methods of Ilizarov (ASAMI). Dror Paley and Stuart Green formed ASAMI-North America, which eventually changed its name to the Limb Lengthening and Reconstruction Society—North America (LLRS-NA), to emphasize the more universal nature of the group's focus, moving beyond Ilizarov's contributions into the twenty-first century.

In 2016, LLRS-NA will celebrate its 25th anniversary in Charlotte, North Carolina. The organization is a member of the Board of Specialty Societies of the American Academy of Orthopaedic Surgeons and offers a full-day program during Specialty Day at the AAOS Annual Meeting, in addition to its own two-and-a-half day meeting each year in the springtime. The LLRS-NA also participates in international gatherings of surgeons interested in limb lengthening and deformity correction, helping to sponsor such a meeting in Miami, Florida, in November 2015.

References

1. Harvey WF, Yang M, Cooke TD, Segal NA, Lane N, Lewis CE, et al. Association of leg-length inequality with knee osteoarthritis: a cohort study. Ann Intern Med. 2010;152(5):287–95.
2. Parkhill CI. Further observations regarding the use of the bone-clamp in ununited fractures, fractures with malunion, and recent fractures, with a tendency to displacement. Ann Surg. 1898;27(5):553–70.
3. Codivilla A. On the means of lengthening, in the lower limbs, the muscles and tissues which are shortened through deformity. Amer J Ortho Surg. 1905;2:353–69.
4. Putti V. Operative lengthening of the femur. Surg Gynecol Obstet. 1934;58:318–24.
5. Abbott LC. The operative lengthening of the tibia and fibula. J Bone Joint Surg Am. 1927;9:128–35.
6. Bost FC, Larsen LJ. Experiences with lengthening the femur over an intramedullary rod. J Bone Joint Surg Am. 1956;38:567–84.
7. Ilizarov GA. A method of uniting bones in fractures and an apparatus to implement this method. USSR Authorship Certificate. 1952. Report No.: 98471.
8. Ilizarov GA. Transosseous osteosynthesis. Heidelberg: Springer-Verlag; 1991.
9. Ilizarov GA. A decade of experience in the application of the author's apparatus for compression osteosynthesis in traumatology and orthopedics. Probl Rehabil Surg Traumatol Orthop. 1962;8:14.
10. Ilizarov GA. Restorative and radical-restorative operations in tuberculous lesions of joints and their sequelae. In: Materials of scientific-practical conferences on tuberculosis of bones and joints in materials of scientific-practical conferences on tuberculosis of bones and joints. Tomsk: USSR; 1963. p. 162–71.
11. Ilizarov GA. General principles of transosteal compression and distraction osteosynthesis. In: Proceeding of scientific session of institutes of traumatology and orthopedics. Leningrad: USSR; 1968. p. 35–9.
12. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Related Res. 1989;239:263–85.
13. Ilizarov GA, Devyatov AA. Operative elongation of the leg with simultaneous correction of the deformities. Ortop Travmatol Protez. 1969;30(3):32.
14. Ilizarov GA, Devyatov AA. Elimination of joint and ankle contractures by means of external fixation apparatuses. Kurgan: USSR; 1971.
15. Ilizarov GA, Gracheva VI. Bloodless treatment of congenital pseudarthrosis of the crus with simultaneous elimination of shortening using dosed distraction. Ortop Travmatol Protez. 1971;32(2):42.
16. Ilizarov GA, Perelitzkich PF. Regeneration of diaphyseal bone tissue in lengthening after closed direct oblique or screw-shaped osteoclasia. Ortop Travmatol Protez. 1978;11:89–93.
17. Ilizarov GA, Schreiner AA, Imerlishvili IA, Bakhlykov YN, Onirkova AM, Marterl II, editors. On the problem of improving osteogenesis conditions in limb lengthening. In: First international symposium on experimental and clinical aspects of transosseous osteosynthesis. Kurgan: USSR; 1983.
18. Ilizarov GA, Trokhova VA. Surgical lengthening of the femur. Ortop Travmatol Protez. 1973;34(11):51.
19. Ilizarov GAD, A.A. Operative elongation of the leg. Ortop Travmatol Protez. 1971;32(8):20.
20. Ilizarov GA, Ledioev VI. Replacement of defects of long tubular bones by means of one of their fragments. Vestn Khir. 1969;102:77.
21. Green SA, Jackson JM, Wall DM, Marinow H, Ishkanian J. Management of segmental defects by the Ilizarov intercalary bone transport method. Clin Orthop Relat Res. 1992;280:136–42.
22. Aronson J, Harrison BH, Cannon DJ, Lubansky HJ. Mechanical induction of osteogenesis: the importance of pin rigidity. J Pediatr Orthop. 1988;8:396–401.
23. Ceballos-Mesa A. Fijacion externa de los huesos. Havana: Editorial Cientifico-Tecnica; 1983.

Further Reading

1. Aronson J, Johnson E, Harp JH. Local bone transport for the treatment of intercalary defects by the Ilizarov technique. Clin Orthop Rel Res. 1989;243:71.
2. Demiralp B, Ege T, Kose O, Yurttas Y, Basbozkurt M. Reconstruction of intercalary bone defects following bone tumor resection with segmental bone transport using an Ilizarov circular external fixator. J Orthop Sci. 2014;19(6):1004–11.
3. Green SA. Skeletal defects a comparison of bone grafting and bone transport for segmental skeletal defects. Clin Orthop Relat Res. 1994; (301):111–7.
4. Griffith MH, Gardner MJ, Blyakher A, Widmann RF. Traumatic segmental bone loss in a pediatric patient treated with bifocal bone transport. J Orthop Trauma. 2007;21(5):347–51.
5. Iacobellis C, Berizzi A, Aldegheri R. Bone transport using the Ilizarov method: a review of complications in 100 consecutive cases. Strateg Trauma Limb Reconstr. 2010;5(1):17–22.
6. Ilizarov GA. A decade of experience in the application of the author's apparatus for compression osteosynthesis in traumatology and orthopedics. Probl Rehabil Surg Traumatol Orthop. 1962;8:14.
7. Ilizarov GA. Restorative and radical-restorative operations in tuberculous lesions of joints and their sequelae. In: Materials of scientific-practical conferences on tuberculosis of bones and joints in materials of scientific-practical conferences on tuberculosis of bones and joints. Tomsk: USSR; 1963. p. 162–71.
8. Ilizarov GA. General principles of transosteal compression and distraction osteosynthesis. In: Proceeding of scientific session of institutes of traumatology and orthopedics. Leningrad: USSR; 1968. p. 35–9.
9. Ilizarov GA, Devyatov AA. Operative elongation of the leg with simultaneous correction of the deformities. Ortop Travmatol Protez. 1969;30(3):32.
10. Ilizarov GA, Devyatov AA. Elimination of joint and ankle contractures by means of external fixation apparatuses. Kurgan: USSR; 1971.
11. Ilizarov GA, Devyatov AA. Operative elongation of the leg. Ortop Travmatol Protez. 1971;32(8):20.
12. Ilizarov GA, Gracheva VI. Bloodless treatment of congenital pseudarthrosis of the crus with simultaneous elimination of shortening using dosed distraction. Ortop Travmatol Protez. 1971;32(2):42.
13. Ilizarov GA, Trokhova VA. Surgical lengthening of the femur. Ortop Travmatol Protez. 1973;34(11):51.
14. Ilizarov GA, Perelitzkich PF. Regeneration of diaphyseal bone tissue in lengthening after closed direct oblique or screw-shaped osteoclasia. Ortop Travmatol Protez. 1978;11:89–93.
15. Ilizarov GA, Schreiner AA, Imerlishvili IA, Bakhlykov YN, Onirkova AM, Marterl II, editors. On the problem of improving osteogenesis conditions in limb lengthening. In: First international symposium on experimental and clinical aspects of transosseous osteosynthesis. Kurgan: USSR; 1983.
16. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263–85.
17. Karargyris O, Polyzois VD, Karabinas P, Mavrogenis AF, Pneumaticos SG. Papineau debridement, Ilizarov bone transport, and negative-pressure wound closure for septic bone defects of the tibia. Eur J Orthop Surg Traumatol. 2013;24(6):1013–7.
18. Lavini F, Dall'Oca C, Bartolozzi P. Bone transport and compression-distraction in the treatment of bone loss of the lower limbs. Injury. 2010;41(11):1191–5.
19. Paley D. Bone transport: the Ilizarov treatment for bone defects. Tech Orthop. 1989;4(3):80–93.
20. Rigal S, Merloz P, Le Nen D, Mathevon H, Masquelet AC. Bone transport techniques in post-traumatic bone defects. Orthop Traumatol Surg Res. 2012;98(1):103–8.

21. Takahashi M, Kawasaki Y, Matsui Y, Yasui N. Fragmental bone transport in conjunction with acute shortening followed by gradual lengthening for a failed infected nonunion of the tibia. J Orthop Sci. 2010;15(3):420–4.
22. Uzel AP, Lemonne F, Casoli V. Tibial segmental bone defect reconstruction by Ilizarov type bone transport in an induced membrane. Orthop Traumatol Surg Res. 2010;96(2):194–8.
23. Zhang Q, Yin P, Hao M, Li J, Lv H, Li T, et al. Bone transport for the treatment of infected forearm nonunion. Injury. 2014;45(12):1880–4.

The Regenerate Bone

2

Histologic Features of the Regenerate

Histology, Early Phases

Under the microscope, the tissue that forms in the widening distraction gap under the influence of steady distraction has some interesting features. In the early phases of distraction osteogenesis, the widening gap contains poorly differentiated connective tissue. This substance slowly organizes into small cones of newly forming bone attached to the fragment ends, separated by a fibrocartilaginous layer. With additional time, distraction, and stability, the entire zone fills with osseous tissue, often with a band of cartilaginous tissue zigzagging across the middle (Fig. 2.1).

Histology, Vascularization

In the early phases of distraction osteogenesis, parallel longitudinal columns of fibrous tissue form. When cut in cross section and viewed under a microscope, these columns have the appearance of honeycombs, whereas when cut longitudinally, they resemble the striations in a stalk of celery [1, 2] (Fig. 2.2).

This new tissue is highly vascularized, with newly formed blood vessels occupying the spaces between the longitudinal fibers.

At the electron microscopic level, the newly forming fibrous tissue, subjected to continuous traction by the elongating mechanism, contains stretched out mitochondria and elongated endoplasmic reticulum (Fig. 2.3).

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_2

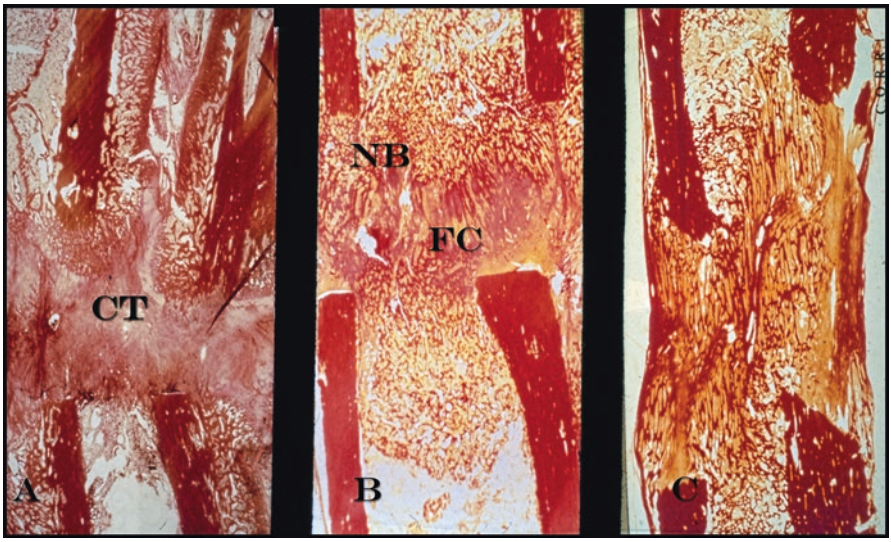


Fig. 2.1 Formation of the bone in a widening distraction zone. (**a**) Initially the zone is filled with connective tissue (CT). (**b**) Next, new osseous tissue (NB) forms at the fragment ends, while fibrocartilage (FC) forms in the middle region. (**c**) Eventually, the bone, oriented longitudinally, consolidates the distraction zone. Reprinted from *Transosseous Osteosynthesis*, "The Tension-Stress Effect on the Genesis and Growth of Tissues," 1992, pp. 137–255, G. A. Ilizarov, Copyright Springer-Verlag Berlin Heidelberg. With permission of Springer

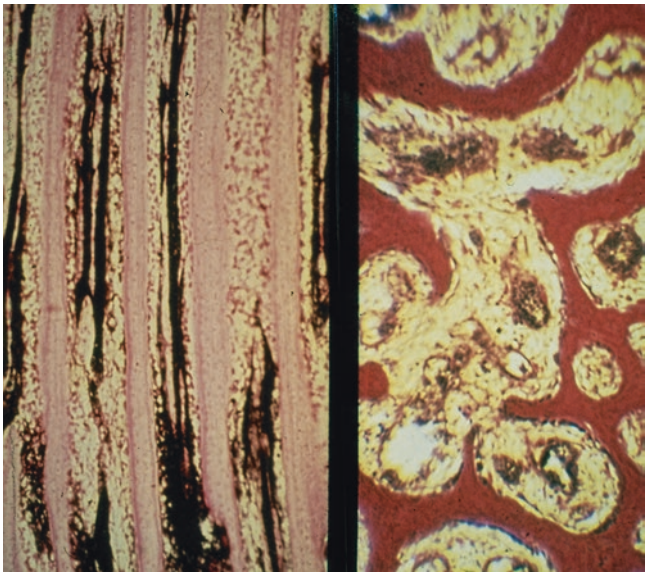


Fig. 2.2 Longitudinal (*left*) and transverse (*right*) section of newly formed regenerate bone with India ink stain. Blood vessels show as black. Note the vertical striations in the longitudinal section and the honeycomb appearance of the transverse section. Reprinted from *Transosseous Osteosynthesis*, "The Tension-Stress Effect on the Genesis and Growth of Tissues," 1992, pp. 137–255, G. A. Ilizarov, Copyright Springer-Verlag Berlin Heidelberg. With permission of Springer

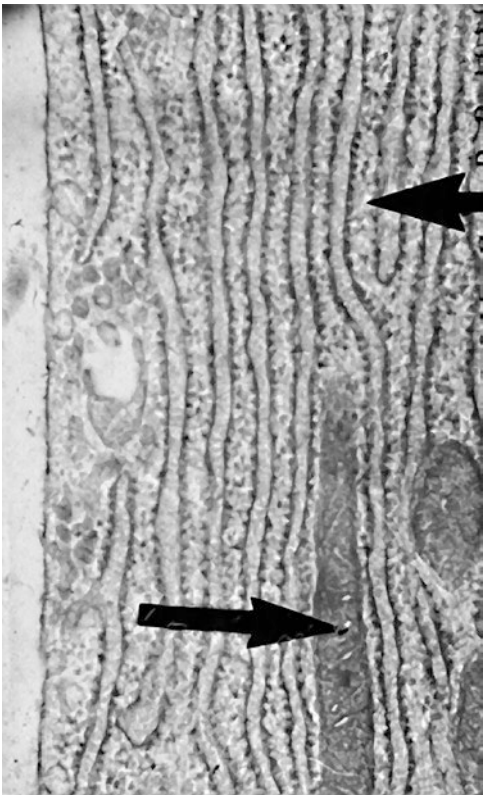


Fig. 2.3 Electron micrograph of a fibrocyte in the distraction zone. Notice the elongated mitochondria (*lower arrow*) and the stretched-out endoplasmic reticulum. 22,000×. Reprinted from *Transosseous Osteosynthesis*, "The Tension-Stress Effect on the Genesis and Growth of Tissues," 1992, pp. 137–255, G. A. Ilizarov, Copyright Springer-Verlag Berlin Heidelberg. With permission of Springer

The Fibrous Interzone

When external fixation is used to create the distraction regenerate, a dark zigzag line often appears traversing the center of the distraction gap [3, 4]. This region is called "the interzone." It is perceived as a kind of growth plate of the distraction regenerate. The interzone would naturally be hard to see when an intramedullary device occupies the middle of the bone (Fig. 2.4).

As the white lines get denser and thicker, they eventually cross the interzone, indicating that ossification has proceeded along the entire regenerate. This usually does not occur until the distraction phase of the process has been completed.

Maturation Phases

The fibrous tissue in the elongating distraction zone consists primarily of collagen. Gradually, tiny calcium hydroxyapatite crystals are deposited within the collagen fibers, stiffening them. Such collagen fibers with embedded calcium hydroxyapatite

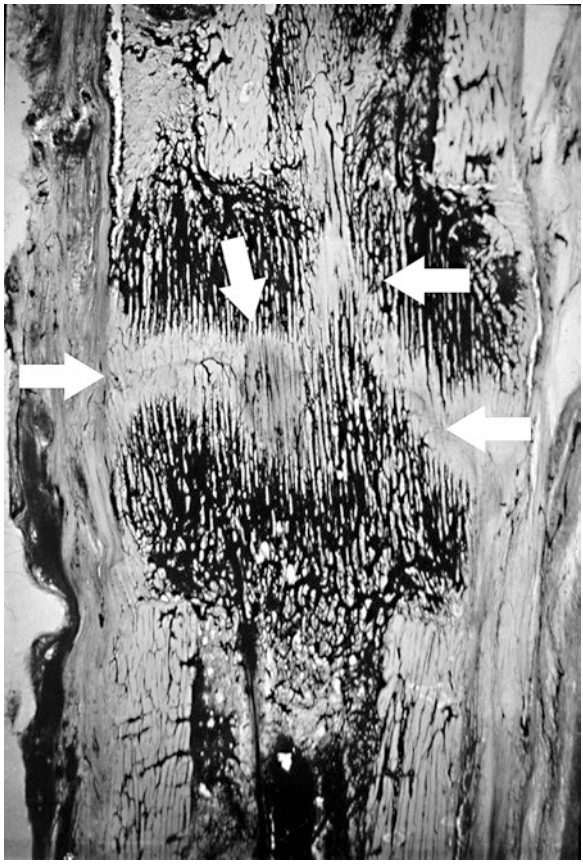


Fig. 2.4 India ink preparation showing the fibrous interzone (*white arrows*) zigzagging across the middle of the regenerate region. Reprinted from *Transosseous Osteosynthesis*, "The Tension-Stress Effect on the Genesis and Growth of Tissues," 1992, pp. 137–255, G. A. Ilizarov, Copyright Springer-Verlag Berlin Heidelberg. With permission of Springer

crystals are the basic ingredients of bone. The calcium hydroxyapatite crystals, because of their density, absorb x-ray beams. These absorbed beams never darken an x-ray film, so they appear white when the film is viewed on a translucent view box.

Thus, increasing maturation of the regenerate bone is characterized by progressive lightening of the x-ray image between the distracted bone ends. Under ideal conditions, the longitudinal orientation of the maturing columns of bone is visible on x-ray studies, seen as thin white bands separated by darker, more radiolucent bands. The white bands, appearing on x-ray studies as though painted against a dark surface with a paintbrush, are most mature at the bone ends and tend to fade toward darkness at the center of the regenerate (Fig. 2.5).

Ossification

Ilizarov believed that he had discovered a new kind of bone formation, differing from both intramembranous ossification (that occurs underneath the periosteum in a growing child) and endochondral ossification (that occurs at both the growth plate

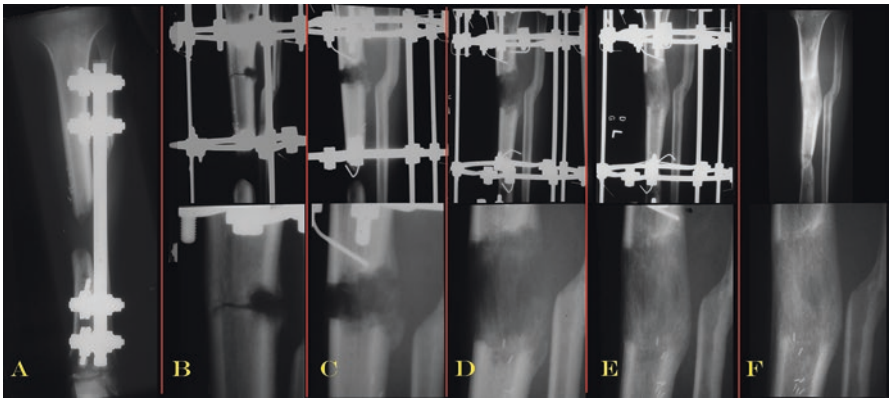


Fig. 2.5 (**a**) Segmental defect of the tibia, stabilized with a monolateral fixator. (**b**–**f**) Placement into a ring fixator and osteotomy through pinhole showing progressive phases of distraction zone elongation and maturation (*top* = whole bone; *bottom* = close-up of regenerate). (**f**) Final image with frame removed. Copyright 2016 Zeeca Publishing Co

of a child and during the process of healing of a fresh fracture treated in a cast). In the latter situation, the blood clot created from the ends of the broken bones first turns into cartilage, which, in turn, matures into bone.

The Ring of Ranvier

Aronson et al. [5], at Little Rock Children's Hospital, has shown with histological studies that regenerate bone forming in a widening distraction zone is indistinguishable from bone that forms in a growing child at the circumferential outer corner of a growth plate, in a zone called "the Ring of Ranvier." In this region, intramembranous bone forms on the undersurface of the periosteum which is simultaneously being stretched by the elongating power of the growth plate. The combination of lengthening and widening of bone characteristic of childhood creates osseous tissue in longitudinal columns with the same appearance as Ilizarov's distraction regenerate.

Thus, Ilizarov's method reactivates a quiescent means of bone formation originally designed by nature for a growing child.

Factors Influencing Regenerate Quality

Stability

Ilizarov employed a canine model to explore factors that optimized outcomes. He and his co-workers learned that the best quality of regenerate bone formation occurs when there is excellent rotational and side-bending bone stability in the ring fixator that, nevertheless, allows axial motion in a trampoline effect [6].

Osteotomy/Corticotomy

Likewise, researchers at Ilizarov's institute determined that good-quality bone formation is more likely to occur when there is a "sparing" (gentle) osteotomy of bone to create the distraction gap [6]. Moreover, they learned that protecting the endosteal blood supply by cracking the cortex without crossing the medullary canal (the "corticotomy") also enhanced bone formation in the widening distraction gap [1].

Rate and Rhythm of Distraction

Scientists in Kurgan determined that the best distraction rate is typically 1 mm per day, divided into multiple doses. The more highly fractionated the distraction, the better the quality of the bone [2]. They also learned that it takes at least twice as long for the bone to mature once distraction is complete than it did to elongate the fragments in the first place, with this being the minimum amount of time required for maturation, generally occurring in children. Regenerate maturation in adult bones takes longer, perhaps three or four times longer than the distraction phase time cycle. In cases where the soft tissues surrounding the regenerate region have been injured, maturation can be slow indeed. A region of the limb that had previously been irradiated to treat malignancy may not support maturation of the regenerate at all.

Ilizarov Terminology

Introduction

"Bilocal consecutive distraction-compression osteosynthesis" is Ilizarov's term for the procedure referred to in the Western orthopedic literature as "bone transport"—the filling in of a segmental osseous defect by pulling a bone fragment through the tissues. Ilizarov's terminology for the numerous strategies of osseous reconstruction helps surgeons communicate the conceptual framework for any treatment plan. For this reason, orthopedists using Ilizarov's methods should employ Ilizarov's terminology to clearly describe treatment tactics to knowledgeable colleagues in North America, Europe, and elsewhere in the world. Furthermore, Ilizarov's publications will be more easily understood and enjoyed if readers are familiar with his descriptive terminology.

Ilizarov usually characterizes procedural strategies by four terms, which form the basis of treatment: location, sequence, action, and objective.

Location

The first term describes the number of locations along a bone where osseous manipulations are occurring. For example, if there is simple compression (or distraction)

at only one level, the procedure is referred to as "monolocal." However, if at one level a segmental defect is being closed while at a second location within the same bone a corticotomy site is being distracted, the strategy is referred to as "bilocal." Likewise, if two corticotomy sites within a bone are being distracted while a skeletal defect between them is being closed, the technique is referred to as "polylocal."

Sequence

The second term in the treatment protocol describes the sequence of maneuvers. Thus, therapy can be either "simultaneous" (when different actions are occurring at the same time) or "consecutive" (when one action precedes a second).

Action

The third term defines the actual maneuver (or maneuvers) used to effect the reconstruction. In most cases involving movement of bone fragments, this action may be either "compression," "distraction," "compression-distraction," and "distraction-compression" or (when correcting deformities) "simple opening wedge," "distractional wedge," and "translational wedge." As a rule, the first term describes the first action is a sequence.

Objective

The last term in the description refers to the goal of therapy. Hence, repair of a fracture or non-union would be called "osteosynthesis," while limb elongation would be referred to as a "lengthening." Limb elongation by traction on a child's growth plate is called "traction epiphysiolysis." Obviously, obliteration of a growth plate by external compression (to treat, e.g., hemihypertrophy) would be "compression epiphysiodesis."

Summary

In a situation where a simple transverse hypertrophic non-union is compressed in an external skeletal fixator to promote union, the Ilizarov terminology for this treatment strategy would be "monolocal compression osteosynthesis." A limb lengthening through a single level (without deformity correction) is called "monolocal distraction lengthening."

When closing a skeletal defect in a bone by transporting an osseous segment through the limb (after performing a corticotomy elsewhere in the bone), the strategy is called "bilocal consecutive distraction-compression osteosynthesis" since the corticotomy site is distracted *before* the defect is compressed. In some situations, a

non-union site is compressed (shortening a limb slightly) at the same time that limb length is restored through a corticotomy elsewhere in the bone. This strategy is called "bilocal simultaneous compression-distraction osteosynthesis." In oblique hypertrophic non-unions associated with shortening, a single location may be compressed for 2 weeks and then distracted to regain length (new bone forms in the non-union site); such a regimen is called "monolocal consecutive compression-distraction osteosynthesis."

Certain pathologic bone diseases—such as diffuse chronic osteomyelitis—can be cured (according to Ilizarov) by performing an oblique S-shaped osteotomy through the region, followed by gradual distraction (after the usual latency interval). The new bone, which forms within the distraction gap, can serve as a highly vascularized cancellous bone graft. Since the limb might end up too long if left in the elongated position, the distraction is stopped and the osteotomy gap gradually compressed until the original limb length is restored. This procedure squeezes the newly formed bone into the microabscesses of the osteomyelitic bone. Such a sequence would be referred to as "monolocal consecutive distraction-compression osteosynthesis."

Conclusion

Surgeons using Ilizarov's method of treatment—or any modification, for that matter, including intramedullary lengthening—will find it convenient to use Ilizarov's terminology to express the location, sequence, action, and objectives of a treatment plan employing external skeletal fixation and the movement of bone fragment.

Applications of the Ilizarov Method

The list of applications of Ilizarov's method has grown considerably since its inception. It now includes:

- Limb lengthening and deformity correction
- Percutaneous treatment of all closed metaphyseal and diaphyseal fractures as well as many epiphyseal fractures
- Repair of extensive defects of bone, nerve, vessel, and soft tissues without the need for grafting and in one operative stage
- Bone thickening for cosmetic and functional reasons
- The percutaneous one-stage treatment of congenital or traumatic pseudarthroses
- Limb lengthening to treat growth retardation through distraction epiphysiolysis or other methods
- The correction of long bone and joint deformities, including resistant and relapsed clubfoot
- The percutaneous elimination of joint contractures
- The treatment of various arthroses through osteotomy and repositioning of the articular surfaces

- Percutaneous joint arthrodesis
- Elongating arthrodesis—a method of fusing major joints without concomitant limb shortening
- The filling in of solitary bone cysts and other such lesions
- The treatment of septic non-unions by the favorable effect of stimulated bone healing on infected bone
- The filling in of osteomyelitis cavities by the gradual collapse of one of the cavity walls
- The lengthening of amputation stumps
- Management of hypoplasia of the mandible and similar conditions
- The ability to overcome certain occlusive vascular diseases without bypass grafting
- Correction of achondroplasia and other forms of dwarfism

References

1. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part I. The influence of stability of fixation and soft-tissue preservation. Clin Orthop Rel Res. 1989;238:249.
2. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Rel Res. 1989;239:263.
3. Ilizarov GA. Some theoretical and clinical aspects of transosseous osteosynthesis. In: Ilizarov GA, editor. In: Abstracts of second international symposium on experimental, theoretical and clinical aspects of transosseous osteosynthesis. Kurgan: USSR; 1986.
4. Ilizarov GA. Transosseous osteosynthesis. Heidelberg: Springer-Verlag; 1991.
5. Aronson J, Good B, Stewart C, Harrison B, Harp J. Preliminary studies of mineralization during distraction osteogenesis. Clin Orthop Rel Res. 1990;250:43–9.
6. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263–85.

Modifications of the Ilizarov Apparatus

3

Half-Pin Mounting

It did not take long for surgeons around the world to realize that the Ilizarov apparatus (consisting of circular rings and tensioned transosseous wires) was not absolutely essential to the biology of distraction osteogenesis. New bone could form in a widening distraction gap in an appropriate biomechanical environment regardless of the method used to establish stability. The first change, therefore, was to substitute threaded half-pins for the tensioned transcutaneous wires, utilizing a standard Ilizarov apparatus [1, 2]. This measure reduced muscle impalement, particularly for the tibia and ulna, because these bones have subcutaneous surfaces ideally suited for half-pin transosseous fixation (Fig. 3.1).

Computer-Controlled Hexapod Fixators

Once surgeons became familiar with the Ilizarov apparatus, they sought ways to simplify the mounting configuration required for complex deformity correction. The hexapod concept, borrowed from flight simulators and other industrial applications, proved popular in this respect because it allowed repositioning of bone fragments in a three-dimensional space by adjusting the lengths of six struts. (With the classic Ilizarov assembly, surgeons had to create separate mechanisms to correct angulation, translation, and rotation deformities.) By applying the hexapod frame, surgeons could correct all planes of deformity with a general frame configuration and computer-generated correction formulation, based on initial mounting and deformity parameters.

Charles Taylor, an orthopedic surgeon in Memphis, Tennessee, created a particularly popular and easy to use device, the Taylor Spatial Frame®. This fixator combined the hexapod system with an attractive computer interface that, once mastered, greatly simplified limb lengthening and deformity correction [3, 4] (Fig. 3.2).

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_3

Fig. 3.1 The Rancho mounting system, incorporating half-pins in place of wires in an otherwise conventional Ilizarov apparatus. Copyright 2016 NuVasive

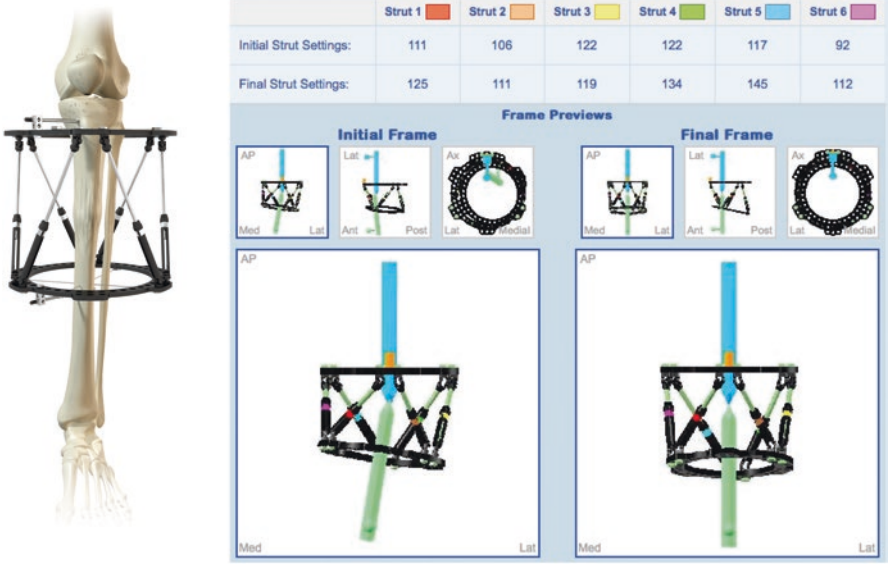


Fig. 3.2 The Taylor Spatial Frame® and corresponding computer interface. Copyright 2016 NuVasive

Monolateral External Fixation

Some surgeons eliminated the circular fixator altogether and used monolateral external devices to secure the bone fragments. DeBastiani (Verona, Italy), for example, used his Orthofix® fracture reduction and stabilization device for limb lengthening after learning about distraction osteogenesis from Italian colleagues who travelled to Siberia, USSR, to learn Ilizarov's methods [5]. As with half-pin mounting strategies, new bone formed in the widening distraction gap. However, some surgeons recognized that the quality and speed of maturation of the regenerate did not quite match what was routinely created with a classic Ilizarov mounting (Fig. 3.3).

All of the aforementioned techniques, however, suffered from the unfavorable features associated with all external skeletal fixators: pin and wire site inflammation and infections, muscle impalement, pain with activities, and the obvious social and functional handicap that is created by a large metallic device affixed to a person's limb.

Fig. 3.3 The Orthofix® monolateral fixator modified for limb lengthening. Copyright 2016 NuVasive

Strategies to Shorten Time in Fixation

Lengthening Over a Nail

With any external skeletal fixator, the rate of pin and wire tract infections increases slowly over the first 150 days and then increases more rapidly thereafter [6]. Therefore, when external fixators used for trauma applications, early bone grafting and other strategies to reduce time in external fixation have evolved. When such frames are used for limb lengthening, however, longer time in fixation is an inevitable consequence of the nature of new bone formation and maturation in the distraction gap.

For this reason, surgeons have developed strategies to shorten fixator time by using intramedullary nails to stabilize the limb and prevent bending or fracture through the regenerate after early frame removal—ideally as soon as the distraction phase of the procedures completed. There are a few approaches available. The earliest, lengthening over a nail (LON), was developed by Paley et al. [7]. He inserts an intramedullary nail at the time for fixator application but secures only the proximal end of that implant to the bone with locking screws. The frame is secured to the same bone with transcutaneous pins and wires, with care taken to place these transosseous pins and wires in locations that do not contact the intramedullary nail.

Typically, this means inserting pins and wires posterior or anterior to the path of the implant with enough bone between the pin or wire and nail to prevent microbes in the always contaminated pinhole from entering the nail pathway.

Likewise, strategic placement of the transcutaneous implants must be arranged in a configuration that allows safe surgical implantation of the transverse locking screws at the end of the distraction phase without contaminating the transverse locking screw entry point.

In spite of these precautions, there exists a risk of approximately 10–15% contamination of the intramedullary nail with microbes from the pin or wire tracks, a complication that usually requires a secondary operation with an antibiotic-impregnated cement-coated intramedullary rod. Nevertheless, the technique has proven popular around the world.

Lengthening and Then Nailing

One of the drawbacks of the LON technique is that the presence of the intramedullary nail during the limb lengthening part of the procedure means that deformity correction using classic Ilizarov methods is generally limited to either axial rotation corrections around the nail or small corrections that can be accomplished at the time of the nail and fixator are applied.

To get around this limitation, Rozbruch and colleagues, to shorten time in external fixation, perform a standard lengthening and deformity correction with a circular external fixator (or even a monolateral external fixator) and then insert the nail when the deformity correction is complete, at the beginning of the consolidation

phase [8]. This allows removal of the frame as soon as the limb is a stabilized by the nail. As with the LON technique, the LTN strategy requires advanced planning for pin and wire placement, lest one or more transcutaneous implants interferes with nail insertion.

Obviously, one would not insert a nail in a patient wearing an external skeletal fixator secured to the same bone if there was any evidence of active pin site infection. Nevertheless, subclinical contamination is always present in every pinhole, so the risk of intramedullary osteomyelitis accompanying nail insertion must be taken into consideration.

Lengthening and Then Plating

Alternatively, the Rozbruch group will occasionally apply plate to the surface of the bone if a nail does not fit properly or provide a sufficient stability at the time of fixator removal, before the bone has consolidated [9]. This technique requires even greater strategic preoperative planning than lengthening and then nailing because one requires a flat surface of the bone, free of transcutaneous implant holes, to secure the plate without risk of infection. The technique is not widely used for this reason.

References

1. Green SA. Ilizarov method: Rancho technique. Orthop Clin N Am. 1991;22(4):677–789.
2. Green SA, Harris NL, Wall DM, Ishkanian J, Marinow H. The Rancho mounting technique for the Ilizarov method: a preliminary report. Clin Orthop Rel Res. 1992;280:104–16.
3. Taylor JC. Ring size selection. Tech Orthop. 1990;5:13–8.
4. Taylor JC. Geometry of hinge placement. Tech Orthop. 1990;5:33–41.
5. DeBastiani G, Aldegheri R, Renzi-Brivio L, Trivelli G. Limb lengthening by callus distraction (callotasis). J Pediatr Orthop. 1987;7:129–34.
6. Burny F, Elbana S, Evrard M, Van der Ghinst M, Degeeter L, Peeters F, et al. Les Fractures Simples du Tibia, Traitement par Fixation Externe Elastique. In: Vidal J, editor. In: Proceedings of the 7th international conference on Hoffmann external fixation. Geneva: Diffinco; 1979.
7. Paley D, Herzenberg JE, Paremain G, Bhave A. Femoral lengthening over an intramedullary nail. A matched-case comparison with Ilizarov femoral lengthening. J Bone Joint Surg Am. 1997;79(10):1464–80.
8. Rozbruch SR, Kleinman D, Fragomen AT, Ilizarov S. Limb lengthening and then insertion of an intramedullary nail: a case-matched comparison. Clin Orthop Relat Res. 2008;466(12):2923–32.
9. Harbacheuski R, Fragomen AT, Rozbruch SR. Does lengthening and then plating (LAP) shorten duration of external fixation? Clin Orthop Relat Res. 2012;470(6):1771–81.

Intramedullary Lengthening Devices

4

Background

Elimination of the External Fixator

It has long been the goal of reconstructive surgeons to eliminate the external fixator altogether and use some method of internal fixation to achieve distraction osteogenesis. The transcutaneous implants used to secure the frame to the limb create portals of entry for bacteria into deeper tissues and bone. This can cause not only pain and inflammation but also risks prolonged or even permanent infection of the bone or surrounding soft tissues. Ilizarov, however, was generally opposed to such notions because he always contended that new bone formation depended upon both periosteal and endosteal blood supply, that is, blood entering the cortical bone from its exterior surface as well as its marrow blood vessels.

Research by Delloye et al. [1], however, showed that the periosteal blood supply was more important for distraction osteogenesis than was preservation of bone marrow vascularity. He did so by filling the marrow with absorbable bone wax in some of their experimental animals but not in others. Bone formed during distraction, provided that the proper conditions of stability, latency interval, and rate and rhythm of distraction were observed. This research, more than any other, confirmed that surgeons could employ an elongating intramedullary nail to create new bone in a widening distraction gap.

The basic principles of the Ilizarov method, however, must be maintained. Indeed, since reaming and nail insertion destroys the endosteal blood supply, careful preservation of the periosteal blood supply (by gentle elevation before osteotomy) is even more critical during intramedullary lengthening than with external fixation lengthening.

However, performing what Ilizarov and his co-workers call a "sparing corticotomy" is not necessary with IM lengthening. During this type of osteotomy, the tip of the chisel or osteotome stays within the cortex and avoids transecting the endosteal blood vessels. It's technically challenging to do so because the far cortex cannot

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_4

be reached by the surgeon's sparing chisel and must therefore be cracked by counterrotation of the fragments or by prying apart the bone through the accessible cortices. Because reaming destroys the marrow vessels, protecting them is not necessary with IM lengthening techniques.

Ratcheting Devices

Bliskunov Intramedullary Nail

The first intramedullary fixation devices used for distraction osteogenesis were all driven by mechanical ratchet systems. The Bliskunov® intramedullary nail fit in the femur like a normal trauma nail but had an extension at the top end connected via a universal joint to a rod that bolted to the pelvis [2]. Rotating the hip in relation to the pelvis twisted the rod, which turned a mechanism within the nail, elongating it (Fig. 4.1).

Fig. 4.1 The Bliskunov® intramedullary nail. Note the connection of the ratcheting mechanism to the pelvis. Copyright 2016 NuVasive

Fig. 4.2 The Albizzia® intramedullary nail. Copyright 2016 NuVasive

The fundamental problem with this implant was that it limits hip mobility while in place. It is still used in Eastern Europe as of this writing.

Albizzia Intramedullary Nail

The Albizzia® intramedullary nail possessed a similar elongating ratchet mechanism but eliminated the connection to the pelvis [3]. The nail resembles the telescoping leg of a camera tripod. Elongation occurs by counterrotating one end of the bone, which rotates the inner telescopic rod in relation to the outer cylinder, while both elements of the device are locked in their respective fragments of the bone (Fig. 4.2).

The basic problem with this device, which appears to successfully elongate the bone during the initial phases of distraction, is that gradual stiffening of the newly formed regenerate bone in the distraction gap makes counterrotation of the

fragments increasingly more difficult. For this reason, some patients must be taken back to the operating room to crack apart the regenerate bone forming in the distraction gap so that elongation can proceed.

Internal Skeletal Kinetic Distractor

The ISKD® (Internal Skeletal Kinetic Distractor) was developed by Dean Cole M.D. of Orlando, Florida [4]. As with the other mechanical limb lengthening devices, a ratchet mechanism elongates the telescopic nail. Unlike the Albizzia nail, which requires 30 degrees of counterrotation to engage the racket mechanism, the ISKD requires only a few degrees of rotation to elongate the device. In ordinary walking, planting the foot on the ground provides sufficient rotation power to lengthen the nail (Fig. 4.3). Indeed, this has proven to be a problem with the device, which can lengthen too rapidly if the patient does too much walking [5]. Excessive elongation can occur between clinic visits, even if they are a week apart.

Fig. 4.3 The ISKD® intramedullary nail. Copyright 2016 NuVasive

In early 2015, the manufacturer Orthofix Inc. removed the ISKD® from the market although it may return.

Rotating Spindle Devices

The next category of the intramedullary lengthening nails includes those that are elongated by a rotating threaded spindle that gradually separates the telescopic portions of the nail, each interlocked into its respective fragment of the bone. Each of these devices requires an external power source that can transfer energy or motive power into the nail.

Fitbone® Intramedullary Nail

The Fitbone®, a German-made intramedullary lengthening nail, incorporates an electric motor to turn the spindle [6]. Rather than place batteries within the nail to power the motor, the device utilizes a subcutaneous antenna attached by wire to the motor (Fig. 4.4). This antenna generates electric current through induction by a

Fig. 4.4 The Fitbone® intramedullary nail and remote controller. An induction coil is placed on the skin over the corresponding subcutaneous induction coil connected to the nail's internal electric motor. Copyright 2016 NuVasive

corresponding electric current passing through a pad placed on the outside of the skin adjacent to the antenna. The current causes the motor to turn, rotating the spindle (Fig. 4.5).

The Fitbone® has been successfully employed to lengthen long bones in many musculoskeletal conditions, including post-trauma shortening, congenital anomalies, and short stature. At present, the motor turns in only one direction, allowing lengthening but no compression. The FDA has recently cleared the nail for marketing in the United States.

Fig. 4.5 (**a**) The Phenix intramedullary lengthening nail. Copyright 2016 NuVasive. (**b**) Handheld magnet used to power the nail

Phenix® Intramedullary Nail

The other category of threaded spindle devices consists of those motorized by an internal rotating magnet responding to an external magnetic field pressed against the body. The Phenix® nail, developed in France by Soubeiran, is just such a device [7].

The external component is a handheld solid magnet. Simply moving the magnet circumferentially around the limb rotates the internal magnet. The device has been in development for many years and has not yet been cleared for marketing in the United States by the FDA (Fig. 4.5).

PRECICE® Intramedullary Nail

The most recent threaded spindle intramedullary lengthening implant, and the only one that is cleared for marketing in the United States, is the PRECICE® intramedullary lengthening nail, manufactured by NuVasive Specialized Orthopedics (formerly Ellipse Technologies). A small magnet in the nail rotates in response to two rotating external rare earth magnets. (The internal magnet can rotate either clockwise or counterclockwise, thereby lengthening or compressing the device.) The two external magnets are housed in a computer-controlled, electrically powered assembly. The external magnets, however, are not electromagnets; instead they are composed of rare earth elements and are thus always "on" (Fig. 4.6).

The PRECICE® intramedullary nail and external remote controller (ERC) are cleared for use by the FDA for bone lengthening. Preliminary reports describing small series of patients treated with the PRECICE® nail have confirmed its reliability and accuracy [8] (Figs. 4.7 and 4.8).

Transferring Energy Through Soft Tissues

All rotating spindle of intramedullary lengthening nails currently available depends on external energy sources to power the internal rotor. None have internal batteries. As a result, just as the light from a bulb decreases in intensity proportional to the square of its distance from the source, so too does electrical and magnetic energy diminish with distance. This limits the possible distance from the source to the receiver. The Fitbone® nail employs a subcutaneous induction coil as a receiver antenna, with the transmitter being another coil pressed against the skin opposite the internal one. Wires connect the internal coil to the motor in the nail. Placing the antenna too deeply in the tissues would inhibit its function. The maximum distance between the two antennae should not exceed 1.0 cm (Fig. 4.9).

The same consideration applies to the PRECICE® and Phenix® implants, both depending on an internal magnet rotating in response to an external magnet field. The interval between the exterior power source and the internal rotating magnet contains not only skin and subcutaneous fat as with the Fitbone® antenna but also

Fig. 4.6 The PRECICE® intramedullary nail in the femur and tibia. Copyright 2016 NuVasive

muscle and cortical bone. None of these tissues interfere with the transmission of magnetic flux, but all of them occupy space, pushing the external magnet way from the device.

With the large diameter PRECICE® nail, the magnets in the external remote controller (ERC) must be about 50 mm (~2.0 in.) from internal magnet to power it predictably. In the lower leg, the tibia is an anterior subcutaneous bone, so the ERC magnets, when placed on the front of the limb, are always reasonably close to the nail. The femur, however, is a deeply seated bone, surrounded by bulky muscles. There will thus be some individuals whose thigh circumferences are too great to permit use of the implant.

Fig. 4.7 (*Right*) The external remote controller (ERC) powers the PRECICE® intramedullary nail. The surgeon programs the device with a lengthening prescription. (*Left*) The ERC contains two rare earth magnets to, when rotated, cause the small magnet within the nail to turn, elongating the nail via three 1:4 planetary gears. Copyright 2016 NuVasive

Fig. 4.8 A patient positioning the ERC over the magnet inside the nail. Marking the limb during surgery helps with location. Future ERCs will automatically use the internal magnet's field to aid location. Copyright 2016 NuVasive

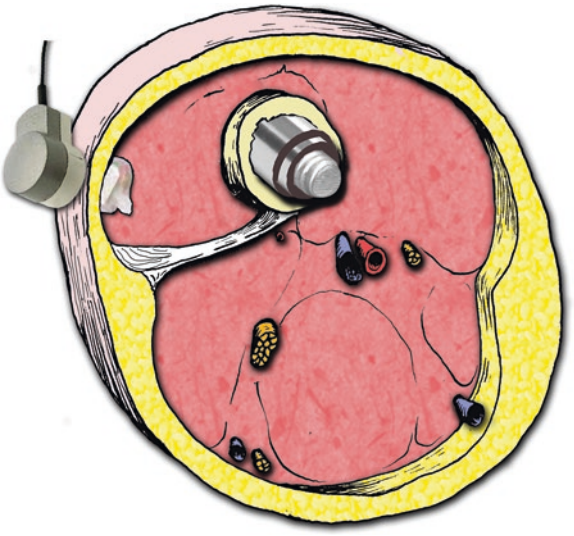


Fig. 4.9 With the Fitbone nail, the transmitting and receiving antennae should be no more than 1.0 cm apart. The receiving antenna is usually placed in the subcutaneous fat, but in thin patients with little subcutaneous fat, the antenna is placed beneath the circular investing fascia. Copyright 2016 NuVasive

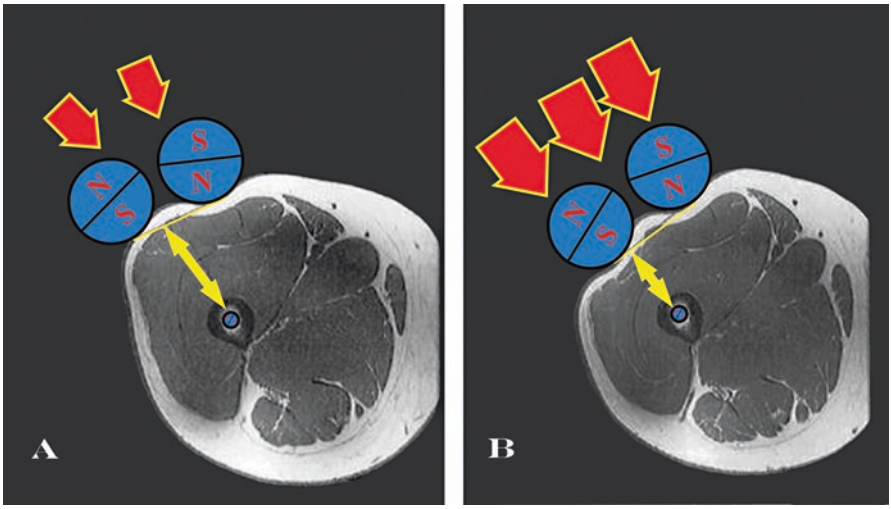


Fig. 4.10 (**a**) With bulky patients, the magnet within the nail may be too far from the external magnets to function properly. (**b**) Pressing downward on the ERC may bring it within range of the implant. Copyright 2016 NuVasive

Fortunately, however, most of the adult population can be successfully treated with the nail, especially if the external device is pressed firmly into the flesh in bulky individuals (Fig. 4.10).

Correcting Deformities with Intramedullary Lengthening Nails

At present, the ability to make substantial deformity corrections with lengthening intramedullary nails is rather limited. Too much manipulation of the osteotomy site risks retarding regenerate bone formation in the distraction zone—a region whose intramedullary blood supply has already been compromised by reaming for the nail. Simple corrections of rotational malalignment with the nail in place are appealing, but excessive rotation at the osteotomy site, beyond 15–20°, may slow maturation of new bone formation especially in the tibia.

Correction of angulation at the distraction zone should also be restricted, with knowledgeable surgeons limiting angular correction to 10–15° in the femur and 5–10° in the tibia.

In some situations, it may be possible to correct a deformity at one end of a bone while simultaneously lengthening the same bone at the other end. In this way, a wedge resection corrective osteotomy, which always shortens a bone, can be neutralized by concomitant elongation. However, the periosteal elevation required to expose the bone, remove a wedge, and apply a plate, when combined with reaming for an intramedullary nail, will devitalize the bone between the nail and the plate, leading to delayed union or non-union. Therefore, these maneuvers should be performed at opposite ends of the bone. Future developments might change this perspective.

The subject of deformity correction requires an understanding of deformities in three-dimensional space, a topic best covered in Paley's monograph "Principles of Deformity Correction" [9].

Achondroplasia and Related Conditions

The discovery by Prof. G. A. Ilizarov that distraction osteogenesis would result in predictable bone formation in a widening distraction gap resulted in the development of a program at his institution in Siberia to treat achondroplasia and similar dwarfing conditions. Indeed, since the clinical departments at his institute are organized by treatment strategies (rather than pathologies), achondroplasia is managed by surgeons in the "Department of Symmetrical Dwarfs."

Achondroplasia

Achondroplasia is the most common and easily recognized dwarfing condition. It is characterized by rhizomelic shortening, meaning that proximal segments are relatively shorter than the distal segments. Thus, the humeri are proportionally shorter than the forearm bones, and the femora are proportionally shorter than the tibia and fibula. Even within the hands and feet, the shortening is more pronounced in the metacarpals and metatarsals than in the phalanges, with the tips of the fingers and toes nearest to normal length.

Likewise, there is a disproportionate relationship between the way bones of the skull and face enlarge during normal childhood growth. This results in a proportionately overlarge forehead and apparent constriction of the central part of the face. Thus, all individuals afflicted with achondroplasia have a similar facial appearance.

Achondroplastic patients typically have a number of orthopedic maladies superimposed on their short stature. These include hyperlordosis of the lumbar spine, increased kyphosis of the thoracic spine, bowleggedness (genu varum), and a hypoplastic odontoid process of the second cervical vertebrae in the neck.

This latter abnormality can cause sudden death on the operating table if an anesthetized patient's head is not handled properly [10, 11].

In 1994, researchers discovered the abnormality that causes achondroplasia. There is a defect in the gene that controls fibroblast growth factor receptor three (fgfr3) [12], which results in a profound abnormality in the growth of bones via enchondral ossification, the process whereby new bone is first created in a cartilaginous growth plate, which turns into bone as the growth plate elongates [13].

The genetic defect often appears as a spontaneous mutation, with parents of normal stature having a child afflicted by achondroplasia. Mutation rate is about one per 100,000. An older male parent appears to be a risk factor for the spontaneous mutation to achondroplasia. Once the achondroplasia gene appears, it is dominant. Thus, mating a person with achondroplasia to a person of normal stature will result in the 50% of the children having achondroplasia, and the other 50% being of normal stature.

If two individuals with achondroplasia mate and have children, 25% of the children will be of normal stature, 50% of the children will be achondroplastic, and 25% of the children will have two genes for the mutant growth factor receptor, a situation incompatible with life.

Intelligence is not adversely affected by the condition.

Certain domestic animals have been bred to become achondroplastic, including dachshunds, basset hounds, bulldogs, and specific breeds of sheep and pigs. The genetic mutation in achondroplastic animals, however, appears to be different from that in achondroplastic humans.

Other conditions generally grouped in the same limb lengthening category as achondroplasia include hypochondroplasia, Turner's syndrome, and constitutional short stature.

Hypochondroplasia

Hypochondroplasia is caused by the same genetic disorder as achondroplasia, but there was less penetrance of the gene [14]. As a result, hypochondroplastic children appeared normal at birth, but the condition becomes obvious over time. Moreover, the amount of dwarfing is not as extreme as in achondroplasia, and, unlike achondroplasia, there may be some stature improvement with human growth hormone [15].

Turner's Syndrome

Turner's syndrome appears when one of the X chromosomes is deficient or absent. Thus, it only occurs in females. Afflicted individuals are short in stature and have web necks, low set ears, and swollen hands and feet [16]. Moreover, there is deficient development of female sexual characteristics. As result, Turner's syndrome patients are not fertile. The condition does not appear to be hereditary but strikes as a spontaneous mutation [17].

Constitutional Short Stature

Constitutional short stature is just with the name implies, a very short individual (2.5 standard deviations from the mean height for their sex [18]) who was otherwise normally proportioned (hence, not dwarfs) and is not suffering from an endocrine or developmental disorder. The condition may be either hereditary or idiopathic [19].

Furthermore, some individuals who appear to have constitutional short stature actually demonstrate a delayed growth spurt and may end up near the 50th percentile of height. Usually, parents of a child who appears to be abnormally short when compared to other family members take the youngster to a pediatric endocrinologist to make sure there are no hormonal causes for the shortness. Likewise, they will inquire about the use of hormones to stimulate growth. Finally, as such individuals reach the end of their growth cycle, they may be referred for stature surgery.

Stature Surgery in Dwarfing Conditions

The matter of providing limb lengthening services to an individual of short stature is a somewhat controversial, although centers for treating individuals with achondroplasia exist in nearly every advanced country on earth. At many facilities, part of such treatment includes elective stature surgery.

In the United States, some indemnity healthcare insurance plans specifically preclude, in their policies, stature surgery in achondroplasia, but they do not decline to pay for deformity correction in the same category of patients.

Likewise, Little People of America, Inc., a support group for families of patients with short stature conditions, neither supports nor condemns limb lengthening surgery for their members. However, they quite clearly state on their website that they consider stature surgery to be a cosmetic choice, rather than an orthopedic necessity. They point out the hazards limb elongation operations and recommend that families interested in such surgery consult with experts at centers where such operations are routinely done.

During the era when external fixators were used for limb lengthening, the pain, pin tract infections, and other issues associated with frames dominated patient and surgeon concerns. As we enter a new epoch with internal lengthening devices, demand for stature surgery in dwarfing conditions will likely increase. While the

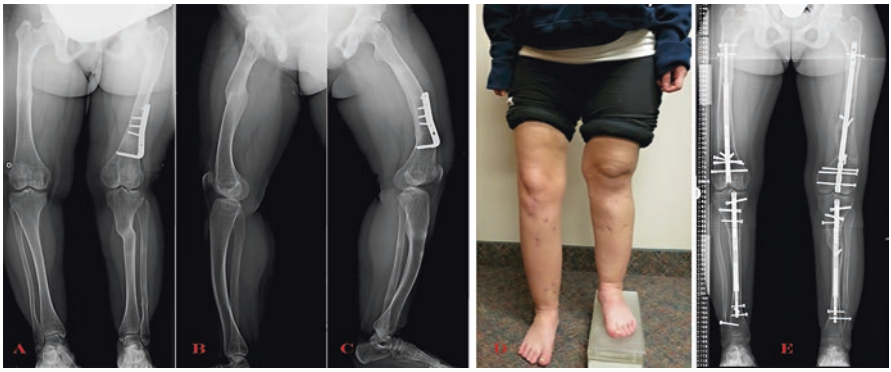


Fig. 4.11 Bilateral four-segment intramedullary lengthening for vitamin D-resistant rickets. (**a**) Pre-op standing AP demonstrating varus right knee and valgus left knee deformity. (**b**) Pre-op lateral view, right lower extremity. (**c**) Pre-op lateral view, left lower extremity. (**d**) Pre-op clinical photograph. (**e**) Post-op standing AP demonstrating consolidation of all segments

process will certainly be more comfortable for patients, most of the serious risks associated with limb lengthening will continue to haunt the procedures.

Surgeons who routinely perform limb lengthening surgeries in achondroplastic patients as well as others afflicted by short stature have a sense that the soft tissues in achondroplasia individuals are naturally more elongated than the bones, as though they are waiting to be stretched out to length. Sure enough, when compared to Turner's syndrome and constitutional short stature, patients within the achondroplasia/hypochondroplasia spectrum have a fewer complications, in spite of the fact that they are subjected to greater lengthenings [20].

Clinical Illustration: Four-Segment Intramedullary Lengthening

This case involves a 22-year-old female with short stature caused by vitamin D-resistant rickets who had already been subjected to multiple childhood osteotomies. Her principle complaint was right medial and left lateral knee pain. She requested deformity correction and limb lengthening (Fig. 4.11).

Technical Considerations

An important technical issue associated with the use of internal lengthening nails involves the size of the implant. When external fixators were used to treat dwarfing conditions, the rings could start out quite close together and gradually be pulled apart by the distraction mechanism. At Ilizarov's center and elsewhere, two levels of osteotomy in the same bone were often used to speed up the entire process [21]. As the limb elongated, it was a simple matter to exchange the longitudinal struts in the construct to allow for progressive lengthening.

With an internal lengthening nail, the initial shortness of the bone limits the size of the implant that can be inserted. A short implant results in a short stroke length for the inner telescopic rod, so not much elongation is obtained with the first procedure. As the bone gets longer, larger implants can be inserted, which improves the situation. Nevertheless, since many insurance companies consider stature surgery in dwarfing conditions to be cosmetic procedures, the cost of so many implants early on may prove prohibitive.

A possible workaround for the situation would be to gain the initial length with the first implant and allow the bone to fully consolidate. After a couple of years, the surgeon could remove the transverse locking screws and the nail and reverse the telescopic portion of the nail back to zero (if the implant has that capacity). Thereafter, a new osteotomy is performed, the device reinserted, and the transverse locking screws repositioned.

While the FDA has not yet approved such a reuse protocol, motorized internal implants are now being introduced for compression osteosynthesis, so the concept of employing of the same implant in both of the distraction and compression modes will not seem particularly unreasonable in the near future. The only question in the minds of regulatory agencies will be the durability and safety of an implant that is left quiescent in the marrow canal for years at a time, to be periodically reactivated for implant shortening and then lengthening.

Except for the shortness of the implant, the operative techniques and protocols, as well as postoperative management principles outlined elsewhere in this book, are applied to individuals with symmetrical short stature conditions in the same manner used for limb length equalization.

Uses of Intramedullary Lengthening Nails

Intramedullary bone lengthening is becoming increasingly popular among orthopedic surgeons for treating a variety of musculoskeletal conditions because elongation of the bone can be accomplished without the application of an external skeletal fixator. Here are some common indications:

- To lengthen bones that are short as a result of congenital, developmental, acquired, or posttraumatic conditions
- Stature surgery for cosmetic reasons
- Equalizing limb lengths after inadvertent overlengthening during total hip replacement (the opposite side is lengthened)
- Lengthening with concomitant deformity correction (small angles only at present)

In the future, intramedullary lengthening devices will likely be used for:

- Compression osteosynthesis of non-unions
- Compression of fresh fracture fragments of the long bones

Bone transport—a method of overcoming gaps in long bones using Ilizarov's distraction osteosynthesis techniques—may, in the near future, be accomplished with internal fixation devices, perhaps in combination with other kinds of internal fixation devices.

A likely limitation in this regard is that many injuries that result in skeletal defects occur as a consequence of open fractures, often followed by bone infections. In these situations, the use of an intramedullary implant may be limited.

Internal lengthening bone plates, still in development, hold the promise of limb elongation without the need to cross a growth plate in a child, as occurs with intramedullary devices.

References

1. Delloye C, Delefortrie G, Coutelier L, Vincent A. Bone regenerate formation in cortical bone during distraction lengthening; an experimental study. Clin Orthop Rel Res. 1990;250:34.
2. Bliskunov AI. Intramedullary distraction of the femur (preliminary report). Ortop Travmatol Protez. 1983;10:59–62.
3. Guichet JM. Leg lengthening and correction of deformity using the femoral Albizzia nail. Der Orthopade. 1999;28(12):1066–77.
4. Cole JD, Justin D, Kasparis T, DeVlught D, Knobloch C. The intramedullary skeletal kinetic distractor (ISKD): first clinical results of a new intramedullary nail for lengthening of the femur and tibia. Injury. 2001;32(Suppl 4):SD129–39.
5. Mahboubian S, Seah M, Fragomen AT, Rozbruch SR. Femoral lengthening with lengthening over a nail has fewer complications than intramedullary skeletal kinetic distraction. Clin Orthop Relat Res. 2012;470(4):1221–31.
6. Baumgart R, Zeiler C, Kettler M, Weiss S, Schweiberer L. Fully implantable intramedullary distraction nail in shortening deformity and bone defects. Spectrum of indications. Der Orthopade. 1999;28(12):1058–65.
7. Thailer PH, Furmetz J, Wolf F, Eilers T, Mutschler W. Limb lengthening with fully implantable magnetically actuated mechanical nail (Phenix)— preliminary results. Injury. 2014;45(S1):S60–5.
8. Kirane YM, Fragomen AT, Rozbruch SR. Precision of the PRECICE internal bone lengthening nail. Clin Orthop Relat Res. 2014;472(12):3869–78.
9. Paley D. Principles of Deformity Correction. Berlin: Springer; 2002.
10. Kaushal A, Haldar R, Ambesh P. Anesthesia for an achondroplastic individual with coexisting atlantoaxial dislocation. Anesth Essays Res. 2015;9(3):443–6.
11. Rahimizadeh A, Soufiani HF, Hassani V, Rahimizadeh A. Atlantoaxial subluxation due to an Os odontoideum in an achondroplastic adult: report of a case and review of the literature. Case Rep Orthop. 2015;2015:142586.
12. Narayana J, Horton WA. FGFR3 biology and skeletal disease. Connect Tissue Res. 2015;56(6):427–33.
13. Richette P, Bardin T, Stheneur C. Achondroplasia: from genotype to phenotype. Joint Bone Spine: revue du rhumatisme. 2008;75(2):125–30.
14. Rousseau F, Bonaventure J, Legeai-Mallet L, Schmidt H, Weissenbach J, Maroteaux P, et al. Clinical and genetic heterogeneity of hypochondroplasia. J Med Genet. 1996;33(9):749–52.
15. Massart F, Miccoli M, Baggiani A, Bertelloni S. Height outcome of short children with hypochondroplasia after recombinant human growth hormone treatment: a meta-analysis. Pharmacogenomics. 2015;16(17):1965–73.
16. Granger A, Zurada A, Zurada-Zielinska A, Gielecki J, Loukas M. Anatomy of turner syndrome. Clin Anat. 2016;29(5):638–42.

17. Gicquel C, Cabrol S, Schneid H, Girard F, Le Bouc Y. Molecular diagnosis of Turner's syndrome. J Med Genet. 1992;29(8):547–51.
18. Moore KC, Donaldson DL, Ideus PL, Gifford RA, Moore WV. Clinical diagnoses of children with extremely short stature and their response to growth hormone. J Pediatr. 1993;122(5 Pt 1):687–92.
19. Lanes R, Lee PA, Plotnick LP, Kowarski AA, Migeon CJ. Are constitutional delay of growth and familial short stature different conditions? Clin Pediatr. 1980;19(1):31–3.
20. Kim SJ, Pierce W, Sabharwal S. The etiology of short stature affects the clinical outcome of lower limb lengthening using external fixation. A systematic review of 18 trials involving 547 patients. Acta Orthop. 2014;85(2):181–6.
21. Burghardt RD, Yoshino K, Kashiwagi N, Yoshino S, Bhave A, Paley D, et al. Bilateral double level tibial lengthening in dwarfism. J Orthop. 2015;12(4):242–7.

Further Reading

1. Al-Sayyad MJ. Lower limb lengthening and deformity correction using the Fitbone motorized nail system in the adolescent patient. J Pediatr Orthop B. 2012;21(2):131–6.
2. Baumgart R. The reverse planning method for lengthening of the lower limb using a straight intramedullary nail with or without deformity correction. A new method. Oper Orthop Traumatol. 2009;21(2):221–33.
3. Baumgart R, Burklein D, Hinterwimmer S, Thaller P, Mutschler W. The management of leg-length discrepancy in Ollier's disease with a fully implantable lengthening nail. J Bone Joint Surg Br. 2005;87(7):1000–4.
4. Bouali H, Latrech H. Achondroplasia: current options and future perspective. Pediatr Endocrinol Rev.: PER. 2015;12(4):388–95.
5. Burghardt RD, Herzenberg JE, Specht SC, Paley D. Mechanical failure of the intramedullary skeletal kinetic distractor in limb lengthening. J Bone Joint Surg Br. 2011;93(5):639–43.
6. Cattaneo R, Villa A, Catagni M, Tentori L. Limb lengthening in achondroplasia by Ilizarov's method. Int Orthop. 1988;12(3):173–9.
7. Dincyurek H, Kocaoglu M, Eralp IL, Bilen FE, Dikmen G, Eren I. Functional results of lower extremity lengthening by motorized intramedullary nails. Acta Orthop Traumatol Turc. 2012;46(1):42–9.
8. Donaldson J, Aftab S, Bradish C. Achondroplasia and limb lengthening: Results in a UK cohort and review of the literature. J Orthop. 2015;12(1):31–4.
9. Karakoyun O, Kucukkaya M, Sokucu S. Intramedullary skeletal kinetic distractor in lower extremity lengthening. Acta Orthop Traumatol Turc. 2014;48(3):307–12.
10. Kenawey M, Krettek C, Liodakis E, Meller R, Hankemeier S. Insufficient bone regenerate after intramedullary femoral lengthening: risk factors and classification system. Clin Orthop Relat Res. 2011;469(1):264–73.
11. Kitoh H, Mishima K, Matsushita M, Nishida Y, Ishiguro N. Early and late fracture following extensive limb lengthening in patients with achondroplasia and hypochondroplasia. Bone Joint J. 2014;96-B(9):1269–73.
12. Kim SJ, Pierce W, Sabharwal S. The etiology of short stature affects the clinical outcome of lower limb lengthening using external fixation. A systematic review of 18 trials involving 547 patients. Acta Orthop. 2014;85(2):181–6.
13. Kim SJ, Agashe MV, Song SH, Choi HJ, Lee H, Song HR. Comparison between upper and lower limb lengthening in patients with achondroplasia: a retrospective study. J Bone Joint Surg Br. 2012;94(1):128–33.
14. Klag KA, Horton WA. Advances in treatment of achondroplasia and osteoarthritis. Hum Mol Genet. 2016;25(R1):R2–8.
15. Kocaoglu M, Bilen FE, Dikmen G, Balci HI, Eralp L. Simultaneous bilateral lengthening of femora and tibiae in achondroplastic patients. Acta Orthop Traumatol Turc. 2014;48(2):157–63.

16. Koczewski P, Shadi M. Surgical treatment of short stature of different etiology by the Ilizarov method. Pediatr Endocrinol Diabetes Metab. 2007;13(3):143–6.
17. Krakow D. Skeletal dysplasias. Clin Perinatol. 2015;42(2):301–19. viii
18. Krieg AH, Lenze U, Speth BM, Hasler CC. Intramedullary leg lengthening with a motorized nail. Acta Orthop. 2011;82(3):344–50.
19. Kucukkaya M, Karakoyun O, Sokucu S, Soydan R. Femoral lengthening and deformity correction using the Fitbone motorized lengthening nail. J Orthop Sci. 2015;20(1):149–54.
20. Lee DH, Ryu KJ, Song HR, Han SH. Complications of the intramedullary skeletal kinetic distractor (ISKD) in distraction osteogenesis. Clin Orthop Relat Res. 2014;472(12):3852–9.
21. Mahboubian S, Seah M, Fragomen AT, Rozbruch SR. Femoral lengthening with lengthening over a nail has fewer complications than intramedullary skeletal kinetic distraction. Clin Orthop Relat Res. 2012;470(4):1221–31.
22. Paley D. PRECICE intramedullary limb lengthening system. Expert Rev. Med Devices. 2015;12(3):231–49.
23. Park KW, Garcia RA, Rejuso CA, Choi JW, Song HR. Limb lengthening in patients with achondroplasia. Yonsei Med J. 2015;56(6):1656–62.
24. Peretti G, Memeo A, Paronzini A, Marzorati S. Staged lengthening in the prevention of dwarfism in achondroplastic children: a preliminary report. J Pediatr Orthop B. 1995;4(1):58–64.
25. Schiedel FM, Vogt B, Tretow HL, Schuhknecht B, Gosheger G, Horter MJ, et al. How precise is the PRECICE compared to the ISKD in intramedullary limb lengthening? Reliability and safety in 26 procedures. Acta Orthop. 2014;85(3):293–8.
26. Singh S, Lahiri A, Iqbal M. The results of limb lengthening by callus distraction using an extending intramedullary nail (Fitbone) in non-traumatic disorders. J Bone Joint Surg Br. 2006;88(7):938–42.
27. Singh S, Song HR, Venkatesh KP, Modi HN, Park MS, Jang KM, et al. Analysis of callus pattern of tibia lengthening in achondroplasia and a novel method of regeneration assessment using pixel values. Skelet Radiol. 2010;39(3):261–6.
28. Tandon A, Bhargava SK, Goel S, Bhatt S. Pseudoachondroplasia: a rare cause of rhizomelic dwarfism. Indian J Orthop. 2008;42(4):477–9.
29. Yasui N, Kawabata H, Kojimoto H, Ohno H, Matsuda S, Araki N, et al. Lengthening of the lower limbs in patients with achondroplasia and hypochondroplasia. Clin Orthop Relat Res. 1997;344(344):298–306.

Surgical Principles of Intramedullary Lengthening

5

General Principles

Similarity to Intramedullary Trauma Nails

An appealing aspect of intramedullary lengthening nails is that the surgical technique for insertion is readily familiar to orthopedic surgeons who use intramedullary nails to treat long bone fractures. Indeed, the portals of entry at the ends of long bones are the same for trauma and intramedullary lengthening nails. Likewise, familiar *contraindications* for certain locations—particularly, avoiding piriformis fossa entry in pediatric cases—apply to both trauma and intramedullary lengthening implants.

Nevertheless, there are distinct differences between the practical surgical use of trauma nails and intramedullary lengthening nails.

Differences from Intramedullary Trauma Nails

Unlike fracture cases, a bone to be lengthened with an intramedullary elongating nail is *intact* prior to commencing the operation. As a result, reaming the bone marrow cavity of an intact bone can potentially push marrow fat into the venous circulation ahead of the reamer. Subsequently, this marrow fat travels via the inferior vena cava into the heart and thereafter into the lungs, occluding the pulmonary circulation. This fat embolism can be fatal. A surprising number of patients have died on the operating table from fat emboli caused by reaming a closed medullary canal in surgical procedures for intramedullary shortening, as well as that in total hip and knee replacement.

Also, trauma nails, without internal mechanisms, are stronger than lengthening nails.

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_5

Guide Wire Issues

Trauma nails are *hollow*, whereas intramedullary lengthening nails are not. A hollow nail permits the use of a guide wire throughout the entire insertion process, thereby simplifying nail insertion once the fracture fragments have been reduced to near-anatomic position and the guide wire crosses the fracture line.

While guide wires are used with intramedullary lengthening nails during the initial bone reaming phases of the surgical procedure, a surgeon must remove the guide wire before inserting the nail. This may result in displacement of the transected adjacent bone ends, suddenly making a seemingly straightforward operation technically challenging. We will describe techniques to prevent loss of alignment during nail insertion.

Short or Long Nails

Another area of ongoing discourse among surgeons with experience in intramedullary bone lengthening deals with the question: What is better for intramedullary lengthening, a short nail or long nail? In brief, a short nail can be inserted without concern for the anterior bow of the femur, a matter discussed in more detail below. The short nail, used with either a proximal or distal femoral osteotomy (with antegrade or retrograde nail insertion, respectively), does not reach into the bone far enough to risk anterior cortex penetration where the bone curves away from a straight-line entry axis.

However, a very proximal or distal osteotomy, performed at or near the metaphyseal/diaphyseal junction, results in substantial space around the nail, enough to allow angulation during lengthening unless blocking screws are used.

A longer nail, on the other hand, permits osteotomy in a region where the nail fits fairly snugly in the bone, meaning that angular displacements are nearly impossible for purely mechanical reasons.

In view of the paucity of published experience with large numbers of intramedullary lengthening nails—after all, the technique is quite new—there is no consensus on this matter yet.

Preoperative Planning

Detailed analysis of the nature and cause of the limb length discrepancy is essential when contemplating operative limb lengthening. Full-length x-ray images, preferably with a lift under the short limb to level the pelvis, will help determine which bone or bones require lengthening (Fig. 5.1). Furthermore, accurate measurement of the width, cortical thickness, and medullary canal diameter of the involved bone is essential. A standardized marker, placed on the skin at the same distance from the x-ray plate as the bone, will permit computation to account for image magnification.

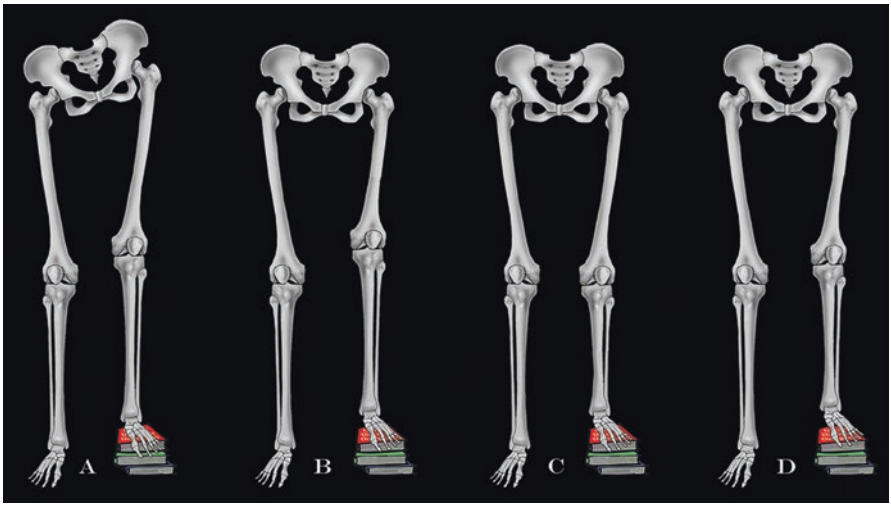


Fig. 5.1 Four cases demonstrating apparently identical shortening, requiring the same lift. (**a**) Supra-pelvic deformity. (**b**) Short femur. (**c**) Short tibia and fibula. (**d**) Shortening of both femur and tibiofibula. Copyright 2016 Zeeca Publishing Co

Careful examination of the patient should include assessment of the range of joint motion in the involved and contralateral limb. It is at this point that the surgeon must decide what surgical releases should accompany the operative lengthening. While it is possible to delay surgical releases in the hope that intensive physical therapy and a certain amount of patient stoicism will be sufficient to prevent contractures, subluxations, and dislocations, return trips to the operating room can be kept to a minimum by anticipating likely deformities and prophylactically releasing appropriate myofascial structures. This matter will be discussed in greater detail in the next chapter.

Assessing Deformities

With Ilizarov's circular external fixation method, limb lengthening is often combined with deformity correction because the hinges, translation rails, rotation constructs allow almost infinity spatial modification of bone fragments with respect to each other. Likewise, with circular hexapod fixators, the six oblique struts, used in conjunction with a properly configured computer program, will do the same.

Intramedullary lengthening nails presently available have none of these adjustable features, but they may be forthcoming as innovative surgeons and engineers come up with ingenious mechanisms to accomplish the seemingly impossible.

As mentioned elsewhere in this monograph, small angular and rotational corrections, perhaps up to 10 degrees of angulation and 20 degrees of rotation, are possible through the osteotomy site at the time of implant surgery. Greater changes risk

injury to the periosteum (specifically, strangulation of microcirculation) that generates the new bone in the widening distraction zone. For this reason, surgeons performing limb elongation surgery must be conversant in the language of deformity correction, which includes an understanding of normal anatomical joint and axis alignment values in both the coronal and sagittal plane, familiarity with methods of measuring both the anatomical and biomechanical axes of bones, and appreciating the difference between them (Figs. 5.2 and 5.3). Additionally, limb lengthening

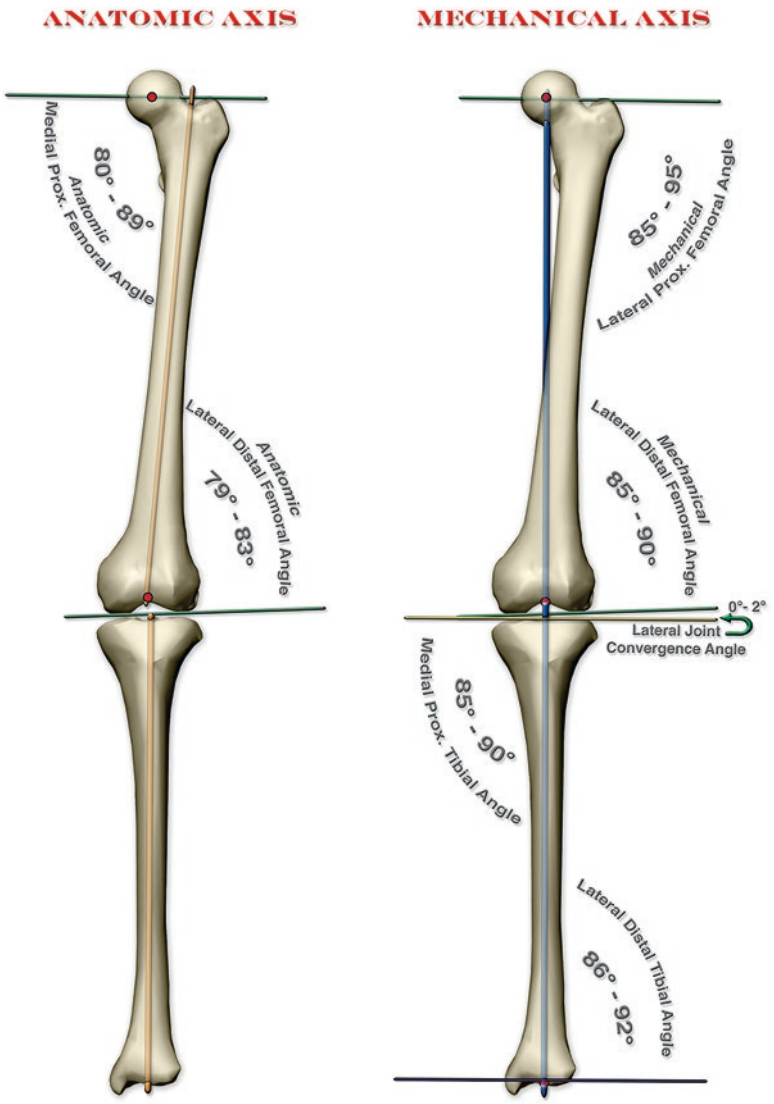


Fig. 5.2 Normal angles, frontal plane, for preoperative planning. Fibulae and patellae omitted for clarity. *Left*: anatomic axes. *Right*: biomechanical axis (After Paley and Herzenberg)

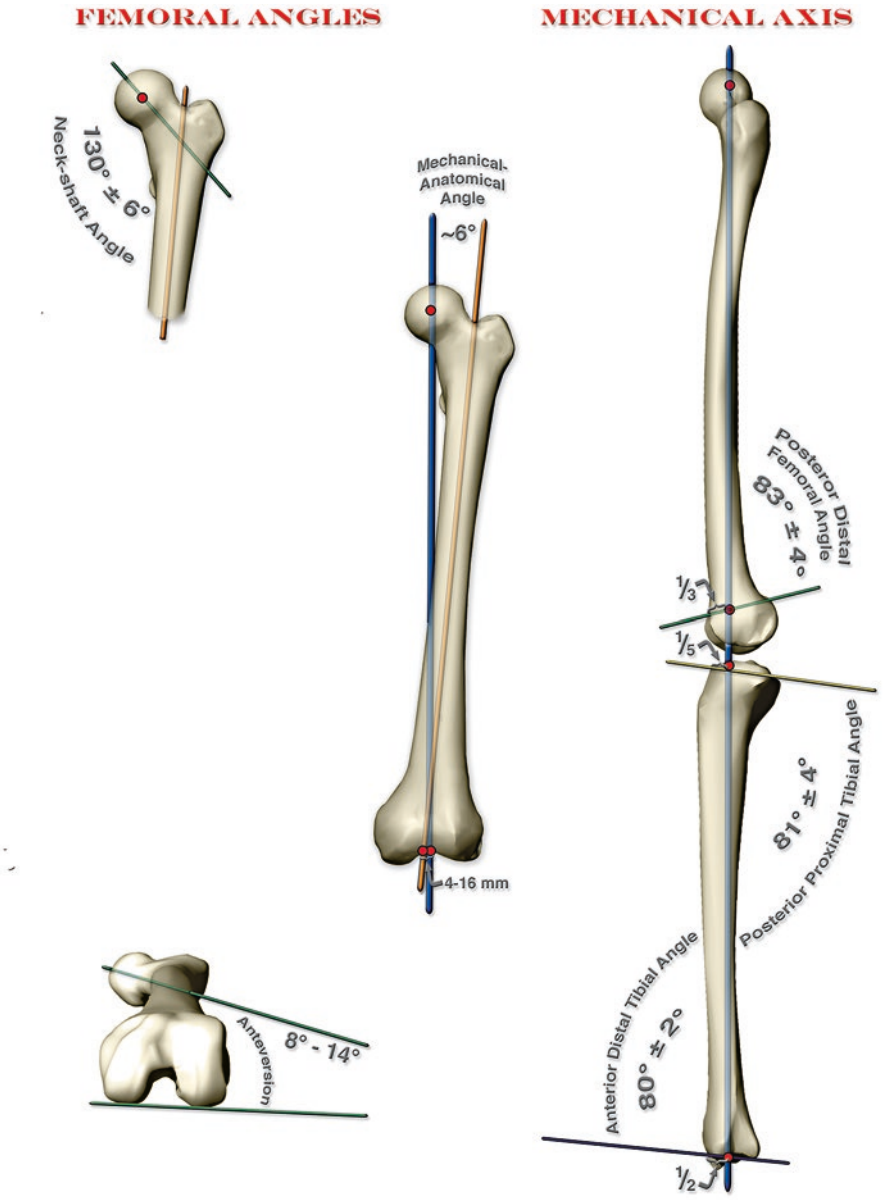


Fig. 5.3 Normal angles, for preoperative planning. Fibulae and patellae omitted for clarity. *Left*: femoral angles. *Right*: biomechanical axis (After Paley and Herzenberg)

surgeons should appreciate the fact that many deformities are not oriented in the radiographic anteroposterior and lateral projection planes but are instead obliquely oriented.

Hidden Deformities

Certain deformities are obvious, evident to anyone looking at a limb. Others, however, are subtler, revealed after measuring the aforementioned angles and axes. Some hidden deformities are compensatory, especially those that develop in a growing child. For example, a partial medial growth arrest of the proximal tibial growth plate will cause the bone to gradually grow into a progressively more varus attitude. The evident bowing of the limb may mask a compensatory valgus overgrowth of the distal femoral physeal plate. Correcting the tibial varus deformation will unmask the femoral valgus because, after correction of the tibia, the limb will stick out to the side, making ambulation impossible (Fig. 5.4).

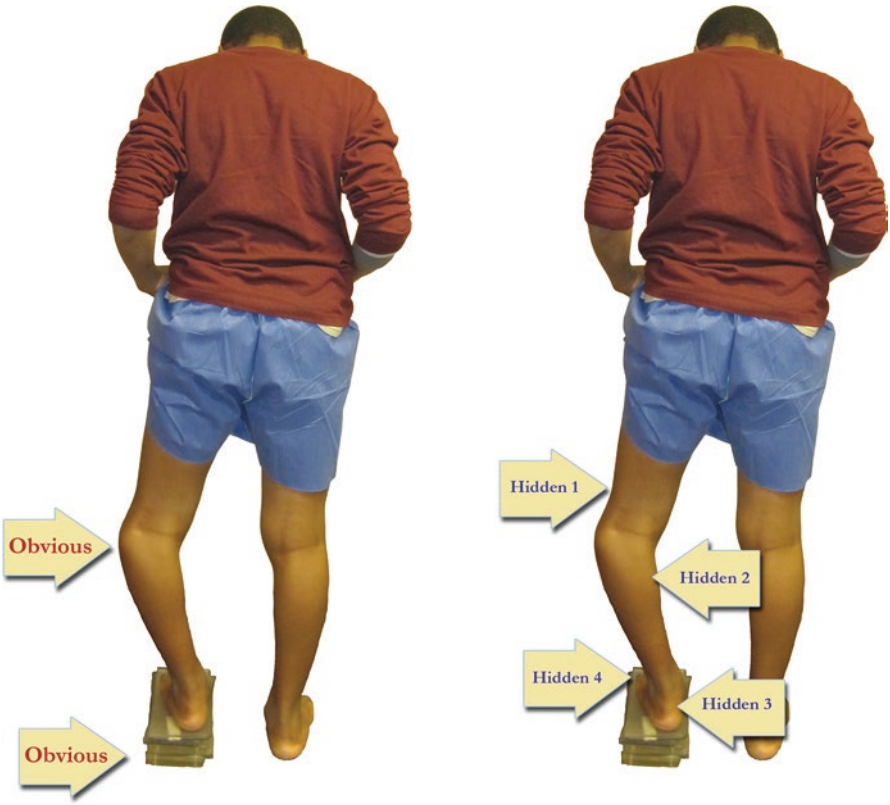


Fig. 5.4 Correcting obvious deformities (*left*) of shortening and tibia varus may unmask hidden deformities including (*1*) distal femur valgus, (*2*) internal tibial torsion, (*3*) heel valgus, and (*4*) foot pronation

Full-Length X-ray Studies

Full-length standing x-ray films are essential for proper assessment of lower limb length discrepancies and deformities. Reproducible positioning helps follow-up x-ray examinations as well. For this reason, the "patella forward" position is commonly used for this purpose. The short limb should be elevated on stacked 1.0 cm blocks until the pelvis is level both clinically and radiographically (Fig. 5.5).

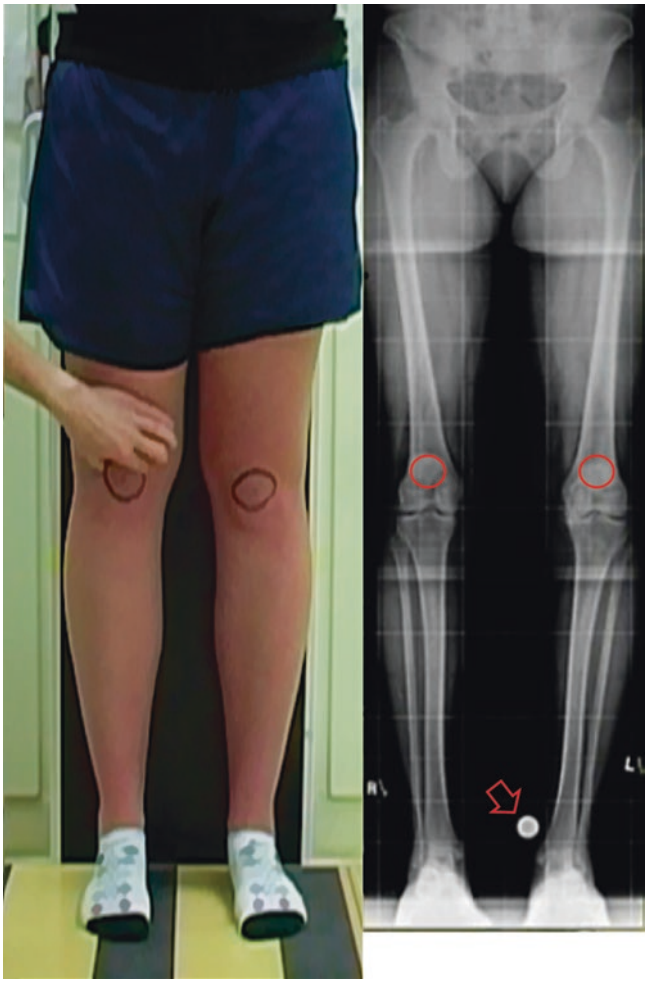


Fig. 5.5 Centering patella will help standardize serial x-ray studies. Note magnification marker (*red arrow*). Courtesy of Janet D. Conway, M.D. Used with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Clinical Illustration: Failure to Identify Hidden Deformity

This case illustrates a femoral deformity in a 14-year-old boy who suffered a partial growth arrest following osteomyelitis of the distal femur. The valgus malalignment and shortening of the femur was corrected with an intramedullary lengthening nail. The surgery and elongation proceeded uneventfully. Final full-length x-ray images demonstrated an unrecognized compensatory varus deformity of the upper tibia, subsequently treated with a hemi-epiphyseodesis (Fig. 5.6).

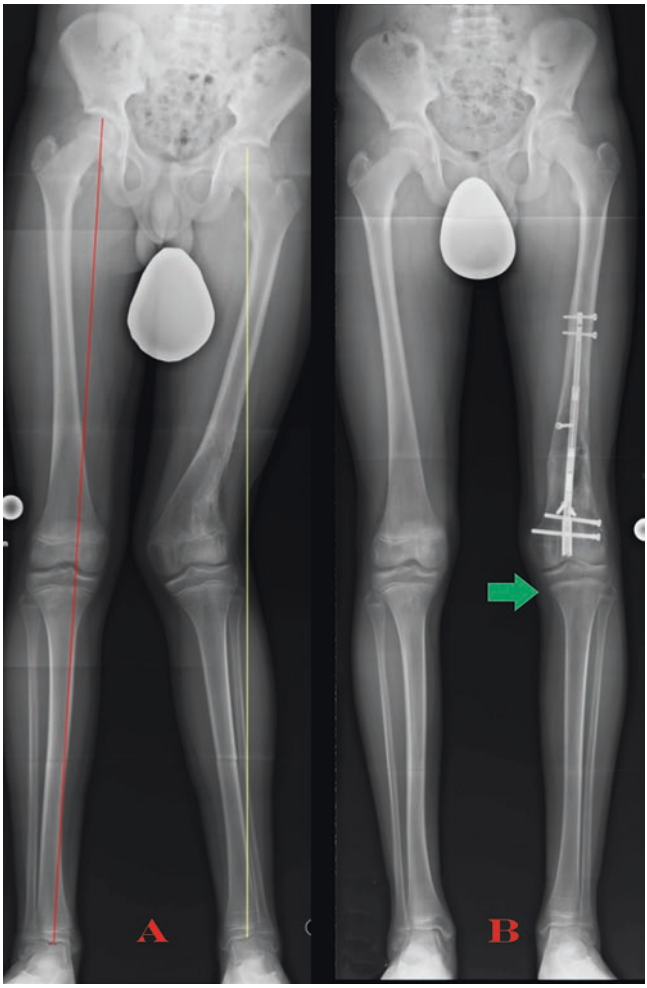


Fig. 5.6 Unmasking a hidden deformity. (**a**) Pre-op AP x-ray image. (**b**) Post-op AP x-ray image. *Green arrow* points to unrecognized compensatory upper tibial varus deformity

Level of the Osteotomy

When elongating a limb with external fixation, osteotomy can be performed anywhere along the length of the bone, provided there exists sufficient bone to safely insert wires or pins into both fragments. When the planned lengthening includes a deformity correction, the location of the osteotomy is dictated by the geometry of the deformity and the anticipated correction. For straight lengthening, when possible, the osteotomy should be performed at the metaphyseal-diaphyseal junction (where the bone begins to flare out as it approaches the adjacent joint). According to Professor Ilizarov, the regenerate develops best in this region.

The Osteotomy Formula

When elongating a limb with an intramedullary lengthening nail, certain recommendations prevail. Such implants contain a nested telescopic rod that extends in response to an externally applied device. The outer tube thus has a larger diameter than the inner rod. Given that stiffness is proportional to the fourth power of the diameter, it stands to reason that the outer tube is stiffer than the inner rod, even though the former is hollow and the latter is solid. Therefore, osteotomy should not be performed at the level of the protruding portion of the nail. Thus, the first consideration in planning the level for osteotomy is to measure the length of the protruding portion of the nail (3.0 cm in the most commonly used PRECICE® nail) and, as a first step, record that length.

Likewise, to place the osteotomy in a manner that assures that the entire widening distraction zone remains along the outer tube of the implant throughout the elongation process, the contemplated length of distraction must be added to the length of the protruding portion of the nail to set the level for osteotomy.

Moreover, to guarantee that the distraction zone does not end near the junction of the inner and outer components of the implant, which would constitute a potential stress riser, the level of the osteotomy should be 4.0–5.0 cm (~2 in.) along the wider component of the implant, away from the telescoping junction.

Therefore, the "osteotomy formula" reads as follows:

Protruding portion (3.0 cm typically) + 5.0 cm (or 4.0 cm) + lengthening

When using a typical PRECICE® nail, therefore, the formula can be shortened to:

8.0 cm + lengthening

Thus, if 7.5 cm of lengthening is planned, the osteotomy level should be 15.5 cm up from the tip of a fully collapsed nail toward the threaded end. In this manner, the entire regenerate zone will be along the thicker portion of the nail, with an additional 5-cm safety margin (Fig. 5.7).

In certain unusual circumstances, it may be necessary to shorten the safety zone to 3.0 or 4.0 cm rather than 5.0 cm. This might occur, for instance, with a very short bone, such as that observed when lengthening the limb of a patient with achondroplastic dwarfism.

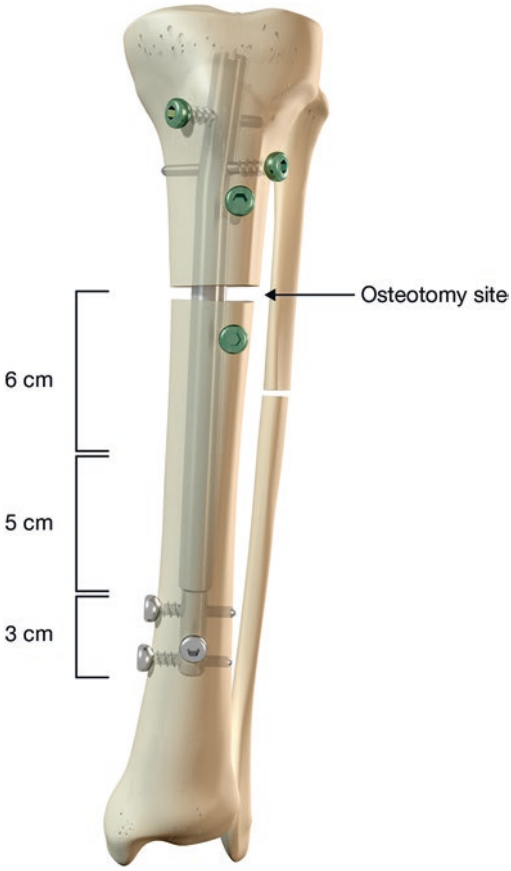


Fig. 5.7 The osteotomy formula helps select an osteotomy level to best prevent nail breakage. The planned lengthening (6.0 cm in this case) is added to the length of the protruding portion of the end of the nail (3.0 cm), which is combined with a "safe zone" at the exterior tube's lower end (usually 4.0–5.0 cm). Add these values and measure upward from the nail's tip. Mark the osteotomy level on the skin after checking with fluoroscopy. Copyright 2016 NuVasive

Fixator-Assisted Nailing

Because intramedullary lengthening nails lack a hollow center, guide wires cannot help maintain alignment of fragments until the implant crosses the osteotomy site. Likewise, oftentimes the lengthening nail is employed to not only elongate a bone but to also acutely correct small angular and/or rotational deformities. Moreover, risk of malrotation of the fragments with respect to each other looms large in the minds of many lengthening surgeons, particularly in femoral lengthening cases.

The tibia's rotation after osteotomy is usually stabilized by performing the fibular osteotomy at a different—typically lower—level on the limb, so external fixation isn't used unless precise control of deformity correction necessitates such a device.

For these reasons, temporary (intraoperative) external skeletal fixation serves to ensure proper final position of the fragments at the end of surgery, especially in femur lengthenings (Fig. 5.8).

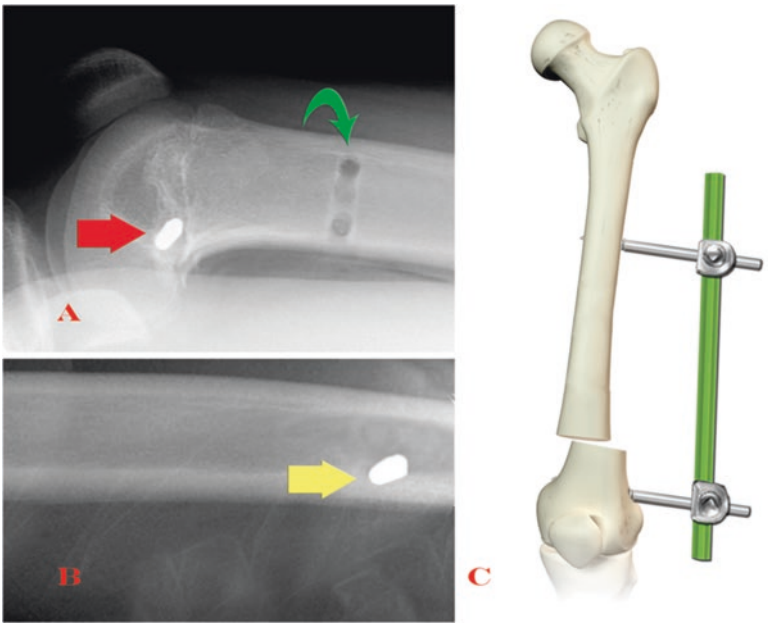


Fig. 5.8 Fixator-assisted nailing. (**a**) Distal external fixation half-pin located behind the nail path (*red arrow*). Note distal femoral osteotomy vent holes (*green arrow*). (**b**) Proximal external fixation half-pin located behind narrower path of telescoping portion of nail (*yellow arrow*). (**c**) Temporary monolateral external fixator used to secure fragments in final alignment prior to nail

Periosteal Elevation

Because intramedullary lengthening involves reaming the marrow thereby destroying endosteal blood supply, formation of the regenerate in the widening distraction region depends exclusively on periosteal blood supply. Therefore, careful elevation of the periosteum from the bone's surface must precede any drilling or osteotomy. Maximum preservation of the periosteum requires approaching the operative region through a small incision, typically the width of an osteotome, made in line with the limb's longitudinal axis.

Approach the bone with a minimal amount of soft tissue dissection. Over the crest of the tibia, for example, a single incision down to the bone will do. Elevate the periosteum with a small elevator as far around the bone as possible on both sides, and extend the elevation for a centimeter up and down the bone. The elevator should be used to protect the periosteum during venting and osteotomy (Fig. 5.9).

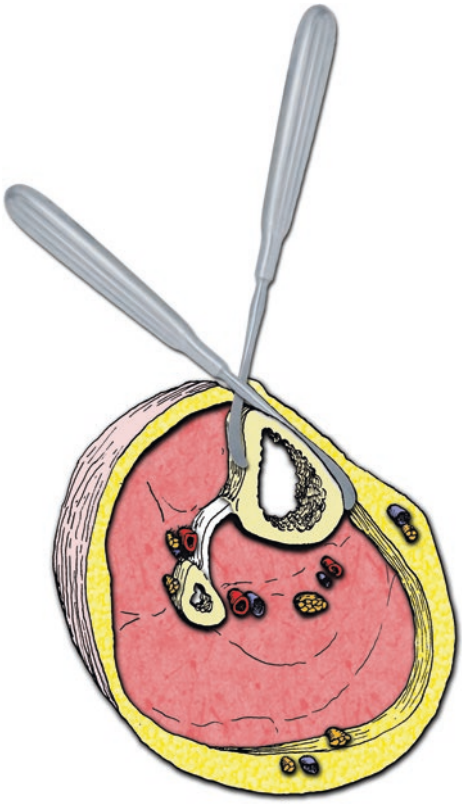


Fig. 5.9 Protection of the periosteum is critically important because it is the only osteogenic tissue remaining after reaming the marrow canal

Venting

To reduce the likelihood of fat embolism, long bone canals must be "vented" prior to reaming. The vent holes, drilled across the involved bone, allow marrow fat to escape into the surrounding soft tissues during reaming. Since a common technique during osteotomy is predrilling at the level where the osteotomy will be performed, these predrilled holes may serve as vent holes for intramedullary reaming. Venting must be performed after first elevating the periosteum to prevent injury to this structure, so important to new bone formation in the distraction zone (Fig. 5.10).

Furthermore, as the vented marrow tissue includes not only fat droplets but also precursor bone cells, reamings that collect around the vent holes can enhance healing at the osteotomy site during lengthening.

Slow reaming, gradually increasing reamer sizes in 0.5 mm steps, will help reduce the incidence of fat embolism syndrome.

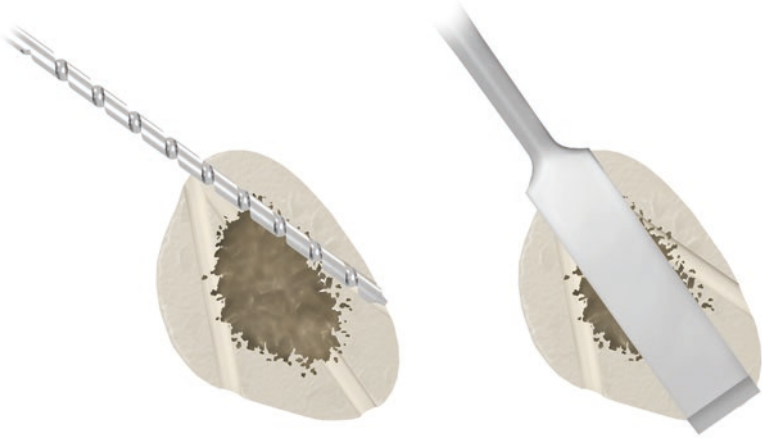


Fig. 5.10 Multiple drill holes (*left*, following periosteal elevation) allow marrow contents to vent out of the bone during reaming of the medullary canal, reducing the risk of fat embolism. The holes also simplify osteotomy of the bone (*right*). Copyright 2016 NuVasive

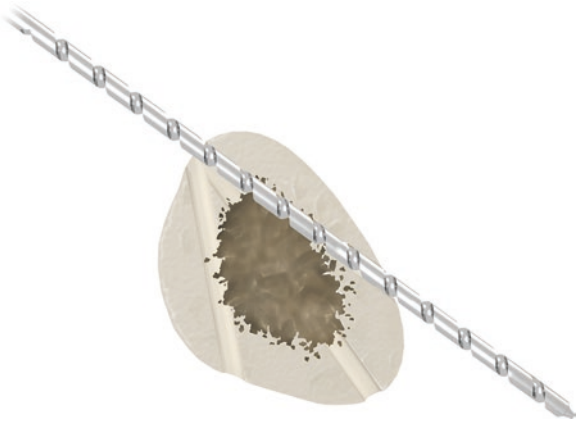


Fig. 5.11 Depth plunging can damage structures on the far side of a bone. See Chap. 11 for discussion of preventive measures. Copyright 2016 NuVasive

Depth Plunging

Over-drilling the far cortex, referred to as "depth plunging," can potentially damage neurovascular structures on the opposite side of bone (Fig. 5.11). The matter is discussed at length in the Chap. 11.

Compartment Syndrome

Certain authorities have recognized that the added volume of such bone marrow reamings can, in the anterolateral tibial compartment, produce a "compartment

syndrome" (a serious form of pressure-induced muscle necrosis). For this reason, it has been suggested that venting should be avoided on the lateral side of the tibia.

Furthermore, as reaming continues beyond the level of the osteotomy site (where the vent holes are located), additional vent holes at the distal end of the bone may be necessary.

Thermal Injury While Drilling Vent Holes

The heat generated by a drill bit spinning in cortical bone has been the subject of considerable research. Temperatures exceeding 55 °C will likely cause necrosis of osteocytes in the lacunae around the drill bit [1]. The radius of this zone of injury is proportional to the heat generated by drilling. Ordinarily, when drilling is part of an internal fixation procedure, the dead bone surrounding a drill hole will, at least initially, hold a screw as securely as do viable bone. With time, however, loosening may occur.

With external skeletal fixation, dead bone surrounding a drill hole subsequently used to secure transcutaneous pins remains in contact with the microflora of the pinhole, a potential nidus of osteomyelitis taking a unique form: the ring sequestrum. In this situation, a zone of separation occurs between the living and dead bone around the hole. Local stresses cause the living bone to convert into strain-tolerant granulation tissue, separating it from the nonviable bone immediately around the hole. Thus, does the burnt bone take the shape of a ring or cylinder surrounding the pin or wire? When the implant is removed, the ring is often left behind, creating a persistent nonviable surface for microorganisms to inhabit.

The matter of heat generated during reaming for an intramedullary lengthening nail is discussed elsewhere in this monograph, but this section deals with heat generated by a drill bit during venting.

The quality of regenerate bone formation in a widening distraction gap depends on many factors described in previous chapters. The new-forming bone grows from both sides of the gap, so it stands to reason that viable bone cells are essential for rapid regenerate formation and consolidation. Hence, thermal injury caused by drilling vent holes will likely degrade the regenerate.

Clearly, a sharp drill bit, a stop-start drilling pattern, irrigation of the drill bit, and a measure of patience will reduce thermal bone damage [2]. The issue of depth plunging, a source of potential damage to structures deep to bone being drilled, is considered in the chapter containing the anatomy cross sections.

Osteotomy Technique

To reduce the likelihood of displacement, experienced surgeons will create an incomplete osteotomy at the level of transection. Subsequently, the surgeon inserts the nail up to, but not across, the osteotomy site. The surgeon next completes the osteotomy with a gentle tap on the chisel or osteotome, placed subperiosteal, taking

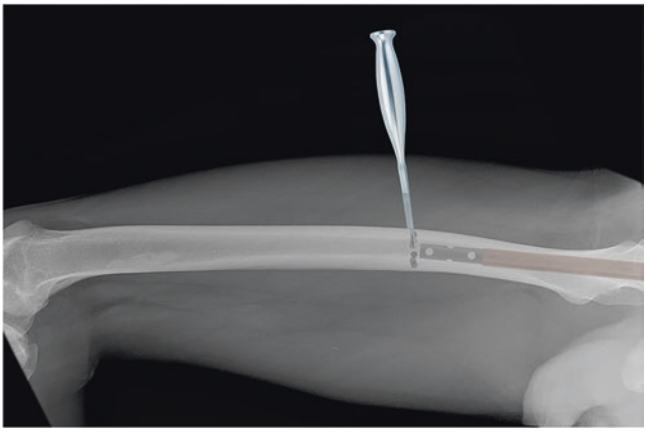


Fig. 5.12 Advance the nail up to the vented osteotomy site after reaming the canal for nail placement. Complete the osteotomy using the percutaneous technique while protecting the periosteum. Copyright 2016 Zeeca Publishing Co

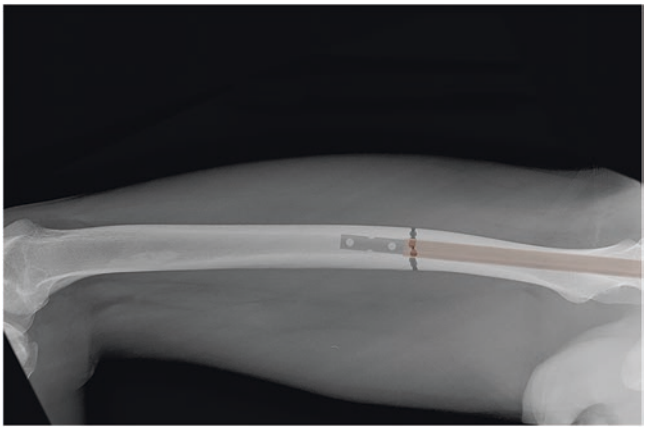


Fig. 5.13 Immediately after completion of osteotomy, advance the nail across the osteotomy site to prevent displacement. Copyright 2016 Zeeca Publishing Co

care not to displace the adjacent bone fragments with respect to each other. The nail is then immediately advanced across the osteotomy site, thereby aligning the fragments (Figs. 5.12 and 5.13).

Transverse Locking Screws: Proximal Guide

Proximal locking of intramedullary lengthening nails is performed in the same manner as the technique developed for trauma nails. The insertion guide always includes holes that line up with the holes in the nail when the guide is securely fixed to the

implant. However, mismatching is possible; the guide could, conceivably, be attached backward or to the wrong device. Therefore, the surgeon should make sure that drill sleeves, drill bits, and perhaps even the screws line up precisely with the locking screw holes of the implant when passed through the holes on the guide handle. This is accomplished by preassembling the proximal guide to the nail and checking that the implant's holes line up with the drill sleeves.

Testing the Implant

Intramedullary lengthening nails incorporate complicated mechanisms that must be tested before the patient leaves the operating room. One or two millimeters of separation should be enough to visually confirm that the device is functioning properly. Observation of a widening of the osteotomy gap or a change within the implant itself can satisfy the need for confirmation. With the PRECICE® nail, for instance, widening of the internal leadscrew gap confirms lengthening.

Preliminary Osteotomy Gap

While some surgeons, to save operating time, leave the bone ends distracted a couple of millimeters after this separation test, the classic Ilizarov strategy calls for reduction of the osteotomy back to a non-displaced fracture. That is, the bone ends should be restored to their anatomic position before the patient is sent to the recovery room, if the device permits retraction of the nail. This author believes that delayed consolidation of the regenerate described by many authors using intramedullary lengthening implants can be traced to the initial unreduced gap at the osteotomy site.

A Spare Implant

Because of the mechanical complexity of intramedullary lengthening nails, the implants cannot be autoclaved for sterilization. Instead, gamma rays are used to kill all potential microbes on or in the device. Therefore, if a surgeon or assistant fails to make a diving grab of an intramedullary lengthening nail before it hits the floor, having a spare nail of the same size nearby will seem like a wise strategy indeed!

Correcting Deformities with Intramedullary Lengthening Nails

At present, the ability to make substantial deformity corrections with lengthening intramedullary nails is rather limited. Too much manipulation of the osteotomy site risks retarding regenerate new bone formation in the distraction zone—a region whose intramedullary blood supply has already been compromised by reaming for

the nail. Even simple corrections of rotational malalignment with the nail in place are appealing, but excessive rotation at the osteotomy site, beyond 15–20°, may slow maturation of new bone formation.

Correction of angulation at the distraction zone should also be restricted; even experienced surgeons limit angular correction to 10–15°. Remember: The more extensive the dissection needed for surgical exposure of the bone, the less blood supply will be available to the bone ends, especially after intramedullary reaming for a lengthening nail. The femur appears to be more tolerant than the tibia of angular correction. Thus, while a 15° angular correction will likely be tolerated in the femur and not interfere with regenerate formation, it is safer to limit acute tibia corrections to 10° or less.

Considerations for Tibiofibula Lengthening

The Fibula

Although surgeons often consider intramedullary lower leg lengthening to be technically similar to treating a tibial fracture with an intramedullary nail, one distinct difference overshadows all others: when lengthening the tibia with any device, the fibula must be pulled along at the same rate and rhythm, with the same final gap size. If this does not occur, the fibula may sublux or dislocate, either at the proximal tibiofibular joint or distally at the ankle.

If the fibular head is pulled downward during tibial lengthening, the lateral collateral ligament of the knee will tighten, restricting knee motion and perhaps resulting in valgus deformity of that joint. Distally, the dense ligaments securing the fibula to the talus and calcaneus usually prevent upward movement of the lateral malleolus during tibial lengthening, but with inappropriate methodology, this upward displacement of the lateral malleolus may occur nonetheless.

For these reasons, a fibular osteotomy must accompany both external and intramedullary tibial lengthening. Furthermore, both ends of the fibula must be secured to a lengthening tibia.

Unless a substantial concomitant deformity correction is anticipated, fibular osteotomy should be performed at a different level to that of the tibial osteotomy. This step will reduce the likelihood of compartment syndrome. Moreover, fibular osteotomy should be at least 13.0 cm below the knee joint to prevent inadvertent injury to the peroneal nerve. If the fibula must be cut more proximally, peroneal nerve exposure, protection, and release should be added to the operative protocol.

Osteotomy of the Fibula

Typically, surgeons expose the fibula subperiosteally through a lateral incision. Retractors are necessary to protect the peroneal artery and its two *venae*

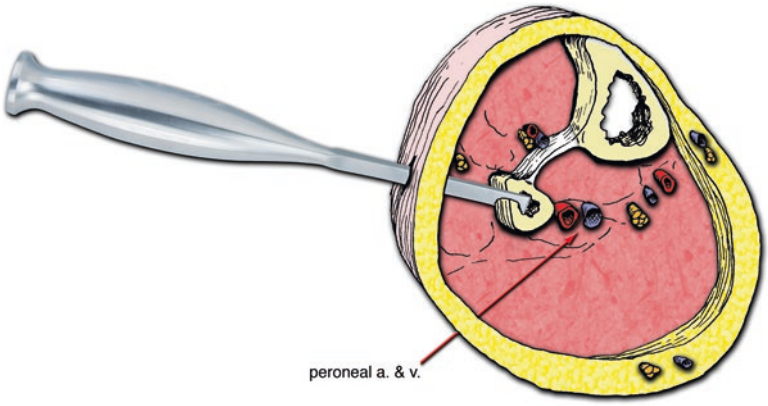


Fig. 5.14 Cross section through middle of the lower leg demonstrates the proximity of the peroneal artery and vein to the medial surface of the fibula. Curved retractors are necessary to protect the structures during fibular osteotomy

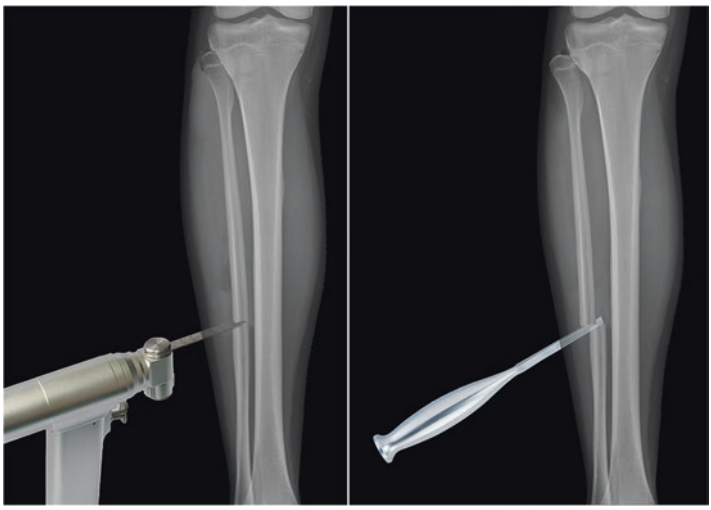


Fig. 5.15 Expose the fibula through a lateral incision and subperiosteal dissection. Partially transect the fibula with an oscillating saw (*left*), and complete and confirm the osteotomy with an osteotome. Copyright 2016 Zeeca Publishing Co

comitantes, during fibular osteotomy (Fig. 5.14). The vessels lie immediately medial to the bone. The best way to avoid transecting these vessels is to begin the osteotomy with a thin-bladed oscillating saw and complete it with a sharp osteotome (Fig. 5.15).

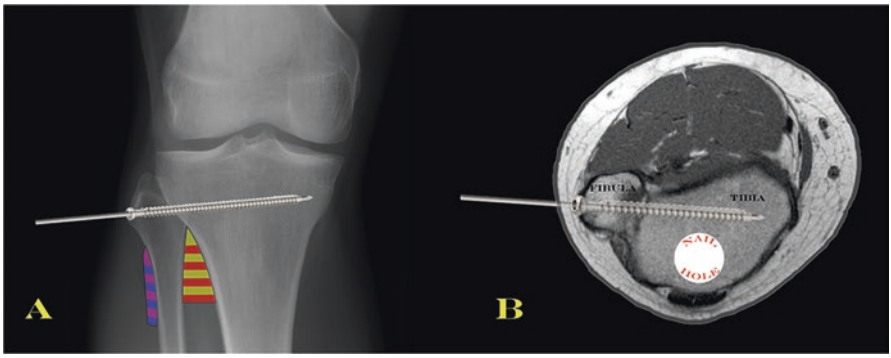


Fig. 5.16 (**a**) Proximal securing of the fibula to tibia; screw over K-wire. (**b**) The screw must pass behind the anticipated nail hole and avoid the peroneal nerve (*blue* and *magenta* stripes) and the hiatus for the anterior tibial artery (*red* and *yellow* stripes). Copyright 2016 Zeeca Publishing Co

Fibular Fixation to the Tibia

The safest and most reliable way to secure the fibula to the tibia for lengthening is to cross both the proximal and distal tibiofibular joints with screws. In this way, structures that inhabit the space between the tibia and fibula are avoided. A Kirschner wire, drilled across the joint, will ensure optimal positioning. Proximally, the technique involves inserting a Kirschner wire into the fibular head (with attention to the position of the peroneal nerve), across the proximal tibiofibular joint, and into the upper tibia behind the location of the intramedullary nail (Fig. 5.16).

Distally, insert a Kirschner wire from the fibula (a narrow bone) into the wider tibia, and use that K-wire to pass a cannulated screw from the tibia to the fibula. A slight tilt of the screw will add stability to the construct (Fig. 5.17).

Be aware of the location of the peroneal tendons immediately behind the lateral crest of the fibula (Fig. 5.18) .

Blocking Equinus

Paley and his co-workers developed an ingenious and deceptively simple way to temporarily block equinus during tibia/fibula lengthening [3]. They suggest insertion of a screw from the calcaneus to the distal tibia, to be left in place until lengthening is complete. The screw is inserted over a K-wire used as a placement guide. The K-wire must be directed toward the posterolateral corner of the distal tibia, immediately above the ankle, lest damage to the flexor hallucis longus tendon or, worse, the posteromedial neurovascular bundle occur (Fig. 5.19).

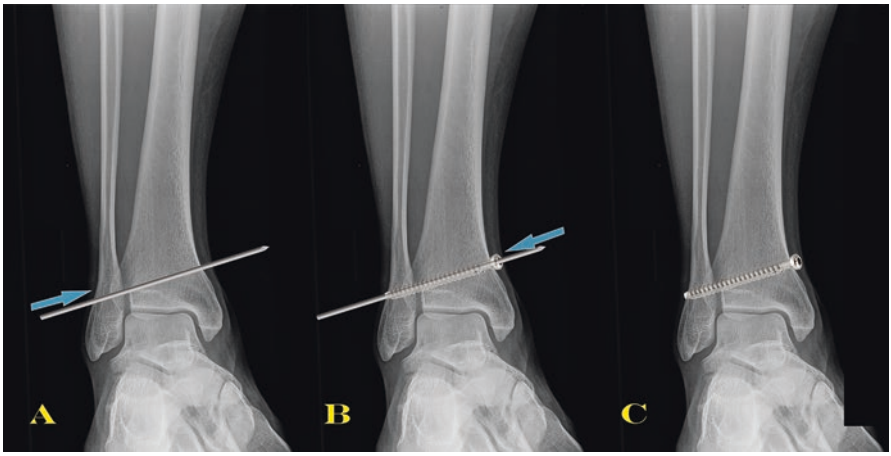


Fig. 5.17 Distal securing of the fibula to tibia. (**a**) A K-wire is inserted from the distal fibula into the tibia across the tibiofibular syndesmosis. (**b**) A screw is inserted along the K-wire from the tibial side. (**c**) The K-wire removed. Copyright 2016 Zeeca Publishing Co

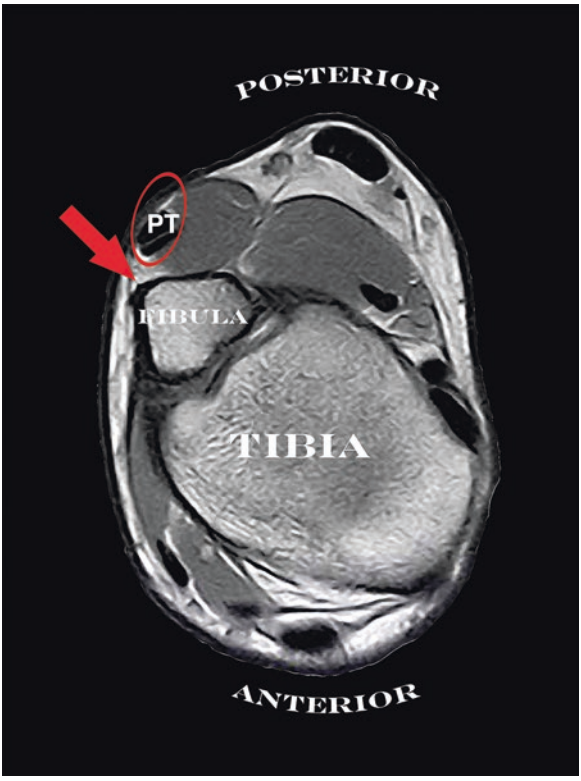


Fig. 5.18 MRI cross section of the syndesmosis. Avoid impaling peroneal tendons (PT) by aiming anteriorly from the lateral crest of the fibula. Copyright 2016 Zeeca Publishing Co

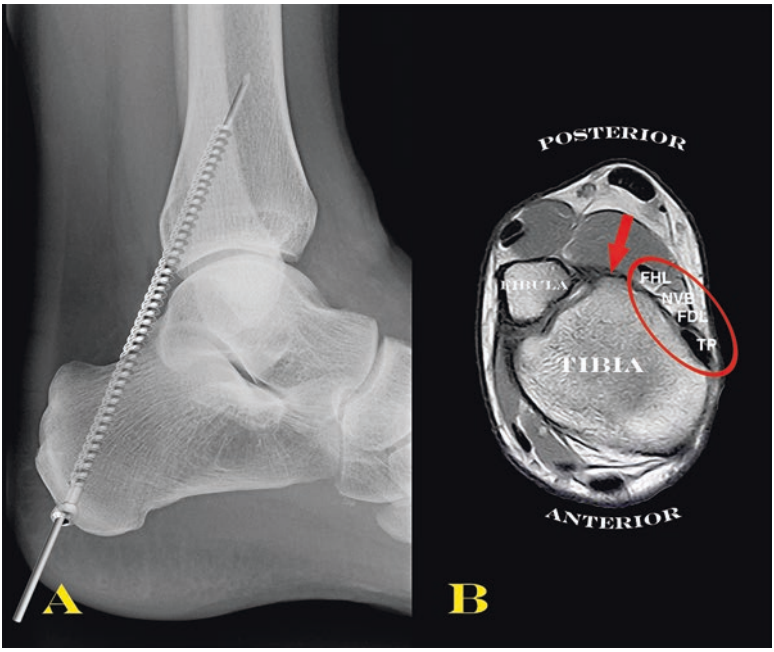


Fig. 5.19 (**a**) A long calcaneal-tibial screw can be used to block equinus during tibia/fibula lengthening, inserted over a guiding K-wire. (**b**) Cross section of the distal tibia and fibula at the ankle. The *arrow* points to the safe insertion point at the posterolateral corner of the tibia, the only place where such a screw can be inserted into the tibia without impaling a tendon, nerve, artery, or vein. Copyright 2016 Zeeca Publishing Co

Tibial Insertion Point

The tibial insertion point for intramedullary lengthening nails is the same as that for intramedullary trauma nails: the bare area anterior to the distal attachment of the anterior cruciate ligament and lateral to the anterior horn of the medial meniscus (some fibers of the meniscal-tibial ligament and ACL may be damaged during insertion) (Fig. 5.20). Several approaches to this area are commonly used, including patella tendon splitting, patella tendon retraction (Fig. 5.21), and via the suprapatellar pouch (Figs. 5.22 and 5.23). In the latter approach, thin-walled tubes must be used to keep drilling debris from ending up in the knee joint after reaming the bone.

Transverse Locking Screws: Distal Targeting

Years ago, intramedullary nail manufacturers developed long targeting devices designed to make distal locking screw insertion as reliable as proximal screw locking. Unfortunately, slight bending of the implant resulted in transverse screws anterior or posterior to the implant in spite of the guide. For this reason, surgeons started

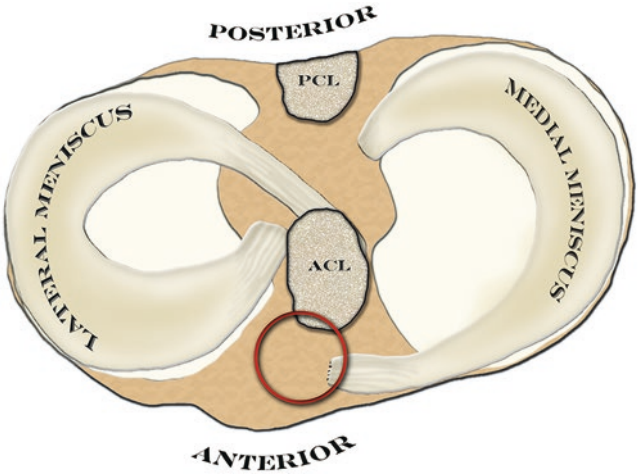


Fig. 5.20 The tibial plateau showing the tibial insertion region (*red circle*) anterior to the anterior cruciate ligament footprint on the tibia. Copyright 2016 Zeeca Publishing Co

Fig. 5.21 The tibial insertion point at the anterior corner of the tibial plateau as seen from the lateral view. Flexing the knee helps with this approach. Copyright 2016 Zeeca Publishing Co

to "freehand" distal locking, using the "perfect circle" technique. Nowadays surgeons experienced in the use of locked trauma nails use an identical technique to lock intramedullary lengthening implants (Fig. 5.24).

Alternatively, surgical instrument makers have created radiolucent right angle drill attachments that simplify distal targeting. Biomet, for instance, has one that connects to a regular electric drill at a right angle to the drill bit. It contains a metal circle around the drill chuck that is easily visible on fluoroscopy. The device is maneuvered into place and aligned until the drill bit is visualized axially as a single

Fig. 5.22 The suprapatellar approach to the proximal tibia entry point. Reaming through a tube will prevent bone fragments from entering the joint. Copyright 2016 Zeeca Publishing Co

Fig. 5.23 AP view of insertion point or tibial nailing. The location is slightly medial to the lateral tibial spine. Copyright 2016 Zeeca Publishing Co

dot centered over the perfect circle of the distal locking hole in the implant. Then the drill bit is pushed through the bone and implant without allowing the drill bit to slide along the bone's cortex (Figs. 5.25 and 5.26).

Fig. 5.24 Perfect circle techniques developed for trauma nails are also ideal for lengthening nails. Copyright 2016 Zeeca Publishing Co

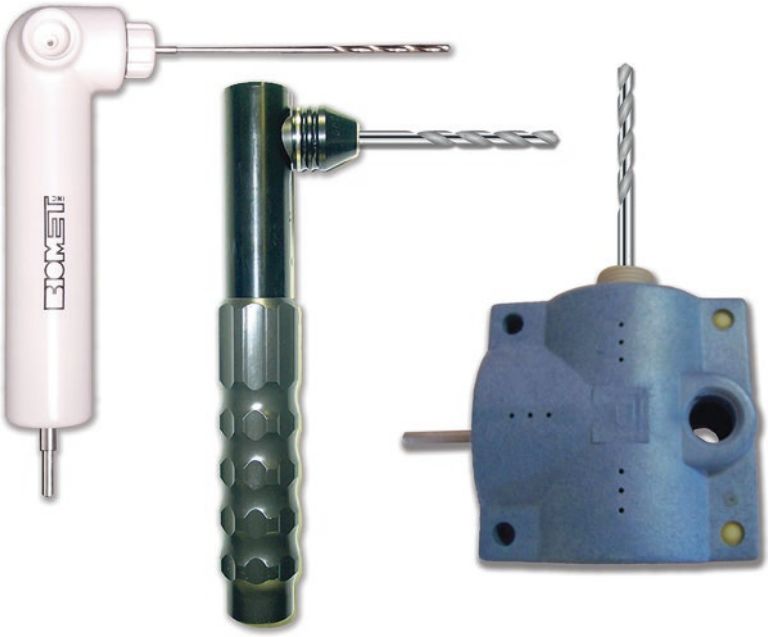


Fig. 5.25 Right angle radiolucent drills, Biomet (*left*), Synthes (*middle*), and Smith & Nephew (*right*—no longer available). Copyright 2016 Zeeca Publishing Co

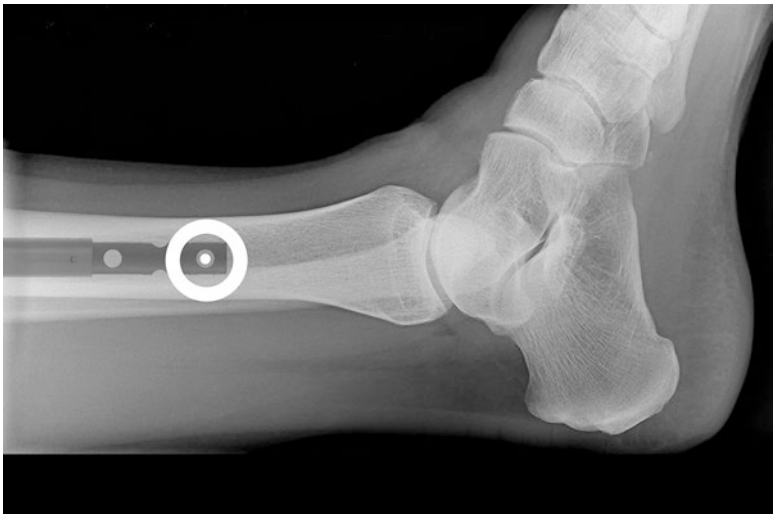


Fig. 5.26 Fluoroscopic image of chuck and drill bit (*white dot*) perfectly aligned with distal screw hole of implant. Copyright 2016 Zeeca Publishing Co

Considerations for Femoral Lengthening

Antegrade and Retrograde Nailing

The femur can be nailed from either the hip region or through the knee joint. Each method has its advantages and disadvantages, as well as passionate proponents and opponents. The debate regarding antegrade or retrograde nailing in limb lengthening echoes an identical discourse in trauma care. In a carefully performed prospective study of this issue, Ostrum et al. [4] came to the following conclusion: "Each insertion technique has its own advantages and disadvantages. The two insertion modes appear to be relatively equal for the treatment of femoral shaft fractures."

With an antegrade approach, irritation of muscles around the hip can result in hip pain that is usually relieved when the nail is removed. The incidence of heterotopic ossification (Fig. 5.27) is rarely reported as a major postoperative problem with intramedullary limb lengthening, although, with hip fractures in general, 2.5% will develop heterotopic ossification after internal fixation sufficient to require treatment [5].

The retrograde insertion point, originally developed by the author [6] and David Seligson [7], violates the articular cartilage of the knee joint in the femoral groove, a potential source of patellofemoral knee pain for the patient. Nevertheless, the principal advantage of the retrograde approach is to allow a small translation and angulation offset in the supracondylar region of the femur to prevent valgization of the knee as an unfavorable consequence of femur lengthening.

Fig. 5.27 Painful heterotopic ossification located around a trochanteric entry portal following intramedullary femoral lengthening. Resection cured the symptoms. Copyright 2016 Zeeca Publishing Co

In children with an open distal growth plate, retrograde nailing risks damaging the distal physeal plate cartilage; therefore, distal nailing is generally contraindicated in children. However, some evidence suggests that growth arrest will not occur when an implant crosses the growth plate if the implant is left in place during growth, thereby preventing the formation of a bony bridge across the growth plate cartilage.

Trochanteric or Piriformis Femoral Entry Portal

Clearly, for growing children, surgeons should avoid piriformis fossa entry for antegrade femoral nailing. The blood supply to the epiphyseal plate of the femoral head can be compromised when performing antegrade femoral nailing via the piriformis fossa, as indicated in the diagram below. Thus, for pediatric antegrade nailing, the trochanteric entry point is the only one available (Fig. 5.28).

Likewise, certain pathologies, such as a very short femoral neck, may require trochanteric rather than piriformis entry. The piriformis fossa is in a direct line with the central axis of the femur. From an anatomic perspective, the piriformis fossa is *not* the superior ridge of the femoral neck as seen on anteroposterior (AP) x-ray views; instead, the fossa lies behind and below that ridge. A K-wire or Steinmann pin used to find the fossa should first make contact with the bone after the tip of the

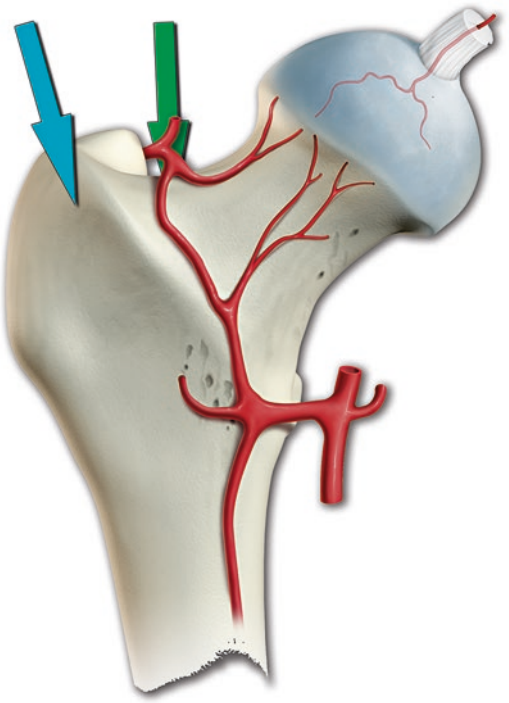


Fig. 5.28 Blood supply to the femoral head. In a growing child, the blood supply of the growth plate of the femoral head is especially vulnerable to injury during piriformis entry (*cyan arrow*) of an intramedullary nail. For this reason, trochanteric entry (*magenta arrow*) is recommended for children. Copyright 2016 NuVasive

implant passes below the superior radiographic ridge of the bone seen on AP fluoroscopic views (Fig. 5.29).

Trochanteric entry can also be difficult, because there is often a tendency to aim medially rather than distally, risking penetration of the medial cortex. Fortunately, orthopedic surgeons who perform limb lengthening procedures are quite familiar with strategies to avoid medial cortex penetration during femoral nailing. Indeed, nowadays, even hip fractures are treated with an implant that enters the canal through the greater trochanter. The entry point for trochanteric nailing is *anterior* to the tip of the trochanter when seen on AP fluoroscopic imaging, because the tip of the trochanter curls posteriorly, above the anatomical axis of the shaft (Fig. 5.30).

For both piriformis and trochanteric entry points, the true lateral projection image on fluoroscopy should be the same: the initial guide pin must be aligned with the center of the femur's medullary canal (Fig. 5.31).

Retrograde Femoral Nailing

The retrograde (distal) portal for femoral intramedullary nailing, as mentioned earlier, was devised by the author and Seligson and Henry [8] in the late 1980s. Originally, the approach was designed to treat distal femoral non-unions, a challenging consequence of comminuted and displaced distal femoral fractures. In the

Fig. 5.29 Trochanteric and piriformis entry points for the femur. The trochanteric entry point is anterior and distal to the highest visualized point of the greater trochanter (*black line*) on the AP x-ray view, while the piriformis entry point is posterior and distal to the ridge of the femoral neck (*white line*). Copyright 2016 Zeeca Publishing Co

Fig. 5.30 Trochanteric (*cyan circle*) and piriformis (*red circle*) entry points. (**a**) *Top* view. (**b**) *Anterior* view. (**c**) *Posterior* view. Notice that the trochanteric entry point is anterior and below the tip of the trochanter (*cyan arrow*), whereas the piriformis entry point is behind and below the high ridge of the femoral neck. Copyright 2016 Zeeca Publishing Co

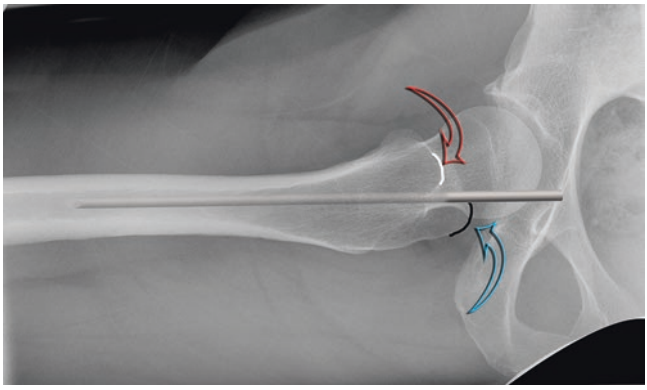


Fig. 5.31 Lateral of hip and femur aligned for IM nail insertion. One the lateral projection, the trochanteric entry point is anterior and distal to the tip of the greater trochanter (*cyan arrow* and *black line*), whereas the piriformis entry point is behind and below the superior edge of the femoral neck (*red arrow* and *white line*). Copyright 2016 Zeeca Publishing Co

1980s, the implants used for preliminary care of such injuries were suboptimal, resulting in loss of fixation, hardware pullout, and the so-called golf club femur, among other catastrophic problems. In such injuries, the primary goal of the procedure in question was salvage of a potential limb loss situation; consequently, some residual knee pain was thought to be a small price to pay for retention of the limb.

Gradually, the simplicity of the retrograde technique proved appealing to surgeons treating fresh trauma. The original implant designed for insertion through the distal portal (the GSH nail) had a 15° bend near the end of the nail, to facilitate a more posterior insertion point than is used at present. Indeed, a surgeon cannot use such a posterior insertion point on an intact femur for anatomic reasons. As this portal was created for treating supracondylar non-unions, the distal fragment in such cases could be extended at the non-union site, allowing direct visualization of the posterior femoral notch region, an area bare of articular cartilage and ligament attachments (despite being very close to the footprint of the posterior cruciate ligament on the femur).

As other manufacturers began creating retrograde femoral nails, the bend disappeared, and a straight nail became the standard for the distal portal. Unfortunately, this culminated in a more anterior entry point into the distal femur than originally contemplated, resulting in a hole in the femoral cartilage of the patellar groove, a possible source of future patellofemoral pain.

Having said that, wide acceptance of the distal trans-cartilage portal for trauma care and now femoral lengthening suggests that a trade-off exists between hip pain with the antegrade entry and knee pain with the retrograde entry. In both cases, equally good and bad choices are present, giving rise to substantial controversy.

Careful attention to initial Steinmann pin placement prior to implant insertion will likely minimize the problems mentioned above. The pin should be inserted at the distal end of Blumensaat's line, along the central axis of the distal femur (Fig. 5.32).

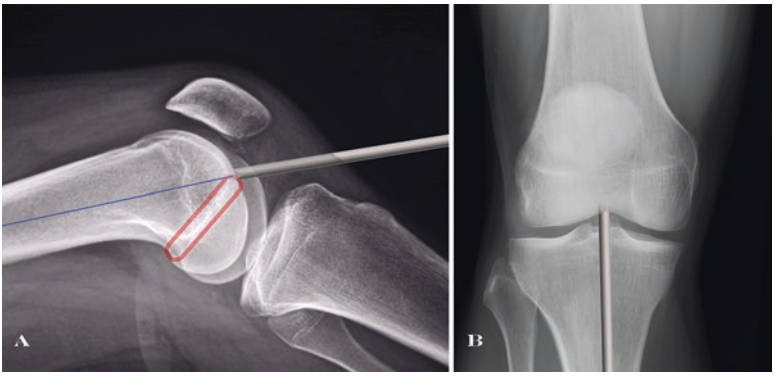


Fig. 5.32 The entry point for distal (retrograde) nail insertion. (**a**) In the lateral projection, the Steinmann pin is inserted where Blumensaat's line (within *red oval*) meets the distal cortex. (**b**) In the AP view, the pin is placed in the femoral notch, at the midline. Copyright 2016 Zeeca Publishing Co

Rozbruch and Fragomen, orthopedic surgeons at the Hospital for Special Surgery in New York City, insert a temporary coronal plane Steinmann pin in the proximal and distal femur prior to osteotomy when performing femoral lengthening [8]. The pins should be placed in the posterior part of the bone, behind the planned nail track. In this manner, they can judge and correct any malrotation that occurs after osteotomy, but before transverse screw locking, by aligning the pins in the coronal plane. Likewise, if any rotational correction is included as part of the operative plan, the pins can be offset by the expected amount of malrotation and aligned with each other in the coronal plane after osteotomy.

Bone Curvature

By necessity, intramedullary lengthening nails are *straight* implants, whereas the femur is a curved bone, with the arch of the curve facing anteriorly. For the telescopic portion of the nail to function properly, the bone must glide smoothly along the tubular part of the implant. To achieve this, a surgeon must over-ream the femoral canal such that the implant does not jam during elongation. Over-reaming must be carefully performed, lest the reamer penetrate the cortex. Usually, the reamed medullary canal is 2 mm wider than the largest diameter of the implant. In a bone with a steep curve, further reaming may be necessary (Fig. 5.33).

Clinical Illustration: Cortical Penetration

One of the distinct advantages of a straight reamer for femoral intramedullary nailing, whether through a proximal or distal portal, is that a tighter nail fit is possible.

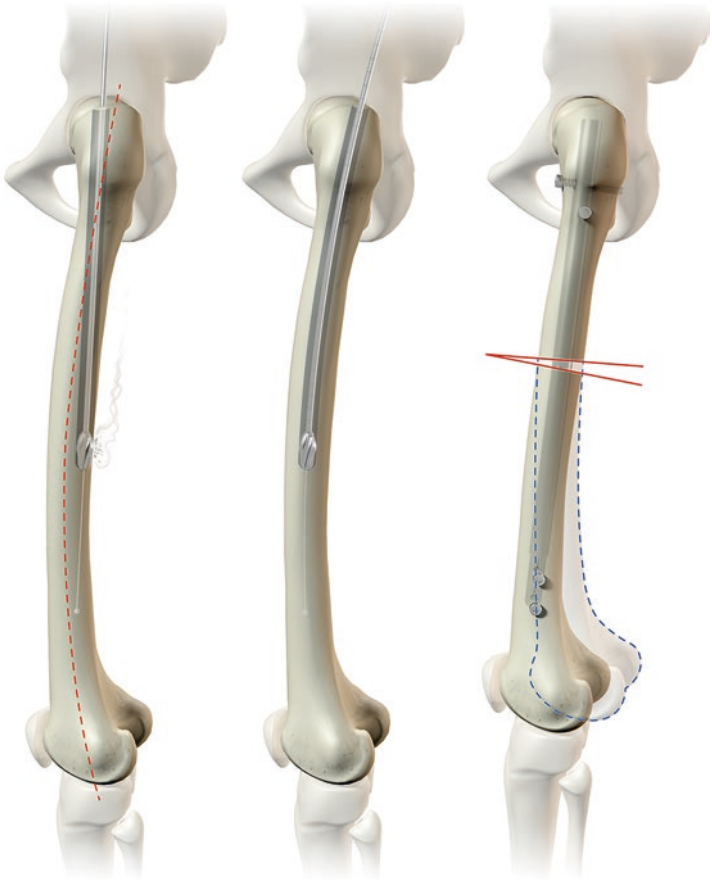


Fig. 5.33 A straight reamer risks penetration of the anterior or posterior cortex of the femur, a curved bone. A flexible reamer follows the central axis of the marrow canal, but over-reaming is necessary to allow a straight nail to fit in the bone. A slight straightening of the bone's curvature will often occur when a straight nail is inserted into a curved, transected bone. Copyright 2016 NuVasive

With flexible reamers, as mentioned above, a surgeon should over-ream 2.0 mm larger than the nail diameter to allow easy gliding of the bone over the implant during limb lengthening. This capacious channel will prevent jamming of the implant against the sidewalls of the nails pathway, especially in curved sections of the femur. The downside risk of such a large canal is instability during elongation, leading to possible varus-valgus or recurvatum-antecurvatum deformities at the osteotomy site. Judiciously placed blocking screws are needed to prevent such occurrences.

A straight reamer allows a tighter nail fit, with over-reaming to only 0.5 mm larger than the nail's diameter. This snugness will do much to prevent osseous deformities, especially in the diaphyseal portion of the bone, although close-fitting

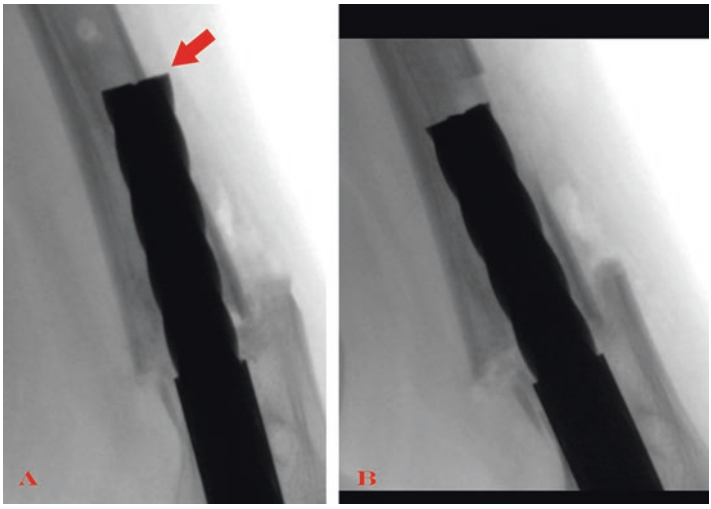


Fig. 5.34 (**a**) Cortical penetration during reaming with a straight reamer (*red arrow*). (**b**) Withdrawing and redirecting reamer demonstrates cortical defect

reaming in the cancellous ends of the bone may not be sufficient to prevent deformities if blocking screws are not used.

A straight reamer, however, can act as a bone eraser, easily penetrating the cortex if use injudiciously (Fig. 5.34).

Considerations for Humeral Lengthening

Antegrade and Retrograde Nailing

The standard portal for the humerus is antegrade, through the rotator cuff, which must be split parallel to its fibers and retracted both anteriorly and posteriorly, to avoid permanent damage to the structure.

The retrograde portal is located along the distal humeral shaft, above the joint, at the roof of the olecranon fossa.

As with femoral IM nailings, antegrade nailing is more likely to create functional deficiencies of the shoulder, and retrograde nailing will more likely produce residual elbow problems [9].

Nail removal may prove especially challenging with retrograde lengthening humeral nails. Indeed, it may be necessary to remove the locking screws and shorten the nail to get it out through the distal portal, particularly after substantial lengthenings. Clearly, only a nail that can be shortened via an external controller can be used in this situation.

Nail Size

Needless to say, careful sizing of the humeral canal, rather narrow in many people, must be done during the preoperative planning phase of any contemplated humeral lengthening. A standard size marker is essential for the step of the procedure.

The Proximal Humeral Portal

Percutaneous insertion of an intramedullary device on any kind into the proximal humerus should be performed through an open incision, splitting the fibers of the deltoid muscle and rotator cuff, and exposing the bone to direct vision (Figs. 5.35 and 5.36). The only structure at risk during the surgical approach is the axillary nerve, which lays on the undersurface of the deltoid muscle (perpendicular to its fibers) two fingerbreadths below the edge of the acromion process laterally. For this reason, the deltoid split should not extend too far distally, just as one would do when performing an open rotator cuff repair (Fig. 5.37).

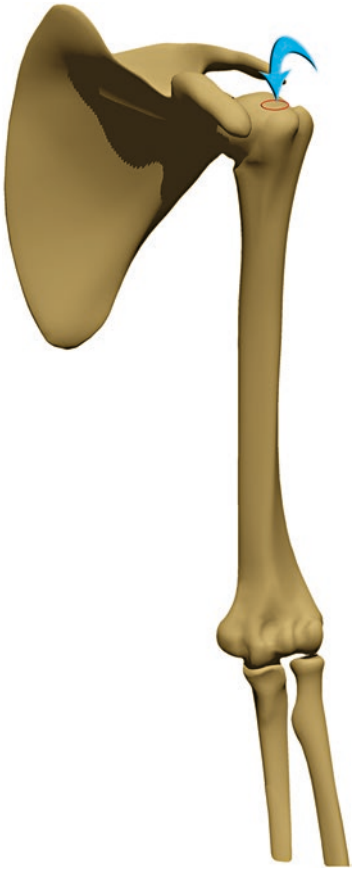


Fig. 5.35 The proximal humeral portal, *anterior* view. Copyright 2016 Zeeca Publishing Co

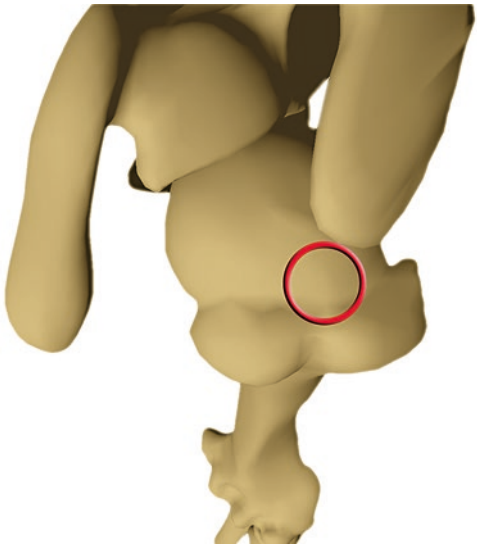


Fig. 5.36 The proximal humeral portal, *superior* view. Copyright 2016 Zeeca Publishing Co

Fig. 5.37 Proximal portal guide pin insertion. Note retractors in rotator cuff. Copyright 2016 Zeeca Publishing Co

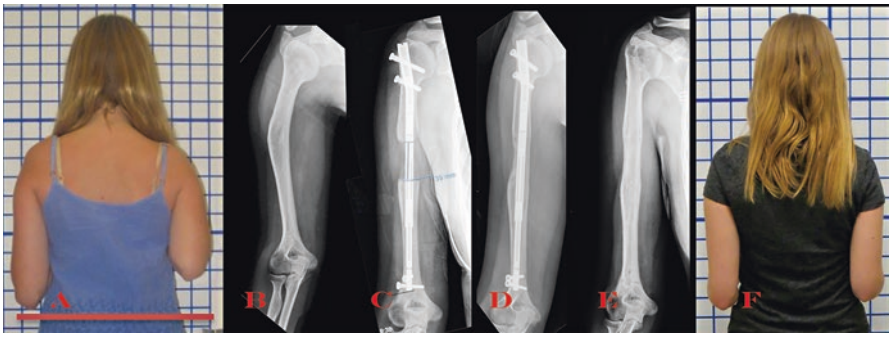


Fig. 5.38 Antegrade humeral lengthening. (**a**) Pre-op clinical condition. (**b**) Pre-op AP x-ray image. (**c**) Distraction phase image. (**d**) Consolidation phase image. (**e**) After nail removal. (**f**) Final clinical condition. Courtesy S. Robert Rozbruch, MD

The osteotomy level and vent holes for reamings must be selected with proper concern for the location of neurovascular structures in close proximity to the bone, especially the radial nerve, which rest directly on the bone's surface during part of its course.

Likewise inserting distal locking screws can damage the radial nerve in the lower arm. For this reason, open exposure and retraction of the nerve are recommended prior to transverse locking screw insertion.

Clinical Illustration: Humeral Lengthening via Proximal Portal

Intramedullary lengthening nails have yet to be approved for humeral lengthening. Nevertheless, surgeons, recognizing the improved patient experience with intramedullary lower extremity limb elongation compared to accomplishing the same objective with an external fixator, have been quick to use existing tibial and femoral nails in off-label applications to the humerus. This case, from the group at the Hospital for Special Surgery in New York, illustrates a combined lengthening and deformity correction of the humerus. The patient had a 6-cm humeral length discrepancy with an apex anterolateral deformity (Fig. 5.38). An 8.5-mm diameter femoral nail (215 mm length) was used off-label to achieve the correction.

The Distal Humeral Portal

The reported incidence of shoulder pain after intramedullary nailing of the humerus led surgeons to look for an alternative entry point for treating humeral shaft fractures with IM nails. The roof to the olecranon fossa, an undercut of the humeral shaft so to speak, seemed ideal for this situation. At first flexible nails were used, but gradually the technique for safe nail insertion from below was developed [10].

Fig. 5.39 Distal portal for insertion of retrograde humeral nail, located by splitting the triceps tendon. The radial nerve must be located and protected. Copyright 2016 Zeeca Publishing Co

The distal humeral portal presents certain anatomic challenges. Flexing the elbow to get a good line with the humeral shaft pulls the triceps tendon tightly over the portal; hence, the tendon must be split in line with its fibers (Fig. 5.39). Likewise, the radial nerve is pulled downward slightly by this maneuver, so it must be identified, looped, and protected during the procedure.

Because the entry site is along the shaft, a bent nail or straight may be used (off-label as of this writing). However, with substantial lengthening, at may be necessary to retract the nail back to its shortened configuration to remove it from the bone.

Clinical Illustration: Humeral Lengthening via Distal Portal

An off-label use of a straight femoral nail for humeral lengthening seems ideal for a volleyball player who wants to retain rotator cuff integrity to continue to play the sport after limb elongation to overcome humeral shortening caused by proximal unicameral bone cyst (Figs. 5.40, 5.41, 5.42, and 5.43).

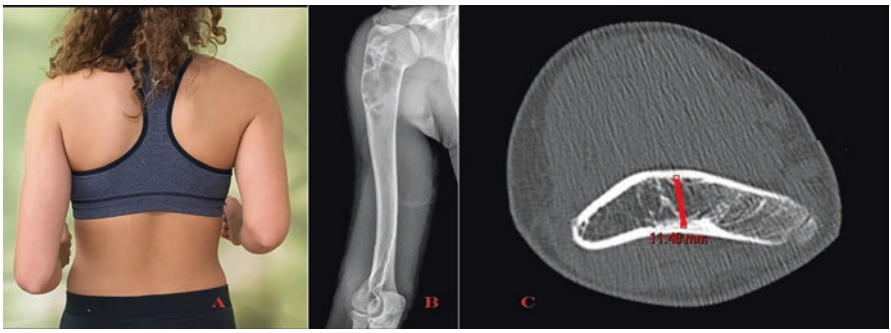


Fig. 5.40 Pre-op situation. (**a**) Right humeral discrepancy. (**b**) Pre-op x-ray showing unicameral bone cyst in proximal humerus. (**c**) AP dimension of distal humerus is large enough for a narrow femoral nail

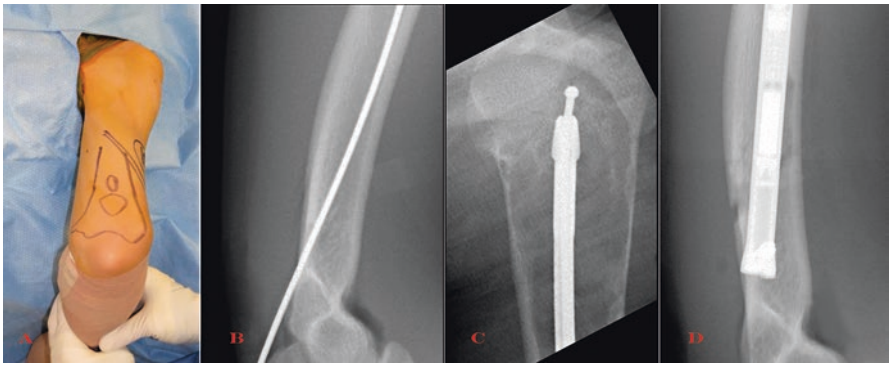


Fig. 5.41 Intraoperative images. (**a**) The distal portal is just above the olecranon fossa. (**b**) Guide wire I place. (**c**) Reaming up into unicameral bone cyst. (**d**) Straight femoral nail inserted. Notice that additional posterior humeral cortex had to be removed to fit the nail in place

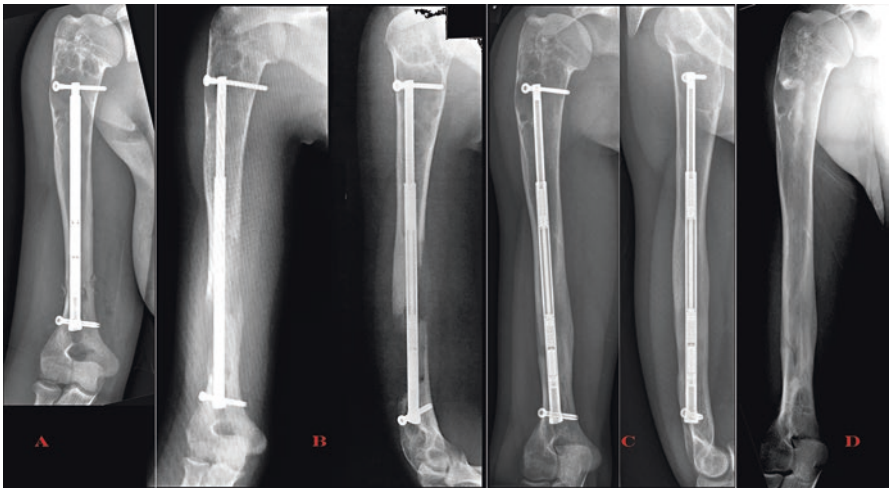


Fig. 5.42 Lengthening images. (**a**) After nail insertion. (**b**) AP and lateral views at end of lengthening. (**c**) AP and lateral views at end of consolidation. (**d**) AP view after nail removal and bone grafting of cyst and distal portal cortical window

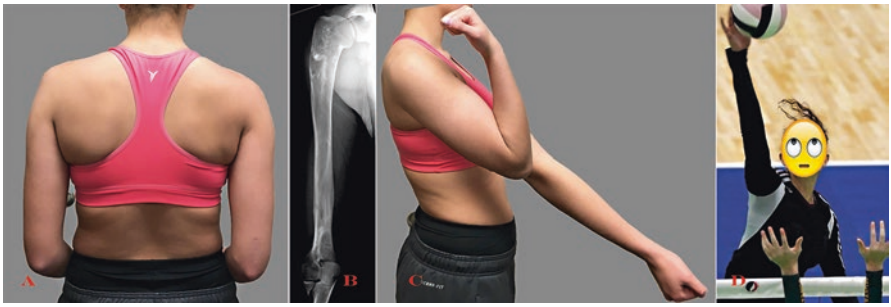


Fig. 5.43 Final condition. (**a**) Humeral lengths equal. (**b**) Bone cyst remodeled. (**c**) Range of motion. (**d**) Spiking a volleyball

Forearm

As of this writing, no intramedullary forearm lengthening nail has been created. Certainly, no nail with a ratcheting mechanism—like the Albizzia or ISKD nails—could be employed in the forearm because the radius would preclude counterrotation of the bones ends for elongation. As for motor-driven nails, such as the Fitbone or PRECICE nails, the small diameter of the forearm bones may make a telescopic nail impractical because the outer nail component would, of necessity, have a thin wall, whereas the internal telescopic portion of such a device must be robust enough to allow transverse screws of sufficient diameter to prevent breaking, a known complication with small diameter screws.

Therefore, any discussion of intramedullary forearm lengthening is not only off-label—it is off-design as well.

Thin intramedullary forearm nails and flexible pins have long been used as part of the treatment of forearm fractures. The portal is located at the flat proximal corner of the ulna after first splitting the triceps muscle and retracting it away from the bone. A straight intramedullary lengthening nail sits well in such cases, because the bone is straight (Fig. 5.44).

The radius is far less amenable to intramedullary nails, although a curved (or flexible) nail could conceivably be inserted through the radial styloid. After all, Rush pins have a long history with the radius. No antegrade insertion is possible in the radius for obvious reasons, only retrograde nailing through the styloid process.

Let us hope that implant engineers will someday offer surgeons a forearm lengthening nail, making this text section valuable indeed.

Reaming the Canal

In most cases, the exterior diameter of size-appropriate intramedullary lengthening nails exceeds the narrowest portion of internal diameter of long bone medullary canals. After all, an entire rotor and gear mechanism resides within the implant.

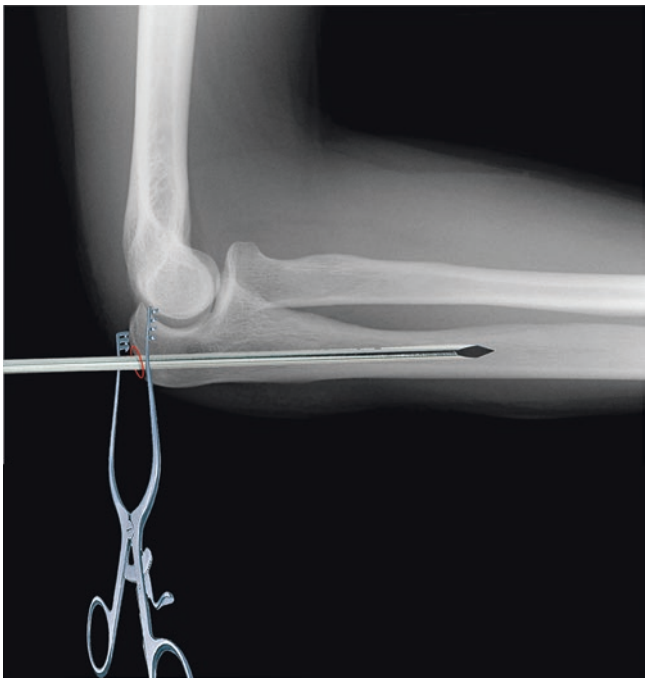


Fig. 5.44 Portal of insertion of intramedullary nail into ulna (antegrade). No internal lengthening nail has yet been devised for such a narrow canal. Copyright 2016 Zeeca Publishing Co

Moreover, while trauma nails should provide snug-fit stability across the zone of injury, lengthening implants risk jamming if implanted too tightly. Thus, unlike trauma applications where an unreamed nail can, at times, be inserted, canal reaming is an important feature of operative bone elongation with lengthening nails.

Hence, an intramedullary lengthening nail is better too loose than too tight within an intramedullary canal. For this reason, lengthening intramedullary nails should be inserted into channels that are over-reamed 1.5–2.0 mm larger that the nail diameter. The ability for the implant to slide freely up and down within the canal is more important than the actual canal diameter. In some situations this to-and-fro sliding motion is easily demonstrated. Straight nails, used for piriformis and distal femoral entry portals, can be pulled in and out to confirm canal glide. With bent nails—femoral trochanteric entry and all tibial nails—the implant typically feels snug as it turns distal in the canal, but it should glide smoothly thereafter.

Having said that, too much reaming is also undesirable. An over-reamed, eggshell-thin cortex may fracture during lengthening nail insertion, while the device is in place, or within days of removal—an especially disheartening experience for both patient and surgeon. For this reason, careful preoperative bone size measurements are essential to a successful intramedullary limb lengthening.

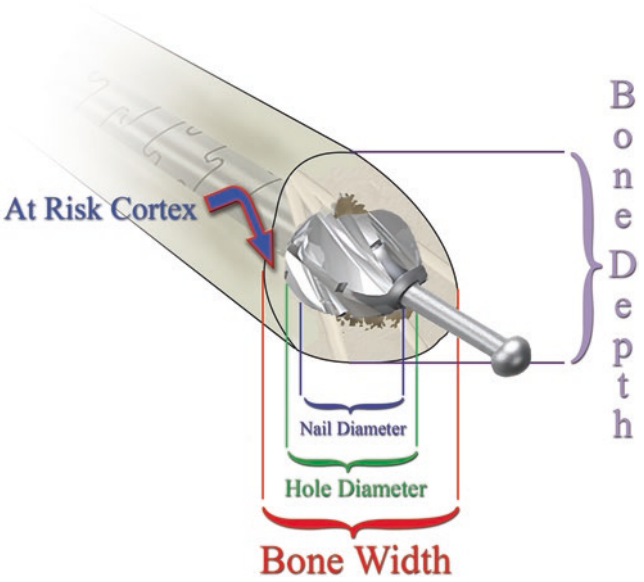


Fig. 5.45 Selection of nail size is constrained by the thickness of the thinnest cortical wall prior to reaming

The bone to be lengthening should be imaged in both the anteroposterior and lateral x-ray projection, with a magnification marker secured to the limb in the bone's projection plane. The imaging mAs and kVp settings should permit clear visualization of inner wall of the medullary canal. Analysis of the x-ray images will suggest the proper nail size (Fig. 5.45).

References

1. Moss RW. Histopathologic reaction of bone to surgical cutting. Oral Surg Oral Med Oral Pathol. 1964;17(3):405–14.
2. Sener BC, Dergin G, Gursoy B, Kelesoglu E, Slih I. Effects of irrigation temperature on heat control in vitro at different drilling depths. Clin Oral Implants Res. 2009;20(3):294–8.
3. Belthur MV, Paley D, Jindal G, Burghardt RD, Specht SC, Herzenberg JE. Tibial lengthening: extraarticular calcaneotibial screw to prevent ankle equinus. Clin Orthop Relat Res. 2008;466(12):3003–10.
4. Ostrum RF, Agarwal A, Lakatos R, Poka A. Prospective comparison of retrograde and antegrade femoral intramedullary nailing. J Orthop Trauma. 2000;14(7):496–501.
5. Johansson T, Risto O, Knutsson A, Wahlstrom O. Heterotopic ossification following internal fixation or arthroplasty for displaced femoral neck fractures: a prospective randomized study. Int Orthop. 2001;25(4):223–5.
6. Green SA. Distal intramedullary fixation of supracondylar fractures of the femur. Tech Orthop. 1988;3:71–7.
7. Henry SL, Trager S, Green S, Seligson D. Management of supracondylar fractures of the femur with the GSH intramedullary nail: preliminary report. Contemp Orthop. 1991;22(6):631–40.

8. Kim HJ, Fragomen AT, Reinhardt K, Hutson JJ, Rozbruch SR. Lengthening of the femur over an existing intramedullary nail. J Orthop Trauma. 2011;25(11):681–4.
9. Rommens PM, Kuechle R, Bord T, Lewens T, Engelmann R, Blum J. Humeral nailing revisited. Injury. 2008;39(12):1319–28.
10. Hollister AM, Saulsbery C, Odom JL, Anissian L, Garon MT, Jordan J. New technique for humerus shaft fracture retrograde intramedullary nailing. Tech Hand Up Extrem Surg. 2011;15(3):138–43.

Further Reading

1. Archdeacon M, Ford KR, Wyrick J, Paterno MV, Hampton S, Ludwig MB, et al. A prospective functional outcome and motion analysis evaluation of the hip abductors after femur fracture and antegrade nailing. J Orthop Trauma. 2008;22(1):3–9.
2. Andrzejewski K, Panasiuk M, Grzegorzewski A, Synder M. Comparison of knee function in patients with a healed fracture of the femoral shaft fixed with retrograde and antegrade intramedullary nailing. Ortop Traumatol Rehabil. 2013;15(5):395–405.
3. Augustin G, Zigman T, Davila S, Udilljak T, Staroveski T, Brezak D, et al. Cortical bone drilling and thermal osteonecrosis. Clin Biomech. 2012;27(4):313–25.
4. Gardner MJ, Robertson WJ, Boraiah S, Barker JU, Lorich DG. Anatomy of the greater trochanteric 'bald spot': a potential portal for abductor sparing femoral nailing? Clin Orthop Relat Res. 2008;466(9):2196–200.
5. Helmy N, Jando VT, Lu T, Chan H, O'Brien PJ. Muscle function and functional outcome following standard antegrade reamed intramedullary nailing of isolated femoral shaft fractures. J Orthop Trauma. 2008;22(1):10–5.
6. Kale SP, Patil N, Pilankar S, Karkhanis AR, Bagaria V. Correct anatomical location of entry point for antegrade femoral nailing. Injury. 2006;37(10):990–3.
7. Karmani S, Lam F. The design and function of surgical drills and K-wires. Curr Orthop. 2004;18(6):484–90.
8. Kuntscher G. Die Marknagelung von Knochenbruchen. Arch Klin Chir. 1940;200:443–55.
9. Kuntscher G. Practice of Intramedullary Nailing. Springfield, IL: Thomas; 1967.
10. Lee J, Ozdoganlar OB, Rabin Y. An experimental investigation on thermal exposure during bone drilling. Med Eng Phys. 2012;34(10):1510–20.
11. Lee J, Rabin Y, Ozdoganlar OB. A new thermal model for bone drilling with applications to orthopaedic surgery. Med Eng Phys. 2011;33(10):1234–44.
12. Misir AF, Sumer M, Yenisey M, Ergioglu E. Effect of surgical drill guide on heat generated from implant drilling. J Oral Maxillofac Surg. 2009;67(12):2663–8.
13. el Moumni M, Voogd EH, ten Duis HJ, Wendt KW. Long-term functional outcome following intramedullary nailing of femoral shaft fractures. Injury. 2012;43(7):1154–8.
14. Nam O, Yu W, Choi MY, Kyung HM. Monitoring of bone temperature during osseous preparation for orthodontic micro-screw implants: effect of motor speed and ressure. Key Eng Mater. 2006;321–323:1044–7.
15. Narasimha K, Osman MOM, Chandrashekhar S, Frazao J. An investigation into the influence of helix angle on the torque-thrust coupling effect in twist drills. Int J Adv Manuf Technol. 1987;2(4):91–105.
16. Oliveira N, Alaejos-Algarra F, Mareque-Bueno J, Ferres-Padro E, Hernandez-Alfaro F. Thermal changes and drill wear in bovine bone during implant site preparation. A comparative in vitro study: twisted stainless steel and ceramic drills. Clin Oral Implants Res. 2012;23(8):963–9.
17. Ostrum RF, Marcantonio A, Marburger R. A critical analysis of the eccentric starting point for trochanteric intramedullary femoral nailing. J Orthop Trauma. 2005;19(10):681–6.
18. Ricci WM, Schwappach J, Tucker M, Coupe K, Brandt A, Sanders R, et al. Trochanteric versus piriformis entry portal for the treatment of femoral shaft fractures. J Orthop Trauma. 2006;20(10):663–7.

19. Stannard JP, Bankston L, Futch LA, McGwin G, Volgas DA. Functional outcome following intramedullary nailing of the femur: a prospective randomized comparison of piriformis fossa and greater trochanteric entry portals. J Bone Joint Surg Am. 2011;93(15):1385–91.
20. Tu Y-K, Lu W-H, Chen L-W, Chiang C-H, Chen Y-C, Tsai H-H. Thermal contact simulation of drill bit and bone during drilling. In: 2010 4th International conference on bioinformatics and biomedical engineering. 2010. p. 1–4.
21. Tupis TM, Altman GT, Altman DT, Cook HA, Miller MC. Femoral bone strains during antegrade nailing: a comparison of two entry points with identical nails using finite element analysis. Clin Biomech (Bristol, Avon). 2012;27(4):354–9.
22. Wiggins KL, Malkin S. Drilling of bone. J Biomech. 1976;9(9):553–9.
23. Winquist RA, Hansen ST, Clawson DK. Closed intramedullary nailing of the femur: a report of five hundred and twenty cases. J Bone Joint Surg Am. 1984;66:529–39.

Preventing Complications During Limb Lengthening

6

Background

All limb lengthening operations and the majority of deformity correction procedures have potential to create secondary deformities in long bones, as well as contractures, subluxations, and dislocations of adjacent joints. This seems paradoxical; after all, how could an operation designed to correct a deformity cause one?

Certain biological tissues do not readily lengthen, whereas others do. For instance, nerves and blood vessels can be easily stretched with slow, steady traction, applied over a period of days, weeks, or even months. Indeed, this is how nerves are repaired in cases of traumatic segmental loss. When, for example, such a loss (a couple of inches) occurs in the anterior forearm, the surgeon bends the elbow sufficiently to approximate the nerve stumps and sutures them together. After preliminary healing of the suture line, the patient's elbow is gradually extended, at the rate of a couple of degrees each week, elongating the repaired nerve.

There are, however, structures of the limb that do not readily elongate, resisting traction. Tough bands of fascia are particularly hard to stretch, as are tendons. Muscle tissue, although somewhat softer, also resists elongation, both actively (by contraction) and passively, primarily through resistance of the perimysium, the membrane that surrounds individual muscle bundles [1].

In general, undesirable *bone* deformities during elongation are a consequence of resistance from longitudinal fascial bands within the limb, whereas secondary *joint* problems are the result of resistance to elongation by musculotendinous structures.

In both situations, knowledge of surrounding anatomy allows the surgeon to predict likely problems and to establish prophylactic strategies to prevent these problems from occurring.

As a principle, techniques designed to prevent the bone from undesirable angulation (or translation) during lengthening are incorporated in the surgical procedure, while methods of preventing joint contractures, subluxations, and dislocations are part of the postoperative regimen.

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_6

Operative Strategies to Prevent Bone Deformities During Lengthening

Bone Deformities During Lengthening

Whenever a surgeon performs limb elongation, the bone has a tendency to deform with its apex opposite to the thickest muscles or densest fascia. For this reason, experienced surgeons apply hardware in strategic locations to prevent such angulation during elongation.

At every bony level, whenever fragments are moving with respect to each other, typical patterns of deformity occur. The surgeon must be ever vigilant, with the aim of preventing such deformities from occurring if possible and dealing with the problems as they happen.

Tibial Deformation

When lengthening a tibia, for example, the power of the calf musculature has a tendency to produce knee flexion contractures (gastrocnemius), ankle equinus (gastrocnemius/soleus), and antecurvatum (apex forward) of the osteotomy site. Simultaneously, resistant anterior compartment components, and especially the interosseous membrane, can cause the deformity apex of a lengthening tibia to point medially. The combination of valgus and antecurvatum forces acting upon a tibia will result in deformation of the elongating bone with its apex anteromedial. Moreover, in view of the observation that thick fascial tissue contributes to angular deformities of long bones during lengthening, it is no surprise that the apex of the deformity of any elongating tibia is opposite to the attachment of the interosseous membrane on the posterolateral edge of the bone.

Femoral Deformation

In the thigh, the hamstrings and linea aspera generally cause the femur to angulate apex anteriorly during lengthening. The proximal femur is particularly prone to anterior angulation, as a product of flexion produced by the iliopsoas at the hip. In the coronal plane, the proximal femur tends to angulate into varus, due to the combined action of the hip abductors attached to the greater trochanter of the femur and the adductors inserting along the distal shaft. This combination of deformities results in a proximal femoral angulation with its apex directed anterolaterally.

An osteotomy of the distal femur may angulate toward either valgus (because of tension from the iliotibial band) or varus (the result of adductor muscle pull) during distraction.

The proximal humerus tends to angulate into varus (with an anterolaterally directed apex) because of the abductor strength of the rotator cuff and the action of

the medial upper arm muscles. Distally, the humerus usually angulates with the apex posteromedial.

Forearm Deformation

In the forearm, the ulna can angulate apex anterolateral, while a distal radial osteotomy may angulate apex medially. With forearm applications, however, muscle tension is likely to be insufficient to overcome the intrinsic resistance to deformation offered by an intramedullary nail.

Preventing Deformities (External Fixator Principles)

Ring Prophylaxis

Because bone deformities during elongation occur with every method of limb lengthening surgery, specialized techniques specific for each type of apparatus have evolved.

Ilizarov and co-workers were, obviously, the first surgeons to observe such problems. To mitigate these issues, they created a tension wire fixator technique called "ring prophylaxis." Based on experience and anatomic considerations, Ilizarov surgeons, when applying a circular frame for a simple longitudinal lengthening, do not mount the rings parallel to each other and perpendicular to the long axis of the bone, as is often shown in illustrations. Instead, they tilt the ring nearest the anticipated level of deformity in a manner that parallels such a deformity if it actually exists at the time of surgery.

In other words, as osteotomies of the upper tibia typically angulate during lengthening with the apex of the evolving deformity pointing anteromedially, the proximal ring of the configuration is tilted higher on the anteromedial corner and lower on the posterolateral corner. Since, in this position, the ring is no longer perpendicular to the longitudinal connecting rods of the frame, there must necessarily be hinges between the ring and the four longitudinal lengthening rods. Moreover, the rotation axes of each of these four hinges must be parallel with each other and perpendicular to the plane of the anticipated deformity.

Once lengthening begins, the patient is instructed to lengthen the short posterior and lateral distracting rods at a greater rate than the anterior and medial ones. In this manner, the hinged ring is gradually tilted downward in the anteromedial corner and upward in the posterolateral corner as elongation proceeds. Thus, the ring and its associated tension wires counteract the evolving deformity by correcting for it as it occurs.

Such ring prophylaxis is a characteristic feature of a properly applied, circular, tensioned wire, Ilizarov external skeletal fixator. The flexibility of Ilizarov's tension wires make such ring prophylaxis necessary. Without this prophylaxis, bones deform predictably during lengthening (Fig. 6.1).

Fig. 6.1 Ring prophylaxis with the Ilizarov method. The frame configuration anticipates the likely deformity during elongation, in this case the apex of deformity, and is tilted as though the deformity already exists (*left*). As the deformity evolves, the frame is gradually squared off (*middle*) to correct changes as they occur. At conclusion of the lengthening process, the frame is squared off, with the rings parallel and the deformity prevented (*right*). Copyright 2016 NuVasive

Strategic Pin Placement

With introduction of stiff half-pins as a substitute for flexible tensioned wires in many locations of an Ilizarov frame configuration, a technique developed by the author while working at Rancho Los Amigos Medical Center, one would think that the necessity for ring prophylaxis would be mitigated. Indeed, the "Rancho technique," as it is known, reduces the tendency for bone deformation during elongation, but does not eliminate it completely. However, the use of half-pins in many anatomic locations allows the surgeon to insert a pin on both sides of an osteotomy in the plane of an anticipated deformity during elongation. In this manner, for a deformity to occur during elongation, the bone must push directly into a transcutaneous implant placed to prevent that deformity. This is usually sufficient prophylaxis, if local neurovascular anatomy allows such pin placement.

The Waypoint Method

With the introduction of hexapod circular external fixation (i.e., the Taylor Spatial Frame), connection of the fixator to bone can be accomplished with either tensioned wires, half-pins, or a combination of both. Therefore, one would think that either ring prophylaxis or strategic pin placement would be required to prevent deformation during lengthening, depending upon the mounting components used to secure the frame to the bone.

Fig. 6.2 Deformity prevention with the waypoint method. The frame is mounted orthogonal to the bone with rings parallel to each other (*left*). As the deformity evolves, the angulation of the deformity is measured and entered into a computer containing the case parameters (*middle*). At the end of elongation, the rings are nonparallel but the deformity is corrected (*right*). Copyright 2016 NuVasive

As it turns out, however, the computer program used to create the prescription for daily strut length changes can also generate a modified prescription to deal with evolving deformities as they occur during limb elongation. This "waypoint" method of dealing with secondary bone deformities is similar to interim recalculation of a ship's planned route to account for changing seas and shifting winds (Fig. 6.2).

Corrections involve reentering the "deformity" parameters that define the evolving angulation and translation problem while maintaining the mounting parameters, existing strut lengths, and other data that went into the original prescription.

Needless to say, waypoint corrections can also be made with classic Ilizarov-type ring fixators and those with various modifications. Typically, however, such corrections require exchanging hinges placed at the apex of the evolving deformity for the longitudinal rods and the attachment of twisted plates and a distraction strut on the opposite side of the configuration; this exchange requires approximately 2 h of office time.

Biomechanical Axis Considerations

The objective of any lower extremity limb lengthening or deformity correction procedure is to maintain, restore, or achieve a natural biomechanical axis. While the details of deformity correction are beyond the scope of this publication, it is worthwhile to consider the effects of lengthening a bone along its own anatomic axis. After all, that is exactly what intramedullary lengthening devices do: they lengthen the marrow canal and surrounding cortex along the device.

Tibia

When elongating an already normally aligned tibia, the anatomic axis (a line following the center of the bone) and the biomechanical axis (a line passing through the middle of the femoral head, the middle of the knee joint, and the middle of the ankle joint) are nearly identical. Therefore, lengthening a tibia, whether using external or internal fixation means, typically maintains both the anatomical and biomechanical axes.

Femur

The femur, however, presents a far more challenging problem. In the coronal plane, the femur slants inward approximately 7° from vertical. Thus, from the anterior view, the anatomic axis of the bone makes a "V" with the biomechanical axis.

In the sagittal plane, when viewed laterally, the femur has a curving anterior bow, resulting in a curved anatomic axis. The biomechanical axis, however, goes straight down to the floor from the center of the femoral head.

Since it is nearly impossible to reproduce the curve of the femur with either external or intramedullary bone lengthening, elongation of the bone has different consequences when comparing external skeletal fixation to intramedullary lengthening devices.

With the external fixator, an osteotomy at the apex of the curve results in a straight segment between the curved fragments.

With an intramedullary lengthening device, an osteotomy at the apex of the curve tends to convert the femur into two half curves meeting at the osteotomy site once the implant is fully inserted. This has the effect of shifting the knee anteriorly at the end of lengthening, in the lateral view.

Sagittal Plane Malalignment

Shifting the mechanical axis of the femur anteriorly probably has little, if any, long-term effect, because, in general, the body is reasonably tolerant of joint deformities in the plane of function of that joint. Since the knee flexes and extends in a plane parallel to walking forward, angulation of the distal femur or upper tibia is better tolerated in the sagittal (laterally viewed) plane than in any other plane. The reason for this seemingly strange observation is that a joint typically uses only a small portion of its total range of motion in day-to-day functioning. Walking on level ground, for instance, involves only 30–40° of motion, whereas the total range of motion at the knee joint is approximately 135°. Therefore, reduced motion of the knee joint is well tolerated in Western culture, especially if the loss is at the extreme of flexion. In Eastern cultures, however, where food is eaten while kneeling or sitting cross-legged on the ground, and excretion is accomplished by squatting, loss of flexion can be a significant problem.

Angular malalignment of the distal femur or proximal tibia in an apex-posterior direction has the effect of increasing apparent knee extension and decreasing flexion. Since, as noted above, knee flexion loss is better tolerated than knee extension loss, a patient with such deformity, if not too great, compensates during gait by preventing full extension of the knee during midstance, assuming the presence of normal muscular control.

Angular malalignment of the distal femur or proximal tibia in an apex-anterior direction effectively reduces knee extension; however, as the calf normally contacts the thigh at full flexion, additional gain in flexion will not be realized. The lack of full extension during midstance causes a flexed-knee gait pattern, which has the effect of shortening the limb and resulting in an uneven gait pattern.

Frontal Plane Malalignment

Malalignment of the distal femur or proximal tibia in the coronal (frontal) plane is far more devastating. Here, varus or valgus angulation typically leads to gradual erosion and osteoarthroses of the weight-bearing cartilage of either the medial or lateral compartment of the knee, respectively. Moreover, as the cartilage and underlying bones erode away, the joint gradually assumes a progressive varus or valgus tendency. This stretches the joint capsule and ligaments opposite the narrowing, a cause of significant pain with activity. Indeed, some authorities consider joint instability and concomitant capsular ligament stretching to be the principal reason for pain in erosive osteoarthroses, overshadowing pain caused by bone-on-bone contact.

Because the femur slants inward when viewed anteriorly, the anatomic axis of the bone (its centerline) deviates approximately 7° from the biomechanical axis—a line from the center of the ball of the hip joint to the center of the ankle. This line also passes through the center of the knee or may be slightly medial.

Femoral Valgization During Elongation

Lengthening the femur along its anatomic axis, whether with intramedullary or external devices, pushes the distal end of the femur progressively medial, thereby increasing the valgus thrust of the knee during weight bearing. The concern, of course, is the potential for lateral compartment osteoarthroses of the knee, as the outer side of the knee bears a disproportionate share of the weight-bearing load.

It is estimated that for each centimeter of femur elongation, the valgus attitude of the knee increases by 1° (Fig. 6.3).

Correction with External Fixators

Ilizarov recognized the significance of increasing valgization during femoral elongation and compensates for this tendency in a unique manner. Because of the modularity of the Ilizarov apparatus, an Ilizarov femoral lengthening frame contains hinges that angulate the elongating regenerate into valgus at the upper end and then into a corresponding varus at the lower end. The net effect of these two complementary angles is to create a zigzag-looking femur with the regenerate new bone parallel to the biomechanical axis, while the upper and lower ends of the bone retain their original angular relation with the biomechanical axis. Hence, valgization of the knee is avoided.

During the 1970s, femoral lengthening with the monolateral external Wagner® device was a popular method of dealing with a unilateral short femur, whether caused by a traumatic growth arrest or a variant of proximal femoral focal deficiency called “congenital short thigh” [2]. The device lengthened the femur, after open osteotomy, at a rate of 1 mm/day in a single step (a parent turned a knob at the

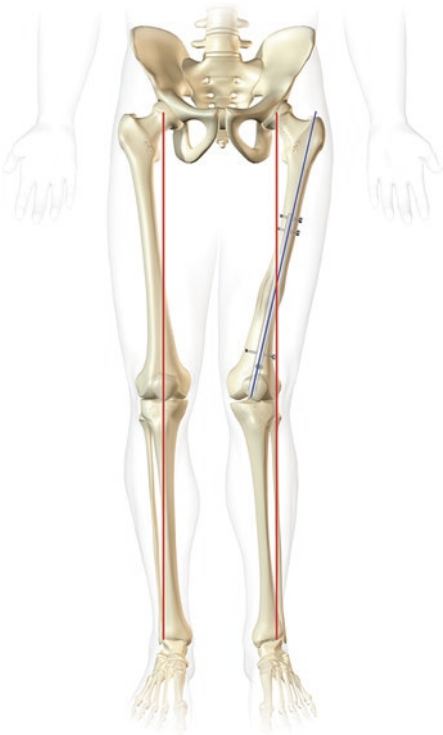


Fig. 6.3 Lengthening the femur along its central anatomical axis pushes the knee joint medially, toward the opposite knee, causing valgization of that knee and lateralization of the limb's biomechanical axis. Copyright 2016 NuVasive

end of the fixator for elongation). Regenerate new bone rarely formed with this protocol, except in very young children. Once the desired femoral length was achieved, the surgeon replaced the fixator with a sturdy internal plate and inserted an autogenous bone graft in the resulting distraction gap.

Because Wagner's fixator was mounted parallel to the femoral shaft, it elongated the bone along its anatomic axis (Fig. 6.4).

Follow-up research, performed years later, showed that many patients subjected to this procedure developed knee joint problems in later life [3].

Valgus Prophylaxis with Intramedullary Lengthening Nails

Of necessity, an intramedullary lengthening nail elongates a femur along the axis of the implant—the centerline of the marrow canal. Thus, as with the Wagner external fixator, the knee and distal femur is pushed medially during lengthening.

To overcome this problem, Baumgart, the developer of the Fitbone® self-lengthening nail, devised a distally based ("retrograde") nail insertion strategy to correct for the anticipated valgus angulation at the time of nail insertion. He calls this method "reverse planning," because Baumgart originally used paper cutouts of the femur and tibia to trace the path of the angular correction needed to restore the knee to its proper relationship with the biomechanical axis of the limb [4].

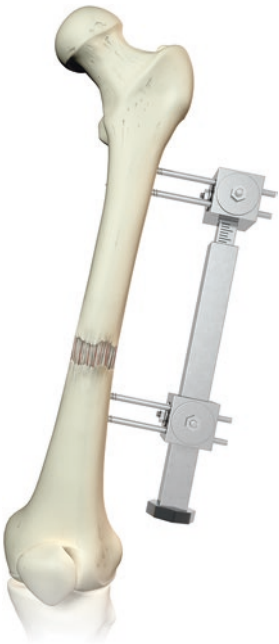


Fig. 6.4 The Wagner® fixator typically lengthened the femur along its anatomical axis. Copyright 2016 NuVasive

His method works with any intramedullary lengthening nail system and should be considered in any femoral lengthening over 3 cm (Fig. 6.5).

Planning the Correction

Preoperative planning is the hallmark of deformity correction surgery. Whereas tracing paper cutouts served well for pioneering Ilizarov surgeons around the world, nowadays digital x-ray studies save time and prove permanent, easily manipulated images to aid planning.

Bone Ninja and Multiplier, created by the limb lengthening team at Sinai Hospital Baltimore, can be downloaded to cell phones and iPads.

TraumaCad is another planning program with which we've had good experience (Fig. 6.6).

Preventing Deformities During Lengthening (Intramedullary Lengthening Nail Principles)

Background

Intramedullary lengthening nails are becoming increasingly popular nowadays because they greatly simplify postoperative management. Indeed, pin tract infections are a thing of the past with such devices. Likewise, muscle impalement from

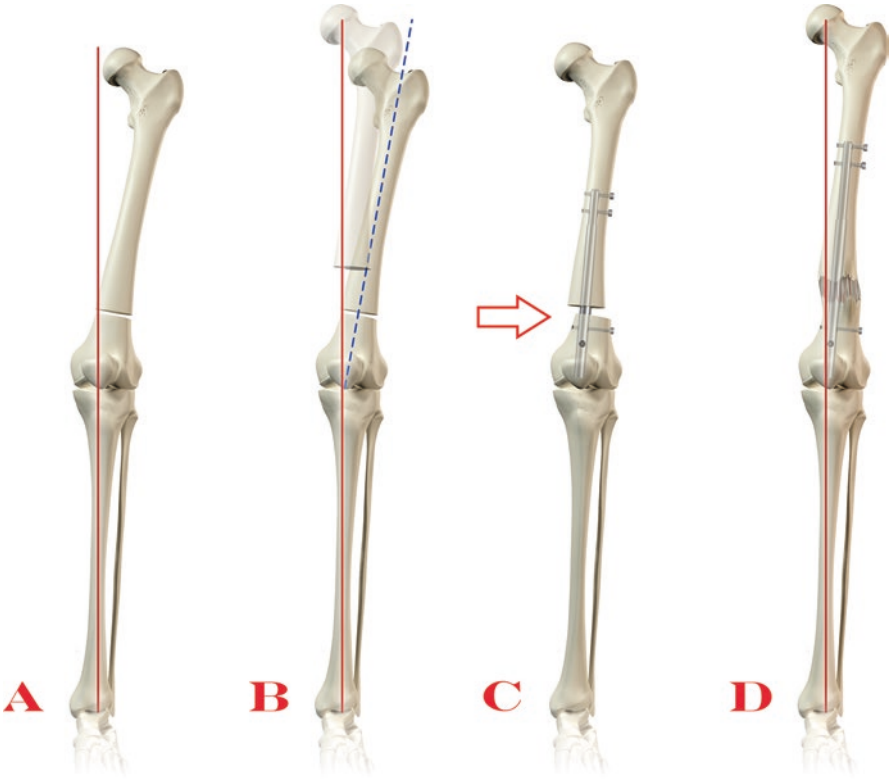


Fig. 6.5 The Baumgart reverse planning method for preventing valgization of the knee during intramedullary femoral lengthening. With paper cutouts or a computer program, transect the femur image at the planned level (**a**). Place the center of the femoral head along the upward extension of the biomechanical axis at the planned final length (**b**). Next, slide the shaft fragment down the biomechanical axis back to the osteotomy level, indicating the offset needed to correctly lengthen the bone (**c**-*arrow*). The completed elongation will have a zigzag in the shaft, but the alignment will be perfect (**d**). Copyright 2016 NuVasive

transcutaneous implants is eliminated, making postoperative physical therapy and activities of daily living much easier for the patient.

Intramedullary lengthening nails, in spite of their obvious advantages, do not eliminate deformities of the bone during elongation, nor do they reduce the likelihood of joint contractures, subluxations, and dislocations. After all, thick fascial bands and powerful muscular tendinous units do not melt away when exposed to a self-elongating nail.

In certain locations, where a snug fit exists between the intramedullary lengthening device and the surrounding cortex, angular and translational deformities are impossible. There is simply no place for the bone to go.

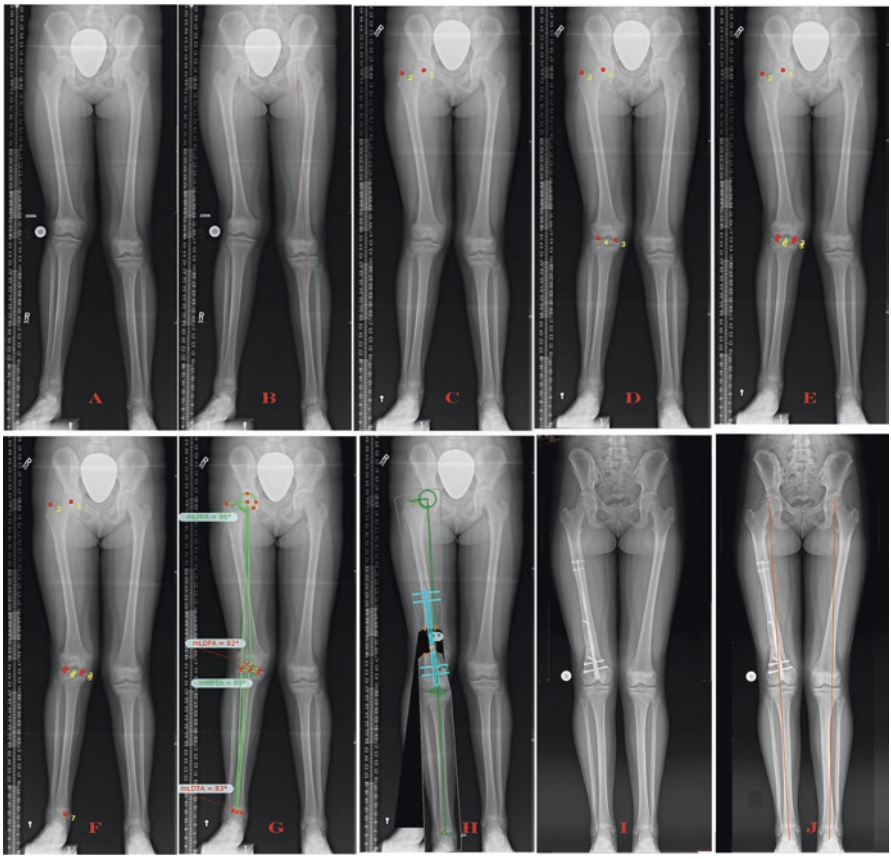


Fig. 6.6 Pre-op planning with TraumaCad software. (**a**) Initial condition, with leveling lift under short limb. (**b**) Center-of-hip to center-of-ankle lines. (**c**) Proximal femur landmarks located. (**d**) Distal femur landmarks identified. (**e**) Proximal tibia landmarks identified. (**f**) Distal tibia landmarks identified. (**g**) Shoe lift removed and Baumgart reverse planning lines drawn. (**h**) Osteotomy planning and implant location identified. (**i**) After osteotomy and lengthening. (**j**) Final biomechanical axes

Blocking Screws

As the cortex of the bone trumpets out in the metaphyseal region, however, the potential for angular deformity becomes a real concern, in spite of the presence of transverse locking screws that seemingly secure the bone fragment to the nail. In certain locations, for instance, where transverse locking screws are perpendicular to each other, the bone fragment can pivot on the screw perpendicular to the plane of likely deformation. The fragment will gradually displace in a direction opposite the thickest, most resistant, fascial band, until the cortex of the bone butts up against the nail (Fig. 6.7).

Fig. 6.7 Common deformity occurring during tibia lengthening, with apex anteromedial. Copyright 2016 NuVasive

Preventing such displacement is relatively simple: use blocking screws [5, 6]. In most cases, the blocking screws are placed in locations that appear, at first glance, counterintuitive. In the upper tibia, for example, such screws are placed adjacent to the intramedullary nail on the side *opposite* the apex of the likely deformity.

Thus, since proximal tibial deformities during lengthening point with their apex anteromedial, blocking screws are placed on each side of the osteotomy along the posterolateral surface of the nail. In this manner, the bone fragments cannot translate or angulate more than a fraction of a millimeter without butting up against the blocking screw. In a sense, we view the nail as a stationary object and the surrounding bone fragment as a moving cylinder. Blocking screw placement thereby stops the cylinder from translating or angulating.

Rather than a single blocking screw inserted perpendicular to the plane of likely angulation, many surgeons use two such screws at right angles to each other—one in the coronal plane and the other in the sagittal plane. The blocking, however, is just as effective (Figs. 6.8, 6.9, 6.10, and 6.11).

In some situations, the direction of angulation during lengthening is unpredictable. In such cases, it is wise to put a blocking screw on both sides of the intramedullary nail, or even on all four sides, at the time of surgery, rather than subject the patient to a return trip to the operating room.

Fig. 6.8 Deformity without blocking screws (*left*) prevented by placing blocking screws adjacent to the bone on the side *opposite* the apex of the likely deformity (*right*). Copyright 2016 NuVasive

Clinical Illustration: No Blocking Screws

It is hard, at first, to understand how a bone fragment securely fixed to the end of an intramedullary nail with two transverse locking screws could possibly angulate in the coronal plane, but it happens all the time (Fig. 6.12). Typically, the displacement occurs gradually, at the cancellous end of the bone. Careful assessment with high-quality imaging studies will often show a halo of radiolucency around either the transverse screws or the nail or both.

In this sense, loosening of the lengthening implant is no different from loss of fixation associated with plates and screws, external fixation pins, and total joint components. High local strain on the bone—a strain-intolerant tissue—stimulates bone resorption by osteoclasts, mostly likely via piezoelectric signals generated by bone's strained calcium hydroxyapatite crystals [7]. The resultant bone deficiency is filled in by loose granulation tissue, a substance far more strain tolerant and much softer.

Once a thin margin of bone is absorbed away in this manner, the implant presses against the next layer of bone, starting the cycle over again. The process continues until the implant is floating loosely.

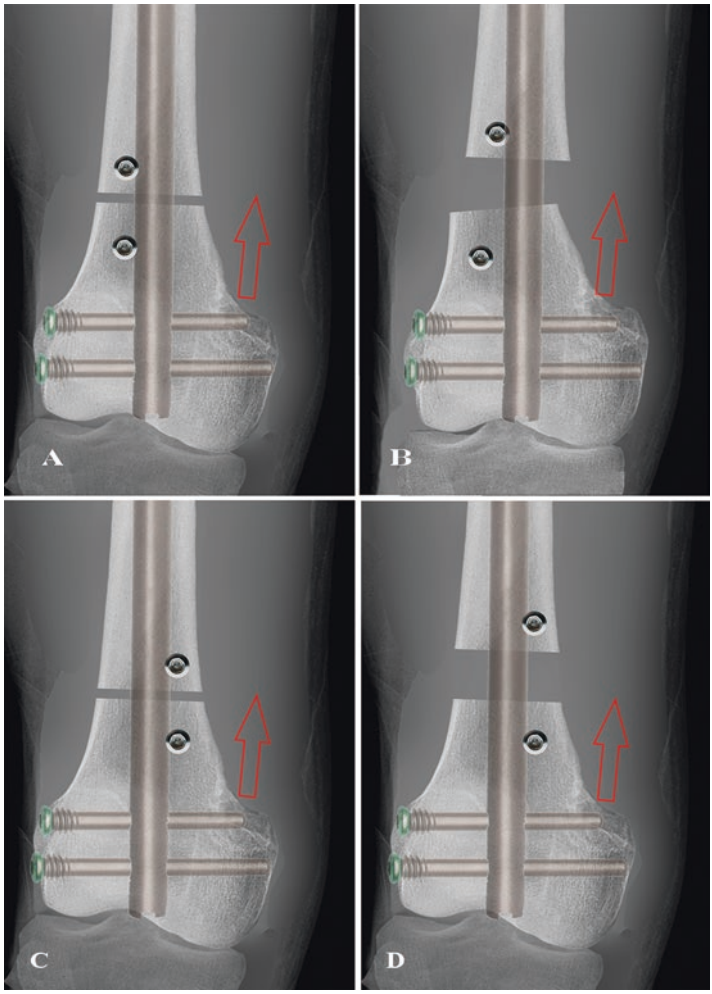


Fig. 6.9 Incorrect and correct blocking screw placement. Tension of the adductors (*red arrows*) will cause apex lateral angulation of the distal femur during lengthening. (**a**, **b**) Placing the blocking screws on the side of the apex of the likely deformity will not prevent the deformity. (**c**, **d**) Placing the blocking screws on the side of the nail opposite the apex of the likely deformity will prevent the deformity during lengthening. Copyright 2016 Zeeca Publishing Co

With these considerations in mind, the most logical way to reduce implant-induced strain on bone is to restrict weight bearing. This measure will not only reduce osseous absorption at the implant-bone interface but will also diminish the risk of implant breakage.

Decreased weight bearing, one might argue, prevents the regenerate from maturing into solid bone. After all, mineralization of the regenerate responds to piezoelectric stimulation generated during weight bearing. In our experience, however, a rhythmic, one step per second pace of supported ambulation—with a cane, crutches, or a walker—is more important than the actual pounds put on the ground.

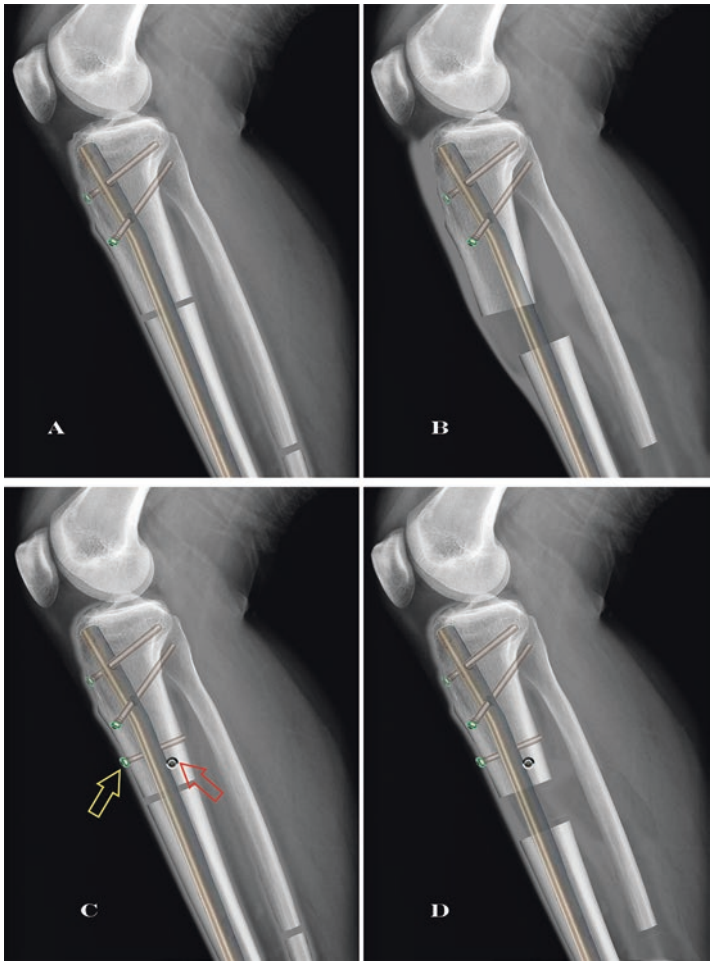


Fig. 6.10 Biplanar blocking screws. (**a**, **b**) Without blocking screws, the proximal fragment of a tibial lengthening angulates anteriorly and medially due to tension of the interosseous membrane. (**c**, **d**) Placing blocking screws posterior (*red arrow*) and lateral (*yellow arrow*) to the nail prevents the angulation. No screws are needed in the distal fragment because the nail fits snugly, so displacement cannot occur. Copyright 2016 Zeeca Publishing Co

Clinical Illustration: Secondary Blocking Screws

Omitting blocking screws will often lead to deformity of a lengthening bone, as described above. In many cases, the problem can be corrected with delayed blocking screw insertion before the regenerate hardens. The 13-year-old girl in this clinical illustration had idiopathic tibial shortening without angular deformity. A Fitbone® was inserted without blocking screws. The distal end of the proximal fragment angulated anteromedial during lengthening, causing the limb to deviate into

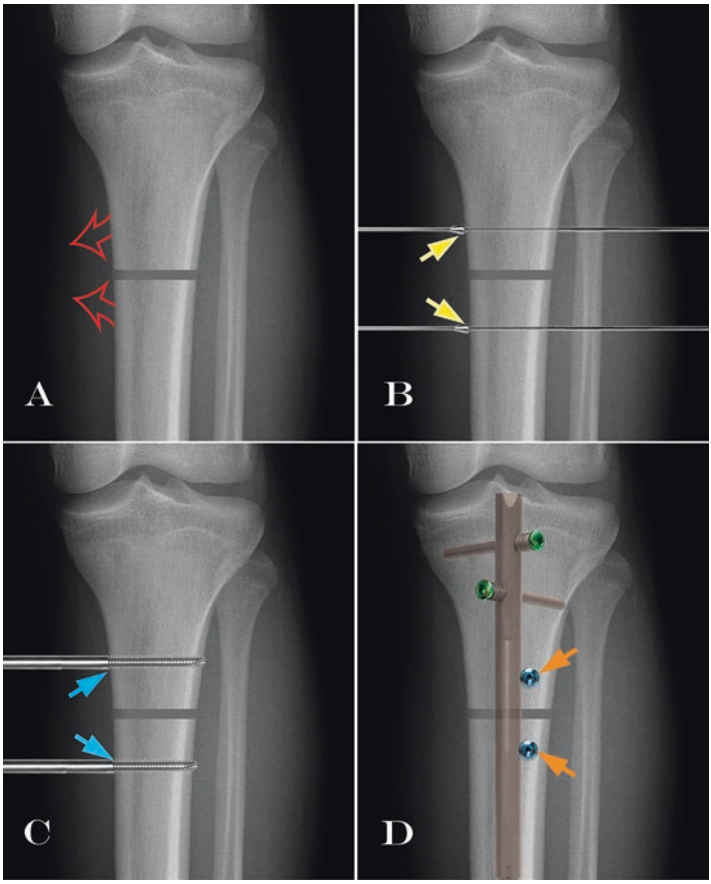


Fig. 6.11 Comparative strategies to prevent medial angulation of the tibia during lengthening. (**a**) The tibia tends to angulate anteromedially (*red arrows*) during lengthening, due to tension of the interosseous membrane. (**b**) With tensioned wire fixation, beaded olive wire (*yellow arrows*) on the medial side of the bone (*cyan arrows*) blocks angulation. (**c**) With half-pin fixation, the shoulders of half-pins on the medial side of the bone block angulation. (**d**) With intramedullary nail lengthening, blocking screws (*orange arrows*) on the lateral side of the nail block angulation. Copyright 2016 Zeeca Publishing Co

valgus deformity. The patient was brought to surgery where blocking screws were inserted, while thumb pressure reduced the displacement. Proper alignment was restored (Fig. 6.13).

Clinical Illustration: One Blocking Screw

When in doubt about whether or not to use blocking screws, use them. In this illustration (Fig. 6.14), a varus femur deformity required both acute angulation correction and translation at the osteotomy site directed by the principles of Baumgart

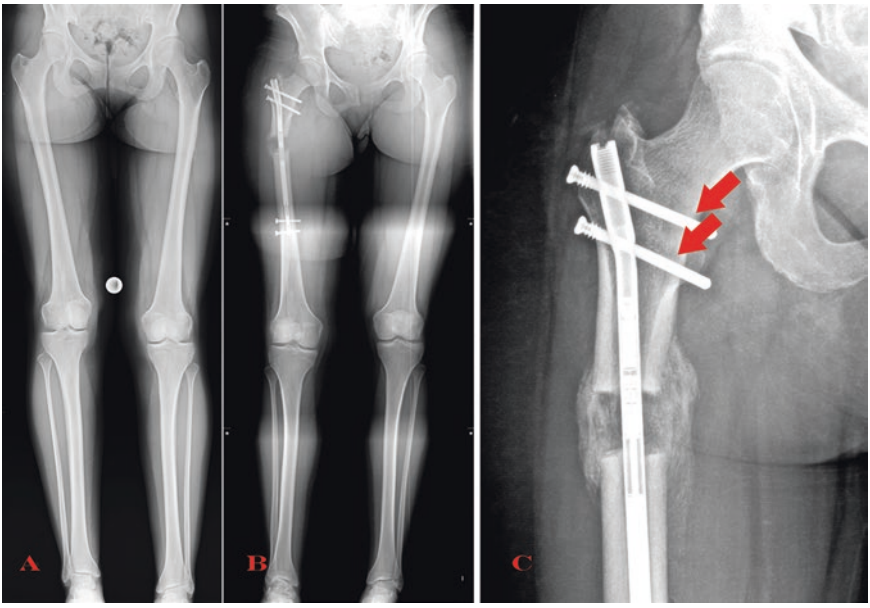


Fig. 6.12 Varus deformation during proximal lengthening. (**a**) Pre-op condition, shortened right femur. (**b**) Lengthening complete, apex lateral (varus) angulation of proximal fragment. (**c**) Close-up: note osteopenia halos around the transverse locking screws (*red arrrows*)

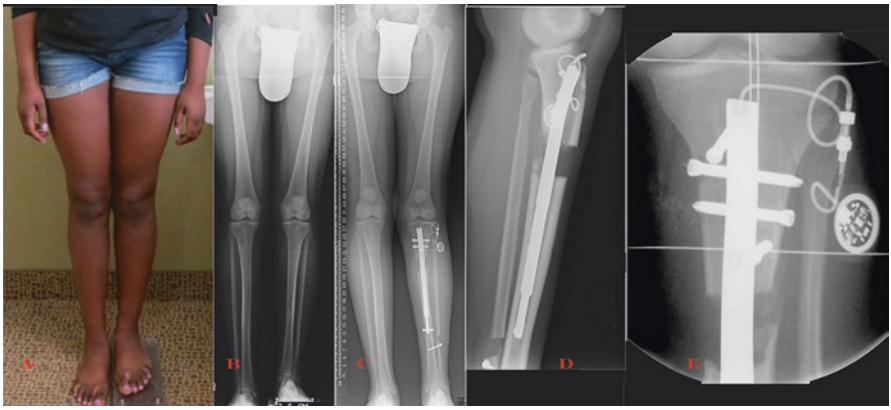


Fig. 6.13 Delayed blocking screw insertion. (**a**) Idiopathic unilateral tibial shortening. (**b**) Standing AP x-ray image with lift to level pelvis. (**c**) Progressive valgus deformity. (**d**) Anterior displacement of proximal fragment. (**e**) Corrected with secondary blocking screws

reverse planning method. A snug fit of the nail in the proximal fragment, a feature associated with the use of a straight reamer and over-reaming only 0.5 mm, eliminated the need for a blocking screw in the proximal fragment.

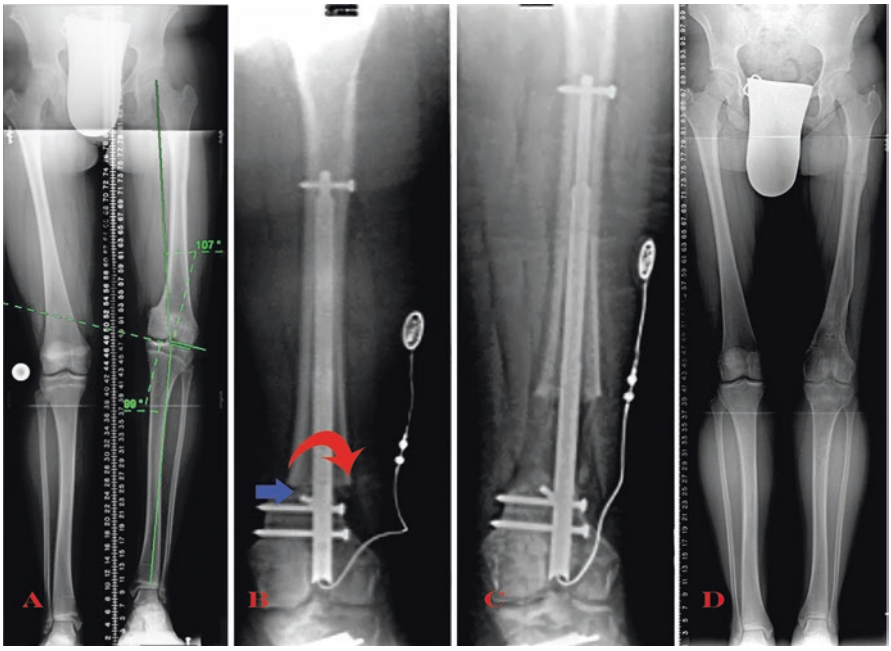


Fig. 6.14 Use of blocking screw to prevent displacement during lengthening. (**a**) Pretreatment situation. (**b**) Fitbone® lengthening nail in place. Note blocking screw on lateral side of nail (*blue arrow*) to prevent rotation of distal fragment (*red arrow*). (**c**) At end of lengthening. (**d**) Result

Notice how the biomechanical axis is correct at the end of treatment even though there is a lateral bump on the bone when fully consolidated.

Clinical Illustration: Many Blocking Screws

If one blocking screw is good, then more are better. In this illustration, we were not sure which way the distal femur would tilt during lengthening, so blocking screws were inserted on both sides of the nail in the distal fragment. In the proximal fragment only one screw is inserted. Notice, however, that this screw presses the nail against the medial cortex of the femur, which functions as the medial stabilizer (Fig. 6.15).

Blocking Screws as Guidance Screws

The primary purpose of blocking screws is self-evident: prevent osseous deformities during limb elongation. However, such screws can also serve to aid positioning of the intramedullary nail within the canal. In this situation, the screws are inserted *before* the canal is reamed.

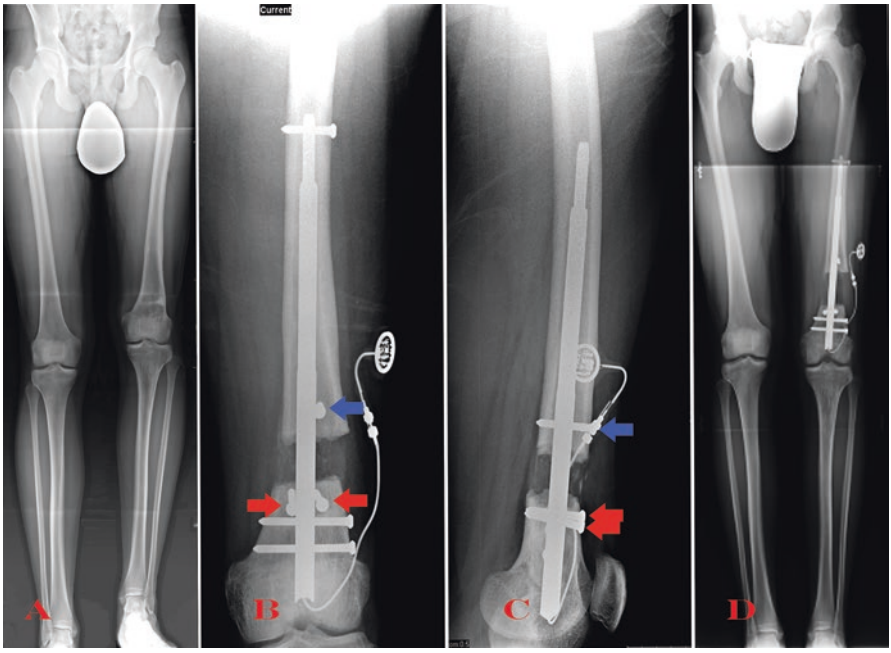


Fig. 6.15 Multiple blocking screws for complete control in sagittal plane. (**a**) Pre-op situation, 5 cm short left femur. (**b**, **c**) AP and lateral images with two distal screws (*red arrows*) and one proximal lateral blocking screw (*blue arrow*). Notice that the proximal screw traps the femur against the medial cortex of the proximal fragment. (**d**) Alignment at the end of lengthening

The surgeon must mentally visualize the desired path of the nail as well as the likely trajectory of the reamer or nail if the guiding screws were not present. The screws are placed in strategic locations to guide the reamer and nail into the desired position. If judiciously done, the guidance screws can be left in place, after inserting the lengthening nail, to act as blocking screws (Fig. 6.16).

Drill Bits for Guidance

Considerable experience is required to properly place guidance screws, so they aid nail insertion but do not, themselves, block the implant during insertion. Likewise if such screws are to be left in place to act as blocking screws, the screwnail fit must be quite snug, lest the bone shift somewhat before the screw blocks further displacement during elongation of the limb. Also, the sharp blades of a reamer can scratch guidance screws, perhaps rather significantly.

For these reasons, we recommend using drill bits for nail guidance, rather than screws. Drill bits can be easily repositioned if necessary. Being temporary, they tolerate damage from the reamer (Figs. 6.17 and 6.18).

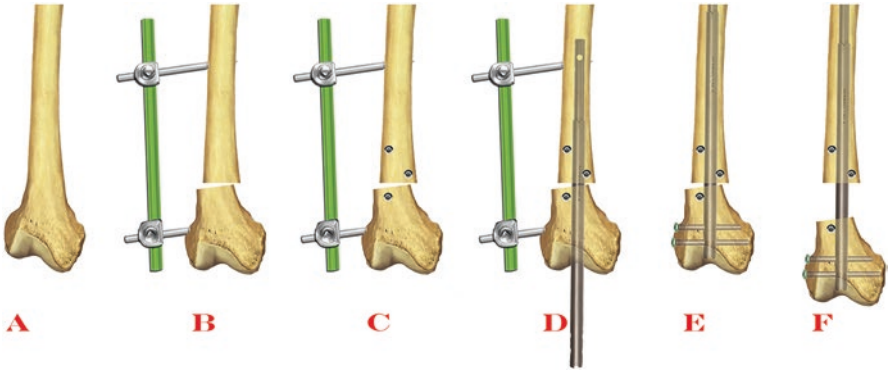


Fig. 6.16 Blocking screws as guidance screws. (**a**) Valgus deformity due to partial lateral growth arrest. (**b**) Fixator-assisted correction of deformity. (**c**) Blocking screws inserted as guidance screws. (**d**) Nail path determined by screws. (**e**) Fixator removed in operating room. (**f**) Guidance screws become blocking screws to maintain alignment of correction. Copyright 2016 Zeeca Publishing Co

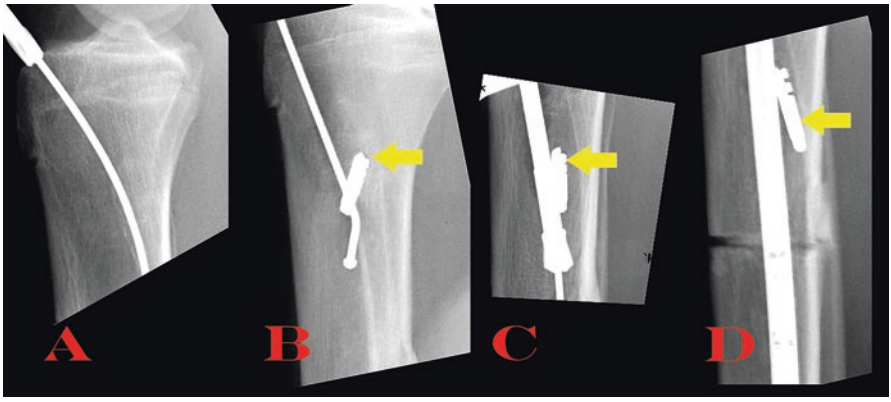


Fig. 6.17 Drill bit for guidance. (**a**) The guide wire's trajectory will likely cause the reamer to scrape along the posterior cortex of the tibia. (**b**) A drill serves (*yellow arrow*) as a guide for the bent-tip guide wire. (**c**) The reamer is directed away from the posterior cortex by the drill bit. (**d**) Lengthening nail in place. The drill bit will be replaced by a blocking screw

Regenerate Enhancement with Blocking Screws

In the 1960s, G. A. Ilizarov conducted experiments with a canine model to assess the importance of stability to the quality of regenerate new bone formation in a widening distraction gap [8]. He applied circular fixators of increasing stability to lengthen limbs of dogs. He learned that the greater the stability, the better the quality of new bone formation during distraction. Looking at his experimental design, however, the most prominent plane of instability that Ilizarov actually studied was translational shear at the osteotomy site.

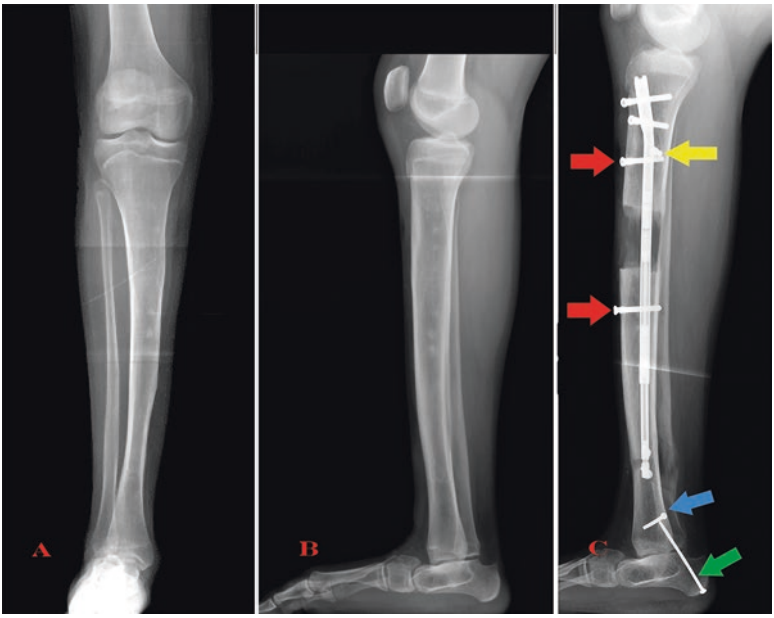


Fig. 6.18 (**a**, **b**) Pre-op clinical situation of patient is previous figure. (**c**) During lengthening. The guidance drill bit has been replaced by a blocking screw (*yellow arrow*) in coronal plane. Two additional blocking screws have been added in the sagittal plane on the lateral side of the nail (*red arrows*) to prevent apex medial angulation. No distal coronal blocking screw is necessary because the nail is flush against the cortex. Note the distal tibiofibula screw (*blue arrow*) and the calcaneal-tibial screw to prevent equinus (*green arrow*)

We have concluded, based on our own clinical experience, that an intramedullary lengthening nail may not eliminate translational shear at the osteotomy level because the canal width typically exceeds the implant's diameter, at least on the metaphyseal side of the construct. Indeed, time-lapse photograph taken by the senior author and staff during early sheep and goat research on the PRECICE® nail clearly demonstrate slight back-and-forth translational motion of the osteotomy site during elongation of the bone.

Therefore, it stands to reason that the judicious use of blocking screws, by eliminating wobble of the cut bone ends, may enhance regenerate formation, leading to earlier maturation and consolidation of the newly formed bone.

Excessive Correction

The Osteotomy Site

The favorable experience patients have with intramedullary lengthening—especially when compared to their prior experience with external fixator limb elongation—has caused surgeons to incorporate deformity correction into limb lengthening

protocols using remotely controlled nails. Since the osteotomy site is the only place available for such corrections, surgeons have been expanding the envelop of indications for such operations.

Angulation

As mentioned earlier in this monograph, it appears that the femur is more tolerant than the tibia to acute angular changes at the time of intramedullary lengthening nail surgery. Although the question has not yet been resolved in the medical literature, we recommend limiting angulation correction in the tibia to 10°degrees or less, and in the femur, 15° or less. Correcting more than these values likely stretches the periosteum and may damage its blood supply. Since reaming for the nail destroys the endosteal circulation, ischemic periosteal necrosis has catastrophic consequences for regenerate formation.

Translation and Rotation

There are two other displacements that can be corrected at an osteotomy site: rotation and translation. As with angulation, too much displacement in either of these planes risks periosteum strangulation and reduced regenerate formation (Figs. 6.19, 6.20, and 6.21).

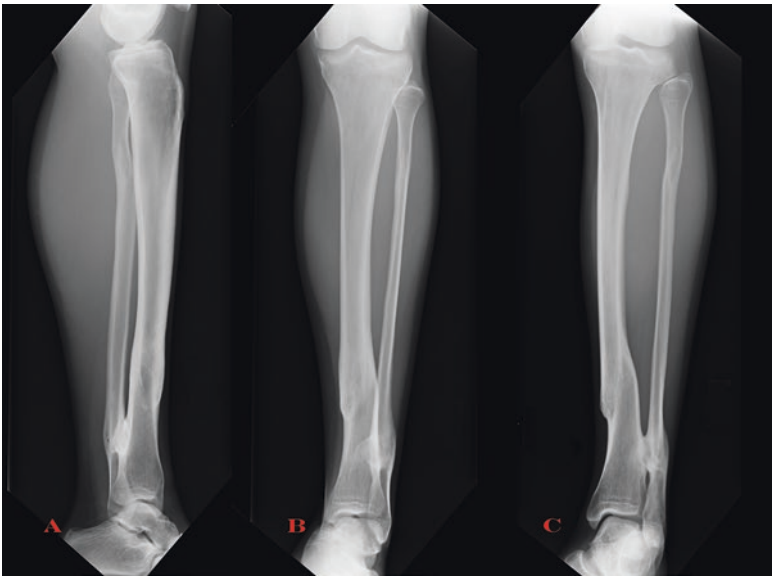


Fig. 6.19 Pre-op situation. (**a**) Lateral view of deformity. (**b**) AP deformity. (**c**) Oblique view of deformity demonstrates true lateralward translation of distal tibia

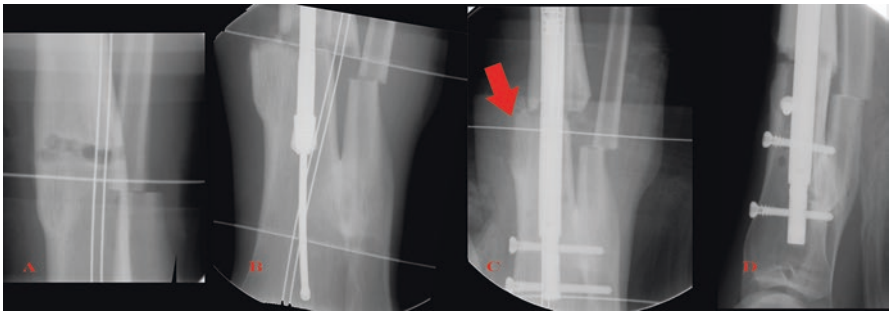


Fig. 6.20 Attempted correction of translation deformity. (**a**) Vent holes at osteotomy level. (**b**) Reaming correctly aligned tibia. (**c**) Lengthening nail inserted. *Red arrow* points to prominent medial tibial cortex tenting skin. (**d**) Prominence shaved off

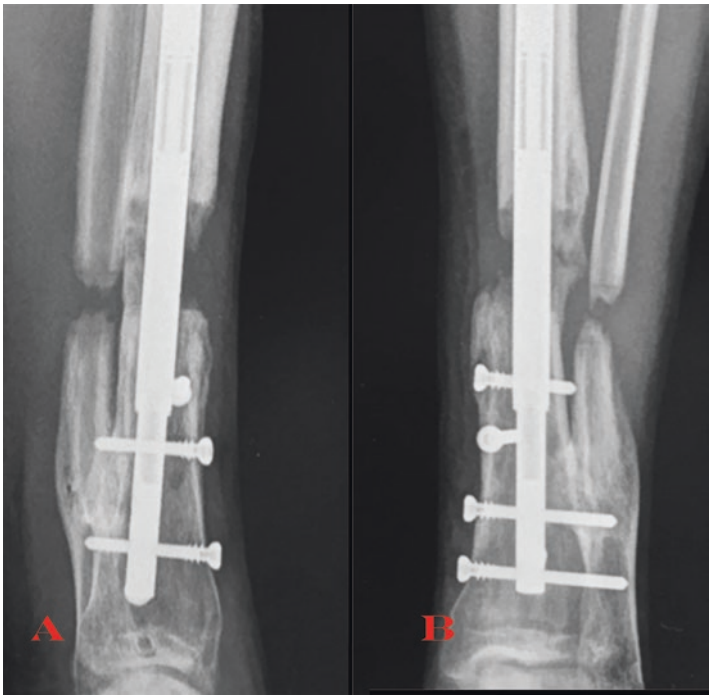


Fig. 6.21 Poor regenerate formation. (**a**) Lateral view. (**b**) AP view

Angulation, Translation, and Rotation

We should never lose sight of the basic tenet of Ilizarov's method: create a non-displaced fracture with a "sparing corticotomy" (preserving both endosteal and periosteal blood supply), which is allowed to begin preliminary healing in an axially

dynamic—but otherwise stable—circular external fixator. Using an intramedullary lengthening nail eliminates the endosteal blood supply, and attempting to correct a deformity though an osteotomy site changes what should have been a non-displaced fracture into something entirely different—a displaced fracture with non-congruent bone ends.

Likewise, the dynamics of the bone's mechanical environment are different with intramedullary lengthening when compared to Ilizarov's frame. Tensioned wires act like an axially lively trampoline at both ends of the Ilizarov's configuration, while beaded wires limit translation at the osteotomy site.

An intramedullary lengthening nail offers the opposite environment: an axially stiff construct that often permits slight translational wobble at the osteotomy level. Under the circumstance, it's a wonder that distraction osteogenesis with an intramedullary nail matures and consolidates at all—but it does.

Any displacement at the osteotomy level that deviates from Ilizarov's dictum to create a non-displaced corticotomy will likely retard regenerate biology. Moreover, if excessive angulation, rotation, or translation inhibits regenerate formation, displacement in all three planes may be too much strain on the microcirculation of the periosteum to support any bone formation in the distraction gap (Fig. 6.22).

At the very least, with any amount of displacement of the bone ends from each other at the osteotomy level, lengthen the latency interval (delay before distraction) to 2 or 3 weeks, reduce the daily lengthening amplitude to 0.5 or 0.6 mm per day, and, if practical, increase the fractional rhythm to q4h or q2h. Based on clinical experience with fractures and osteotomies in general, we have concluded that the more distal the osteotomy in the tibia, the more likely will be tardy regenerate ossification in all circumstances, but especially when there is a correction of malalignment in any plane.

Finally, keep this in mind: A lengthening intramedullary nail cannot substitute for a circular external fixator to correct substantial deformities. Either use a

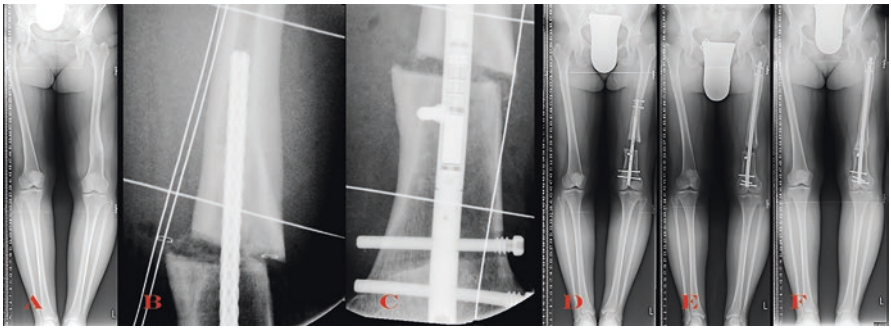


Fig. 6.22 Too much correction. (**a**) Initial condition, with angular, translational, and rotation deformities of left femur. (**b**) Reaming after alignment with intraoperative temporary fixator. Note the displacement of osteotomy site. (**c**) PRECICE® nail in place. (**d**) Minimal regenerate formation. (**e**) Established non-union 10 months after end of distraction. (**f**) Healing with trauma nail

multiplanar or hexapod fixator or correct the deformity first with internal fixation (typically a subtraction closing wedge osteotomy), with a second surgery to achieve lengthening.

Latency, Rate, and Rhythm Modifications

Rationale

As Ilizarov surgeons, we naturally assume that the standard distraction rate of 1.0 mm per day in four doses of 0.25 mm every 6 h is a reasonable rate and rhythm of limb elongation for lengthening intramedullary nails. As emphasized repeatedly in this volume, such a prescription should be modified in many situations and perhaps in all tibial lengthenings, with 0.5 or 0.6 mm/day (in divided doses) being more appropriate.

A surgeon has three variables in the postoperative management of a patient that deserve consideration: the latency interval, the rate of distraction, and the rhythm of distraction.

Latency

Latency, to reiterate, is the time interval between surgery and commencement of distraction. While in pediatric cases, we typically start lengthening at 5–7 days, there is no such need for a rush in adults. It's hard to image any osteotomy pattern, especially one followed by a canal-filling intramedullary nail, that could be at risk of premature consolidation if the surgeon decides to wait 10–14 days or even longer, up to 3 or even 4 weeks, before commencing limb distraction.

Therefore, the surgeon should consider prolonging latency if the osteotomy caused substantial displacement of the bone ends with respect to each other during surgery (even if only temporary), if there was *any* amount of deformity correction immediately after the osteotomy. With the substantial corrections, or those involving more than one plane, greater latency intervals are required.

Rate

The term "rate" in limb lengthening jargon refers to the amount of elongation per day. Although often assumed to be 1.0 mm/day, we are learning that intramedullary lengthening is more easily tolerated and forms better quality regenerate in the femur than in the tibia. Therefore, as mentioned above, a good starting rate for tibial lengthenings might be 0.5 or 0.6 mm/day.

The matter of modifying the rate of lengthening during treatment will be discussed at length in Chaps. 7 and 8.

Fig. 6.23 Good-quality regenerate created with a 0.6 mm/day rate and q2h rhythm

Rhythm

The term "rhythm" in limb lengthening refers to the fractionation of the rate into equal steps in a day. As mentioned in Chap. 2, Ilizarov determined that the more highly fractionated the rhythm, the better the quality of bone that forms in a distraction gap.

Consider, for a moment, the hassle of performing a single 0.25 mm elongation using a classic Ilizarov fixator. All the nuts on one side of a ring must be released, after which each nut on the inside of the configuration must be turned one-fourth the way around to elongate the frame by 0.25 mm. After that, the loose nuts must be tightened down again.

Italian surgeons, in the early 1980s, developed ratcheted "clickers" to simplify distraction. The external controllers of fully implantable motorized lengthening devices make the task easy indeed, but the process is still time consuming. Therefore, our suggestion to fractionate the rhythm to a greater frequency than every 6 h may seem burdensome. However, it appears, in preliminary analysis, that a q2h schedule (at least during waking hours) created regenerate bone that resembles the best regenerate morphology produced with the classic Ilizarov method (Fig. 6.23).

Surgical Soft Tissue Release

Rationale

One of the most widely used methods to reduce the likelihood of bone deformity and soft tissue contractures, subluxations, and dislocations is to perform a prophylactic surgical release of the soft tissues likely to cause such problems.

Experience has shown that certain tough fibrous bands and musculotendinous structures most commonly deform elongating structures. Prior to any lengthening surgery, a careful examination of the limb should be conducted to identify abnormally tight structures.

Thigh

In the thigh, adduction of the hip with the knee extended will reveal any tension of the tensor fascia lata/iliotibial band (TFL/ITB) complex that necessitates release at the time of femur lengthening surgery (Fig. 6.24).

Fig. 6.24 Distal tensor fascia lata release. Copyright 2016 NuVasive

A tight TFL/ITB can cause a valgus angulation of the knee. This thick layer of condensed fascia extends from the iliac crest to the outer lateral tibial condyle.

The common location for release of this structure is within 1.0 cm of the level of the proximal edge of the patella. The iliotibial band is easily identified as a condensed portion of the circular investing fascia of the thigh and should be cut transversely. Often, surgeons also transversely incise the lateral intramuscular septum down to the femur.

Examination of the hip in extension (with the knee flexed) will reveal the presence of a tight rectus femoris, suggesting release of the rectus femoris tendon at the time of femur lengthening surgery. Likewise, if during femoral lengthening, knee flexion becomes increasingly restricted, and the problem cannot be overcome with intensive physical therapy, proximal release of the rectus femoris tendon from the pelvis often solves the problem.

If a knee flexion deformity is anticipated, recession of the biceps femoris is recommended. A quadriceps contracture may require a modified quadriceps as described by the group at the Hospital for Special Surgery [9].

Lower Leg

The two most common deforming structures that impede tibia and fibula lengthening are the interosseous membrane and the gastrocnemius/soleus complex (Fig. 6.25).

The interosseous membrane, a thick sheet of fibrous tissue spanning the space between the tibia and fibula, extends from the knee to the ankle. It is so deeply seated that it cannot be easily or safely released during typical limb elongation surgery. For this reason, surgeons insert blocking screws to prevent anteromedial tibial deformation caused by the interosseous membrane.

The gastrocnemius/soleus complex (the calf muscles) pulls on the heel through the Achilles tendon. This structure rarely deforms bone during lengthening; instead, increasing calf tension during limb elongation causes progressive ankle plantarflexion and an equinus attitude of the foot, reported in from 1 to 7% of tibial lengthenings [10]. At its most extreme manifestation, such calf tightness may also produce a

Fig. 6.25 Patient with fibular hemimelia; equinus and knee contractures during tibial lengthening

knee flexion deformity, because the gastrocnemius muscle crosses behind the knee and attaches to the posterior surface of the distal femur.

Therefore, careful examination of the limb, by dorsiflexing the foot with the knee alternately flexed and extended, will reveal any preoperative tightness in this structure. This is a common phenomenon in numerous pathologies that require lower leg lengthening.

If excessive calf tightness is identified or anticipated during lower leg lengthening, a gastrocnemius recession (Fig. 6.26) is commonly incorporated into the surgical plan for many lower leg lengthening procedures [11].

Subluxations and Dislocations

As mentioned earlier, if left untreated or if lengthening continues as contractures develop, subluxation and even frank dislocation can occur (Fig. 6.27). Typically these complications are more common in femur lengthening than in tibia lengthening. If recognized early, subluxation can often be reduced with a combination of measures, including reverse to distraction to relieve tension on soft tissues, intensive exercises, and possible surgical releases. In the knee joint, sagittal plane subluxation combined with rotation of the tibia on the femur is a particularly ominous combination, possibly precluding full recovery of knee motion.

With the hip joint, subluxations are often associated with a shallow acetabulum, but can occur with a normal hip socket (Fig. 6.28). If not recognized, a subluxation leads to frank dislocation, a particularly difficult problem to treat, necessitating, at times, extensive soft tissue releases and perhaps even femoral shortening to reposition the hip (Fig. 6.29).

Fig. 6.26 Gastrocnemius recession. Copyright 2016 NuVasive

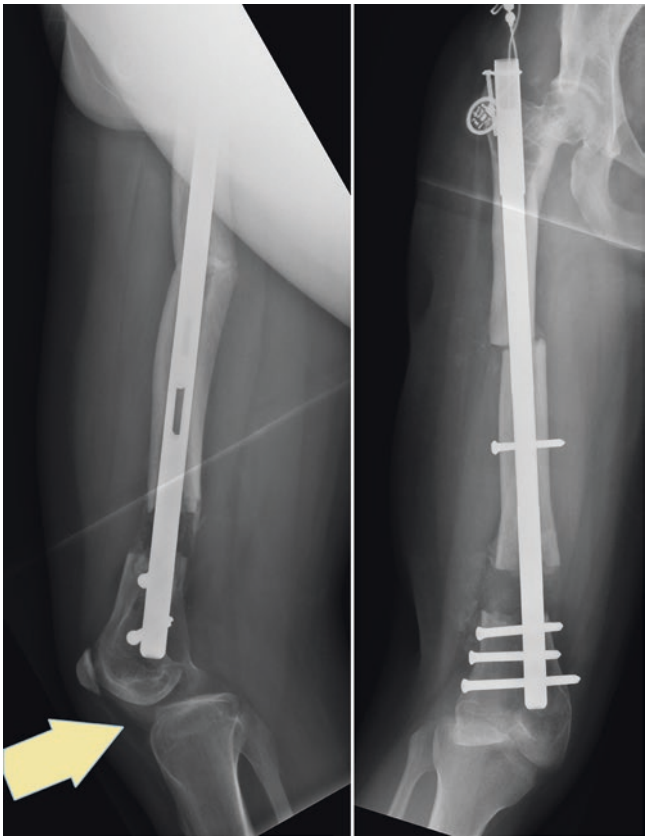


Fig. 6.27 Dislocation of knee during intramedullary bone transport. Contractures, if left untreated, may progress to subluxation (*yellow arrow*) and then frank dislocation

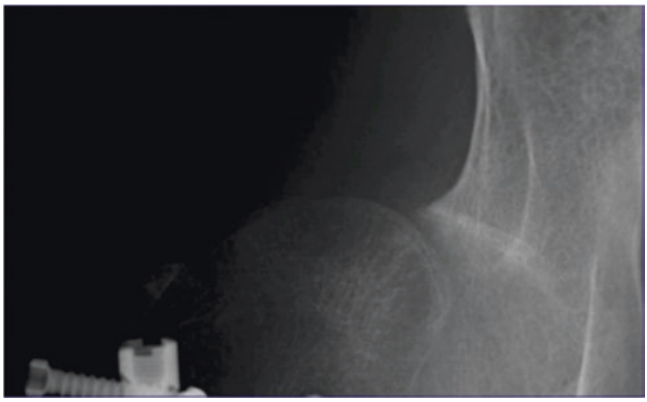


Fig. 6.28 Hip subluxation during lengthening

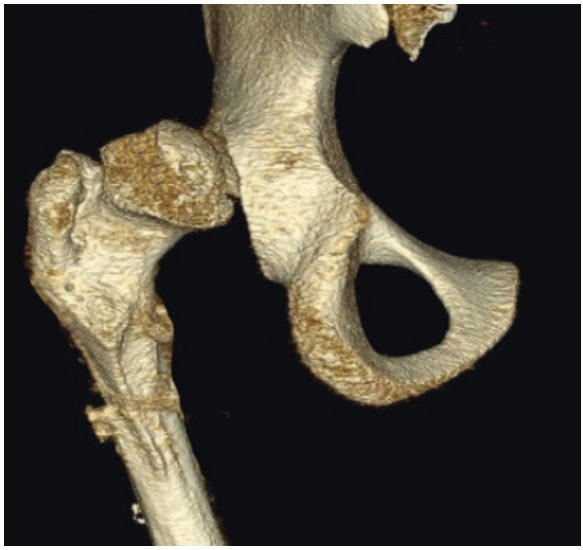


Fig. 6.29 3D reformat of hip subluxation during femur lengthening

References

1. Caiozzo VJ, Utkan A, Chou R, Khalafi A, Chandra H, Baker M, et al. Effects of distraction on muscle length: mechanisms involved in sarcomerogenesis. Clin Orthop Relat Res. 2002;403(Suppl):S133–45.
2. Wagner H. Operative Beinverlangerung. Chirurgie. 1971;42:260–71.
3. Zarzycki D, Tesiorowski M, Zarzycka M, Kacki W, Jasiewicz B. Long-term results of lower limb lengthening by the Wagner method. J Pediatr Orthop. 2002;22(3):371–4.
4. Baumgart R. The reverse planning method for lengthening of the lower limb using a straight intramedullary nail with or without deformity correction. A new method. Oper Orthop Traumatol. 2009;21(2):221–33.
5. Krettek C, Stephan C, Schandelmaier P, Richter M, Pape HC, Miclau T. The use of Poller screws as blocking screws in stabilising tibial fractures treated with small diameter intramedullary nails. J Bone Joint Surg Br. 1999;81(6):963–8.
6. Rosbruch SR, Kleinman D, Fragomen AT, Ilizarov S. Limb lengthening and then insertion of an intramedullary nail: a case-matched comparison. Clin Orthop Rel Res. 2008;466:2923–32.
7. Perren SM, Huggler A, Russenberger M, Straumann F, Muller ME, Allgower M. A method of measuring the change in compression applied to living cortical bone. Acta Orthop Scand. 1969;125:7–16.
8. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263–85.
9. Khakharia S, Fragomen AT, Rozbruch SR. Limited quadricepsplasty for contracture during femoral lengthening. Clin Orthop Relat Res. 2009;467(11):2911–7.

10. Bilen FE, Kocaoglu M, Eralp L, Balci HI. Fixator-assisted nailing and consecutive lengthening over an intramedullary nail for the correction of tibial deformity. J Bone Joint Surg Br. 2010;92(1):146–52.
11. Rozbruch SR, Zonshayn S, Muthusamy S, Borst EW, Fragomen AT, Nguyen JT. What risk factors predict usage of gastrocsoleus recession during tibial lengthening? Clin Orthop Relat Res. 2014;472(12):3842–51.

Further Reading

1. Gulabi D, Erdem M, Cecen GS, Avci CC, Saglam N, Saglam F. Ilizarov fixator combined with an intramedullary nail for tibial nonunions with bone loss: is it effective? Clin Orthop Relat Res. 2014;472(12):3892–901.
2. Kim SJ, Mandar A, Song SH, Song HR. Pitfalls of lengthening over an intramedullary nail in tibia: a consecutive case series. Arch Orthop Trauma Surg. 2012;132(2):185–91.
3. Yoshino A, Takao M, Innami K, Matsushita T. Equinus deformity during tibial lengthening with ankle orthoses for equalization of leg-length discrepancies. J Orthop Sci. 2011;16(6):756–9.

Postoperative Management Considerations

7

Basic Principles

Initial Management

The initial postoperative management for patients with self-lengthening intramedullary nails parallels that following trauma nail insertion. The surgical team manages the postoperative pain while being cognizant of the potential for evolving compartment syndrome or fat embolism syndrome.

Unlike most other surgical operations where the work is done when the patient leaves the operating room, a limb lengthening "procedure" takes months to complete. For this reason, success or failure depends upon postoperative management.

The principles of postoperative management described herein apply to any treatment protocol that slowly moves bone fragments, whether lengthening, correction of deformity, or bone transport for a skeletal defect. The suggestions for patient management apply to both intramedullary lengthening nails and external skeletal fixators.

Complications associated with the movement of bone fragments or the stretching of limbs have not been eliminated with Ilizarov's methods of bone lengthening. To the contrary, surgeons, no longer bound to the need for bone grafting, now attempt to elongate limbs by up to 100% or more of the original length, a likely source of severe complications. Indeed, problems occur whenever elongation of a limb is attempted.

Postoperative Neurological Problems

Nerve or vessel stretching occurs infrequently with high-frequency/small-step elongation strategies. Nerves and vessels can tolerate up to 2 mm of distraction a day in many locations around the body.

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_7

The earliest manifestations of excessive nerve traction are paresthesia (tingling in the nerve distribution) followed by (or accompanied by) numbness. Stop the distraction immediately, and rest the limb for a day whenever a patient complains of tingling or numbness during limb lengthening. Usually, this solves the problem. Ilizarov, in fact, recommends stopping distraction 1 or 2 days for every 10 days of lengthening, in all cases. If the sensory abnormality does improve by stopping elongation, reverse the distraction back past the length where the altered sensation began. This should eliminate the problem.

If the tingling or numbness returns with resumption of distraction, something is wrong. With external fixation devices, a pin or wire is likely pressing against a neurovascular bundle. Usually, the patient will be able to identify the specific implant causing the problem. With intramedullary lengthening nails, nerve compression by a tight fascial band is the likely culprit. Typically, this occurs where the peroneal nerve crosses the fascia just beyond the neck of the fibula. Fascial release is, therefore, the best way to deal with the problem (Fig. 7.1).

Postoperative Vascular and Circulatory Problems

Hypertension or limb ischemia is unlikely to occur when a high-frequency distraction plan is followed.

Pulmonary emboli have occurred with patients subjected to all manners of limb reconstruction, including lengthening; however, the incidence is very low. Moreover, such cases may be related to inactivity, rather than a consequence of the procedure itself.

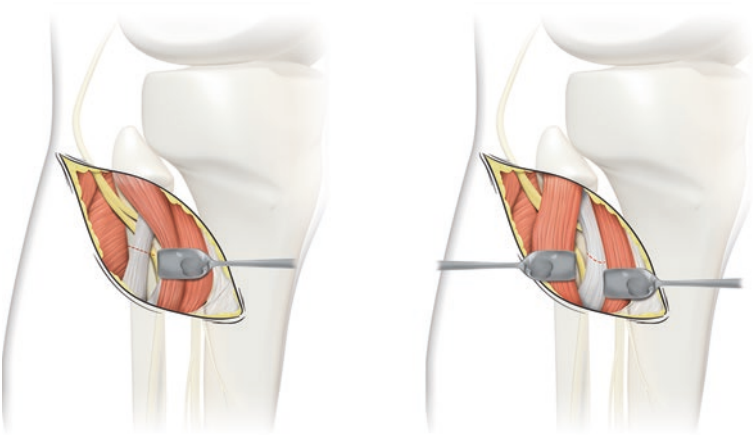


Fig. 7.1 Fascia release of the peroneal nerve near the fibular head. Both the overlying fascial band and the intramuscular septum where the nerve enters the anterior compartment should be released. Copyright 2016 NuVasive

Compartment Syndrome

Compartment syndrome is always a risk when limbs are submitted to surgical manipulation, or any kind of trauma, for that matter. Undue pain is the hallmark of the condition, requiring prompt compartment release.

Typically, compartment syndrome will occur in tibial lengthening procedures, especially when marrow canal reamings extrude through the pre-osteotomy drill holes into the lateral compartment of the tibia. Some surgeons avoid drilling into this region of the limb during preliminary osteotomy site preparation, whereas others prophylactically release the compartment. The most important step of all, however, is postoperative vigilance.

Contractures, Subluxations, and Dislocations

Every limb lengthening operation can result in contracture, subluxation (incomplete dislocation), or even complete dislocation of an adjacent joint. This occurs because myofascial tissue resists elongation. It is generally believed that the fascial tissues surrounding muscle bundles, rather than the muscle cells themselves, are responsible for such problems. Hence, joint contractures during limb elongation tend to occur in the direction opposite the greatest muscle mass or thickest fascial structures.

During limb lengthening, myofascial tissues, by resisting elongation, create joint contractures, which limit the range of motion. This occurs either temporarily, or, if the problem is not addressed, permanently (Fig. 7.2). Furthermore, if left untreated during progressive limb lengthening, joint contractures worsen as elongation proceeds. The contracted joint gradually subluxes, whereby the articular surfaces of the joint are not perfectly congruent (subluxation *means* incomplete dislocation).

The last phase of this process occurs when the joint dislocates completely, a worrisome consequence of limb lengthening that can occur with either internal or

Fig. 7.2 A not uncommon deformity after distal tibia and fibula lengthening: extension of the great toe and flexion of the lesser toes, caused by the arrangement of muscle origins near the osteotomy site. Copyright 2016 NuVasive

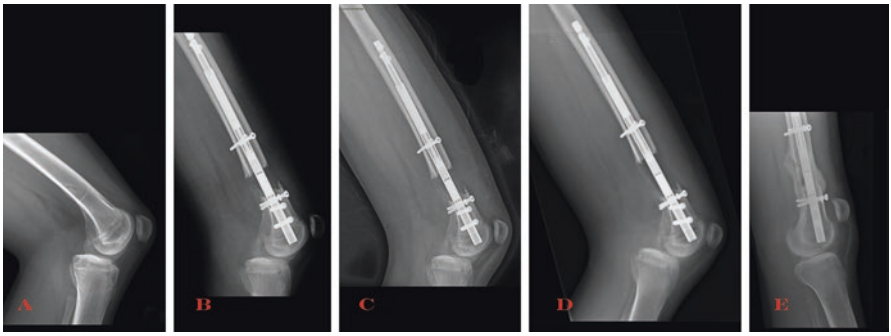


Fig. 7.3 Posterior subluxation of the knee. (**a**) Pre-op status after two previous femur lengthenings. (**b**) Gradual posterior subluxation of the knee during PRECICE® nail lengthening. (**c**) Further subluxation. (**d**) Rotation and subluxation. Nail reversed 10 mm; dynamic splint applied; 3-week intensive daily physical therapy. (**e**) Reduction without surgery, but 10 mm shorter than planned

external devices. Returning such a joint to its anatomic position typically involves extensive reconstructive surgery that includes shortening the elongated bone to its preoperative length, complex capsular reconstruction of the joint itself, and often angulation osteotomies adjacent to the joint. Moreover, such a joint is rarely, if ever, normal thereafter.

Ankle equinus and knee flexion contractures are the two most common types of deformities that occur with lower limb elongation. Likewise, hip flexion contractures can develop during femoral lengthening. These deformities should not be allowed to persist or worsen while the bone is being lengthened. If a knee flexion contracture is not corrected, progressive tightening of the hamstrings can lead to posterior subluxation of the knee (Fig. 7.3). If this subluxation is not recognized and corrected, a frank dislocation may occur.

In the hip, a dislocation is most likely to happen in a patient with femoral neck valgus or a shallow acetabular roof at the start of femoral lengthening.

It is possible for contractures, subluxations, or dislocations to occur at both ends of a bone simultaneously. Thus, a progressive knee flexion deformity during femur lengthening is often accompanied by a flexion contracture of the hip as well. As the hamstring muscles tighten, the patella and rectus femoris muscle are pulled distally by increased knee flexion. This combination causes the patient to stand with a flexed knee and hip, balancing the limb on his or her toes (Fig. 7.4).

Avoiding Complications

Physiotherapy

Important elements of every postoperative physical therapy treatment plan designed to prevent joint contractures, subluxation, and dislocations include muscle stretching, elastic and static splinting, appropriate nighttime positioning, and active use of the limb during the entire lengthening process.

Fig. 7.4 A worrisome combination of joint contractures during femoral lengthening. Hamstring tightness causes knee flexion and posterior displacement of the tibia on the femoral condyles ("ski-slope knee"). This pulls the quadriceps distally, tightening the rectus femoris and resulting in hip flexion. If untreated, both the hip and knee can dislocate with further elongation of the bone. Copyright 2016 NuVasive

As a rule, however, physiotherapy cannot prevent deformities that angulate a lengthening bone through the regenerate new bone; instead, the surgical strategy should include techniques to prevent deformation (with blocking screws, for instance) or ways with which to deal with the problem if it occurs.

Whenever bone fragments are moved with respect to one another, soft tissues are placed under tension; the greater the movement, the greater the tension. Therefore, physiotherapy strategies have been developed to maintain joint mobility during limb lengthening. Indeed, constant stretching of tightening tissues is the hallmark of proper postoperative management of a patient using any device that is lengthening bone and soft tissues.

Stretching

Passive stretching of tightening tissues is the basis of the postoperative management of every patient receiving limb lengthening. Intensive "hands-on" physiotherapy is necessary to prevent contractures, joint subluxations, and dislocations associated with limb elongation.

Fig. 7.5 Calf stretching exercises are essential with any tibia or fibula lengthening procedure. Copyright 2016 NuVasive

Interestingly, active muscle exercises do not help much in preventing contractures. For example, active dorsiflexion of the ankle is not nearly as effective as passive stretching of the calf musculature in limiting equinus contractures (Fig. 7.5). Nevertheless, active exercises are important for a patient during limb lengthening, as the nutrition of the elongating tissues depends upon local circulation, which, in turn, is dependent upon functional use of the muscles.

The physical therapist must teach the patient and family members how to stretch the calf, hamstrings, and other muscle groups (Fig. 7.6). At least 2 or 3 h a day should be devoted to this activity, especially in cases involving substantial lengthening. Indeed, the greater the anticipated elongation, the more time per day must be devoted to passive muscle stretching.

Splinting

Both static and dynamic splinting have a role in the management of patients undergoing limb lengthening and elongation of resistant soft tissues. Static splints include

Fig. 7.6 Knee extension exercises are required with both thigh and lower leg lengthening. Copyright 2016 NuVasive

fixed-position orthoses and other nonelastic devices that hold a limb in a neutral position (Fig. 7.7). Such devices are especially helpful at night, when the constant pressure of a dynamic splint can become uncomfortable for the patient.

Dynamic splinting incorporates a spring-like mechanism that counteracts deforming forces. Such splinting techniques are very useful, especially where less expensive orthoses have not proven successful (Fig. 7.8).

Both kinds of splints, while effective, may also prove dangerous, because a constant pressure on the skin, particularly when overlying bony prominences, can cause skin ulceration. Typically, however, a conscious patient will note discomfort (burning of the skin) in regions of cutaneous compression. Padding, cutouts, and other orthotic tricks usually can address this problem.

Night Positioning

The 7 or 8 h a patient spends in bed may be the most important hours of the day for a patient undergoing limb lengthening. At nighttime, joints are allowed to fall into suboptimal positions that will affect correction during the day.

Fig. 7.7 A static splint to prevent equinus contracture (Orliman®). Attach to distal most lacing. Copyright 2016 NuVasive

Fig. 7.8 Spring-loaded ankle dorsiflexion brace (Dynasplint®). Copyright 2016 NuVasive

For most lower extremity applications, the foot must be supported and prevented from dropping into plantarflexion. Likewise, the knee joint should be gently forced into full extension, usually by the proper placement of pillows under the foot. Nurses generally prop pillows under a limb that has been operated upon. Almost invariably, this propping is under the operative site, where the bleeding is noted. In lower leg applications, the pillow will thus cause the knee to flex and the foot to fall into an equinus attitude (Fig. 7.9).

The only effective way to support a lower limb during lengthening is with the pillow behind the ankle and heel, a measure that extends the knee.

If a knee flexion contracture appears to be developing, placing a sandbag on the knee when the patient is in bed may aid knee extension.

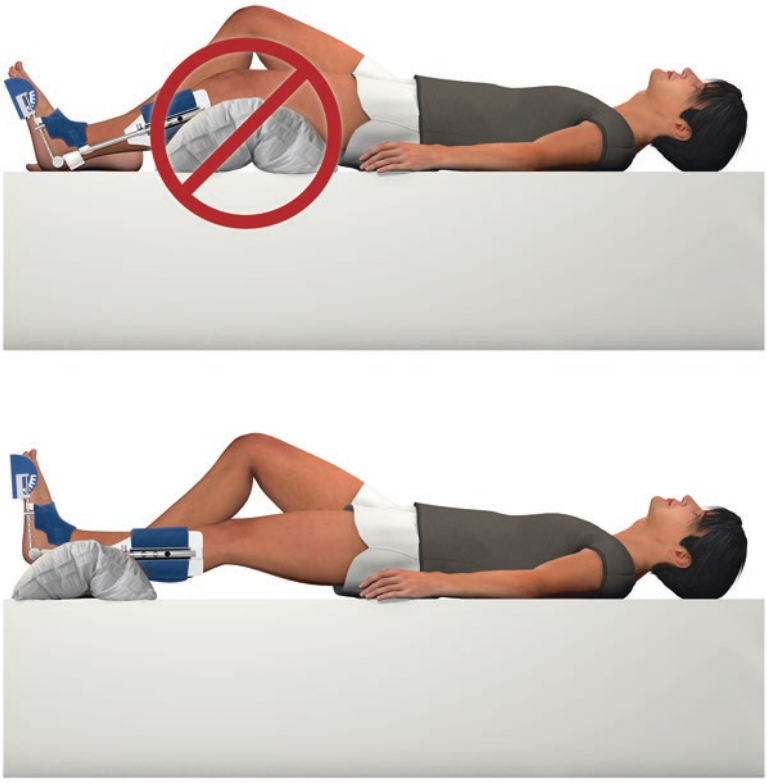


Fig. 7.9 Place pillow underfoot, not behind the knee. Copyright 2016 NuVasive

Fig. 7.10 Knee extension position. Note weight attached to foot. Copyright 2016 NuVasive

At times, especially with extended lengthening procedures, an ankle-foot orthosis (AFO) may be required.

With thigh lengthening, the tendency for hip flexion at night can be overcome by placing the patient prone. A pillow under a prone patient's knee and upper thigh extends both the hip and knee (Fig. 7.10).

In any lower extremity lengthening or procedure that causes tension in the tissues, the patient should not be permitted in bed with the back and knee gatched-up, the so-called "semi-Fowler's" position. In these cases, a combination of hip flexion, knee flexion, and ankle plantarflexion will cause problems that are difficult to overcome.

Ankle equinus is not the only problem associated with lower leg lengthening. There is a tendency for the toes to curl during distal lengthening of the tibia. At times, it may be necessary to insert K-wires into the toes to prevent this problem.

Functional Use of the Limb

For a successful application of distraction osteogenesis, a limb must be used in a physiologic manner throughout the course of treatment. Mechanical stimulation is essential for the proper ossification of the newly formed regenerate bone in a distraction gap and for the optimum maturation of healing fractures and pseudarthroses.

To achieve this goal, a patient must partially bear weight on a lower limb and use the upper limb as normally as possible.

Graduated gait training begins on the first postoperative day. Encourage the patient to bear weight as tolerated on the operated limb, aided by crutches or a walker. Likewise, active range of motion of the joints should be encouraged.

A natural rhythmic walking pattern is probably more important than the actual amount of weight on the limb at the beginning of the rehabilitation program. With time, the patient must progressively increase the load on the limb. Toward the end of treatment, the patient should be able to move around with one crutch or a cane.

If, during the course of postoperative limb elongation surgery management, a patient's walking ability decreases, the surgeon must immediately determine the cause. Usually, there is a clear-cut reason why the patient is having difficulty with ambulation. One example is the development of a deep infection. The surgeon must not ignore such a development. A full work-up of the problem may be required, including blood tests, bone scans, and so forth.

Ambulation and upper extremity use not only promote ossification of the regenerate but also help prevent contractures, subluxations, and dislocations. Weight bearing, for example, serves as a means of passive calf muscle stretching while maintaining tone and stimulating circulation in the limb. With upper extremity lengthening, eating, hair combing, gymnastics, dance therapy, and other similar activities are also useful adjuncts to therapy. The rhythmic movements involved with swimming, cycling, and walking are among the best therapeutic exercises available. Constant encouragement by the physical therapist and surgeon will do much to ensure rapid ossification of the regenerate new bone and prevent contractures, subluxations, and dislocations.

Failure to Control Progressive Contractures

If the surgeon and physiotherapist cannot overcome an evolving joint contracture with splinting, hands-on passive stretching, or other such strategies, the wisest course of action is to stop whatever bone movement is occurring and commence a course of intensive physiotherapy. For this purpose, the patient may have to be admitted to hospital for supervised care. If the contracture does not improve, the best course of action is to abandon the treatment goal and postpone completion of the original treatment plan. If not, and the distraction continues, contractures lead to subluxations, which may result in a dislocation of one or both adjacent joints. This typically occurs with femoral lengthening, and the consequences of simultaneous hip and knee dislocations may be catastrophic. The limb must be usually be shortened to permit relocation of the dislocated joints.

Dhawale et al. [1] reported three hip dislocations during femoral lengthening. The average amount of elongation was 9 cm. in these cases. This suggests that the most effective way to reduce the possibility of hip dislocation during femoral lengthening is to resist the temptation to elongate a limb more than 6 or 7 cm. at a time.

It is evident that physiotherapy is the key to a successful application strategy involved with the creation of a distraction regenerate. Ambulation and functional loading are essential for ossification of the regenerate new bone, while stretching and range of motion preservation are the keys to preventing contractures, subluxations, and dislocations.

Reference

1. Dhawale AA, Johari AN, Nemade A. Hip dislocation during lengthening of congenital short femur. J Pediatr Orthop B. 2012;21(3):240–7.

Further Reading

1. Al-Sayyad MJ. Lower limb lengthening and deformity correction using the Fitbone motorized nail system in the adolescent patient. J Pediatr Orthop B. 2012;21(2):131–6.
2. Barker KL, Lamb SE, Simpson HR. Recovery of muscle strength and power after limb-lengthening surgery. Arch Phys Med Rehabil. 2010;91(3):384–8.
3. Bilen FE, Kocaoglu M, Eralp L, Balci HI. Fixator-assisted nailing and consecutive lengthening over an intramedullary nail for the correction of tibial deformity. J Bone Joint Surg Br. 2010;92(1):146–52.
4. Blair JA, Owens JG, Saucedo J, Hsu JR, Skeletal Trauma Research C. Functional rehabilitation with a foot plate modification for circular external fixation. Foot Ankle Int. 2013;34(6):890–7.
5. Coglianese DB, Herzenberg JE, Goulet JA. Physical therapy management of patients undergoing limb lengthening by distraction osteogenesis. J Orthop Sports Phys Ther. 1993;17(3):124–32.
6. Dhawale AA, Johari AN, Nemade A. Hip dislocation during lengthening of congenital short femur. J Pediatr Orthop B. 2012;21(3):240–7.

7. Ferguson JY, Sutherland M, Pandit HG, McNally M. The rate of symptomatic venous thromboembolism in patients undergoing elective Ilizarov surgery and the cost of chemical prophylaxis. Bone Joint J. 2014;96-B(3):426–30.
8. Green SA. Physiotherapy in Ilizarov fixation. Tech Orthop. 1990;5(4):61–5.
9. Green SA. Post-operative management during limb lengthening. Orthop Clin N Am. 1991;22(4):723–34.
10. Hazra S, Song HR, Jajodia N, Biswal S, Modi HN, Srinivasalu S. Hip and knee dislocation during femoral lengthening in congenital short femur: a rare case report. Arch Orthop Trauma Surg. 2009;129(4):425–9.
11. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: Part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263–85.
12. Ippolito E, Peretti G, Bellocci M, Farsetti P, Tudisco C, Caterini R, et al. Histology and ultrastructure of arteries, veins, and peripheral nerves during limb lengthening. Clin Orthop Relat Res. 1994;(308):54–62.
13. Karakoyun O, Kucukkaya M, Sokucu S. Intramedullary skeletal kinetic distractor in lower extremity lengthening. Acta Orthop Traumatol Turc. 2014;48(3):307–12.
14. Kim SJ, Mandar A, Song SH, Song HR. Pitfalls of lengthening over an intramedullary nail in tibia: a consecutive case series. Arch Orthop Trauma Surg. 2012;132(2):185–91.
15. Koplin SA, Su L, Salamat S, Torrealba J, McCarthy J, Mitchell J, et al. Distraction osteogenesis-induced muscle fibrosis may not be associated with TGF-beta1. J Pediatr Orthop. 2011;31(4):413–20.
16. Lee DY, Han TR, Choi IH, Lee CK, Chung SS. Changes in somatosensory-evoked potentials in limb lengthening. An experimental study on rabbits' tibiae. Clin Orthop Relat Res. 1992;(285):273–9.
17. Lee DH, Ryu KJ, Shin DE, Kim HW. Botulinum toxin a does not decrease calf pain or improve ROM during limb lengthening: a randomized trial. Clin Orthop Relat Res. 2014;472(12):3835–41.
18. Makarov MR, Samchukov ML, Birch JG, Cherkashin AM, Sparagana SP, Delgado MR. Somatosensory evoked potential monitoring of peripheral nerves during external fixation for limb lengthening and correction of deformity in children. J Bone Joint Surg Br. 2012;94(10):1421–6.
19. Makarov MR, Delgado MR, Samchukov ML, Welch RD, Birch JG. Somatosensory evoked potential evaluation of acute nerve injury associated with external fixation procedures. Clin Orthop Relat Res. 1994;(308):254–63.
20. Martin BD, Cherkashin AM, Tulchin K, Samchukov M, Birch JG. Treatment of femoral lengthening-related knee stiffness with a novel quadricepsplasty. J Pediatr Orthop. 2013;33(4):446–52.
21. Rozbruch SR, Fryman C, Bigman D, Adler R. Use of ultrasound in detection and treatment of nerve compromise in a case of humeral lengthening. HSS J. 2011;7(1):80–4.
22. Rozbruch SR, Zonshayn S, Muthusamy S, Borst EW, Fragomen AT, Nguyen JT. What risk factors predict usage of gastrocsoleus recession during tibial lengthening? Clin Orthop Relat Res. 2014;472(12):3842–51.
23. Strong M, Hruska J, Czyrny J, Heffner R, Brody A, Wong-Chung J. Nerve palsy during femoral lengthening: MRI, electrical, and histologic findings in the central and peripheral nervous systems—a canine model. J Pediatr Orthop. 1994;14(3):347–51.
24. Suzuki S, Kasahara Y, Seto Y, Futami T, Furukawa K, Nishino Y. Dislocation and subluxation during femoral lengthening. J Pediatr Orthop. 1994;14(3):343–6.

Postoperative Care, Day by Day

8

The following step-by-step guide to the postoperative management of patients undergoing limb lengthening also applies to patients who are having a deformity corrected or a bone segment transported to overcome an osseous defect.

In Operating Room

After Surgery

Use gauze wraps to apply pressure on the operative sites.

Be sure to obtain final roentgenograms before the patient leaves the operating table. The limited image size of intraoperative intensified fluoroscopy often fails to reveal malalignment of the limb as a whole.

Day-by-Day Management

Postoperative Day 1

Physical therapy is started on the first postoperative day. The patient must "work" on preventing contractures before lengthening begins.

The patient's bed must be flat, not elevated behind the knee. Place a pillow under the ankle to force the knee into extension.

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_8

Postoperative Days 2–4

The patient's physical therapy program continues with progressive weight bearing and range of motion of the joints. As mentioned earlier, passive stretching is an important part of the physiotherapy program.

We often discharge the patient from the hospital on days 2, 3, or 4 postoperatively. At the time of discharge, the patient should be off parenteral pain medication, taking only oral painkillers.

Postoperative Days 5, 6, and 7 or Later

One of the most important days for the patient postoperatively is the day that distraction begins. This may occur while the patient is still in the hospital, or it may take place at the first outpatient visit. Remove sutures when appropriate. The latency interval (delay before beginning distraction) after insertion of the implant has allowed the first stage of fracture healing to commence. During distraction, the osteotomy site fracture begins to heal. The newly formed fracture callus attempts to "catch up" with the distracting bone ends, but, under most circumstances, does not consolidate the regenerate bone within the distraction gap until the neutral fixation period following elongation.

Latency

In general, the delay (latency) prior to distraction is 5–7 days, for the femur, but may be longer or shorter under certain circumstances. The latency for the tibia should be 10–14 days, since it does not form regenerate bone as readily as the femur.

Shorten latency:

- In pediatric cases. The rapidity of bone growth means that the latency period should be only 4 or 5 days following osteotomy through healthy bone.
- Where the corticotomy is oblique. The latency should be shortened by 1–2 days because oblique osteotomies heal more rapidly than transverse ones.

Prolong latency:

- If there has been considerable comminution at the site of osteotomy (the latency interval should be lengthened by 3 or 4 days)
- If there has been substantial displacement of the major fragments during osteotomy
- If fragments were counterrotated (during torsional osteoclasis of the posterior cortex) more than 30°

If the bone is of poor quality—either extremely dense or osteopenic—the latency interval should be lengthened up to 14 days (or perhaps even longer), especially if the soft tissues surrounding the bone are also of suboptimal quality.

Distraction

Following the latency interval, the patient is taught to distract the corticotomy gap 0.25 mm every 6 h. This rate and frequency may be altered, depending upon the clinical circumstances.

With an intramedullary lengthening nail, reaming of the marrow canal has the effect of reducing the rate of regenerate ossification. While the femur, surrounded on all sides by thick muscle, responds well to distraction at a speed of 1.0 mm/day (divided into three or four doses of 0.33 or 0.25 mm each), the tibia, whose anterior surface is subcutaneous, often has deficient maturation of the regenerate anteriorly. For this reason, experienced surgeons are now recommending tibial distraction at a rate of 0.75 mm/day in three doses of 0.25 mm each.

For an adult with dense bone and suboptimal surrounding tissues, a more appropriate initial rate and frequency would be 0.25 mm every 12 h. In pediatric cases, however, such a slow rate of distraction might result in premature osseous consolidation, especially if the corticotomy is oblique and through healthy tissues. The fastest rate of distraction, however, is usually 1.0 mm per day at each widening distraction gap.

Have the patient (or responsible individual) practice distraction at the first postoperative visit, making sure that everything is understood.

Visualizing the Regenerate New Bone

One can easily misinterpret new bone formation in the widening distraction gap if the central beam of the x-ray tube is not directly over the middle of the distraction zone, especially if the tube is close to the patient. Likewise, if the limb is not perpendicular to the x-ray beam (as can happen if a knee flexion contracture is present) and parallel to the image receiver (either film or sensitive plate), distortion and cortex overlap may give the false impression that the bone is forming during distraction, when, in fact, it is not. Repositioning the tube and plate for orthogonal imaging, or repositioning the patient, may be necessary (Figs. 8.1 and 8.2).

After One Week of Distraction

The next important contact with the patient comes after 1 week of distraction. Usually, the patient is at home by this time. The first set of roentgenograms should show a gap between the bone fragments that corresponds in width to the rate and frequency of distraction. Thus, if the patient has been lengthening at a rate of 1.0 mm per day for a week, the measured bone gap should be 7.0 mm.

Fig. 8.1 To best judge the regenerate, the x-ray beam must be perpendicular to the bone and film (*left*). Bone angulation, in relation to beam and film, can cause a false reading of regenerate bone in the gap (*right*). Copyright 2016 NuVasive

Fig. 8.2 Methods to correct the issue of parallax: either tilt the beam and film (*left*) or reposition the patient (*right*). Copyright 2016 NuVasive

Plan the roentgenographic views to obtain maximum information with the least amount of x-ray exposure for the patient. As a rule, the central x-ray beam must be perpendicular to the osteotomy gap. To be helpful, such a view must show the bone lengthening in the profile of the anticipated deformity. For example, a tibia will deform with its apex anteromedially; hence, the anterolateral oblique view will demonstrate such a deformation before any other projection.

If the bone fragments are not separating, consider the osteotomy incomplete (it is unlikely that the bone will have healed by this time). At times, a residual bridge of the bone holding the fragment together can be pulled apart by continuing distraction, thereby disrupting the bridge. The patient must be warned that he or she may experience acute severe pain in the limb if the bone suddenly yields to the forces generated by the elongated implant. In any case, do not distract the intramedullary lengthening nail for more than 5.0 mm if the bone is not separating, because the sudden elongation can damage nerves or vessels.

The absence of progressive widening of the distraction gap usually means a repeat trip to the operating room for completion of the corticotomy or osteotomy.

Do not expect to see any new bone formation in the gap as soon as 1 week after distraction starts, although a cloudy regenerate may be observed in young children.

Two Weeks of Distraction

After 2 weeks of distraction, the gap should be 14 mm wide (or less if the limb is being elongated at a rate slower than 1 mm per day). Regenerate new bone may not be visible at this stage of distraction. For this reason, the patient should stay on the course and continue distraction at the same rate.

Three Weeks of Distraction

By the third week of distraction, some regenerate new bone should be visible in the distraction gap, usually, as a cloudy haze in adults or a fully formed regenerate (with striations and an early interzone) in children. At this stage, the absence of any evidence of regenerate new bone means that the rate of distraction should be slowed—perhaps to 0.25 mm every 8 or 12 h.

Four Weeks of Distraction

At the end of the fourth week of distraction, the gap will be 28 mm wide at a rate of 1 mm per day. By this time, regenerate new bone must be clearly evident in the distraction gap. If not, reverse distraction—closing the gap at a rate that is tolerable to the patient—generally about 1 or 2 mm per day in divided doses of 0.25–0.5 mm every 6 h. Usually, regenerate new bone will form and be visible before the gap is completely closed, especially if the patient has been bearing weight on the limb.

If no bone forms by the time the gap is fully closed, wait a longer latency interval than initially employed (Ilizarov recommends doubling the latency interval), and begin distracting again at a slower rate (half speed). Follow the post-distraction strategy as before.

Visits During Distraction

With good regenerate formation, the patient is evaluated on a weekly basis. Assess the quality of the regenerate new bone roentgenographically, slowing down, speeding up, or even stopping distraction depending on the quality of bone forming in the distraction gap. Professor Ilizarov recommends resting the limb (stopping distraction) 1 or even 2 days for every 10 days of distraction, although this is rarely done nowadays.

During the weekly visits, check the range of joint motion of the limb. Any progressive loss of motion must be dealt with immediately. In some cases, intensifying the frequency of physiotherapy will overcome the problem. If not, the patient may have to be admitted to the hospital for treatment. It may be necessary to stop distraction altogether during this period, in an attempt to regain motion.

It is critically important to check the joints for any evidence of subluxation—a problem that could lead to complete dislocation if left unnoticed. Subluxation will mostly involve the knee. Typically, a flexion deformity has preceded the subluxation. The patient will display a "ski-slope knee," the outward appearance of posterior tibial subluxation on the femur. Obtain a lateral roentgenographic view of the knee. On a roentgenogram of a normal knee, the center of the tibial plateau will be directly under the center of the femoral condyles.

As mentioned in the section dealing with general principles, it is both safer and wiser to stop lengthening a limb that develops a contracture and plan a second-stage procedure at a later time to complete limb elongation.

During lengthening, the bone may deform. When this occurs, correct the deviation or it will progressively become worse. The tactics for deformity correction vary, with blocking screws used for intramedullary lengthening nails.

Judging the Regenerate

Assessment

The progress and success of a limb lengthening protocol depends, to a considerable extent, on assessing the quality of regenerate bone in a widening distraction gap. Most commonly, this is done with serial standard AP and lateral x-ray studies taken at frequent intervals while bone fragments are moving with respect to each other, and less often during the neutral fixation phase, when the regenerate matures and hardens.

When external fixators are used for limb lengthening, the matter of corticalization of the regenerate is critical, because removal of the frame before the new bone can support weight leads to either bending or breaking of the regenerate.

Although classic Ilizarov teaching is to leave the frame on a limb until the risk of bending or breaking is nil, Western surgeons, Paley in particular, [1] pressed by their patients for premature fixator removal, have devised devise schemes to support the regenerate with an intramedullary device inserted at the time of fixator application.

Alternatively, a nail or plate can be inserted at the time the frame is removed [2].

To some extent, lengthening with an intramedullary nail has reduced, but not eliminated, concern about the risk of regenerate bending or fracture because the nail should prevent either eventuality. However, motorized nails are not nearly as strong as trauma nails, so deficient bone formation during elongation, combined with early weight bearing, risks nail breakage and loss of limb alignment—a potentially worse problem that bending or breaking of a bone through newly formed regenerate with no implant in place.

For this reason, the criteria employed with external fixator limb lengthening cases to determine when to allow frame removal and unprotected weight bearing also apply to limb elongation with intramedullary lengthening nails. As mentioned elsewhere, at a minimum, full corticalization of three of four cortices (seen on AP and lateral x-ray views) is required before full weight bearing is allowed. Moreover,

the incompletely ossified cortex should be nearly completely corticalized, with, at most, a small triangular defect (called a rat-bite) seen on imaging studies.

Experienced Ilizarov surgeons frequently receive x-ray studies from colleagues asking if the bone in the distraction gap is solid enough to permit frame removal. The bone is rarely ready. Solid-looking regenerate is easy to identify, and anything questionable isn't ready.

Classification of Regenerate

Li et al. created a classification system based on the quality of ossification and the shape of bone formed in the regenerate zone [3]. The authors identify five distinct shapes (fusiform, cylindrical, concave, lateral, and central) and three different levels of density (low, intermediate, and normal). Likewise, they categorize four patterns of distribution of bone formation (sparse, homogeneous, heterogeneous, and lucent). Combining the latter two features, Li et al. describe ten types of features: soft, stripe, speckle, adjacent, halftone, uniform, irregular, sawtooth, solid, and cystic defects.

Bone Mineral Density

Because such assessments are subjective, a number of researchers have tried to establish quantitative methods of determining when the bone in a regenerate zone is solid enough for fixator removal and/or full weight bearing. Most of these techniques employ some quantitative comparison between the bone along the edge of the regenerate and the bone in adjacent normal region [4].

Pixel Value Ratio

With digitalized x-ray images, one can compare the intensity of pixels at the edge of the regenerate to pixels in the same location in the adjacent normal bone and create a ratio of such intensities, the "pixel value ratio" (PVR). Such ratios correspond quite well to relative bone mineral density (BMD) [5].

In many ways, such quantitative measures correspond to what experienced surgeons do visually when assessing regenerate ossification. We look at the whiteness of the bone along the edge of regenerate cortices and compare it in our mind's eye to the whiteness of the cortical bone above and below the distraction zone. This visual whiteness comparison is, in reality, a pixel value ratio. If the pixels that make up the image of the regenerate cortex equal in brightness the pixels that make up the adjacent normal cortex, then the ratio value is 1.0 and the cortex can be considered solid.

Thus, using PVR improves regenerate maturation assessment compared to simple BMD measurements with a DEXA scan device (Markel, 1993 #1629).

Ultrasound

Diagnostic ultrasound, used increasingly in physician's offices for joint aspiration, injections, and diagnoses, has the potential for reducing x-ray exposure to patients undergoing limb lengthening. The modality is most often used therapeutically for stimulating maturation of the regenerate [6] but only rarely has diagnostic ultrasound been proposed for assessing the regenerate. Luk et al. used ultrasound for quantifying mineralization of the regenerate in rabbits subjected to limb elongation [7]. Acoustic reflection in 2D and 3D ultrasonography and ultrasonometry proved more sensitive to *early* mineralization of newly forming regenerate than did computerized radiography.

Clinicians have not, as yet, taken up this proposal, perhaps because early mineralization is not as important clinically as end-stage calcification of the periphery of the regenerate mass, which shows up so well on ordinary x-ray images and can be quantified with the technology described above.

All of the above techniques were developed during an era when the only means of predictably elongating a bone was to use an external skeletal fixator and Ilizarov principles. Nowadays, however, lengthening with an intramedullary motorized nail is becoming increasingly popular among surgeons in the field. With such a device, the metallic nature of the implant may interfere with pixel quantification and thus reduce the value of such determinations. The problem, as of this writing, has not been fully explored.

The Regenerate Around a Lengthening Nail

As more surgeons use intramedullary lengthening nails, a pattern of regenerate formation that was infrequently observed with external fixator lengthening is now becoming more common, namely, the eggshell > hollow > fusiform archetype. In the past, this pattern, when observed during fixator lengthenings, was considered a sign of instability at the widening distraction zone. After all, a bulging regenerate resembles normal fracture callus when healing of an intrinsically unstable long bone fracture occurs in a cylindrical cast unaccompanied by internal or external fixation. In essence, nature creates a scaffold of new bone at the periphery of a fracture hematoma where the effect of moving bone fragments is least likely to disrupt osteogenesis. The healing bone matures from the outside inwards.

A bulging regenerate, hardening on the outside first, was initially viewed with some concern by limb lengthening surgeons, but time has shown that most regenerates displaying this pattern consolidate nicely, although it takes a while. Protected weight bearing must continue until the well-established three cortices rule is obeyed.

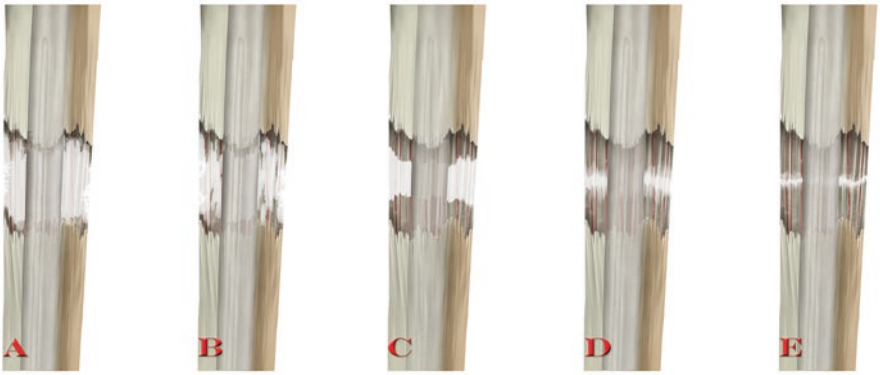


Fig. 8.3 Regenerate at 30 mm: (**a**) sparse, (**b**) patchy, (**c**) fair, (**d**) good, (**e**) excellent

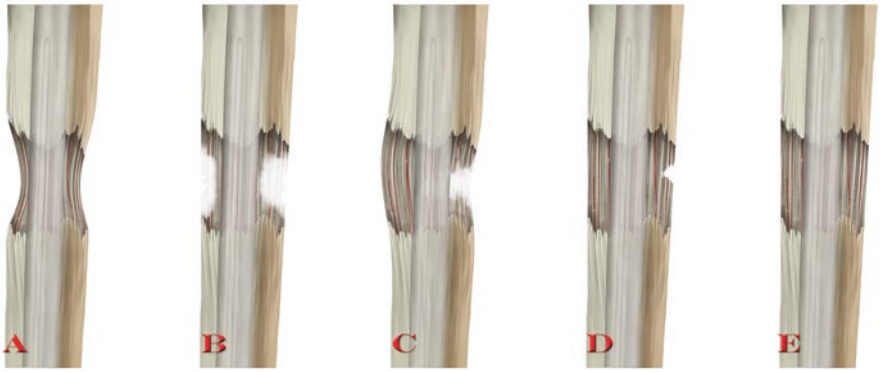


Fig. 8.4 Regenerate at 30 mm: (**a**) concave, (**b**) central, (**c**) lateral, (**d**) rat bite, (**e**) cylindrical

Patterns of Regenerate

We have modified Li et al.'s [3] classification scheme for regenerate ossification patterns to include variants seen with intramedullary lengthenings as well as other patterns characteristic of normal and rapid maturation of the regenerate (Figs. 8.3, 8.4, and 8.5).

Biology of the Regenerate

Gardening

Regenerate new bone in a widening distraction gap is like any rapidly multiplying living thing: its growth can be either retarded or enhanced by environmental factors.

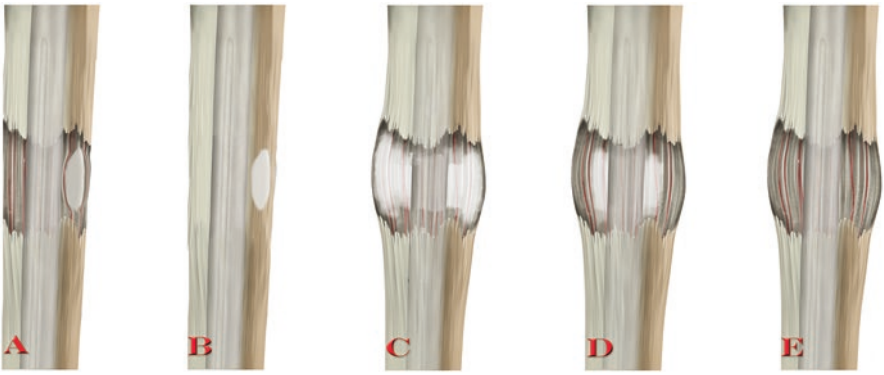


Fig. 8.5 Regenerate at 30 mm: (**a**) cystic, (**b**) mature cystic, (**c**) eggshell, (**d**) hollow, (**e**) fusiform

In this sense, a limb lengthening surgeon is more a gardener than a carpenter. Thus, anything that slows or accelerated normal fracture healing will have a similar effect on the regenerate in a distraction gap. After all, an osteotomy that creates a regenerate is a non-displaced fracture that heals in an ordinary and natural manner for the 5–7-day latency interval.

Biological Factors Retarding Regenerate

Environmental influences that slow fracture callus and regenerate maturation can be divided into biological, chemical and mechanical factors. Among the biological factors are those directly and indirectly impacting the site of fracture or osteotomy, including soft tissue damage (either acute or old), local circulation, the degree of comminution, anemia, severe malnutrition, vitamin D deficiency, diabetes, hypothyroidism, and past or present radiation of the local tissues.

Chemical Factors Retarding Regenerate

The chemical factors that adversely influence bone and regenerate healing are those that reduce inflammation, the first phase of fracture healing. Thus, any anti-inflammatory medication, steroidal or nonsteroidal, has this adverse effect. Likewise, commonly used over-the-counter pills, while not as powerful as prescription medications, should not be used during distraction osteogenesis.

Nicotine, no matter how delivered, slows bone healing and thus regenerate maturation [8–14].

Mechanical Factors Retarding Regenerate

Mechanical factors that retard regenerate ossification center around weight bearing activities. With external fixation, weight bearing to tolerance is promoted. Unfortunately, when intramedullary lengthening nails are employed, concern about nail breakage makes surgeons exceedingly caution about unprotected weight bearing. Nevertheless, partial weight bearing, as discussed below, remains a hallmark of proper postoperative care during limb elongation.

Maturation (Consolidation)

Throughout the course of regenerate maturation, the patient must be encouraged to bear weight on the limb, lest the regenerate fail to ossify (Fig. 8.6).

Visits During Maturation

Evaluate patients monthly during regenerate maturation, checking the quality of the maturing bone with roentgenograms. The patient's weight-bearing and functional capacity should increase steadily during this period. Investigate any decline in the patient's ability to use the elongated limb.

Tardy Regenerate Ossification

At times, maturation of the regenerate can be maddeningly slow. This problem is more distressing when an external skeletal fixator has been used for the procedure, when compared to an intramedullary lengthening nail. This is because the patient,

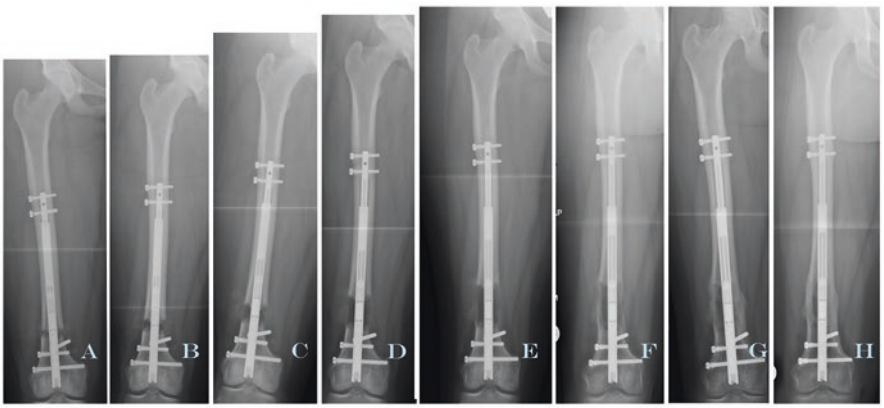


Fig. 8.6 Patience is a virtue. (**a**–**d**) Gradual distraction. (**e**) Wispy new bone formation at the periphery of the regenerate, a common pattern seen in intramedullary lengthening. (**f**–**h**) With partial weight bearing, the regenerate matures

perhaps expecting a short fixator application, comes to hate the device surrounding his or her limb. Indeed, strategies such as lengthening over an intramedullary nail (a combination of an intramedullary nail and external skeletal fixator) or lengthening and then nailing (substituting an intramedullary nail for an external fixator before the bone is fully mature) have become popular with many surgeons [15–17].

An intramedullary lengthening nail is far more tolerable than a cumbersome external device secured to the limb with transosseous pins or wires. Thus, patient-doctor negotiations, so common during external fixation treatment, are rarely as intense with intramedullary devices.

Moreover, it appears that the regenerate matures faster with IM lengthening, in a case matched study, when compared to circular external fixator lengthening [18].

Stimulating Regenerate Ossification

On occasion, regenerate maturation slows progressing or stops altogether, with a radiolucent defect where bone should be forming. Needless to say, any factor—biological, chemical, and mechanical—that inhibits bone formation should be eliminated. Inquiring about over-the-counter anti-inflammatory medication, for instance, may yield a surprising affirmative answer. Poor nutrition, low vitamin D, concomitant diseases, smoking, and other adverse factors will defeat any surgeon waiting for regenerate to mature (Fig. 8.7).

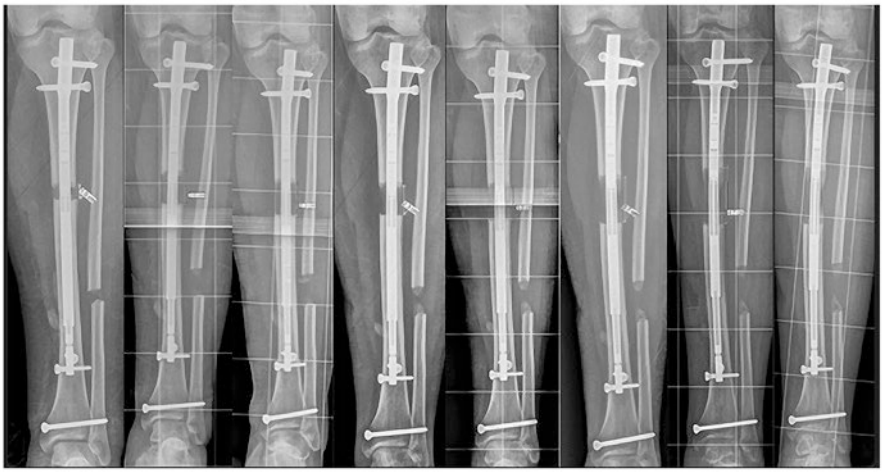


Fig. 8.7 Absent regenerate ossification. At a 30 mm gap size, when faced with absence of early evidence of regenerate new bone in a distraction gap, slow, stop, or even reverse distraction, advancing at a much slower rate thereafter. A bone graft was needed. Courtesy of John E. Herzenberg, M.D. Used with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

Mechanical Enhancement

A modality that has been proven to enhance fracture healing will likely do the same for a distraction gap regenerate. Needless to say, weight bearing is first on the list of mechanical factors that can enhance bone formation within the regenerate. The amount of weight is not as important as the rhythmic pattern, one step per second, typically supported with crutches or a walker at first and then a cane later. The patient must experience what 30 pounds of weight bearing feels like on a bathroom scale, to get the proper sensory feedback from the lower extremity. As the weight increases, the patient needs to check again on a scale, for the same reason as above.

With external fixation lengthening, pain is often a serious inhibitor of ambulation, almost always relate to transcutaneous implant inflammation. Since no through-skin implant is used with intramedullary lengthening nails, such pain will not occur. Likewise, the pain of limb stretching settles down once length goal is achieved, so weight bearing must be encouraged.

Patchy osteopenia of the bone in adjacent regions of the bone surrounding the regenerate is a sign of inadequate weight bearing. After all how could the regenerate ossify if the rest of the limb is de-ossifying?

Pulsed Electromagnetic Field

While the value of electromagnetic stimulation of bone formation is well established in the treatment of non-unions, the results with this modality when applied to regenerate bone formation are less impressive. Although it has been shown in a rabbit limb distraction model that pulsed electromagnetic field (PEMF) reduces osteoporosis in bone adjacent to the distraction gap, the field effect on the regenerate is disappointingly nil [19].

Luna Gonzalez and co-workers in Malaga, Spain [20], applied PEMF to one side of patients undergoing simultaneous bilateral external fixation limb lengthening, starting the tenth day after surgery. They found faster regenerate maturation and greater regenerate bone density on the stimulated side than on the control side, allowing fixator removal 1 month sooner as a result of PEMF stimulation. If this observation is repeated with intramedullary lengthening, widespread use of PEMF in such cases will soon follow.

Pulsed Ultrasound

Low intensity pulsed ultrasound (LIPUS) has been shown to enhance maturation and consolidation of regenerate new bone formation is rabbits [21, 22]. Likewise, a small number of clinical studies have used LIPUS on human subjects, all reports involving external fixation lengthenings. El-Mowafi et al. [6] applied 30 mW/cm^2 to

ten patients after the end of the distraction phase and compared to an equal number of patients without LIPUS. They noted substantial improvement in the healing index, meaning a shortened time in external fixation. Other groups have had similar results with external fixator lengthenings [23, 24].

AS with PEMF studies, LIPUS appears to accelerate normal maturation of the regenerate, but so far, no series has studied the effect of the modality on problematic regenerate.

Chemical Enhancement

Several therapeutic approaches have been proposed to stimulate regenerate formation that utilize purified chemicals including bone morphogenic protein and diphosphonates [25]. Burkhart and Rommens [26] published a case report describing the use of BMP-7 to treat tardy regenerate ossification in a case of external fixator lengthening over an IM nail. They reported successful consolidation after inserting the substance into the marrow canal. Their results are confounded by the fact that they also performed a concurrent exchange nailing—a therapeutic approach known to stimulate healing without the use of BMP.

Kiely et al. [25] reported on seven pediatric patients (average age 13.8) undergoing external fixator lengthenings whom at an average of 170 days into their treatment demonstrated tardy regenerate ossification. Three patients were treated with intravenous pamidronate and four with IV zoledronic acid. Six of the seven cases involved the upper tibia and the seventh case involved the femur. The average lengthening was 4.8 cm. Bone mineral density (BMD) was studied both before and after the diphosphonate intervention. Six of the seven children had an average increase in BMD that was impressive, coming up to 85.6% of the non-operated side. The substances were well tolerated by the children. One patient did not heal with the diphosphonates and required a bone marrow + BMP-7 injection into the regenerate to obtain union.

Biological Enhancement

As noted above, bone marrow and BMP-7 were used to rescue a case of failed diphosphonate treatment of tardy regenerate ossification, a reasonable salvage option. For the same reason, bone marrow cells and platelet-rich plasma combinations have been used by Kitoh and co-workers, reportedly with good success. In one study [27] aspirated mesenchymal stem cells (MSCs) from the iliac crests of 11 patients (achondroplasia and hypochondroplasia) culture-expanded the cells and injected them with platelet-rich plasma (PRP) into the distraction zone in external fixator lengthening cases. The MSCs and PRP were in a thrombin-calcium carrier at

the time of injection. The authors reported significantly enhanced healing and short fixator times when compared to nine patients with similar demographics treated without MSCs and PRP.

MSCs have been transfected with BMP-7 in a rat mandibular distraction study and compared to MSCs without the BMP-7 and mandibles injected with inert saline [28]. The BMP-7-mediated ex vivo gene transfer into MSCs significantly accelerated callus formation in the regenerate and facilitated consolidation.

Bone Grafting

Fresh autogenous bone grafting is the logical choice for deficient regenerate ossification when all else fails. Indeed, surgeons treating skeletal defects with Ilizarov's bone transport protocol often find it necessary to insert autogenous bone graft into the docking site, either prophylactically or as treatment for deficient consolidation of the intercalary transport segment with the target bone. This is done in spite of the risk that surgery with an external fixator still on a limb might spread pin or wire site sepsis into the operative region.

With intramedullary lengthening, the risk of postoperative infection after bone grafting a defect in the regenerate is the same as any clean surgery. Thus, the only reasonable drawback to prompt autogenous bone grafting is donor site morbidity. This feature of orthopedic surgery seems to be technique dependent, with some surgeons claiming minimal (2%) donor site morbidity even when massive iliac bone grafts are obtained [29] and others describing as much as 31% long-term donor site issues [30].

Alternative means of obtaining autogenous bone graft material without a surgical approach to the pelvis include the reamer-irrigator-aspirator (RIA) technique that has become increasingly popular among surgeons. As with any surgical intervention, however, the RIA technique introduces its own set of problems.

Needle aspiration and cell concentration of bone marrow have become another alternate to operative harvesting of autogenous bone graft. Needle marrow aspiration holds great promise for dealing with deficient regenerate, although it is technique sensitive. Over all, the complication rate is one tenth that of open iliac crest bone graft harvesting [31].

Hyperplastic Regenerate

Rarely, the bone forms in the distraction zone too rapidly, a phenomenon occasionally seen in children and achondroplastic patients undergoing limb lengthening. In such cases, the elongation rate must be increased to 1.5 mm per day or occasionally even faster. However, it is noted that rates approaching 3.0 mm per day endanger soft tissue structures, especially nerves and arteries (Fig. 8.8).

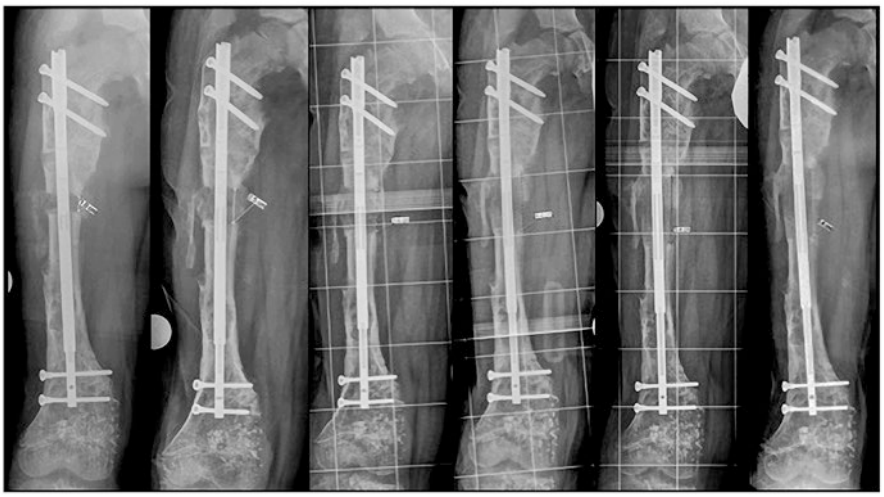


Fig. 8.8 Rapid formation of new bone in the distraction zone. Close follow-up of patients during limb lengthening will allow a surgeon to speed up the rate and rhythm of distraction. Courtesy of John E. Herzenberg, M.D. Used with permission from the Rubin Institute for Advanced Orthopedics, Sinai Hospital of Baltimore

When Length Is Achieved

When the patient's limb has reached the lengthening goal, physical activity must increase, thereby transferring weight-bearing stresses to the newly formed regenerate osseous tissue. At this point, Ilizarov recommends "training the regenerate" by overlengthening the limb 7–10 mm and then compressing the consequent overlong regenerate back down to proper length at a rate of 0.25–0.5 mm every 12 h. In this manner, extra bulk of regenerate new bone will be available to participate in consolidation. This is rarely performed with intramedullary lengthening nails, but is certainly worth considering.

Planning Implant Removal

When lengthening a limb with an external fixator, we leave the frame in place until the regenerate bone in the distraction gap has formed a complete cortex on three sides. Patients are usually anxious to get the fixator off, wanting an end to torn bed sheets, funny-looking clothes, and pin site infections.

With intramedullary limb lengthening, however, there is no rush to remove the nail. While lengthening with an external fixator creates a solid regenerate, filling the entire cylindrical space between the bone ends, the osseous tissue that forms around an intramedullary lengthening nail is a hollow tube of the bone. Therefore, after the nail is removed, any cortical defect could serve as a stress riser, leading to a fracture. For this reason, it is wise to allow the bone to fully mature before nail removal. This takes at least 1 year, and more likely 2 years, after elongation is completed (Fig. 8.9).

Fig. 8.9 Delayed maturation of the regenerate can be very frustrating for the patient. From LLRS Scientific Exhibit, AAOS Annual Meeting, 2011. Copyright 2016 Zeeca Publishing Co

Post-Removal Management

After the implant is removed, it should not be necessary to apply a splint, orthosis, or cast to the patient's limb. The patient should not resume sports, however, until the bone appears normal on roentgenographic evaluation in multiple views.

Conclusion

It is evident from the foregoing narrative that the postoperative management of a patient having his or her limb lengthened by the formation of regenerate bone requires frequent contact and close monitoring by the surgeon. Deformities and contractures cannot be allowed to persist or progress. The patient must be encouraged to partially bear weight on the involved limb lest the newly formed bone fails to mature and corticalize properly.

By following the principles outlined above, a surgeon and his or her patient will have the gratifying experience of elongating a limb to an amount never before thought possible and without undue problems or residual complications.

References

1. Paley D. Problems, obstacles and complications of limb lengthening by the Ilizarov technique. Clin Orthop Rel Res. 1990;250:81–104.
2. Rosbruch SR, Kleinman D, Fragomen AT, Ilizarov S. Limb lengthening and then insertion of an intramedullary nail: a case-matched comparison. Clin Orthop Rel Res. 2008;466:923–32.
3. Li R, Saleh M, Yang L, Coulton L. Radiographic classification of osteogenesis during bone distraction. J Orthop Res. 2006;24(3):339–47.

4. Eski M, Ilgan S, Cil Y, Sengezer M, Ozcan A, Yapici K. Assessment of distraction regenerate using quantitative bone scintigraphy. Ann Plast Surg. 2007;58(3):328–34.
5. Song SH, Agashe M, Kim TY, Sinha S, Park YE, Kim SJ, et al. Serial bone mineral density ratio measurement for fixator removal in tibia distraction osteogenesis and need of a supportive method using the pixel value ratio. J Pediatr Orthop B. 2012;21(2):137–45.
6. El-Mowafi H, Mohsen M. The effect of low-intensity pulsed ultrasound on callus maturation in tibial distraction osteogenesis. Int Orthop. 2005;29(2):121–4.
7. Luk HK, Lai YM, Qin L, Huang Y-P, Zheng Y-P. Computed radiographic and ultrasonic evaluation of bone regeneration during tibial distraction osteogenesis in rabbits. Ultrasound Med Biol 2012;38(10):1744–58.
8. Al-Hadithy N, Sewell MD, Bhavikatti M, Gikas PD. The effect of smoking on fracture healing and on various orthopaedic procedures. Acta Orthop Belg. 2012;78(3):285–90.
9. Donigan JA, Fredericks DC, Nepola JV, Smucker JD. The effect of transdermal nicotine on fracture healing in a rabbit model. J Orthop Trauma. 2012;26(12):724–7.
10. Hastrup SG, Chen X, Bechtold JE, Kyle RF, Rahbek O, Keyler DE, et al. Effect of nicotine and tobacco administration method on the mechanical properties of healing bone following closed fracture. J Orthop Res. 2010;28(9):1235–9.
11. Lynch JR, Taitsman LA, Barei DP, Nork SE. Femoral nonunion: risk factors and treatment options. J Am Acad Orthop Surg. 2008;16(2):88–97.
12. Scolaro JA, Schenker ML, Yannascoli S, Baldwin K, Mehta S, Ahn J. Cigarette smoking increases complications following fracture: a systematic review. J Bone Joint Surg Am. 2014;96(8):674–81.
13. Sloan A, Hussain I, Maqsood M, Eremin O, El-Sheemy M. The effects of smoking on fracture healing. Surg J R Coll Surg Edinb Irel. 2010;8(2):111–6.
14. Jeffcoach DR, Sams VG, Lawson CM, Enderson BL, Smith ST, Kline H, et al. Nonsteroidal anti-inflammatory drugs' impact on nonunion and infection rates in long-bone fractures. J Trauma Acute Care Surg. 2014;76(3):779–83.
15. Paley D, Herzenberg JE, Paremain G, Bhave A. Femoral lengthening over an intramedullary nail. A matched-case comparison with Ilizarov femoral lengthening. J Bone Joint Surg Am. 1997;79(10):1464–80.
16. Rozbruch SR, Birch JG, Dahl MT, Herzenberg JE. Motorized intramedullary nail for management of limb-length discrepancy and deformity. J Am Acad Orthop Surg. 2014;22(7):403–9.
17. Burghardt RD, Manzotti A, Bhave A, Paley D, Herzenberg JE. Tibial lengthening over intramedullary nails: a matched case comparison with Ilizarov tibial lengthening. Bone Joint Res. 2016;5(1):1–10.
18. Horn J, Grimsrud O, Dagsgard AH, Huhnstock S, Steen H. Femoral lengthening with a motorized intramedullary nail. Acta Orthop. 2015;86(2):248–56.
19. Eyres KS, Saleh M, Kanis JA. Effect of pulsed electromagnetic fields on bone formation and bone loss during limb lengthening. Bone. 1996;18(6):505–9.
20. Luna Gonzalez F, Lopez Arevalo R, Meschian Coretti S, Urbano Labajos V, Delgado Rufino B. Pulsed electromagnetic stimulation of regenerate bone in lengthening procedures. Acta Orthop Belg. 2005;71(5):571–6.
21. Chan CW, Qin L, Lee KM, Zhang M, Cheng JC, Leung KS. Low intensity pulsed ultrasound accelerated bone remodeling during consolidation stage of distraction osteogenesis. J Orthop Res. 2006;24(2):263–70.
22. Tobita K, Ohnishi I, Matsumoto T, Ohashi S, Bessho M, Kaneko M, et al. Effect of low-intensity pulsed ultrasound stimulation on callus remodelling in a gap-healing model: evaluation by bone morphometry using three-dimensional quantitative micro-CT. J Bone Joint Surg Br. 2011;93(4):525–30.
23. Gold SM, Wasserman R. Preliminary results of tibial bone transports with pulsed low intensity ultrasound (Exogen). J Orthop Trauma. 2005;19(1):10–6.
24. Salem KH, Schmelz A. Low-intensity pulsed ultrasound shortens the treatment time in tibial distraction osteogenesis. Int Orthop. 2014;38(7):1477–82.

25. Kiely P, Ward K, Bellemore CM, Briody J, Cowell CT, Little DG. Bisphosphonate rescue in distraction osteogenesis: a case series. J Pediatr Orthop. 2007;27(4):467–71.
26. Burkhart KJ, Rommens PM. Intramedullary application of bone morphogenetic protein in the management of a major bone defect after an Ilizarov procedure. J Bone Joint Surg Br. 2008;90(6):806–9.
27. Kitoh H, Kitakoji T, Tsuchiya H, Katoh M, Ishiguro N. Distraction osteogenesis of the lower extremity in patients with achondroplasia/hypochondroplasia treated with transplantation of culture-expanded bone marrow cells and platelet-rich plasma. J Pediatr Orthop. 2007;27(6):629–34.
28. Hu J, Qi MC, Zou SJ, Li JH, Luo E. Callus formation enhanced by BMP-7 ex vivo gene therapy during distraction osteogenesis in rats. J Orthop Res. 2007;25(2):241–51.
29. Loeffler BJ, Kellam JF, Sims SH, Bosse MJ. Prospective observational study of donor-site morbidity following anterior iliac crest bone-grafting in orthopaedic trauma reconstruction patients. J Bone Joint Surg Am. 2012;94(18):1649–54.
30. Sasso RC, LeHuec JC, Shaffrey C. Iliac crest bone graft donor site pain after anterior lumbar interbody fusion: a prospective patient satisfaction outcome assessment. J Spinal Disord Tech. 2005;18(Suppl):S77–81.
31. Hernigou P, Desroches A, Queinnec S, Flouzat Lachaniette CH, Poignard A, Allain J, et al. Morbidity of graft harvesting versus bone marrow aspiration in cell regenerative therapy. Int Orthop. 2014;38(9):1855–60.

Further Reading

1. Aldegheri R, Volino C, Zambito A, Tessari G, Trivella G. Use of ultrasound to monitor limb lengthening by callotasis. J Pediatr Orthop B. 1993;2(1):22–7.
2. Al-Hadithy N, Sewell MD, Bhavikatti M, Gikas PD. The effect of smoking on fracture healing and on various orthopaedic procedures. Acta Orthop Belg. 2012;78(3):285–90.
3. Babatunde OM, Fragomen AT, Rozbruch SR. Noninvasive quantitative assessment of bone healing after distraction osteogenesis. HSS J. 2010;6(1):71–8.
4. Beitzel K, McCarthy MB, Cote MP, Durant TJ, Chowaniec DM, Solovyova O, et al. Comparison of mesenchymal stem cells (osteoprogenitors) harvested from proximal humerus and distal femur during arthroscopic surgery. Arthrosc J Arthrosc Relat Surg Off Publ Arthrosc Assoc North Am Int Arthrosc Assoc. 2013;29(2):301–8.
5. Burghardt RD, Manzotti A, Bhave A, Paley D, Herzenberg JE. Tibial lengthening over intramedullary nails: a matched case comparison with Ilizarov tibial lengthening. Bone Joint Res. 2016;5(1):1–10.
6. Calori GM, Colombo M, Mazza EL, Mazzola S, Malagoli E, Mineo GV. Incidence of donor site morbidity following harvesting from iliac crest or RIA graft. Injury. 2014;45(Suppl 6):S116–20.
7. Chan CW, Qin L, Lee KM, Zhang M, Cheng JC, Leung KS. Low intensity pulsed ultrasound accelerated bone remodeling during consolidation stage of distraction osteogenesis. J Orthop Res. 2006;24(2):263–70.
8. Cho TJ, Choi IH, Lee SM, Chung CY, Yoo WJ, Lee DY, et al. Refracture after Ilizarov osteosynthesis in atrophic-type congenital pseudarthrosis of the tibia. J Bone Joint Surg Br. 2008;90(4):488–93.
9. Crist BD, Stoker AM, Stannard JP, Cook JL. Analysis of relevant proteins from bone graft harvested using the reamer irrigator and aspirator system (RIA) versus iliac crest (IC) bone graft and RIA waste water. Injury. 2016;47(8):1661–8.
10. Davis RL, Taylor BC, Johnson N, Ferrel JR, Castaneda J. Retrograde versus antegrade femoral bone graft harvesting using the reamer-irrigator-aspirator. J Orthop Trauma. 2015;29(8):370–2.

11. Devine DM, Arens D, Thalhauser M, Schiuma D, Zeiter S, Nehrbass D. Healing pattern of reamed bone following bone harvesting by a RIA device. Eur Cells Mater. 2015;29:97–104. discussion
12. Donders JC, Baumann HM, Stevens MF, Kloen P. Hemorrhagic induced cardiovascular complications during Reamer-Irrigator-Aspirator assisted femoral nonunion treatment. J Orthop Trauma. 2016;30(9):e294–8.
13. Donigan JA, Fredericks DC, Nepola JV, Smucker JD. The effect of transdermal nicotine on fracture healing in a rabbit model. J Orthop Trauma. 2012;26(12):724–7.
14. Donnan LT, Saleh M, Rigby AS, McAndrew A. Radiographic assessment of bone formation in tibia during distraction osteogenesis. J Pediatr Orthop. 2002;22(5):645–51.
15. Dudda M, Hauser J, Muhr G, Esenwein SA. Low-intensity pulsed ultrasound as a useful adjuvant during distraction osteogenesis: a prospective, randomized controlled trial. J Trauma. 2011;71(5):1376–80.
16. Fu XM, Fan SG, Li SL, Chen YS, Wu H, Guo YL. Low 25(OH) D serum levels are related with hip fracture in postmenopausal women: a matched case-control study. J Transl Med. 2015;13:388.
17. Paley D. Problems, obstacles, and complications of limb lengthening by the Ilizarov technique. Clin Orthop Relat Res. 1990;(250):81–104.
18. Gorter EA, Krijnen P, Schipper IB. Vitamin D deficiency in adult fracture patients: prevalence and risk factors. Eur J Trauma Emerg Surg Off Publ Eur Trauma Soc. 2016;42(3):369–78.
19. Gruson KI, Aharonoff GB, Egol KA, Zuckerman JD, Koval KJ. The relationship between admission hemoglobin level and outcome after hip fracture. J Orthop Trauma. 2002;16(1):39–44.
20. Han F, Peter L, Lau ET, Thambiah J, Murphy D, Kagda FH. Reamer irrigator aspirator bone graft harvesting: complications and outcomes in an Asian population. Injury. 2015;46(10):2042–51.
21. Harbacheuski R, Fragomen AT, Rozbruch SR. Does lengthening and then plating (LAP) shorten duration of external fixation? Clin Orthop Relat Res. 2012;470(6):1771–81.
22. Harshwal RK, Sankhala SS, Jalan D. Management of nonunion of lower-extremity long bones using mono-lateral external fixator--report of 37 cases. Injury. 2014;45(3):560–7.
23. Hastrup SG, Chen X, Bechtold JE, Kyle RF, Rahbek O, Keyler DE, et al. Effect of nicotine and tobacco administration method on the mechanical properties of healing bone following closed fracture. J Orthop Res. 2010;28(9):1235–9.
24. Hazra S, Song HR, Biswal S, Lee SH, Lee SH, Jang KM, et al. Quantitative assessment of mineralization in distraction osteogenesis. Skelet Radiol. 2008;37(9):843–7.
25. Hegde V, Shonuga O, Ellis S, Fragomen A, Kennedy J, Kudryashov V, et al. A prospective comparison of 3 approved systems for autologous bone marrow concentration demonstrated nonequivalency in progenitor cell number and concentration. J Orthop Trauma. 2014;28(10):591–8.
26. Hendrich C, Franz E, Waertel G, Krebs R, Jager M. Safety of autologous bone marrow aspiration concentrate transplantation: initial experiences in 101 patients. Orthop Rev. 2009;1(2):e32.
27. Hernigou P, Homma Y, Flouzat Lachaniette CH, Poignard A, Allain J, Chevallier N, et al. Benefits of small volume and small syringe for bone marrow aspirations of mesenchymal stem cells. Int Orthop. 2013;37(11):2279–87.
28. Hernigou J, Picard L, Alves A, Silvera J, Homma Y, Hernigou P. Understanding bone safety zones during bone marrow aspiration from the iliac crest: the sector rule. Int Orthop. 2014;38(11):2377–84.
29. Hernigou J, Alves A, Homma Y, Guissou I, Hernigou P. Anatomy of the ilium for bone marrow aspiration: map of sectors and implication for safe trocar placement. Int Orthop. 2014;38(12):2585–90.
30. Hernigou P, Desroches A, Queinnec S, Flouzat Lachaniette CH, Poignard A, Allain J, et al. Morbidity of graft harvesting versus bone marrow aspiration in cell regenerative therapy. Int Orthop. 2014;38(9):1855–60.
31. Hughes TH, Maffulli N, Green V, Fixsen JA. Imaging in bone lengthening. A review. Clin Orthop Relat Res. 1994;(308):50–3.

32. Jeffcoach DR, Sams VG, Lawson CM, Enderson BL, Smith ST, Kline H, et al. Nonsteroidal anti-inflammatory drugs' impact on nonunion and infection rates in long-bone fractures. J Trauma Acute Care Surg. 2014;76(3):779–83.
33. Kemper O, Herten M, Fischer J, Haversath M, Beck S, Classen T, et al. Prostacyclin suppresses twist expression in the presence of indomethacin in bone marrow-derived mesenchymal stromal cells. Med Sci Monit Int Med J Exp Clin Res. 2014;20:2219–27.
34. Kitoh H, Kitakoji T, Tsuchiya H, Mitsuyama H, Nakamura H, Katoh M, et al. Transplantation of marrow-derived mesenchymal stem cells and platelet-rich plasma during distraction osteogenesis--a preliminary result of three cases. Bone. 2004;35(4):892–8.
35. Kitoh H, Kitakoji T, Tsuchiya H, Katoh M, Ishiguro N. Distraction osteogenesis of the lower extremity in patients with achondroplasia/hypochondroplasia treated with transplantation of culture-expanded bone marrow cells and platelet-rich plasma. J Pediatr Orthop. 2007;27(6):629–34.
36. Kuehlfluck P, Moghaddam A, Helbig L, Child C, Wildemann B, Schmidmaier G. RIA fractions contain mesenchymal stroma cells with high osteogenic potency. Injury. 2015;46(Suppl 8):S23–32.
37. Lee JH, Hutzler LH, Shulman BS, Karia RJ, Egol KA. Does risk for malnutrition in patients presenting with fractures predict lower quality measures? J Orthop Trauma. 2015;29(8):373–8.
38. Ko KI, Coimbra LS, Tian C, Alblowi J, Kayal RA, Einhorn TA, et al. Diabetes reduces mesenchymal stem cells in fracture healing through a TNF alpha-mediated mechanism. Diabetologia. 2015;58(3):633–42.
39. Lowe JA, Crist BD, Pfeiffer F, Carson WL. Predicting reduction in torsional strength by concentric/eccentric RIA reaming normal and osteoporotic long bones (femurs). J Orthop Trauma. 2015;29(10):e371–9.
40. Lowenberg DW, Buntic RF, Buncke GM, Parrett BM. Long-term results and costs of muscle flap coverage with Ilizarov bone transport in lower limb salvage. J Orthop Trauma. 2013;27(10):576–81.
41. Lynch JR, Taitsman LA, Barei DP, Nork SE. Femoral nonunion: risk factors and treatment options. J Am Acad Orthop Surg. 2008;16(2):88–97.
42. Mansour J, Conway JD, et al. Am J Orthop. 2015;44(5):202–5.
43. Markel MD, Chao EY. Noninvasive monitoring techniques for quantitative description of callus mineral content and mechanical properties. Clin Orthop Relat Res. 1993;(293):37–45.
44. Miller AN, Deal D, Green J, Houle T, Brown W, Thore C, et al. Use of the reamer/irrigator/aspirator decreases carotid and cranial embolic events in a canine model. J Bone Joint Surg Am. 2016;98(8):658–64.
45. Muzaffar N, Hafeez A, Modi H, Song HR. Callus patterns in femoral lengthening over an intramedullary nail. J Orthop Res. 2011;29(7):1106–13.
46. Narbona-Carceles J, Vaquero J, Suarez-Sancho S, Forriol F, Fernandez-Santos ME. Bone marrow mesenchymal stem cell aspirates from alternative sources: is the knee as good as the iliac crest? Injury. 2014;45(Suppl 4):S42–7.
47. Nauth A, Lane J, Watson JT, Giannoudis P. Bone graft substitution and augmentation. J Orthop Trauma. 2015;29(Suppl 12):S34–8.
48. Oh CW, Apivatthakakul T, Oh JK, Kim JW, Lee HJ, Kyung HS, et al. Bone transport with an external fixator and a locking plate for segmental tibial defects. Bone Joint J. 2013;95(12):1667–72.
49. Paley D. Problems, obstacles and complications of limb lengthening by the Ilizarov technique. Clin Orthop Rel Res. 1990;250:81–104.
50. Paley D, Herzenberg JE, Paremain G, Bhave A. Femoral lengthening over an intramedullary nail. A matched-case comparison with Ilizarov femoral lengthening. J Bone Joint Surg Am. 1997;79(10):1464–80.
51. Paley D. Principles of deformity correction. Berlin: Springer; 2002.
52. Rankine JJ, Hodgson RJ, Tan HB, Cox G, Giannoudis PV. MRI appearances of the femur following bone graft harvesting using the Reamer-Irrigator-Aspirator. Injury. 2015;46(Suppl 8):S65–7.

53. Rigal S, Merloz P, Le Nen D, Mathevon H, Masquelet AC. Bone transport techniques in post-traumatic bone defects. Orthop Traumatol Surg Res: OTSR. 2012;98(1):103–8.
54. Sabharwal S. Enhancement of bone formation during distraction osteogenesis: pediatric applications. J Am Acad Orthop Surg. 2011;19(2):101–11.
55. Sala F, Marinoni E, Miller AN, Pesenti G, Castelli F, Alati S, et al. Evaluation of an endoscopic procedure for the treatment of docking site nonunion. J Orthop Trauma. 2013;27(10):569–75.
56. Sandberg O, Aspenberg P. Different effects of indomethacin on healing of shaft and metaphyseal fractures. Acta Orthop. 2015;86(2):243–7.
57. Scolaro JA, Schenker ML, Yannascoli S, Baldwin K, Mehta S, Ahn J. Cigarette smoking increases complications following fracture: a systematic review. J Bone Joint Surg Am. 2014;96(8):674–81.
58. Singh S, Song HR, Venkatesh KP, Modi HN, Park MS, Jang KM, et al. Analysis of callus pattern of tibia lengthening in achondroplasia and a novel method of regeneration assessment using pixel values. Skelet Radiol. 2010;39(3):261–6.
59. Sloan A, Hussain I, Maqsood M, Eremin O, El-Sheemy M. The effects of smoking on fracture healing. Surg J R Coll Surg Edinb Irel. 2010;8(2):111–6.
60. Song SH, Sinha S, Kim TY, Park YE, Kim SJ, Song HR. Analysis of corticalization using the pixel value ratio for fixator removal in tibial lengthening. J Orthop Sci. 2011;16(2):177–83.
61. Song SH, Agashe M, Kim TY, Sinha S, Park YE, Kim SJ, et al. Serial bone mineral density ratio measurement for fixator removal in tibia distraction osteogenesis and need of a supportive method using the pixel value ratio. J Pediatr Orthop B. 2012;21(2):137–45.
62. Sun X-T, Easwar TR, Stephen M, Song S-H, Kim S-J, Song H-R. Comparative study of callus progression in limb lengthening with or without intramedullary nail with reference to the pixel value ratio and the Ru Li's classification. Arch Orthop Trauma Surg. 2011;131(10):1333–40.
63. Tashjian RZ, Horwitz DS. Healing and graft-site morbidity rates for midshaft clavicle nonunions treated with open reduction and internal fixation augmented with iliac crest aspiration. Am J Orthop. 2009;38(3):133–6.
64. Taylor KF, Inoue N, Rafiee B, Tis JE, McHale KA, Chao EY. Effect of pulsed electromagnetic fields on maturation of regenerate bone in a rabbit limb lengthening model. J Orthop Res. 2006;24(1):2–10.
65. Tobita K, Ohnishi I, Matsumoto T, Ohashi S, Bessho M, Kaneko M, et al. Effect of low-intensity pulsed ultrasound stimulation on callus remodelling in a gap-healing model: evaluation by bone morphometry using three-dimensional quantitative micro-CT. J Bone Joint Surg Br. 2011;93(4):525–30.
66. Urabe K, Hotokebuchi T, Oles KJ, Bronk JT, Jingushi S, Iwamoto Y, et al. Inhibition of endochondral ossification during fracture repair in experimental hypothyroid rats. J Orthop Res. 1999;17(6):920–5.
67. Warner SJ, Garner MR, Nguyen JT, Lorich DG. Perioperative vitamin D levels correlate with clinical outcomes after ankle fracture fixation. Arch Orthop Trauma Surg. 2016;136(3):339–44.
68. Young JW, Kovelman H, Resnik CS, Paley D. Radiologic assessment of bones after Ilizarov procedures. Radiology. 1990;177(1):89–93.
69. Zhao L, Fan Q, Venkatesh KP, Park MS, Song HR. Objective guidelines for removing an external fixator after tibial lengthening using pixel value ratio: a pilot study. Clin Orthop Relat Res. 2009;467(12):3321–6.

Alternatives to Limb Lengthening

9

Pelvic Tilting

Children and teenagers, both boys and girls, often omit corrective footwear and hide a short-limb limp as best they can. This entails walking with their pelvis tilted toward the short side, with compensatory curvature of the spine. While such postural scoliosis disappears when lying down, long-term damage to the spinal facet joints may doom the youngster to a lifetime of spinal problems (Fig. 9.1).

For these reasons, the indications for surgical limb elongation have gradually become more liberal, especially with the introduction of intramedullary lengthening nails. Sadly, health insurance providers, whose TV and print advertisements tout some variant of "We Care," do not seem particularly mindful about issues important to their enrollees.

Shoe Lifts

Classically, shoe lifts proved the simplest solution to limb length discrepancies. Although long shoe lifts measuring several centimeters are clunky, socially embarrassing, and risk ankle injury, lifts in the 2.5 cm (1 in.) range should prove well tolerated, especially by men (Fig. 9.2). A 2.5 cm lift—half inside the shoe and half outside—is barely detectable when seen from above.

For women, whose dressy shoes are often subject to close inspection by others, self-consciousness accompanies even the smallest visible lift. A 1.0 cm heel lift inside a shoe may be tolerably hidden from view, but even an insert this small tends to push the heel up and out of the shoe, especially when wearing pumps and similar non-orthopedic footwear.

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_9

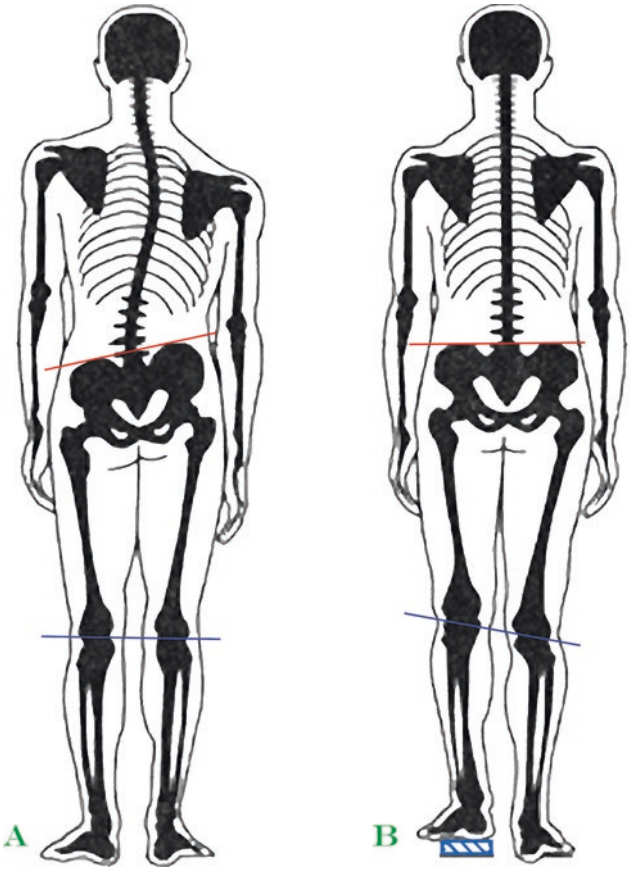


Fig. 9.1 (**a**) Hiding a short femur by pelvic tilting (*red line*). This creates a compensatory spinal curvature. The knees are the same height from the ground because the tibias are the same length. (**b**) Compensation with a shoe lift (*blue box*). The pelvis is now level and the spine is straight, but the knees are not at the same level (*blue line*)

Surgery on the Normal Side

Growth Arrest

Before the advent of distraction osteogenesis, the most common way of dealing with a modest lower extremity length discrepancy in a growing child was to retard or stop the growth of the opposite limb. Using growth charts to predict the difference in limb length at skeletal maturity, a surgeon could span a still-growing epiphyseal plate with thick bone staples, thereby slowing its growth. The staples could either be left in place until the growth plate closed completely or, if some additional growth was needed on the normal side, the staples could be removed before maturity, anticipating further growth. The results, however, are not always predictably successful.

Fig. 9.2 Exterior shoe lifts as small as 1 in. (*left*) can be a source of social embarrassment for many people

When growth charts indicate that complete growth arrest is the best way to equalize limb length, permanent closure of the epiphyseal plate is easily accomplished by drilling out its cartilaginous center. This allows a bone bridge to form across the plate, ending its growth.

These two methods of epiphysiodesis (surgically stopping growth) are relatively simple outpatient procedures, a feature that makes them appealing to surgeons and parents alike. Sadly, the still-growing patient, a minor, has no legal status in the decision-making process, although it is the youngster that bears the lifelong consequences of the decision.

What adult has not, at one time or another during their lifetime, wished to be taller? In the case of an adult whose growth was *intentionally* stopped before reaching full height potential, much wishful thinking and resentment might occur—especially now that intramedullary lengthening nails have eliminated the most distressing features of external fixator limb elongation.

Shortening the Normal Limb

Once a child transitions into adulthood, the potential for stopping growth no longer exists, so the only option for equalizing limb length without lengthening the stunted limb is to shorten the normal one. Years ago, the standard shortening procedure involved open surgical removal of a predetermined length of bone, acutely closing the resultant defect and stabilizing the involved bone with a plate and screws.

Intramedullary Segmental Shortening

More recently, techniques have evolved to perform the entire shortening operation with intramedullary instruments [1]. First, the surgeon reams out the medullary canal and inserts a saw that cuts the cortex transversely from the inside. Two such bone cuts, separated by the requisite distance, create a floating bone segment equal to the length of the planned shortening. Next, the surgeon uses a back-cutting chisel, shaped like a hook, to split the floating segment in half longitudinally. The surgeon uses instruments to maneuver the two half-cortices outward—into the surrounding soft tissues—and collapses the bone to its final length. Placement of a standard (locking) intramedullary nail completes the surgery.

The x-ray image following such an operation is odd-looking, indeed: two half-bone segments—floating forever in the soft tissues—surround the shortened bone.

Shortening any normal bone in an adult also shortens the surrounding soft tissues. Although shortened skin may shrink somewhat, shortened muscles no longer function optimally, weakening them. While 2.5 cm (1 in.) of muscle shortening is reasonably well tolerated, a 5.0-cm (2-in.) decrease in muscle length weakens it. Nevertheless, in the days before intramedullary lengthening nails, some residual weakness seemed preferable to an unpleasant external fixation experience on the stunted limb.

Now, however, since an operation to elongate the short limb with an intramedullary lengthening nail offers a similar perioperative patient experience to shortening a normal limb with intramedullary techniques, we anticipate the eventual relegation of the intramedullary saw and back-cutting chisel to the museum of historic orthopedic devices.

From a philosophical perspective, many of us in the limb lengthening community are loath to operate on a normal limb to equalize it to an abnormal one. We see it this way: Would a farmer with a healthy chicken and a sick chicken kill the healthy chicken to make soup for the sick chicken? We doubt it.

Amputation

Major Limb Deficiencies

In the past, before Ilizarov developed a reliable method of overcoming substantial limb length inequalities, certain conditions—especially those manifesting at birth or developing shortly thereafter—were best managed with amputation and prosthetic fitting, often starting in early childhood. Clearly, a well-functioning artificial limb is superior to a shoe lift that measures 12.5 cm (5 in.) or more. The greater the discrepancy with the opposite side, the more desirable the prosthesic option becomes.

Ilizarov and his co-workers have developed remarkable limb lengthening strategies utilizing his circular frame's adaptability to overcome many birth defects and early developmental maladies previously considered incurable. Proximal femoral focal deficiency, fibular hemimelia, tibial agenesis, and related disorders can now be successfully treated. Unfortunately, the therapeutic approach proves quite an ordeal for the patient, often requiring several separate frame applications during the growing years.

With the introduction of a reliable intramedullary lengthening nail, surgeons can now offer patients and their families hope that serious limb deficiencies can be treated without the inconvenience of external fixators and the concomitant risk of potentially long-lasting—even permanent—pin or wire site infections.

In some congenital limb deformities, the limb must first be shortened to reconstruct an abnormal joint. While this seems paradoxical, relieving tight soft tissues constitutes a requisite first step for bone repositioning osteotomies. Paley, of Miami, Florida, is a pioneering developer and advocate of this approach.

The availability of fully implantable lengthening devices means that, with sufficient ingenuity and long-term planning, surgeons will increasingly abandon external fixators and use intramedullary lengthening methods to compliment joint reconstruction operations, offering hope for previously untreatable conditions.

Assessing the Medical Literature

The Limb Lengthening and Reconstruction Society-North America

The North America's limb lengthening surgeons meet annually to share new techniques devised to deal with the aforementioned conditions—and others as well. The excitement generated by intramedullary lengthening nails is unprecedented among these practitioners. Any American or Canadian surgeon planning to become involved with the exciting developments in limb lengthening and deformity correction should join the Limb Lengthening and Reconstruction Society-North America (LLRS-NA). Comparable organizations exist in most countries and regions of the world.

Problems with Publications

Surgeons are anxious to apply new diagnostic tools to their clinical armamentarium if the measures improve patient outcomes. Innovative practitioners who develop such concepts are usually the first to introduce them to the healthcare community. Often, developer enthusiasm blinds innovators to the limitations of their brainchild. Thus, the medical literature is replete with examples of therapies that first appear with great fanfare, but don't pan out over time. Rarely does the innovator hearken the demise of his or her own creation. Instead, it is other practitioners who do the deed.

Realistic assessment of techniques and devices to treat limb length deficiencies and deformities has proven difficult because of the wide range of pathological conditions that cause shortening and angulation. Therefore, comparison series whereby surgeons assess the merits of one device against another are very sensitive to complexity factors. Obviously, the surgeon with the most challenging and difficult deformities will likely have poorer outcomes than those who treat simpler conditions. Likewise, age and comorbidity factors also weigh heavily on outcomes, making comparisons exceedingly difficult.

The LLRS-AIM Complexity Score

To overcome these difficulties, LLRS-NA formed an ad hoc committee to solve the complexity problem. They created a complexity score that considers the number of planes of deformity; the amount of lengthening required; the clinical risk factors; the character of soft tissue defects; the greatest angular deformity to be corrected; the quality of the bone, e.g., osteoporotic and infected; and adjacent joint stability. The values for each of these seven domains are added together to yield a final complexity score, which ranges from 0 (least complex) to 28 (high complexity). The authors of the study, led by James McCarthy [2], are given the acronym LLRS-AIM, which stands for location, length, risk, soft tissues, angulation, infection, and motion. It is reproduced on the following page .

Limb Lengthening and Reconstruction Society AIM severity score	
Location (number of deformities per limb of ≥10° in separate planes and rotation all count as separate deformities)	
No deformity	0
One deformity	1
Two deformities	2
Three deformities	3
More than three deformities	4
Leg length inequality (estimate at skeletal maturity)	
0 to 2 cm	0
>2 to 5 cm	1
>5 to 10 cm	2
>10 to 15 cm	3
>15 cm	4
Risk factors (assess clinically)	
None	0
Age of less than 5 or more than 40 years	Add 1 point
Smoker	Add 1 point
Obesity	Add 1 point
Other diseases (e.g., diabetes)	Add 1 point
Soft tissue coverage	
Normal	0
Bruising or contusion	1
Scarring (open grade I)	2
Poor coverage (open grade II)	3
Inadequate coverage (open grade III)	4
Angular deformity (measure and assign greatest primary deformity)	
0° to 10°	0
>10° to 20°	1
>20° to 40°	2
>40° to 60°	3
>60°	4
Infection and bone quality (select the most severe)	
Normal	0
Osteoporotic	1
Dysplastic	2
Infection	3
Combination	4

Limb Lengthening and Reconstruction Society AIM severity score	
Motion and stability of the joints above and below	
Normal	0
Decreased motion (<60% of normal)	1
Subluxation of joint	2
Dislocation of joint	3
More than one joint affected	4
LLRS-AIM index scoring (scores range from a minimum of 0 points to a maximum of 28 points)	
Normal	0
Minimal complexity	1–5
Moderate complexity	6–10
Substantial complexity	11–15
High complexity	16–20

Judging Patient Wishes

Patient-Reported Outcomes

To scientifically quantify the value to patients of limb elongation and deformity correction, the author and other members of LLRS-NA have developed a patient-reported outcome (PRO) questionnaire. The PRO is administered to patients both before and after surgery to eliminate limb length inequalities and deformities. The questionnaire was modeled after a similar document created by the Scoliosis Research Society [3].

Unlike questionnaires that measure the value of operations like joint replacement surgery (that focus on pain and function), our Limb Deformity-Scoliosis Research Society (LD-SRS) questionnaire emphasizes issues like social embarrassment, reaction to meeting strangers, and the like, as well as pain and function, both before and after correction.

Preliminary results, recently published, have validated the reliability of the LD-SRS questionnaire. Such evidence-based data should overcome resistance by insurance companies to pay for treatment designed to eliminate even small limb length differences—an issue extremely important to patients.

The entire questionnaire follows. It is in the public domain and may be used free of charge to assess a patient's outcome from his or her perspective.

The Limb Deformity Modified SRS (LD-SRS) Score

Examination: ☐Pre–treatment ☐3 mos. ☐6 mos. ☐1 year ☐2 years

In the following questionnaire, the term "limb" refers to the body part currently being treated, including the joint above it and the joint below it. If more than one body part is involved in the current treatment plan, please fill out a separate questionnaire for each one.

Side: ☐Right ☐Left

Body Part: ☐Upper Arm ☐Forearm ☐Hand/Wrist ☐Thigh ☐Lower Leg ☐Foot/Ankle

Please select the one best answer to each question unless otherwise indicated. If you already have had surgery, please complete sections 1 and 2. Otherwise, just complete section 1.

All results will be kept confidential.

Section I: All patients

1. Which one of the following best describes the amount of limb pain you have experienced during the past 6 months?
   ☐None ☐Mild ☐Moderate ☐Moderate to severe ☐Severe

2. Which one of the following best describes the amount of limb pain you have experienced over the last month?
   ☐None ☐Mild ☐Moderate ☐Moderate to severe ☐Severe

3. During the past 6 months have you been a very nervous person?
   ☐None of the time ☐A little of the time ☐Some of the time
   ☐Most of the time ☐All of the time

4. If you had to spend the rest of your life with your limb shaped as it is right now, how would you feel about it?
   ☐Very happy ☐Somewhat happy ☐Neither happy nor unhappy
   ☐Somewhat unhappy ☐Very unhappy

5. What is your current level of activity?
   ☐Full activities without restriction
   ☐Moderate manual labor and moderate sports, such as walking and biking
   ☐Primarily no activity
   ☐Light labor, such as household chores
   ☐Bedridden/wheelchair

6. How do you look in clothes?
   ☐Very good ☐Good ☐Fair
   ☐Bad ☐Very bad

7. In the past 6 months have you felt so down in the dumps that nothing could cheer you up?
   ☐Never ☐Rarely ☐Sometimes
   ☐Often ☐Very often

8. Do you experience limb pain when at rest?
   ☐Never ☐Rarely ☐Sometimes
   ☐Often ☐Very often

9. What is your current level of work/school activity?
   ☐100% normal ☐75% normal ☐50% normal
   ☐25% normal ☐0% normal

10. Which of the following best describes the appearance of your limb:
    ☐Very good ☐Good ☐Fair
    ☐Bad ☐Very Bad

11. Which one of the following best describes your medication usage for your limb?
    ☐None
    ☐Non-narcotics weekly or less (e.g., Tylenol, Ibuprofen)
    ☐Narcotics weekly or less (e.g., Percocet, Lorcet, Codeine, Darvocet)
    ☐Other (please specify below)
    Medication:
    Usage: (weekly or less or daily):

12. Does your limb limit your ability to do things around the house?
    ☐Never ☐Rarely ☐Sometimes
    ☐Often ☐Very often

13. Have you felt calm and peaceful during the past 6 months?
    ☐All of the time ☐Most of the time ☐Some of the time
    ☐A little of the time ☐None of the time

14. Do you feel that your limb condition affects your personal relationships?
    ☐None ☐Slightly ☐Mildly ☐Moderately ☐Severely

15. Are you and/or your family experiencing financial difficulties because of your limb?
    ☐None ☐Slightly ☐Mildly ☐Moderately ☐Severely

16. In the past 6 months have you felt downhearted and blue?
    ☐Never ☐Rarely ☐Sometimes
    ☐Often ☐Very often

17. In the last 3 months have you taken any sick days from work/school due to limb pain and, if so, how many?
    ☐0 ☐1 ☐2 ☐3 ☐4 or more

18. Do you go out more or less than your friends?
    ☐Much more ☐More ☐Same
    ☐Less ☐Much less

19. Do you feel attractive with your current limb condition?
☐Yes, very
☐Yes, somewhat
☐Neither attractive nor unattractive
☐No, not very much
☐No, not at all

20. Have you felt calm and peaceful during the past 6 months?
☐All of the time ☐Most of the time ☐Some of the time
☐A little of the time ☐None of the time

Section 2: After Completion of Treatment

21. Are you satisfied with the results of your limb management?
☐Very satisfied ☐Satisfied ☐Neither satisfied nor unsatisfied
☐Unsatisfied ☐Very unsatisfied

22. Would you have the same management again if you had the same condition?
☐Definitely yes ☐Probably yes ☐Not sure
☐Probably not ☐Definitely not

23. On a scale of 1 to 9, with 1 being very low and 9 being extremely high, how would you rate your self-image?
☐1 ☐2 ☐3 ☐4 ☐5 ☐6 ☐7 ☐8 ☐9

24. Compared with before treatment, how do you feel you now look?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

25. Has your limb treatment changed your function and daily activity?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

26. Has your limb treatment changed your ability to enjoy sports/hobbies?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

27. How has your limb treatment changed your limb pain?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

28. Has your treatment changed your confidence in personal relationships with others?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

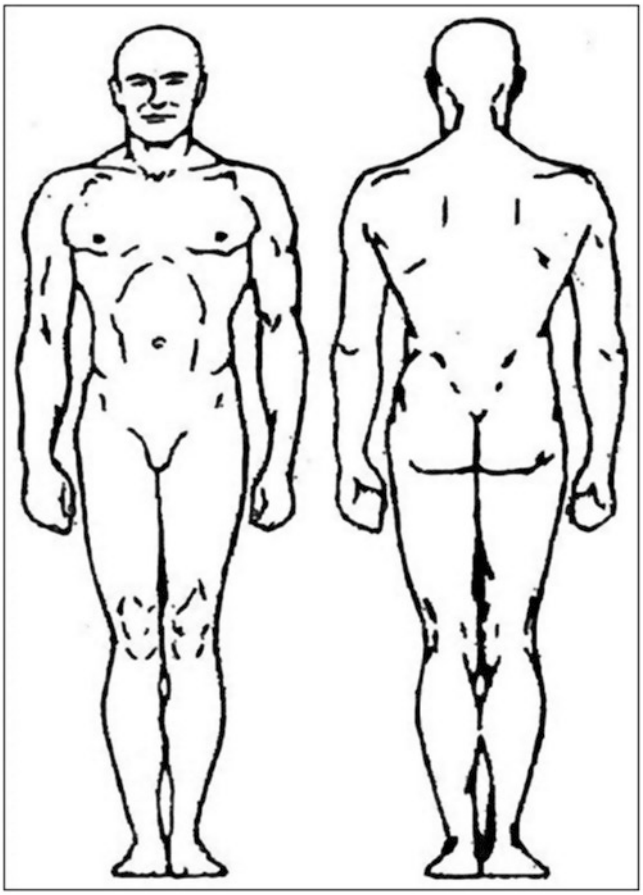
29. Has your treatment changed the way others view you?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

30. Has your treatment changed your self-image?
☐Much better ☐Better ☐Same ☐Worse ☐Much worse

Please mark on the drawings any areas where you feel pain. If you are not having any pain, leave blank and initial.

Use the following key to show particular types of pain

KEY: Pins & needles = 000000
Burning = XXXXXX
Stabbing = IIIIIII
Deep ache = ZZZZZZ



References

1. Winquist R, Hansen S, Pearson R. Closed intramedullary shortening of the femur. Clin Orthop Relat Res. 1978;136:54–61.
2. McCarthy JJ, Iobst CA, Rozbruch SR, Sabharwal S, Eismann EA. Limb Lengthening and Reconstruction Society AIM index reliably assesses lower limb deformity. Clin Orthop Relat Res. 2013;471(2):621–7.

3. Fabricant PD, Borst EW, Green SA, Marx RG, Fragomen AT, Rozbruch SR. Validation of a modified Scoliosis Research Society instrument for patients with limb deformity: the limb deformity-Scoliosis Research Society (LD-SRS) score. J Limb Lengthen Reconstr. 2016;2(2):86–97.

Further Reading

1. Babu LV, Evans O, Sankar A, Davies AG, Jones S, Fernandes JA. Epiphysiodesis for limb length discrepancy: a comparison of two methods. Strategies Trauma Limb Reconstr. 2014;9(1):1–3.
2. Blane CE, Herzenberg JE, DiPietro MA. Radiographic imaging for Ilizarov limb lengthening in children. Pediatr Radiol. 1991;21(2):117–20.
3. Dutton DB. Worse than the disease: Pitfalls of medical progress. New York, NY: Cambridge University Press; 1988.
4. Foote SB. Managing the Medical Arms Race. Berkeley, CA: University of California Press; 1992. p. 228.
5. Freund DA, Dittus RS, Fitzgerald J, Heck D. Assessing and improving outcomes: total knee replacement. Health Serv Res. 1990;25(5):723–6.
6. Freund DA, Katz BP, Callahan CM. Patient outcomes research teams: examples from a study of knee replacement. Ann N Y Acad Sci. 1993;703:86–93.
7. Ghamen I, Karam JA, Widmann RF. Surgical epiphysiodesis indications and techniques: update. Curr Opin Pediatr. 2011;23:53–9.
8. Ilharreborde B, Gaumetou E, Souchet P, Fitoussi F, Presedo A, Pennecot GF, et al. Efficacy and late complications of percutaneous epiphysiodesis with transphyseal screws. J Bone Joint Surg Br. 2012;94(2):270–5.
9. Lykissas MG, Jain VV, Manickam V, Nathan S, Eismann EA, McCarthy JJ. Guided growth for the treatment of limb length discrepancy: a comparative study of the three most commonly used surgical techniques. J Pediatr Orthop B. 2013;22(4):311–7.
10. Sass H-M. Moral aspects of risk and innovation. Artif Organs. 1997;21(11):1217–21.
11. Schroerlucke S, Bertrand S, Clapp J, Bundy J, Gregg FO. Failure of Orthofix eight-Plate for the treatment of Blount disease. J Pediatr Orthop. 2009;29(1):57–60.
12. Stewart D, Cheema A, Szalay EA. Dual 8-plate technique is not as effective as ablation for epiphysiodesis about the knee. J Pediatr Orthop. 2013;33(8):843–6.
13. Wiemann JM, Tryon C, Szalay EA. Physeal stapling versus 8-plate hemiepiphysiodesis for guided correction of angular deformity about the knee. J Pediatr Orthop. 2009;29(5):481–5.

The Future of Intramedullary Limb Lengthening

10

Once a reliable mechanism for intramedullary lengthening nails was developed, surgeons immediately began to conceive of other ways to eliminate external fixation—with its pain, inconvenience, and pin site infections—with the same ingenious technology. The pattern of innovation resembles the evolution of total knee, shoulder, elbow, and ankle arthroplasties after Sir John Charnley's monumental development of total hip replacement.

Device Modifications

Lengthening Plates

One limitation of intramedullary lengthening nails, when used for pediatric patients, concerns potential damage to the growth plate by a device that crossed such a sensitive structure. This has led surgeons to hope that someday lengthening plates will be forthcoming. Development in this area is likely to be slow because of two issues: concern about cantilever bending by an off-axis implant and the thickness of such a device in a child's juxtacortical soft tissues.

The junior author, Mark Dahl, has used the PRECICE® nail in a far off-label application as an internal lengthening implant (Fig. 10.1).

Trauma Nail

An individual submitting to elective limb lengthening understands in advance the requirements for limited weight bearing while the distraction regenerate matures. Traumatically injured patients, perfectly healthy and functional before their accident, are typically far less cooperative in this regard. Nevertheless, they too will someday soon benefit from motorized intramedullary nails, used as either a

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_10

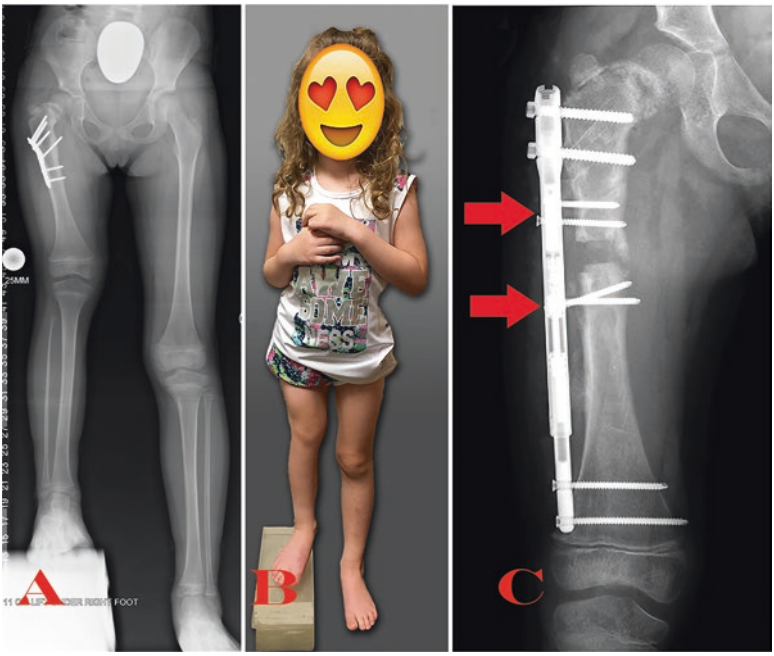


Fig. 10.1 Intramedullary lengthening nail as extra-osseous lengthener in pediatric patient. (**a**) Standing AP x-ray image of patient after valgizing osteotomy of hip in proximal femoral focal deficiency. (**b**) Clinical photograph. (**c**) Off-label use of lengthening nail as internal distractor. *Red arrows* point to cortical screws that surround nail to secure it in place. The proximal and distal locking screws have been tapped into the implant for added stability

distraction device (for post-trauma shortening and bone loss) or in a compression mode (for interfragmentary compression of fresh fractures and non-unions).

The illustrated example consists of a comminuted humeral shaft fracture treated by Dr. J. Tracy Watson of St. Louis. After inserting a pre-lengthened PRECICE® intramedullary nail, he used the External Remote Controller to compress the fracture site until the fracture gap is eliminated. Thereafter, Dr. Watson reassesses the x-ray image and adds additional compression to close residual fracture gap, if present. Once callus begins to appear, Dr. Watson compresses the implant 0.3 mm every 3 weeks to maintain preload and tension in the construct (similar to re-tensioning wires in an Ilizarov circular external fixator) (Figs. 10.2 and 10.3).

High Tibial Osteotomy

Osteoarthrosis of the knee is an increasingly common problem in every society where aging athletes—professional and amateur—continue to participate in sports long past their prime. Likewise, obesity, previous arthroscopic knee surgery, occupational and avocational injuries, and natural propensities all contribute to the epidemic

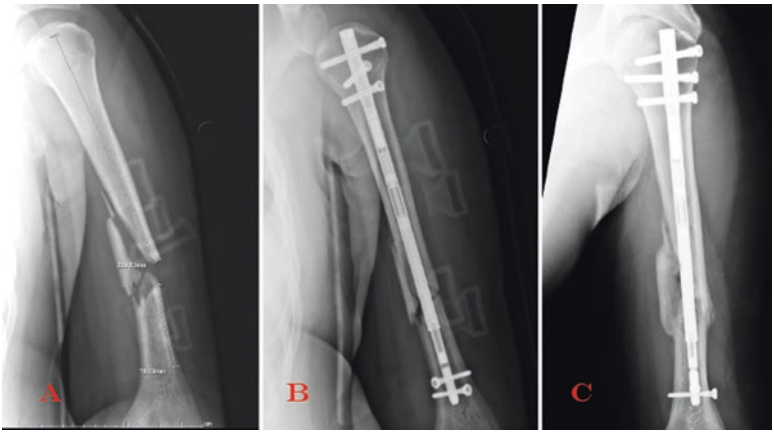


Fig. 10.2 Comminuted humerus fracture treated with PRECICE® intramedullary nail. (**a**) Initial condition. (**b**) After nail insertion and interfragmentary compression via External Remote Controller. (**c**) Union at 4 months

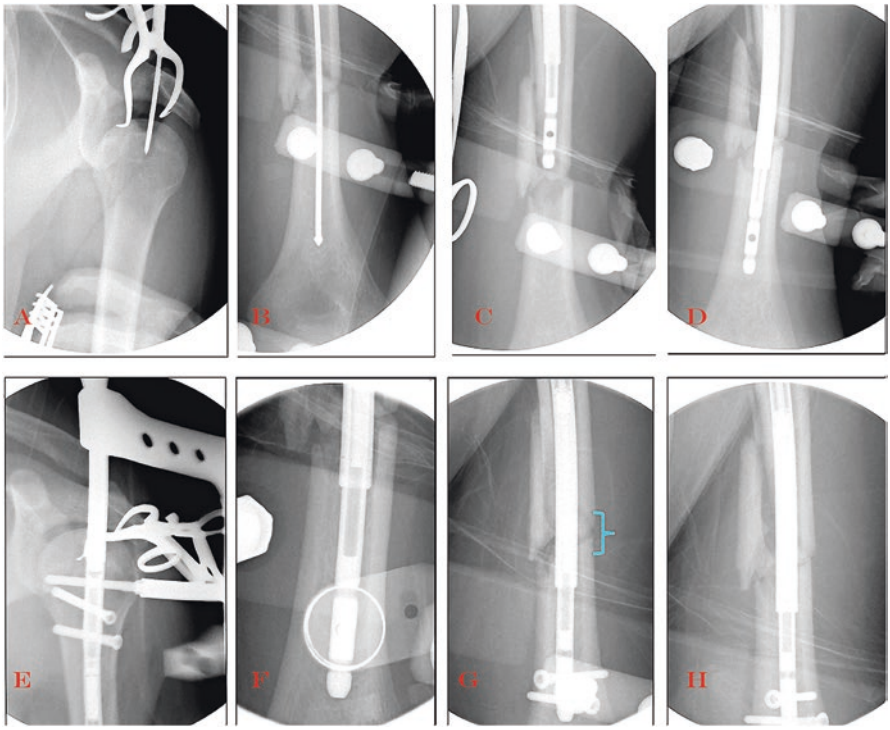


Fig. 10.3 Technique details of Fig. 10.2: (**a**) entry point. (**b**) Guide wire in distal fragment. (**c**) Nail insertion at fracture site. (**d**) Insertion completed. Small diameter nail fits well in distal fragment. (**e**) Proximal locking with guide. (**f**) Distal locking with alignment circle. (**g**) Nail fully locked, but gap persists at fracture site (*blue bracket*). (**h**) Gap compressed with External Remote Controller

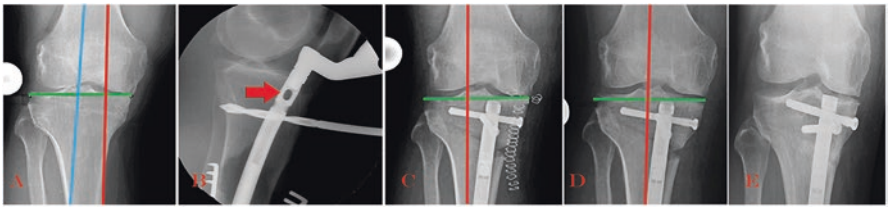


Fig. 10.4 High tibial osteotomy with specialized intramedullary lengthening nail. (**a**) Initial condition, comparing actual weight-bearing line (*red*) to ideal weight-bearing line (*blue*). (**b**) Image during nail insertion. Note oval proximal hole that allows proximal tibial to angulate around the implant (*red arrow*). (**c**) After distraction, slight overcorrection (*red line*). (**d**) Nail compressed slightly to eliminate overcorrection. (**e**) Result (Courtesy Matt Dawson, FRSC (Tr & O), ESSKA Osteotomy Committee)

of gonarthrosis. Total knee replacement (or hemiarthroplasty), while remarkably effective in dealing with the problem, is better suited for older individuals.

For this reason, the high tibial osteotomy has gained popularity as a temporizing procedure, gaining up to 10 years of additional knee function before knee replacement surgery is required. In the past, opening and closing wedge osteotomies have each had their proponents. Likewise, distraction osteogenesis has been employed in conjunction with external fixation to gradually effect opening wedge correction while creating new regenerate bone in the widening distraction zone.

With the advent of intramedullary lengthening, the prospect for a fully implantable alignment device especially designed by the junior author for high tibial osteotomy is on the near horizon, although, as of this writing, not yet approved for marketing by the FDA (Fig. 10.4).

Residual Limb Elongation

A short residual limb proves a serious challenge for any amputee. Prosthetic fitting, especially around the hip, may be impossible with a very short remaining femur. Such amputees are often restricted to ischial weight-bearing prostheses—with the associated limp—rather than a suction quadrilateral socket that is suspended from a longer stump.

Ilizarov developed a method of elongating residual limbs, but the process is painful because the external fixation frame must be suspended from the remaining bone, while a fragment of the bone's tip is pulled through the floppy soft tissues at the end of the stump.

Mark Dahl of Minneapolis employed (on a compassionate use basis) a very short Fitbone® intramedullary nail for residual limb lengthening (Fig. 10.5). The product, however, is not yet FDA cleared for marketing in the United States. Dahl therefore proposed a double telescoping nail for lengthening very short residual femurs, based on the PRECICE® nail technology. Called the Freedom® nail, the device has been cleared for marketing by the FDA and is now available for elongating short femoral residual limbs (Fig. 10.6).

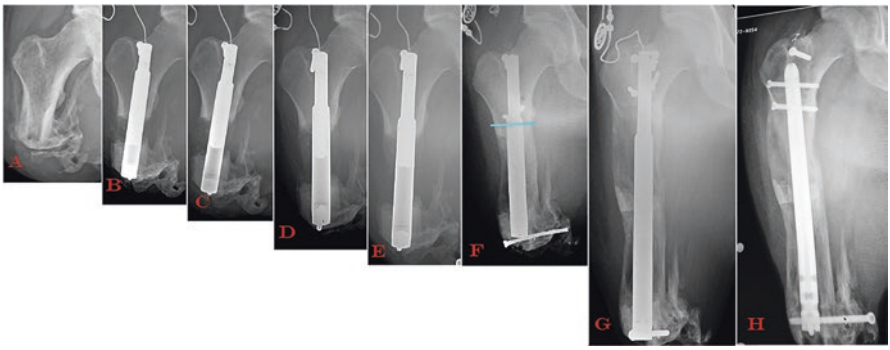


Fig. 10.5 Residual limb elongation with intramedullary lengthening nail. (**a**) Initial situation. (**b**–**e**) Progressive elongation with Fitbone® nail. (**f**) Insertion of longer Fitbone® nail. New osteotomy (*cyan line*). (**g**) At completion of second lengthening. (**h**) Trauma nail exchange to begin weight bearing with prosthesis

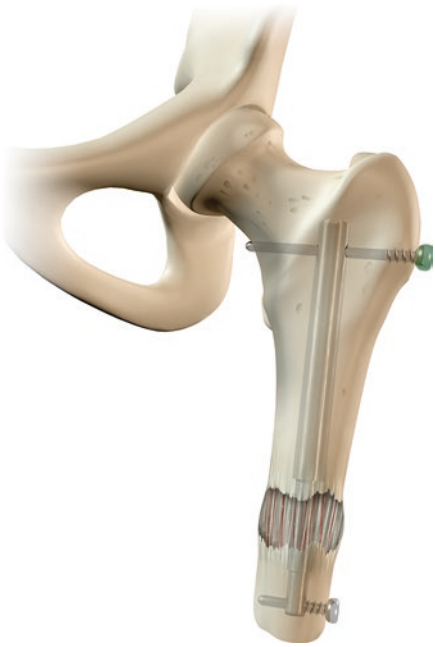


Fig. 10.6 The Freedom® nail, a double telescopic nail for elongating short residual femora. ©2016 NuVasive

Smart Remote Controllers

At the time of writing, the only commercially available externally controlled intramedullary lengthening nail in the United States was the PRECICE® nail, manufactured by NuVasive Specialized Orthopedics (formerly Ellipse Technologies) of Aliso Viejo, California. The External Remote Controller (ERC), which powers the intramedullary device, must be manually aligned directly over the rotating magnet

to work properly. The company is finalizing development of a new ERC that can detect the location of the nail's internal magnet as an aid to positioning the ERC. Likewise, by detecting north-south pole changes, the ERC's future sensor will record—and provide feedback—about the nail magnet's functioning.

More remarkably, the ERC's sensor will judge the nail magnet's resistance to rotation, thereby quantifying the progressive stiffness of the evolving regenerate bone in the widening distraction gap. Ultimately, this information will be conveyed to the treating surgeon wirelessly, thereby allowing a change in the lengthening or compression prescription from a distance.

Bone Transport

Eliminating a segmental bone defect has long been one of the most impressive features of Ilizarov's method. In essence, the Ilizarov method can enable lengthening bone on one or the other side of the defect to gradually close the space while regenerate bone forms in the distraction zone. Most Western surgeons insert a bone graft at the docking site (where the intercalary moving fragment contacts the opposite side of the defect, thereby reducing the incidence of docking site non-unions).

Biomedical engineers and orthopedic surgeons hope to achieve bone transport using intramedullary bone lengthening technology. In this way, the most distressing aspect of Ilizarov bone transport—long wire-cut skin scarring—would be eliminated (Fig. 10.7). Likewise, wire or pin site infections and their inevitable pain would disappear with such a device.

Logically, an intramedullary bone transport nail would securely affix to both ends of the deficient bone, with the motorized part of the implant moving the intercalary segment through the soft tissues. Unfortunately, the placement of such an intramedullary nail reams out the marrow blood supply of all fragments, a source of bone healing at the docking site. Engineers and clinicians are testing several proposals to use intramedullary nails to eliminate skeletal defects without resorting to external skeletal fixators. In 2014, for instance, Kold and Christensen, working in

Fig. 10.7 Wire tract created during bone transport. Many become infected during the process

Denmark, published a case report of bone transport using the Fitbone® system [1]. The authors employed a custom-made nail with a longitudinal slot to permit the intercalary bone segment to slide along the nail after it was secured to the implant with a transfixion screw. Four weeks after surgery, they injected recombinant BMP-7 into the docking site. Both the regenerate and docking sites matured appropriately, eliminating a 3 cm defect (Figs. 10.8 and 10.9).

An alternative strategy for eliminating a segment defect with an intramedullary lengthening nail was devised by Matthew Gardner of Springfield, Illinois. Rather than cross the defect with the implant, Gardner, for a tibial defect case still in progress as of this writing, spanned the gap with a subcutaneous plate and screws, being careful to place the screws in the proximal fragment posterior enough to preclude interference with subsequent nail insertion.

Thereafter, he used a PRECICE® tibial nail to push an intercalary segment of bone across the defect until docking was complete. He obtained excellent regenerate formation in the widening distraction gap, but had usual docking site union issues. An autogenous bone graft is maturing nicely at the contact point between the intercalary segment and the target segment (Fig. 10.10).

Fig. 10.8 Mal-non-union of the tibia in 53-year-old male, from a ski injury. Reprinted from [1]

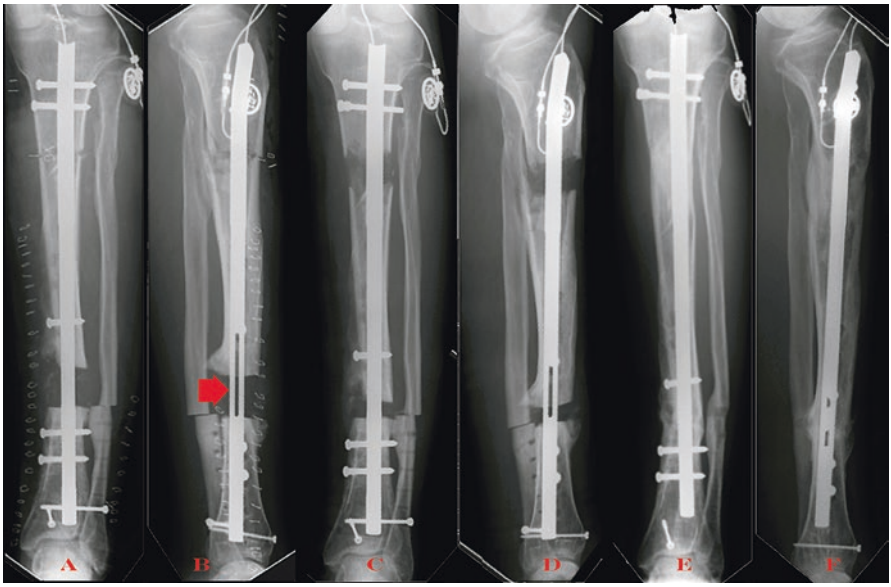


Fig. 10.9 Treatment of mal-non-union with Fitbone® custom-made slotted transport nail. (**a**) AP view after nail insertion. (**b**) Lateral view after nail insertion. Red arrow points to slot in nail. (**c**) AP view during transport of intercalary segment. (**d**) Lateral view of intercalary segment. (**e**) AP view of result after docking and BMP-7 insertion. (**f**) Lateral view after docking and BMP-7 insertion (the Fitbone® nail has not been approved for sale in the United States) Reprinted from [1]

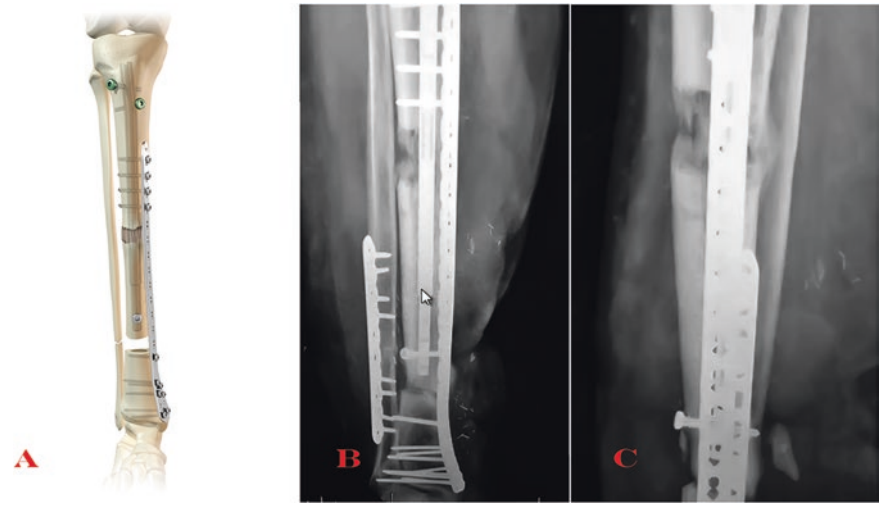


Fig. 10.10 Gardner technique of bone transport. (**a**) Schema of plan: spanning subcutaneous plate and intramedullary nail. (**b**) AP view at docking time. (**c**) Close-up lateral of regenerate showing good vertical striations of newly formed bone

Bifocal Treatment

One of the distinct advantages of the Ilizarov method with circular fixators is the ability to solve two problems simultaneously in the same bone. Ilizarov referred to such protocols as "bifocal." When originally introduced, intramedullary lengthening nails were viewed as simple bone elongating devices, with the potential for slight deformity correction at the site of osteotomy for lengthening. Once surgeons became comfortable with the reliability of externally controlled lengthening nails, they started to devise strategies to eliminate external fixators for applications requiring surgery at both ends of the bone, either simultaneously or sequentially (Figs. 10.11 and 10.12).

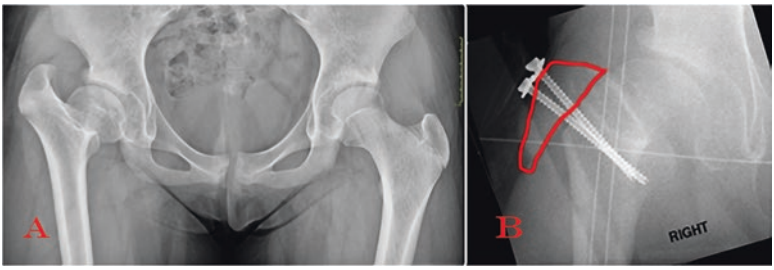


Fig. 10.11 (**a**) Perthes disease with overgrown greater trochanter and femoral shortening. (**b**) Transfer of trochanter after partial resection

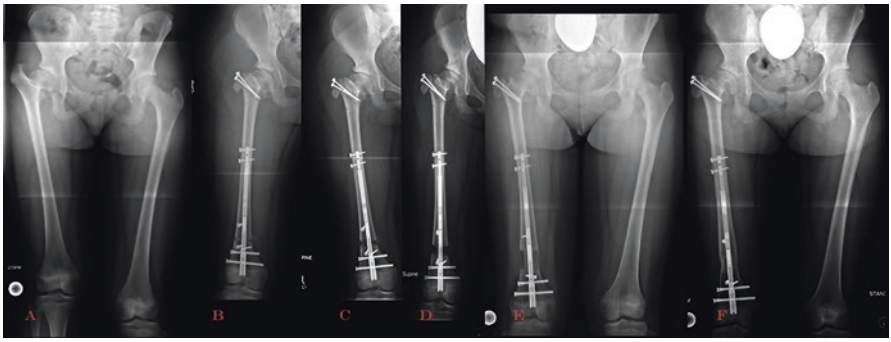


Fig. 10.12 Bilocal treatment of Perthes disease with greater trochanter overgrowth. (**a**) Initial situation. (**b**) Simultaneous transposition of greater trochanter and intramedullary lengthening. (**c–e**) Progressive lengthening. (**f**) Result

Expanded Indications

Cosmetic Stature Surgery

As soon as limb lengthening became predictably successful, certain individuals, self-conscious about their height, began to request stature surgery by Professor Ilizarov. He relates the story of a woman who wanted to join Moscow's legendary Bolshoi Ballet Company, but was 1.0 cm shorter than the minimum height required to dance in the *Corps de Ballet*. But for that centimeter, she assured Ilizarov, she would certainly be selected. So Ilizarov lengthened her tibiae 1.0 cm.

Although the woman now could present herself as the proper height for a ballerina in a major Russian dance company, she failed her audition, having lost just enough flexibility in her lower extremities to handicap her dancing.

A recent report from Ilizarov's facility [2] offers an honest assessment of the problems associated with external fixator cosmetic lengthening in 138 "somatically normal" patients treated in Kurgan between 1983 and 2006. (Professor Ilizarov died in 1991.) One hundred thirty-one were available for at least one-year follow-up. In spite of optimum care in the most experienced hands available in the world, 48 patients (37%) had 59 complications: 37 were soft tissue related and 22 were bone related. Sixteen patients needed unplanned returns to the operating room for surgery. At the end of treatment, however, 130 patients felt subjectively satisfied.

Other reports, reviews, and meta-analyses in the literature of stature surgery during the external fixation era report a similar constellation of problems and complications.

In the past, the potential complications and pain associated with the use of external fixation for limb lengthening dissuaded most people concerned about their height from ever considering stature surgery. Now, however, intramedullary lengthening nails have shifted the balance scales in the eyes of some surgeons, and many patients.

As any reader of this book must by now realize, changing the mechanics of limb elongation from external fixators to lengthening nails has not eliminated many of the serious complications of limb lengthening.

Paley, for instance, was among the first to use the PRECICE® intramedullary lengthening nail for cosmetic stature surgery. In 2015, in a review of the market potential of the device [3], Paley reported on 15 patients who underwent stature surgery for either achondroplasia or cosmetic reasons. The mean length gain was 5.64 cm in this group of patients (average age, 29.7 years). Eight of the 15 stopped lengthening before reaching their goals for "personal" rather than "medical" reasons. Three patients ("all congenital") required bone grafting. Three nails broke and had to be replaced. Seven nails in six patients stopped lengthening during distraction, two due to operator error. In five other nails, the internal mechanism gave way, perhaps due to stiffening regenerate and/or large muscular thighs. In all, there were 18 unplanned surgeries in 16 patients.

In many of Paley's patients, the first-generation PRECICE® nail was inserted. It had several welds that proved weak spots when the nail was stressed, leading to implant breakage. The second-generation nail, the PRECICE 2 (P2), has not had

this problem. Likewise, the company has incrementally improved the internal mechanism, so motor failures are now quite rare.

Few topics elicit as much emotion among limb lengthening surgeons than the matter of cosmetic stature surgery. Both authors of this monograph have concluded that the risk/benefit ratio is sufficiently unfavorable to preclude performing such operations unless the surgeon has considerable experience lengthening congenitally or traumatically shortened limbs. After all, adding five to eight centimeters to someone's height is fraught with greater risks than placing a pair of silicone-filled bags into a woman's chest or chiseling off a couple of millimeters from a teenager's nose.

So why would any orthopedic surgeon consider performing vanity stature surgery? The question has been the subject of intense debate at the several gatherings of limb lengthening surgeons recently. The matter of surgical ethics and responsibility always comes up.

Although surgeons with extensive limb lengthening experience are anecdotally reporting that their patients who have come through vanity stature procedures unscathed by serious complications are happy that they chose cosmetic lengthening surgery, the pages of this monograph offer adaquate warning that such operations should probably not be performed by surgeons lacking significant limb lengthening experience.

Why? Because a serious complication, one that deprives a patient of life or limb function, will prove devastating for both the patient and the surgeon.

Conclusion

In the 1970s, a therapeutic paradigm shift occurred when external skeletal fixation reentered the surgical armamentarium after having all but disappeared from use after Second World War. The senior author's *Complications of External Skeletal Fixation: Causes, Prevention and Treatment* [4] focused attention on technical details necessary to assure successful treatment of recalcitrant non-unions, malunions, and post-trauma osteomyelitis with external fixation. Subsequent adaptation and improvement of fixators have been a remarkable process to witness.

Starting in 1951 and extending to his 1991 death, G. A. Ilizarov's astounding discoveries about bone's capacity to form new osseous tissue in a widening distraction gap created one of the most transformative paradigm shifts in the history of deformity correction and limb lengthening. Orthopedic surgery and maxillofacial surgery have been changed forever.

Children with crippling birth defects, who previously would have been subjected to amputation of deformed body parts, can now experience limb alignment and elongation procedures to an extent never before thought possible. Patients with post-trauma non-unions and malunions could now enjoy full restoration—with equal limb lengths.

Exactly 30 years ago, to the day, of this writing, the senior author flew to Kurgan, Siberia, Soviet Union, and landed in a place where human bones appeared to be made of wax—to be bent, twisted, and pulled on without apparent limits—to cure all manner of maladies. Green subsequently assisted Professor Ilizarov present his

work to Western surgeons with publications in Clinical Orthopaedics and Related Research [5, 6] and in his 1991 opus magnum, *Transosseous Osteosynthesis* [7].

Over the past three decades, Ilizarov's distraction osteogenesis techniques have been at the heart of increasingly common operative procedures. Nevertheless, pin and wire site sepsis continue to plague all external skeletal fixation devices, whether applied for dynamic corrections or static immobilization.

Now, we are witnessing yet another paradigm shift in orthopedic surgery as intramedullary lengthening devices, elongated by mechanical, electrical, or magnetic means, supplant external fixators for reconstructive strategies requiring dynamic distraction and compression techniques to stimulate osseous healing and new bone growth.

With time, imaginative surgeons and knowledgeable engineers will create innovative intramedullary distraction and compression devices—accompanied by a new vocabulary of reconstructive terminology—to help patients overcome the adverse consequences of genetics, development, or trauma without spending months or years in external fixators.

The senior author ended his 1981 book *Complications of External Skeletal Fixation* as follows:

> It is obvious from the preceding chapters that complications continue to haunt external fixation in spite of the superbly designed frames and components currently available. For these reasons, fixators should be reserved for those situations where other treatment modalities are likely to fail.

New intramedullary distraction and compression devices, including those currently available, are already supplanting external fixation as a prime orthopedic reconstruction modality—a process that will continue into the foreseeable future.

References

1. Kold S, Christensen KS. Bone transport of the tibia with a motorized intramedullary lengthening nail—a case report. Acta Orthop. 2014;85(2):211–3.
2. Novikov KI, Subramanyam KN, Muradisinov SO, Novikova OS, Kolesnikova ES. Cosmetic lower limb lengthening by Ilizarov apparatus: what are the risks? Clin Orthop Relat Res. 2014;472(11):3549–56.
3. Paley D. PRECICE intramedullary limb lengthening system. Expert Rev Med Dev. 2015;12(3):231–49.
4. Green SA. Complications of external skeletal fixation: causes, prevention and treatment. Springfield, IL: Charles C. Thomas; 1981.
5. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: Part I. The influence of stability of fixation and soft-tissue preservation. Clin Orthop Relat Res. 1989;238:249.
6. Ilizarov GA. The tension-stress effect on the genesis and growth of tissues: Part II. The influence of the rate and frequency of distraction. Clin Orthop Relat Res. 1989;239:263.
7. Ilizarov GA. Transosseous osteosynthesis. Heidelberg: Springer-Verlag; 1991.

Further Reading

1. Catagni MA, Lovisetti L, Guerreschi F, Combi A, Ottaviani G. Cosmetic bilateral leg lengthening: experience of 54 cases. J Bone Joint Surg Br. 2005;87(10):1402–5.
2. Kim SJ, Pierce W, Sabharwal S. The etiology of short stature affects the clinical outcome of lower limb lengthening using external fixation. A systematic review of 18 trials involving 547 patients. Acta Orthop. 2014;85(2):181–6.
3. Koczewski P, Shadi M, Napiontek M, Dorman T, Faflik J, Grzegorzewski A, et al. Short stature treatment by lower limb lengthening–multicenter study from five centers. Chir Narzadow Ruchu Ortop Pol. 2002;67(2):197–206.
4. Koczewski P, Shadi M. Surgical treatment of short stature of different etiology by the Ilizarov method. Pediatr Endocrinol Diabetes Metab. 2007;13(3):143–6.

Cross-Section Atlas

11

Development of the Atlas

Background

In 1981, the senior author's book *Complications of External Skeletal Fixation: Causes, Prevention and Treatment* [1] contained a cross-section atlas to help surgeons avoid injuring nerves, arteries, and veins while inserting transcutaneous pins during external skeletal fixation. Originally, the atlas was designed to aid in the application of Hoffmann-type full- and half-pin fixators, as well as those frames that used similar mounting strategies. Needless to say, the book subsequently proved useful for the application of the Ilizarov external fixation method, which typically employed even more transcutaneous implants (specifically, tensioned wires) than did a complex quadrilateral multi-pin Hoffmann frame in the Vidal configuration [2].

Indeed, the senior author was not surprised when, while attending the first English-language course on the Ilizarov method in Kurgan, Siberia, USSR (1988), cross-section diagrams were affixed to the walls of the workshop where frame application principles were taught. Moreover, the concept of utilizing cross-section anatomy for safe transcutaneous implant insertion is hardly new. During the Second World War, Kreuz and Shaar produced a manual of fracture treatment for use by medics in the US Navy [3] that contained cross-section diagrams derived from a classic 1911 atlas of cross-section anatomy by Eycleshymer and Schoemaker [4].

Although *Complications of External Skeletal Fixation* is out of print after three successful press runs (copies being sold on rare medical book websites) because the publisher stopped selling medical books, the pin and wire atlas lives on in recent editions of Browner's *Skeletal Trauma* [5].

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4_11

Derivation of the Atlas

In 1911, Eycleshymer and Schoemaker created a cross-section atlas of human anatomy by freezing 11 human cadavers solid and band-sawing the bodies into 1-inch thick slices. They then mapped out the position of every structure on a piece of tracing paper preprinted with grid marks. They next averaged the size and position of each structure and created a composite transverse section drawing of these averaged elements, the unique feature of the atlas. A talented medical illustrator, Michele Predesik, used the Eycleshymer and Schoemaker images to create 3D-appearing slices in the senior author's external fixation monograph. These were recently updated by the senior author for inclusion in the present monograph.

The Need for a Cross-Section Atlas for Intramedullary Lengthening

One could reasonably assume that intramedullary limb lengthening has eliminated the need for a cross-section atlas because, after all, the main implant is an intramedullary nail, with well-defined entry portals developed to avoid injury to neurovascular structures. Such, however, is not the case: both the osteotomy and the insertion of transverse locking screws and pillar blocking screws are percutaneous procedures, with risks similar to, if not identical to, external fixation pins (Figs. 11.1, through 11.16).

Plunge Depth

Alajmo and coauthors [6] studied the depth a drill bit plunges after traversing the marrow canal and the far cortex of a long bone. They considered three variables: bone density, drill bit sharpness, and surgeon's experience. The research was conducted at an international trauma course. The authors found that a sharp drill bit plunges less than a dull one, that a dense bone causes more plunging than osteoporotic bone, and that an experienced surgeon plunges less than an inexperienced one. The amount of plunge ranges from 5.1 mm when an experienced surgeon drills through osteoporotic bone with a sharp drill bit to 22 mm when an inexperienced pushes a dull drill bit through dense bone. The atlas on the following pages takes this observation into account.

The Atlas

Principles of the Atlas

In modifying the senior author's 1981 cross-section atlas for intramedullary lengthening, structures are indicated with *red arrows* if they are at risk of injury with either drill bits, transverse locking screws, pillar blocking screws, or a percutaneous osteotomy. Other structures are indicated with *green arrow*.

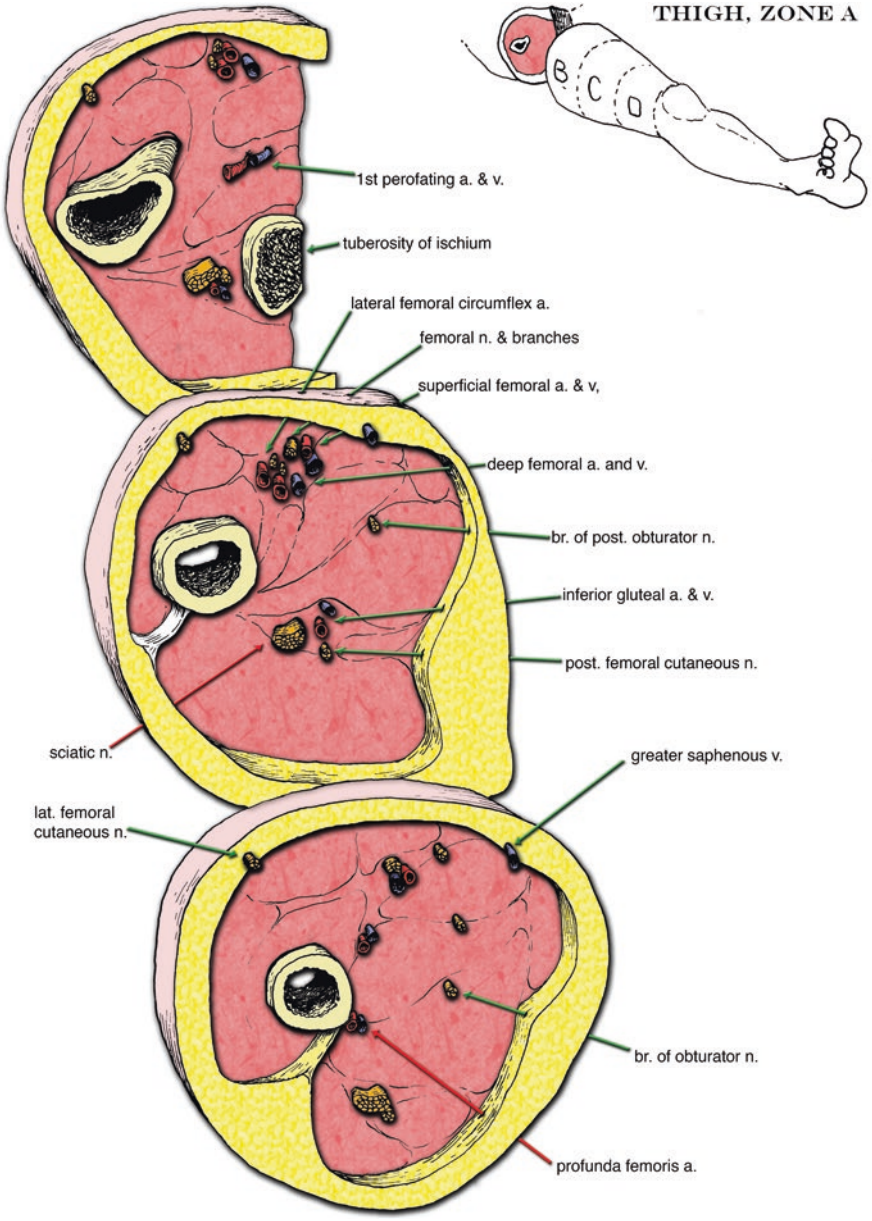


Fig. 11.1 The sciatic nerve is posteromedial to the femur throughout Zone A, more than a bone diameter away. The deep femoral artery comes to lie medial to the femur in the lower end of Zone A, separated from it by the origin of the vastus medialis muscle, but only one half bone width away

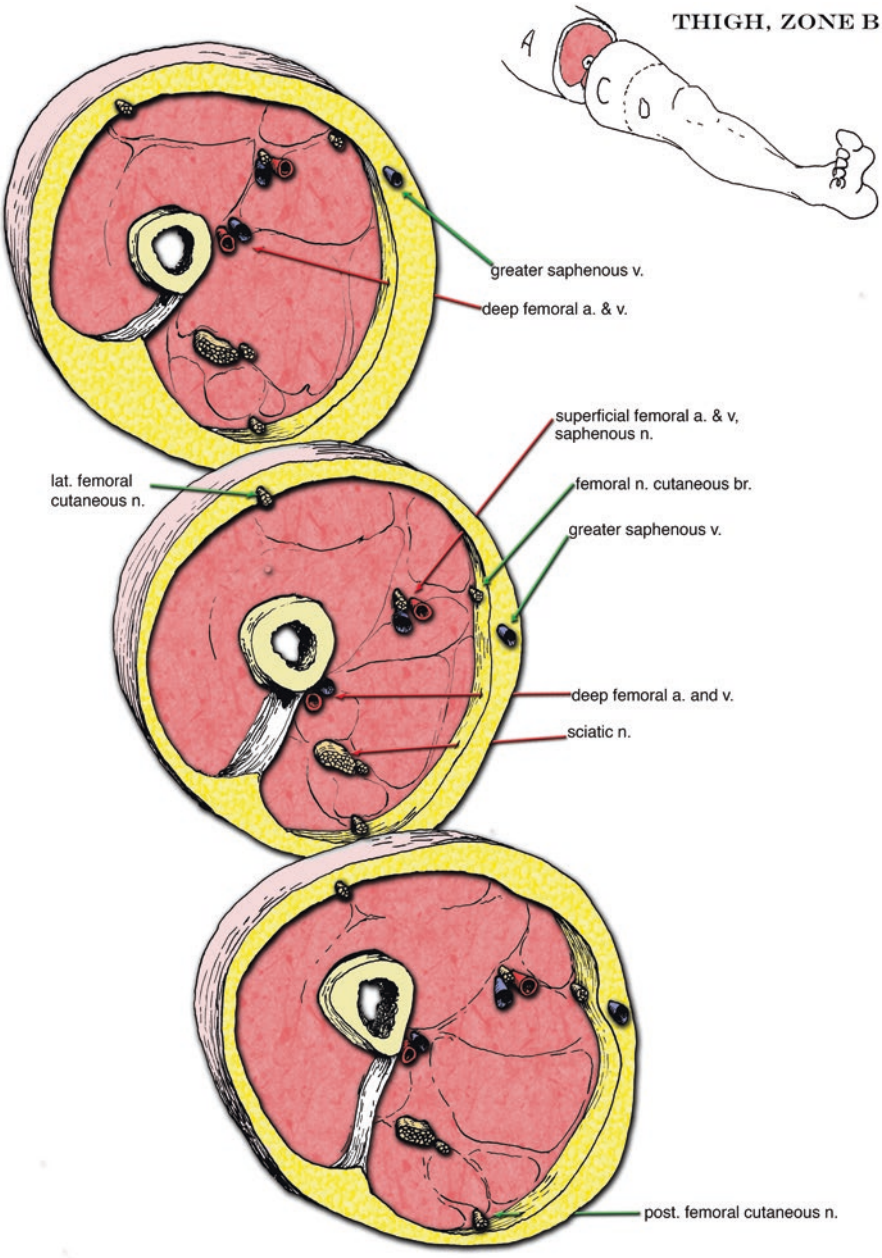


Fig. 11.2 The sciatic nerve is posteromedial to the femur, separated by one bone diameter. The superficial femoral artery crosses the coronal plane of the femur between Zone B and Zone C. The deep femoral artery and vein are medial to the femur in proximal Zone B and posterior to the femur in distal Zone B. Caution is necessary in proximal Zone B, because the superficial and deep femoral vessels are in a straight line and can both be injured with a plunge

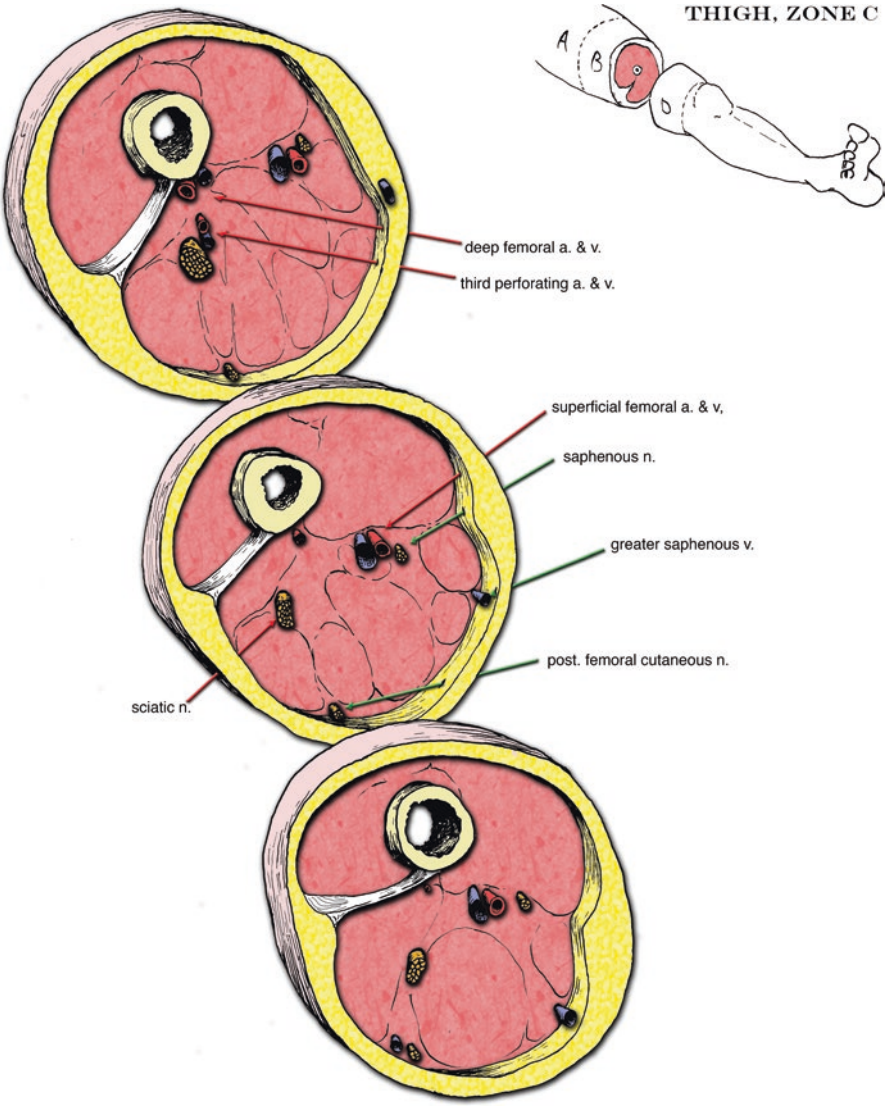


Fig. 11.3 The sciatic nerve passes from medial to lateral behind the femur, approximately one bone width away. The superficial femoral artery passes the coronal plane of the femur in Zone C and is posterior to the bone at the lower end of this zone. The deep femoral artery and vein are adjacent to the posterior surface of the femur but terminate at the lower end of Zone C

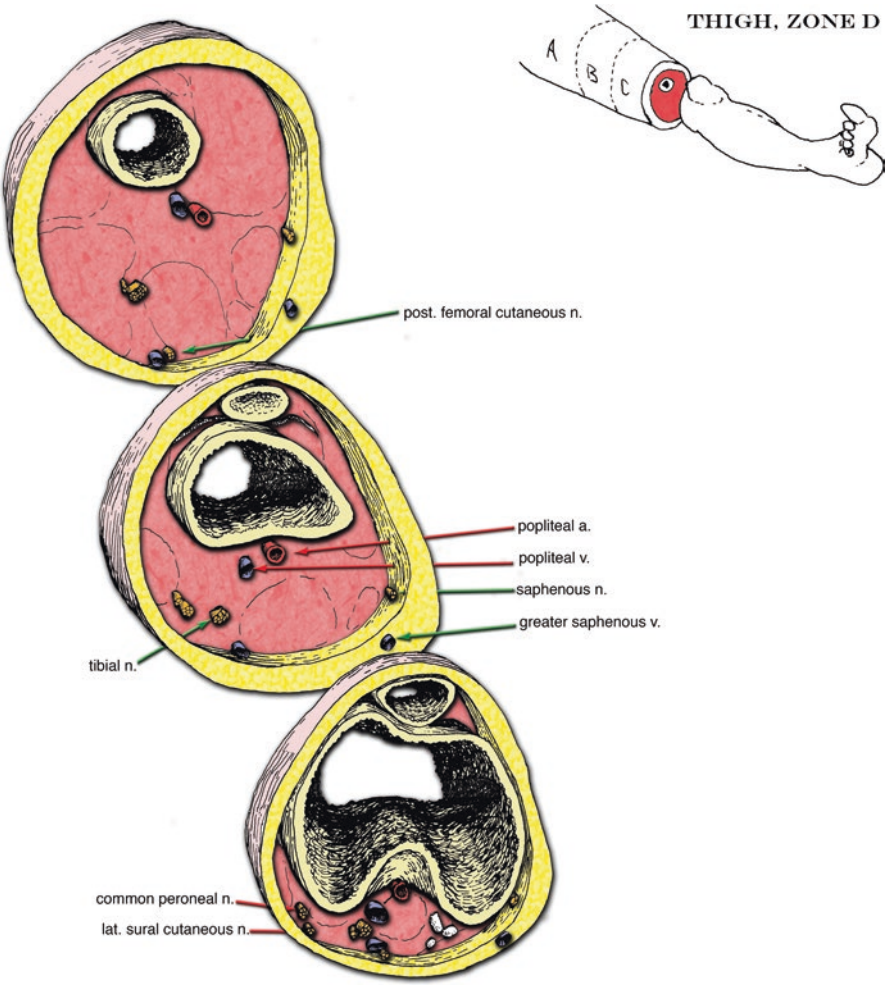


Fig. 11.4 The femoral artery becomes the popliteal artery and, with the popliteal vein, is immediately posterior to the femur in Zone D. The synovial cavity of the knee joint enlarges to encompass the anterior half of the femur immediately above the joint line

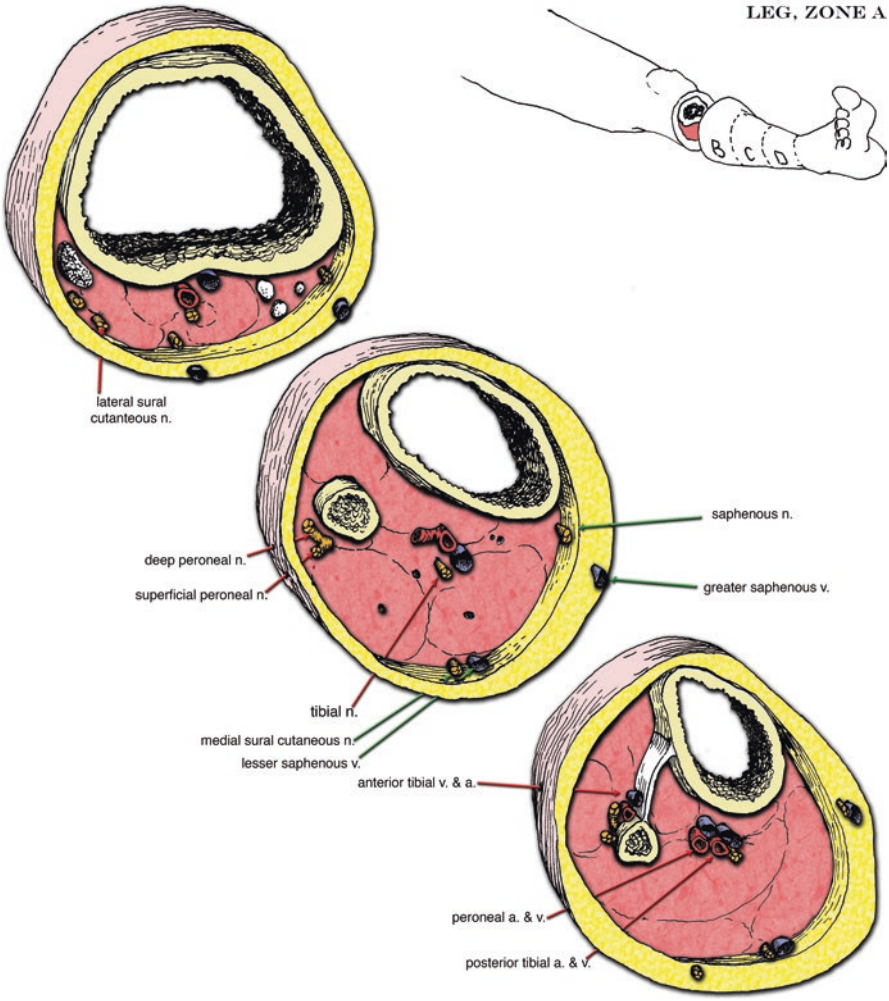


Fig. 11.5 The shape of the tibia changes rapidly through this zone. The popliteal artery is posterior to the tibia where it divides into its terminal branches. The superficial and deep peroneal nerves are lateral to the fibula as they wind around the fibular neck. The saphenous nerve and greater saphenous vein are posterior to the tibia on the medial side of the limb. In distal Zone A, the anterior tibial artery is on the anterior surface of the interosseous membrane, and the peroneal and posterior tibial arteries are posterior to the tibia, accompanied by their associated veins

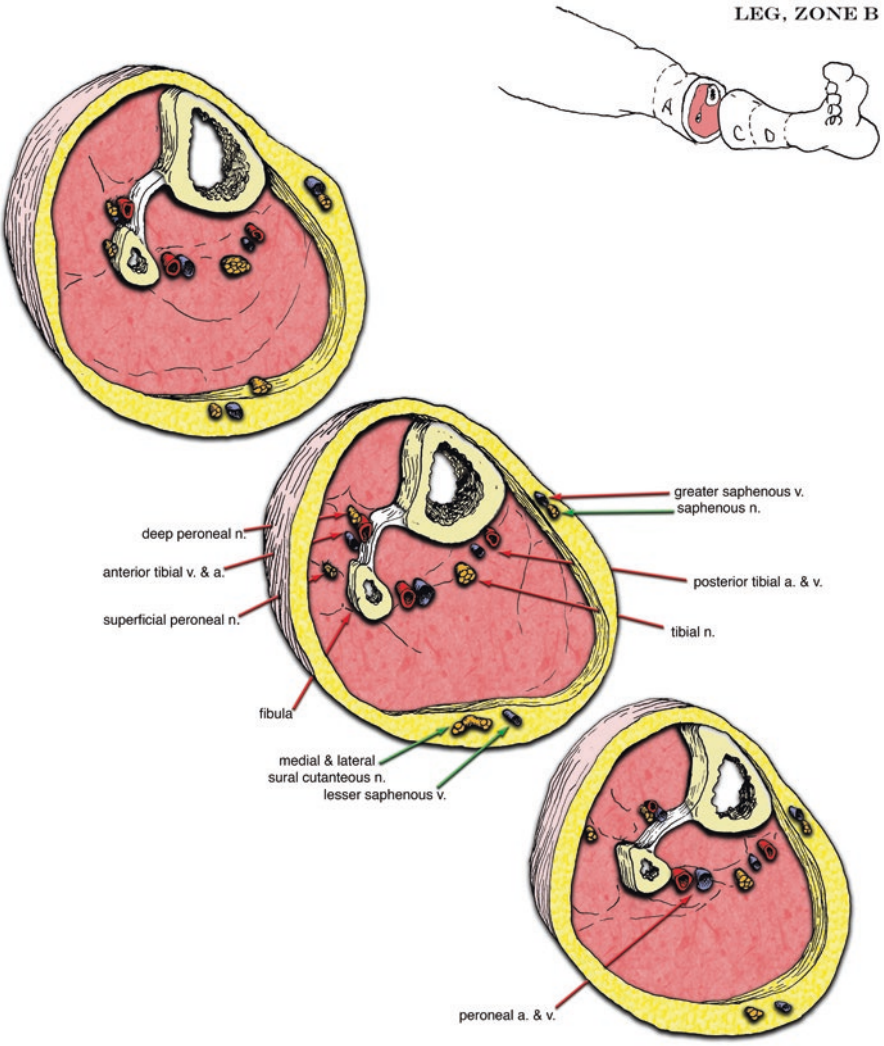


Fig. 11.6 The tibia has a triangular cross section throughout Zone B, with the lateral surface relatively vertical and the medial surface oblique. The posterior tibial vessels, the tibial nerve, and the peroneal vessels maintain a constant relationship throughout Zone B with respect to the posterior surface of the tibia and the medial surface of the fibula. The anterior tibial artery and vein, and the deep peroneal nerve, lie on the anterior surface of the interosseous membrane in Zone B, traversing from the anterior ridge of the fibula toward the lateral ridge of the tibia

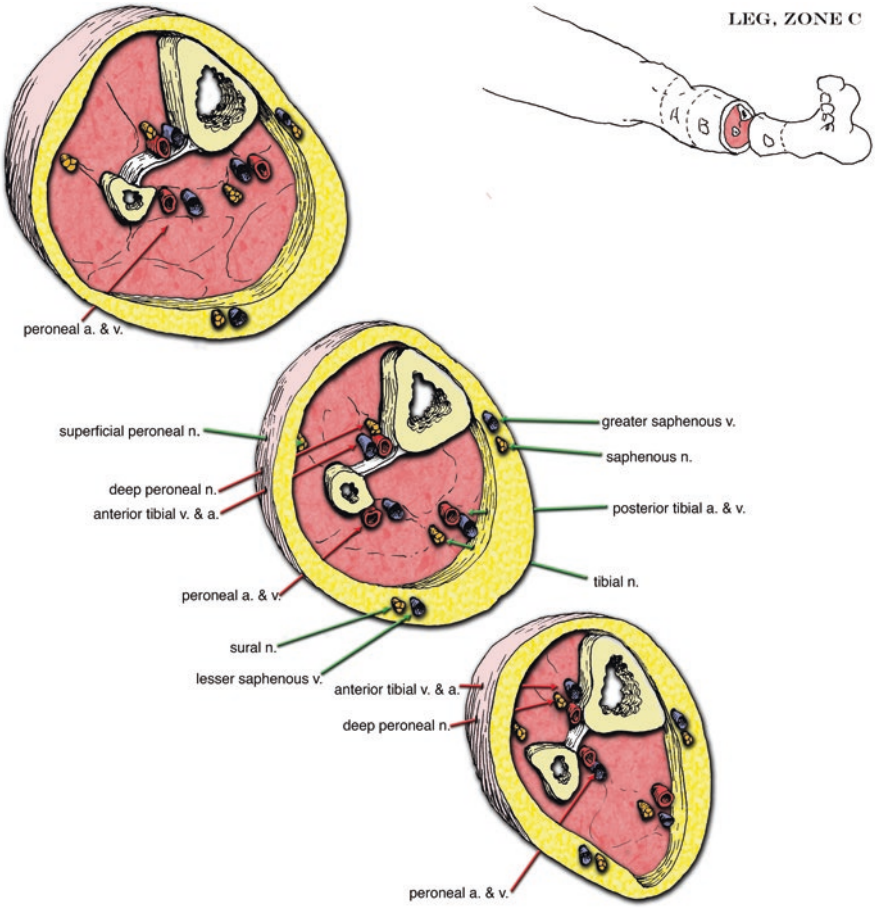


Fig. 11.7 The posterior tibial artery and vein and the tibial nerve remain posterior to the tibia, and the peroneal vessels remain slightly medial to the fibula. The anterior tibial artery and vein and the deep peroneal nerve have completed their traverse of interosseous membrane and are adjacent to the posterolateral corner of the tibia throughout Zone C. These structures begin to traverse the lateral surface of the tibia in distal Zone C

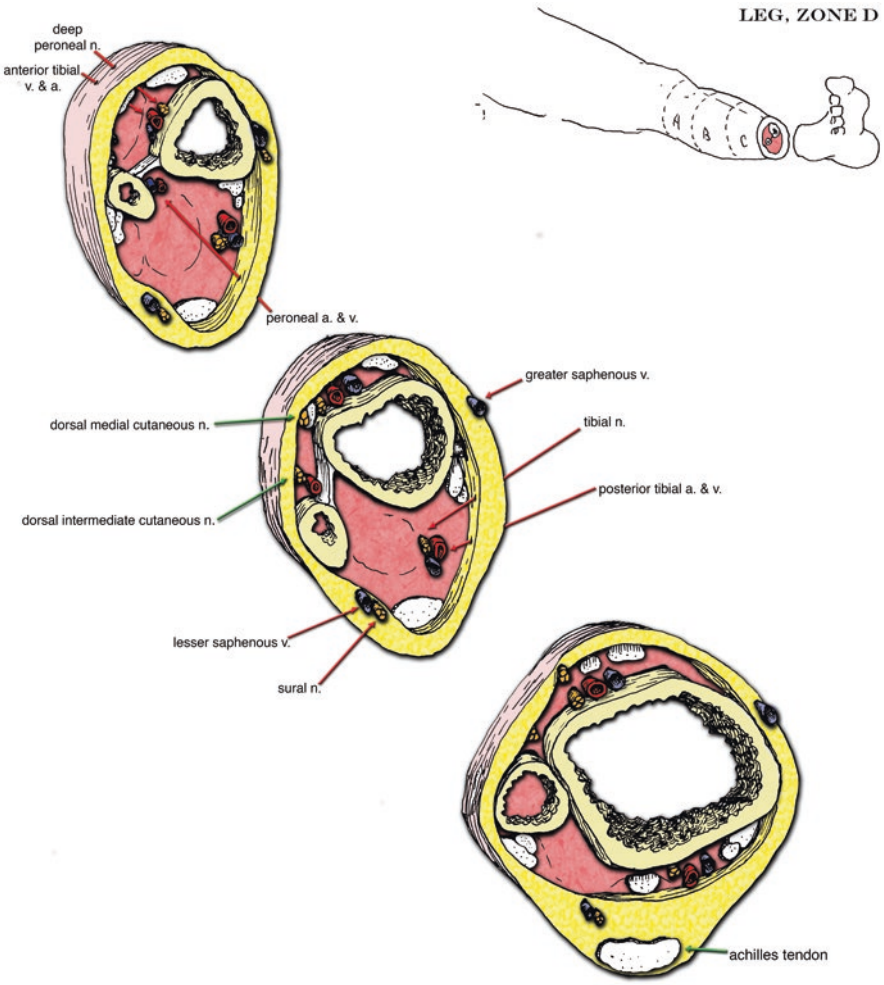


Fig. 11.8 The posterior tibial artery and vein and the tibial nerve remain posterior to the tibia, and the peroneal vessels remain slightly medial to the fibula. The anterior tibial artery and vein and the deep peroneal nerve have completed their traverse of interosseous membrane and are adjacent to the posterolateral corner of the tibia throughout Zone C. These structures begin to traverse the lateral surface of the tibia in distal Zone C. In leg Zone C, the peroneal artery and its two venae comitantes are along the deep surface of the fibula and can be injured during fibular osteotomy, especially if an oscillating saw is used all the way across the bone. In distal Zone C, transverse screws can endanger the anterior tibial artery and deep peroneal nerve on the lateral tibial surface

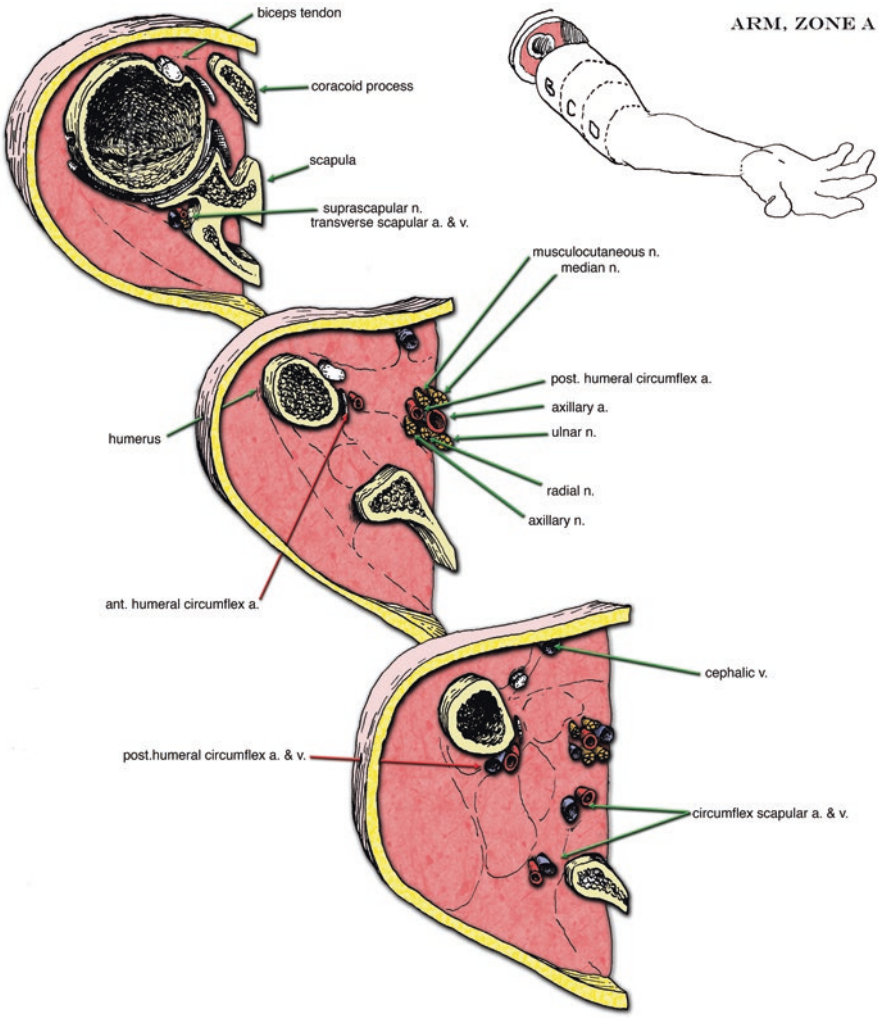


Fig. 11.9 The humeral head is largely intrasynovial, being surrounded by a joint cavity medially and posteriorly and by the subacromial bursa anteriorly. The main neurovascular bundle containing the brachial plexus is medial to the humerus, separated from it by a distance equal to the width of the bone. The anterior and posterior humeral circumflex vessels surround the upper humerus slightly below the surgical neck, accompanied by the axillary nerve

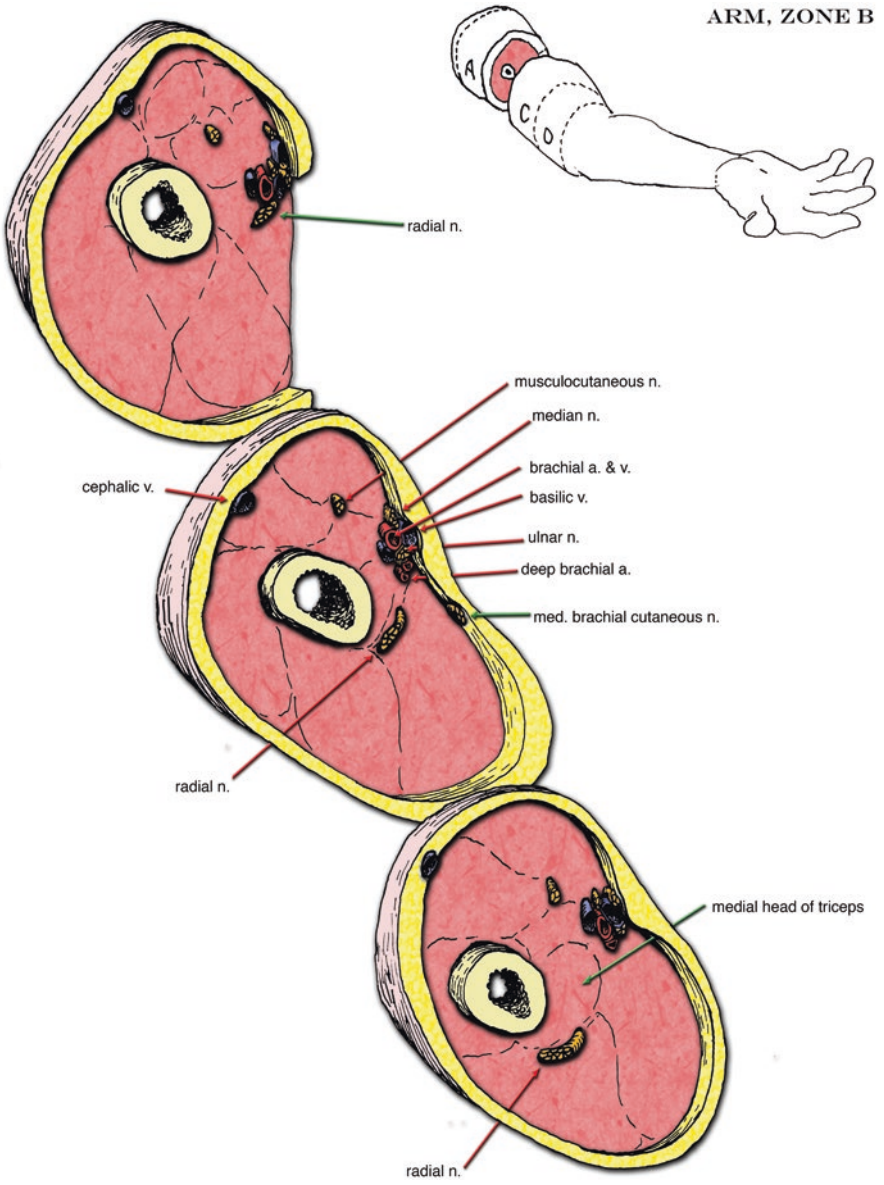


Fig. 11.10 The brachial artery and veins and the brachial plexus remain medial to the humerus in this zone. The radial nerve separates from the main neurovascular bundle and passes posterior to the humerus in Zone B, separated from the bone by the medial head of the triceps. The musculocutaneous nerve and cephalic vein are anterior to the humerus in Zone B

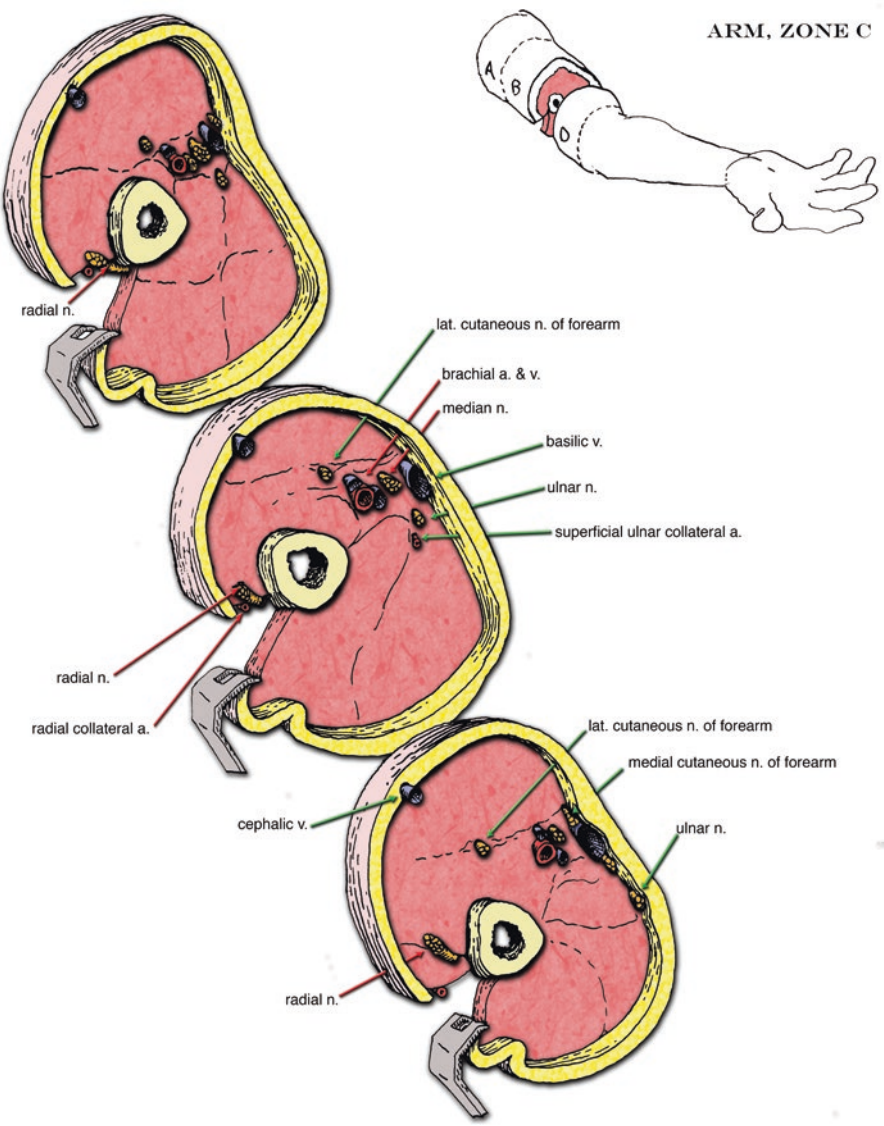


Fig. 11.11 The radial nerve winds around the lateral side of the shaft of the humerus in contact with the bone. The brachial artery, veins, and branches of the brachial plexus remain medial to the humeral shaft. The ulnar nerve separates from the main neurovascular bundle in this zone. The musculocutaneous nerve becomes the lateral cutaneous nerve of the forearm and remains anterior to the humerus

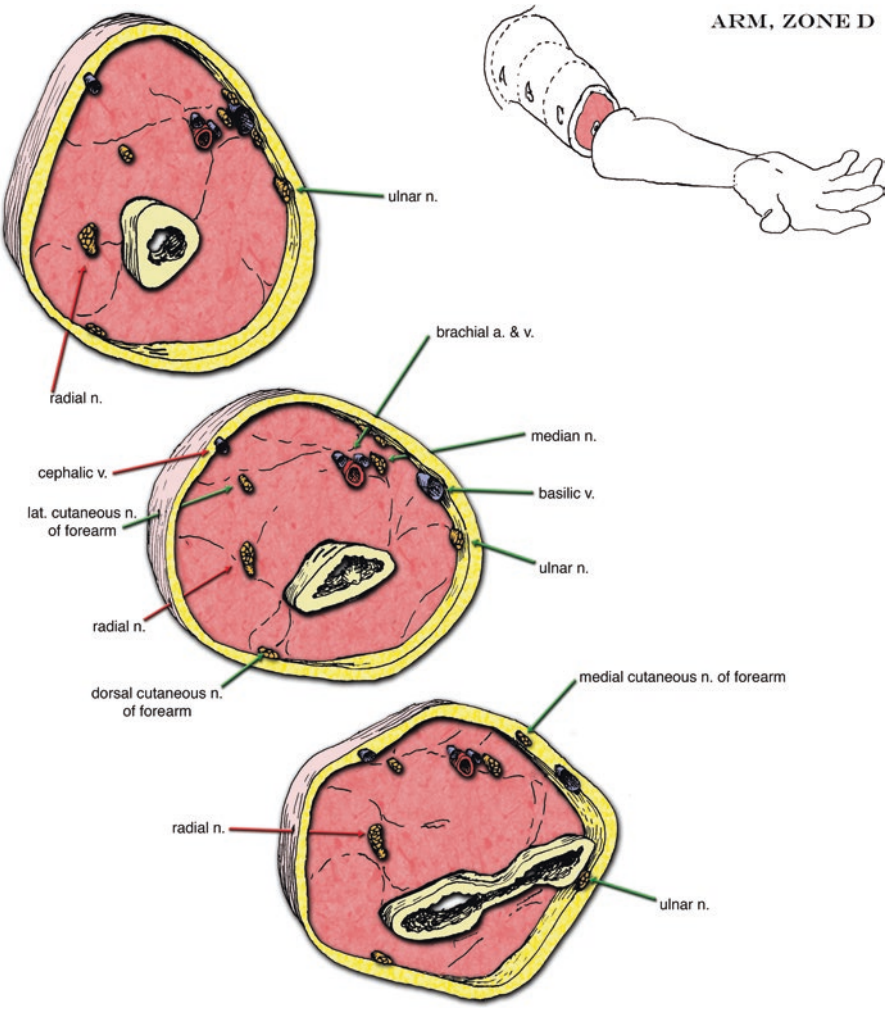


Fig. 11.12 The distal humerus flattens and is rotated with the lateral epicondyle 30° posterior to the medial epicondyle. The radial nerve lies on the lateral side of the radius in proximal Zone D but is anterior to it in the distal portion of the zone. The median nerve remains anterior and medial to the bone throughout this zone. The ulnar nerve passes posterior to the plane of the distal humerus and lies in contact with the posteromedial corner of the bone immediately above the elbow

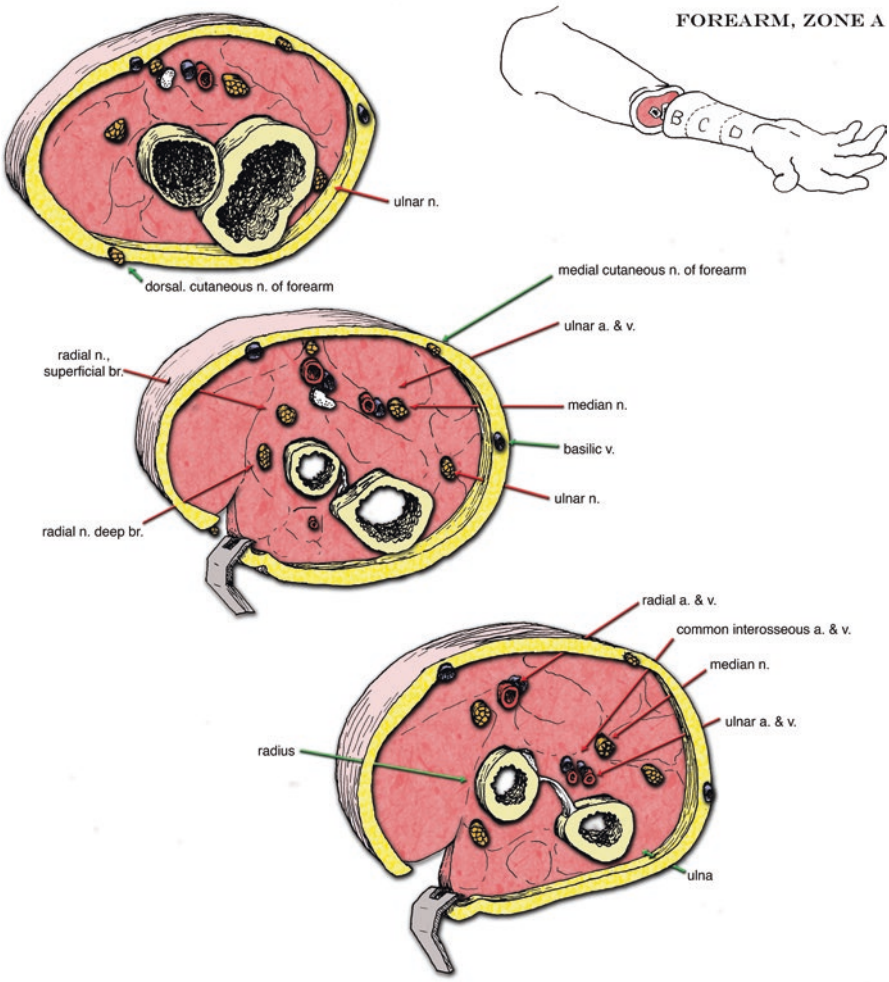


Fig. 11.13 The deep branch of the radial nerve winds around the lateral side of the humerus within the substance of the supinator muscle. The brachial artery divides into its terminal branches (the common interosseous artery and the ulnar artery) in Zone A and is anterior to the proximal ulna distally. Transverse screw placement into the proximal radius is dangerous because of the location of the deep branch of the radial nerve

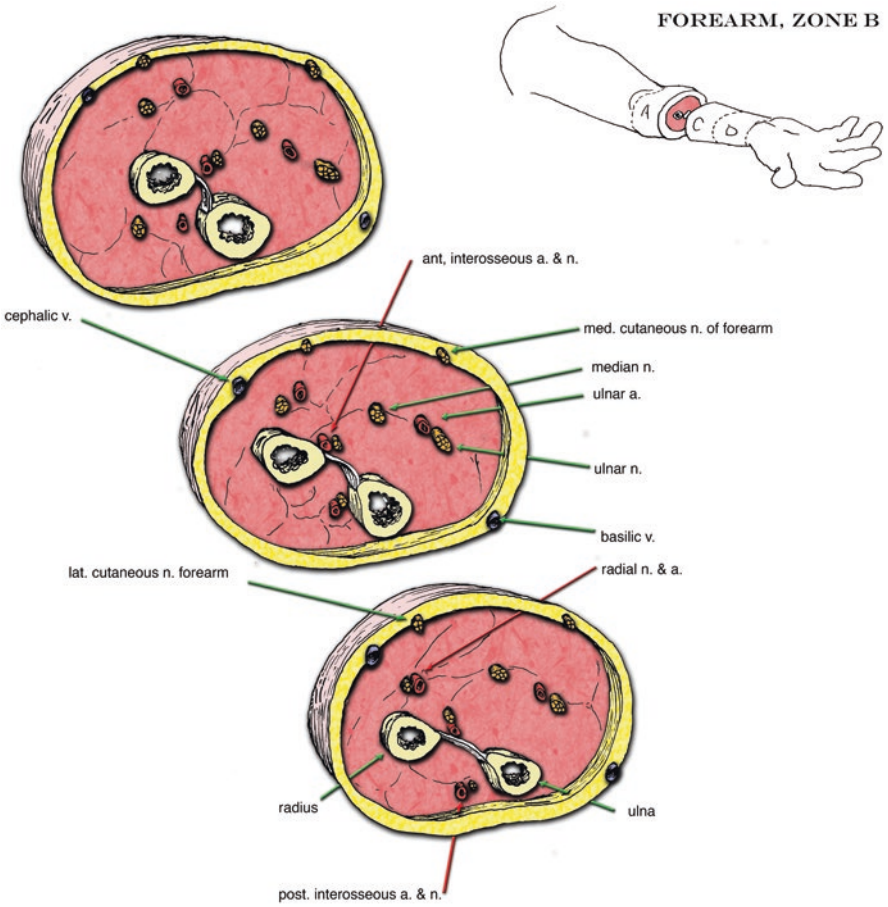


Fig. 11.14 The radial, ulnar, and median nerves remain in relatively constant position throughout Zone B. The anterior interosseous artery and nerve lie on the anterior surface of the interosseous membrane. The deep branch of the radial nerve lies adjacent to the posterior interosseous artery, posterior to the interosseous membrane, and separated from it by muscle

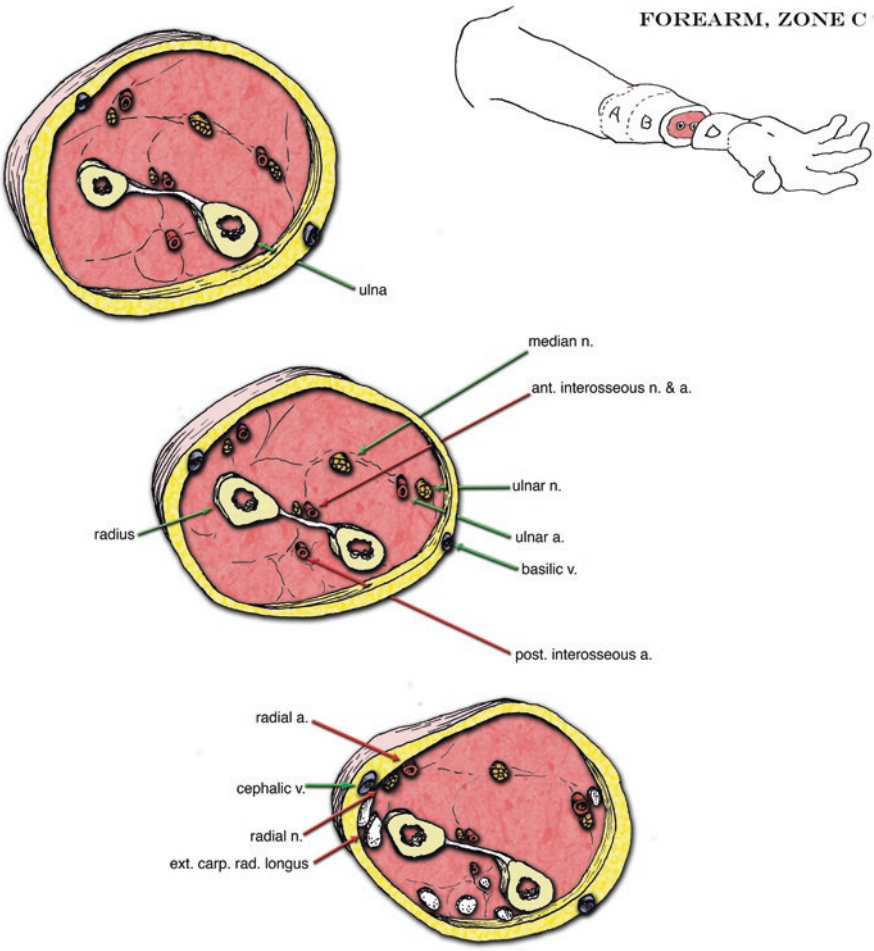


Fig. 11.15 The superficial branch of the radial nerve and radial artery is anterior to the radius in Zone C, becoming more lateral and superficial in the distal part of this zone. The median nerve remains in the middle of the forearm, surrounded by muscle. The ulnar nerve and ulnar artery remain anteromedial to the ulna throughout Zone C

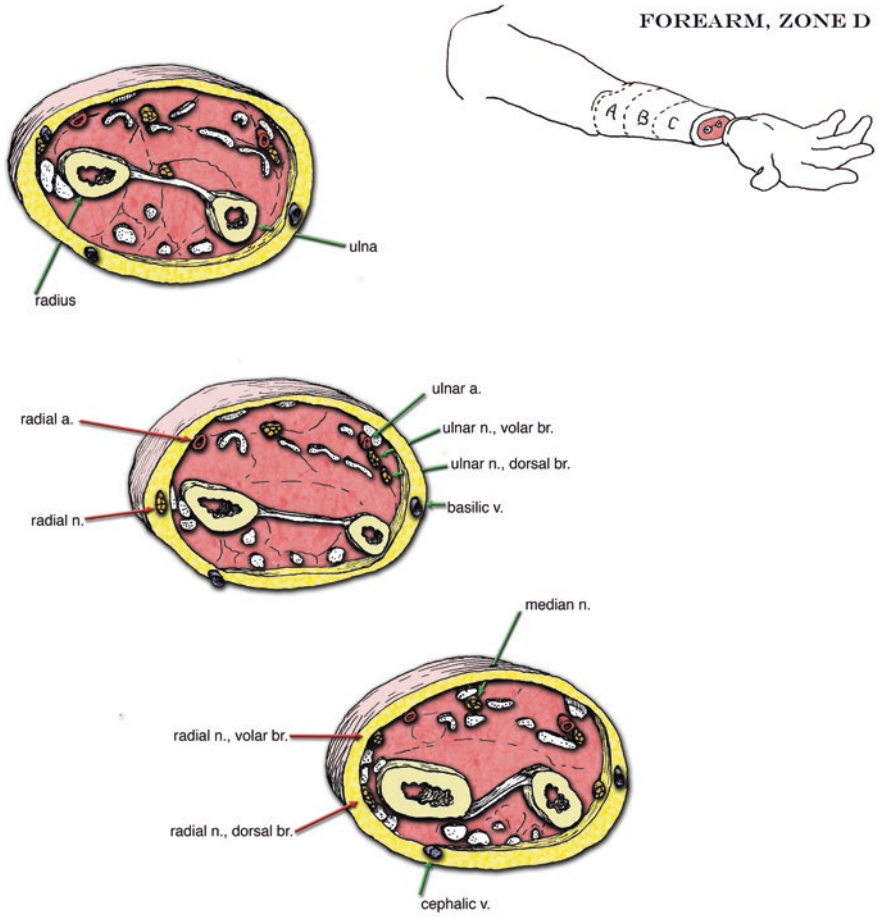


Fig. 11.16 The radius and ulna are posteriorly located in the cross section of the forearm. The radial nerve is lateral to the shaft of the radius, dividing into dorsal and volar branches in Zone D. The median nerve remains within the volar muscle mass. The ulnar nerve divides into dorsal and volar branches, the dorsal branch passing to the posterior aspect of the distal forearm. The extensor and flexor muscles become tendinous in Zone D

As a rule, when a structure is within one bone diameter (typically 25 mm) from an osseous structure on the side opposite from drilling or implant insertion, it could conceivably be injured by inadvertently plunging past the far cortex of the bone. It will be noted, however, that not all structures near bones are flagged with red arrows. In many locations, certain pathways for transcutaneous implants are not commonly used; hence, some structures near the bone are not flagged red.

Likewise, even though upper extremity intramedullary bone lengthening is only now starting for the humerus and, for technical reasons, no FDA approved arm or forearm lengthening device yet exists, both regions are included in the atlas in anticipation of future developments.

References

1. Green SA. Complications of external skeletal fixation: causes, prevention and treatment. Charles C. Thomas: Springfield, IL; 1981.
2. Vidal J, Buscayret C, Paran M, Fischbach C, Brahin B. Utilisation de la technique de papineau dans le traitment des fractures ouvertes. Acta Orthop Belg. 1976;42:49.
3. Kreuz CM, Shaar FP. Manual of fractures; treatment by external skeletal fixation. Philadelphia, PA: Saunders; 1943.
4. Eycleshymer AC, Schoemaker DM. A cross-section anatomy. New York, NY: D Appleton-Century; 1911.
5. Browner B, editor. Skeletal trauma. Philadelphia, PA: Saunders; 1991.
6. Alajmo G, Schlegel U, Gueorguiev B, Matthys R, Gautier E. Plunging when drilling: effect of using blunt drill bits. J Orthop Trauma. 2012;26(8):482–7.

Index

A
Abbott, L., 4–6
Achondroplasia, 43, 44
Albizzia®, 35
Amputation, 162, 163

B
Bifocal treatment, 179, 180
Bliskunov®, 34
Blocking screws
  biplanar, 107
  bone fragment, 105
  delayed, 109
  drill bits, guidance, 111, 112
  guidance screws, 110–111
  incorrect and correct, 106
  loosening, 105
  medial angulation, tibia lengthening, 108
  metaphyseal region, 103
  multiple, 111
  prevention, 110
  regenerate enhancement, 112–113
  secondary, 107–108
  single, 104
  tibia lengthening, 104
  transverse, 105
  varus deformation, 109
  varus femur deformity, 108
Bone deformation
  femoral, 94, 95
  forearm, 95
  limb elongation, 94
  longitudinal fascial bands, limb, 93
  patterns, 94
  ring prophylaxis, 95, 96
  tibial deformation, 94
Bone mineral density (BMD), 143
Bone transport, 48, 176–179

C
Circular external fixation, regenerate bone, 19
Codivilla, A., 4
Compartment syndrome, 63, 64, 127
Computer-controlled hexapod fixators, 27–29
Constitutional short stature, 45
Corticotomy, 22–24
Cosmetic stature surgery, 180, 181
Cross-section atlas
  brachial artery, 196
  external skeletal fixation, 185
  femoral artery, 190
  osteotomy, 186
  plunge depth, 186
  radial nerve, 197–202
  sciatic nerve, 187–189
  tibial artery, 191–194

D
Deformity correction
  angles and axes, 56
  Ilizarov's circular external fixation method, 53
  ingenious mechanisms, 53
  intramedullary lengthening nails, 66, 67
  lateral projection planes, 56
  limb elongation surgery, 54
  preoperative planning, 55
  rotating spindle devices, 43
  small angular and rotational, 53
  tibial varus, 56
Depth plunging, 63
Dislocations, lower leg, 120–123
Distraction osteogenesis, 7, 8, 17, 27
  modifications, Ilizarov apparatus (*see* Ilizarov apparatus)
Drilling vent holes, thermal injury, 64

S.A. Green, M.T. Dahl, *Intramedullary Limb Lengthening*,
DOI 10.1007/978-3-319-60297-4

E
Excessive correction, limb
  angulation, translation and rotation, 114–117
  osteotomy site, 113–114
  translation and rotation, 114–115
External fixator, elimination, 33, 34
External remote controller (ERC), 175

F
Fat embolism, 51, 62, 63
Femoral deformation, 94, 95
Femoral lengthening
  advantages, 80
  antegrade nailing, 75
  bone curvature, 80, 81
  flexible reamers, 81
  reamers, 82
  retrograde (distal) portal, 77, 79, 80
  risk, 81
  trochanteric/piriformis, 76–78
Femoral valgization
  corrections, external fixators, 99–100
  deformities prevention
    blocking screws, 103–104
    joint contractures, 102
    muscle impalement, 101
    postoperative management, 101
    snug fit, 102
  excessive correction (*see* Excessive correction, limb)
  latency, 117
  preoperative planning, 101
  rate, 117
  rationale, 117
  rhythm, 118
  surgical soft tissue release, 118–123
  valgus prophylaxis, 100–101
  Wagner® fixator, 101
Fibrous interzone, 19
Fitbone®, 37–39
Fitbone nail, 42
Fixator-assisted nailing, 60, 61
Forearm deformation, 95
Forearm lengthening nail
  bones, 88
  Fitbone®/PRECICE, 88
  flexible pins, 88
  portal insertion, 89
  ratcheting mechanism, 88
Four-segment intramedullary lengthening, rickets, 46

G
Growth arrest, 160, 161
Guide wire, 52

H
Half-pin mounting, 27, 28
Hexapod fixators, 27–29
High tibial osteotomy, 172–174
Humeral lengthening
  antegrade and retrograde nailing, 82
  distal portal
    condition, 88
    flexing, elbow, 86
    images, after nail insertion, 87
    intraoperative images, 87
    pre-op x-ray, 87
    retrograde, 86
    shoulder pain, 85
    straight femoral nail, 86
  nail size, 83
  proximal portal
    antegrade, 85
    guide pin insertion, 84
    intramedullary lower extremity limb, 85
    neurovascular structures, 85
    percutaneous insertion, 83
    performance, 83
Hypochondroplasia, 44

I
Ilizarov apparatus
  bone transport, 8–9
  computer-controlled hexapod fixators, 27–29
  external skeletal fixator, 31
  half-pin mounting, 27, 28
  intramedullary nail, 30
  lengthening over nail, 30
  limitation, 30
  monolateral external fixation, 29
  plating, 31
Ilizarov, G.A., 6, 7, 10–12
Ilizarov's method
  actual maneuver, 23
  applications, 24, 25
  bone transport, 22
  closure, skeletal defect, 23
  diffuse chronic osteomyelitis, 24
  locations, 22
  monolocal compression osteosynthesis, 23
  objectives, 23
  osseous reconstruction, 22

procedural strategies, 22
sequence of maneuvers, 23
treatment, 24
vascularized cancellous bone graft, 24
Intramedullary lengthening devices, 34–43
achondroplasia (*see* Achondroplasia)
bone transport, 48
constitutional short stature, 45
elimination, external fixator, 33, 34
hypochondroplasia, 44
implant, 47
indications, bone lengthening, 47
internal, 46, 47
limitations, 48
protocol, 47
ratcheting devices (*see* Ratcheting devices)
rotating spindle (*see* Rotating spindle devices, intramedullary lengthening nail)
stature surgery, dwarfing conditions, 45, 46
Turner's syndrome, 45
vitamin D-resistant rickets, 46
Intramedullary lengthening nails
bone canals, 88, 90
compartment syndrome, 63–64
correcting deformities, 66, 67
deformities
assessment, 53–56
hidden, 56–57
depth plunging, 63
femoral (*see* Femoral lengthening)
fibula, 67, 69, 70
fixator-assisted, 60, 61
forearm (*see* Forearm lengthening nail)
full-length standing x-ray films, 57
humeral (*see* Humeral lengthening)
implant, 66
osteotomy, 59, 60, 64–66
periosteal elevation, 61, 62
preoperative planning, 52, 53
short/long nails, 52
spare implant, 66
thermal injury, 64
thickness, 90
transverse locking screws, 65, 66, 71–75
trauma, 51
trauma nails (*see* Trauma nails)
venting, 62
ISKD®, 36

L
Latency, limb lengthening, 117
Lengthening plates, 171
Limb Deformity-Scoliosis Research Society (LD-SRS), 165
Limb lengthening
anatomical modifications, 1
deformities prevention, blocking screws, 113
development, 2
distraction osteogenesis, 7–8
femoral valgization, elongation (*see* Femoral valgization)
femur, 98
frontal plane malalignment, 99
hind limb walking, 1
lower extremity, 97
lower leg, 119–123
marrow canal, 97
middle of the last century, 2
pin placement, 96
sagittal plane malalignment, 98, 99
thigh, 118, 119
tibia, 98
waypoint method, 96, 97
Limb Lengthening and Reconstruction Society-North America (LLRS-NA), 163–165
LLRS-AIM score, 164, 165
Lower leg
deforming structures, 119
fibular hemimelia, 120
gastrocnemius recession, 121
subluxations and dislocations, 120–123
Low intensity pulsed ultrasound (LIPUS), 149, 150

M
Mesenchymal stem cells (MSCs), 150
Monolateral external fixation devices, 29

N
Night positioning, 131–134

O
Opening wedge, 174
Orthofix®, 29
Ossification
biological enhancement, 150–151
bone grafting, 151
chemical enhancement, 150
defect, 148
external skeletal fixator, 147
hyperplastic regenerate, 151, 152

Ossification (*cont.*)
LIPUS, 149, 150
mechanical enhancement, 149
nail removal, 152, 153
PEMF, 149
post-removal management, 153
rate and rhythm, 152
Osteotomy, 21, 22, 24
formula, 59, 60
gap, 66
levels, 59
technique, 64, 65
tibiofibula lengthening, 67–69

P
Paresthesia, 126
Parkhill, C., 3–4
Pelvic tilting, 159, 160
Phenix®, 39
Piriformis fossa, 51, 76
Pixel value ratio (PVR), 143
Platelet-rich plasma (PRP), 150
Postoperative management
compartment syndrome, 127
contractures, 127, 128
control progressive contractures, 135
dislocation, 127, 128
distraction, 139–142
latency, 138–139
neurological problems, 125, 126
night positioning, 131–134
osseous defect, 137
osteogenesis, 134
paresthesia, 126
physiotherapy, 128, 129, 137
splinting, 130, 131
stretching, 129, 130
subluxations, 127, 128
trauma nail insertion, 125
vascular and circulatory problems, 126
PRECICE®, 39, 40
Preoperative planning
frontal plane, 54
limb lengthening, 52, 53
normal angles, 55
Pulsed electromagnetic field (PEMF), 149
Putti, V., 4

R
Rancho mounting system, 28
Ratcheting devices
Albizzia®, 35, 36
Bliskunov, intramedullary nail, 34–35
internal skeletal kinetic distractor, 36
Rate, limb lengthening, 117
Regenerate
assessment, 142, 143
biological factors, 146
BMD, 143
chemical factors, 146
classification, 143
fusiform, 143
gardening, 145–146
lengthening nails, 144
maturation, 147
mechanical factors, 147
new bone formation, 139, 140
ossification (*see* Ossification)
patterns characteristic, 145
PVR, 143
ultrasound, 144
Regenerate bone
distraction zone, 18
early phases, distraction osteogenesis, 17
fibrocyte, distraction zone, 19
fibrous interzone, 19
fibrous tissue form, 17
Ilizarov terminology (*see* Ilizarov's method)
longitudinal and transverse section, 18
maturation phases, 19–21
microscope, 17
ossification, 20–21
quality
osteotomy/corticotomy, 22
rate and rhythm of distraction, 22
stability, 21
Ring of Ranvier, 21
vascularization, 17
Residual limb elongation, 174, 175
Rhythm, limb lengthening, 118
Ring fixator, 21
Ring prophylaxis, 95, 96
Rotating spindle devices, intramedullary lengthening nail
deformities correction, 43
Fitbone®, 37
Phenix®, 39
PRECICE®, 39, 40
transferring energy, soft tissues, 39–43

S
Shoe lifts, 159, 161
Shortening procedure, 161, 162
Splinting, 130–132

Stretching, 129–131
Subluxations, lower leg, 120–123

T
Tardy regenerate ossification, 116
Taylor Spatial Frame®, 27, 28
Tensor fascia lata/iliotibial band (TFL/ITB), 118
Thermal injury, 64
Tibial deformation, 94
Tibiofibula lengthening
  block equinus, 69
  distal securing, 70
  insertion point, 71
  intramedullary lower leg, 67
  Kirschner wire, 69
  lateral collateral ligament, knee, 67
  MRI, syndesmosis, 70
  osteotomy, 67–69
  proximal securing, 69
Transverse locking screws, 65–66
Trauma nails
  fat embolism, 51
  guide wire issues, 52
  intact bone, 51
  long bone fractures, 51
  surgical technique, 51
Turner's syndrome, 45

V
Valgus prophylaxis, limb lengthening, 100, 101
Venting, 62
Vitamin D-resistant rickets, 46

W
Wagner®, 101