Burton J. Kushner

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*The gods split these creatures into two parts, creating male and female humans. The belief is that each one of us, on a deeply subconscious level, knows that something is missing within ourselves, and we seek wholeness.*

*—Rephrased from Plato,*
*The Symposium*

*Again, for Dale*
*My missing half*

# Foreword

This is a new and different type of book on strabismus. It is not your typical textbook but instead is a journey in mentorship. Within these pages, Dr. Kushner has the reader at his side as he discusses questions, issues, problems, and solutions. He does so with the great benefit of a master's long-time experience, drawing on accumulated notes and recollections from countless discussions with colleagues during his 40-year career. Both trainees and experienced practitioners will benefit from Kushner's pearls of wisdom displayed in a unique, frank, and engaging style.

There are many ways of learning strabismus diagnosis and management. This book is a virtual apprenticeship, a method of doing, seeing, and thinking, that textbooks, however well presented, typically do not fully convey to the reader.

The story is told of a question asked of the master surgeon, even though this was not encouraged during surgery. The story relates that the surgeon briefly paused—looked up at the ceiling for a second or two—and then continued his surgery. It was as if God had spoken to him, or him to God—so the story goes. Sadly, and frequently, questions asked of the master teacher are silently ignored or delayed. Not so herein. Kushner has obviously enthusiastically encouraged questions and on-the-spot discussions throughout his career, as manifested in this book. The author is a master teacher, and his notes and discussions reveal excellence in every facet of examination and management. Kushner has cleverly used icons, set in various side border positions, in order to emphasize certain points. These signaling icons were first revealed in his well-known handbook on strabismus from previous years. Some such icons are labeled "myths" or "trash cans," a clever way of highlighting points of interest. Such icons reveal, in a polite and masked way, what words may not.

In short, this author has shared his vast storage of clinical strabismus management in this new and different non-textbook style of teaching and has made his unique way of teaching available to you, the reader. Kushner's method of teaching is a true apprenticeship. It combines an amazing collection of his discussions, notes, and teachings and presents it with considerable entertainment for the reader. This book is *sui generis*—a masterpiece from a true master.

San Francisco, CA Arthur Jampolsky, MD

# Foreword

The former chief of my department was certain that understanding strabismus was a simple matter; after all, "there are only six muscles." That this is a minority opinion goes without saying. In fact, most trainees usually endorse the opposite view: that strabismus is one of the most difficult disciplines in ophthalmology to understand. Many of them find it completely off-putting and cannot wait to move on to some other subspecialty. Those of us in the field of pediatric ophthalmology and strabismus do not deny that there is much to be learned in order to care for the patient with strabismus: extraocular muscle anatomy and physiology, certain aspects of optics, the neurophysiology of eye movements, as well as surgical techniques. However, a systematic and logical framework can be constructed for the management of the various strabismus disorders, be they the everyday mundane or the vexing rarity. Burt Kushner, in this remarkable book, does just that. He shares his over four decades of experience, his thoughtful and critical analysis of his own results, and his synthesis of the pertinent literature. In doing so, he provides a carefully constructed framework within which he approaches strabismus disorders. This is not the usual attempt at a comprehensive textbook, but a carefully selected group of management problems, illustrated by a wide-ranging group of cases, thoroughly discussed from various points of view, and with a resulting set of recommendations from the author on how to best handle them.

For many years, Burt Kushner has been known as the consultant's consultant when it comes to addressing complex strabismus disorders, surgical complications, and new approaches to strabismus management. Many of us routinely seek his counsel about problem or puzzling cases in our practices. In this book, he distills his own experience and combines it with many of the fascinating questions posed to him from ophthalmologists throughout the world. He successfully compresses all of this information into a format that is orderly, logical, and practical. He is wise to describe certain key principles that are fundamental to his approach to strabismus and to reiterate them frequently, as appropriate, when discussing relevant clinical problems. There is no doubt that readers will frequently return to this book when they have specific questions concerning strabismus that need to be answered. This will be one of those well-used "go-to" books that assume a central position in the

library of those interested in strabismus. In no small part, this is because Kushner does not simply proclaim his approach to a problem but considers other approaches and arguments and why he does not prefer them. Kushner has written an invaluable book that will become a standard reference. However, I would be remiss if I did not point out that in addition to being a carefully documented scholarly work, this book is a downright good read. In this book, a recognized authority on strabismus informs and provokes with his lucid reasoning, but he also entertains.

Sausalito, CA Creig Hoyt, MD, MA

# Preface

*One of the symptoms of an approaching nervous breakdown*
*is the belief that one's work is terribly important.*
*—Bertrand Russell*

## Why Did I Write This Book?

Near the beginning of his *tour de force*, *The Art of Surgery*, the brilliant writer and Yale surgeon, Richard Selzer, wrote, "Someone asked me why a surgeon would write. Why, when the shelves are already too full? They sag under the deadweight of books. To add a single adverb is to risk exceeding the strength of the boards. A surgeon should abstain."

I am sitting now in my office, gazing at my bookshelves laden with strabismus textbooks. There are indeed many excellent ones. Some are basic "how to do it" manuals. Some are encyclopedic and thoroughly reference the classic literature and contemporary publications. There are beautifully illustrated surgical atlases, texts that are collections of case presentations, and even books dedicated to the more esoteric aspects of complex strabismus. So I am asking myself, "Does the world need another textbook about strabismus?" Given the fact that you are holding this book in your hands indicates my answer must be "yes." But I feel it is not just another strabismus text that is needed, but a different and novel kind of book.

In a sense, I have been preparing to write this book for many years, but was probably unaware of that fact. In the daily course of teaching residents and fellows, and replying to e-mail requests for my opinion, I would frequently mention some relevant important point (or "pearl," as we so often call it) that never appeared in the textbooks. Some of these pointers reflected my own experience gained in over 40 years of clinical practice. Sometimes they reflected a critical reading of the existing literature that went beyond the published abstract. Often they evolved after meaningful discussion with colleagues and trainees. I have come to learn that there is no one best way to manage most complex strabismus problems. Most often my choosing a treatment option involves assessing and weighing various trade-offs. For example, one approach may be simple and safe and fairly predictably lead to a satisfactory outcome. But an alternative approach may be technically more difficult

and have a higher chance of an adverse outcome. Yet when it works well, the result is better than what can ever be achieved with the simpler procedure. As I have often said, "There are many roads to orthophoria!"

Trainees working with an experienced and thoughtful mentor have the luxury of having discussions about these concepts in real time. Unfortunately, strabismologists and pediatric ophthalmologists who do not still have (or never had) that kind of relationship with a mentor do not have the benefit of this type of learning. Which gets me back to my reason for writing this book. I want to simulate in print, to the best of my ability, the stimulating and challenging discussions that occur regularly between a strabismus mentor and mentee. I hope you will have as much fun reading this book as I had writing it.

## About This Book

This book is not intended to be a comprehensive strabismus primer, of which there are many excellent ones from which to choose. Although novices may benefit from the discussions herein, it is not intended to replace a basic textbook. It was written with the assumption that the reader knows the basics of eye muscle and sensory physiology, the strabismus exam, and standard treatment protocols. I assume the readers already know the appearance of most strabismic conditions. Consequently, this book is intentionally sparse on clinical photographs of most motility disorders. There are many excellent comprehensive strabismus textbooks to provide them.

As a catalyst for discussion, I have drawn heavily from e-mail and curbside consultation requests I have received, as well as patient case histories presented to me by trainees. I have been saving the notes from many of these for years, and upon reviewing my files I find that many themes are recurrent. So in many instances the cases I write about are hypothetical conflations of multiple patients' case histories, and whenever I personalize the patient, it is always fictitious. I have chosen each case because it illustrates a particular principle (or several) that I feel is important and relevant. So for this reason, as well as the fact that most of the cases are de-identified hypothetical constructs, I cannot say exactly how they were treated and what the result was, in most cases. I feel the principles they represent are more important than the outcome of an individual patient. This is a similar approach I used while editing the Grand Rounds section for *Binocular Vision Quarterly.* Although that section was very popular, I was often asked why I did not give outcomes—what I did and how it turned out. My reason then is the same as I offer now. Any given patient may have a good outcome with less than ideal treatment, and a good treatment choice may not always work well in a particular patient. The natural instinct is to conclude a good outcome meant the approach was optimum and, perhaps, any other approach would not have been, and a bad outcome means the converse. That, as most experienced strabismologists know, is not the case.

There are many roads to orthophoria

Of course, in the end, the treatment approaches I recommend are the ones that have worked best for me and my patients. As Frank Sinatra's lyrics go, "I did it my way." But that is not to imply that is the best way, or even the only way. I am particularly fond of one aphorism that hangs on my office wall, and I look at it each day. It reads, "The teacher whose students do not disagree with him is not a teacher. And the teacher who fears his students' disagreement is also not a teacher." These words, which are a modification of a Talmudic saying, reflect my most firmly held belief about the role of teachers. I encourage you, the reader, to think through the issues presented here in this book for yourself. I do not expect you to agree with everything I wrote, and hopefully that will make me a better teacher.

The teacher whose students do not disagree with him is not a teacher

Reviewing the material in this book I see two separate themes frequently repeated. (1) If the existing scientific literature confirms the superiority of an approach, I adopt it. If data are unconvincing, I do what makes sense to me. And, (2) If I have a choice between two good procedures, one of which is easily reversible and other is not, I choose the reversible option.

Another principle that will come up frequently with respect to managing incomitant strabismus is that of anticipating iatrogenic problems and acting preemptively. Often a surgical plan to address a primary position deviation may predictably worsen alignment in sidegaze or downgaze. This can be mitigated by recognizing in advance that this will occur and incorporating remedial measures in the surgical plan. This is a concept I have heard Ed Buckley repeatedly emphasize, and he was wise for doing so.

## How to Use This Book

Throughout this book, I use the icons below to identify and categorize topics and material. I used a similar approach in my online handbook "Eye Muscle Problems in Children and Adults: A Guide to Understanding" (https://www.ophth.wisc.edu/education/eye-muscle/), where this style was used and well received.

**Basic Information.** Information that is considered basic but important for you to read in order to understand a given topic is accompanied by the symbol of stacked books.

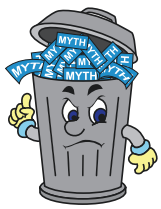
**Advanced Information.** A graduate identifies information that delves more deeply into the technical aspects of a subject that are of an advanced nature. They may be of more interest to the tertiary care strabismologist.

**Important Point.** A string around an index finger denotes an important point. This identifies a particularly useful fact or concept that is worth remembering and perhaps frequently not recognized.

**Pearl.** The pearl in a shell denotes an important clinical "pearl on strabismus." These typically are clinical pointers that have not been stressed in standard textbooks. In some cases, if deemed appropriate, I may repeat material from the body of the adjacent text, labeled with this icon.

**Question.** A question mark will identify material that was initiated by an e-mail or verbal curbside consult request. The question, as presented here, may be a conflation of several related questions I received, and hence may be hypothetical, not actually describing a real-life patient or clinical situation.


**Reply.** This icon denotes my reply to a question. Will Rogers once said, "A humorist entertains, and a lecturer annoys." I feel I would have succeeded in part of my mission in writing this book, if you, the reader, find it pleasurable as well as informative.




**Myth.** There are many myths and misconceptions that relate to strabismus management. These can represent misleading and incorrect information that may confuse your understanding of the subject. The trashcan (or perhaps Facebook or Twitter) is the appropriate place for these incorrect ideas.

**Try This Experiment.** To help you understand certain subjects, I may suggest you perform simple experiments. These will be identified by this scientific symbol.

## Abbreviations

I believe reading and comprehension is impeded when one the reader is confronted with an unfamiliar abbreviation, even if it has been defined earlier in the text. I limited my use of abbreviations to those that should be readily familiar to the strabismologist. In all cases I define them the first time they appear. For the benefit of those readers who may not read this text in its entirety, I also list all the abbreviations used in this book in the following table.

| Abbreviation | Definition |
|---|---|
| AC/A | Accommodative convergence to accommodation ratio |
| Adv | Advancement |
| AES | Anti-elevation syndrome |
| AFG | Active force generation |
| AHP | Anomalous (abnormal) head position |
| ARC | Anomalous retinal correspondence |
| ASI | Anterior segment ischemia |
| AT | Anterior transposition |
| BI | Base in |
| BO | Base out |
| CI | Convergence insufficiency |
| D | Diopter |
| DVD | Dissociated vertical divergence |
| E | Esophoria |
| EOM(S) | Extraocular muscle(s) |
| ET | Esotropia |
| FD | Forced ductions |
| H | Hyperphoria |
| HT | Hypertropia |
| HYPO | Hypotropia |
| IO | Inferior oblique muscle |
| IPD | Interpupillary distance |
| IR | Inferior rectus muscle |
| L | Left |
| LR | Lateral rectus muscle |
| MED | Monocular elevation deficiency |
| MR | Medial rectus muscle |
| NPA | Near point of accommodation |
| NRC | Normal retinal correspondence |
| Nys | Nystagmus |
| OA | Over action |
| OD | Right eye |
| OS | Left eye |
| PACT | Prism and alternate cover test |
| PAN | Periodic alternating nystagmus |
| Parentheses ( ) | Denotes an intermittent deviation |
| PAT | Prism adaptation test |

| Abbreviation | Definition |
|---|---|
| PD (Δ) | Prism diopter |
| PF | Posterior fixation |
| PI | Phospholine iodide (ecothiopate) |
| Prime sign (′) | Denotes a measurement at near |
| PVR | Proliferative vitreoretinopathy |
| PUCT | Prism under cover test |
| R | Right |
| Recess | Recession |
| Res | Resection |
| R&R | Recess-resect procedure |
| ROP | Retinopathy of prematurity |
| ScPh | Scobee phenomenon (*formerly called* tenacious proximal fusion or TPF) |
| SOP | Superior oblique palsy |
| SO | Superior oblique muscle |
| SPCT | Simultaneous prism and cover test |
| SR | Superior rectus muscle |
| TED | Thyroid eye disease (Graves' orbitopathy, endocrine ophthalmopathy) |
| UA | Under action |
| VEP | Visually evoked potential |
| X | Exophoria |
| XT | Exotropia |

# Acknowledgments

This book would not have been possible without the numerous residents and fellows I have been privileged to train. Their hunger to learn, manifested as insightful questions, challenged me daily to consider and reconsider what I thought about strabismus, how it should be managed, and above all—why. I am indebted to them, and similarly to the numerous colleagues from around the world who regularly fill my e-mail in-basket with requests for advice on management of their complex strabismus patients. This book is a product of those in-person discussions and written exchanges. I wish to thank those people who stretched my mind in these ways.

I also have to add a special note of thanks to Creig Hoyt, MD, MA. He was my first reader and editor of this book. His careful reading of the manuscript and insightful comments were invaluable. He made numerous suggestions for expansion, deletion, or clarification of material that greatly improved the final product. I, as well as all readers of this book, reap the benefits of his expertise.

# Contents

# History

# 1

*That men do not learn very much from the lessons of history.*
*is the most important of all the lessons of history.*

—Aldous Huxley

## Basic Information

Most strabismus textbooks state the obvious with respect to history taking: Ask when the problem began? Is there a family history of similar problems? Did anything precede or precipitate the issue? What is the nature (frequency and severity) of the problem and in what circumstances does it occur, etc. All of this information is important. However, I feel that there are some additional questions that can be helpful to ask, and crucial in sorting out the patient's problem. *Lagniappe* is a word of French Creole origin that roughly means "a little something extra." It is still commonly used on the streets of New Orleans where street vendors will throw in something extra with your purchase. I want to offer *lagniappe* with respect to history taking.

## Basic Information

After years of caring for patients we often find ourselves repeating the same words which can almost sound like scripts. This is not a bad thing. What works for one may work for many. For all new patients, or established patients with new symptoms, I always ask, "What were you anticipating I would recommend for XXX (your symptom), and *also* what were you hoping I would say. I recognize the two are not the same. That way if I recommend something different than what you were expecting, I want to be sure to spend time explaining why I am not saying what you hoped for or anticipated." In addition, I ask patients what they were most afraid I would say. I have found this extremely useful. If the patient expected me to say, "it is nothing to worry about," and I in fact am recommending an involved treatment plan, I want to be sure that I and the patient are not blindsided by differing expectations.

## Pearl

*Listen to Patients with Five Ears*

I learned about this from a Zen master [1]. The five ways to listen are for (1) the facts, (2) the emotions, (3) the body language, (4) how the conversation is affecting you, and (5) what is not being said. Always keep in mind that what the patient most wants us to hear is exactly what may remain nonverbalized. It is up to us to hear those unspoken words. This is particularly challenging since the advent of electronic medical records, which require us to spend so much time facing the computer monitor.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_1

## Pearl

*Verbal Discourse Is Like a Scattering of Birds*

We speak our words and they fly off to their own personal destiny. What we think we say may not be what is heard. Mark Twain once said, "The difference between the almost right word and the right word is really a large matter—'tis the difference between the lightning bug and the lightning." Words embody connotative and denotative meaning, each of which has singular power and energy. Therefore, choose your words with care. For example, it is so much better to tell a patient that dilating drops will *relax*, rather than *paralyze* their pupils. *Relaxation* has good energy; *paralysis* sounds ominous. Tell patients that after dilation they may be *sensitive* to light, rather than *bothered* by light. If you suggest they will be bothered, they probably will be [1, 2].

## Basic Information

Years ago I examined 1-year-old Matthew whose parents felt that his one eye had started crossing. His mother was a pediatrician who regularly referred patients to me, and I considered her a good observer; her father and grandfather were also physicians who both observed the same crossing of one eye. All of his caretakers, however, felt that the problem was mild and infrequent. I found his motility to be normal and he had a bilateral and symmetric low hyperopic refractive error. I suggested that we just reevaluate him in the future. When Mathew returned, his mother said that the crossing had gotten much worse and they were seeing it frequently. Even after working hard to break down his control using accommodative targets at near, I could not document any esotropia. Because his mother was so insistent the eye was crossing, I decided to try a diagnostic trial of ecothiopate iodide (phospholine iodide) eyedrops [3]. I reasoned that if the Matthew's caretakers all felt that the problem improved on the medication, I would know I was dealing with an accommodative esotropia that he was simply controlling while in my office; his mother thought this made sense. When Mathew returned after several weeks on the drops his mother said that the crossing was even worse. To my surprise, I saw he manifested a constant **EX**otropia of about 30 PD (prism diopters). I asked "When did Matt's eye start deviating laterally?" "That's what I've been telling you it does," his mother said, "It crosses *to the outside*." Needless to say, I stopped the drops, but he never regained control. I had to operate on him for his exotropia. Fortunately, his outcome was good (and his mother continued to send me patients until she retired). But I learned something very useful from this experience. Patients (even if they are medically sophisticated) do not speak the same language as physicians with respect to medical terms. Ever since my experience with Mathew, I have found it useful to say to patients, "Describe in your own words exactly what you see wrong with the eye position. Is it the right eye, left eye, or both? Does it go in, out, up or down? Is the misalignment always present, and if not, when does it occur?" I will then ask if they feel the problem is occurring at the present moment, and if so, reflect back to them if I see the same thing they are describing. This is particularly useful in cases of pseudostrabismus. I can reassure the parent that what they are seeing is something I also see, and is normal.

> Patients do not speak the same language as physicians with respect to medical terms.

## Important Point

*Obtain Prior Records Including Op-Notes, Pre-op Measurements, and Refraction Data*

It always surprises me how often I will hear that prior records were not available, only to learn that with persistence and the right approach, I can get everything I need. Typically operative reports are not included in doctors' office records.

This also surprises me, but it is a reality. Usually it takes a call to the hospital or surgery center to ask specifically for operative reports. If they are from the distant past, the immediate response is usually that they are unavailable. I then ask them to check archived files, including microfilm, and more often than not the records can be found. I find these crucial in many circumstances. A few examples include the following:

1. Consider the adult patient with unilateral amblyopia, a large exotropia, and who refuses surgery on his dominant eye. There is a history of prior surgery for what he thinks was exotropia, as he only remembers himself being exotropic. If prior records show that he actually had undergone surgery for esotropia, there is a good likelihood I can get a lot of correction and a good outcome with an advance-recess procedure on his amblyopic eye. But if he had a generous recess-resect procedure for exotropia in the past, I will be much more guarded in what I tell him I can accomplish, if I must confine surgery to the amblyopic eye.
2. For the older patient with a new onset of symptoms such as diplopia, or a change in deviation, it is invaluable to know what the alignment was, as well as the refraction findings and management, just prior to the onset of symptoms. Usually the key to eliminating symptoms lies in replicating the pre-symptom state (see Chap. 9).
3. There are times when it may not be evident, even on surgical exploration, that an irreversible surgical procedure had been previously performed. For example, I have seen cases of consecutive exotropia in which a second surgeon explored and resected the medial rectus muscle and got little effect. He did not realize that previously a posterior fixation suture was placed in the medial rectus muscle 12 mm posterior to the insertion. This was not evident on exploration, and operating on the muscle anterior to the posterior fixation suture was not effective.
4. I was referred a patient who reportedly underwent a superior oblique tuck for a superior oblique palsy, and was worse immediately after surgery. I read the operative report which was titled "superior oblique tuck"; however on reading the steps described, it was evident that the surgeon inadvertently did a superior oblique tenotomy. This was an example of his mixing up the two different planned surgical procedures for two patients he was operating on the same day, and doing the operative report by rote.

## Pearl

*Always Ask to Have Prior Records Sent to You in Advance of a Consultation, and Read Them*

I once read an article for the lay public on how to successfully navigate the medical delivery system and get good care. It specifically advised patients to hand deliver prior records to a consultant at the time of the consultation, and it advised against having them sent in advance. I totally disagree. When I get records in advance and read them thoroughly, I almost always discover that important information is missing. As stated above, usually office records do not include operative reports. If the referring doctor generated a narrative summary instead of sending actual office notes, there are often key pieces of information that I need which were omitted. Getting them in advance allows me time to request what is needed. Also, it gives me the opportunity to schedule any special tests that I may want that may need to be anticipated in advance. e.g., orbital imaging, extra time with an orthoptist, and diplopia fields.

## Important Point

*If Prior Images Were Obtained, Have the Actual Scans—Not Just the Report—Sent in Advance*

I really like to review outside scans in person together with one of our neuroradiologists. I cannot do this if the scans are brought to the appointment, and certainly cannot if I just get the report. Scans done on the outside, especially if the area of interest is the extraocular muscles

(EOMs), are usually of insufficient quality to make a good diagnosis. Even if performed at prestigious institutions, most of the time the radiology department does not have a protocol in place for studying the EOMs and the resolution is poor. More often scans are of the brain, and the views of the EOMs are incidental. Reviewing the scans with radiology in advance of the consultation may result in a recommendation that added imaging done differently—perhaps a dynamic scan—may be useful. I never considered myself particularly expert in assessing images of the brain, and usually defer to the judgment of our neuroradiologist. But when it comes to assessing the eye muscles, I trust my own assessment more. Many neuroradiologists receive little or no formal training in assessing EOMs. Many have trained before our understanding evolved about EOM paths and orbital pulleys, and have little knowledge thereof. In fact, scans of the orbit are typically read by neuroradiologists by default because the orbits show up on scans of the brain, and neuroradiologists typically interpret images of the head and brain. See Chap. 17, Fig. 17.3, for a relevant example.

Always review orbital images personally in conjunction with neuroradiology.

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# The Examination

# 2

*The power of accurate observation is commonly called cynicism by those who have not got it.*

*—George Bernard Shaw*

This chapter is structured with the assumption that the reader knows the basics of vision testing, measuring strabismus, and assessing ocular rotations. As in the prior chapter, it offers *lagniappe*—a little something extra.

## Vision Testing

### Recording Visual Acuity

#### Basic Information

When recording visual acuity findings for young patients, especially if they are amblyopic, always record the optotype used, and if its testing was linear or isolated. Subsequent vision testing may appear to suggest deterioration or improvement, when in fact it might just reflect the use of a harder or an easier optotype. This is particularly important if the subsequent testing is being done by you for a second opinion or referral.

### Testing the Preliterate Child

I prefer the term "preliterate" to the more commonly used "preverbal," as the latter is often used to describe children who can talk (are not preverbal) but cannot yet read (are preliterate).

#### Basic Information

*Fixation Preference and the Induced Tropia Test*

Here I would like to introduce a concept that I call a "one-way test." By that I mean, if the test gives one result it is very meaningful. But if it gives the opposite result, it may not be as useful. There are many patients who have a very strong fixation preference despite having equal or near-equal acuity. Examples may include a patient with significant anisometropia but is not or no longer amblyopic, a unilateral aphakic or pseudophakic child post-amblyopia treatment, and even some patients with essentially symmetric refractive errors. These children may not ever alternate on the induced tropia test or by fixation preference if they are tropic. So the absence of alternation does not necessitate a diagnosis of decreased acuity in the non-preferred eye. On the other hand, there would never be free alternation in the presence of amblyopia. This is an example of a "one-way test"; alternation is very meaningful, but lack of alternation is not diagnostic. Still, these are useful tests because they are low-tech ways of possibly ruling out amblyopia.

#### Important Point

Lack of alternation by fixation preference or induced tropia testing may, but does not necessarily, mean acuity is unequal.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_2

### Basic Information

*Grating Acuity Tests Will Overestimate Acuity in Some Circumstances and Underestimate It in Others*

Nevertheless, they are useful for comparing acuity in the same child from one visit to another, comparing right (R) and left (L) eyes, and for comparing average responses in population groups as has been done in the retinopathy of prematurity clinical trials. Increasingly we are driven by metrics in a culture that values the precept "You cannot manage what you cannot measure" (sic). Grating acuity tests have the advantage of allowing us to put a number on visual acuity, something which fixation preference testing does not provide. However, it is most important to know that the Snellen fractions provided by grating acuity tests do not have the same meaning as acuities obtained by recognition acuity tests (e.g., reading the eye chart).

### Try This Experiment

The principle underlying grating acuity tests is that the brain will recognize there being a pattern present if stripes are of a certain width and contrast. Preferential looking tests, such as Teller Cards, assume that an infant would rather look at a pattern than a homogeneous gray card. As such, a child's brain needs to only determine a pattern is present in order to be said to have "seen" it. Visually evoked potential tests that use spatial frequency gratings as targets are based on the assumption that the brain will recognize that the pattern is present, and this recognition results in neural activity in the visual cortex. However, being able to merely detect that a pattern is present is an easier task than telling what that pattern is. The latter is what recognition tests rely on. Do the experiment depicted in Fig. 2.1. This is one reason I strongly advocated that grating acuity test should not be used for social service purposes [1, 2]. This was shortly after the commercial introduction of Teller Cards, and I was seeing many children who were losing social service benefits because they were found to have better acuity with this test than had been previously suspected, yet clearly they had reduced acuity levels that should have allowed them to qualify for aid.

Grating acuity tests should not be used for social service purposes.

### Advanced Information

*Preferential Looking Versus Sweep Visually Evoked Potential (VEP) Testing*

Preferential looking tests not only require the infant to identify the presence of a pattern, but they also require the child to make a motor response by looking toward the pattern. Sweep VEP testing does not require the motor response, but only the awareness that the pattern is present. So children with neurologic abnormalities involving the higher eye movement control centers may have Teller Acuity responses that underestimate what they actually see as compared to responses with sweep VEP. An extreme example would be a child with ocular motor apraxia who has not yet developed neck muscle strength to make head thrusts. These children are often thought to be blind as infants.

## Measuring Strabismus

### Basic Information

*The Exam Lane*

With the increased cost of office space, there has been a push from clinic administrators and office managers to have all ophthalmologists, including strabismologists working in shortened lanes or mirrored lanes that optically simulate 20 ft. I have heard many colleagues say that they feel this does not make a difference in their management of strabismus as compared to the standard 20-ft

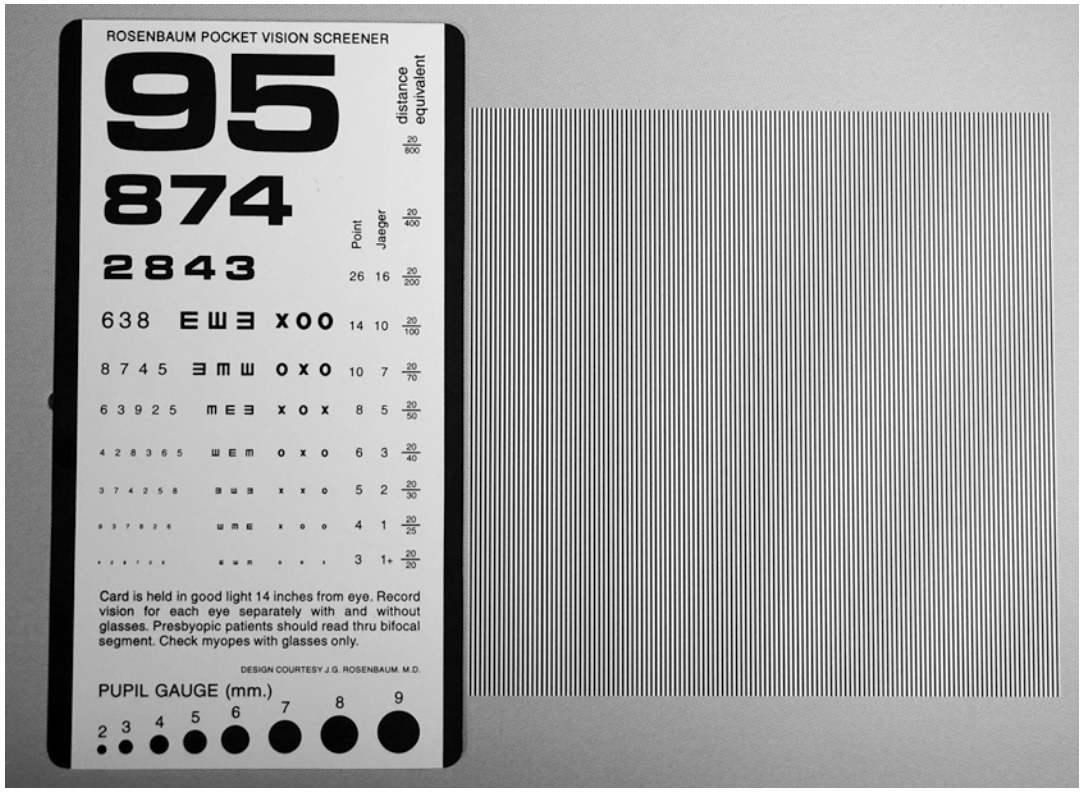



**Fig. 2.1** Rosenbaum near-vision card (*right*) and Teller Acuity Card (*left).* The Snellen equivalents of the angle subtended by the Teller Card gratings are approximately the same as the J-3 line on the Rosenbaum card. If you optically defocus the figure by viewing it through a plus lens of sufficient power that you can just read the J-16 line on the left, you will still be able to detect that stripes are present on the Teller Card, which subtend a much smaller angle than the J-16 optotypes on the Rosenbaum card. In this scenario, your acuity would be 10/200 with the Rosenbaum card but at least 20/40 with the Teller Card

lane. The reality is, however, that the one study that compared strabismus measurements in these different settings found that shortened or mirrored lanes overestimate the angle of misalignment and underestimate control at distance fixation in esotropes, and underestimate the angle and overestimate control in exotropes [3]. The previously described study utilized a standard mirror, about 24 in. square for the mirrored lane. Theoretically if the entire far wall of the exam room was a mirror, measurements might more closely approximate those from an actual 20-ft lane. With such a mirrored arrangement, all visual targets projected on the retinal periphery would also travel 20 ft, and not just those projected near the macula as is the case when a small mirror is used. A very large mirror may not alter the convergence drive as much as a small mirror. Such an arrangement was tested and the measurements were found to be closer with the large mirror to those obtained in a 20-ft lane than had been reported with the small mirror [4].

### Important Point

If you are compelled to use a shortened room with mirrored system, use one in which the entire end wall is a mirror.

### Basic Information

*Estimating the Deviation by Light Reflex*

Sometimes you cannot measure a deviation with the gold standard, the prism and

alternate cover test (PACT), either due to the patient's age, cooperation, or presence of unsteady fixation in the deviating eye. In such cases you need to resort to a light reflex test, either the Hirschberg or the Krimsky test. Keep in mind that these tests are subject to extremely variable interpretation and are not very accurate. One study assessed the accuracy and consistency of 16 very experienced strabismologists in grading the angle of strabismus using the light reflex tests in patients with known angles of misalignment [5]. The results showed a high degree of error and inconsistency, with the Hirschberg test being more inconsistent than the Krimsky test. The Krimsky test is more accurate than the Hirschberg test, because it is easier to tell when the light reflex is centered, than to estimate by how many millimeters it may be decentered. Also, the Hirschberg test relies on the Hirschberg ratio, which is a conversion formula that assumes that each millimeter of decentration corresponds to a given number of prism diopters (Δ) of misalignment. Studies to accurately quantify the Hirschberg ratio have resulted in widely differing conversion ratios, ranging from 15 Δ per millimeter to 22 Δ per millimeter [5]. The Krimsky test does not have a similar source of error. Light reflex tests may provide the best measurement we can obtain for a very young or uncooperative patient, but they are not ideal. Certainly, when evaluating any study that relies on measurements obtained this way, be wary of any conclusions. As the expression goes, "garbage in, garbage out."

See Chap. 21, Case 21.6, on **page 315** for a representative example of some errors that can stem from relying on light reflex tests for measurements.

### Important Point

Light reflex tests are highly inaccurate. The Krimsky test is far superior to the Hirschberg test.

### Basic Information

*The Krimsky Test—Over Which Eye Should You Put the Prism*

One argument says to put the prism over the fixing eye, as it is supposedly easier to observe the centration of the light over the deviating eye if it is not blocked by the prism. The contrary argument is that the prism should go over the deviating eye, as that is the deviation you want to measure. Which is correct? The fact is that as long as the strabismus is completely comitant, it probably does not matter. However, if there is any horizontal incomitance, placing the prism over the fixing eye will introduce more error in an intrinsically error-prone test. With the prism over the fixing eye, that eye is no longer in the primary position when viewing the fixation target, and you will actually be obtaining a side-gaze measurement. I never felt that the presence of the prism over the deviating eye impeded my ability to see the light reflex, so I always default to placing it there.

### Advanced Information

*If You Stack Two Horizontal or Two Vertical Prisms, the Result Is Not the Same as One Prism Equal to the Sum of the Two*

The value of a prism is a trigonometric function of the angle of the apex. Twice the sine or cosine of any angle $X$ is not equal to the sine or cosine of $2X$ (Fig. 2.2). So if you are measuring a deviation that exceeds the maximum prism in your prism box, do not stack two together. It is two prisms theoretically possible to calculate the sum of two added prisms. But the resultant final prism power is greatly influenced by the angle between the two opposing surfaces of the stacked prism, and that is hard to control when measuring patients. Instead, you must split the prism between the two eyes. This does introduce two different but similar errors. One is


**Fig. 2.2** On the bottom is a 40 Δ prism. On top of that is a sandwich of two 20 Δ prisms. The top prism sandwich has a much greater value than the 40 Δ prism, as evidenced by it displacing the image of the pencil much further

similar to that described above with the Krimsky test. Namely, the fixing eye is no longer in the primary position. The second occurs because of the nonlinear definition of a prism diopter. Table 2.2 provides conversion data for two prisms, one placed over each eye [6]. This error starts becoming clinically meaningful (more than about 5 Δ) when the sum of the value of the two prisms is 50 Δ or more.

### Important Point

If you measure a patient by using two horizontal prisms, one before each eye, an error of about 5 Δ or more starts to occur when the sum of the two prisms is more than about 50 Δ due to the nonlinear definition of a prism diopter. For these large deviations, you should convert the sum using Table 2.1.

### Advanced Information

*The Orientation of the Back Surface of a Prism to the Patient's Face Will Affect Its Effective Power*

Ideally loose plastic prisms should be positioned so that the back surface is approximately parallel with the frontal plane of the patient's face. This is called the frontal plane position. If the orientation of the prism deviates from this, measurement error is introduced (Fig. 2.3).

### Basic Information

The manifest deviation is measured by the simultaneous prism and cover test (SPCT), and the total (manifest plus latent) by the prism and alternate cover test (PACT). Both are important and both should be recorded and noted separately. The SPCT tells me *whether* further intervention is needed, and the PACT drives *what to do* if further intervention is needed. For example, if the SPCT is under 10 Δ in an esotropic patient, I might choose not to intervene further, even if the deviation builds to a large amount on PACT. But if the SPCT is more than 10 Δ, the PACT measurement tells me how much surgery to do, or perhaps if further optical measures may be helpful.

The SPCT tells me IF more intervention is needed, and if so, the PACT tells me WHAT to do.

### Pearl

*High-Powered Spectacles Will Induce a Prism Effect over a Deviated Eye That Requires an Adjustment of the Value Measured with the PACT*

This usually becomes clinically significant if the spherical equivalent is 5 diopters or more in the meridian of the deviation, e.g., horizontal meridian for horizontal strabismus and vertical meridian for vertical strabismus. This is because

**Table 2.1** Addition of two prisms with one placed over each eye [8, 9]

| Left eye prism (labeled value in PD) | Right eye prism (labeled value in PD) | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | **10** | **12** | **14** | **16** | **18** | **20** | **25** | **30** | **35** | **40** | **45** | **50** |
| **10** | 20 | 22 | 24 | 26 | 29 | 31 | 36 | 41 | 47 | 52 | 58 | 63 |
| **12** | 22 | 24 | 26 | 29 | 31 | 33 | 38 | 44 | 49 | 55 | 60 | 66 |
| **14** | 24 | 26 | 29 | 31 | 33 | 35 | 40 | 46 | 52 | 57 | 63 | 69 |
| **16** | 26 | 29 | 31 | 33 | 35 | 37 | 43 | 48 | 54 | 60 | 66 | 72 |
| **18** | 29 | 31 | 33 | 35 | 37 | 39 | 45 | 51 | 57 | 63 | 69 | 75 |
| **20** | 31 | 33 | 35 | 37 | 39 | 42 | 47 | 53 | 59 | 65 | 71 | 78 |
| **25** | 36 | 38 | 40 | 43 | 45 | 47 | 53 | 59 | 66 | 72 | 79 | 86 |
| **30** | 41 | 44 | 46 | 48 | 51 | 53 | 59 | 66 | 73 | 80 | 87 | 94 |
| **35** | 47 | 49 | 52 | 54 | 57 | 59 | 66 | 73 | 80 | 87 | 95 | 103 |
| **40** | 52 | 55 | 57 | 60 | 63 | 65 | 72 | 80 | 87 | 94 | 104 | 113 |
| **45** | 58 | 60 | 63 | 66 | 69 | 71 | 79 | 87 | 95 | 104 | 113 | 123 |
| **50** | 63 | 66 | 69 | 72 | 72 | 78 | 86 | 94 | 103 | 113 | 123 | 133 |

*PD* prism diopters


**Fig. 2.3** Two 40 Δ prisms are stacked with the back surfaces not parallel. The orientation of the back surface influences the effective power of the prism, as evidenced by the pencil's image being displaced differently through the two identical prisms

the deviating eye is looking through a lens that in profile has the configuration of a prism (Fig. 2.4). The adjustment is a function of the spherical equivalent of the spectacle lens and the measured deviation. I keep a conversion table in each of my exam rooms and make this adjustment on a regular basis, especially when planning surgery (Table 2.2).

**Important Point**

Keep in mind that overelevation or overdepression in adduction are not necessarily due to "overaction" or "underaction "of the inferior or superior oblique muscles. Many factors such as inferior restriction (thyroid eye disease, orbital fracture, etc.), dissociated vertical divergence (DVD), pulley heterotopia, orbital anomalies, amongst other causes, can cause the clinical picture we commonly and incorrectly refer to as oblique muscle overaction or underaction. True dysfunction of the oblique muscles should have the appropriate accompanying objective torsional abnormality, and should be discernible on exaggerated forced duction testing.

**Pearl**

*Whenever a Patient Has Unequal Vision and Incomitant Strabismus, Always Consider the Possibility That They May Be Fixing with the Affected Eye*

I have personally seen patients with orbital fractures, thyroid eye disease, Brown syndrome, Duane syndrome, and third, fourth, and sixth cranial nerve palsy, who fixed with the involved eye leading initially to incorrect diagnosis.

**Pearl**

*Many Patients Show Redress and the Deviation Is Never Completely Neutralized During the PACT*

Specifically, when the correct prism is in front of the covered deviating eye, and the cover is shifted to the other eye, they still show movement despite the prism correctly neutralizing the deviation. Consider, for example, a patient with a 30 Δ R esotropia (ET). Prior to the introduction of a prism, when the left eye is covered the right eye abducts to take up fixation. Then, with a 30 Δ prism base out over the deviating right eye, there should be no movement if an occluder is moved back and forth over the two eyes. But in a patient with redress, it is as though the right eye has an established reflex to abduct when the left eye is covered, and still does so despite the neutralizing prism. As a result of it overshooting fixation, it makes a corrective adduction saccade to pick up fixation. In the presence of redress, the examiner has to estimate if the abduction and adduction movements were of equal magnitude, and this can make accurate measurement of the deviation difficult. I have found three things that can minimize or eliminate redress. First, have the patient slowly read optotypes of a size near threshold acuity while doing the PACT, instead of having them just fixate on one letter. If redress is still present, repeat the PACT in aforementioned manner; however instead of shifting the cover directly from one eye to the other, allow a

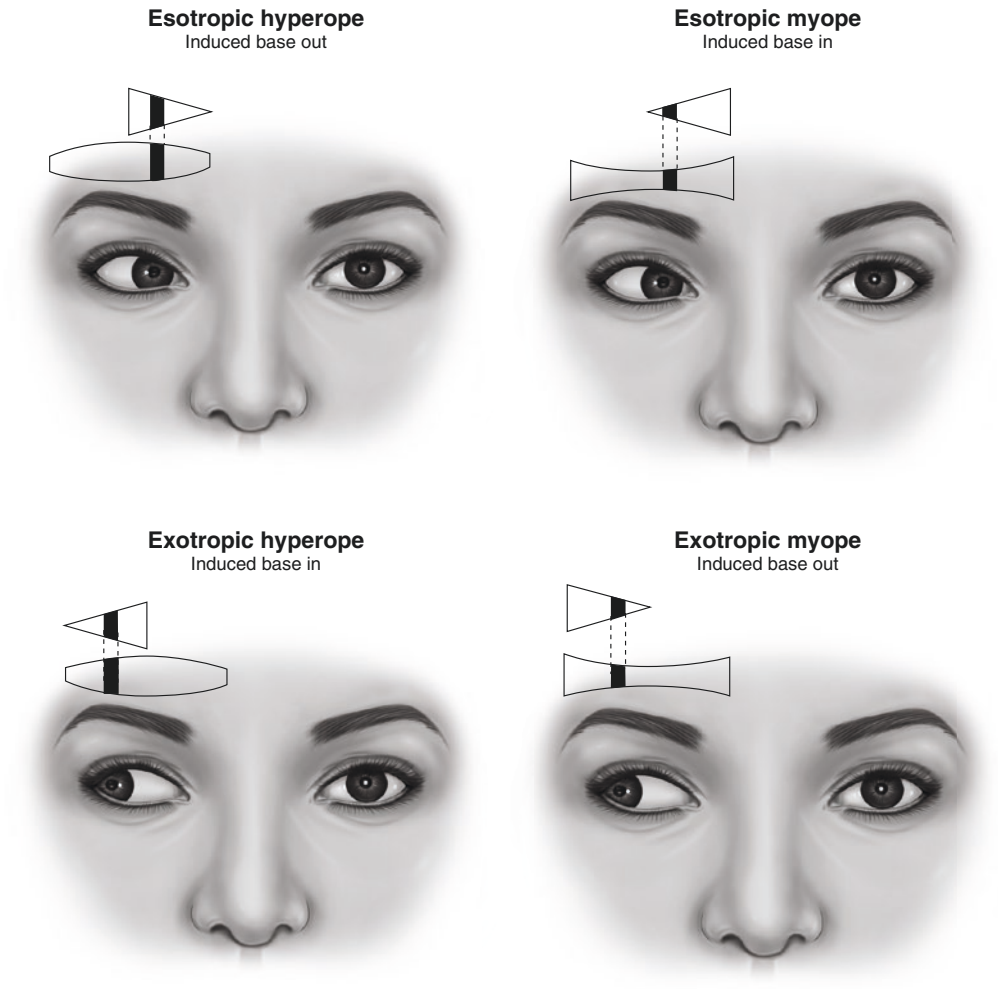



**Fig. 2.4** For each combination of deviation (esotropia or exotropia) and refractive error (hyperopia or myopia) there will either be base-in or base-out induced prism from a high spectacle lens

few moments of binocularity. I do this by lowering the cover to around the mouth area leaving both eyes open for several seconds, and then bring it up to occlude the other eye. Finally, if you ever so slightly overcorrect the deviation with prism, the aforementioned reflex saccade is circumvented and redress may disappear. This, of course, requires you to estimate the tiny amount of overcorrection movement you will observe, and subtract that from your final measurement.

## Question

I have a patient who had surgery for infantile ET and later inferior oblique muscle weakening. Now she has a small R hypotropia (HYPO) when fixing with her left eye, and a rather large L hypertropia (HT) when fixing with her right eye, and the LHT is the cosmetic problem. How can I tell if this is a left DVD versus fixation duress to the left eye driven by inferior restriction in the right eye?

**Table 2.2** Adjustment for spectacle-induced prism: Actual deviation in PD

| Measured deviation in PD | Myopic spectacle power | | | | | | | | | | | | | |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| | **−1** | **−2** | **−3** | **−4** | **−5** | **−6** | **−7** | **−8** | **−9** | **−10** | **−12** | **−15** | **−20** | **−30** |
| **5** | 5 | 5 | 5 | 4 | 4 | 4 | 4 | 4 | 4 | 4 | 4 | 4 | 3 | 3 |
| **10** | 10 | 10 | 9 | 9 | 9 | 9 | 9 | 8 | 8 | 8 | 8 | 7 | 7 | 6 |
| **15** | 15 | 14 | 14 | 14 | 13 | 13 | 13 | 12 | 12 | 12 | 12 | 11 | 10 | 9 |
| **20** | 20 | 19 | 19 | 18 | 18 | 17 | 17 | 17 | 17 | 16 | 15 | 15 | 13 | 11 |
| **25** | 24 | 24 | 23 | 23 | 22 | 22 | 21 | 21 | 20 | 20 | 19 | 16 | 17 | 14 |
| **30** | 29 | 29 | 28 | 27 | 27 | 26 | 26 | 25 | 24 | 24 | 23 | 22 | 20 | 17 |
| **35** | 34 | 33 | 33 | 32 | 31 | 30 | 30 | 30 | 29 | 28 | 27 | 25 | 23 | 20 |
| **40** | 39 | 38 | 37 | 36 | 36 | 35 | 34 | 33 | 33 | 32 | 31 | 29 | 26 | 23 |
| **45** | 44 | 43 | 42 | 41 | 40 | 39 | 38 | 37 | 37 | 35 | 35 | 33 | 30 | 26 |
| **50** | 49 | 48 | 47 | 45 | 44 | 43 | 43 | 42 | 41 | 40 | 38 | 36 | 33 | 29 |
| **60** | 59 | 57 | 56 | 55 | 53 | 52 | 51 | 50 | 49 | 48 | 46 | 44 | 40 | 34 |
| **70** | 68 | 67 | 65 | 64 | 62 | 61 | 60 | 58 | 57 | 56 | 54 | 51 | 46 | 40 |
| | **Hyperopic spectacle power** | | | | | | | | | | | | | |
| | **+1** | **+2** | **+3** | **+4** | **+5** | **+6** | **+7** | **+8** | **+9** | **+10** | **+12** | **+15** | **+20** | **+30** |
| **5** | 5 | 5 | 5 | 6 | 6 | 6 | 6 | 6 | 6 | 7 | 7 | 8 | 10 | 20 |
| **10** | 10 | 11 | 11 | 11 | 11 | 12 | 12 | 13 | 13 | 13 | 14 | 16 | 20 | 40 |
| **15** | 15 | 16 | 16 | 16 | 17 | 18 | 18 | 19 | 19 | 20 | 21 | 24 | 30 | 60 |
| **20** | 21 | 21 | 22 | 22 | 23 | 24 | 24 | 25 | 25 | 27 | 29 | 32 | 40 | 80 |
| **25** | 26 | 26 | 27 | 28 | 29 | 29 | 30 | 31 | 32 | 32 | 36 | 40 | 50 | 100 |
| **30** | 31 | 32 | 32 | 33 | 34 | 35 | 36 | 38 | 39 | 40 | 43 | 48 | 60 | 120 |
| **35** | 36 | 37 | 38 | 39 | 40 | 41 | 42 | 44 | 45 | 47 | 50 | 56 | 70 | 140 |
| **40** | 41 | 42 | 43 | 44 | 46 | 47 | 48 | 50 | 52 | 53 | 57 | 64 | 80 | 160 |
| **45** | 46 | 47 | 49 | 50 | 51 | 53 | 55 | 56 | 58 | 60 | 64 | 72 | 90 | 180 |
| **50** | 51 | 53 | 54 | 56 | 57 | 59 | 61 | 63 | 65 | 67 | 71 | 80 | 100 | 200 |
| **60** | 62 | 63 | 65 | 67 | 69 | 71 | 73 | 75 | 77 | 80 | 87 | 96 | 120 | 240 |
| **70** | 72 | 74 | 76 | 78 | 80 | 82 | 85 | 88 | 90 | 93 | 100 | 112 | 140 | 260 |

*PD* prism diopters


### Reply

This can be sorted out by first estimating or measuring the size of the R HYPO. Then put a prism of a few prism diopters more than the estimated amount base up over the right eye under a cover. When the cover is switched to the left eye there should be a tiny downward movement of the right eye, or at least no movement. Then, switch the cover back to the right eye. If the L HYPO was due to DVD, the left eye will still be up under cover and will infraduct when the cover is shifted. But if the large LHT was all due to fixation duress, the overcorrecting prism right eye will eliminate the fixation duress and the left eye will not elevate under the cover.

### Important Point

Be clear and consistent in your notation of dissociated deviations. It is insufficient to just write "DVD" as a descriptor. There are four components that each need to be identified. (1) Is the deviation right eye, left eye, or both eyes? (2) Is it manifest or latent? If manifest it is a tropia and if latent it is a phoria (nothing new here). (3) If manifest, is the deviation intermittent or constant? (4) Is the deviation dissociated, e.g., not following Hering's law with respect to vertical or horizontal movement? A dissociated deviation of the right eye that is only latent should be called either a dissociated R hyperphoria or alternatively a latent R DVD. A dissociated deviation of the R eye that is intermittently manifest should be called an intermittent dissociated RHT or, alternatively, an intermittent manifest R DVD. I feel using the term "occlusion hyperphoria" to describe a latent DVD, as is sometimes done, is confusing and incomplete. All latent deviations are phorias by definition, and this term does not identify the deviation as being dissociated. The term occlusion hyperphoria might also correctly be applied to a latent non-dissociated vertical phoria and, therefore, is not specific for dissociated deviations.

### Pearl

*Forced Ductions May Be a "One-Way Test"*

If forced ductions done in the office are normal, that must mean there is no restriction. But abnormal forced ductions may not be meaningful if the patient is resisting or squeezing. I have found several ways to maximize patient cooperation. First I instill several anesthetic drops. Then I wet a Q-tip with the anesthetic and tell the patient I am going to gently apply more anesthetic. I then apply the moistened Q-tip to the conjunctiva near the limbus on the side I suspect restriction; for example testing for a temporal restriction to adduction in the right eye, I put the Q-tip at the 9 o'clock. Using the Q-tip, I push the eye into adduction. Sometimes this is all one need to feel a restriction. Often more information is needed, in which case I say I am going to do the same maneuver, but with a delicate instrument. I then grasp the same area with my forceps and do the test in the usual manner. I find that by gradually increasing the invasiveness of the test in this manner, most patients can comply.

### Pearl

*Active Force Generation Is a Simple Test That Is Useful and Underutilized*

It often surprises me how often I am referred a patient in whom the prior ophthalmologist may have done forced duction testing but did not do active force generation. It was not until I started doing the test (and modified it slightly) that I realized how easy it is to interpret. If you are not doing it regularly, I encourage you to try. Classically, the test is done by grasping the anesthetized conjunctiva with forceps near the limbus and having the patient attempt a saccade by the muscle being tested. A problem with this method is that if the muscle generates a strong contraction, the sudden pulling against the forceps can result in a tearing of the conjunctiva. I prefer to do this a bit differently. I have the patient first attempt to look in the direction of the muscle

being tested; for example, for testing the right medial rectus muscle the patient attempts to look left. I then grasp the anesthetized conjunctiva at the 9 o'clock limbus and gently pull against the isometric contraction that is generated by the right medial rectus muscle (Fig. 2.5). The test is easy to perform, but moreover it is surprisingly easy to interpret.

## Basic Information

*"Marlowe Occlusion" or a "Patch Test"*

In 1922 Marlowe described the benefits of doing a prolonged period of monocular occlusion for bringing out the full latent deviation in patients with small manifest deviations. Today most people use 45–60 min of occlusion for this purpose [7]. When doing a patch test, it is important that you do the PACT after the period of occlusion, without allowing the patient to be binocular. Even a moment of binocular viewing can negate the effect of the patch. I do this by either having the patient close both eyes while I remove the patch and then hold the paddle occluder over one eye before he opens his eyes. Alternatively, I may just hold the paddle occluder in front of either eye while I remove the patch. In my practice the most common use for this test is in intermittent exotropia (XT) where I use it to uncover a masked or latent larger deviation at near or distance (see Chap. 6). I also use it if symptoms suggest a larger deviation (most commonly vertical) than the patient manifests.

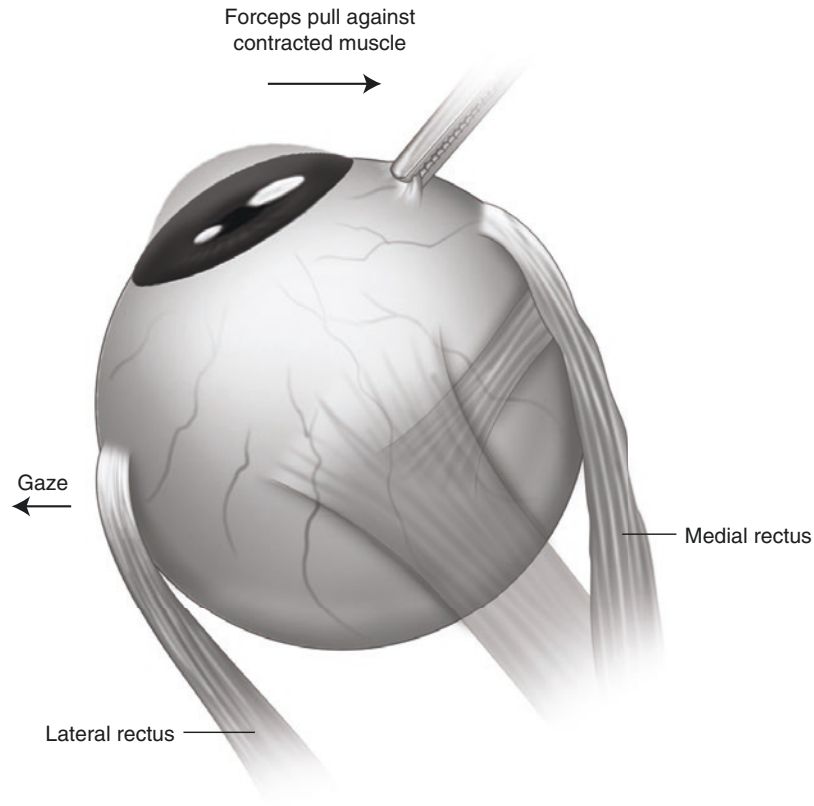
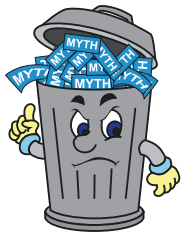



**Fig. 2.5** Preferred technique for active force generation testing, in this case to determine if there is lateral rectus muscle weakness in the presence of a limitation of abduction. The patient is first instructed to look in the direction of the lateral rectus muscle, and then the examiner grasps the nasal limbus with forceps. By pulling against the contracting lateral rectus muscle the strength of the active force can be felt




**Myth**

Marlowe actually described 1 week of monocular occlusion for his prolonged occlusion test.

**Fact**

Technically, Marlowe did recommend a week of occlusion. However, he carried this out by having the patient wear spectacles that occluded one eye for a week's time. To prevent them from experiencing moments of binocularity during this time, he instructed them to keep their eyes closed when they woke up in the morning, until they put on the occluding spectacles. It is hard to imagine this worked. I don't really know I am awake until I open my eyes.

**Pearl**

Sometimes a patch test can bring out what is called a "false hyper." This is when a small vertical deviation, always in the form of a hyperphoria of the occluded eye, is found only after prolonged occlusion. It is thought that this is a residual tonicity to the superior rectus muscle, resulting from the prolonged Bell's phenomenon that occurs during occlusion. Usually these are small, in the range of several. If you uncover a small hyper of the occluded eye when testing a horizontal deviation, and if its presence does not fit with the motility pattern, e.g., oblique dysfunction, other possible etiologies for a vertical deviation, you might repeat the patch test with the other eye occluded. If you do not observe the same hyper, or if you find a hyper of the other eye (the one occluded on the repeat test), you can be confident that you are dealing with a false hyper that can be ignored. If you are doing the patch test for the purpose of uncovering a hyper, or getting one to build, it can be harder to interpret. The best advice is to repeat the test with the other eye occluded, and average the findings of the two tests.

**Advanced Information**

Dynamic retinoscopy is one of the most underutilized tests in pediatric ophthalmology [8, 9]. Dynamic retinoscopy is not a technique for determining refractive error; it is not an objective dry refraction. Instead, it is a test of accommodation and has some really useful applications. The test is performed by first having the patient fixate in the distance while the examiner retinoscopes the eye being tested. Then the subject shifts fixation to an accommodative target that is affixed to the retinoscope (Figs. 2.6 and 2.7). If accommodation is normal, there should be a brisk change in the retinoscopic reflex to that of neutrality, regardless of the refractive error. If you observe that, it means that the subject is fully accommodating on the fixation target. Of course in an infant, you may not be able to direct their fixation to change from distance to near. But as long as you can get them fixing on the near target you will obtain useful information about their accommodation ability. This test can tell which nonverbal children may be having visual blur at near due to decreased accommodation and would benefit from a bifocal. This is a common finding in children with


**Fig. 2.6** For dynamic retinoscopy a fixation target should be affixed to the retinoscope


**Fig. 2.7** For dynamic retinoscopy the examiner is at about arm's length from the patient

cerebral palsy or Down syndrome. But there is another circumstance in which I find dynamic retinoscopy valuable. Consider the 6-month-old child who is found on examination to be orthophoric, but has a high hyperopic refractive error of, let's say +5.00 oculus utero (OU) both eyes. How closely should this child be followed, and what is her risk for developing ET? It appears, according to an unpublished study by Deniz Somer and coworkers, that if dynamic retinoscopy shows that the patient is fully accommodating at near and is not esotropic, there is very little likelihood they will subsequently develop ET. But if dynamic retinoscopy shows decreased accommodation, there is a high likelihood that they will subsequently develop an accommodative ET. It is as though the child with the former findings has a good fusional mechanism that allows him to compensate for accommodative convergence. The child with the latter findings is not initially esotropic simply because they are not accommodating fully. When they get a bit older and start accommodating more fully, they start going esotropic. Such a child should be followed more closely and advised to come in for an examination at the first sign of strabismus.

Dynamic retinoscopy can predict which orthophoric hyperopic infants will later develop esotropia.

## Basic Information

*Torsion Should Be Measured Objectively and Subjectively, and the Two Measurements Reflect Different Things*

Classically, subjective torsion is measured with the double Maddox rod test using one red and one white lens. A study that I found convincing reported that there is a color dissociation artifact if the test is done in this classical manner. Patients may subjectively localize the torsion to the eye with the red lens, even if the other eye is the one with the torsional problem [10]. The authors recommend using two red lenses for the test. Subsequent to this publication, I have modified my technique according to their recommendations. In spite of that, I find the double Maddox rod test is mainly useful for telling the total amount of subjective torsion in the two eyes, but still can be inaccurate as to localizing which eye is the one that is torted. Objective torsion is assessed by looking at the fundus. Normally the fovea lies between an imaginary line level with

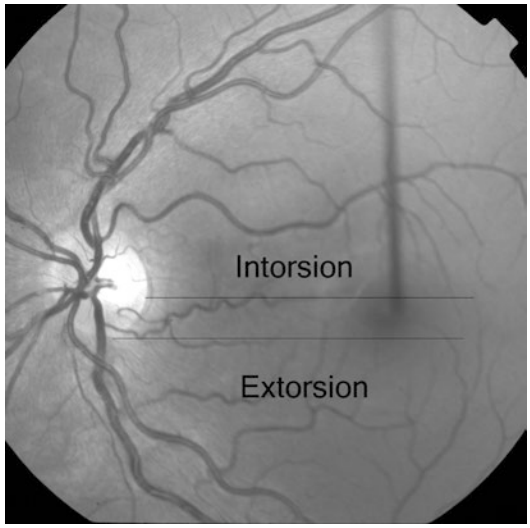



**Fig. 2.8** The fovea lies between an imaginary line level with the bottom of the optic nerve, and a second line at the junction of the lower one-third and upper two-thirds of the disk. Lying above the upper line indicates intorsion and below the lower line indicates extorsion

the bottom of the optic nerve and a second line at the junction of the lower one-third and upper two-thirds of the disk (Fig. 2.8). If it is below the lower line the eye is extorted, and if it is above the upper line the eye is intorted. You do need to keep in mind that the image seen with the indirect ophthalmoscope is inverted. Objective torsion is more useful for determining which eye or if both eyes are actually torted. However, patients with sensory adaptations to strabismus, like infantile esotropes, may be asymptomatic despite marked objective torsion. My thoughts about the relative role and behavior of objective and subjective torsion have been previously described in detail [11].

### Important Point

The double Maddox rod test for subjective torsion is better at telling the total subjective torsional misalignment between the two eyes, than it is for localizing which eye is actually torted. Fundus torsion (objective torsion) is better for telling how much torsion is in each eye, but is less useful for determining if the torsion needs to be treated.

## Sensory Testing

> The man who is swimming against the stream knows the strength of it.
>
> —Woodrow Wilson

### Basic Information

*In My Mind I Divide Sensory Tests into Two Categories*

There are some tests, the results of which will greatly alter or influence your treatment, and there are others that only provide information about how a patient is doing (compared to before, or compared to other patients, etc.). I do feel that many of the tests we do routinely are in the latter category. That is not to say they are useless or should not be performed. But you should be aware of the importance of the information gleaned from them.

### Basic Information

*The Worth 4-dot Test Is Primarily Informational*

This test can be useful in determining in a patient with small-angle strabismus, if their suppression scotoma is getting smaller or breaking up during treatment, and if binocularity is improving or worsening. In general, although the results of this test provide some interesting information, it rarely influences my treatment of a given patient.

### Basic Information

*Stereopsis Testing Is Also Primarily (But Not Exclusively) Informational*

That is not to say it is unimportant. Some patients specifically want to know about their three-dimensional vision. An improvement in stereopsis after treatment or surgery certainly speaks to a significant improvement

in binocular function, and it is good to know if this occurred. Random dot stereo tests are more sensitive for detecting true stereopsis than the ones with visible circles and animals, which have monocular clues at higher angles of disparity; they may falsely imply that stereopsis is present when it is not. There are some circumstances in which the results of stereopsis testing may influence treatment. If one is certain that stereopsis deteriorated in an amblyope as treatment is being tapered or after it has ceased, one should consider this to be a sign of recurrence, and you should consider upping or resuming the treatment. Also, deterioration of stereopsis in a patient with intermittent XT, particularly at distance, is a strong sign of deteriorating control over the XT. This may be a sign for further intervention.

## Advanced Information

*Testing for Anomalous Retinal Correspondence (ARC) or Suppression*

Although it is a bit of a simplification, if a patient has a manifest strabismus and is not diplopic, they are either suppressing or corresponding anomalously, both of which are compensation mechanisms to avoid diplopia. However, both ARC and suppression are not "all or nothing." A patient may be using different compensation mechanisms in different circumstances, e.g., eyes open in normal illumination vs. with colored glasses on or after images with eyes closed. The one simple test that tells what a patient is doing in everyday seeing circumstances is the Bagolini lens test. If a patient with a tropia sees the major part of both arms of the "X" with that test (simultaneous perception), and both arms are running through the fixation light (even if parts of one line are missing), the patient is compensating by corresponding anomalously. If they do not see both arms of the "X" they are suppressing. Test for ARC that is more dissociating by using after images and colored lenses may be useful for investigating the nature or depth of ARC, but do not necessarily address what a patient is doing day in and day out.

## Pearl

*Use Bagolini Lenses to Demonstrate a Child's Binocularity to Parents*

Most patients, if approximately aligned and not significantly amblyopic, will have simultaneous perception with the Bagolini lens test [12]. If a parent is distressed that their child "doesn't use her eyes together" either because they had been told that by a prior eye doctor or due to her having deficient stereopsis, I find it helpful to have the child describe their simultaneous perception while undergoing the Bagolini lens test. Then I have the parent perform the test and observe the two lines. I then have them close either eye to see how true monocular vision would be perceived. They are typically relieved and pleased to see that their child in fact "sees with both eyes together."

## Question

What is the clinical importance of ARC being harmonious or unharmonious?


## Reply

If ARC is present, there is a retinal point (or area) in the deviating eye that localizes images in space as being in the same location as the object on the fovea of the fixing eye—at least at the time and in the visual environment the testing for ARC was performed. ARC is said to be harmonious if distance from the fovea to retinal point of correspondence in the deviating eye equals the angle of misalignment. For example, if a patient with ARC has a 15 Δ RET and the anomalous retinal point of correspondence is 15 Δ nasal to the fovea in the deviating eye, ARC is said to be

harmonious. It is called that because the adaptation is *in harmony;* it is perfectly constituted to prevent diplopia. On the other hand, if the anomalous retinal point of correspondence is 35 Δ nasal to the fovea in the deviating eye, the ARC would be unharmonious. The ARC alone is not constituted to prevent diplopia. Either the patient would experience diplopia or there would need to be substantial suppression of the deviating eye. In many cases unharmonious is an artifact of testing. As stated earlier, ARC and suppression are not "all-or-nothing" phenomenon. A patient may have unharmonious ARC with a dissociating means of testing, but have harmonious ARC with a test that more closely mimics everyday seeing circumstances. In other cases, ARC may be unharmonious due to a change in the angle of strabismus. If a patient has harmonious ARC, and there is a subsequent change in the angle of strabismus, they may appear to have unharmonious ARC if they are tested before their sensory systems adapt to the new angle of strabismus. This is common immediately after strabismus surgery.

See Chap. 21, Case 21.21, on **page 327** for a representative example of these principles.

## Basic Information

*All Patients with Constant Diplopia Should Undergo Prism Testing to See if They Are Diplopia Free When the Deviation Is Offset with Prism*

If prism testing eliminates their diplopia, you know they will be symptom free if you get them aligned. If they cannot fuse with prisms, two further tests are indicated. All such patients should have testing for torsion with the double Maddox rod test. Subjective torsion can occur in circumstances where it is not expected, and significant torsion can be an obstacle to fusion. In a prior publication I reported this to occur in patients without obvious oblique muscle involvement [13]. They included patients with prior scleral buckling, prior penetrating keratoplasty, severe corneal scarring, monocular aphakia, long-standing strabismus, or prior vertical offsets of the horizontal rectus muscles. If subjective torsion is present, a synoptophore evaluation can tell if the patient will be able to fuse if the torsion is corrected. If there is no fusion when the torsion is offset on the synoptophore, there is a central disruption of fusion. See Chap. 21, Case 21.7, on **page 315** for a representative example of these principles. If subjective torsion is not present, you should also test the patient for aniseikonia. This can occur in the absence of anisometropia and can be an obstacle to fusion. The most common cause of this is an epiretinal membrane [14]. In some patients who cannot fuse when the deviation is offset with prisms, the synoptophore or prisms of a different value can be used to find a suppression scotoma that is eccentric to the fovea in the deviating eye. In my experience, this is infrequently successful. Nevertheless, it is worth attempting as it may offer a way to help rid some patients of diplopia even if they are not going to fuse. This is accomplished by putting the second image in the scotoma using prisms or surgery. If neither aniseikonia nor subjective torsion are present and a suppression scotoma cannot be identified, the patient probably has a central disruption of fusion or horror fusionis. Strabismus surgery may not be successful in eliminating diplopia.

See Chap. 21, Case 21.21, on **page 327** for a representative example of how putting the second image in a suppression scotoma can be successful.

## Important Point

*All Non-diplopic Adult Patients Contemplating Surgery for a Constant Tropia Should Undergo Prism Testing Preoperatively to Help Predict Intractable Postoperative Diplopia*

I have heard many speakers (ophthalmologists and orthoptists) say this, but few have backed up their recommendations with any data. I do the test as follows. In the exam room, with normal room illumination, I offset the patient's deviation

with prisms (no colored filters and of course with spectacles on). Next I ask if they see double. I repeat this just slightly overcorrecting and also just slightly undercorrecting the deviation. If the patient does not see double with this prism testing, in my experience they never see double if aligned surgically. However, if they are diplopic with prism testing, that does not mean they will be diplopic if aligned. There is only a very small chance of diplopia [15]. If the patient is concerned about postoperative diplopia, I then offer them a trial of a week or so with a Fresnel prism to offset the deviation. In many such patients, the diplopia subsides in a short time. If it does, the patient has essentially no risk of intractable diplopia if surgically aligned, in my experience. In some patients the diplopia will persist with the Fresnel prism. Even in those patients, the vast majority will not be diplopic if surgically aligned. In a series of 424 adult patients I tested this way, 143 (34%) were diplopic with prism testing. But only 3 (0.8%) had intractable diplopia after surgery [15]. This testing had a high sensitivity (100%) for detecting who will not be diplopic after surgery, but a very low positive predictive value (2%). In other words, this is in effect a "one-way" test. If the test says that a patient will not be diplopic, they almost certainly will not be. But if it suggests that they might be diplopic, there is still a very low chance of diplopia. Consider the following patient:

**Case 2.1** I examined a 36-year-old woman who had a consecutive XT of 30 Δ that was comitant. Detailed records were not available, but her parents recalled that at about 3 years of age she underwent bilateral surgery for an ET. She was not diplopic in free space, but reported diplopia with either 25 Δ, 30 Δ, or 35 Δ base in. With each of the three prisms she localized the images as being far apart and uncrossed, indicating she had ARC. Because she was very interested in restorative strabismus surgery, I had her wear a 30 Δ Fresnel prism base in I over her nondominant eye. After wearing the prism for 2 weeks she was still diplopic. I advised her there was a small, but definite chance of her experiencing postoperative diplopia, but that this could be managed with a fogging or occluding contact lens. This would still allow her to have the aesthetic benefit from surgery. She continued to wear the Fresnel prism for the subsequent 5 weeks, until her surgery could be performed. On the day of her surgery, she was still diplopic with the Fresnel prism in place. However, immediately upon awakening from anesthesia she was diplopia free. She remained free of diplopia and maintained good alignment during the subsequent 6 years I followed her.

Why prism testing does not always predict the postoperative situation is unclear.

Of course, if you don't align the patient they might be diplopic; for example an overcorrected exotrope will be diplopic. But this in not intractable diplopia. You just need to get them aligned.

## Advanced Information

If a patient has torsional diplopia that cannot be eliminated with prisms, you may want to consider a synoptophore evaluation to see if they can fuse with the torsion offset. In my mind, this is the one main use for the synoptophore. Some patients with torsional diplopia, especially after closed head trauma, may also have a central disruption of fusion. It is disappointing to you and the patient when you get them well aligned with surgery, only to find that the diplopia persists. Prisms, of course, will not correct for torsion. But the synoptophore can, and I find it useful in this setting.

## Pearl

The discussion of sensory tests above summarizes my daily use and understanding of this subject. However, I have been blessed to have partnered with a number of wonderful certified orthoptists over my entire career. My patients and I have been

well served by these relationships, and I regularly defer to their judgement when it comes to the sensory aspects of strabismus. If you have the opportunity to have one or more certified orthoptists on your team, you will be doing yourself and your patients a great service by seizing that opportunity.

If at all possible, partner with a certified orthoptist.

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# Refraction

# 3

## Basic Information

*It Can Be Useful to Obtain a Cycloplegic Refraction on All Patients with Strabismus—Even Adults*

I am aware that many colleagues do cycloplegic refractions on all children, but in adults they only perform dry (manifest) refractions. There are several reasons for this. In patients with early nuclear sclerosis, dilation may compromise visual acuity testing. Also, in hyperopic patients it can be hard to predict how much latent hyperopia is present, and how much of the plus they will tolerate. But I find that many adult patients with strabismus (usually esotropes but not exclusively so) do "funny" things with respect to accommodation; they may not relax it during a dry refraction in the same way as non-strabismic patients do. I recall one patient who was referred to me for strabismus surgery in her late 40s. She had a history of accommodative esotropia (ET) that had been controlled for many years, but over the prior 10 years her eye began to increasingly cross. She had undergone five refractions over the prior 10 years, having seen five different comprehensive ophthalmologists, all of whom performed dry refractions; they did dilate her for fundoscopy. Although my dry refraction was close to what she was currently wearing in her spectacles, my cycloplegic refraction revealed three additional diopters of hyperopia. She was incrementally given more hyperopic correction which eliminated her ET and brought great relief from the asthenopia she was experiencing with visual tasks.

## Basic Information

Cyclopentolate takes a full 40 min to achieve maximum cycloplegic effect. I have observed a trend among colleagues to perform cycloplegic refractions only 20–30 min after the instillation of cyclopentolate. Often the child's pupils may appear fully dilated and it is assumed that the drops have peaked. However, the reality is that full mydriasis often precedes the full cycloplegic effect of cyclopentolate. Perhaps some blue-eyed patients may experience full cycloplegia in less than 40 min; how-


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ever one cannot be certain. So for the patient in whom it is important to uncover the full amount of hyperopia, e.g., a partly accommodative esotrope who is possibly facing surgery, waiting the extra 10–20 min is in the patient's best interest.

## Advanced Information

Strabismic patients who alternate between spectacles and contact lenses may need to undergo two completely separate refractions. This is particularly true for the astigmatic patient who reports better strabismic control with spectacles than their contact lenses. After I do my cycloplegic refraction for spectacles, I have the patient reinsert their contact lenses. I then retinoscope them over their contact lenses. It is surprising how frequently I find that there is a substantial uncorrected cylinder, often at a different axis than their de novo astigmatism axis. This usually reflects an error in how the toric contact lenses were made, in how they are sitting on the patient's eyes, or in the conversion from the spectacle prescription to the toric contact lens prescription. Doing two refractions is admittedly time consuming, but I have yet to find a better way to sort out these problems.

## Pearl

*If a Patient Describes More Strabismus with Contact Lenses than Glasses (or Vice Versa), You Should Perform Two Refractions as Described Above*

The most common cause of this difference is that either the patient has inadvertently mixed up their right and left soft contact lenses or there is uncorrected cylinder with the contact lenses as described above. This can only be discovered by doing a refraction over the contact lenses.

## Pearl

*I Frequently Do "Over-refractions"*

In young children, or older patients with high refractive errors, I often objectively refract by retinoscoping over their existing spectacles, and subjectively refract by adding lenses to their spectacles in a Halberg clip (Fig. 3.1). If the change is spherical, simple addition or subtraction gives the resultant prescription. The same is true if the change is in the cylinder power, and if the axis stays the same. However, if the over-refraction shows a change in both cylinder power and axis, the math is not as simple. There are, however, several ways to get the resultant prescription. Many of the electronic medical records programs have a built-in function for calculating an over-refraction. Alternatively, the patient's spectacles with the over-refraction lenses held sandwiched over them can be put in a lensometer and read, which will give the resultant prescription. The advantages in performing an over-refraction are many. Doing so eliminates possible vertex error, which can be significant if the refractive error is high. I find that many young children do not sit well behind a phoropter. They tend to tilt their head and/or drift away. Many trial frames


**Fig. 3.1** Over-refraction is carried out with the use of loose lenses, a trial lens clip, and the patient's spectacles

are uncomfortable and don't stay in place well. None of these factors are issues if the patient is wearing their own glasses.

Consider doing over-refractions in young children and all patients with high refractive errors.

## Important Point

*Oblique Muscle Surgery Will Alter the Axis of Astigmatism, and the Change May Be Permanent*

I stumbled on this observation after I performed bilateral superior oblique tucks in a boy with bilateral high astigmatism [1]. On his 6-week post-op visit he reported that his vision was blurred. He found he could see clearly out of his right eye if he rotated his spectacles one way, and similarly he could see clearly out of the left eye if he rotated the lenses in the opposite direction. Oblique muscle surgery causes a torsional rotation with will alter the orientation of the astigmatism axis. Whenever you do oblique muscle surgery in an eye with significant astigmatism, consider repeating a refraction about 6 weeks after surgery. This is particularly important in a patient with amblyopia, as they may not complain of blur in their nondominant eye, which of course is the eye that is more likely to have the greater refractive error.

## Pearl

*Consider Asking, "Which Is Worse?"*

All ophthalmologists have experienced the really negative patient. You are refracting him and ask, 'Which is better, one or two?" He replies, "Neither." You persist, "Pick the one that is better." He responds, "None!" Art Jampolsky taught me to then ask, "Which is worse?" And *voilà,* he is able to choose. Although this pearl may seem humorous, it actually is quite useful and really works. Some people with a negative world view are always ready to point out that which is worse.

## Important Point

Some children with high hyperopia may not fully relax their accommodation during their first cycloplegic refraction, even if they appear to have achieved adequate cycloplegia. Consequently, if there is a residual ET when they return wearing their first pair of glasses, a repeat refraction a month or so later may uncover as much as several diopters of additional hyperopia. I find that this is more likely to occur when the initial refraction revealed more than about 5 diopters of hyperopia. I assume that because the child had been constantly accommodating a large amount prior to getting glasses, there was more tone or spasticity of the ciliary muscle which was not fully overcome with the cycloplegic agent. After the child is put in spectacles that correct most of the hyperopia, the ciliary muscle relaxes can be more effectively pharmacologically put at rest.

## Advanced Information

*Anticipate and Preemptively Answer Two Questions*

When I recommend glasses for young child, I say to parents that there are two questions they probably are thinking of, or if not probably will shortly. The first is

"How will I ever get him to wear glasses?" I go on to say that in most cases children will love their glasses and not want to take them off. This is of course more likely to be the case if the refractive error is high. But I tell parents that if he has difficulty accepting glasses, there are things we can do to help. "The second question is 'How long will she need to wear glasses?'" If I have learned one thing in my 40+ years of practice, it is to never make predictions in this regard. I have seen young children who only had several diopters of hyperopia who never outgrew the need for glasses. Conversely, I have seen children who started with 6 or more diopters of hyperopia that outgrew their hyperopia completely, albeit this happens very infrequently. Finally, I have seen about a dozen children who initially had about 5 diopters of hyperopia as infants, and who ended up with over 9 diopters of hyperopia as teenagers. So I tell parents that on average, children get more hyperopic until about age 7 years, at which time they start losing hyperopia until the end of growth. The average toddler is several diopters hyperopic, and if all works well they lose this by about age 16–17. So I indicate in advance that the second pair of glasses for a toddler is typically stronger than the first; I am trying to head off disappointment stated by the parents as "she's getting worse." I relate these average numbers to the child's refractive error; for example a 5-diopter hyperope will be on average left with about 3 diopters of plus by the end of growth. I stress that averages are just that—an average—and not what is "expected." It never hurts to throw in "of course your child is above average," which always evokes a smile and lightens the discussion.

## Pearl

If a child is rejecting wearing glasses, I put them on a short course of atropine. I recommend atropine 1% daily for a week, which will take another week to wear off. During the time on atropine the child almost invariably wears the glasses, and typically continues to do so after the medication wears off. If this form of behavior modification does not result in good compliance with spectacle wear, I will repeat the refraction.

## Basic Information

*Bifocal Contact Lenses Often Are Not Effective for Accommodative Esotropes with a High Accommodative Convergence/Accommodation (AC/A) Ratio*

In a prospective masked study coworkers and I compared alignment and fusion in bifocal spectacles to that with either a simultaneous vision bifocal contact lens or an aspheric multifocal contact lens [2]. All of the patients showed poorer alignment at near in the contact lens as compared to spectacles, and many showed poorer fusion. It was as though the patient did not know which near image to focus on.

## Basic Information

*Many Adults with a History of Accommodative ET May Go Many Years with Good Alignment, Only to Have the ET Decompensate As They Approach Presbyopia*

It is thought that this is due to increased convergence tonus due to increased accommodative demand [3]. Many of these patients have decreased accommodative ability. In theory, prescribing a bifocal for them early in the development of presbyopia, rather than

allowing them to put off getting bifocals as long as possible (as many patients want to do), may prevent the decompensation of their alignment. I have the impression that this is more likely to occur in patients who never developed bifoveal fusion and monofixators, at best. In my experience, it is unusual for this to occur in patients who developed bifoveal fusion in childhood.

> But there comes a day when your eyes are all right but your arm isn't long enough to hold the telephone book where you can read it.
> And your friends get jocular, so you go to the oculist,
> And of all your friends, he is the joculist,
> So over the facetiousness let us skim.
> Only noting he has been waiting for you ever since you said
> Good evening to his grandfather clock under the impression it was him.
> And you look at his chart … and he says one set of glasses won't do,
> You need two.
> —Ogden Nash, *Peekaboo, I Almost See You*

## Advanced Information

*Monovision May Be Detrimental to Patients with Strabismus* [4]

Monovision occurs when one eye is optically in focus and the other for near. It occurs either de novo due to anisometropia or from an intentional strategy to treat presbyopia either with spectacles, contact lenses, refractive surgery, or choice of intra-ocular lens power. About three-quarters of patients fit with contact lenses that create monovision to treat presbyopia wear them successfully, despite there being well-documented deleterious effects of monovision on binocular function. However, patients with strabismus may have difficulty with monovision usually for one of the three reasons.

1. Unequal vision input from the two eyes is one of the most destabilizing conditions for patients with shaky fusion, e.g., intermittent exotropia, or X(T) [5]. With monovision, at least one eye is always out of focus, which disrupts fusion in these patients.
2. Patients with paretic strabismus may have a secondary (larger) deviation when they fixate with the paretic eye. West and I found that almost half of our strabismic patients who had difficulty with monovision had fourth cranial nerve palsy [4]. We theorized that they were accustomed to fixing with the non-paretic eye and had developed vertical vergence amplitudes sufficient to control the deviation. However once in monovision they would fixate with the paretic eye at either distance or near, and they were not accustomed to controlling this larger secondary deviation so their control broke down.
3. Some strabismic patients with a strong fixation preference may suppress their nondominant eye while fixing with their dominant eye. However, the suppression scotoma does not transfer to the dominant eye if they fix with the nondominant eye. With monovision they will be diplopic. (See Chap. 9, for more about "fixation switch diplopia.") Monovision may be detrimental to patients with strabismus.

Monovision may be detrimental to patients with strabismus.

West and I found that slightly less than half the time, putting patients in proper optical correction, e.g., eliminating the monovision situation, was successful in restoring the prior satisfactory alignment. West coined the term "optical rescue" for this treatment modality. It is worth trying. Consider this patient:

This woman had a history of X(T) which was well controlled until age 45 when she began having symptoms of asthenopia and diplopia. Her refractive error was −2.50 sph (OU) *oculus utro,* both eyes, and around this time she

had been fit with a contact lens for her dominant left eye only, because she wanted monovision for her early presbyopia. At 46 years of age she underwent lateral rectus recessions OU which was initially successful. However, by age 47 her X(T) returned to its original angle and her symptoms recurred. She then underwent bilateral medial rectus resections which resulted in initial improvement; however, within 6 months the effect had worn off. A third strabismus operation was advised at which time she came to me for a second opinion. For me she had a poorly controlled X(T) of 25 Δ at distance and 30 Δ at near (with her monovision contact lens). I advised her against monovision and prescribed bifocals as well as bilateral contact lenses over which single-vision readers could be worn; I advised against further surgery until optical rescue had been tried. Six months later she had an exophoria of 10 Δ at distance and near, and she was asymptomatic. Her motility remained stable for the subsequent 14 years I followed her.

## Important Point

*Balance Refractions to Eliminate Anisometropia*

Consider an exotropic patient with low hyperopia in her fixing R eye and perhaps more hyperopia and hyperopic astigmatism in her deviating eye. If you decide to cut the plus in her fixing eye on the grounds that you do not want to push plus in an exotrope, you need to cut the plus an exactly equal amount in her deviating eye. If you (incorrectly) only cut the plus in her fixing eye, she will be fogged in her left eye when fixing with the right. This is because accommodation is thought to be symmetric and driven by the fixing eye. Consider this patient:

I examined a young boy with a poorly controlled intermittent left XT of 25 prism diopters (Δ) and a refractive error of OD +0.50 sph and OS −2.50 + 1.00 × 180. I prescribed that refraction exactly although I realize one can make a case for cutting the plus a bit. The optical shop mixed up the prescription and reversed the right and left eye lenses. They dispensed OD −2.50 + 1.00 × 180 and OS +0.50sph. At first glance one might think that his angle and control would be better with the fixing eye being over-minused by several diopters. In fact, he returned having poorer control; his XT was now constant, and his angle increased to 30 Δ. The error was spotted when we neutralized his glasses, and the optical shop corrected it. He then returned wearing his prescribed spectacles. His deviation had decreased to 10 Δ and his XT converted to an exophoria. He remained stable for 8 years.

## Advanced Information

*When Patients Specifically Complain of Problems with Near Vision, There Are Ten Common Causes to Consider* [6]

Most often these relate to either the increased accommodative demands at near or the fact that reading usually occurs in downgaze, and there are some clinical situations for which downgaze poses some unique optical problems. You should consider the following issues, many of which were discussed above:

Usually problems are due to increased need for accommodation or the fact that reading involves downgaze.

1. There may be increased accommodative demands in myopes switching to contact lenses.
2. Patients who need a bifocal because of a high AC/A ratio ET may be symptomatic with multifocal contact lenses.
3. Adults with a history of accommodative ET may decompensate as they approach presby-

opia because of decreased accommodation and may need to start using a bifocal at an earlier age than expected.
4. Many adults with a history of accommodative ET continue to need cycloplegic refractions. They do not relax accommodation in the usual way, and a manifest refraction may leave substantial uncorrected hyperopia.
5. Adult patients with a strong fixation preference, incomitant strabismus, or shaky fusion may have difficulty with monovision.
6. A change in refractive error, refractive management, or institution of monovision may cause symptoms of fixation switch diplopia/asthenopia (see Chap. 9 for further discussion).
7. Patients with monocular aphakia or pseudophakia will need to have a bifocal lens prescribed only for the aphakic eye if they have any binocularity. Either not prescribing a bifocal for the aphakic or pseudophakic eye or prescribing a bifocal for both eyes will result in one eye being out of focus for near. This can cause symptoms with near tasks.
8. Anisometropia may induce prism in downgaze requiring slab-off prism in the glasses to compensate. In brief, slab-off prism adds base up and reverse slab-off adds base down.
9. Patients with vertically incomitant strabismus may be symptomatic in downgaze if they are presbyopic, and they may need special optical management for reading.
10. Progressive (no-line) bifocals may not work well for fusing patients with vertically incomitant strabismus.
11. See Chap. 9 for further discussion of aforementioned items 7, 8, and 9.

## Important Point

All patients with asthenopia or diplopia at near should have an assessment of accommodation either by testing near point of accommodation (NPA) and/or dynamic retinoscopy, as well as fusional convergence amplitudes.

## Important Point

*If a Strabismic or an Amblyopic Patient Shows Deterioration of Their Visual Acuity or Deviation, They Should Have a Repeat Refraction*

Also, you should check via the lensometer to see that the glasses they are wearing are the same as you expect them to be. It is surprising how often patients come in for a return visit and do not report that the glasses had been replaced (and they were made incorrectly) or they inadvertently resumed wearing an outdated prescription.

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# Functional Amblyopia

# 4

*With his right eye,*
*God looks after all he created,*
*except for the poor.*
*For them he uses his left eye, which is blind.*

*—Dale M. Kushner, Black Loaves.*

*With nothing more than a simple eye patch, we have brought amblyopia to its knees.*
*—Charles Schultz (Sally talking to Charlie Brown).*

## Overview

### Basic Information

Few things in medicine are as gratifying as the successful treatment of amblyopia. Not only the treatment is inexpensive and noninvasive, but also the patient's own effort and compliance play an important role in determining the outcome. The successfully treated patient can feel a great sense of triumph and accomplishment, which can be empowering for children and their parents.

> The treatment of amblyopia is extremely gratifying.

The term *amblyopia* was derived from Greek, *ambluōpia* "dim-sightedness." It originated with Hippocrates around 450 BCE, when he wrote "If the doctor does not find any alteration in the eyes of a child, and if the child does not see for one of them, the diagnosis is ambliopy." Later, Albrecht von Graefe famously rephrased this as "The condition in which the observer sees nothing and the patient very little." [1] A more thorough definition of amblyopia was offered by von Noorden when he described amblyopia as "a decrease of visual acuity in one eye when caused by abnormal binocular interaction or occurring in one or both eyes as a result of pattern vision deprivation during visual immaturity, for which no cause can be detected during physical examination of the eye(s) and which in appropriate cases is reversible by therapeutic measures." [1] To be thoroughly twenty-first century one can look on Wikipedia, where amblyopia is defined as a disorder of sight that "results in decreased vision in an eye that otherwise appears normal" [2]. In fact there are structural organic abnormalities present in amblyopia, but they lie in the lateral geniculate body and visual cortex in the brain, and indeed cannot be seen in the course of an eye exam. Importantly, these microscopic abnormalities can be reversed as a result of the brain's plasticity. I prefer the term "functional amblyopia" to refer to this potentially reversible form of vision deficit.

## Treatment Principles

### Basic Information

Put in reductionist terms, the treatment of amblyopia is simple: Correct for any significant optical abnormality and then force the patient's brain to pay attention to the formerly neglected eye. The classic way to accomplish this has been to occlude the preferred


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_4

eye with a patch. This treatment dates back to about 900 AD when the Sabian physician, mathematician, and astronomer, Thabit Ibn Qurrah of Harran (a city in upper Mesopotamia), recommended patching to treat lazy eye. However, credit is usually given to the Frenchman, George Louis Leclerc, Comte de Buffon (1743), for recommending occlusion therapy. As Creig Hoyt pointed out, both Thabit Ibn Qurrah and Comte de Buffon only treated patients with strabismus, and it is possible that they were recommending occlusion for the purpose of straightening the deviated eye—a treatment that may have been an outgrowth of the various facial masks that had been used to force a deviated eye into proper alignment [3]. In the 17th Costenbader Lecture, "Amblyopia Revisited," John Flynn pointed out that our basic treatment approach to amblyopia has not changed in hundreds of years [4]. Although this is true, recent knowledge has led to refinement of treatment that has made it less onerous. In addition, I feel that we are on the cusp of some radically different treatment modalities that show promise. However, as of this writing, none seems ready for prime time.

## The Nature of Amblyopic Vision

### Advanced Information

We tend to equate a patient's "vision" with their visual acuity. This does not tell the whole story. Patients with different cause for decreased visual acuity may be experiencing a totally different view of the world; their visual acuities may even be identical. A patient with 20/200 visual acuity due to uncorrected myopia sees the world very differently than the patient who is 20/200 secondary to macular degeneration or optic neuritis. I have come to view amblyopic vision as one of the worst forms of vision deficit. Typically, a patient with visual acuity of 20/40 in one eye secondary to amblyopia will refer to that eye as their "bad eye" or "blind eye." I once cared for a 67-year-old woman who demonstrated this dramatically. Her left eye was amblyopic with a best corrected acuity of 20/40; she had a large left exotropia (LXT). Prior to seeing me she developed a spontaneous macular hole in her right eye, which reduced her acuity from 20/20 to 20/400. Nevertheless, she continued to always fixate with the 20/400 right eye. When I asked why she did not prefer to use her 20/40 left eye, which in my mind had better vision, she replied, "I cannot see with my left eye."

> Visual acuity alone paints an incomplete picture of vision of amblyopia.

### Advanced Information

Adults with amblyopia have described what they see with their amblyopic eye as being like the shimmer effect of hot air over a highway. There is a continuous wavy motion to the environment. Parts of the object of regard do not remain stationary but come in and out of focus or intermittently fade from view. One artistically talented patient of mine drew for me what he experienced visually in his amblyopic eye (Fig. 4.1). Consequently, an amblyopic

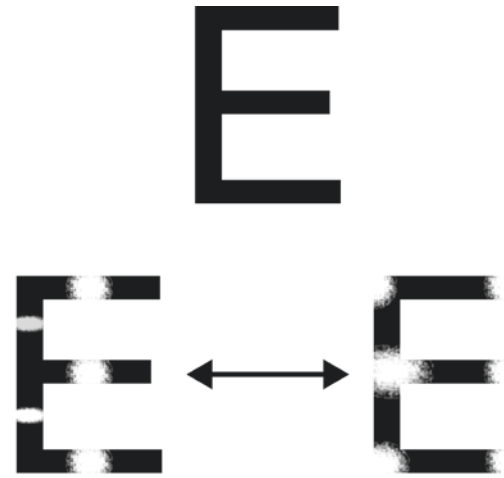


**Fig. 4.1** Artist's depiction of how the letter E appears with an amblyopic eye. There is a continuous sense of motion, with different parts of the E coming in and out of focus

eye may have a much better visual acuity if allowed to read slowly, which allows the patient to figure out the shape underlying the motion. This, in part, is the basis for the crowding phenomenon, e.g., better acuity if tested with isolated optotypes. Multiple optotypes surrounding the test object tend to blend into one another and are harder for the amblyopic eye to distinguish.

### Advanced Information

Standard visual acuity tests involve identifying shapes for which there is an abrupt interface between the black optotype and a white surround, which is a situation of high contrast. However, the world we live in is not limited to these high-contrast visual stimuli. For most objects in our visual environment, there is a more gradual change in contrast and also varying degrees of illumination. Eyes with certain types of functional amblyopia may actually have a better visual acuity than normal eyes in certain conditions of reduced illumination. This is why some eyes with amblyopia may show an improvement in visual acuity when tested with a neutral density filter. Currently, most exam rooms are equipped with acuity charts that are randomly generated on a TV monitor. Alternatively, some use a projected chart. One can make a theoretical argument that with either testing system, vision in amblyopes should be tested with the room lights on. After all, that represents the world the child lives in, and in which they must patch.

## Classification of Functional Amblyopia

### Basic Information

Functional amblyopia is typically classified according to five general categories, the first two of which are decidedly more common than the other three.

1. **Strabismic Amblyopia**
   In some forms of strabismus there is an active inhibition of the image coming from the fovea of the deviating eye, which gives rise to strabismic amblyopia.
2. **Amblyopia Caused by Unilateral Distortion of Form**
   If there is asymmetry of the clarity of the form projected on the retinas of the two eyes, the eye with the less clear image may be suppressed, resulting in amblyopia. Anisometropia is by far the most common cause of this condition. However, eyes in which there is distortion of the form of the retinal image due to problems within the media (corneal scar, partial cataract, vitreous opacity, etc.) may also develop some element of functional amblyopia superimposed on the organic vision loss [5–7]. Although many patients with amblyopia secondary to an asymmetry of the clarity of the image formed on the retina may have straight eyes, many others develop strabismus. In some cases, it may be impossible to tell clinically if the amblyopia is strabismic or secondary to the asymmetric clarity of the formed image. Fortunately, for all practical purposes the distinction is moot, as the treatment is the same for both conditions.
3. **Ametropic Amblyopia**
   Patients with significant bilateral refractive errors that have not been previously corrected with appropriate spectacles may develop a reversible functional amblyopia in both eyes. This is decidedly less common than the two previously described forms of amblyopia.
4. **Deprivation Amblyopia**
   This term describes amblyopia caused by substantial blockage of the image formed on the retina in early infancy (congenital cataract, vitreous hemorrhage, corneal opacity, etc.). Derivation amblyopia is usually more severe than the three aforementioned types of amblyopia and needs to be treated earlier. The critical period for treating it is generally assumed to be within the first 2–3 months of life.
5. **Occlusion Amblyopia**
   This is an iatrogenic form of amblyopia resulting from prolonged occlusion or pharmaco-

logic penalization of a previously normal eye for treating contralateral amblyopia. It is almost always reversible.

## Criteria to Diagnose Amblyopia

### Basic Information

The standard definition of amblyopia requires a best corrected visual acuity of less than 20/40 on a standard visual acuity chart, or at least a two-line difference between the two eyes.

### Advanced Information

I feel that there are some problems with the aforementioned definition, but fortunately they are minor and do not really alter our treatment—just our understanding. Consider a patient with a condition that predisposes to amblyopia (anisometropia, strabismus, etc.) and who has a best corrected visual acuity of 20/40 + 3 in the nondominant eye. Can one not say there is mild amblyopia, even though it does not meet the definition thereof? If not, what is the physiological explanation for the decreased vision? In my experience, eyes fitting this description have a lot of characteristics of amblyopia. They show the crowding phenomenon, may perform better if given more time for testing, and sometimes improve when tested with a neutral density filter. Furthermore, the acuity will often improve with further amblyopia therapy. Presumably the same physiological process is at work as is seen in eyes with amblyopia. Similarly, an eye that can eke letters on the 20/20 line, but takes much longer to do so than the fellow eye, probably also has the same physiologic process that affects amblyopic eyes as the cause. I consider the standard definition to be somewhat arbitrary, yet useful for interpreting reports on the treatment of amblyopia; I shall use it in this book.

### Basic Information

Some of the characteristics of amblyopic vision include improved vision in mesopic conditions, the crowding phenomenon, variability of spatial frequency responses, and a mild afferent pupillary response in some cases, but normal color vision.

### Important Point

The threshold of 20/40 acuity for diagnosing amblyopia is arbitrary and probably not completely accurate. Eyes with acuity that is better than 20/40 may still exhibit many of the characteristics of amblyopia.

### Basic Information: Diagnosing Amblyopia in a Patient

We tend to incorrectly use the term *preverbal* to describe patients who cannot yet identify optotypes, when in fact we mean *preliterate*. A 1-year-old child may be able to talk but cannot identify letters. For the literate patient, diagnosing amblyopia is easy. If the patient's best corrected visual acuity is below the arbitrary threshold of 20/20, and the eye is structurally normal, *voilà,* the diagnosis of amblyopia is made. For the preliterate child, there are an array of tests in our toolbox that can be helpful.

1. **Fixation Preference**
   If a child with strabismus will hold fixation with either eye, you can be sure that she is not amblyopic. If there is no strabismus, you can induce strabismus by introducing a 10 prism diopters (Δ) vertical prism, or a 25 Δ base-in prism over one eye, and assess fixation behavior as you would do in a strabismic patient. But fixation preference is another "one-way test." If a patient does not alternate fixation, one can only conclude that they might be amblyopic.

There are certain circumstances in which a patient will not hold fixation with their nonpreferred eye, even if their two eyes have approximately equal visual acuity. These include substantial anisometropia, optically corrected aphakia, pseudophakia, monofixation syndrome, and minor unilateral media opacity. In addition, some strabismic patients simply maintain a strong fixation preference even after amblyopia is overcome.

The absence of alternation only indicates that amblyopia *may* be present.

2. **Preferential Looking: Teller Acuity Cards**
Preferential looking tests are based on the principle that an infant's visual cortex would rather look at a pattern than at a homogeneous background, provided that the visual system can resolve that a pattern is present. Teller Acuity Cards (Teller Acuity Cards II, Stereo Optical, Chicago IL, USA), developed at the University of Washington, Seattle, Washington, are a commercially available set of grating acuity cards for performing preferential looking according to a specific testing algorithm. In the 1980s and 1990s, Teller Acuity Cards were becoming all the rage for solving the problem of visual acuity testing in preliterate children. Thirty plus years later it appears that this promise has not really borne fruit for several reasons [8, 9]. To start with, Teller Acuity Cards are a test of visual resolution; the subject only has to be able to determine that a pattern is present as opposed to homogeneous gray. However, reading a Snellen chart is a test of visual recognition; the subject must not only be able to determine that a pattern is present, but he or she also must recognize what form or shape the pattern has, e.g., identify a letter. Numerous studies have shown that ocular diseases affect grating acuity differently than recognition acuity, and in fact the original developers of Teller Acuity Cards never intended them to be used for diagnosing amblyopia [8, 9]. This in part was due to the fact that grating acuity tends to overestimate acuity in many of the ocular conditions that affect children, and specifically amblyopia. Conversely, because preferential looking tests require the child to make a motor response in the form of looking toward the pattern they detect, Teller Acuity Cards may underestimate acuity in children who have motor delays affecting their saccadic eye movement system. They may "see" the pattern but not look toward it. In one study involving 69 patients with amblyopia of mixed cause, I found that Teller Acuity Cards had only a 66% sensitivity for detecting amblyopia (acuity less than 20/40) [9].

### Important Point

If Teller Acuity Cards indicate normal visual acuity, there is still a high likelihood that it may be abnormal. If Teller Acuity Cards suggest abnormal vision in a developmentally normal child, there is a high likelihood that it is abnormal.

### Try This Experiment

The reason grating acuity overestimates acuity compared to recognition acuity can be appreciated by referring to the experiment (Fig. 2.1) shown in Chap. 2, "The Examination." If you optically defocus the figure by viewing it through a plus lens of sufficient power so that you can just read the J16 line on the card on the left, you will still be able to easily detect the stripes on the Teller Card, which subtends a much smaller angle than the J16 line.

### Important Point

*Because Grating Acuity Tends to Overestimate Visual Acuity as Compared to Recognition Tests, Grating Acuity Values Should Not Be Used for Social Service Purposes* [8].

I have seen preliterate children with clinical conditions incompatible with acuity better than 20/200, e.g., extensive posterior pole toxoplasmosis scars, who had acuity values in the normal range with Teller Acuity Cards. Based on clinical findings they would clearly qualify as being *legally blind* and qualify for various social service benefits. These would be denied based on their normal Teller Card acuities.

**Important Point**

*In Spite of the Above-Mentioned Limitations of Teller Acuity Cards, They Are Not Without Value*

If they do detect an interocular difference in acuity in a suspected amblyope, they can be useful for monitoring treatment progress. They also can be useful for comparing mean visual acuities in different treatment arms of a study, as has been done in various retinopathy of prematurity (ROP) clinical trials.

3. **Sweep Visually Evoked Potential (VEP) Testing**
   This is also a grating acuity test, but in my mind has distinct advantages over preferential looking tests. Because it does not rely on a motor response from the child to look toward the pattern, I find it much more accurate than Teller Acuity Cards in children with multiple system delay. Also, and importantly, interpretation of the test is more objective and less influenced by examiner subjective interpretation; it requires less cooperation from the child, and takes less time. However the acuity values obtained with sweep VEP are also not identical to Snellen acuity on recognition tests, and should also not be used for social service purposes. The main downside to sweep VEP testing is that it requires a skilled technician to perform and interpret the test correctly. Most sweep VEP tests are done in labs that perform electroretinography, and those performing the test may not be familiar with the nuances of VEP testing. Although there is need for a "turnkey" sweep VEP testing unit that can be performed by a relatively unskilled technician, such a unit is not currently available. I find sweep VEP the most practical and helpful way to diagnose amblyopia and follow treatment progress in preliterate children who are not yet old enough for letter matching tests, given appropriate technical support.
4. **Letter Matching Tests**
   I find that many young children can perform a letter matching test at a younger age than the tumbling-E test, particularly if they have had no prior introduction to testing. I prefer the Sheridan Gardner test to the HOTV test, as it has seven letters to choose among. In both tests the child is shown letters of various sizes and she responds by pointing to the same letter on a card that she holds during the test. In its commercial form, the Sheridan Gardner test is designed as an isolated-letter test, which is a substantial disadvantage. However, it is easy to modify the test by adding black lines (crowding bars) around each letter in order to introduce the crowding phenomenon. This permits the test to simulate a linear test in accuracy (Fig. 4.2) [10].
5. **Other Optotypes**
   There are numerous other choices of optotypes for recognition acuity tests for children who are not quite ready to read letters, yet are verbal. The most useful and accurate in my opinion are the Lea symbols, which consist of an apple, a square, a pentagon, and a circle. These can be described to children and identified as an apple, a window, a house, and a ring (Fig. 4.3). I find other picture charts, such as Allen figures, to be substantially less accurate. The tumbling-E test, or the Landolt C test, both of which require the child to identify the orientation of the "legs" of the "E," or the opening in the "C," respectively, comes close in accuracy to the standard Snellen chart, but is a bit more

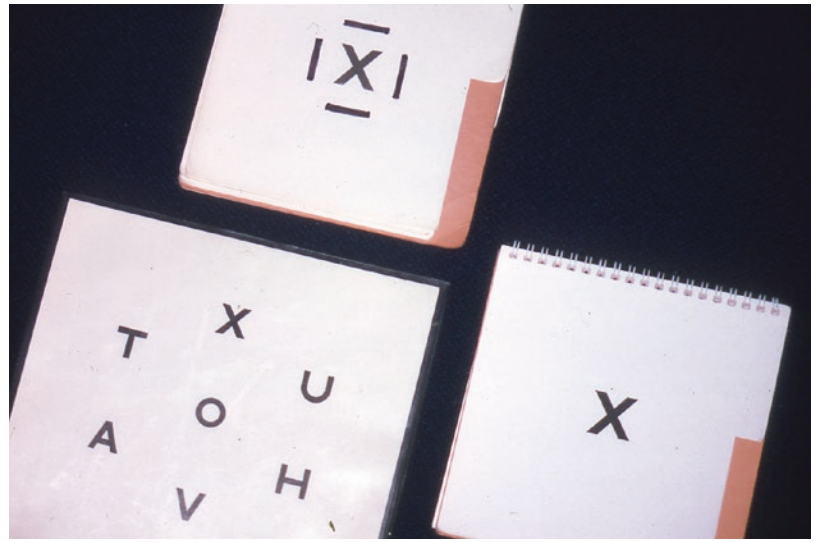



**Fig. 4.2** Sheridan Gardner test: *Bottom left:* Card held by patients for matching optoptype. *Bottom right:* Isolated optotypes presented to the patients. *Top:* Crowding bars have been placed around the isolated optotype to introduce the crowding phenomenon (from Kushner [10], with permission)

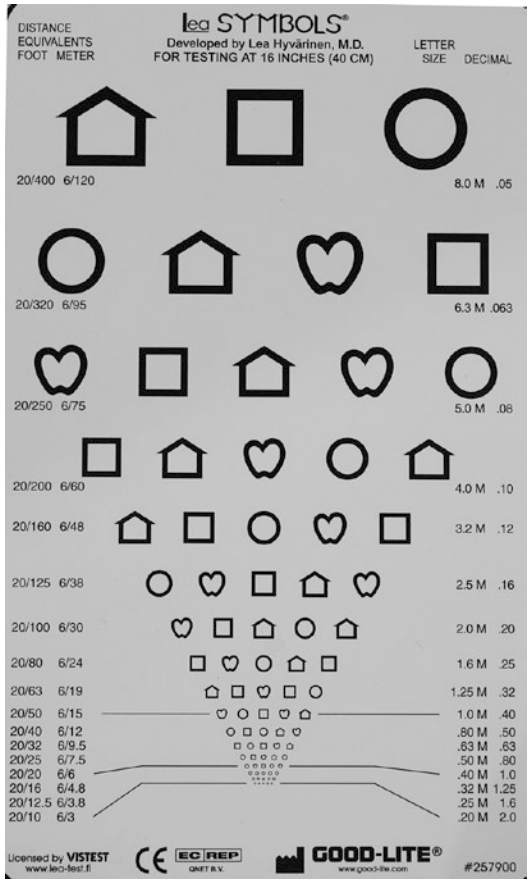



**Fig. 4.3** The Lea symbols consisting of an apple, a square, a pentagon, and a circle

difficult for the child than the Lea symbols or the Sheridan Gardner test. The ability to distinguish right from left is often not established in the cortex until 5 or 6 years of age.

### Important Point

The degree to which lack of the crowding phenomenon invalidates visual acuity tests in eyes with mblyopia is substantial. When I reviewed a series of 847 amblyopes, 44 had a recorded visual acuity of 20/20 with an isolated-optotype test (either the tumbling-E or the Sheridan Gardner without crowding bars) and on their subsequent visit 1 month later were testable for the first time with Snellen letters [10]. Thirty-six of the 44 patients showed a drop in visual acuity when graduating from the isolated-optotype to the linear test, with an average drop being four lines on the chart (range 1–8 lines). In general, any child who can perform the tumbling-E test can be tested with a linear-E chart. It is almost never necessary to test a child with isolated optotypes.

It is almost never necessary to test a child with isolated optotypes.

## Refracting an Amblyopic Eye

### Basic Information

Accurately refracting an amblyopic eye is one of the most critical steps toward successful management. The refraction must be done objectively, either with streak retinoscopy under full cycloplegia or using an auto-refractor. I personally have more confidence in my findings obtained with retinoscopy than with any auto-refractor I have used, but that may reflect my age and when I trained. What is important is that any attempt to subjectively refine the refraction in an eye with significant amblyopia will be frustrating and inaccurate. Amblyopic eyes have decreased ability to discriminate changes in image quality. It is not uncommon for patients with amblyopia to be unable to distinguish the difference between changes in trial lenses in increments of over 1 diopter.

> Amblyopic eyes must be refracted objectively, either with retinoscopy or an auto-refractor.

### Important Point

*Retinoscopy*

The greatest source of error in retinoscopy occurs when the examiner is retinoscoping off axis, which results in an error in determining the magnitude of astigmatism. It is essential that the patient look at the retinoscope with the amblyopic eye; consequently, full cycloplegia is needed (Fig. 4.4).

### Pearl

*If The Visual Acuity in the Amblyopic Eye Is Poor Enough That the Eye Will Not Fixate on the Retinoscope Light, I Repeat the Refraction After the Child Has Occluded the Unaffected Eye for a Week*

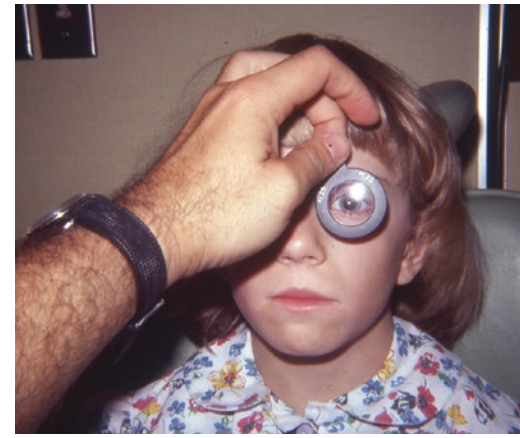


**Fig. 4.4** To insure that the amblyopic eye is being refracted on-axis, loose lenses can be held in such a manner as to occlude the unaffected eye and permit fixation with the amblyopic eye (from Kushner [10], with permission)

I have the child come in to the office wearing the patch, and have the parents instill cycloplegic drops prior to bringing the child to the office.

### Important Point

*I Rarely Use a Phoropter for Younger Children*

I believe that it is often more accurate to use a pediatric trial frame for most children between approximately 2 and 10 years of age. Young children often do not maintain adequate head position behind the large phoropter. They often assume head tilts, which can produce errors in the interpretation of the axis of astigmatism. A trial frame will automatically compensate for this. For children under 2 years of age, I generally use loose lenses. When I refract a young child who is already wearing spectacles, particularly if there is a high refractive error, I typically perform an over-refraction as described in Chap. 3 (Fig. 3.1). By using a trial lens clip over the patient's spectacles, the amount of additional sphere and cylinder needed can be obtained with retinoscopy. This technique eliminates potential errors in vertex distance correction as well as errors induced by the patient tilting his or her head during retinoscopy. If the

axis of the added cylinder needed differs from the axis of the cylinder of the existing spectacles, the final resultant refractive correction can be obtained by one of the three methods. The patient's spectacles, combined with the over-refraction lenses, can be sandwiched together and placed in a lensometer. Neutralization of that combination will give the final net refraction. Alternatively, some of the electronic medical record systems are equipped to calculate an over-refraction result with the click of a mouse (I never thought I would ever have anything positive to say about electronic medical records).

### Important Point

*If a Patient Has Anisometropia, the Amblyopic Eye Should Be the More Hyperopic (If Both Eyes Are Hyperopic) or the More Astigmatic Eye*

If not, you should strongly suspect the presence of organic pathology in the eye suspected of having amblyopia. I have seen very few exceptions to this generalization. Also, if one eye is hyperopic and other is mildly to moderately myopic, the patient will typically use one eye for distance and the other for near and not be amblyopic. This latter generalization breaks down if the more myopic is highly myopic, e.g., 5–6 diopters or more. In that case the near point of focus in the more myopic eye may be so close that the patient will use the other eye at all common viewing distances.

If the eye suspected of being amblyopic is not the more ametropic eye, look again for organic pathology.

### Important Point

*The Conditions That Lead to Amblyopia (Strabismus and Anisometropia) Are Frequently Hereditary*

Always suggest that siblings of amblyopic patients be examined.

## Prescribing Spectacles for Amblyopia

### Basic Information

*Image Degradation Is a Major Cause of Amblyopia, and as Such an Amblyopic Eye Should Have the Most Accurate Optical Correction Possible*

You should never arbitrarily cut the size of the cylinder or rotate the cylinder axis to bring it closer to 90 or 180°. Sometimes correction of what seems like a trivial refractive error may be needed to successfully treat an amblyope.

When we prescribe glasses for children that are not esotropic, it is common to prescribe less than the full amount of hyperopia. But for patients with amblyopia, some important rules should be followed. Because eyes accommodate symmetrically, and it is the fixing eye that determines the amount of accommodation, any decrease in plus must be done symmetrically. Consider a hypothetical patient with amblyopia in the right eye and the following cycloplegic refraction: OD -1.00 + 1.75 × 90 and OS +0.50 sphere. If one wished to cut the plus in the left eye and prescribe plano sphere, the prescription for the amblyopic eye should also reflect less plus (or in this case more minus) and be OD −1.50 + 1.75 × 90. Otherwise when fixing with the left eye and accommodating 0.50 diopters, the amblyopic eye would also be accommodating +0.50 diopters and be fogged by that amount.

Cut plus or over-minus symmetrically.

### Important Point

*Amblyopic Eyes Have Decreased Accommodative Amplitudes if They Are Presented with the Stimulus to Accommodate*

Consequently, you should be cautious in excessively cutting plus in an amblyopic eye. As a general rule, in a patient without an accommodative esotropia (ET), cutting plus by 0.75–1.00

diopter will prevent the amblyopic eye from being fogged, yet still allow for good vision in the amblyopic eye.

## Question

I was asked to consult on the management of a 7-year-old boy regarding his amblyopia management. At 4 years of age he was diagnosed with amblyopia in his right eye, given spectacles, and patched for all his waking hours for 4 months. His parents said that during that time he behaved "like he was blind," and his visual acuity did not improve. Past records showed that he had been refracted by his prior ophthalmologist as being OD +8.75 + 0.75 × 90 and OS +1.50 sphere. The doctor had prescribed OD +5.00 sphere and OS plano sphere. He reasoned that a 4-year-old could easily accommodate 3.75 diopters, and hence he cut the plus by that amount in the right eye, and that the +0.75 D cylinder was trivial. Also, he did not need the plus corrected in the left eye. My examination revealed essentially the same refractive error as had been originally found by the prior doctor, and a best corrected visual acuity of 20/200 OD. I was questioned if anything different should be done.


## Reply

My treatment consisted of prescribing my refraction for both eyes, cutting the plus by 1 diopter in each. I then reevaluated him a month later, because some amblyopic eyes may improve with appropriate spectacle wear alone. His acuity did not improve after 1 month, so I resumed occlusion therapy. This time he responded well and 4 months later was 20/30 in his right eye. Patching was tapered and then discontinued, and he remained stable for the subsequent 8 years that I followed him. In this case, the prior doctor's management was problematic in several ways. Firstly, any cutting of the plus should have been done symmetrically. By cutting the plus 1.50 in the left eye and 3.75 in the right eye, the right eye would be 2.25 diopters out of focus when the left eye was fixing. This, however, does not explain the failure to improve, as the child was patched essentially full time and not fixing with the left eye. Secondly, cutting the plus by 3.75 diopters is too much for an eye with amblyopia due to its decreased accommodative amplitude. This is probably the major source of treatment failure. Finally, not correcting the astigmatism may have contributed to the treatment failure, as any amount of blur or distortion is undesirable when treating amblyopia.

Although it is classically taught that accommodation is equal and symmetric, with the amount being determined by the fixing eye, the evidence for this is scant. In fact, Horwood has evidence that by measuring accommodation in both eyes simultaneously by photo-refraction, many anisometropic amblyopic children can aniso-accommodate by a considerable amount, with the amblyopic eyes under-accommodating while the other eye does just fine (Anna M. Horwood, personal communication, 15 Nov 2016). If this observation is valid, one can make a strong case for giving full hyperopic correction with a bifocal to all amblyopes while they undergo amblyopia treatment. This is an untested approach, but could easily be validated with a clinical trial.

## Advanced Information

*Occlusion Esotropia*

Rarely, patients with amblyopia and straight eyes may develop an ET as a result of occlusion therapy—a condition appropriately called *occlusion.* In theory this is more likely to occur in patients who have a substantial amount of their hyperopia left uncorrected while being patched. This makes sense. If a patient has to accommodate several diopters, and one eye is occluded, her binocular fusional mechanism cannot prevent accommodative convergence from causing an ET. By giving close to the full hyperopic correc-

tion to patients with amblyopia, this would be prevented. Theoretically, occlusion ET should be less likely to occur with the trend toward less than full-time occlusion (more on this to come). I have seen an ET develop in a 4-year-old boy who was treated by his family practitioner for a corneal abrasion, with 3 days of full-time occlusion. He had a symmetric uncorrected hyperopic refractive error of 3 diopters.

### Question

I am treating an anisometropic amblyope with a refraction of OD -8.00 + 1.00 × 70 and OS +2.00 + 1/00 × 90. Do I need to fit him with a contact lens because of concern about aniseikonia?


### Reply

Admittedly there is controversy about this, but I have been happy with the following approach. I fit these patients initially with spectacles incorporating the appropriate correction and begin occlusion therapy. Aniseikonia should not be an issue while the patient is being patched. If the amblyopia is successfully treated, and if the patient then shows signs of aniseikonia as the visual acuity improves in the amblyopic eye (complaints of diplopia, closing one eye, etc.), I then switch to a contact lens. Even if the child is only patching several hours a day, aniseikonia should not be a problem until he or she improves to the point he or she no longer suppresses. Knapp's Rule says that aniseikonia will not occur if the anisometropia is axia [10]. Although congenital unilateral high myopia is often axial, this is not always the case. Although obtaining a quantified axial length measurement will sort this out, I find it easiest to start with spectacles in all patients of this type. In a review of 847 amblyopic patients, 18 were high anisometropes (greater than 5 diopters difference in spherical equivalent) who had a successful result from occlusion therapy [10]. Of the 18, 10 did not show signs of aniseikonia and therefore stayed in spectacles. The other 8 did experience aniseikonia and were fitted with a contact lens which eliminated their symptoms. Using this two-stage approach, ten patients were spared the expense and inconvenience of contact lens fitting.

## Amblyopia Therapy

### Basic Information

*Instituting Therapy*

If a patient's refractive error is substantial enough in the amblyopic eye that I feel he or she will not obtain good vision without spectacles, I do not begin occlusion therapy until after spectacles have been obtained. Generally, this is in the range of more than 1.5 diopters of plus sphere and/or 1 diopter of astigmatism. If the refractive error is less than that, I may begin occlusion without waiting for the spectacles.

### Important Point

*Many Amblyopes Will Show Some Improvement in Their Visual Acuity with Spectacles Alone [11]*

This is particularly true if they do not have a significant angle of manifest strabismus, but do have a substantial refractive error (anisometropic amblyopia). It is my routine with these patients to see them at regular intervals and defer patching until the vision stops improving. Of the 847 amblyopes I reviewed, 68 were anisometropic amblyopes [10]. Nearly half of them improved at least two lines as a result of just wearing spectacles. Eighteen patients never needed occlusion therapy and were successfully treated with just spectacles alone.

Many anisometropic amblyopes will improve with spectacles alone.

Although many ultimately needed occlusion therapy, it seems logical that patching is easier, and need not be carried out as long, if one starts with a better visual acuity.

## Basic Information

*Patching vs. Penalization*

I think one of the major take-home points from the numerous Pediatric Eye Disease Investigator Group (PEDIG) studies on amblyopia is that pharmacologic penalization worked better than many people thought [12, 13]. To my mind, however, the studies did not prove that the treatments are equally effective. Like all well-designed studies, they answered a narrowly defined question—What was the percent of patients achieving 20/40 or better after 6 months of treatment? With respect to this question, the results were equivalent in the two groups. However, that is not the only question worth asking, or necessarily the best. Did one group reach peak acuity earlier? Did one group have more patients achieving acuity of 20/20? With respect to these other questions, patching seemed to do better.

Both patching and penalization have relative advantages and disadvantages over one another. Pharmacologic penalization has the main advantage of taking poor patient compliance out of the picture. Once the drop is instilled, it cannot be removed. For this reason, parents often find this treatment easier. Patching has the advantage of allowing for more titration. You can increase or decrease the number of hours a day the child patches, and can have days with no treatment if that is desired. I personally like to taper treatment after peak acuity is reached. This can be done with more finesse with patching. I recommend patching as a first line of treatment, and save penalization for those patients where compliance is a problem or in patients with nystagmus.

> "And watch Betsy's eye patch."
>
> Betsy had one strong eye and one weak eye that tended to cross. She didn't use the weak one because she couldn't see very much through it. The patch was meant to block the good eye so that she would use the weak eye and make it stronger. But whenever she thought no one was looking, Betsy switched the patch from the good eye to the bad eye so she could see better. You had to pay attention.—Terry Ryan, *The Prize Winner of Defiance, Ohio*

## Question

What is your protocol for patching?


## Reply

This has undergone a shift in recent years as studies have shown that many amblyopes will respond to fewer hours of patching than was previously thought necessary. I will usually start with something like 2–4 h a day of occlusion of the dominant eye, depending on several factors. If there are findings that put the patient in the poorer prognosis category (older age, poorer vision, or marked anisometropia), I tend to start with a higher patching dose. In the absence of any of these factors my initial dosage is at the lower end. The issue as to whether it is important to prescribe near work while patching, as has been done in PEDIG trials, has not been proven [14]. Nevertheless, it seems logical that this might make a difference. My personal approach is to suggest that patching be done when the child is engaged in visual tasks, e.g., reading, watching television, and using a computer, rather than while just running around—if possible. This makes sense to me, but is not of confirmed benefit. There is an old rule of thumb that patients undergoing patching should be seen at intervals of a week per year of age. A 1-year-old should be seen weekly, a 2-year-old every 2 weeks, and so on up to a maximum interval of every 4 weeks. This was primarily to monitor for occlusion amblyopia, and was proposed at a time that most amblyopes were being patched full time, or close thereto. With the trend toward starting with fewer hours a day of patching, this protocol seems overly intense [15]. If I am starting with 2–4 h a day of patching, I typically see the child back for the first time between 1 and

4 weeks, in part because I want to address any problems or issues that may have arisen from the treatment, and to get a sense as to how rapidly or slowly the child is responding. Depending on the response, age, depth of amblyopia, and parent's ability to come in for appointments, I may spread out the visits. If coming in to the office frequently is problematic for the family, the patching dose can be decreased and the time between visits increased. When peak acuity is reached I prefer to slowly taper patching. If a child is patching 4 h a day when peak acuity is reached, I cut that to 2 h a day for a month, 2 h every other day for a month (or 1 h daily), and perhaps 2 h every 3 days for a month, and then stop. If a child has a recurrence, I reintroduce patching and taper more slowly. I know of no data that this is beneficial, but it makes sense to me. In my review of 847 amblyopic patients, 802 had a successful outcome from their amblyopia treatment. Fifty-two of the 802 had some recurrence of their amblyopia as occlusion was tapered or discontinued. Nineteen of them had high anisometropia, 14 had some organic pathology coexisting with amblyopia, and 19 had a manifest tropia of 15 Δ or more at the time of recurrence. If you prefer not to routinely taper patching, I recommend you at least do so in patients in any of the three mentioned categories (older age, poorer vision, or marked anisometropia).

## Important Point

*Always Neutralize the Patient's Glasses on Each Visit and Be Sure That They Are Correct*

It is surprising how often patients will come in having replaced a broken pair of glasses, and the parents do not mention it as they assumed that the prescription used was supposedly the same as what I last prescribed. Often they filled an old prior prescription. Also, it is not uncommon for children to come in wearing an older pair of glasses with a prior prescription, and the patents assumed that it was the same as the current prescription. Finally, I see several patients a year who are wearing glasses with round or at least symmetrically shaped lenses. The lenses had popped out and the parent replaced them reversing the right and left lenses. It did not occur to them that the prescription was different in the two eyes. For children wearing this type of lens, it is prudent to mark one of the lenses with a dot of nail polish or in some manner that allows the parents to know which lens goes with which eye.

## Question

How much deterioration in vision is necessary before you will say the vision "slipped"?


## Reply

I will always allow for the fact that certain optotypes are harder to discern than others, so I am usually forgiving of a child missing a few more letters on their threshold line than on a prior exam. Certainly, if the child is older, cooperative, and attentive, one half of a line poorer than before is meaningful. If the child is younger, not completely cooperative, or attentive, I need to factor those issues into my verdict with respect to slippage.

## Important Point

*If an Amblyope Shows a Slippage in Vision, And You Are Testing Them with Similar Optotypes As Were Used in the Prior Exam, You Should Re-Refract Them Unless the Last Refraction Is Very Recent*

A change in refractive error is a major cause of slippage.

## Important Point

Always record what test was used for vision testing—what type of optotype and if it was linear or isolated. If a child's vision appears to have slipped, it may solely be a function of their being tested with a more difficult test than on the prior visit.

## Pearl

If an amblyope has several episodes of recurrence, and they have a small manifest tropia that exceeds 10 Δ, surgery to reduce their alignment to less than 10 Δ may prevent further recurrence. I know of no solid data to confirm this, but it makes sense. We know that many anisometropic amblyopes may improve with spectacles alone, and it makes sense that this improvement would not occur if there were a significant manifest tropia. There is a study that suggests that amblyopia may resolve as a result of strabismus surgery [16]. I know that this is anecdotal, but I have cared for a number of patients who repeatedly had recurrence of amblyopia, who then underwent surgery for deviations between 10 and 15 Δ, and their vision never slipped again.

## Question

I care for a 14-year-old girl who had anisometropic amblyopia. Amblyopia treatment when she was younger was not successful due to poor compliance and irregular follow-up. She underwent surgery in the past for an intermittent left exotropia, or LX(T), and now she has a well-controlled LX(T) of 10 Δ. Her current cycloplegic refraction and best corrected acuity are OD −1.25 + 1.00 × 90 20/20 and OS −2.25 + 5.00 × 70 20/200. She, of course, wears glasses because of the myopic astigmatism in her right eye. Is it necessary to correct the astigmatism in the left eye? Can I prescribe a balance lens, given that her refractive correction does not seem to make much difference?


## Reply

There are no good data on this subject, but I have some convincing anecdotal evidence that leaving the refractive error uncorrected may lead to a deterioration of the strabismus. So other than the cosmetic issue of an asymmetric pair of spectacles, I see no reason not to give proper correction. Many years ago I cared for a girl who had a monocular congenital cataract that I operated on at 3 months of age. It wasn't until after the cataract was removed that I discovered mild optic nerve hypoplasia in the eye. Nevertheless, I fit her with a contact lens and began patching for amblyopia. This proved unsuccessful and she ended up 10/200 in that eye. However, during the time she was patching, she developed an LX(T). Her parents observed that she controlled the XT much better when she was wearing the contact lens, so they elected to continue using it after we gave up on patching. She is now 35 years old and still wears the contact lens. While wearing it, the XT is rarely manifest. But within an hour or so of removing the contact lens, the XT is almost constant (Fig. 4.5).

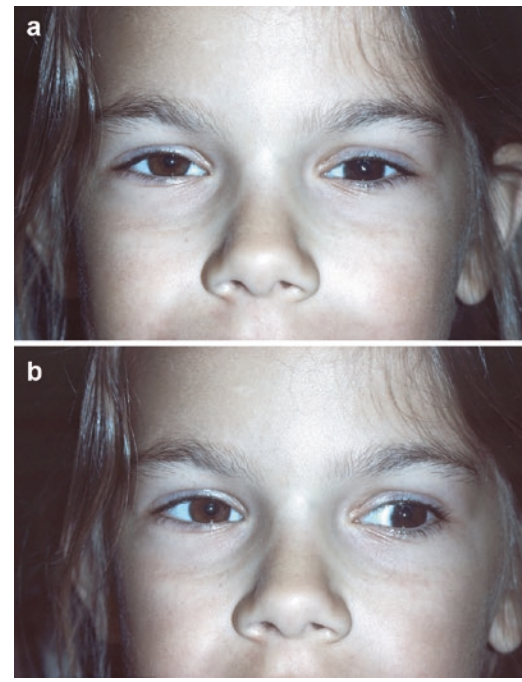



**Fig. 4.5** (**a**) This girl is aphakic OS and has a best corrected acuity in that eye of 10/200. Wearing a contact lens OS she looks well aligned. (**b**) Shortly after removing the contact lens a large intermittent left exotropia becomes manifest

## Basic Information

*Technical Aspects to Patching*

It probably goes without saying that patching is best with an occlusive patch, rather than the *pirate patch* or patch that fits on the glasses. These latter make it too easy for the child to peek. If skin irritation is a problem, I first suggest that parents try a different brand of patch. There are several to choose from, and I find that no one is uniformly superior. Children may be sensitive to the adhesive or material in one brand, and not another. If that does not solve the problem, I have parents spread milk of magnesia on the skin before applying the patch. This usually works wonders. If compliance is a problem, there are many tools that may help, depending on the child's age. Reward systems like gold stars on a calendar work for some. There are an increasing number of books parents can read to their child, geared for children of different ages, to ease the feeling of "being different" because of the need to patch. Allowing the child to play act by patching a doll or toy can be helpful (Fig. 4.6).

## Basic Information

*Time to Give Up*

If a patient is not responding to part-time patching, I will incrementally increase the patching dose until it is close to full time, meaning all waking hours. If a child has gone 3 consecutive months with nearly full-time occlusion and has not shown any improvement, I think it is time to consider treatment a failure and discontinue occlusion. It is important to differentiate between improvement and cure. I have seen a number of patients in whom occlusion was stopped after 3 months because the patient was still amblyopic; however they were improving. In this case treatment should be continued. It is also important to try and assess compliance before deciding to discontinue treatment. If parents claim that a child has been faithfully wearing the patch, but one does not see the telltale sign of pale skin around the eye that was to have been occluded, one can suspect that compliance is a problem.

**Fig. 4.6** Encouraging play acting with a child's favorite toy can decrease the stress of patching

## Pearl

If noncompliance is a problem, and if the non-amblyopic eye has a significant amount of hyperopia (typically 3 diopters or more), I often combine patching with pharmacologic penalization in this manner. I have the parents occlude the spectacle lens of the non-amblyopic eye in a manner the child cannot undo. I recommend either a heavy coating of clear nail polish or adhesive tape on the lens. In addition, the non-amblyopic eye is cyclopleged with atropine 1%, once daily. Usually the child will then leave the glasses on and effectively will be patched. If they take the glasses off, the atropine causes them to prefer fixation with the amblyopic eye. I usually use this type of combined treatment as a form of behavior modification, and switch to my usual patching protocol when compliance and vision have improved.

## Basic Information

*Pharmacologic Penalization*

As stated earlier, my personal preference is to start with occlusion therapy for the reasons stated. However, if compliance is a problem, I do not hesitate to switch to penalization with atropine 1%. When optimum acuity is obtained, I still prefer to taper treatment in some form. I may decrease the atropine to twice weekly followed by once a week. In some circumstances I may switch to cyclopentolate 1% several times a week, which effectively penalizes only on the day it is instilled.

## Important Point

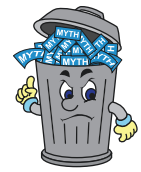
Early in the use of pharmacologic penalization it was felt that if the patient did not switch fixation to the amblyopic eye when the dominant eye was penalized, the treatment would not be effective. We now know this is not the case. How and why a switch in fixation is not needed is not clearly understood. It probably relates to the importance of binocular interaction in causing and curing amblyopia, which is altered by the pharmacologic degradation of the image from the sound eye, even without a fixation switch.

## Basic Information

*Bangerter Filters*

These paper-thin "foils" of varying opacity are a popular alternative to part-time patching. Many ophthalmologists and orthoptists find them particularly useful for mild amblyopia or maintenance therapy. They have the advantage of being less cosmetically noticeable than complete occlusion. I consider these a perfectly acceptable treatment option, but one which I use very infrequently. I have always preferred part-time occlusion when indicated.

# Patient Selection and Prognostic Factors




## Myth

A school-age amblyope is too old to treat.

## Fact

An interplay of several factors impact the prognosis for successful treatment of amblyopia. In general, the younger a patient is when treatment is first instituted, and the better the visual acuity in the amblyopic eye, the better the prognosis. I prefer to avoid rigid boundaries such as setting an arbitrary age above which amblyopia cannot be treated. A 10-year-old with amblyopia and an acuity of 20/50 may be successfully treated, whereas a 6-year-old with acuity of 20/800 might not be. The presence of high anisometropia tends to worsen the prognosis, as does the presence of coexisting organic pathology or eccentric fixation. My own experience is that many children of school age undergoing amblyopia therapy for the first time can be successfully treated, even up to

the age of 14 [10]. I recall one girl who came to see me the day before she turned 12. She had just been discovered to be amblyopic due to high hyperopic anisometropia, and had an acuity of 20/200 in her amblyopic eye. The ophthalmologist who diagnosed her said that she was too old to treat. I put her in spectacles and began patching. Within 2 months she was 20/40 and after 3 months of patching was 20/25. I recently saw her at age 35. Her vision has remained stable and she has 100 s of stereopsis.

### Important Point

Although not everyone agrees on what the age is, most authorities indicate that amblyopia cannot be successfully treated after some stated age. It is important to differentiate between treatment being instituted for the first time and treatment for recurrent amblyopia. In general, most amblyopic children can be brought back to their previous best corrected visual acuity if amblyopia recurs, regardless of their age. I treated a 5-year-old boy with anisometropic amblyopia secondary to 8 diopters of myopia in his amblyopic eye. He improved from 20/200 to 20/30 with spectacles and patching, and later he wore a contact lens in his amblyopic eye. I continued to follow him, and when I saw him at 16 years of age he had gone the entire prior year without wearing optical correction. His best corrected acuity slipped to the pretreatment level of 20/200 best corrected. He then resumed wearing optical correction and started part-time patching. Within 2 months his acuity was back to 20/30.

### Pearl

*Prior Treatment Failures May Be Successfully Treated*

Sometimes it is worth trying amblyopia therapy in a child who presents with a history of previous treatment failure either under your care or elsewhere. Several factors influence my decision on whether or not to begin treatment. Certainly, if there is a change in the degree of motivation on the part of the patient or parents and the prior failure was due to poor compliance, retreatment is indicated. Changes in the domestic situation such as divorce and remarriage can sometimes have a positive impact on the likelihood of success. For patients who were treated elsewhere, I always review prior treatment records to see if the treatment was adequate. In my review, I identified 30 patients in whom a previous patching trial was inadequate and unsuccessful, despite the parents having thought that an adequate treatment course had been carried out [10]. I identified five specific common errors in the prior treatment. Frequently, the patching was discontinued too soon or involved too few hours a day. Often patients were not put in spectacles, or the spectacle prescription was inappropriate. Failure to taper patching seemed to be associated with recurrence. In some circumstances the patching was discontinued after several months because the amblyopia was not cured, despite there being significant improvement.

Recurrent amblyopia can usually be successfully treated at an age.

## Special Forms of Amblyopia

### Functional Amblyopia with Organic Ocular Pathology

### Advanced Information

It has been theorized that mild organic causes of optical blur may induce functional amblyopia in the same manner as a refractive error [5–7]. This is the basis for reports of successful treatment of amblyopia associated with organic pathology. It may be difficult to diagnose the presence of amblyopia in a patient who had some obvious structural abnormality in one eye. I find that improvement in visual acuity with isolated letters or with a neutral density filter suggests the presence of functional amblyopia. In patients where the organic problem is that of media opacity, one can assess if the level

**Table 4.1** Organic pathology coexisting with functional amblyopia

| |
|---|
| Corneal leukoma |
| Extensive pupillary membrane |
| Partial cataract |
| Vitreous opacity (includes PHPV) |
| Perimacular chorioretinal scar |
| Iris/choroidal coloboma |
| Albinism |
| Congenital glaucoma |
| Extensive myelinated nerve fiber |
| Mild cicatricial retinopathy of prematurity |
| Optic nerve hypoplasia |
| Disk coloboma |
| Optic atrophy |
| Optic nerve glioma |
| Congenital nystagmus |

of vision deficit is proportional to the degradation of the view one has looking at the fundus with the direct ophthalmoscope. However, in cases of optic nerve hypoplasia, nystagmus, albinism, or other situations that are not specifically associated with media opacity, only a trial of amblyopia therapy can confirm the diagnosis. Table 4.1 lists many conditions that I have found to be associated with functional amblyopia and that often successfully responded to treatment.

**Important Point**

When commenting on pornography, the United States Supreme Court Justice Potter Stewart famously said that he could not define it, "But I know it when I see it." I like to describe functional amblyopia associated with organic disease as Potter Stewart reversed, "I can define it, but I don't always know it when I see it."

## Occlusion Amblyopia

### Basic Information

An iatrogenic amblyopia can occur in the formerly unaffected eye if it is occluded too long. In general, occlusion amblyopia will not occur until the amblyopia in the previously amblyopic eye has been overcome. Thus the key to preventing occlusion amblyopia is to check the visual acuity at regular intervals while the patient is undergoing amblyopia therapy. Occlusion amblyopia is less common now that many ophthalmologists patch fewer hours a day than was done in the past. Nevertheless, it still can and does occur. Similarly, it can occur with pharmacologic penalization, so regular monitoring of visual acuity is important with that treatment modality.

Even though the development of occlusion amblyopia is typically very disturbing to parents, it is almost always reversible. I always tell parents that near the end of amblyopia management we may need to do some patching of either eye, to be sure that we end up with good visual acuity in both eyes. If parents are forewarned, they are usually more accepting if this need occurs.

If occlusion amblyopia is relatively mild and the patient has a moderate-to-high amount of anisometropia in the previously amblyopic eye, one can often treat occlusion amblyopia by merely discontinuing the occlusion or penalization. If the occlusion amblyopia is more profound and there is not substantial anisometropia, one may need to occlude the previously amblyopic eye.

**Important Point**

If occlusion amblyopia develops before the amblyopic eye has developed normal vision, you should suspect either the coexistence of organic pathology in the eye initially presumed to be amblyopic or that the refractive error in the amblyopic eye is not accurately corrected.

## Ametropic Amblyopia

### Basic Information

Ametropic amblyopia occurs when there is a high refractive error in both eyes that has not been corrected at an early age. It is usually correctable by the wearing of appropriate spectacles. If the refractive error is substantial bilaterally, but

significant anisometropia exists, the visual acuity in one only, the less ametropic eye, may improve with spectacles alone. Occlusion therapy may be needed to treat the amblyopia in the more affected eye. In this situation, one should be particularly attentive to the occurrence of occlusion amblyopia, because it appears to occur more frequently in an eye that previously had ametropic amblyopia.

Ametropic amblyopia is not reported to occur as frequently as unilateral amblyopia. Patients with poor visual acuity in both eyes are frequently brought to the ophthalmologist at a younger age than patients with unilateral amblyopia, because parents note that they are having trouble seeing. The latter may go undetected until a routine vision screening test is performed at an older age.

Ametropic amblyopia may be treated successfully at older ages. I once examined a 29-year-old woman who came from a developing country. She had never had an eye exam and was not bothered by her blurred vision, because in her native country she had no need to read or drive a car. She came to my college town with her husband who was a graduate student, and now she found that she had need for better vision. I found her to have a best corrected visual acuity of 20/200 with her refractive correction of approximately 8 D of myopia and 7 D of with the rule astigmatism, which was essentially the same in both eyes. I prescribed her full correction and followed her monthly. At each visit her visual acuity improved approximately one line from that of the prior exam, until after approximately 8 months she attained 20/25. At that time she returned to her home country and I had no further follow-up.

## Amblyopia Prophylaxis

### Important Point

*Any Child Up to About 6 Years of Age Who Sustains an Injury to an Eye That Results in Occlusion of the Visual Axis, Either By a Hyphema or a Surgical Dressing, Is at Risk for Developing Amblyopia*

Similarly, any child who sustains a corneal laceration, or develops a partial or traumatic cataract, is at risk for developing functional amblyopia superimposed on whatever visual deficit may be caused by the structural abnormality. It is disheartening to see a patient several years after successful treatment of an ocular injury in early childhood, only to find that there is profound amblyopia that could have been prevented. All too often, follow-up is not continued long enough to adequately document visual acuity after successful treatment of the initial ocular trauma in preliterate children. It has been my practice to follow visual function closely in preliterate children after serious ocular trauma. As soon as the eye is healed from the trauma, I consider part-time occlusion of the uninjured eye if they are preliterate and there is any suggestion that the visual acuity is decreased in the injured eye. All such children should be followed until they are old enough to perform objective visual acuity tests..

Ametropic amblyopia may respond well to treatment at an older age.

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# Esotropia

# 5

*Don't cross your eyes or they'll get stuck that way.*
*—Common bubbe-meise, or grandmother's fable.*

## Overview

The term *esotropia* is derived from the Greek, *ésō*, meaning within, and *trópos*, meaning a turn. There are countless jokes about crossed eyes, none of which will be repeated here. But it is no joke that having an esotropia (ET) has a subtle and not so subtle effect on the impression someone makes. Diane Arbus was a photographer who became famous by photographing marginal people, many of whom seemed somehow off-putting or surreal. Scrutiny of her work reveals that a disproportionately large number of subjects who just seemed "unsettling" to view had strabismus, with ET being substantially more frequent than exotropia (XT). Although permission was not obtainable for me to reproduce any of her photographs, interested readers can search the Web for examples. Search for "Girl With Cigar in Washington Square Park," "A Young Brooklyn Couple Going for a Sunday Outing," and "Young Couple on a Bench in Washington Square Park."

Esotropia is intimately linked to accommodation and accommodative convergence. This relationship is important in all forms of ET, and is not limited to pure accommodative ET. Proper management of hyperopia and control of accommodation are crucial in treating all forms of ET. You need to know the full amount of hyperopia as well as have an estimate of the accommodation-to-accommodative convergence (AC/A) ratio.

## Examination

> It's a good thing says Frankie so I'll know he's talking to me because he has crossed eyes and you never know who he is looking at … He says it's a gift to have crossed eyes because you're like a god looking two ways at once and if you had crossed eyes in the ancient Roman times you had no problem getting a good job.
>
> —Frank McCourt, *Angela's Ashes.*

## Refraction

### Important Point

*If There Is one Mantra That Needs Constant Repetition in Managing ET It Is "Refract Often"*

If doing well, a child with ET needs a cycloplegic refraction once a year. Detecting a change in anisometropia or hyperopia and making appropriate changes in the child's refractive management may prevent a deterioration in alignment or recurrence of amblyopia before it occurs. Similarly, if ever a patient experiences a recurrence of amblyopia or deterioration of alignment, she should have a cycloplegic refraction even if her current prescription is less than a year old.

Refract often.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_5

## Basic Information

*All Children with ET Need to Be Refracted After Full Cycloplegia*

Years ago it was standard to do atropine refractions in all esotropic children. Subsequent studies have shown that proper use of cyclopentolate can achieve similar results in most children. This usually involves cyclopentolate 1% administered twice, or cyclopentolate 2% administered once for most children. Children with blue eyes may achieve adequate cycloplegia with cyclopentolate 1% given once, and children with darkly pigmented irides may need tropicamide as a supplement to cyclopentolate. However, as discussed in Chap. 3, the cycloplegic effect of cyclopentolate peaks after the mydriatic effect and takes a full 40 min on average; it can take up to 75 min in darkly pigmented individuals. Many ophthalmologists refract esotropic children prior to this peak of the cycloplegic effect, because they either routinely wait less than 40 min or use mydriasis as a surrogate for assessing cycloplegia. They consequently may not uncover all of the child's hyperopia. It is important to note during retinoscopy if cycloplegia is complete, as evidenced by a stable endpoint of the retinoscopic reflex. If cycloplegia is not complete, waiting longer for the drops to reach peak effect, supplementing the drops with additional cycloplegic agents, or doing a repeat refraction using atropine is indicated. This is particularly true if glasses were prescribed and there is a residual deviation. Be advised that cyclopentolate can have serious central nervous system effects, particularly in children with seizures or other neurologic issues. Cyclopentolate 2% is decidedly more likely to cause problems, in small children, than cyclopentolate 1% administered twice. It may seem that one drop of the 2% concentration delivers as much medication as the 1% administered twice, yet it does not. It is the concentration of the drop itself that determines the systemic diffusion gradient, and consequently the 2% concentration does deliver more medication systemically.

## Important Point

Cyclopentolate 2% can have serious central nervous system side effects in children who are small, have seizures, or have other neurologic problems.

## Important Point

Cyclopentolate takes 40 min to achieve its peak cycloplegic effect.

## Pearl

*A Wide "D-Segment" Is Preferable to an Executive Bifocal for Esotropes with a High AC/A Ratio*

Older textbooks suggest that strabismic children needing a bifocal should be put in an executive-style segment. This was predicated on the idea that a round-top segment is not adequate because it is harder for the optician to determine how to center the segment when the corrected near alignment is not known at the time of dispensing. However, a D-style segment is a flat-top segment that can come in widths almost equal to the horizontal dimension of the lens. D-segments are substantially thinner and lighter than an executive segment with the same prescription. Fig. 5.1 shows two pairs of spectacles with identical prescription +5.00 OU with a +2.50 add. The edge thickness of the D-segment lens is 6 mm as compared to 12 mm for the executive seg, and the weight is 35 g versus 60 g, respectively.

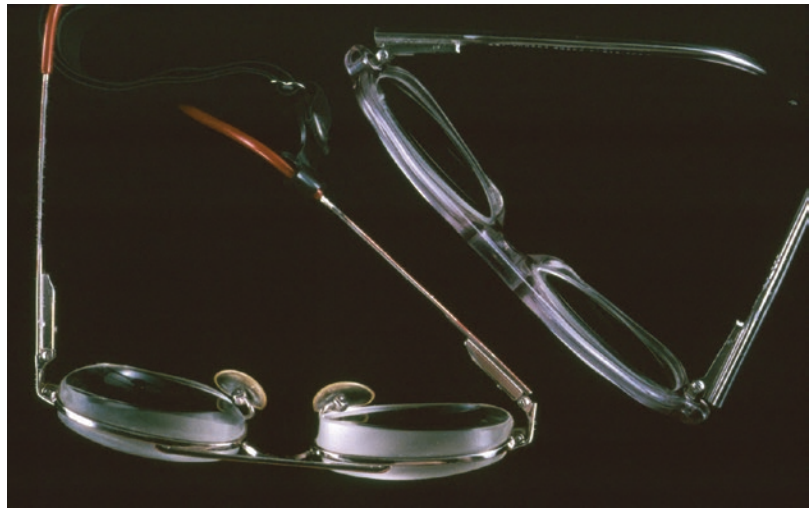


**Fig. 5.1** Two pair of spectacles with identical prescriptions, +5.00sph OU with add +2.50. The pair on the lower left is an executive bifocal which is thicker and heavier than the pair on the upper right, which is a wide D-segment

## Pearl

*Start with a Bifocal Strength of +2.50 Diopters for Young Children*

Commonly, I hear that colleagues routinely order a +3.00 add for esotropic children with a high AC/A. I prefer a +2.50 diopters add for several reasons. If the distance refraction is spot-on, a +3.00 diopters add will fog anything farther than one-third meter from the child's eyes. This may be a bit too close for many near tasks, and may result in the child looking over the seg. Prescribing +2.50 diopters allows for a small error in my distance retinoscopy (yes, this can occur) and still not have the child fogged for near. Also, if the child starts to lose hyperopia, a +3.00 add will become "too strong" sooner. Thus, I start with a +2.50 add, and if a follow-up exam reveals that I need a little more correction at near, I can always order more correction. I almost never have had to do that.

## Basic Information

*You Need to Have at Least a Qualitative Assessment of the AC/A Ratio to Properly Manage Esotropes*

There are two conceptually different methods for calculating the AC/A—the gradient method and the heterophoria method. They give quantitatively different results but usually are qualitatively similar, e.g., high, low, or normal. The gradient method involves measuring the alignment before and after the introduction of either 1–2 diopters of minus lens power for a distance measurement or +2 or +3 diopters of added plus lenses for a near measurement. Relying on the assumption that the patient's additional accommodation is equal to the amount of minus lens power added at distance, or that the relaxation of accommodation at near is equal to the added plus lens power at near, and attributing the change in deviation to that change in accommodation, one can calculate how much accommodative convergence occurs per diopter of accommodation. For example, if a patient has a 25 Δ ET at both 6 m and 40 Δ at one-third meter, and if the near deviation decreases to 10Δ when measured through +3 diopters lenses, the heterophoria AC/A is 10:1. This is arrived at by dividing the change in the deviation from the added plus lenses, 30 Δ, by 3, the added plus power. The heterophoria method involves comparing a distance deviation to a near

deviation and is based on the assumption that the difference is completely due to accommodative convergence. If the distance and near deviation are the same, the AC/A ratio by this method equals the interpupillary distance (ipd) in cm. The derivation of this can be found in most comprehensive strabismus textbooks. If the near deviation differs from the distance deviation, the AC/A ratio equals the difference between the distance and near deviation divided by the dioptric equivalent of the near testing distance. This would be 3 if the near test distance was one-third meter, which is then added to the ipd in centimeters. If the near deviation is more esotropic or less exotropic than the distance finding, the quotient of the fraction should be added to the ipd. If the near deviation is less esotropic or more exotropic than the distance finding, the quotient should be subtracted from the ipd. For the same hypothetical patient described above, if the ipd is 5 cm, the heterophoria AC/A is also 10. This is arrived at by adding the difference between the distance and near ET, 15 Δ, by the dioptric equivalent of the near testing distance, 3, which equals 5. This is added to the ipd of 5, for a final result of an AC/A of 10:1. In practice, you can assume that the ipd is about 5 cm in children and 6 cm in adults.

### Important Point

An AC/A can be said to be "perfect" if the amount of accommodative convergence is exactly equal to the amount needed to maintain the same alignment at distance and near fixation. In that case it would be equal to the ipd in centimeters (about 5 for a child and 6 for an adult). In fact, the average AC/A in normal subjects is insufficient to provide for that amount of convergence. This is in part why many normal people have a small exophoria at near. Established normal values for the AC/A are just under 4/1 with the heterophoria method and just over 2/1with the gradient method.

### Question

Which is the preferred way to measure the AC/A, and why do the two methods give somewhat differing results?


### Reply

In clinical settings, both the gradient and heterophoria methods produce what is called a stimulus AC/A. This means that the answer is predicated on the assumption that the change in accommodation is exactly equal to the stimulus used. For example, a distance gradient AC/A assumes that if −2 diopters lenses were used, the patient in fact accommodates 2 diopters more; if +3 diopters lenses were used at near, it is assumed that the patient relaxed accommodation by 3 diopters. This may not be what happens. A young child may not exert the expected accommodative effort if tested through added minus lenses at distance, and a presbyope may not be able to. On the other hand, a heterophoria AC/A assumes that all differences in the distance and near measurement are due to accommodative convergence and do not take into account the role of other factors like the *Scobee phenomenon* (ScPh, or *tenacious proximal fusion*) in exotropes. (see Chap. 6). Unlike stimulus AC/A ratio calculation, a response AC/A is calculated based on measurements using laboratory techniques that actually determine how much accommodation has occurred. This is outside the scope of clinical use. Despite their limitations, both gradient and heterophoria methods for calculating AC/A have their uses. If, for example, I want to know how much reduction I will get in an XT using over-minus lens therapy, I am interested in a gradient method AC/A using minus lenses at distance. If I want to know whether glasses, perhaps with a bifocal, will be useful managing

an esotrope, I am interested in a heterophoria AC/A and juxtaposing the answer with the refractive error.

Both Gradient and Heterophoria AC/A calculations have their uses.

### Pearl

*Adults with a History of ET May Continue to Need Cycloplegic Refractions*

Many ophthalmologists use cycloplegia for refracting esotropic children, but start doing a manifest (dry) refraction when they become adults. As long as they are doing well with that approach, it may be satisfactory. But some adults with a history of accommodative ET are atypical in how they relax their accommodation during a refraction, and cycloplegia may be needed to uncover their full amount of hyperopia. I once was referred a 45-year-old woman who had shown a progressive increase in her ET over the prior 10 years. In addition, she was asthenopic. As a child she had worn hyperopic glasses for ET. For each of the prior 10 years she had undergone a refraction, each time by a different ophthalmologist. For every examination the refraction was manifest (dry), uncovering about 3 diopters of hyperopia OU. She was, however, dilated for fundoscopy. On my exam she had about 30 Δ of ET. Refracting her with cycloplegia I found 2.50 diopters of residual uncorrected hyperopia OU. Incrementally increasing her hyperopic correction eliminated her ET and asthenopia.

### Advanced Information

*Start Paying Attention to Accommodative Amplitudes in Esotropes in Their Mid-30s and Do Not Delay Pushing Plus or Ordering Bifocals*

Although many people know that bifocals or reading glasses are inevitable, they also often like to delay passing that aging milestone as long as possible. Patients with a history of ET have a tendency to decompensate their alignment as they approach presbyopia [1]. Presumably, as accommodative amplitudes decrease, accommodative effort increases, resulting in more accommodative convergence. I recommend routinely measuring accommodative amplitudes in esotropic patients when they reach their middle 30s, and ordering a reading add as soon as the amplitudes become borderline. I find most patients accept this if it is explained as a way to ward off a recurrence of the ET.

> I had friends, once … who had a daughter. Beautiful, beautiful child — but all you had to do was take one look at her to see there was something wrong. She had one crossed eye … Her left eye was locked in place, staring only to the right … The parents knew, but they kept saying it would fix itself. She was very young, this child — not yet two years old. But nothing was done … By the time she was eighteen, the eye was so badly damaged it had to be removed. God's will.
>
> No, dammit … Not God's will. Parental stupidity … If they had taken her to a specialist before she was three or four years old, the could have been corrected and today she would be whole.
>
> —Timothy Findley, *Spadework*

## Motility Exam

### Basic Information

*Pseudostrabismus*

When I observe completely normal findings on a child suspected of having an ET, I tread cautiously in attributing it to pseudostrabismus. I do keep a photograph of a child with pseudostrabismus in my exam room, show it to the parents, and point out how the symmetric corneal light reflexes in the photograph mean the eyes are aligned. I juxtapose this with a photograph of another baby who actually has an ET, and point out the difference. If this discussion produces an

"aha" reaction in the parents, and they feel I have shown them what they are seeing in their child, I am more comfortable telling them to return only if they observe changes. However, if they feel this is not the same thing they are observing, I always schedule a follow-up examination 4–6 months later. I tell them that if the problem stops occurring, they can cancel the appointment. In my discussion I tell them that I can say several things for certain: (1) The eyes are structurally healthy, meaning no birth defects, tumors, or other scary things. (2) There is no significant refractive error, meaning no optical distortion for which glasses would be needed. (3) At least during my examination, the eye did not cross, even though at casual glance it may have appeared to be misaligned. But I also say that the child may be in the very early stages of developing an eye muscle problem and is merely controlling it well. Even if that were the case, right now it is not a problem. But it should be followed up if it continues.

### Question

Why do you even schedule a follow-up examination if the first examination was normal, and you thought the child had pseudostrabismus?


### Reply

Over the years I have seen a number of children who previously had been diagnosed with pseudostrabismus, who presented later with obvious strabismus. Recently I examined a 2-year-old girl who was found to have pseudostrabismus at 6 months of age by an excellent pediatric ophthalmologist and orthoptist. She had a 70 Δ ET with all the concurrent findings to suggest infantile ET, e.g., dissociated vertical divergence (DVD), optokinetic nystagmus (OKN) asymmetry, and true inferior oblique overaction. Although this stunning case is an outlier, patients with smaller esotropic angles due to accommodative ET are not infrequently suspected as having pseudostrabismus on their initial exam.

### Pearl

*Two Additional Tests to Always Do in Cases of Suspected Pseudostrabismus*

Getting an infant to track a vertically moving OKN target will stimulate them to accommodate and cause a well-controlled latent accommodative ET to become manifest. Whenever I suspect a child of having pseudostrabismus, I get them to watch an OKN drum with the stripes horizontal (rotating vertically). When I start to see the child make vertical tracking movements, I look closely at the light reflex. In cases of a well-controlled early accommodative ET, I will see one eye progressively cross. I also perform dynamic retinoscopy according to the technique described earlier for the reasons outlined in Chap. 2.

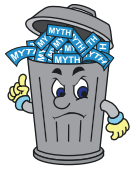
> If a cross-eye person looks at you, you will have ill-luck all day,
> for such people can see right through you and know your thoughts.
> —Iona Opie and Moira Tatem
> *A Dictionary of Superstitions (Oxford Quick Reference).*

## Treatment

### Basic Information

*There Are two Types of ET That Are Related to Accommodation, and They Should Be Thought of Differently*

I like to think of one as being caused by abnormal anatomy of the eye (high hyperopia, which after all is a function of the size and shape of the eye) but with normal physiology—specifically a normal coupling of accommodation to convergence. The second has normal anatomy (normal refractive error) but abnormal physiology—a high AC/A. We would be better served by referring to the former as a refractive ET and the latter as an accommodative ET. However, historically both have been called accommodative ET. Finally, there are patients who have a combination of both, e.g., high hyperopia and a high AC/A.




## Myth

All ET presenting before 6 months of age is infantile ET and requires surgery.

## Fact

Infantile accommodative ET is a real entity. If an infant with ET has more than about +2.50 diopters of hyperopia, spectacles should be instituted. In my experience, infantile refractive ET is much more common than infantile ET due to a high AC/A ratio, but both do occur. So it is crucial to obtain a distance as well as near measurement and make an assessment of the AC/A before deciding that a low hyperopic refractive error does not need to be corrected in an esotropic infant.

## Question

I examined a 6-month-old boy with 40 prism diopters (Δ) of ET and a +6.00 spherical refractive error OU. I put him in spectacles and the deviation did not decrease. I plan to operate. Should I leave him in glasses? I assume not, as they make no difference.


## Reply

I think it is a big mistake to discontinue the glasses for the following reason. Right now the glasses are probably not making a difference because the child is young and simply not accommodating very much. But as he gets a bit older and starts to accommodate to clear up the visual image, accommodative convergence will kick in. If this child has a normal AC/A of about 5, his alignment will vary by about 40 Δ, depending on whether he is accommodating versus relaxing accommodation. This will introduce considerable instability in his oculomotor system.

## Question

I care for a 5-year-old boy with accommodative ET. He is well controlled with his single-vision glasses that incorporate his full plus. When should I start decreasing his hyperopic correction? What is your approach for weaning such children out of glasses?


## Reply

My approach is not age specific, but more tailored to clinical findings. After a year of good alignment with spectacles, I try to make an assessment as to how the child would fare if the plus were decreased on each subsequent exam. In attentive cooperative patients who are fusing, I test their divergence amplitudes with base in prism. I juxtapose that result with their AC/A to determine if, and by how much, the plus can be decreased. For example, if divergence amplitudes are 8 Δ base-in-prism (BI), and the AC/A is 3, I feel I can easily cut the plus by 1 diopter. This would leave him a divergence fusional reserve of 5. This method is only applicable to patients with good fusion. For all monofixators and patients who are insufficiently cooperative for divergence amplitude testing, I assess their alignment and control with −0.50 diopters to −1.00 diopters held in front of their spectacles. If they maintain good alignment (less than 10 Δ of ET) and/or their control of a latent deviation still seems good, I decrease their plus accordingly. In practice I find I use this latter method more often than the former, due to patient selection criteria. I care for more monofixators than bifoveal fusers in this population. Other colleagues use an arbitrary formula and cut the plus by a standard amount at regular age intervals. I have no experience with that approach.

## Question

I examined a 2-year-old girl with an ET measuring 15 Δ at distance and 30 Δ at near. Her refractive error was +3.00 SPH (sphere) OU. I calculated that with this AC/A of 10 using the heterophoria method, and 3 diopters of hyperopia, single-vision spectacles should correct the 30 Δ at near. However, when she returned in the glasses, she was orthophoric at distance and still had about 15 Δ of near ET. The near deviation does decrease with added plus lenses at near, so I know it is accommodative. Why were my calculations not predictive of her response at near?


## Reply

This is actually a common scenario. I find that if there is a significant convergence excess due to a high AC/A, single-vision lenses usually do not correct the deviation at near. As discussed earlier, our clinical calculations of the AC/A are based on the assumption that accommodation is equal to the stimulus. In this situation, the child initially was probably not accommodating the full 6 diopters when shown a near target (3 diopters for the hyperopia and 3 diopters for the extra accommodation needed at 1/3 meter). She was probably accepting some degree of blur.

## Question

In the aforementioned example, what are my treatment options when the child is seen for the first time in glasses and the residual ET at near observed? The parents just spent hundreds of dollars on the glasses, and their insurance will not pay for another prescription this soon. How should I handle this?


## Reply

It is not widely known, but Fresnel lenses come in spherical powers as well as prism diopters. It is very easy and inexpensive to make a bifocal with a +2.50 diopters or +3.00 diopters spherical Fresnel lens. You can use this to temporize until the child can obtain another pair of glasses. Alternatively, the child can be put on phospholine iodide (PI) drops as a temporizing measure until new glasses can be obtained.

## Question

Do you prescribe a bifocal if it decreases the near deviation but not to orthophoria and does not result in stereopsis, and if so, why?


## Reply

I prescribe a bifocal if doing so decreases the near deviation to less than 10 Δ, even if there is no increase in stereopsis. I know that this approach is controversial but it makes sense to me. Stereopsis is not the only test of binocular function, albeit one of the harder ones to obtain. However, most patients will show a more normal binocular response with Bagolini lens test if their ET is decreased to less than 10 Δ, and this is a sign of peripheral fusion. This goes along with an increase in the size of the binocular visual field, which I presume to be beneficial, and may help contribute to stability of alignment [2, 3]. In addition, many (not all) of these children will go out of their way to raise their chin when looking at a near object, in order to use the bifocal segment. To me this means that on some level they functionally benefit.

Stereopsis is not the only important test of binocularity.

## Question

I prescribed a bifocal for a 3-year-old girl with a high AC/A ET. Although her near deviation is nil when she looks through the add, she does not spontaneously use it. I think perhaps the segment is a bit lower than I prescribed. What should I do?


## Reply

This problem is more common when the bifocal is not positioned properly. Nevertheless, you can often get a child in the habit of spontaneous bifocal use with pharmacologic behavior modification. Put the child on atropine 1% drops or ointment once daily for about a week; the effect will last well into the next week after the medication is discontinued. Typically, the child will use the bifocal well while they are cyclopleged, and most of the time they continue to do so after the medication has worn off, unless the bifocal is substantially too low. Although most colleagues specify bifocal height when writing a prescription in this setting, there is much additional information you can give the optician to facilitate a successful outcome. Frame selection is important. Ideally, the frame should be of a style so the pupil is approximately in the geometric center of the lens, both vertically and horizontally (Fig. 5.2). The frames should have bows that wrap around the ears (called a riding bow) to prevent slippage and are preferable to bows that have little support behind the ear. It is desirable to have a frame with a solid nose piece such as a form-fit or saddle bridge. Frames with non-joined adjustable nose pads are not satisfactory, as they rarely stay in place firmly enough, although opticians often recommend them on the assumption that they can be adjusted to make up for fitting errors.

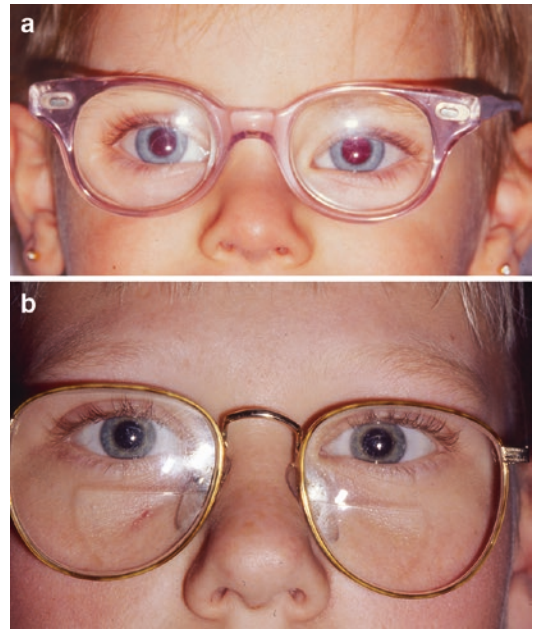



**Fig. 5.2** (**a**) A good frame style for an infant. The pupil is near the geometric center of the lens. (**b**) A poor frame choice for a child in need of a bifocal. The frame has the pupil positioned disproportionately near the top of the lens

Finally, I like to avoid frames with a sweat bar, as that often pushes the frame down on the child's face. I typically order a wide D segment to bisect the pupil for a child's first pair of bifocals. But as they get older, and are habituated to using it, I often order it positioned at the bottom of the pupil, and even later at the lower eyelid margin. I have a handout (Fig. 5.3) on which I can check the desired position of the segment and also instruct the optician on the aforementioned fitting parameters. The most important entry on the form is near the bottom. It reads, "If you are unable to provide a frame which approximates the above criteria, please do not dispense."

There is more to fitting a bifocal than just specifying segment height.

**Fig. 5.3** Handout giving opticians instructions for frame selection and positioning of the bifocal segment

# U. W. Pediatric Eye Clinic

## To: Opticians and Optometrists

The need for bifocals in accommodative strabismus problems carries some specific and unique requirements which are different than the usual adult applications.

Please dispense according to the following recommendations:

The top edge of the bifocal segments should be place so as to:

- ☐ Bisect both pupils
- ☐ Be at the bottom of both pupils
- ☐ Be at the level of the lower lid margin

when the eyes are looking at a distant object in the straight ahead position.

A flat top segment such as a wide "D" or an executive bifocal should be used.

Frames with non-joined adjustable nose pads are not satisfactory as they rarely stay in place firmly enough. This results in the bifocal segment being positioned too low. A solid nose piece such as a formfit or a saddle bridge is recommended. The Unifit is also acceptable.

A style should be chosen in which the eye is near the geometric center of the lens, both vertically and horizontally.

It is often desirable to dispense a frame with a single bridge. Frames with a sweat bar often ride too low, as the sweat bar causes the spectables to be pushed down. Also this type of frame style usually results in the visual axis being substantially above the geometric center of the lens. Please do not dispense a frame with a sweat bar unless the eye is near the geometric center of the lens, both vertically and horizontally when the frame is in place.

Bows that have a wrap-around effect such as a riding bow, are preferable to blows that have little support behind the ear. It is important that the spectacles not slip down.

**If you are unable to provide a frame which approximates the above criteria, please do not dispense.**

Your cooperation is appreciated.

### Question

Are you concerned that having a child use a bifocal during their growth years may lead to ciliary muscle atrophy and premature presbyopia?


### Reply

von Noorden has observed that some esotropic children who had worn bifocals during childhood had decreased accommodative amplitudes when he tested them in their early 20s [4]. This raised the question that childhood bifocal wear might lead to premature presbyopia. But another explanation seems likely. Costenbader observed that some patients with ET have hypoaccommodation [5]. He speculated that because it is the effort to accommodate that drives convergence, hypoaccommodation could be associated with ET secondary to the excessive innervation needed to accommodate. These patients might show premature presbyopia regardless of bifocal wear in childhood. Although it is not a proven conclusion, I think it is unlikely that bifocal wear for ET causes premature presbyopia. The vast majority of patients who wear a bifocal in childhood for ET do not show problems with accommodation after they have outgrown the need for the bifocal.

### Question

I put a 4-year-old child in a +2.50 add for a high AC/A ET. How long should I keep her in a bifocal, and what protocol should I follow for weaning out of the add?


## Reply

I have a strong (but unproven) impression that a child is more likely to successfully outgrow the need of a bifocal if it is incrementally decreased in strength a little bit at a time rather than going "cold turkey" from a +2.50 add to no bifocal all at once. I keep a child in the full-strength bifocal for 2–3 years and then try to decrease its strength. On each subsequent yearly exam, I see the effect of −0.50 or −0.75 diopters held over the bifocal on the alignment at near. If control remains good, and the increase in the angle is small (usually 10 Δ or less), I decrease the bifocal strength accordingly. If I have a hypothetical patient who is controlling the ET well, and appears to be able to take a decrease in their plus correction of 0.50 diopters at both distance and near, my preference is to try and decrease the difference between the distance and near-optical correction. Rather than decreasing the distance correction by 0.50 diopters and leaving the bifocal power unchanged, I would opt for leaving the distance correction the same and decreasing the bifocal power. That way I am working toward eliminating the need for different correction distance and near.

Some people report good results using an arbitrary approach like decreasing the power of the bifocal by 0.50 or 0.75 diopters each year, regardless of what effect that appears to have during the office exam. I have not been comfortable with that approach.

Perhaps the most scientific approach is to measure the patient's divergence amplitudes at near through the bifocal segment. Then, armed with an estimate of the AC/A, you can calculate how much the bifocal power can be decreased and still leave the patient with some fusional reserve. This method is accurate, but a bit more involved than the simple observational method described above, and in my experience is usually not necessary.

## Question

Can I safely switch my well-controlled refractive esotropes to contact lenses when they become teenagers. Similarly, do refractive esotropes have problems if they alternate between contact lenses for sports and spectacles the rest of the time?


## Reply

In theory, contact lenses create an increased accommodative demand (compared to spectacles) because of the difference in vertex distance, and may result in poorer alignment for refractive esotropes. In practice, I have not found this to be the case. When I care for a patient who makes this switch, I evaluate their alignment about 3 months after they start wearing contact lenses. I cannot recall a single refractive esotrope who was well controlled in spectacles and who had poorer alignment in contact lenses due to increased accommodative demand. Similarly, I have not seen patients deteriorate as a result of switching back and forth between spectacles and intermittently wearing contact lenses. However, I do find that often refractive esotropes with moderate-to-high astigmatism are not as well aligned with toric soft contact lenses as they are with spectacles. The problem is that the toric lenses often do not adequately correct for the astigmatism. The only way you can determine this is to retinoscopy the patient over his or her contact lenses (with cycloplegia) and see if there is uncorrected astigmatism. Admittedly, this is time consuming, but it is necessary.

## Pearl

If a refractive esotrope has poorer alignment with contact lenses than with spectacles of the same power, consider that the contact lenses do not adequately correct for

astigmatism. This is more likely the cause than the increased accommodative demand with contact lenses. You need to refract the patient over the contact lenses and specifically look for uncorrected astigmatism.

## Question

What can I offer to my teenage patients who are well controlled with bifocals, but want to wear contact lenses? Should I operate to decrease their AC/A and eliminate their need for a bifocal?


## Reply

My bias is to not operate on teenagers solely for the purpose of getting them out of bifocals. It stems from my own experience, which indicated that almost all will outgrow the need for bifocals. I reviewed my experience of 252 patients with a high AC/A ET who were initially controlled with bifocals [6]. In my analysis, 140 (56%) remained aligned with bifocals; 112 (44%) lost control at distance and underwent surgery. Of the 140 who stayed aligned in bifocals, 138 (99%) normalized their AC/A and no longer needed a bifocal by 18 years of age. But notably, 32 (23%) of the patients who did not need a bifocal by 18 years of age still needed it by 13 years of age. So if you do operate on an early teenager for the purpose of eliminating the need for a bifocal, you should be aware that almost all of them will outgrow that need if given a bit more time.

## Advanced Information

*Phospholine Iodide (PI) Is an Underutilized Arrow in Your Quiver for Treating ET*

I find PI useful in three clinical settings [7]. If a child has a small angle variable ET, and relatively low hyperopia, a diagnostic trial of PI may tell if the strabismus is accommodative. I will put a child on one drop of PI at bedtime and reevaluate them in several weeks. If the deviation is less, I can comfortably order glasses, knowing that I am dealing with an accommodative ET. Secondly, there are some children with relatively well-controlled ET who break down only occasionally, yet are symptomatic—sometimes diplopic. If the refractive error is small, one may want to keep such a child out of glasses. I will put them on PI at bedtime for a month. If that eliminates the ET, I will gradually taper the drops by going to every other night for a month or so, then every third night, and finally stopping. I have seen children in whom this treatment program sufficiently improved their fusion that they obtained a long-term cure from one or several treatment courses with PI. Finally, I find PI extremely useful in treating postoperative ET. It seems equally effective in treating prolonged overcorrection after surgery for XT, and undercorrection after surgery for ET. Moreover, it is often effective in esotropic patients who did not have an accommodative ET initially. Even nonaccommodative esotropes have an AC/A ratio (presumably normal) and PI will decrease the accommodative convergence. Side effects of PI can include brow ache and fussiness. I find that giving the drop at bedtime minimizes this. My preference is to use 0.06% PI; however, currently it is only available in a 0.125% concentration. Thus I have established a relationship with a pharmacy that will dilute the commercial preparation to my desired strength. I find that the aforementioned side effects are less with the 0.06% concentration, and it is usually effective at that strength. If it is showing some effect, but the magnitude is insufficient, I prescribe the 0.06% concentration twice daily. Iris cysts are reported to occur with PI. These can be prevented with concurrent use of phenylephrine drops. However, I have never seen iris cysts occur with the 0.06% concentration. Finally, patients on PI need to be warned about the drop interfering with the breakdown of succinyl choline, which should be avoided in patients on PI. Fig. 5.4 is the handout I give to patients for whom I prescribe PI.

**Fig. 5.4** Handout for patients using phospholine iodide

# U. W. Pediatric Eye Clinic

## Phospholine Iodide

**Your child has been placed on Phospholine Iodide, which is a drop that often helps straighten eyes that cross. One drop is to be placed in each of your child's eyes prior to bedtime.**

**There are several things you should be aware of with respect to the use of Phospholine Iodide.**

1. **The drops works in the opposite manner of a dilating drop and consequently**

**makes the pupils small. Frequently, it makes the pupils unequally small. It is of no concern if one pupil is smaller than the other while your child is using Phospholine Iodide.**

2. **Phospholine Iodide drops interfere with the body's ability to metabolize,**

**Succinyl Choline, one specific medication sometimes used during the general anesthesia. If for any reason your child needs to undergo a surgical procedure while using Phospholine Iodide, be sure to tell the anesthesiologist that your child is taking Phospholine Iodide. If they are aware of the use of Phospholine Iodide, they will probably use an anesthetic agent other than Succinyl Choline, and there should be no problem elated to the use of Phospholine Iodide.**

3. **On rare occasions, a small child may develop an upset stomach or diarrhea**

**while using Phospholine Iodide. If this occurs, discontinue the use of the drops and contact your ophthalmologist.**

**Burton J. Kushner, M.D.**

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### Basic Information

*Reevaluate a Patient About 4 Weeks After Putting Them in Glasses for ET with an Accommodative Component*

That is usually sufficient time to determine the effect of the glasses, before deciding on the next treatment step.

### Surgery for Esotropia

> Of goggle eyes: If yet he begynne to goggle, then set the cradell after such a fourme that the light may be on the contrary side: that is, on the same side from whence he turneth hys eyes, so that for desire of lyghte he maye directed them to the same part … .
>
> —Thomas Phaer,
> *The Boke of Chyldren* (1544)

## Basic Information

*The Optimum Time to Operate on Infantile ET Is An Evolving Issue*

One large meta-analysis found that sensory results are better if good alignment is obtained prior to 24 months of age as opposed to later [8]; however, it found no difference in outcomes if alignment was obtained by 6 months, 12 months, or prior to 24 months of age, respectively. At best, however, these patients achieved only subnormal fusion. In spite of this, many pediatric ophthalmologists prefer to do surgery at or shortly after 6 months of age due to a belief that the sensory outcome will be better. Another study found that high-grade stereopsis could be obtained in some patients with surgery between 13 and 19 weeks of age [9]. However, another study involving multiple surgeons did not find a benefit to operating this early [10].

## Question

Why then is this issue far from settled?


## Reply

The argument in favor of very early surgery (prior to 6 months of age) is based on the assumption that if satisfactory alignment is obtained with the early surgical procedure, there will be a better sensory outcome. Thus far, there has yet to be a sufficiently large study that addresses the success rate of obtaining good alignment with very early surgery, and there is theoretical reason to believe that very early surgery may be less predictable. A study by Swan in 1984 compared anatomic landmarks of the extraocular muscles in newborns to older children and adults [11]. He found that the most dramatic changes in the growth of the eye occur in the first 6 months of life, and most of the growth occurs posteriorly. At birth, the insertion of the medial rectus muscle (MR) is almost as far from the limbus as it is in a 2-year-old. However, in the newborn, that insertion may be at the equator. The distance between the MR insertion and equator increases markedly by 2 years of age. Swan concluded by saying. "The predictability and safety of extraocular muscle surgery, therefore, would appear to be greater if the surgical correction could be deferred until … about 6 months of age; however, additional anatomical and clinical studies are needed, especially on eyes of three to five-month-old infants." As of this writing, no one knows the long-term effect on motility of recessing MRs in infants 3 to 5 months old. Moreover, the issue as to the timing of surgery for infantile ET should be viewed in light of the British orthoptists' study that assessed the occurrence of strabismus in infants. In a study by Horwood [12], 214 infants were observed by their mothers, who were orthoptists, for neonatal ocular misalignments for up to 15 years of age. The study concluded that neonatal ocular misalignments occur frequently in the first 2 months of life and usually reflect a normally developing vergence system. Emerging infantile ET was indistinguishable from frequent neonatal ocular misalignments before 2 months of age.

In addition, concern has been raised recently about anesthesia in young children leading to learning and behavior difficulties [13, 14]. Much more needs to be learned about how the length and depth of anesthesia influence these findings. This is probably only a risk in children who undergo multiple anesthetics in infancy, but this has yet to be defined clearly. I believe the optimum age to operate on an infantile esotrope is a matter of weighing the aforementioned tradeoffs. My personal preference, taking these factors into account, is to operate shortly after 6 months of age.

## Question

What role should preoperative alternate occlusion play in treating infantile ET?


## Reply

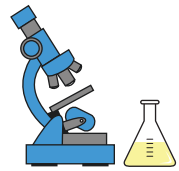
Historically, it has been advocated by some that full-time alternate eye occlusion prior to the time of surgery would "keep the sensory slate clean" and prevent the development of suppression. The one prospective study that looked at this treatment approach found no benefit from preoperative alternate occlusion with respect to alignment at 6 weeks and 12 months after surgery [15]. However, I personally find a role for alternate occlusion according to a different protocol and for different reasons. If a child has a large-angle ET and cross fixates, they typically present with limited abduction. I presume this is due to MR contracture secondary to the limited abduction, and it seems intuitive that varying degrees of contracture can add to the variability of surgery. So if a child does show limited abduction in this setting, I recommend alternate patching for about 2–3 h a day. Typically, within a week or so, abduction improves, which I take to mean the MR contracture is loosening. There are no data about this approach, but it makes sense to me.

## Question

I have many patients whose parents are convinced that there is a surge in passing developmental milestones after surgery for infantile ET, and I have concurred. But I have also seen this after surgery that left the child moderately undercorrected or overcorrected. Is this possible, and if so why does this occur?


## Reply

As stated above, many children with infantile ET cross fixate and develop MR contracture. Or, they may have nystagmus that increases in abduction. In either case they fixate with a face turn toward the fixing eye, and their nose blocks their nasal field. I believe that this blockage of almost half of the visual environment limits an infant's exploring his or her environment. I actually had a patient who stood up for the for first time and took his first step on the day after surgery for infantile ET. These are milestones that are typically months apart. To understand this better, do the experiment below.



## Try This Experiment

Close one eye and fixate on a distant object with the open eye. Now turn your head about 30–40° toward the open eye, and to simulate the alternate suppression that accompanies infantile ET, close the other eye. Note how your nose blocks your nasal visual field. Now imagine you are an infant starting to explore the world with that compromised view. It is easy to imagine how elimination of the head turn after surgery for infantile ET can expand an infant's visual world and curiosity, even if she does not obtain bifoveal fusion.

## Basic Information

*For Partly Accommodative ET, Operate for the Nonaccommodative Component at Distance or Near Fixation, Targeting Whichever Angle Is Greater*

This means that you should operate for the deviation found with the patient wearing their full cycloplegic correction. If the near angle exceeds the distance angle, operate for the near angle as measured through the distance correction [16].

## Question

If the patient is hyperopic, why not operate for somewhat more than the deviation obtained with glasses, and plan to cut the plus a bit after surgery if there is an overcorrection?


## Reply

Although at first glance there is appeal in this approach, it does not make sense to me for several reasons. Uncontrolled accommodation is the enemy of success in esotropes. Unless there is minimal hyperopia, the patient will need to stay in glasses after surgery for maintaining a stable angle. There are advocates of operating for various combinations of the accommodative and nonaccommodative component, e.g., an average of the near deviation with and without correction [17], or similar permutations, and they report good results. However, they invariably reported only short-term outcomes. Also, they considered outcomes successful if the patient was taken out of her plus correction postoperatively, often high hyperopia, to control an overcorrection. Although I do not intentionally overcorrect esotropes and cut the plus after surgery, I do manage inadvertent overcorrections by decreasing plus. I found that this rarely results in a long-term success [18]. With the exception of patients with less than 2.5 diopters of hyperopia it was invariably a temporizing measure.

## Question

If you operate for the near angle as measured through the distance correction, do you not fear an overcorrection at distance?


## Reply

Surprisingly, no. When I first went in practice in 1974 I followed Marshall Parks's advice for high AC/A esotropes. He recognized that this subset of esotropes were prone to undercorrection, and he recommended recessing the MRs by an amount appropriate for the distance deviation, and then arbitrarily adding 1 mm of recession to each MR because of the high AC/A. In 1980 I reviewed my results and was surprised to find that not only did I have a high percentage of undercorrections, I observed that the response to surgery (Δ per mm of MR recession) at *distance* was inversely related to the amount of convergence excess [16, 19]. In other words, if one hypothetical patient measured 15 Δ at distance and 25 Δ at near, and another had 15 Δ at distance and 45 Δ at near, both would get bilateral MR recessions of 4.5 mm (3.5 mm for the 15 Δ at distance plus 1 mm for the high AC/A). I found that the surgery might be successful in the first patient but would routinely undercorrect the 15 Δ at distance in the second patient. This led me to a formula for that took the near deviation into account [16]. A 15-year outcome of patients treated this way in a prospective randomized clinical trial showed that only 5% had to stay in a bifocal, 86% had less than 10 Δ of ET, and none was overcorrected [19].

## Advanced Information

*Nonaccommodative Convergence Excess Can Be Treated the Same Way*

Not all esotropes with a near deviation that exceeds the distance have a high AC/A. Uncommonly, there are patients who look exactly like high AC/A esotropes, except that the near deviation does not decrease with additional plus lenses. They are said to have a nonaccommodative convergence excess, for which the pathophysiology is unclear. In my experience this difference is moot, because they can be treated the same as high AC/A esotropes, e.g., operating for the near angle measured through the distance correction.

## Basic Information

*The Optimum Time to Operate on Partly Accommodative ET*

As with infantile ET, studies have shown that the shorter the duration of a constant ET, the better the final sensory results [20]. For me this means that once a child is wearing

full hyperopic correction for at least a month, amblyopia has been overcome, and measurements are accurate and stable, it is time to invite the child to the operating room for surgery on the nonaccommodative component only. This does need to be balanced by the fact that there are a small number of patients who may still be tropic after 1 month of spectacle wear, who may straighten on their own given more time [20]. The first year I was in practice I scheduled a 4-year-old boy for surgery for the 20 Δ ET that he manifested after 1 month of spectacle wear. When I examined him on the day prior to surgery an additional month later, he was straight and fusing. I cancelled his surgery, of course. One needs to temper the above timing recommendations with the fact that you need to be sure that the deviation is not still improving. This is one reason I always re-examined patients the day prior to surgery, and encourage you to do so.

### Question

Why do you feel amblyopia should be completely overcome before surgery? I thought it has been shown that this is not necessary.


### Reply

Traditionally, it was taught that amblyopia should always be overcome prior to surgery. In 1993 a group from Johns Hopkins Wilmer Eye Institute published a study suggesting that this was not necessary [21]. This was in part predicated on the observation that some amblyopes may continue to have improvement in vision after successful alignment. There were also practical benefits by permitting more flexibility with the timing of surgery based on parents' and surgeons' preferences. There is also the theoretical advantage that earlier surgery may result in improved binocularity afterwards. In spite of this, I feel that there are two overriding issues that lead me to prefer completing amblyopia therapy prior to operating. First, I have found that compliance with amblyopia drops off after the eyes are aligned. To the parents, it may look as though the problem has been taken care if the eyes look straight. Unfortunately, some patients are lost to follow-up once the eyes look well aligned. Secondly, in the preliterate child, it may be harder to assess if amblyopia is still present if the eyes are aligned. Although induced tropia testing is useful in this setting, it still requires more cooperation than observing fixation preference in the face of a substantial deviation. In addition, I feel one cannot extrapolate from the studies that show better sensory outcomes with earlier surgery, as they were all done in patients who were not amblyopic. One cannot tell from existing data the degree to which residual amblyopia negates the beneficial effect that earlier surgery has on the sensory outcome. I think that in select circumstances, operating prior to overcoming amblyopia may be acceptable. If there are important personal factors, e.g., imminent loss of health insurance coverage, family situations like the upcoming birth of a sibling, or parents soon to undergo surgery themselves, etc., strabismus surgery while amblyopia is still present may be acceptable. The most important factor for me in deciding to operate early is that the patient and parents fully understand the importance of continued amblyopia therapy after surgery.

### Question

What do you see as the role for the prism adaptation test (PAT) in partly accommodative esotropes both with and without a high AC/A? Didn't the PAT study prove it was helpful?


### Reply

I essentially never use PAT for the following reasons. The original PAT study sought to address two questions [22]. (1) If you prism adapted a patient, he was a "responder"

(e.g., had sensory fusion at the adapted angle), and the angle built during PAT, should you operate for the original angle or the built-up angle? And (2) do patients have a better outcome if they undergo the process of prism adaptation? In my mind the study did not answer either question, although the abstract implies otherwise. With respect to the first question, although there were better outcomes in prism-adapted "responders" who had surgery for the angle to which they built, the results were clearly not statistically significant ($P = 0.23$). However, nowhere in the study or presentation of the data did the words "not statistically significant" appear. Instead it was stated that the results were "significant at the 'point 23 level.'" Most people hearing this interpreted it to mean "significant." With respect to the second question, the study reported better results if PAT was used with a $P$ value of 0.04. However, this was a complicated two-tiered randomization that required some "creativity" in determining the proper control group, and I personally believe that the statistical methodology was flawed. For the treatment group, they used only the prism-adapted responders who had surgery for the built-up angle. This might have been appropriate if they had "proven" that surgery should be based on the built-up angle. But with a $P$ value of 0.23, this was not proven. Had they included all the patients who underwent PAT, the success rate would have been lower and the results not significant. With respect to using PAT of the near angle in high AC/A esotropes, the often-cited study investigating this compared patients undergoing PAT to a control group that had surgery for the distance angle, after PAT but without augmentation [23]. Because of the aforementioned indications that high AC/A esotropes need some augmentation of the surgical dose to account for the high AC/A, it is not surprising that the patients undergoing PAT for the near deviation did better; I think they used the wrong control group. In my own series of patients with convergence excess ET, none of the patients underwent PAT [16, 19].

> In the Middle Ages, the rapidity with which the Plague could spread led to the theory that it was transmitted by eye contact. This led to the practice of scientists wearing blinders. After the discovery that the Plague was in fact caused by a microbe, the theory of spread by eye contact was set aside. However, it is widely felt that the practice of academicians wearing blinders has stood the test of time. —Author unknown.

## Question

I have a patient with 20 Δ of ET. I did PAT, and he built to 35 Δ with fusion. He is now on my surgical schedule, but he stopped wearing the Fresnel prisms due to blur, and his angle is back to 20 Δ. For what angle should I operate.


## Reply

There are no data that I know of that address this question specifically, so I think one needs to do what makes sense. That depends on what you think is happening when a patient builds up their angle with PAT. Some feel prism adaptation unmasks a larger latent deviation that does not become manifest with a standard cover test. If that is indeed the case, then one should operate for the larger deviation. My personal opinion is that prism adaptation, in most cases, "creates" a larger deviation, rather than "unmasks" a larger latent deviation. I feel that this is most often the case in patients with a constant deviation to which they have developed a sensory adaptation. They are accustomed to having the image of regard a certain distance from the fovea, and when a prism moves the image to the fovea, they respond by increasing the deviation to put the image again closer to where they are accustomed to it being. That is different than the larger deviation actually being present but latent. My reason, in part, is based on the observation that a prolonged patch test rarely replicates the larger deviation seen with PAT. In this setting, I think it is important to operate for the deviation the person has at the time of surgery. So, I think he ideally should be left in the Fresnel prism up to the time of surgery, or at least have it reapplied a few days prior to surgery, so he goes into surgery with the

larger deviation. In this case surgery should be done for the larger deviation. I think if such a patient is not left in prism and goes to surgery manifesting the smaller deviation, overcorrection is more likely if the prior larger angle is targeted.

### Question

What is your thinking about posterior fixation for treating convergence excess?


### Reply

I recognize that good results have been reported using posterior fixation in this setting [24]. Specifically, people usually advocate recessing the MRs, targeting the distance deviation, and add bilateral posterior fixation to address the convergence excess. To me this is illogical. It is thought that posterior fixation works by decreasing the muscle's lever arm as the eye moves progressively into its field of action [25]. This effect does not start to occur until the eye is about 30° into the field of action of the muscle. At near fixation, the eyes are each about 9° further into adduction than at distance fixation, due to convergence. At this small angle of movement into adduction, the posterior fixation suture should not have any effect. Nevertheless, it does appear that posterior fixation does add something to a standard recession in this setting, albeit variable and unpredictable [16]. Another mechanism must be at play. I have speculated that the effect is due to differential shortening of the sarcomeres posterior to the posterior fixation suture, which can best be appreciated by referring to Fig. 10.8. However, this effect is a function of how far the eye is rotated into abduction in order to place the posterior fixation suture, and hence is variable and unpredictable. In addition, whenever I have a choice of a reversible or irreversible procedure, I opt for the reversible one. Reversing posterior fixation suture surgery is unpredictable and cannot always be effectively done. Often at the time of reoperation, the entire muscle between the insertion and the posterior fixation suture is atrophic and fibrotic. In other cases it looks like normal muscle. In the former situation, there is no predictable way to reverse the procedure.

### Question

I will be operating on an esotropic child. I do not see over-elevation in adduction (inferior oblique overaction), and there is no V-pattern. There is, however, marked fundus extorsion. Should I weaken the inferior oblique muscles?


### Reply

Currently, there are no good data documenting the appropriateness of prophylactically weakening the inferior oblique muscles in this setting. However, there are some theoretical reasons to suggest that this may be appropriate. One prospective study found that fundus extorsion occurs in advance (sometimes by years) of over-elevation in adduction and a V-pattern, and was highly predictive thereof in infantile ET [26]. Logic might suggest that weakening the inferior oblique muscles would prevent those findings from occurring and perhaps prevent the need for reoperation. If you do choose to weaken the inferior oblique muscles in this setting, you should be sure to recess them rather than do a myectomy. That way the procedure could later be reversed if need be, or the muscles transposed anteriorly if DVD develops.

### Question

I care for a 2-year-old boy who presented with an acute benign L sixth cranial nerve palsy of childhood. The left lateral rectus function completely recovered, but he now has a convergence excess pattern ET. How can this be, and how should he be treated?


## Reply

This is an unusual scenario that I have seen on several occasions. Initially it perplexed me until I saw a child with a known high AC/A accommodative ET who later presented with a classic postviral sixth cranial nerve palsy that subsequently recovered. I am assuming that your patient had a preexisting subclinical ET of a convergence excess pattern, and developed the sixth cranial nerve palsy secondarily. Treating him now as a routine convergence excess ET should be successful.

## Question

I care for a 60-year-old woman who had a long-standing ET for distance only. I recessed one MR. She was initially exophoric after surgery and drifted back to the same preoperative findings. I recessed the other MR, and the same thing happened; she is now the same as before surgery. What should I do next and why did this happen?


## Reply

I think that there are three likely explanations, which require good orbital imaging to sort out. Pulley issues in the form of a degeneration of the connection between the superior rectus muscle and the lateral rectus muscle (LR) will cause the LR to slip inferiorly. This age-related change is different than the *heavy eye syndrome* that occurs in patients with high axial myopia [27]. It can be diagnosed using high-quality coronal orbital imaging, and can be corrected by raising the LR belly to the normal position and fixating it to the sclera to keep it from slipping inferiorly. One nonabsorbable myopexy suture incorporating about one-eighth to one-fourth of the superior aspect of the LR should be fixated to the sclera at about the equator with the LR elevated to its normal position. The second possibility is severe chronic sinus disease causing an inflammation and contracture of the MRs. When this is the cause, forced ductions should be abnormal, but only mildly so. One must have a high index of suspicion to detect the tightness. Treating the sinus disease first is important, or the strabismus will recur after surgery. However, in my experience, managing the sinus disease alone never corrects the strabismus. Finally, this is not an atypical clinical course for patients with a Chiari malformation, especially when the patient is younger than yours. According to Hoyt (Creig Hoyt, M.D., personal communication, 29 Nov 2016), age is a big factor in deciding what type of imaging to obtain. In his review of a large series of Chiari I patients with strabismus, none was more than 40 years of age, and most were children or young adults. His guidelines for imaging patients with a ET at distance but not at near are as follows:

1. If the patient is an older adult, an orbital magnetic resonance image (MRI) may be helpful, but a brain MRI is almost never required.
2. If the patient is a child or young adult, an orbital MRI is almost never helpful but a brain MRI is probably indicated.

## Advanced Information

Heavy eye syndrome, sagging eye syndrome, and divergence insufficiency pattern ET are discussed in detail in Chap. 10.

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# Exotropia

# 6

*The boy had what people termed a traveling eye. As he looked directly at someone, his left eye would often follow some extraneous moving object that might be just to the side … Then the eye would return to the person in front of the boy … It was a cruel thing God had done … people in that part of Virginia thought a traveling eye a sign of a dishonest or inattentive man.*

*—Edward P. Jones, The Known World*

*Divergent strabismus creates the impression of suffering together with absent mindedness*
*—Friedrich Dieffenbach*

## Cultural and Social Implications

In some circumstances, however, the presence of an exotropia (XT) was viewed positively. From the Middle Ages to the present, it has been common for artistic representations of Christ to depict him with an XT (Fig. 6.1). This metaphorically represents a wide or panoramic view. They even chose an actor with an XT to play Christ in the movie *Jesus Christ Superstar.* Although I am unable to reproduce images from the movie's promotional material (it is copyright protected), you can easily see it for yourself by doing a Web search for images of Ted Neely in *Jesus Christ Superstar*.

von Noorden described an exotropic patient who was a mail carrier. He was able to drive down the street watching the road with one eye while simultaneously being able to read addresses on mailboxes with his other eye [1]. Although he was pleased with his improved cosmesis after strabismus surgery, he missed having this useful panoramic vision.

I don't know why they call my eye lazy. It's traveling all the time.
—8-year-old boy with intermittent exotropia.

**Fig. 6.1** Late thirteenth-century Deesis mosaic of Christ from the Hagia Sophia, Istanbul, Turkey. There is a prominent left exotropia (Robert Preston, Alamy Stock Photo, with permission)


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_6

## Overview

### Basic Information

Exodeviations occur about one-third less frequently than esodeviations in North America and Europe, are more common in females [2], and occur more frequently in regions with higher levels of sunlight [3, 4]. There is a genetic component to XT; however, it is most likely multifactorial [5]. If one sibling from a multiple birth has an XT, there is a 17-fold increase in the chance of another sibling having a similar problem; there is no such increase if the sibling is from a separate birth [6]. Like all forms of strabismus, exotropic deviations can be manifest or latent, and if the former, intermittent or constant. Intermittent exotropia is by far the most common form of manifest XT.

## Intermittent Exotropia

### Clinical Characteristics

#### Basic Information

*Presentation*

Intermittent exotropia X(T) may begin as a latent deviation (exophoria) that over time deteriorates to an intermittently manifest deviation. Although X(T) can appear in the first year of life, the majority of cases present between 2 and 4 years of age [7]. Although amblyopia is uncommon in patients with X(T) unless there is significant anisometropia, there is typically a strong fixation preference. In the vast majority of patients only one eye is spontaneously misaligned intermittently; this should not be taken as evidence of amblyopia in the deviating eye.

Often a child with X(T) will intermittently close the nondominant eye in bright sunlight. This light sensitivity, called "photalgia," was thought to be due to avoidance of diplopia by auto-occlusion when the eye starts to deviate. The reason for eye closure is probably more complex. It has been shown that bright light decreases fusional convergence amplitudes in patients with X(T) [3, 8]. Also, many patients will continue to demonstrate this phenomenon despite successful surgical intervention.

Lack of alternation does not confirm that amblyopia is present.

#### Important Point

Before operating on a patient for X(T) who manifests photalgia, be proactive in telling parents that the monocular eye closure may persist after surgery, even if the surgery is successful. Doing so will help prevent parental disappointment.

#### Pearl

Sometimes parents may not remember or agree upon which eye habitually deviates, and if the child is controlling well during an exam you may not see it spontaneously deviate. You can always determine which eye is nondominant by observing the behavior of the eyes after a cover-uncover test. Consider a hypothetical patient who is left eye dominant and has an intermittent right (R) XT. First cover either eye—let's assume the nondominant right eye. It will become exotropic under the cover. When the cover is removed, it will make a slow vergence movement to refuse. Now cover the dominant left eye, and of course it will deviate under the cover. If you then remove the cover, the left eye will make a saccadic movement to pick up fixation. The nondominant right eye will first make a saccade to the right, putting it in an exotropic position, followed by a slow convergence movement to fuse (Fig. 6.2).

#### Basic Information

*Natural History*

The natural history of X(T) is unclear because it tends to change slowly over time, and there are no long-term longitudinal prospective natural history

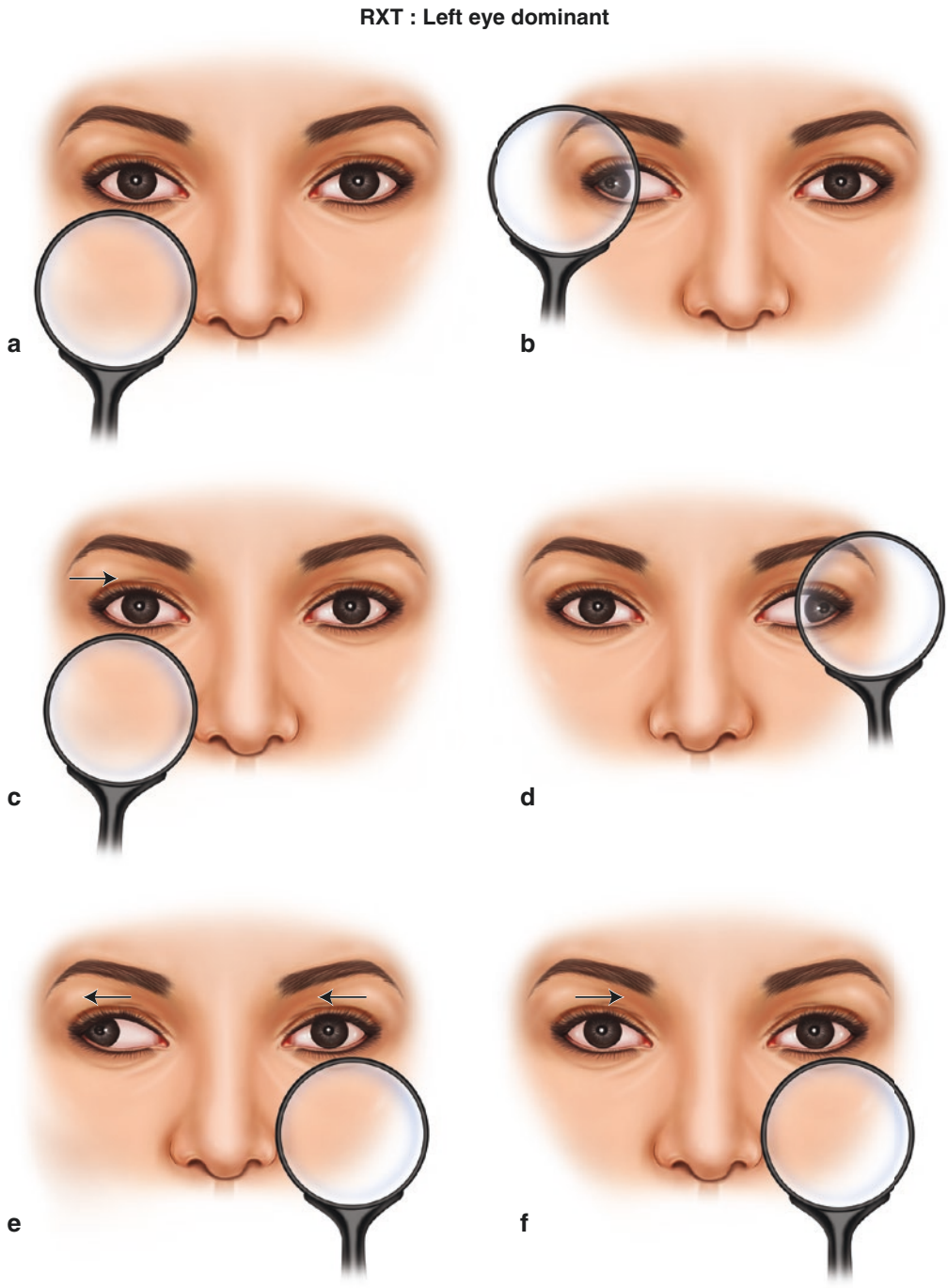



**Fig. 6.2** (**a**) Patient with dominant left eye and well-controlled intermittent right exotropia shown while fusing. (**b**) If the nondominant right eye is occluded, it drifts exo. (**c**) If the cover is removed, the right eye makes a vergence movement to refuse. (**d**) If the dominant left eye is occluded, it will go exotropic. (**e**) If the occluder is removed, both eyes make saccades to the right, allowing the left eye to pick up fixation and the right to become exotropic. (**f**) Finally, the right eye makes a vergence to fuse

studies. Some patients progress from an exophoria to an X(T), which later progresses to a constant XT. This evolution can be slow. There are even some patients who improve over time. von Noorden found that in a series of 51 intermittent exotropes who were treated conservatively over a period of 3.5 years, 16% improved, 9% did not change, and 75% got worse [1].

## Basic Information

*Sensory Adaptation*

When X(T) develops in a young child with an immature visual system, bitemporal suppression develops to avoid diplopia and visual confusion. This suppression is facultative, in that it is present when the XT is manifest, and it is absent when the eyes are aligned. Most patients with X(T) avoid diplopia by suppressing, and hence do not develop anomalous retinal correspondence. Many have normal stereopsis if tested when the eyes are aligned. Although early in the evolution of an X(T) a child may report diplopia, typically this is short-lived, as the suppression develops rapidly.

## Advanced Information

*The Suppression Scotoma Is Not Absolute*

The size of a scotoma from neurologic or retinal damage is inversely related to stimulus used to map it. The brighter the stimulus, the smaller the scotoma. The facultative bitemporal scotoma with X(T) is an adaption to stimulation of noncorresponding retinal points in the two eyes to avoid diplopia, and it behaves in an opposite manner. The brighter the stimulus to the fixing eye, the greater the stimulus to suppress, and hence the scotoma is denser. Conversely, as the intensity of the stimulus to the fixing eye decreases, the less the stimulus to suppress and the less dense the scotoma. This is the reason some patients with X(T) may describe diplopia for night driving, where the stimulus to the fixing eye is less intense.

## Try This Experiment

*Demonstrating the Scotoma Is Not Absolute*

Have a patient with X(T) fixate a light at the end of the exam lane while tropic. Typically, they will not be diplopic. Then introduce a neutral density filter bar in front of the fixing eye and gradually increase the density. You will reach a density at which the patient will report diplopia. If you treat this patient with antisuppression orthoptics as described later in this chapter, the patient will progressively begin to appreciate diplopia with increasingly less dense filters. This experiment is best done with a Bagolini filter bar, which consists of red filters of increasing density (Fig. 6.3); however, a standard neutral density filter bar will suffice.

> God and Satan were walking. God sees a bright shiny object and picks it up. "It's truth," God says. "Give it to me says Satan, and I'll organize it."
>
> —Tale told by Ram Dass.

## Basic Information

*Classification of X(T)*

Over 60 years ago, Burian proposed a classification system, the use of which has become the standard way of understanding X(T) [9–13]. He classified X(T) as being *divergence excess* if the distance deviation exceeded the nearby 10 prism diopters (Δ) or more, *basic* if the distance and near deviations were within 10 Δ of being the same, and *convergence insufficiency* if the near XT exceeded the distance by 10 Δ or more. In addition, he categorized patients as having a *simulated divergence excess* if they presented as having a *divergence excess* pattern, but in fact were merely masking a larger deviation at near which can be brought out by 45–60 min of monocular occlusion. Subsequently Brown observed that measuring through +2.50 or +3.00 diopter (D) lenses at near may also bring out a larger near deviation, and he felt that this test would also identify patients with a *simulated divergence excess XT* [14, 15]. Based on his classification system, Burian made treatment recommendations that evolved from certain

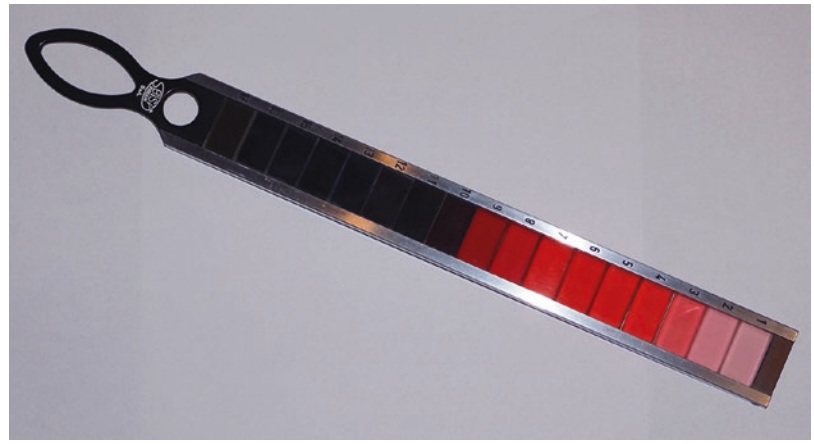
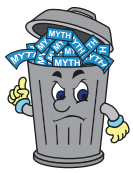


**Fig. 6.3** Bagolini filter bar consists of a column of increasingly dense red lenses

unproven and probably false assumptions. For the *divergence excess* pattern, he recommended lateral rectus muscle (LR) recessions bilaterally on the assumption that it would give more correction at distance than near; for the *basic* pattern he recommended a unilateral recess/resect (R&R) procedure based on the assumption that it would give equal correction at distance and near; and for the *convergence insufficiency* pattern he advocated bilateral medial rectus muscle (MR) resections based on the assumption that it would give more correction at near than at distance. He considered the *simulated divergence excess* patients to really be masking a *basic* pattern and hence he treated them as such—with a unilateral R&R.

There are numerous clinical observations that are inconsistent with Burian's classification system and surgical recommendations.




### Myth

The so-called divergence excess pattern is caused by an excess of divergence.

### Fact

The divergence excess pattern of XT is probably not caused by an excess of divergence. Typically, patients with X(T) will be approximately 30 Δ more exotropic under deep anesthesia than they are in the awake state at distance fixation. I would contend that if the deviation increases when all neuromuscular control to the extraocular muscles is pharmacologically blocked by anesthesia, one cannot attribute the awake distance deviation to an excess of divergence. Moreover, there is no evidence for a divergence center in the brain, so "divergence" is really inhibition or dampening of convergence.

### Basic Information

Also, approximately one-third of patients presenting with a greater distance XT than at near will have the same response at near to both prolonged monocular occlusion and plus lenses [12]. But monocular occlusion disrupts fusional convergence, and plus lenses decrease accommodative convergence, so the tests should not be interchangeable. In addition, bilateral LR recessions typically have a major effect on the distance deviation and a negligible effect on the near deviation of a *divergence excess* intermittent exotrope, but will have an almost equal effect on the distance and near deviation in a *basic* intermittent exotrope [16, 17]. If Burian's underlying premise that bilateral LR recessions affected the distance more than the near deviation was correct, this should not be the case.


### Myth

Most distance/near differences reflect the accommodative convergence/accommodation (AC/A). Jampolsky attributed most dis-

tance/near differences in X(T) to the AC/A ratio [18]. At first glance, the common intermittent exotrope with a large distance deviation and minimal at near could be thought of as having a high AC/A ratio. This is certainly the case if the heterophoria method of calculating the AC/A is employed. However, in such a patient bilateral LR recessions will typically minimize the distance deviation without affecting the near. This would mean that bilateral LR recessions *decreased* the AC/A, which is counterintuitive.

### Fact

Clearly other explanations for explaining distance/near differences in X(T) are called for.

> Gandhi was leading a protest march in India when he changed his mind several days into the march and called it off. His aides objected, saying many people had planned on marching. Gandhi replied, "I only know relative truth, and relative truth keeps changing. I have a commitment to the truth, not consistency."
>
> —Ram Dass.

## New Classification

In the 18th Annual Scobee Memorial Lecture and subsequent publications I outlined a new functional classification of X(T) that attributed most distance/near differences to the presence or absence of a strong fusional mechanism at near that often masked a larger deviation [16, 17, 19].

### Basic Information

In 1952 Scobee described the common scenario in which patients with X(T) may have a large deviation at distance fixation, a much smaller (or no) deviation at one-third meter, yet the near deviation will increase to approximate the distance deviation after a period of prolonged monocular occlusion [20]. Initially, Scobee described using 24 h of monocular occlusion to bring out the near deviation, but later he and others realized that 45–60 min was sufficient [9, 12]. In my Scobee Lecture [19] I described this phenomenon as being due to what I called "tenacious proximal fusion" (TPF), because it appeared as though these patients had a strong fusional mechanism at near that did not break down with a simple cover test. I subsequently realized that the term TPF is not adequately descriptive for the following reason: Consider a patient who manifests a large RXT at distance and is orthophoric at near, but who has a larger near deviation after 1 h of monocular occlusion (has TPF). If you observe this patient while the distance XT is manifest, then occlude the exotropic eye, and have the patient shift fixation to one-third meter while keeping the exotropic eye covered, you will invariably find them to be ortho at near. In other words, they mask the near deviation without having been allowed to fuse. Consequently, I feel it is incorrect to attribute this phenomenon to fusion that is slow to dissipate. Subsequent studies suggest that the masking of the larger near deviation is due to a preponderance of binasal or uncrossed retinal disparity that typically occurs with near fixation [21]. For simplicity, I now prefer to replace the term TPF with "the Scobee phenomenon" (ScPh). The full explanation as to how this complex phenomenon accounts for the near deviation is beyond the scope of this book. I refer interested readers to the source [21].

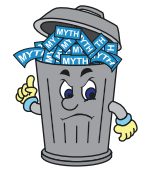
According to this new classification, patients in whom the distance angle exceeds the near angle by 10 Δ or more, and do not increase the near deviation after a patch test, most often have a strong proximal convergence. I suspect that in most of these patients a larger near deviation would be uncovered either with a longer patch test or some form of prism adaptation; however, this is speculation. In addition, there is a quite rare patient in whom this pattern can be seen due to a truly high AC/A ratio, which will be discussed in detail later in this chapter. Patients in whom the distance XT exceeds the near, and in whom the near approximates the distance after a patch test (e.g., exhibits ScPh) do not have a high AC/A, but often have a pseudo-high AC/A (also discussed later in this chapter). Those patients in whom the distance and near XT are initially with 10 Δ of being equal are also called basic exotropes (for lack of a better term) in this new classification. Finally, those patients in whom the initial near XT

exceeds the distance either have a low AC/A ratio, a fusional convergence insufficiency, or a pseudo-convergence insufficiency. Table 6.1 lists the different patterns of XT according to Burian's classification and the new classification.

## Basic Information

*The AC/A and the New Classification of X(T)*

The ScPh—a strong drive for convergence at near that masks a larger deviation—will "contaminate" any near measurement used for the calculation of the AC/A. Consequently, any near measurement for AC/A calculation must be one that is made after the ScPh has been suspended with a prolonged patch test. Thus a gradient AC/A calculation using minus lenses at distance can be done in the usual manner. A gradient AC/A using plus lenses at near must be done after a patch test, and a heterophoria method calculation must use a post-patch test near measurement.




## Myth

A large increase in the XT at near with +3 D lenses means there is a high AC/A.

**Table 6.1** Burian vs. Kushner classification of intermittent exotropia

| Burian's classification | Kushner's classification | Comments | Surgical recommendations according to Kushner's classification |
|---|---|---|---|
| Divergence excess | Proximal convergence or high AC/A | High AC/A is present if AC/A is high with gradient testing at distance, an ESOphoria is present at very near fixation, and near increases with +3 lenses but *not* with patch test | Recess LR OU if proximal convergence. If true hi AC/A, defer surgery with minus lenses and bifocal,[a] or recess LR OU, and posterior fixation MR OU |
| Simulated divergence excess based on patch test | ScPh or pseudo high AC/A | Pseudo high AC/A is present if XT at near increases with +3 lenses but NOT with patch test, distance gradient AC/A is normal, and EXOphoria is present at very near fixation | Recess LR OU[a] or R&R per surgeon's preference |
| Simulated divergence excess based on +3 lenses at near | ScPh with pseudo high AC/A or true high AC/A | True high AC/A is present if patch test does not increase the near XT, distance gradient AC/A is high, and ESOphoria is present at very near fixation | If pseudo high AC/A, recess LR OU[a] or R&R per surgeon's preference. If true high AC/A, defer surgery with minus lenses and bifocal,[a] or recess LR OU and posterior fixation MR OU |
| Basic | Basic | — | R&R or recess LR OU[a] with increased surgical dose |
| Convergence insufficiency | Low AC/A, or fusional convergence insufficiency, or pseudo-convergence insufficiency | Low AC/A shows low response to distance gradient testing and no change with patch test. Fusional convergence insufficiency has normal AC/A on distance gradient testing and no change with patch test. Pseudo-convergence insufficiency has normal AC/A on gradient testing and increases the distance XT with a patch test to approximate the near XT | Recess LR OU[a], or resect MR OU, or R&R targeting distance XT. Consider infraplacing LRs or supraplacing MRs to induce a V-pattern.[a] If pseudo-convergence insufficiency either recess LR OU[a] or R&R |

*AC/A* accommodative convergence/accommodation ratio, *LR OU* lateral rectus muscle, both eyes, *MR OU* medial rectus muscle, both eyes, *ScPH* Scobee phenomenon, *R&R* recess/resect procedure, *XT* exotropia

[a]Author's personal preference when multiple options are acceptable

### Fact

Most such patients have a "pseudo-high AC/A (43% of intermittent exotropes in my experience) [22]. Consider a typical patient who initially measures 30 Δ XT at distance and is ortho at near. With +3 D at near he measures 30 Δ XT. If one does a heterophoria calculation using these numbers, the AC/A will be approximately 15–16/1, and with the gradient method it would be 10/1—high with both methods. But most of these patients are masking a larger near XT because of the ScPh. If a patch test is done, the near measurement is found to be 30 XT'. (The prime sign denotes a near-gaze measurement.) If +3 lenses are then used at near, there is only a modest increase in the XT with a measurement of 38 Δ XT'. Applying the heterophoria method using the post-patch test near measurement of 30, the AC/A is about 5–6 (normal). Using the gradient method at near incorporating the post-patch measurements, the AC/A is about 3 (also no longer high). This hypothetical patient has a pseudo-high AC/A.

### Question

Are there intermittent exotropes that actually do have a high AC/A, and if so, do they need to be managed differently?


### Reply

Yes there are, but they are rather uncommon patients (7% of intermittent exotropes in my experience) [22]. The key finding in this group is that initially the distance XT exceeds the near XT, and the latter does not increase with a patch test. The near XT will increase significantly, however, *after* a patch test with +3 lenses. In addition, a gradient AC/A calculation at distance with minus lenses will show a high AC/A. Finally, if tested at very near range, e.g., 3–4 in., they may show an ESOphoria. However for most intermittent exotropes this is closer than their near point of convergence, and they show an EXOphoria. It is important to identify this relatively uncommon patient, because unlike most exotropes in whom the distance exceeds the near deviation, they will typically keep their high AC/A after LR recessions and show an esotropia (ET) at near postoperatively, requiring a bifocal. In my experience the aforementioned diagnostic criteria had a sensitivity, specificity, and positive and negative predictive values, all of 100% in identifying those intermittent exotropes who would show a postoperative ET if surgically aligned at distance [22]. The good news is that because they have a high AC/A, you get a lot of "bang for your buck" with overminus spectacles. One or two D of minus will control a large deviation because of the high AC/A. They uniformly will require a bifocal for control of a near. However, in my experience 100% of these patients normalized their AC/A by 18 years of age and could be operated upon in the usual manner without developing a near-gaze ET afterwards.

### Question

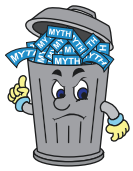
I have heard that both +3 lenses at near and a patch test are equally satisfactory ways of identifying simulated divergence excess XT. Is one test preferable?


### Reply

It is important to recognize that these two tests work on different mechanisms that affect near gaze. A patch test suspends fusional convergence, and +3 lenses decrease accommodative convergence. So the patch test is more appropriate. The misconception that they are interchangeable stems from the observation that approximately one-third of patients will have the same results with either test [12, 14]. How then is it possible that the two tests give the same results if they work on different mechanisms? Consider the hypothetical patient with 30 Δ XT at distance and orthophoria at near, but a masked 30 Δ near XT' via the ScPh. Even with a normal AC/A, one would expect the total near XT to

increase some when tested through +3 lenses—perhaps an added 8–9 Δ. The ScPh had been accustomed to masking 30 Δ of XT, but not 38–39 Δ. The system's convergence drive is exceeded and breaks down, and the entire near deviation (that which was masked plus that added by the +3 lenses) becomes manifest. It is as though the extra 8–9 Δ of XT brought out by the +3 lenses is the straw that broke the camel's back—in this case the camel being the near fusional control mechanism.

With his classic classification, Burian made surgical recommendations based on the mistaken assumptions about the effect of different surgical procedures on the distance/near relationship.




### Myth

Symmetric LR recessions will give more correction at distance than near, recess/resect procedures will have equal effect at distance and near, and MR resections will give more affect and near than distance.

### Fact

Unless the size of LR recessions is large enough to cause an underaction, symmetric LR recessions may affect distance and near equally. For most patients in whom the distance exceeds the near, LR recessions will affect distance more than near and you "get what you need" at both distances. This is understandable if you think of these patients as masking a larger near deviation. Recess/resect procedures will give equal correction at distance and near, but notably will give more correction in the field of the recessed LR, and less in the field of the resected MR. This is because resections do not "strengthen" a muscle but do "tighten" it, causing a relative restriction in the opposite gaze field. A small resection may have some strengthening affect, but typically resections cause a tightening or restriction (see Chap. 10, and Figs. 10.1 and 10.2 for further explanation). Hence, symmetric MR resections may cause most correction in side gazes, and more at distance than at near due to their restricting effect.

### Basic Information

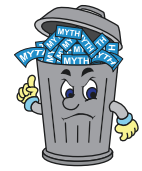
*Distance/Near Differences According to the New Classification with Surgical Recommendations*

1. **Distance XT exceeds the near XT' initially, after a patch test, and with +3 lenses at near**
   First and foremost, these patients do not have an "excess of divergence" for the reasons outlined above. The majority really have a larger deviation that is masked by a very strong convergence tone at near, which even a patch test does not bring out. I suspect that many of these will manifest a larger near deviation either with a patch test that is longer than an hour, or with prism adaptation. This, however, is speculation on my part. These patients can be treated with symmetric LR recessions according to both the new classification and Burian's [16].
2. **Distance XT exceeds the near XT' initially, but the near XT increases after a patch test. The near XT may or may not increase with +3 lenses.**
   This patient exhibits the ScPh and is what Burian called a simulated divergence excess. Whereas Burian felt that they should be considered *basic* with respect to surgical management and receive a recess/resect procedure, the new classification suggests that they can be treated equally well with symmetric LR recessions, or a recess/resect procedure, according to the surgeon's preference [16]. If the XT does increase at near with +3 lenses, the patient has, by definition, a pseudo-high AC/A.
3. **Distance XT exceeds the near XT' initially, and the near XT does not increase after a patch test but does with +3 lenses prior to the patch test.**
   This patient may have a true high AC/A. Further testing should include a distance gradient AC/A calculation with minus lenses, and testing at about 3–4 in. to look for

an esophoria to confirm the diagnosis. If the patient is confirmed to have a true high AC/A, standard surgery will result in an ET at near if the distance XT is corrected surgically. Treatment can be with over minus spectacles, which will need to include a bifocal add, until the child outgrows the high AC/A. Good results have been reported in this type of patient with LR recessions and simultaneous MR posterior fixation [23]; however, I have little personal experience with that approach.

4. **Distance XT equals near XT.**
This is Burian's *basic* XT, which, for lack of a better term, is also called *basic* in the new classification system. It is an XT that does not manifest the ScPh. Think of the ScPh as a helpful finding indicating a stronger fusional mechanism, which improves the prognosis. Conversely, patients lacking this strong fusional mechanism have a poorer prognosis and may need a more "stabilizing" surgical procedure. In fact, studies have shown that patients who do not manifest the ScPh have a poorer surgical prognosis [16, 19]. Although my personal bias is for symmetric surgery when possible, in a prospective and randomized clinical trial I found that patients with a *basic* XT pattern did do better with a recess/resect procedure than with symmetric LR recessions [16]. This was contrary to what I had hoped to find; thus I did in fact change my practice pattern. Whereas Burian felt that a recess/resect procedure was indicated in these patients based on the mistaken assumption that it would be more likely to produce equal effects at distance at near, I believe that it is the stabilizing effect of a recess/resect procedure that produced the better results. A recess/resect procedure produces incomitance and more of an ET in the field of the recessed LR, which limits the eye from drifting outward. Whereas with LR recessions, if there is a drift toward undercorrection in primary gaze, the patient will be undercorrected in all gazes. The image in the deviating eye will be on temporal retina that is suppressed. Subsequent to my randomized clinical trial, it has been suggested that by augmenting the amount of LR recession by 1.5–2.5 mm per muscle, results can be as good as with recess/resect surgery [24]. To my mind this approach shows promise.

The absence of the ScPh indicates a weaker fusional mechanism.




**Myth**
LR recession affects the distance deviation more than the near in contradistinction to a R&R, which affects distance and near equally.

**Fact**
LR recessions will affect the distance and near angle equally in a basic pattern intermittent exotrope.

## Convergence Insufficiency

I think it is somewhat unfortunate that the term *convergence insufficiency* is used to describe two different clinical conditions—fusional convergence insufficiency and accommodative convergence insufficiency. They actually have different clinical findings, and require different treatment.

Fusional convergence insufficiency is characterized by a minimal (or no) exophoria at distance, and a somewhat larger exophoria at near, which in some patients may be an X(T). It is rare for the distance deviation to be an XT, either intermittent or constant. The size of the near deviation is typically not more than 10–15 Δ greater than the distance deviation, although there may be exceptions. These patients have decreased fusional convergence amplitudes, and symptoms of headache, asthenopia, and/or diplopia with near work. Fusional convergence insufficiency responds well to fusional convergence orthoptic exercises.

Fusional vs. Accommodative Convergence Insufficiency are physiologically different.

Accommodative convergence insufficiency is characterized by a manifest XT at distance, either an intermittent or, less frequently, constant, and a larger exo deviation at near. The near deviation is fre-

quently 15–18 Δ greater than the distance exo, implying that there is essentially no convergence occurring with a shift from distance to near gaze. Recall that the amount of convergence needed at one-third meter to maintain the same alignment as at distance fixation is equal to the interpupillary distance in millimeters × 3 (about 15–21 Δ). If you test the AC/A in these patients with either a gradient test at distance or near, or a heterophoria method after a patch test, you will find that they invariably have essentially no accommodative convergence. The AC/A typically ranges from 0 to 1 [17, 19]. These patients do notoriously poorly with surgery. In most cases if the near XT is surgically corrected, they end up overcorrected at distance. This is not surprising if one realizes that they have little or no accommodative convergence. Various proposed surgical formulae involve doing a recess/resect procedure with more of a MR resection and less LR recession, on the assumption that the medial resection has more effect on the near deviation [25], or slanted recessions of the LRs or biased resection of the MR muscles (MRs) [26]. I am unconvinced by these published reports and doubt biasing a R&R toward a greater resection, or slanting procedures, which I feel are based on unsound theoretical constructs [27], are any different than standard recessions or resections. Similarly, I have been unimpressed that symmetric MR resections selectively give more correction at near as Burian proposed. For many years I have tried to manage these patients with prisms if I could, because so many do not do well with surgery. If I did perform surgery, I would aim to correct the distance deviation, which would leave the near improved, but undercorrected. I managed the improved exo at near with prisms if it was symptomatic. In 2004, Buckley described an interesting surgical approach to these patients [28]. He recommended intentionally transposing the rectus muscles to create a V-pattern, because most near work (reading) involves downgaze. I have used this approach in about a dozen patients. I found that by transposing the LRs downward or the MRs upward one tendon each, I predictably created 7–8 Δ of V-pattern shift. This was helpful with respect to symptoms in the reading position.

## Pseudo-Convergence Insufficiency

Relatively infrequently, a patient presents with what looks like a convergence insufficiency pattern, but for some unknown reason, she is masking a larger deviation at distance. A 45–60 min patch test will unmask this. It is worth doing a patch test on suspected convergence insufficiency exotropes, because their prognosis is better if they really have a pseudo-convergence insufficiency. They can be treated like a basic exotrope. These are really uncommon patients.

### Basic Information

Table 6.1 gives the recommended surgical procedures for the different types of X(T) according to the new classification.

## Examination

### Basic Information

*Refractive Correction*

Visual blur, particularly if it is more in one eye than in the other, is extremely destabilizing for patients with X(T) [29, 30]. Consequently, it is crucial to correct any significant refractive error, but equally important to cut plus or over-minus symmetrically. That will keep the image in both eyes equally in focus. However, because accommodation can help control an XT, you should not push plus the way you should with ET. In a hyperopic child, I will typically cut the plus by about 1–1.5 D, provided that I am not intentionally planning to use over-minus therapy. My rationale for these numbers is that a normal non-strabismic child is typically 1–1.5 D hyperopic, and I want the exotropic child to accommodate this approximately normal amount. You should be cautious about cutting plus (or over-minusing) more than 3 D, as that will introduce instability in child's alignment as he or she varies from accommodating 3 D or more, or relaxing his or her accommodation. A variation in

accommodation of 3 D will cause about 15–18 Δ of variability in horizontal alignment.

### Advanced Information

There are some intermittent exotropes with high hyperopia (greater than 3 D) who may improve their alignment by wearing all or most of their hyperopic correction [31]. Obviously, this improvement has nothing to do with manipulating accommodation or accommodative convergence. Instead, it is testimonial to the fact that clearing up the image quality may improve control. These children are simply not accommodating the 3–8 D (or so) prior to glasses, due to the excessive effort needed to overcome this much hyperopia, and they are blurred. I prescribe the cycloplegic refraction, cutting the plus about 1 D. Again, the rationale is to have them accommodating an amount that would be normal for their age. Others prefer to give the full plus.

### Important Point

For intermittent exotropes with high hyperopia, you should prescribe most or all of the plus combined with all of the cylinder.

### Question

I care for a presbyopic patient with X(T), who is interested in monovision. Is this OK?


### Reply

No. By definition, one eye or the other is always fogged with monovision. Because of the deleterious effect of unequal vision inputs, monovision may cause a worsening of control and be an obstacle to fusion [32]. I saw a 45-year-old woman for a second opinion. She had a long-standing well-controlled X(T). When she became presbyopic, she opted for managing her −3 D refractive error with monovision, wearing plano over one eye and −3 D over the other. After 1 year of monovision she noted frequent diplopia and a worsening of the control and magnitude of her XT. Her ophthalmologist performed bilateral LR recessions, and she had the initial desired small overcorrection, but within a short time the XT returned. Her doctor then performed bilateral MR resections, again having an initial small-angle ET immediately after surgery. However, within several months her deviation was again bothersome, and a third operation was recommended. She then saw me in consultation. I recommended she discontinue monovision and wear her proper myopic correction with a bifocal. Within 1 year she was no longer manifesting a tropia and only had an asymptomatic 10 Δ exophoria. She remained symptom free without further surgery over the subsequent 11 years. I have seen many similar patients.

### Important Point

It may take several months for a deviation to be controlled after discontinuing of monovision.

### Basic Information

*Motility Information Specifically Unique to XT*

In addition to the usual measurements you want to obtain on strabismus patients, intermittent exotropes call for some additional tests. Put in reductionist terms, the frequency of the deviation dictates *if* treatment is indicated, and the size of the deviation determines *what* the treatment should be, if treatment is necessary. So you need to develop some system for assessing control. It is not so important what that system is, as long as it is something you can use consistently from one exam to the next. Nevertheless, there have been attempts to develop a classification system for assessing control in intermittent exotropes [33]. I feel that they fall short because they conflate assessing how easily a patient drifts exotropic, with their ability to regain control once dissociated. There are some patients who rarely drift exotropic, but may have difficulty recovering once dissociated. Others may frequently become tropic spontaneously, but have a great ability to recover. My own system

**Table 6.2** Classification of control of intermittent exotropia

| Grade | Control | Recovery |
|---|---|---|
| Excellent | Is not tropic | Within seconds after cover and prior to a blink |
| Good | Briefly tropic only after cover and not spontaneously | Quick recovery in less than 5 s after cover or spontaneous exo |
| Fair | Occasional spontaneous tropia | With a blink or change in fixation distance |
| Poor | Tropic frequently or all of the time spontaneously | May stay exo through a blink and/or with change in fixation distance |

separately categorizes the ease of drifting exotropic with the ability to recover (Table 6.2). These two separate categories should be noted at distance and near fixation.

Many patients with X(T) will manifest a larger deviation if tested while looking out a window at a far distant target [12, 13]. I have found this to occur in 17% of intermittent exotropes, and the phenomenon can occur in all of the differing distance/near patterns of X(T) [19]. Studies have shown that it is a combination of the increased light outdoors and the increased distance that is responsible for the larger deviation [8, 17]. Thus a measurement in a very long indoor hallway, or in a standard exam lane under floodlights that simulate outdoor illumination, will not suffice. I reported that patients who manifest increased deviation when looking out a window, and underwent surgery for their initial measurement at 6 m, had an undercorrection rate of 40% [8, 19]. Whereas those who underwent surgery targeting the larger out-the-window measurement had an undercorrection rate of only 18% [8].

### Important Point

All patients with X(T) who are undergoing surgery should have a measurement made while they fixate a far distant outdoor target, regardless of their distance/near classification. This can be obtained with them standing close to and looking out a window.

### Basic Information

In a series of 118 patients with X(T), 19% showed an increase in their deviation measured at 6 m after a 45–60-min patch test (mean increase 10.3 Δ ± 5, range 5–30 Δ) [8]. This phenomenon occurred in all classes of distance/near differences and was not necessarily present in the same patients who showed a deviation increase while looking at an outdoor target.

### Important Point

All patients with X(T) who are undergoing surgery should have a measurement made at 6 m after a 45–60-min patch test.

### Basic Information

In a prospective randomized clinical trial [8], 40 intermittent exotropes who showed an increase in their deviation at 6 m either after a 1-h patch test or while looking out a window had surgery for their initial measurement at 6 m and had a satisfactory outcome rate of 62%. This contrasted with a significantly higher satisfactory outcome rate of 86% in 43 patients who underwent surgery for the largest deviation found (e.g., outdoor target or after 1-h patch test), $P < 0.001$.

A patch test and outdoor measurement are not interchangeable tests. Both need to be done in all pre-surgical intermittent exotropes.

### Important Point

For all patients with X(T) who are undergoing surgery, the surgery should target the largest deviation obtained.

### Question

My patient has no more than 5 Δ at 6 m, but measures 35 Δ XT with poor control while looking out a window. The parents see the deviation very

frequently outdoors, and say that it is getting worse. If I operate for 35 Δ, how likely is it that he will be overcorrected afterwards?


### Reply

Patients like yours are relatively uncommon, but certainly are out there. I have operated on approximately a dozen similar patients, and none was overcorrected despite my targeting the largest deviation. I suspect that most such patients are just masking a bigger deviation at 6 m, which might come out with a prolonged patch test or prism adaptation.

### Advanced Information

*Lateral Incomitance*

In 1969, Moore reported that intermittent exotropes who had lateral incomitance in the form of a decrease in their deviation of 5 Δ or more in both right and left gaze were very prone to surgical overcorrection [34]. Others felt that the decrease had to be greater than 10 Δ to be considered diagnostic of lateral incomitance [1, 35]. I found this type of lateral incomitance to be uncommon, occurring in about 5% of intermittent exotropes. Although others reported a much higher incidence [34, 36], some of this higher incidence may be due to artifacts of measuring [37]. The accepted explanation for lateral incomitance is that the MRs are tight, thus decreasing the deviation in side gazes. However, I have not felt that intraoperative forced ductions confirmed excessive tightness to the MRs in these patients. Another explanation for this phenomenon is that the MRs are "overly strong" (e.g., generate a greater than normal contractile force when stimulated; this mechanism might explain why these patients are prone to overcorrection). Although we usually think in terms of extraocular muscles being weak or tight, there are theoretical constructs that explain how they can become "overacting," e.g., overly strong [38]. Of course, weak or underacting LRs may result in a decrease in the deviation in right and left gazes. However, it would be counterintuitive to assume that in the absence of prior surgery, an exotrope would have underacting LRs. Underacting LRs can, however, result in lateral incomitance after prior LR recessions.

### Question

My patient had previously undergone bilateral LR recessions of 7 mm for an X(T), and the deviation has returned. He now measures 30 Δ XT in primary, and the deviation drops to 18 Δ in both right and left gazes. Do I need to be as worried about an overcorrection as I would with an X(T) with de novo lateral incomitance?


### Reply

I think not. To my knowledge, none of the prior reports on lateral incomitance specifically looked at the overcorrection rate in patients in whom the incomitance resulted from prior surgery. In the absence of firm data, I will argue for what makes sense. De novo lateral incomitance is presumably due to tight or "overacting" MRs, which presumably overpower surgically weakened LRs, resulting in an overcorrection. There is no reason to suspect that this will occur if the lateral incomitance is caused by recessed LRs. In my own experience, patients with secondary lateral incomitance did not experience higher numbers of overcorrections in primary gaze than were seen in patients without lateral incomitance. There is another concern, however. If the new deviation is fully corrected with secondary surgery, there is likely to be an overcorrection in right and left gazes, especially if the LRs are recessed further. Small medial resections may avoid side-gaze overcorrections. However, keep in mind that moderate-to-large resections induce restrictions, and in this setting will also cause side-gaze overcorrections. Depending on the size of the deviation in primary, and the amount of lateral incomitance, you may not be able to fully correct the primary position deviation without having some degree of ET on right and left gazes. This may be an acceptable result, depending on how far into side gaze the diplopia occurs.

### Pearl

Patients who develop lateral incomitance as a result of prior LR recessions are not particularly prone to surgical overcorrection in the primary position. They may, however, be more prone to overcorrection in right and left gazes.

### Question

If a patient had large bilateral LR recessions for X(T), and years later develops a recurrence, what factors influence your decision about resecting the MRs, or re-recessing the LRs?


### Reply

I am influenced by the size of the initial LR recessions and the amount of lateral incomitance. My guidelines include the following:

1. If the prior LR recessions were 8 mm or more, I usually resect the MRs.
2. If the XT in side gaze is 10 Δ or less, I usually resect the MRs, regardless of the size of the prior LR recessions.
3. I consider the presence of a convergence insufficiency pattern to represent some degree of LR weakness and will not recess the LRs further.
4. If the prior LR recessions were less than 8 mm, the XT in side gaze is greater than 10 Δ, and there is not a convergence insufficiency, I re-recess the LRs.

### Advanced Information

*Prism Adaptation*

The role of prism adaptation has not been adequately studied. It seems plausible that prism adaptation might bring the larger latent deviation that otherwise is seen with a patch test or by fixation on a far outdoor target. I say this, because in my own investigations I found that a very rapid form of prism adaptation would often uncover the larger near deviation that was masked in patients with the ScPh [17]. This is a fertile area for investigation.

### Basic Information

*Sensory Tests*

In most patients with X(T), the results of sensory tests depend on the alignment and control at the moment the test is performed. The majority are sensorially normal when aligned, with normal retinal correspondence and normal stereopsis. But when they deviate, they suppress the deviating eye and lose stereopsis. Consequently, although a progressive trend toward deteriorating binocularity may be important in making a treatment decision, you should always keep in mind that sensory testing is a function of alignment at the moment the test is performed, and may be a moving target. That said, there are data to suggest that deterioration in distance stereopsis may closely parallel a deterioration in control of the deviation [39]. I have found this to be of limited practical utility because clinically available tests of distance stereopsis are cumbersome and awkward to use. Another study has suggested that a decrease in distance binocular acuity may reflect over-accommodation to control the XT, and parallels deteriorating motor control [40]. Although this makes sense, I usually rely on my clinical assessment of motor control to influence my treatment decisions.

## Treatment

### Basic Information

*Timing of Intervention*

The decision to begin treatment of X(T) is based primarily on the frequency of the deviation. A widely used convention is that if the deviation is manifest 50% of the time, treatment is indicated. Realistically speaking, this can be very difficult for parents to assess. If the eye deviates many times during the day, but recovers readily, the parents may perceive it as being worse than if the

eye drifts a small amount exotropic and stays in the deviated position. So you need to pay attention to trends. Is the frequency getting worse? Is the control based on your office assessment deteriorating? Try and assess if the eye is deviated a substantial part of the day.

### Important Point

The decision as to *whether* to begin or change treatment is determined by the *frequency* of the deviation. The *magnitude* of the deviation influences only what the treatment should be; if it is determined that treatment is indicated. I have seen some patients who maintained excellent control over deviations that were as large as 60 Δ and remained symptom free, for as long as 30 plus years; they never needed treatment. Conversely, there are some patients who are symptomatic from poorly controlled 12–15 Δ deviations; they require intervention.

### Basic Information

*Nonsurgical Treatment—Occlusion*

When I see a child for the first time with a newly diagnosed X(T), I may just observe them if control is good. I tell parents that sometimes the problem remains infrequent and asymptomatic for many years. Once I make the decision to intervene, I will usually begin with nonsurgical measures.

I have found that some form of alternate occlusion for anti-suppression almost always results in initial improvement. In the relatively rare patient who is a free alternator, I will patch alternate eyes on alternate days for about 3 h per day. More commonly, there is a strong fixation preference (but no amblyopia). In this situation I will patch the dominant eye for 3 h 2 or 3 days in a row, followed by 1 day over the nondominant eye, and keep repeating. If they are amblyopic, which is uncommon in the absence of anisometropia in intermittent exotropes, I just patch the dominant eye. I will see the child in about a month. Almost invariably his control will be improved and, interestingly, the angle smaller. Depending on the response, I will continue patching but begin tapering the hours, ultimately discontinuing the patching. If the deviation subsequently deteriorates, I will reinstitute patching and consider adding overminus lens therapy. In some patients, admittedly infrequently, patching will be curative. I have some patients whom I treated this way who never needed further intervention and remained well controlled for many years.

### Question

What is the rationale behind patching a nonamblyopic intermittent exotrope?


### Reply

Suppression in X(T) is an "active phenomenon" and its depth is inversely related to the stimulus to the fixing eye. Eliminating the stimulus to the fixing eye removes the stimulus to suppress, and over time the depth of the suppression lessens. In theory, eliminating suppression increases diplopia awareness and hence allows for better control of the deviation. This makes sense. However, I typically also see a reduction in the angle of deviation after patching, and this cannot be explained by the aforementioned mechanism. Perhaps there is a vergence adaptation that occurs as a result of the deviation occurring less often, but this is speculation. In many cases occlusion therapy is just a temporizing measure, but this is not necessarily bad. It can allow for repeated and more accurate measurements, and possible defer surgery until a child is older. This may be important in young children who are more prone to losing bifoveal fusion if temporarily overcorrected after surgery for X(T).

### Question

What is the role for using patching prophylactically to prevent deterioration in a well-controlled intermittent exotrope?


### Reply

I personally do not use patching in this manner. Many well-controlled intermittent exotropes never deteriorate, or if they do it happens slowly. I prefer to avoid intervention in those patients that are doing well. A Pediatric Eye Disease Investigator Group (PEDIG) study assessing the role of patching in this manner found that the deterioration rate was low in both the treated and untreated patients [41].

### Important Point

Although pharmacologic penalization has been shown to be an effective choice for treating amblyopia, I am unaware of any studies suggesting that it is an alternative to occlusion for anti-suppression in X(T). Intuitively, it would not appear to be useful. Unequal vision input to the two eyes (unilateral blur) is extremely destabilizing, and, unlike occlusion, it increases suppression [30]. With pharmacologic penalization one eye is always fogged at near range, and depending on the refractive error may also be fogged at distance.

### Basic Information

*Nonsurgical Treatment—Minus Lenses*

Over-minusing a child will cause them to accommodate for distance viewing, and accommodate more than the normal amount for near viewing. This in turn will trigger accommodative convergence and decrease an XT. Jampolsky advocated 3 D of overcorrecting minus lens therapy [5, 42], but I have tended to use somewhat less. To minimize asthenopia and increase acceptance of the over-minus therapy I typically prescribe 2 D of over-minus correction, combined with 2½ or 3 Δ of base in prism in each lens. This amount of prism is approximately equivalent to the amount of correction that one added diopter of over-minus correction would provide. It is important, of course, to over-minus symmetrically in both eyes; add an equal amount of minus lens power to the cycloplegic refraction.

I have found this treatment most useful in children under 7–8 years of age. Older patients may be bothered by the increased accommodative demand it requires, and it is rare that I am utilizing it in older patients. Often patients who have good control with about 2 D of over-minus correction and prism return at a later date still controlling well despite having become more myopic. If so, easing them toward correction with their actual refractive error is the way by which I wean them out of over-minus therapy. Although some colleagues have expressed concern that over-minus therapy may cause myopia to progress, I have found this to not be the case [43]. Myopia and myopic progression are common in children with XT, and that probably is the reason for the perception that their treatment causes the myopia.

Like occlusion therapy, overcorrecting minus lens therapy may be temporizing. But I have a small number of patients in whom I utilized this treatment modality many years ago, and never needed further intervention.

### Basic Information

*Nonsurgical Treatment—Exercises*

Convergence exercises, either in the form of orthoptics or computer-based convergence training, is effective in treating a fusional convergence insufficiency. However, most patients with X(T) do not have diplopia awareness when they are tropic; they suppress. For this reason, most will not benefit from exercises, which require diplopia awareness to be effective.

### Basic Information

*Surgical Treatment—Timing*

The indication for surgery is poor or deteriorating control, and/or the deviation being manifest a substantial part of the day—50% of waking hours is the most commonly suggested threshold, and nonsurgical means have been unsuccessful. Nonetheless, there are compelling reasons to

defer surgery in children under 2½–3 years of age, provided that acceptable control can be maintained nonsurgically. Studies have shown that surgery in very young intermittent exotropes is more likely to result in subnormal fusion than when surgery is done at an older age. The common explanation for this is that, although an initial overcorrection is desired after surgery, very young children may rapidly lose the ability for bifoveal fusion if left esotropic for even a short time. This argument is one of the main ones for temporizing in very young patients. But another explanation is possible. Perhaps the children who have subnormal fusion to start with (e.g., monofixation XT) are more likely to present with poor control at a younger age. The poorer sensory outcome is not a result of the early surgery but rather selection bias.

### Question

I care for a girl who just turned 3 and has an X(T). She has "moderate" control at distance over a 35 XT. At near she has excellent control over a small deviation, which builds to 20 Δ after a 20-min patch test. Cycloplegic refraction is +1.00 sph OU. Her mom is very interested in surgery, but my obvious fear is that she will be overcorrected at near, requiring a bifocal, or that she will become amblyopic. When should I operate on her and what are the concerns?


### Reply

You raise two separate issues. One relates to the timing of surgery and the other the issue of developing an ET at near due to the distance/near difference. With respect to the first issue, if control is "moderate" at distance and "good" at near, there is no rush to intervene. Things deteriorate slowly with X(T). There is the aforementioned concern that very early surgery for X(T) can result in more patients ending up monofixators, but probably by age 3 that is less a concern than in a younger child. If you prefer to wait, you might explain to mom that even in 6 months or a year the risk of losing bifoveal fusion is less. You might try anti-suppression patching, which almost always improves control, at least temporarily, making an additional 6–12 months' wait easier. You certainly could consider surgery now, and if your only hesitation is about her age, I would go ahead. With respect to the second issue, high AC/A ET after XT surgery is uncommon but can be predicted with appropriate AC/A testing pre-op. The fact that she increased her XT at near after a 20-min patch tests suggests that she manifests ScPh, in which case she will not develop a high AC/A ET after surgery. But 20 min is not really adequate to fully uncover the larger latent deviation; I suspect that the XT at near would be greater with a longer patch test. A near measurement *after* the patch test with +3 D lenses would be useful in determining if she has a high AC/A. But, regardless, the likelihood of developing a high AC/A after surgery is not increased due to her relatively young age.

### Basic Information

*Surgical Treatment—Targeted Angle*

As stated earlier, all patients should have a 45–60-min patch test and a measurement made while they fixate a far distant outdoor target. Surgery should be done for the largest deviation found.

### Basic Information

*Surgical Treatment—Choice of Procedure*

This has been discussed in detail earlier and in Table 6.1.

### Basic Information

*Surgical Treatment—Post-op*

In my experience the ideal alignment for children on the day after surgery is between 5 and 15 Δ of ET, because there is an exo drift in the weeks after surgery. If an initial overcorrection is achieved, most patients will be diplopic. It is good to warn patients of this eventuality in advance.

In my experience, adults behave differently and do not have as marked an exo drift; if they are at all overcorrected, they often stay that way—and are quite unhappy. In adults I aim to have them perfectly orthophoric in primary, which usually is accompanied by a slight ET on right and left gazes (if LR recessions were done), with diplopia in side gazes.

**Advanced Information**

*Surgical Treatment—Post-op Persistent Overcorrection*

If the overcorrection is initially in the desired range, but lasts more than several weeks, and particularly if symptoms of diplopia are bothersome, I will intervene in one of several ways. Part-time alternate occlusion will eliminate diplopic symptoms. In young children, particularly if they have uncorrected hyperopia, the daily use of ecothiopate iodide 0.06 or 0.125% can be curative. Finally, Fresnel prisms to offset the deviation can be useful. If I have operated on a very young patient (under 2 years of age), I will start treating an overcorrection at 1 week after surgery because they can rapidly lose bifixation ability if left esotropic even for a modest amount of time. If I had performed a R&R, and the overcorrection is because of limited abduction, I will start passive rotation exercises if the patient is old enough. This simply involves having him or her follow a finger or target into abduction with the operated eye (the fellow eye is covered) 5–10 times several times a day to stretch out the resected muscle. If this is unsuccessful, Botox injection of the resected muscle is highly effective.

The very best long-term results after surgery for X(T) are in patients who had a persistent small-angle overcorrection for a long time after surgery—even a year or so. Consequently, if the patient can be kept comfortable, it is best to defer reoperating as long as practical. This is obviously easier to do in a patient who wears glasses and may be more accepting of long-term prisms. An exception would be if you suspect a slipped muscle, in which case drift to orthophoria would be unlikely.

**Advanced Information**

*Surgical Treatment—Post-op Overcorrection and the Blind Spot Syndrome*

There are some intermittent exotropes who respond to the initial desired 5–15 Δ overcorrection with diplopia, by over-converging to put the second image on the blind spot and hence get rid of the diplopia. You should think of this as a possibility whenever there is a postoperative alignment of about 25–35 Δ ET, but be particularly suspicious if the initial postoperative overcorrection was smaller and then built to about 25–35 Δ. It is important to recognize this uncommon but real syndrome, because it requires special treatment. This consists of full-time alternate occlusion to break up the drive to overconverge. Then you need to measure the patient after about 1 week, without letting them be binocular prior to the measurement. If this reveals a substantially smaller ET, that will represent the patient's underlying deviation. You should then either offset that deviation with Fresnel prisms to eliminate diplopia or, alternatively, initiate a trial with peripheral iridotomy, which may be curative.

If a post-op XT is manifesting about 25–35 Δ of ET you should consider the blindspot syndrome.

**Question**

I operated on a 5-year-old large-angle intermittent exotrope, having done bilateral LR recessions of 8.5 mm. It is now 1 year after surgery, and there is still a 10 Δ ET in primary, which increases to 15 Δ in right and left gazes. Both LRs are −1 underacting. I wonder if there is a slipped muscle. I will explore both, and if either or both slipped, advance it/them. But if not slipped, should I just advance one LR? The primary position deviation is small.


**Reply**

I suspect this is not a case of slipped muscle(s), as it would be unlikely for both

to slip the same amount after the same operation. The overcorrection probably reflects the size of the recessions. Although advancing one LR (if not slipped) would correct the 10 Δ ET in primary and the 15 Δ in the field of the advanced LR, I am concerned that it will not address the ET and limited abduction in the field of the other LR. I would prefer to advance both LRs by smaller amounts.

### Question

I did a left R&R on a 6-year-old girl with a basic pattern XT. I ended up with a persistent overcorrection with slightly limited abduction OS (left eye), so I recessed the left (L) MR 5 mm. Initially the patient looked good, but she rapidly developed an incomitant XT measuring.

25Δ LXT←18Δ LXT→Ortho

She is −2 to adduction OS and I suspect a slipped LMR. She has a big face turn to the right. How should I approach her, given the fact she is orthophoric in left gaze?


### Reply

Obviously, you need to explore the LMR to determine if it is slipped or has an elongated scar. Depending on findings, the cause of the limited adduction must be fixed by advancing the LMR and eliminating any empty capsule or elongated scar. However, the fact that she is orthophoric in left gaze suggests that just operating to correct primary and right gaze will result in an ET in left gaze postoperatively. This is an example of how you should plan surgery to correct primary gaze, and anticipate what this will do to eccentric gaze alignment, and proactively address that. In this situation you could either advance the recessed LLR, recess the RMR, or apply posterior fixation to the left MR, depending on findings.

### Advanced Information

*Surgical Treatment—Post-op Undercorrection*

Although surgically undercorrected intermittent exotropes are more likely to need further surgery than those that are initially overcorrected, not all have poor outcomes. There are fewer interventions in the immediate postoperative period that are helpful, but some can be tried. Alternate occlusion as described above is useful in some patients, as is the use of Fresnel prisms.

### Basic Information

*Surgical Treatment—Reoperation for Undercorrection or Recurrence*

If the initial operation consisted of a R&R, usually the best choice for an "enhancement" is to perform a R&R on the other eye. If the initial operation consisted of bilateral LR recessions, the decision whether to re-recess the LRs vs. resecting the MRs is a function of how large the initial recessions were, and the side-gaze measurements. If there is lateral incomitance and the deviation decreases by 10Δ or more in both right and left gazes, further LR recession will increase this phenomenon and probably result in ET with diplopia in right and left gazes. In this case MR resections may be preferable. If lateral incomitance is not present, and if the initial recessions were 6 mm or less, I will usually re-recess the LRs, because I think recessions are in general preferable to resections—even if the recessions involve operating on previously operated muscles.

## Other Types of Exotropia

### Basic Information

*Primary Infantile XT*

A constant XT that is present by 6 months of age that is not associated with orbital anomalies or vision deficit is called *primary infantile XT*. Like infantile ET it usually requires surgery to correct and rarely develops better than subnormal fusion. Unlike infantile ET, infantile XT is a soft sign for developmental delay or other neurologic problems. I do not routinely institute a neurologic investigation if I diagnose infantile XT. I do, however, alert the pediatrician to monitor carefully for signs of developmental delay or neurologic problems, and

to investigate accordingly. In most cases the developmental or neurologic problems are obvious.

### Advanced Information

*Monofixational XT*

The idiopathic *monofixation syndrome* occurs far more frequently in esotropes than exotropes [44]. In both situations it is characterized by a small manifest deviation (8 Δ or less), a larger latent deviation, and subnormal fusion. In esotropes, the latent deviation is more likely to remain latent, but in exotropes it often decompensates to an intermittently manifest deviation. When this occurs, it can be difficult to distinguish from an X(T) relying on the motor exam alone. Often the constant manifest deviation is tiny (2–4 Δ), and unless one carefully looks for it on the cover-uncover test, it will be overlooked. It can be important to recognize this entity prior to surgery, as its presence will alter expectations for the sensory outcome after surgery; it will be subnormal fusion at best.

There is controversy as to whether or not surgery in very young intermittent exotropes can predispose to their losing bifoveal fusion and end up with monofixators. My approach is to temporize and defer surgery until a child is 2½–3 years of age, unless I cannot get them to control satisfactorily with nonsurgical means.

### Basic Information

*Consecutive XT*

A patient who develops an XT after surgery for ET has a consecutive XT. Nonsurgical management options are similar to those for primary XT, with a few exceptions. Anti-suppression patching is less effective, because these patients do not have the same underlying sensory adaptation that occurs with X(T). Because many esotropes are hyperopic, one does have the option of cutting plus in the spectacles to stimulate accommodative convergence and decrease the XT. Although I often do that with hyperopic consecutive exotropes, I have found in most cases that it is only a temporizing measure [45]. Cutting plus by 3 D will only correct 15–18 Δ of XT, and cutting it more introduces a lot of instability in the alignment, as the patient varies from accommodating to relaxing accommodation. If I have done this with a consecutive exotrope, and they are either not well aligned or asthenopic with the plus cut, I will then put them in close to their full plus, cutting it by about 1 D, and operate for the angle the measurement obtained with those glasses.

The choice of surgical procedures depends mainly on what was done previously, and if there is a limitation of adduction. If adduction is full, and a R&R had previously been done, I recommend a R&R in the other eye. If prior MR recessions were performed, I recommend recessing the LRs. In both scenarios, no adjustment need be made in the surgical numbers because of the prior surgery. However, if there is an underaction of adduction, this must be addressed. If the prior surgery was a R&R, forced ductions are needed to tell if the lateral rectus muscle is too tight. The MR must be explored to see if it slipped or has an elongated scar. In either situation, the underlying problem should be addressed. See Chap. 11 for further discussion.

### Pearl

I have found that if a consecutive exotrope has a near deviation that exceeds the distance by even a few prism diopters, results with subsequent LR recessions produce very disappointing results with a high undercorrection rate—even if my clinical assessment of adduction was normal [30]. I now take this pattern of mild convergence insufficiency to be a soft sign for MR underaction. These patients do much better with MR advancement.

> Even a mild convergence insufficiency pattern with consecutive exotropia is a soft sign of medial rectus muscle underaction.

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# A-Pattern, V-Pattern, and Other Alphabet Pattern Strabismus

7

## Basic Information

*Overview and Definitions*

The terms *A-pattern* and *V-pattern* describe horizontal strabismus that is vertically incomitant and has a substantial difference in the horizontal deviation between the midline upgaze and midline downgaze positions. A patient with a V-pattern is more esotropic or less exotropic in downgaze than upgaze and an A-pattern is characterized by the converse. By convention, the difference between upgaze and downgaze must be 15 prism diopters (Δ) or greater to diagnose a clinically significant V-pattern and 10 Δ to diagnose an A-pattern. Less commonly, there are variations of pattern strabismus in which there is minimal change from downgaze to the primary position, but the eyes diverge in upgaze, resulting in a Y-pattern. Converse to a Y-pattern, the main exo shift may be between the primary position and downgaze to form a "λ" (lambda) pattern. Divergence in both upgaze and downgaze constitutes an X-pattern.

## Basic Information

*Occurrence*

Because ethnic and systemic factors influence the incidence of pattern strabismus, estimates range from 12 to 50% of patients with strabismus [1–4]. Alphabet pattern strabismus occurs much more frequently with congenital or paralytic strabismus than with non-paralytic acquired strabismus. Duane syndrome often presents with a V- or Y-pattern; less commonly an A- or λ-pattern can occur. There is also a high incidence of alphabet pattern strabismus in patients with craniofacial abnormalities.

## Advanced Information

A-patterns with overdepression in adduction are very frequently seen in patients with spina bifida and/or hydrocephalus. Frontal bossing in children with hydrocephalus may result in anterior displacement of the trochlea, mechanically enhancing the vertical vector of the tendon of the superior oblique muscle (SO), thereby strengthening the depressing action of the SO, and this may be the cause of the A-pattern. However, the exact mechanism that causes A-patterns to occur with hydrocephalus is unclear.

## Basic Information

*Etiology*

Because different mechanisms may be responsible for alphabet pattern strabismus in different patients, there are multiple theories as to etiology.


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## Oblique Muscle Dysfunction

Probably the most popular theory that accounts for the majority of cases of A- or V-patterns implicates oblique muscle overaction (OA) or underaction (UA) as the cause. If the inferior oblique muscles (IOs) are overacting and the antagonist SOs are underacting, one would expect a relative convergence in downgaze and divergence in upgaze, resulting in a V-pattern. The converse occurs if the SOs are overacting and the IOs are underacting, resulting in an A-pattern. One typically finds the oblique muscles to be dysfunctional in this manner in most patients with pattern strabismus.

## Torsion as a Cause of A- and V-Patterns

Patients with a V-pattern typically have an excyclotropia, which is most likely secondary to the accompanying IO OA. This excyclotropia results in rotation of the rectus muscles (Fig. 7.1) [5]. The superior rectus muscles would become partial abductors and the inferior rectus muscles (IRs) partial adductors, which will contribute to a V-pattern. The medial rectus muscles (MRs) will also have elevating force vectors and the lateral rectus muscles (LRs) depressing force vectors, contributing to the elevation seen in adduction with overacting IOs (Fig. 7.2). Thus the torsion that accompanies oblique muscle dysfunction should theoretically cause or contribute to A- and V-patterns, but not necessarily be the main cause [6].

## Question

If torsion is present, is it the main cause of an A- or V-pattern. If so, can just correcting the torsion correct the pattern?


## Reply

I believe that torsion is only a minor contributing cause of pattern strabismus rather than the main cause, for several reasons [5]. First, the rise or fall of an eye with oblique muscle OA, as it moves

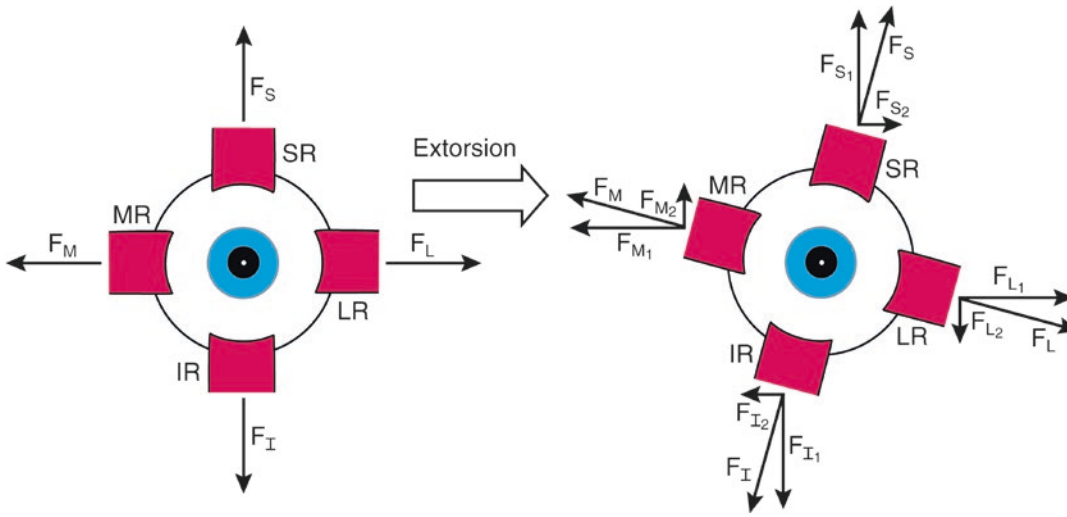


**Fig. 7.1** Effect of torsion on individual muscle function. An excyclorotation of the left eye will result in a clockwise rotation of the insertion of the muscles. This will create a vector for elevation for the muscle, abduction for the superior rectus, depression for the lateral rectus, and adduction for the inferior rectus (from Kushner [5], with permission. © 2010 American Medical Association. All rights reserved)

into adduction, is curvilinear increasing exponentially, rather than linearly, as the eye moves into adduction. If the rise or fall were primarily caused by the change in force vectors of the rectus muscles as seen in Fig.7.1, one would expect a linear trajectory (Fig. 7.3). Secondly, it has been shown that objective extorsion may precede the development of overelevation in adduction and a V-pattern in patients with infantile esotropia, by as much as several years [7]. If the torsion caused the overelevation in adduction and the pattern, they should occur concurrently. Thirdly, surgery in the form of vertical transposition of the horizontal rectus muscles that successfully eliminates

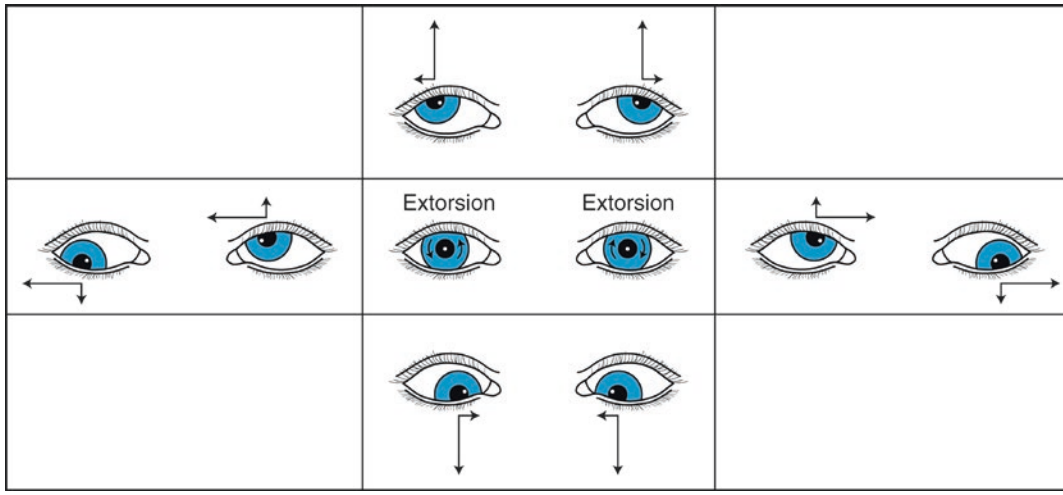


**Fig. 7.2** Effect of torsion on motility pattern. If the torsional changes that are depicted in Fig. 7.1 occurred in both eyes, the new force vectors would cause divergence in upgaze and convergence in downgaze. In addition, there would be an elevation of the adducting eye and depression of the abducting eye. Thus, these torsional changes that occurred as a result of extorsion contribute to both the V-pattern and the elevation seen in adduction (from Kushner [5], with permission. © 2010 American Medical Association. All rights reserved)

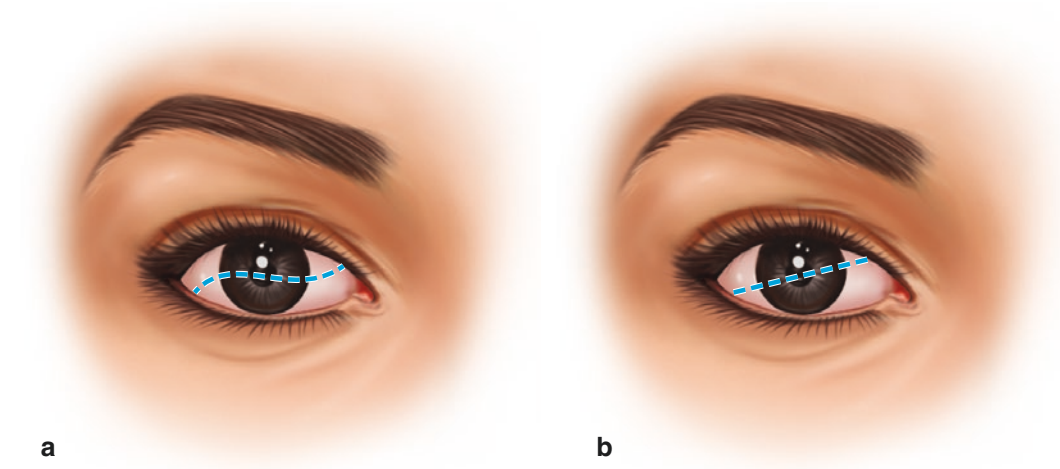


**Fig. 7.3** (**a**) Typical rise of the eye with "inferior oblique muscle overaction" as it moves into adduction is curvilinear as denoted by the *dotted line*. (**b**) Expected rise of the eye with "inferior oblique overaction" should be linear if the overelevation was due to the force vector changes shown in Fig. 7.1 as denoted by the *dashed line* (adapted from Kushner [5], with permission. © 2010 American Medical Association. All rights reserved)

an A- or a V-pattern will predictably worsen the underlying torsion (see section *Advanced Information: Horizontal Transposition of Vertical Rectus Muscles*, below) [8]. Finally, surgery like the Harada-Ito procedure, which mainly corrects torsion, has a negligible effect on the overelevation in adduction in patients with fourth cranial nerve palsy, even when it eliminates the excyclotropia. Thus correcting the torsion alone will not correct pattern strabismus if the surgical plan does not address the vertical and horizontal components of the extraocular muscles.

## Advanced Information:

*Orbital Structural Anomalies*

Orbital anomalies are often associated with A- and V-patterns. In addition to the high occurrence of alphabet patterns in craniofacial syndromes, there is a frequent occurrence of A-pattern esotropia accompanied by IO UA in patients with upslanting palpebral fissures and an association of V-pattern exotropia with IO OA. In patients with downslanting palpebral fissures the opposite occurs. There is also a very high incidence of A- and V-patterns in patients with craniofacial syndromes. Clark et al. [9] and Demer [10] have attributed some cases of A- and V-patterns to orbital pulley heterotopia or laxity. This is an evolving concept with no good data as to the frequency that pulley issues might be causative of pattern strabismus.

## Basic Information:

*Iatrogenic Causes*

An A- or a V-pattern may develop as a result of prior strabismus surgery. A common scenario is the development of a marked Y-pattern that can occur in the form of the anti-elevation syndrome after anterior transposition of the IOs [11]. There is overelevation on attempted elevation in adduction, which looks like IO OA. In fact it, is due to fixation duress secondary to limitation of elevation of the abducted eye.

A second common iatrogenic cause for alphabet pattern strabismus is the development of an A-pattern following large bilateral recessions of the IRs, commonly in thyroid eye disease. This occurs from a loss of the adducting effect of the IRs in downgaze secondary to surgical weakening, and by an increase in innervation to the yoke SOs.

Iatrogenic alphabet patterns can also result from an overcorrection from prior treatment of A- or V-pattern strabismus.

## Advanced Information:

*Horizontal Rectus Muscles*

Urist felt that OA or UA of the horizontal rectus muscles was responsible for A- and V-patterns [1, 12] and that the MRs were more active in downgaze and the LRs were more active in upgaze. His surgical recommendations involved weakening the offending muscles. Although this theory is less compelling than others, it may explain the occurrence of some cases of A- or V-pattern where there is no other apparent cause. It can also explain the small decrease in V-pattern observed after bilateral MR recessions. Recent work by Demer and coworkers has described distinct and separate innervation to the superior and inferior compartments of the horizontal rectus muscles [13–15]. In theory this could give rise to A- or V-patterns if the superior and inferior compartments are recruited separately on upgaze or downgaze. At this point the role of compartmental innervation on A- or V-patterns is not clear and still evolving.

## Advanced Information:

*Other Less Popular Theories*

Gobin suggested that the primary event that causes a V-pattern is an abnormal sagittalization of the IOs, which decreases its vector for torsion, resulting in an

incyclotropia. Subsequently, to correct this abnormal incyclotropia, there is increased innervation to the excyclorotary muscles, resulting in IO OA [16]. In my opinion, this theory is inconsistent with the clinical findings of torsion in patients with pattern strabismus. According to the sagittalization theory, a patient with a V-pattern should have no fundus torsion, as the IO OA developed to serve the purpose of eliminating an abnormal intorsion. In fact, these patients typically have objective extorsion.

Brown felt that A- and V-patterns were caused by a weakness of the IOs and SOs, respectively, resulting in apparent OA of the yoke oblique muscles due to fixation "duress" [17]. This theory is not widely accepted.

## Basic Information

*Clinical Presentation*

The manner of presentation of a patient with an A- or a V-pattern depends on the underlying strabismus and the size of the deviation. If there is a sufficiently large deviation in the primary position such that, despite the presence of an A- or a V-pattern, there is no head position in which fusion is possible, the pattern may not affect the presentation. If the deviation is small enough that fusion is possible in upgaze or downgaze, the patient may assume a chin-down or chin-up head posture in order to fuse. Some adults with an A- or a V-pattern may not become symptomatic until they become presbyopic, and they need to get their eyes into downgaze to read through a bifocal segment. Prior to becoming presbyopic they may have unconsciously held reading material closer to the primary position. Similarly, some presbyopic patients with strabismus that is only significant in downgaze may become symptomatic if they switch from a flat top to a progressive bifocal which has a wide transition zone that requires the eyes to move further into downgaze in order to permit vision through the prescribed add.

## Examination

### Motor Exam

The diagnosis of an A- or a V-pattern must be made by measuring with the prism and alternate cover test 25−30°above and below the midline. Testing should be done at 6 m to eliminate the near reflex. It should be done with full optical correction in place and while fixation is maintained on an accommodative target. If this is not done, a pseudo A- or V-pattern may be observed.

Special care should be taken to assess any relative overelevation or overdepression in adduction, and the presence or absence of fundus torsion should be assessed with indirect ophthalmoscopy.

## Important Point

You can be misled by assessing the size of the deviation on version testing using a light reflex, as this will not bring out the latent deviation. Particularly in patients with intermittent exotropia, the deviation may be controlled in the primary position; however, fusion may break down when the eyes are rotated into extreme fields of gaze giving the false appearance of a Y-, λ-, or X-pattern.

## Sensory Findings

Sensory findings depend on whether the patient is orthotropic in any field of gaze. Patients who are well aligned in the primary position may have surprisingly good fusion. If the patient is tropic in all fields of gaze, suppression and varying depths of abnormal retinal correspondence (ARC) may be found. In many patients with ARC and pattern strabismus, the angle of anomaly varies with the angle of deviation, thus resulting in ARC that is harmonious in all fields of gaze. This speaks to the fluidity of ARC.

## Surgical Treatment of Alphabet Patterns

If a pattern needs to be eliminated, the treatment is surgery. However, first you need to determine whether the pattern itself needs to be addressed. If the patient has a significant chin-up or chin-down head posture in order to fuse, surgery that addresses the pattern is appropriate. A continuous change in the size of a deviation that varies with change in gaze direction is destabilizing for binocular fusion. Thus if an A- or a V-pattern is clinically significant, it should be addressed in any child undergoing horizontal strabismus surgery, in whom there is any likelihood of some degree of binocularity being gained. This is probably the case in all children unless there is dense amblyopia. You should also pay attention to the location of the pattern. The primary position and downgaze are the two most important fields of gaze. You might ignore the pattern completely in an asymptomatic patient with a Y-pattern who only manifests a deviation in upgaze. This is particularly true if treating a pattern might be at the expense of the good alignment in the primary position and downgaze. If the strabismus is cosmetically unacceptable because of an A- or a V-pattern, surgery for the pattern is also appropriate. For most adults undergoing surgery for other reasons, a pattern should be treated if present. Exceptions include patients in whom treating the pattern might adversely affect the outcome in the primary position or downgaze, or if dense amblyopia is present. In these situations, one may decide to ignore the pattern.

First you need to determine if the pattern needs to be treated.

### Important Point

Do not sacrifice good alignment in primary and midline downgaze by correcting an upgaze misalignment.

### Basic Information

If you decide to treat a pattern in a patient in whom there is substantial oblique muscle OA, the overacting oblique muscles should be weakened because doing so:

1. Decreases the excessive abducting force in the gaze direction in which the eyes diverge (upgaze for a V-pattern, downgaze for an A-pattern)
2. Decreases the torsion that contributes to the pattern and may be an obstacle to fusion
3. Corrects any cosmetically unacceptable upshoot or downshoot occurring in adduction

Oblique muscle surgery should be combined with any necessary horizontal surgery, the latter based on the angle of misalignment in the primary position. No allowance is necessary for the loss of abducting force of the oblique muscles, because it is negligible in the primary position. Weakening both IOs will result in 15–25 Δ of esotropic shift in upgaze, depending on the degree of OA of the IOs; the greater the degree of OA, the more effect is obtained. There will be little or no effect on the horizontal deviation in the primary position. Initially, there will also be no horizontal effect in downgaze from weakening the IOs. Later, however, there may be some increased divergence in downgaze because the previously underacting SOs may recover function after the antagonist IOs have been weakened. This effect is frequently in the range of 10–15Δ.

The effect of weakening the SOs depends on the technique. Weakening the SOs nasally, by any technique, has a large effect and can correct up to 40 Δ of exotropia in downgaze. Weakening the SOs temporally is a less powerful operation, but is also less likely to cause complications. A posterior 7/8 tenectomy will cause a reduction of approximately 15–20 Δ of exotropia in downgaze [18, 19]. A somewhat more powerful operation than a posterior tenectomy is a complete disinsertion of the SO tendon. A still greater

effect can be obtained with a tenectomy of the SO tendon at the insertion or a graded recession. Weakening both SOs has no effect on the horizontal deviation in upgaze, both short-term and long-term. In the primary position it will only correct between 0 and 3 Δ of exotropia [20]. You need no adjustment, or perhaps up to 3 Δ, in your horizontal surgery if you do bilateral SO weakening.

If you weaken both IOs or SOs, do not alter your surgical dose for concurrent horizontal surgery.

When treating a pattern by weakening oblique muscles, it is important to perform the surgery symmetrically unless you are also trying to correct a vertical deviation in the primary position. Otherwise you will create an unwanted vertical strabismus. Complete weakening procedures of the SOs, such as nasal tenectomy or tendon expansion, have a powerful effect on torsion. These procedures may lead to a disruption of fusion in a patient with bifoveal fusion and can create postoperative torsional diplopia in a fusing patient. Exercise extreme caution when weakening the SOs in such patients! A posterior tenectomy procedure may be adequate for addressing the pattern and is safer than doing a complete SO tenectomy or tendon expansion. Alternatively, it might be prudent to address the pattern with vertical transpositions of the horizontal rectus muscles and just decrease, rather than eliminate, the pattern if it is large.

Strong weakening procedures of the SOs can lead to torsional symptoms in patients with bifoveal fusion.

If oblique muscles are not overacting, they should not be weakened when treating an A- or a V-pattern. Finally, V-patterns with SO can be treated with SO tucks.

## Important Point

The IO is more forgiving than the SO. Consequently, my threshold for weakening IOs with a V-pattern is lower than for weakening SOs with an A-pattern. If a V-pattern mandates treatment, I might weaken IOs if they are only +1 or +2 overacting. However, I tend to want more significant SO OA before I will weaken them when treating an A-pattern.

## Basic Information

*Vertical Transpositions of the Horizontal Rectus Muscles*

If there is no significant oblique muscle dysfunction, many patients with A- or V-pattern can be treated effectively with transpositions of the horizontal rectus muscles combined with the usual recession or resection you would perform based on the primary position deviation. You should not make any adjustment in your standard surgical formula because of the transposition.

Horizontal rectus transpositions are based on the principle that, when a rectus muscle is transposed, its primary action is decreased when the eye rotates into the field of gaze toward which the muscle was moved, and its action increases when the eye moves in the opposite direction (Fig. 7.4) [21]. For example, if an MR is transposed inferiorly, it becomes a weaker adductor when the eye is in downgaze and a greater adductor when the eye is in upgaze. This occurs because the insertion of the transposed muscle has a new relationship to the center of rotation of the globe as seen in Fig. 7.4. Thus, for treating a V-pattern esotropia, you can recess and infraplace the MRs, because downgaze is the field in which you want them to have less adducting action. However, when transposing rectus muscles, it is important to consider that two additional changes in their function occur simultaneously (Fig. 7.5). One effect is the creation of a force vector in the direction in which the muscle is moved. Thus, if an MR is infraplaced, an additional force vector for depression is established. For this reason, it is important to always use this treatment symmetrically; otherwise, an unwanted vertical deviation in the primary position will be induced. The exception might be those circumstances in which you want

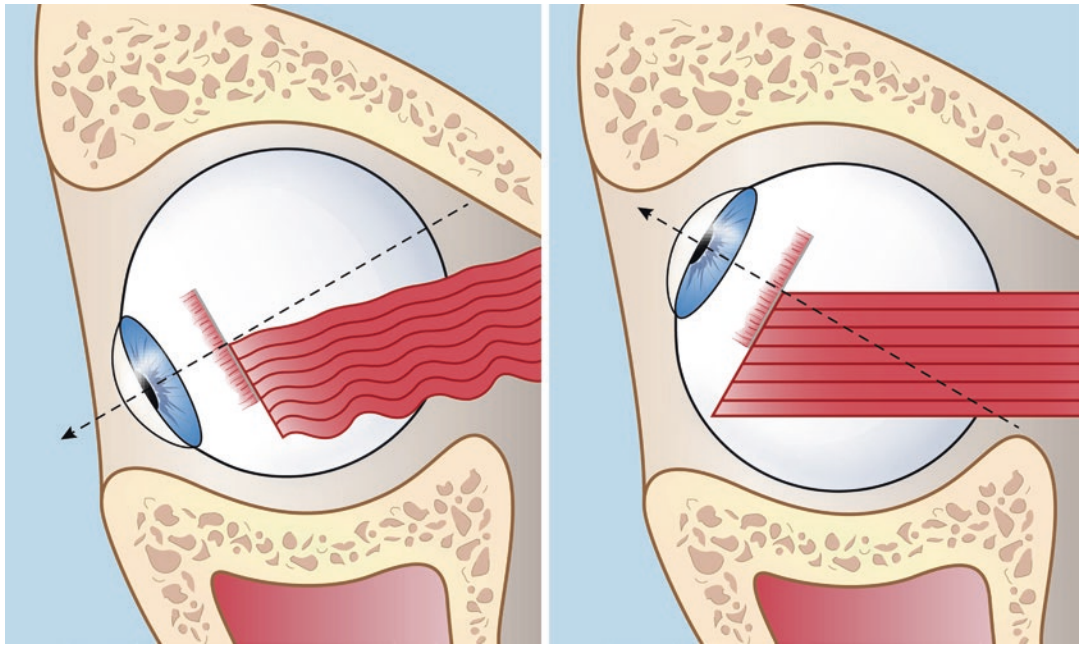

**Fig. 7.4** Effect of vertical transposition of rectus muscle on its primary action. If a muscle is infraplaced, there will be more slack in the muscle when the eye is in downgaze (*left*) than in upgaze (*right*). This weakens the primary action of the muscle more in downgaze than in upgaze (from Kushner [21], with permission. © 2010 American Medical Association. All rights reserved)

to treat a preexisting primary position vertical deviation, in which case you might perform a unilateral transposition or a bilateral transposition that is asymmetric.

The other effect of transposition of rectus muscles relates to torsion. When a muscle is transposed, a torsional vector is created in the direction from which the muscle is moved. For example, moving an MR inferiorly creates a force vector that results in extorsion as shown in Fig. 7.5. Importantly, this torsional rotation is in the opposite direction to the torsional correction one desires when treating most A- or V-pattern. A V-pattern esotropia is typically associated with IO OA and excyclotropia. Transposing the MRs inferiorly successfully treats a V-pattern but makes excyclotropia worse [8]. This rarely results in adverse symptoms, probably because most patients with A- and V-patterns do not have bifoveal fusion, and also because the amount of torsional change is small.

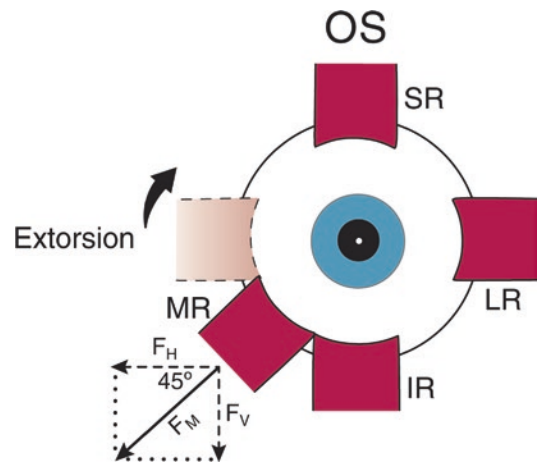


**Fig. 7.5** Multiple effects of transposition of rectus muscle. If a medial rectus muscle is infraplaced, a new force vector for depression will be created. In addition, a torsional force vector will be created in the direction toward the original insertion. With infraplacement of the medial rectus, a vector for extorsion is created (from Kushner [5], with permission. © 2010 American Medical Association. All rights reserved)

## Important Point

Three effects always occur when you transpose a rectus muscle: (1) The muscles' primary action is weakened when the eye rotates in the direction the muscle is transposed; (2) a force vector is created in the direction the muscle is moved; and (3) a torsional vector is created in the direction of the original insertion of the muscle.

## Important Point

When you transpose to treat torsion, you will typically worsen the accompanying alphabet pattern; when you transpose to treat an alphabet pattern, you will worsen the accompanying torsion.

## Question

My patient had an A-pattern with moderate SO OA. She had a negligible exophoria in primary but was 12 Δ exotropia in downgaze at near. She was asymptomatic until she became presbyopic and needed a bifocal for reading, and she was diplopic in the segment because it required downgaze. I was hesitant to weaken the SOs in this woman with bifoveal fusion, so I transposed both IRs nasally to treat the downgaze exotropia. This eliminated the exotropia, but now she has a symptomatic incyclotropia. What happened, and what should I do?


## Reply

This is not an unexpected outcome. Nasal transposition of the IRs will decrease the downgaze exotropia but predictably will create a vector for intorsion. I advised this colleague to undo the transpositions and do posterior SO tenectomies. This corrected the problem.

## Basic Information

*Which Way to Transpose?*

A simple mnemonic for remembering the direction in which to transpose the horizontal muscles for treating pattern strabismus is presented in Fig. 7.6. The MRs are always transposed toward the apex of the pattern (down for a V-pattern and up for an A-pattern), and the LRs are always transposed toward the open space of the pattern (up for a V″ pattern and down for an A-pattern). This holds true whether the muscles are recessed or resected, and if the patient is esotropic or exotropic. Vertical transposition of the horizontal rectus muscles is generally effective for treating pattern strabismus if there is no significant oblique dysfunction, but is less effective if the obliques are significantly overacting.

## Question

I prefer to avoid oblique muscle surgery whenever possible, especially on the SO. Why are rectus muscle transpositions less effective if there is oblique muscle dysfunction?

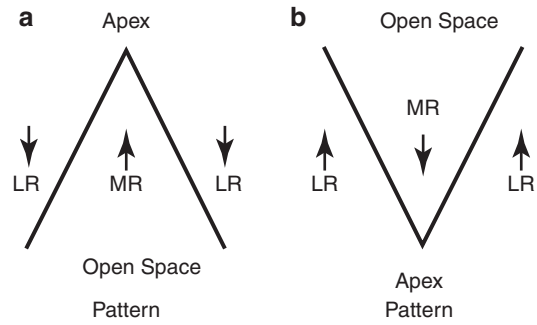


**Fig. 7.6** Mnemonic for transposing horizontal rectus muscles. This diagram depicts the directions to move the horizontal rectus muscles for the treatment of an A-pattern (*left*) and a V-pattern (*right*). The lateral rectus (LR) muscles are always moved toward the open space and the medial rectus (MR) muscles always move toward the apex of the pattern


## Reply

If there is oblique muscle dysfunction, there is typically objective torsion (fundus torsion)—excyclo with V-patterns and IO OA and incyclo with A-patterns and SO OA. There usually is no subjective torsion due to a torsional sensory adaptation. Transposing the horizontal rectus muscles to treat the pattern will worsen the already existing torsion. And as outlined earlier, torsion itself will contribute to an alphabet pattern.

## Basic Information

I find that, in most cases of A- or V-pattern without oblique muscle dysfunction, a symmetric vertical transposition of the appropriate horizontal rectus muscles by one-half tendon width (5 mm) is effective in reducing an average of 15–20 Δ of pattern. In most cases in which the pattern is more than 20 Δ, there is usually significant oblique muscle dysfunction, and I usually address the pattern with oblique muscle surgery. If oblique muscle surgery is not indicated in a patient with a large pattern (perhaps it has already been performed but a pattern persists), transposition of the appropriate horizontal rectus muscles three-quarters or a full tendon width can be effective for patterns that are greater than 20 Δ. Also, transpositions of the horizontal rectus muscles can be combined with oblique muscle surgery. A patient with IO OA plus SO UA might benefit from bilateral MR recessions with infraplacement and bilateral IO weakening.

## Advanced Information

You can also apply the principles of transposition when doing0 unilateral surgery in the form of a recess/resect procedure. One muscle can be raised and the other lowered. Keep in mind, however, that doing so does not create an equal balance of forces. If you raise a weakened (recessed) muscle the same amount as you lower a tightened (resected) muscle, you are creating an increased force vector in the direction the resected muscle was moved. I have seen unwanted vertical deviations created from this surgical approach. Although unilateral transposition is often successful, I prefer symmetric surgery when performing transpositions.

> Raising a horizontal rectus muscle and lowering its antagonist an equal amount do not balance the vertical forces.

## Advanced Information

*Horizontal Transposition of the Vertical Rectus Muscles*

Horizontal transposition of the vertical rectus muscles can treat A- and V-patterns [22, 23]. The theoretical efficacy of this approach is based on a different principle than that of vertical transposition of the horizontal rectus muscles. It is predicated on one of the effects depicted in Fig. 7.5— specifically that a force vector is created in the direction a muscle is moved. This same principle is the basis for transposition procedures to treat paralytic strabismus. Thus a V-pattern esotropia can be treated by temporal transposition of the IRs along with appropriate recessions of the MRs; typically, the vertical rectus transpositions are 7 mm. A summary of the directions to transpose the vertical rectus muscles for treating A- and V-patterns is presented in Table 7.1.

Notably, horizontal transposition of the vertical rectus muscles may create a torsional shift that exacerbates a preexisting cyclotropia for the reasons depicted in Fig. 7.5. This is particularly relevant in treating patients with thyroid eye disease with large bilateral IR recessions, which often results in a postoperative A-pattern with intorsion. I have seen such patients in whom the IRs were transposed nasally at the time of recession in hope of preventing the occurrence of an A-pattern, and an unexpectedly large and

**Table 7.1** Summary of directions in which to transpose vertical rectus muscles

| Strabismus | Transposition |
|---|---|
| V-pattern esotropia | IR OU temporally |
| V-pattern exotropia | SR OU nasally |
| A-pattern esotropia | SR OU temporally |
| A-pattern exotropia | IR OU nasally |

*IR* inferior rectus muscle, *SR* superior rectus muscle, *OU* both eyes

symptomatic incyclotropia was created [8]. I prefer to combine large IR recessions with posterior tenectomy of the SO tendons to prevent an A-pattern in this setting.

Horizontal transposition of the vertical rectus muscles never gained popularity for treating routine cases of A- or V-pattern strabismus. I feel this is largely because it involves operating on an additional rectus muscle in each eye, which increases the risk of anterior segment ischemia. Doing so is not necessary when oblique muscle surgery or vertical transposition of the horizontal rectus muscles can be performed.

## Basic Information

*Pulley Abnormalities*

If pulley heterotopia or laxity is the cause of an A-pattern, surgery could be performed to stabilize or reposition the orbital pulleys. Diagnosis of pulley abnormalities requires precise orbital imaging [9, 10].

## Advanced Information

*Insertion Slanting Procedures*

Mathematical modeling of extraocular muscle mechanics indicates that one can theoretically consider an extraocular muscle as inserting at a point in the middle of its insertion (Fig. 7.7a) [21]. If there is unequal tension along the edges of the muscle, one can still consider the muscle as inserting at a point which

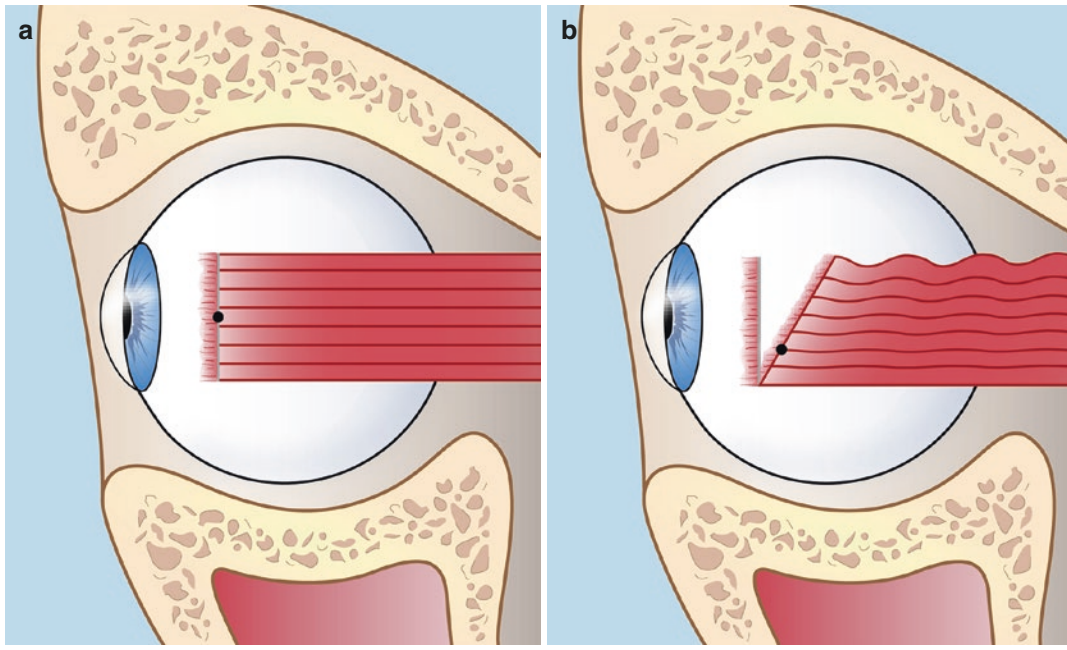


**Fig. 7.7** (**a**) Mathematically, one can consider a rectus muscle to insert at a point (*black dot*) at the middle of its insertion. (**b**) If there is unequal tension along the edges of a muscle, as occurs with a slanting procedure, the muscle still can be thought of as inserting at a point (*black dot*). However, the point shifts toward the edge under greater tension. If the superior pole of a horizontal rectus muscle is slanted back, the point shifts toward the inferior edge, which is akin to infraplacing the muscle (from Kushner [21], with permission. © 2010 American Medical Association. All rights reserved)

is shifted toward the edge with greater tension (Fig. 7.7b). In theory, this principle can be used to treat pattern strabismus by selectively slanting the superior or inferior pole of the recessed muscles to simulate transposition. For example, for a V-pattern esotropia the superior poles of the MRs are recessed more than the inferior poles, which simulates the effect of an infraplacement (Simonsz's method) [24]. This approach has been used successfully to treat pattern strabismus. Paradoxically, good results have been reported with slanting the muscles in an opposite manner by numerous authors [21, 25]. For a V-pattern esotropia these authors would recess the inferior pole of the MRs more than the superior poles (Bietti's method). The rationale for this approach is based on the erroneous theory that the inferior edge of a horizontal rectus muscle is more taut in downgaze and the superior edge more taut in upgaze. The authors arrived at this conclusion through a misinterpretation of known facts about extraocular muscle mechanics [21]. In fact, the opposite is true. This leads to a conundrum: How can opposite methods of slanting both produce good results (see Fig. 7.8)? For the same V-pattern esotropia some investigators would slant in the manner depicted in Fig. 7.8a and others would use the configuration shown in Fig. 7.8b. I believe that, despite their popularity, insertion slanting procedures in either manner should have a negligible effect on pattern strabismus. Sarcomere remodeling should rapidly occur and even out the edge tension after an insertion slanting procedure (Fig. 7.9). Probably the good outcomes reported with these procedures were the result of the recession or resection performed.

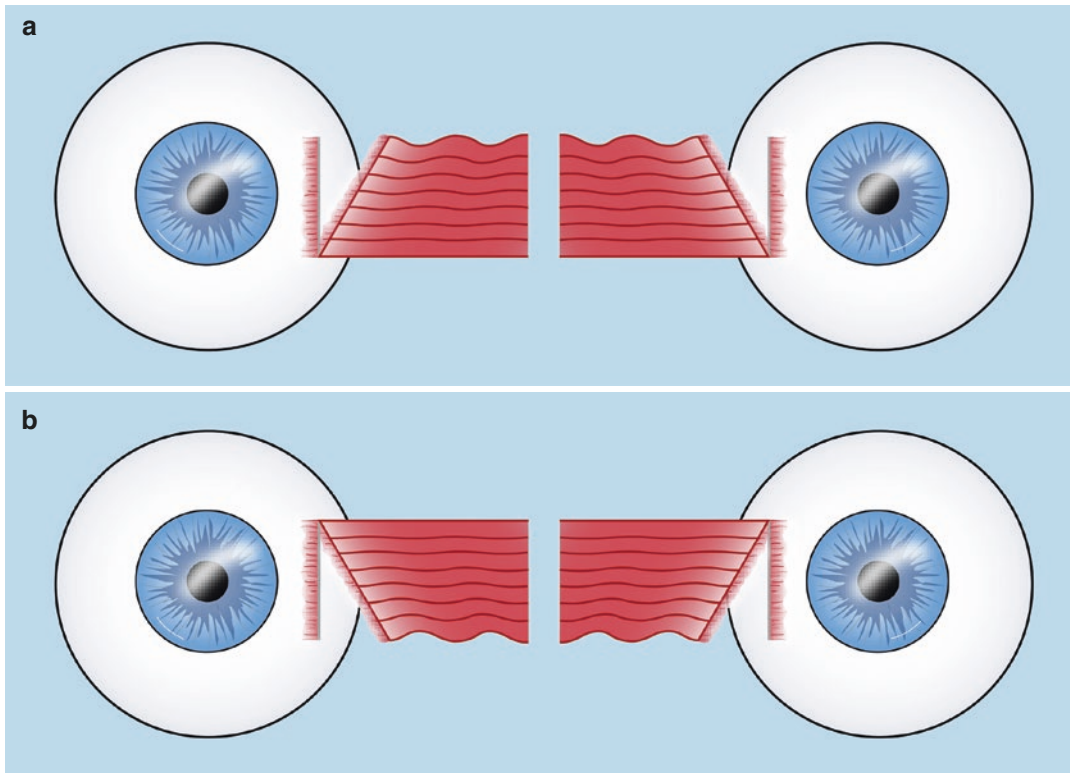


**Fig. 7.8** (**a**) The treatment of a V-pattern esotropia with insertion slanting according to the principles shown in Fig. 7.7. The superior poles of the medial rectus insertions are slanted back, which simulates infraplacement. (**b**) The treatment of a V-pattern esotropia according to the incorrect concept that the inferior fibers are more taut in downgaze (from Kushner [21], with permission. © 2010 American Medical Association. All rights reserved)

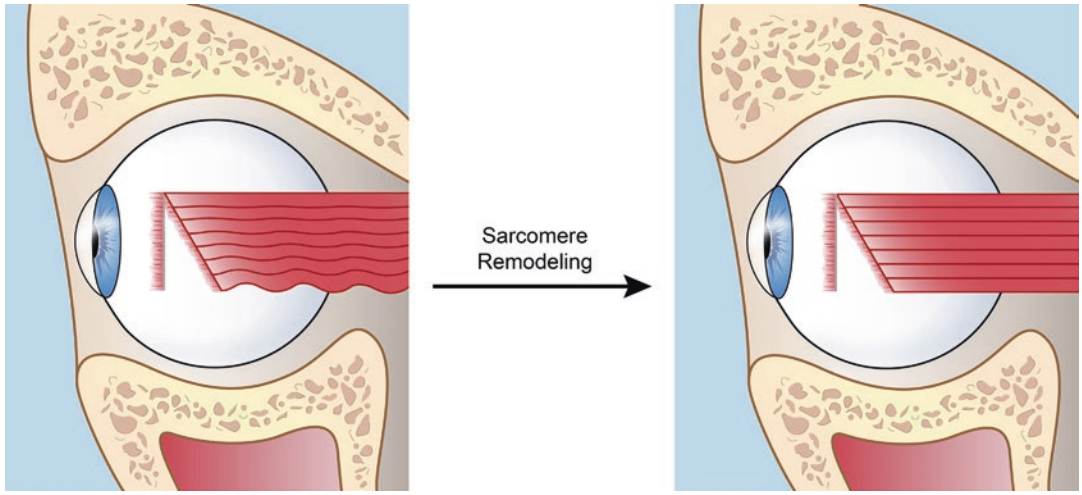


**Fig. 7.9** Initially after slanting back the inferior pole of a rectus muscle, there is substantial slack induced in the inferior muscle fibers (*left*). After a period of several weeks, sarcomere remodeling should even out the differential edge tension and largely negate the effect of the slanting procedure (*right*) (from Kushner [21], with permission. © 2010 American Medical Association. All rights reserved)

## Question

Don't the findings of compartmentalization of the innervation to the MR and LR provide justification to the concept of slanting procedures as being effective for A- and V-patterns, as well as for distance near differences like high AC/A esotropia or convergence insufficiency exotropia [13–15]?


## Reply

Not really. I know that the description of compartmentalization of the innervation to the MR and LR has led to speculation that this provides justification for slanting surgery. However, in order to close the circle, we need to not only know that there is compartmentalization, but also that there is selective recruitment to the upper and lower halves of the muscles on upgaze vs. downgaze (important for A- and V-patterns) and also between distance and near (important for high AC/A esotropia and convergence insufficiency exotropia correction). This is not what has been borne out. Demer and Clark found that although the inferior half of the MR recruits more on downgaze, this is not the case for the LR [26, 27]. Advocates of slanting profess that it works as well when done on the LR as on the MR. That cannot be explained by compartmental innervation if the recruitment to the inferior half of the MR and LR is different on shifts of vertical gaze. Similarly, no selective recruitment was seen for horizontal gazes, which would negate this concept as explaining the effect of slanting procedures for treating distance/near differences. Furthermore, Scott reported that he put electromyography (EMG) electrodes into the upper and lower halves of MR and LR on several occasions and found no difference in innervation as shown by EMG in these muscle areas with vertical frontal plane versions (Alan B. Scott, MD, personal communication, 18 Jul 2016). Consider the following:

1. Any theoretical justification of slanting needs to take into account that equally good results are reported with slanting in exactly opposite ways, e.g., slanting back the inferior pole of the MR for V-pattern esotropia (Bietti's approach) or slanting back the superior pole (Simonsz's approach).

2. As stated above, people are using slanting for both distance near differences (high AC/A esotropia) and convergence insufficiency exotropia, and up/down differences (A- and V-patterns). For compartmentalization to be the rationale behind slanting surgery, one would need to see similar changes in the inferior pole of both the MR and LR on vertical gazes, as well as on gaze going from distance to near and vice versa. Indeed, that was not found by Demer and Clark, whose speaks against compartmentalization being the justification for slanting.
3. Sarcomere remodeling should rapidly negate any effect of slanting.

My feeling is that slanting is nothing more than a recession or resection in which the dose is equal to an average of the recession or resection of the top and bottom poles. This ends up increasing the dose a bit by targeting something between the lesser and greater of the two measurements for distance/near differences.

## Question

When I see patients with large-angle exotropia, there is often an X-pattern with all four oblique muscles overacting. How does this occur, and is it necessary to weaken all four oblique muscles?


## Reply

There are two commonly held explanations for this phenomenon, both of which fail to explain many cases. Most commonly, this is attributed to tight LRs causing a leash effect simulating oblique muscle OA. The other explanation postulates that both SOs and IOs become contractured because they are shortened when an eye is in an exotropic position. Although both explanations are plausible, they fail to explain why this apparent oblique muscle dysfunction is typically present in both eyes, even when the strabismus is not alternating. This observation can, however, be explained by a mechanism proposed by Capo et al. [28]. They attributed the overelevation and overdepression of the adducting eye on side gaze to the shape of the orbit. If there is a large exotropia, the adducting eye is positioned closer to the horizontal center of the orbit on when the abducting eye is in far lateral gaze, and the oval shape of the orbital allows it to move both higher and lower than the abducting eye on elevation and depression, respectively, as seen in Fig. 7.10.

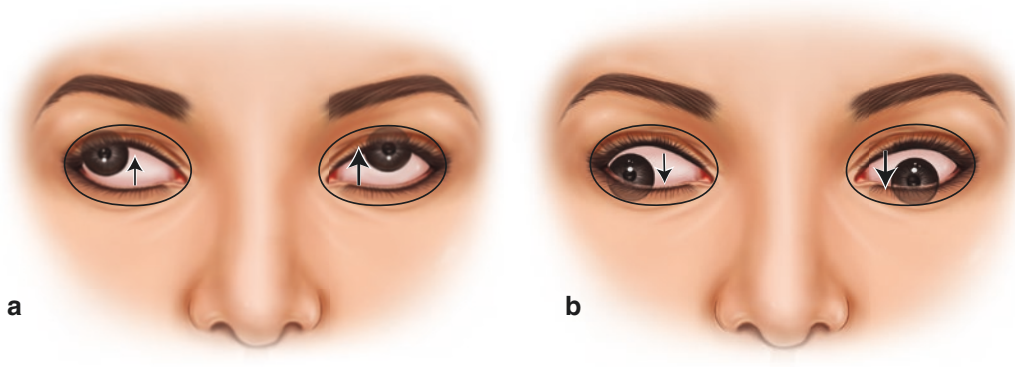


**Fig. 7.10** A patient with a large-angle exotropia looking to the right. (**a**) On attempted elevation (*top*) the left eye can move higher than the right due to the greater height of the orbit centrally than laterally, simulating L inferior oblique overaction. (**b**) On attempted depression (*bottom*) the left eye can move lower than the right simulating L superior oblique overaction

If this mechanism is at play, the oblique muscles do not need to be weakened. Intraoperative forced ductions to test for bilateral LR tightness and oblique muscle tightness should guide the surgical intervention.

## Summary of Surgical Planning

In all cases, I do the standard horizontal surgery based on the primary position measurement, except perhaps allowing for up to 3 Δ of esotropia shift in the primary position with SO weakening. Since my surgical dose table increases in 5 Δ increments, this has me either making no allowance in my horizontal surgery if I also do oblique muscle surgery or simply rounding up or down. For example, if I recessed MRs for a 23 Δ esotrope in whom I also weakened the SOs, I would target 25 Δ for my MR recession. Table 7.2 summarizes my recommendations for the surgical management of A- and V-patterns.

### Important Point

You should not make any allowance in your horizontal surgery for concurrently doing IO surgery, and no more than minimal allowance for doing concurrent SO surgery.

## Optical Management

Some patients with A- or V-pattern may have good alignment in the primary position but have an esotropia or exotropia in downgaze. As stated earlier, such patients may first become symptomatic as they become presbyopic. Some patients of this type can be managed by either using single-vision reading glasses, or having a separate bifocal for prolonged reading with a segment that is higher than is typically dispensed.

**Table 7.2** Summary of surgical recommendations

| If oblique dysfunction is present[a] | |
|---|---|
| V-pattern ET with IO OA | Recess MR or resect LR and weaken IO OU |
| V-pattern XT with IO OA | Recess LR or resect MR and weaken IO OU |
| A-pattern ET with SO OA | Recess MR or resect LR and weaken SO OU[b] |
| A-pattern XT with SO OA | Recess LR or resect MR and weaken SO OU[b] |
| Without oblique dysfunction[a] | |
| V-pattern ET | Recess MR and infraplace or resect LR and supraplace OU |
| V-pattern XT | Recess LR and supraplace or resect MR and infraplace OU |
| A-pattern ET | Recess MR and supraplace or resect LR and infraplace OU |
| A-pattern XT | Recess LR and infraplace or resect MR and supraplace OU |

*ET* esotropia, *IO* inferior oblique muscle, *OA* overaction, *MR* medial rectus muscle, *LR* lateral rectus muscle, *OU* both eyes, *XT* exotropia, *SO* superior oblique muscle

[a]Perform the usual amount of recession or resection of the horizontal rectus muscles based on the deviation in the primary position

[b]Avoid a powerful superior oblique weakening procedure in patients with bifoveal fusion

### Advanced Information

*Pseudo IO OA with Y- or V-Pattern*

There is a syndrome that is characterized by a Y- or less commonly a V-pattern with what appears to be marked IO OA [29]. In fact this is pseudo IO OA probably caused by aberrant innervation to the LR on elevation, or perhaps pulley heterotopia. The hallmarks of this syndrome are that in spite of a marked splaying out of the eyes on upgaze (+4), there is no SO muscle OA, no hypertropia (HT) on direct horizontal side gaze, and no fundus torsion (Fig. 7.11). It is important to recognize this syndrome because weakening the IOs will have absolutely no effect on the pattern or apparent IO OA. If there is no deviation in primary, you can treat conservatively. I have followed some

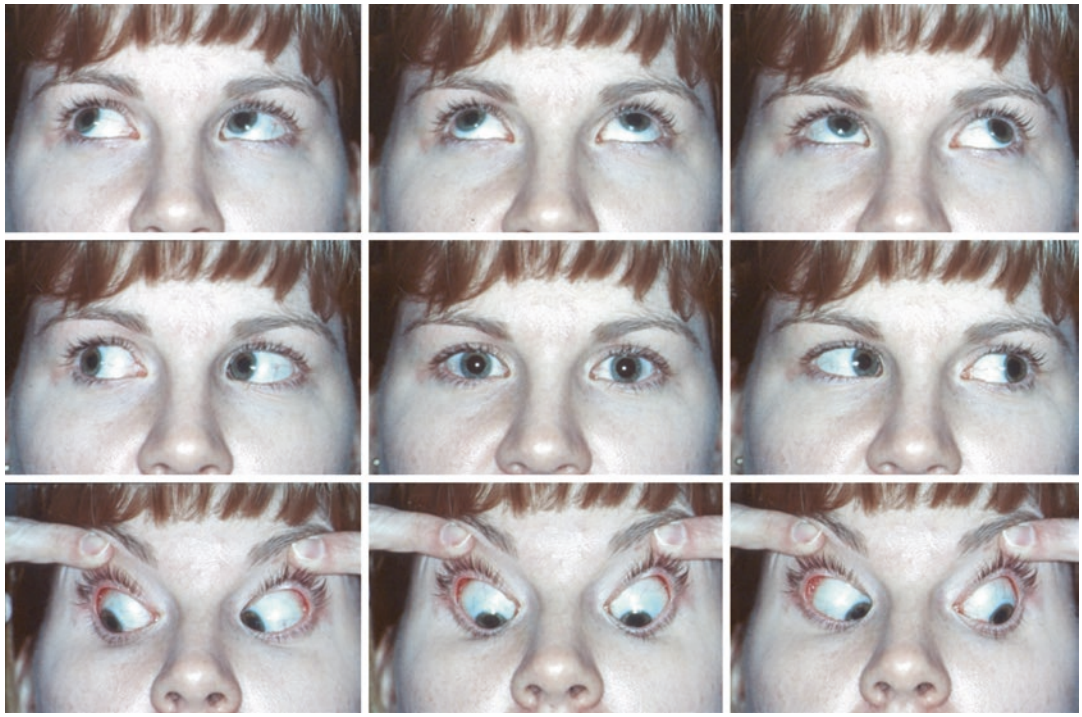

**Fig. 7.11** Patient with pseudo-inferior oblique overaction and a Y-pattern. There is a marked splaying out on upgaze, yet no superior oblique underaction, no hypertropia on side gaze, and no fundus torsion (from Kushner [29], with permission)

patients of this type for as long as 35 years, and they all remained stable. If there is a primary position deviation that mandates treatment, you can collapse the pattern by supraplacing the LRs three-fourths of a tendon width, combined with whatever horizontal surgery is indicated.

## Pearl

Weakening the IOs will have absolutely no effect on the motility pattern in patients with pseudo IO OA.

## Question

I have a patient that has pseudo IO OA. A prior ophthalmologist anteriorly transposed both IOs, apparently having failed to recognize this syndrome. That made the pattern worse. I think this is caused by fixation duress that increased the aberrant innervation to the LRs, which now makes the Y-pattern worse. I was thinking of converting the IO anterior transpositions to simple recessions. Would you agree?


## Reply

I think if you proceed as you suggest, the patient will be right back where she started. Simple IO recessions have no effect on this syndrome. I agree that the IO anterior transpositions should be reversed, as you suggest. But I would also supraplace the LRs three-fourths of a tendon each. That is the treatment for this condition.

## Question

How can you tell if a Y-pattern is caused by pulley heterotopia as opposed to aberrant innerva-

tion to the LRs? Is orbital imaging always indicated in patients with Y-patterns?


## Reply

The original description of Y-patterns as a form of aberrant innervation was from 1991 and hence before the possible role of pulley heterotopia was understood [29]. The relative frequency of these two etiologies causing Y-patterns is not yet understood. An interesting observation made by my colleague Struck may be useful in determining which of these two mechanisms may be causing a patient's Y-pattern (Michael C. Struck, MD, personal communication, 30 Nov 2016). Struck observed that if you have a patient with a Y-pattern look rapidly from downgaze to the primary position, some will show a momentary divergence of one or both eyes, and then rapidly converge to eliminate the exotropia. Not all patients with Y-patterns demonstrate this. I have subsequently observed this in patients who clearly have aberrant innervation, and found it absent in patients with pulley heterotopia as a cause of their Y-patterns. It makes sense that if there is aberrant innervation from the superior rec to the LR, there may be a sudden burst of abduction with saccadic movement from downgaze to the primary position. This may prove to be a useful sign for sorting out the etiology of Y-patterns.

## Pearl

A patient with a Y-pattern due to aberrant innervation may manifest a momentary exotropia on one or both eyes when making a saccadic movement from downgaze to the primary position.

## Question

I plan to operate on a 4-year-old boy with a moderate V-pattern esotropia. There is marked bilateral fundus extorsion; however, I see no more than a trace of IO OA, e.g., trace overelevation in adduction. Should I weaken the negligibly overacting IOs, correct the torsion with Harada-Ito surgery, or infraplace the MR (which I plan to also recess)?


## Reply

I understand the dilemma in this relatively uncommon patient. I presume you wonder about Harada-Ito surgery because you are considering that the torsion is the cause of the pattern due to the mechanism depicted in Fig. 7.2. However, if this mechanism is at play, and the rotation of the muscle insertions caused the V-pattern, it should also cause elevation in adduction and depression in abduction, which would be indistinguishable from what we call IO OA. Given that you do not see overelevation in adduction, I am hard-pressed to attribute the pattern to the torsion. In my experience, operating for torsion alone will not significantly help the pattern. In most cases, if there is a V-pattern and no IO OA, infraplacing the MRs is the procedure of choice. However, that will worsen the torsion, which you describe as marked. In this situation I would opt for weakening the IOs, in spite of their OA being minimal. That will still help the pattern and the torsion. If he does end up with a little underelevation in adduction, that would probably not be a clinically significant problem. I suspect it will not occur.

## Question

I have a patient with a V-pattern exotropia with +3 IO OA in the right eye and +4 IO OA in the left eye. There is a 10 Δ LHT in primary. I plan to do LR recessions and bilateral asymmetric IO recessions. Is this a good plan?


## Reply

I agree you need to do something asymmetrically to address the primary position. I find that smaller

HTs can be collapsed by asymmetric IO recessions. But with +3 in the right eye you will want to do a minimum of an 8 mm recession, and even with a 12 mm recession in the left eye you will probably not correct more than 6 Δ of HT in primary, with the difference between the two being only 4 mm. Another option would be to do standard (large) IO recessions, and also recess a vertical rectus muscle for the HTtropia.

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# Vertical Deviations

# 8

*What goes up, must come down.*

*—Origin unknown, circa early 1800s*

## Diagnosis

### Basic Information

Like horizontal deviations, vertical deviations can be paretic or non-paretic. Proper diagnosis of non-paretic deviations relies on history (infantile onset, prior surgery, trauma, etc.), and relevant systemic illness (thyroid eye disease [TED], etc.). The diagnosis of paretic deviations relies on the pattern of misalignment and ductions and versions. In general, the greatest deviation will be in the field of action of the paretic muscle, unless secondary changes have occurred in other muscles, e.g., contracture causing a spread of comitance. The Parks three-step test relies on this principle [1], taking the actions of the muscles on forced head tilt into account according to the teachings of Bielschowsky [1, 2]. The three-step test has become the mainstay in diagnosing vertical strabismus. However, it is only designed to tell which of the eight cyclovertical muscles might be palsied. It does not tell if you are in fact dealing with a palsy of one cyclovertical muscle. And when patient presents to you, it is your job to determine whether or not that is what your patient has. There are numerous causes of vertical strabismus for which the three-step test may incorrectly implicate one muscle as being paretic.

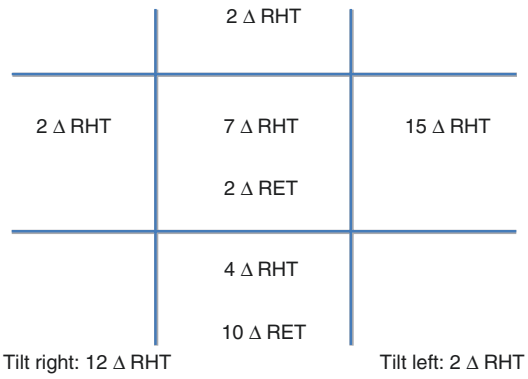
### Basic Information

*Errors in the Three-Step Test* [3]

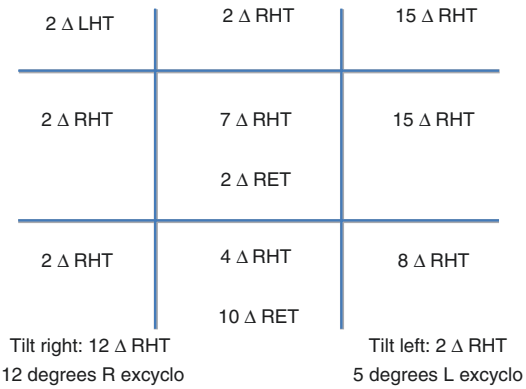
1. **Restrictions:** Although the majority of contractured or otherwise restricted muscles do not have a positive head tilt test, some do. I have seen patients with a restricted or contractured inferior rectus muscle (IR) that behaved as though it was overacting. There was an increase in the hypotropia (HYPO) with tilt to the contralateral side [3]. If the patient had decreased vision in the contralateral eye and fixated with the eye with the restriction, their motility pattern would mistakenly be attributed to a superior oblique muscle (SO) palsy (or SOP) )of the non-restricted contralateral eye. For example, if a patient had a restriction/overaction (OA) of his right IR, and fixated with the right eye due to amblyopia of the left eye, he would have a left hypertropia (LHT) that would increase in right gaze and on left head tilt. This would meet the three-step test criteria for a paretic left SO.
2. **Multiple muscle involvement:** The three-step test assumes that there is a palsy of one cyclovertical muscle. It will not tell you if more than one muscle is involved. Consider


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_8

the set of measurements (*RHT* right hypertropia, *RET* right-eye torsion, Δ prism diopters).




These measurements meet all the three-step test criteria for a right SO (RSO) palsy. But important clinical information is missing. If we add oblique field measurements and subjective torsional findings, the picture of a bilateral SO palsy becomes evident.




You can see that the RHT reverses up and right and becomes a LHT. If a patient with a RHT reverses and has a LHT in any field of gaze, including on head tilt, one cannot attribute the LHT to a palsy of the RSO. Something else must be going on. Also the three-step test does not take torsion into account. This patient has a total of 17° of excyclotropia. In general, a unilateral SOP will not have more than 8–10° of subjective excyclotropia.

**Pearl**

Oblique field measurements and torsion are important in diagnosing vertical strabismus.

If there is a reversal of the HT in any field of gaze or direction of head tilt (e.g., a RHT converts to a LHT), you cannot be dealing with a palsy of one cyclovertical muscle.

**Pearl**

A unilateral SOP will not have more than 8–10° of subjective excyclotropia. If there is more than 10, suspect a bilateral SOP.

3. **Prior vertical muscle surgery:** If prior vertical muscle surgery involved recessing a muscle, there would now be an iatrogenic ally weakened muscle in addition to the one that originally may have been paretic. Thus for the same reason that multiple muscle involvement may render the three-step test inaccurate, so can prior surgery.
4. **Dissociated vertical divergence (DVD):** DVD will increase in adduction if there is associated inferior oblique muscle (IO) OA. In some patients it may just appear to increase in adduction on version testing because the nose may act as an occluder. Classically, it is said that the DVD increases on contralateral head tilt, e.g., left DVD increases on right head tilt (see Fig. 8.1) [4]. In this respect the classic response of DVD to forced head tilt is the opposite of a SOP, which increases in ipsilateral tilt; for example, LHT would increase on left head tilt. If this classic behavior is followed, a left DVD would be diagnosed as a right superior rectus muscle (RSR) palsy by the three-step test. However, in

Some patients with DVD will assume a head tilt to control the DVD.

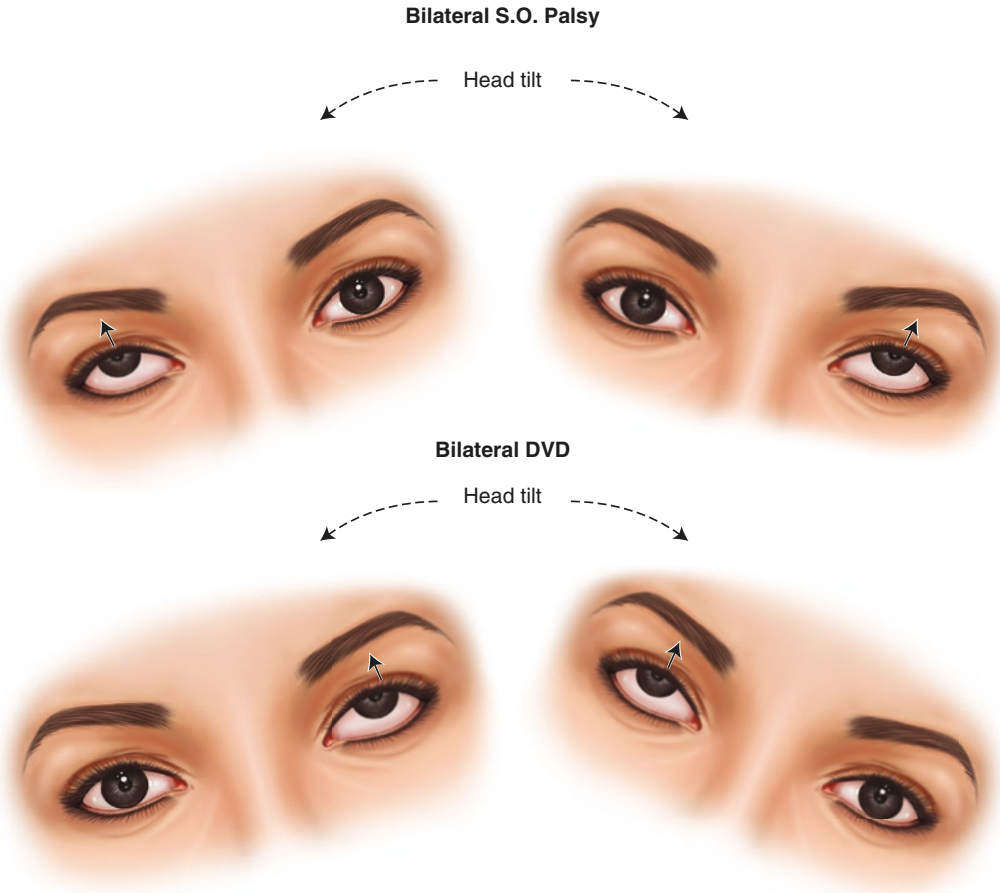



**Fig. 8.1** (*Top*) The hypertropia associated with superior oblique palsy (SOP) increases on ipsilateral tilt. This depicts bilateral SOP, which has a right hypertropia (RHT) on right tilt and an LHT on left tilt. (*Bottom*) DVD classically behaves the opposite of SOP with respect to head tilt. This depicts the classic response of bilateral dissociated vertical divergence to head tilt showing a left HT on right tilt and a right HT on left tilt. The three-step test would diagnose this as superior rectus muscle palsies

a prospective study I found that only 57% of patients with DVD showed an increase in the deviation on contralateral head tilt, and 19% had an increase on ipsilateral tilt [5]. The rest either did not increase in either direction, or had equivocal results. Thirty-five percent of the patients with DVD and a strong fixation preference had a spontaneous head tilt that served the purpose of controlling the DVD; none of the patients with no fixation preference and bilaterally manifest DVD assumed a spontaneous head tilt. DVD accounted for 9% of 100 consecutive patients with spontaneous head tilts.

5. **Skew deviation:** A skew deviation is a vertical strabismus caused by a demyelinating disease, or trauma with lesions in the brain stem or posterior fossa. In demyelinating disease, a skew deviation often accompanies a bilateral internuclear ophthalmoplegia, and thus the site of pathology is brain stem in this circumstance. Because skew deviation may have a positive Bielschowsky head tilt test, the three-step test will give an erroneous diagnosis. A useful test for skew deviation is to observe the change in the HT from the upright to supine position [6]. A decrease of 50% or more is strongly suggestive of a skew.

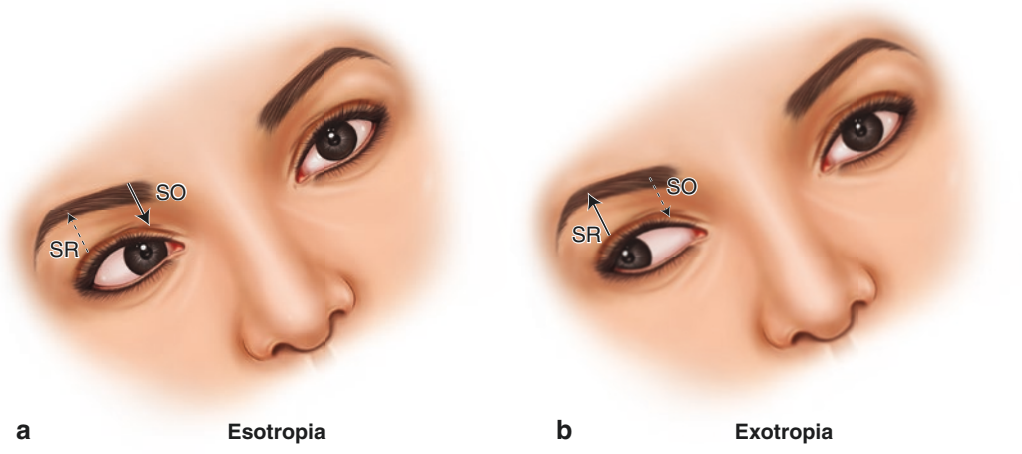



**Fig. 8.2** (**a**) For an esotropic patient, the eye is more in the field of vertical action of the right superior oblique (RSO) than the right superior rectus (RSR). With normal innervation to both muscles, there will be a force vector advantage for depression from the RSO, thus decreasing a right hypertropia (RHT) or increasing a right hypotropia. (**b**) For an exotropic patient, the eye is more in the field of vertical action of the RSR than the RSO. So with normal innervation to both muscles, there will be a force vector advantage for elevation from the RSR, thus increasing an RHT or decreasing a right hypotropia

6. **Myasthenia:** Because myasthenia can mimic a palsy of any of the extraocular muscles, the three-step test may give misleading results. Although in this setting the three-step test may correctly identify which muscle is weak, it is misleading because it will not identify the true cause of the vertical deviation, which is myasthenia. Correctly identifying myasthenia as the cause is essential for proper treatment.
7. **Small vertical deviations with horizontal strabismus:** Often patients with large-angle horizontal strabismus may have small vertical deviations that have a positive Bielschowsky head tilt test. In the orthophoric patient, on tilt to the right the RSR and RSO (both intorters) are stimulated. The elevating action of the RSR is cancelled by the depressing action of the RSO, and no vertical shift occurs. Moore and Cohen observed that exotropes typically show an increase in the HT on tilt toward the eye with the HT, and esotropes show a decrease in the HT on tilt in that direction [7]. They hypothesized that an exotropic eye is more in the field of the SR than of the SO, so the eye elevates, even though the SR function is normal. The converse is true with esotropia. The eye depresses because it is more in the field of action of the SO, even though its function is normal (Fig. 8.2). Although this mechanism will not cause horizontal incomitance (step 2 of the three-step test), most patients with vertical have some degree, however small, of horizontal incomitance. Such patients would therefore be misdiagnosed by the three-step test.
8. **Vertical rectus muscle palsy:** The three-step test is notoriously inaccurate for diagnosing vertical rectus muscle palsies [3, 8]. Jampolsky has shown that the IO cannot elevate the eye above the midline by itself, and the SO cannot depress the eye below the midline by itself [9]. The three-step test would rely on the IO muscle to raise the hypertropic eye with an IR palsy with tilt toward the unaffected eye. Conversely, the SO would be required to depress the hypotropic eye with a SR palsy with tilt toward the paretic eye.

**Table 8.1** Conditions in which the three-step test will be inaccurate

| |
|---|
| Restrictions |
| Multiple muscle involvement |
| Prior surgery |
| Dissociated vertical divergence |
| Skew |
| Myasthenia |
| Small hyper with horizontal strabismus |
| Vertical rectus palsy |
| Muscle overaction |
| Overcorrection of unilateral superior oblique palsy |

9. **Vertical muscle OA:** Because the three-step test assumes that one is dealing with an eye muscle palsy, it will be inaccurate in cases of muscle OA. Many clinical situations we incorrectly call muscle "OA" are in fact cases of muscle contracture [10]. The muscle is stiff (has increased elastic force) but does not have an increased contractile force. There are, however, situations in which muscle may have an increased contractile force, and the three-step test will be misleading if applied in those situations. Examples include the type of SR OA that results from Marcaine toxicity, or patients with muscle hypertrophy due to myositis [11, 12].
10. **Surgical overcorrection of unilateral SOP:** It is widely known that SOP is often bilateral, and if it is asymmetric, the deviation in the more affected eye may mask the palsy on the lesser affected side [13]. It is less widely known that a surgical overcorrection of a unilateral SOP may meet all the three-step test criteria for an SOP in the unoperated eye [14]. This mechanism has been elegantly fleshed out by Ellis and colleagues [15]. An important diagnostic finding to pay attention to is fundus torsion. In a true masked bilateral SOP that becomes "unmasked" after unilateral surgery, there should be fundus excyclotorsion in the eye that is hypertropic after the unilateral surgery. In a simple overcorrection, there should not be.

Table 8.1 lists the situations in which the three-step test will be misleading.

**Fig. 8.3** Overelevation in -dduction of the right eye. There are many etiologies that could cause this, many of which have nothing to do with the function of the right inferior oblique muscle

**Table 8.2** Causes of over- or underelevation or depression in adduction

| Condition | Helpful diagnostic hints |
|---|---|
| **Overelevation in adduction** | |
| Pseudo IO OA [16] | No elevation on side gaze, no SO UA, no fundus torsion |
| Anti-elevation syndrome [17] | History of prior IO anterior transposition. Lower eyelid bulging and flattening of eyelid margin on upgaze [18] |
| Dissociated vertical divergence | Hypotropia of adducted eye seen on alternate cover test. History of infantile strabismus |
| Duane syndrome | Limited abduction and lid fissure narrowing on adduction. Often upshot with adduction above midline and downshoot with adduction below midline |
| Pulley heterotopia or laxity | Orbital imaging shows anomalous pulley position [19, 20] |
| Craniofacial syndromes | Dysmorphia. Orbital imaging will diagnose |
| SO tendon incarceration syndrome [21] | Prior superior rectus or oblique surgery. Hypertropia with incyclotropia. Forced ductions abnormal |
| SO muscle palsy | Diagnosed by three-step test. Hypertropia with excyclotropia. History of closed head trauma with acquired. Facial asymmetry with congenital [22, 23] |
| IO OA/ contracture | Exaggerated force ductions [24] |
| Inferior restriction in contralateral eye | History of prior surgery or trauma. Forced ductions abnormal |
| **Underelevation in adduction** | |
| Inferior oblique muscle palsy | Three-step test should diagnose. Underelevation in adduction with A-pattern. Incyclotropia |

(continued)

**Table 8.2** (continued)

| Condition | Helpful diagnostic hints |
|---|---|
| Brown syndrome | Underelevation in adduction but with V-pattern. Positive forced ductions. Negligible Bielschowsky head tilt test |
| IO adherence syndrome [25] | Prior lateral rectus or inferior oblique muscle surgery. Hypotropia with excyclotropia |
| **Overdepression in adduction** | |
| Duane syndrome | Limited abduction and lid fissure narrowing on adduction. Often upshot with adduction above midline and downshoot with adduction below midline |
| Brown syndrome | Underelevation in adduction but with V-pattern. Positive forced ductions. Negligible Bielschowsky head tilt test |
| IO palsy | Three-step test should diagnose. Underelevation in adduction with A-pattern. Incyclotropia |
| SO "OA" | A-pattern, fundus incyclotropia. Horizontal strabismus. High association with spina bifida |
| **Underdepression in adduction** | |
| Craniofacial syndromes | Dysmorphia. Orbital imaging will diagnose |
| SO palsy | Diagnosed by three-step test. Hypertropia with excyclotropia. History of closed head trauma with acquired. Facial asymmetry with congenital [22, 23] |
| IO overaction/ contracture | Exaggerated force ductions [26] |

*IO* inferior oblique muscle, *OA* overaction, *SO* superior oblique muscle, *UA* underaction

## Basic Information

*Overelevation or Underelevation in Adduction*

Historically, we have tended to refer to overelevation in adduction as IO "OA" (Fig. 8.3). Conversely, underelevation in adduction is called inferior muscle "underaction" (UA). In fact, there are many causes of overelevation or overdepression in adduction, as listed in Table 8.2 [16–26], many of which have nothing to do with dysfunction of an oblique muscle.

## Overelevation in Adduction

1. **Pseudo IO OA:** This syndrome was discussed in detail in Chap. 7.

2. **Anti-elevation syndrome:** Anti-elevation syndrome (AES) occurs as a complication of IO anterior transposition [17, 19]. That procedure changes the IO from being an elevator to an anti-elevator, which can restrict elevation in abduction. If this effect is excessive, on elevation in side gaze the abducting eye is restricted due to the neural input to the IO, which now has a depressing vector. This results in overelevation of the adducting eye due to fixation duress, which looks like an "overacting" IO (Fig. 8.4). There is typically marked fundus extorsion. Treatment involves retroplacing the anteriorly transposed IO and converting it to a standard recession. Particularly if this occurs unilaterally, it is important to recognize that the problem is not in the eye that has overelevation in adduction, but rather it is in the eye that has limited elevation in abduction.
3. **Dissociated vertical divergence:** DVD can often have overelevation in adduction. This will occur when there is coexisting IO "OA," or sometimes just due to the nose acting as an occluder and dissociating the eyes on side gaze. In the former situation there would be a HYPO of the abducting eye on side gaze when it is occluded during alternate cover testing as well as fundus extorsion. IO anterior transposition is a good surgical option in this situation. In the latter situation (just DVD and no IO OA), there is no HYPO of the abducting eye on side gaze when it is occluded during alternate cover testing and typically no fundus extorsion. Also, although it appears that the HT (DVD) is bigger in adduction on version testing, quantification of the deviation with the prism under cover test often shows that it is the same across the horizontal gaze fields. It only appears to increase in adduction because the eyes are dissociated and the HT becomes manifest. In this situation, SR recessions are indicated.

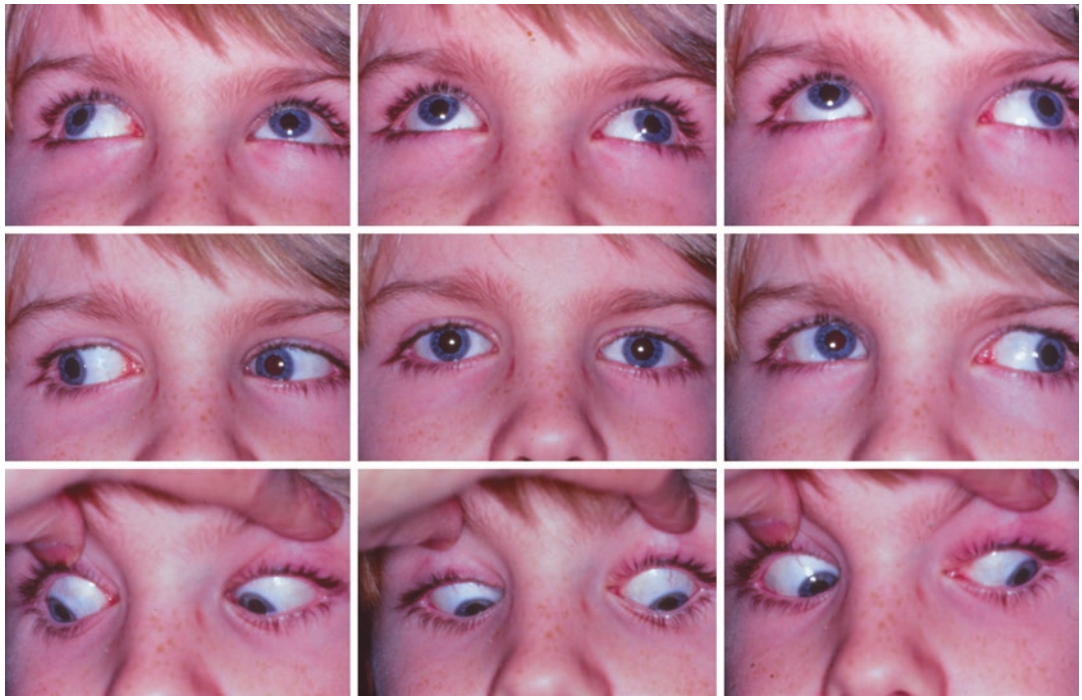


**Fig. 8.4** Anti-elevation syndrome after prior bilateral inferior oblique anterior transposition. What appears to be overelevation in adduction is actually a secondary deviation due to fixation duress. The abducting eye is restricted to elevation in abduction. Note the elevation and bulging of the lower eyelids on attempted elevation as well as the flattening of the lower eyelid margin

### Pearl

Overelevation in adduction in a patient with DVD could indicate associated IO OA. It also might be a manifestation of the DVD becoming manifest in adduction due to the nose acting as an occluder. Cover testing in side gaze will sort this out.

4. **Duane syndrome:** The upshoot on adduction in *Duane syndrome* can mimic IO OA but typically is not caused by the IO. It is caused by anomalous contraction of the lateral rectus muscle (LR) on adduction, which causes its belly to slide superiorly on the globe creating an elevating force vector. Often if adduction is attempted across the lower gaze fields there is a downshoot, as the LR slides down on the globe in this situation. It is important to understand this mechanism because weakening the IO will typically have no effect. Instead, the co-contracting LR must be generously weakened or disabled by suturing it to the periosteum.

### Pearl

Overelevation or overdepression in adduction in patients with Duane syndrome is usually not caused by oblique muscle dysfunction, and weakening the suspected oblique will have no effect.

5. **Pulley heterotopia or laxity:** If the LR pulley is inferiorly displaced (pulley heterotopia), or if it slips inferiorly on upgaze (pulley laxity) the affected eye may infraduct on abduction [20, 21]. If the patient fixes with that eye, it will appear to be overelevation of the contralateral eye on adduction. Orbital imaging is needed to diagnose pulley heterotopia. Dynamic orbital imaging comparing primary gaze to upgaze and side gaze is needed to

diagnose pulley laxity. See Chap. 21, Case 21.15 on page 322, for a representative example of these principles.

6. **Craniofacial syndromes:** Many of the more severe craniofacial syndromes (Crouzon, Apert, etc.) may have marked upshoots on adduction that are not related to the IOs. Most often there are anomalies of the rectus muscles, especially the SR; in some cases, the SR may be absent or severely attenuated. This can cause overelevation of the adducting eye due to fixation duress. Orbital imaging is important in these patients in order to formulate a sound surgical plan. See Chap. 17 for further discussion.
7. **Contralateral inferior restriction:** Any cause of inferior restriction that results in limited elevation can cause overelevation of the other eye due to fixation duress. Depending on the nature and location of the restriction, it may cause more limitation of elevation in abduction, resulting in more fixation duress to the contralateral eye in adduction, mimicking "IO OA."
8. **Superior oblique tendon incarceration syndrome:** After prior surgery on the SO tendon or on SR, the SO tendon can sometimes get scarred into the SR, causing a restriction [27]. This has been referred to as the SO tendon incarceration syndrome. It most commonly causes a HT of the affected eye, an incyclotropia, and limitation of depression. Although the pattern can vary, I see it most commonly present with more a greater HT in adduction than abduction. A history of prior surgery superiorly is the tip-off for considering this entity.
9. **Superior oblique muscle palsy:** In this situation the overelevation in adduction is in fact caused by the IO, which is opposed by a weakened antagonist SO. There is typically a head tilt toward the unaffected side, and the three-step test is positive for that diagnosis.
10. **Primary IO "OA"/contracture:** This usually accompanies primary horizontal strabismus, and is associated with a "V"-pattern and fundus extorsion. However, unlike the "IO OA" that occurs secondary to SOP, the Bielschowsky head tilt test is typically negative. Exaggerated forced ductions show IO contracture.

The Bielschowsky head tilt test can differentiate primary IO "OA" from IO "OA" that is secondary to SOP. Typically, there is no significant difference in the vertical alignment on head tilt right and left with primary IO "OA," but there is a difference if the "OA" is secondary to a paretic antagonist SO.

### Important Point

Overelevation in adduction does not mean the IO is "overacting" or even causative.

## Underelevation in Adduction

1. **IO palsy:** This will typically have SO "OA," fundus (and perhaps subjective) intorsion, and a positive three-step test for confirming the diagnosis. It is associated with an "A"-pattern.
2. **Brown syndrome:** On version there is limitation of elevation in adduction mimicking IO palsy. However, the three-step test is typically not positive. Most importantly, because the eye cannot adduct in elevation there is a V-pattern. This is important in distinguishing it from IO palsy, which has an A-pattern. Forced ductions are diagnostic.
3. **Superior oblique "OA":** With SO "OA" there is an A-pattern and bilateral fundus intorsion. It is typically associated with horizontal strabismus. There is a high incidence of this pattern in patients with spina bifida and hydrocephalus.

### Basic Information

*Overdepression or Underdepression in Adduction*

Historically, we have tended to refer to overdepression in adduction as SO "OA." Conversely, underdepression in adduction is called superior muscle "UA." Table 8.2 is also a partial list of causes of over- and underdepression in adduction.

## Overdepression in Adduction

1. **Duane syndrome:** A downshoot on adduction in Duane syndrome can mimic SO OA but typically is not caused by the SO. It is caused by co-contraction of the LR and medial rectus muscles on adduction, which causes the LR belly to slide inferiorly on the globe, creating a depressing force vector. Often if adduction is attempted across the upper gaze fields there is an upshoot, as the LR slides up on the globe in this situation. It is important to understand this mechanism because weakening the SO will typically have no effect. Instead, the co-contracting LR must be generously weakened or disabled by suturing it to the periosteum.
2. **Brown syndrome:** See description above in the section on underelevation in adduction.
3. **IO palsy:** See description above in the section on underelevation in adduction.
4. **Superior oblique muscle OA:** See description above in the section on underelevation in adduction.

## Underdepression in Adduction

1. **Craniofacial syndromes:** See description above in the section on overelevation in adduction.
2. **Superior oblique muscle palsy:** See description above in the section on overelevation in adduction.
3. **IO "OA"/contracture:** See description above in the section on overelevation in adduction.

### Advanced Information

*Measurement Pointers for Vertical Strabismus*

The gold standard for measuring the angle of strabismus is the prism and alternate cover test. There are some circumstances in which this is not sufficient. They include the quantifying of DVD, sorting out fixation duress from a manifest HYPO and contralateral DVD, and measuring torsion.

1. **Dissociated vertical divergence:** Classically, DVD is quantified by the prism under cover test (PUCT). This involves first estimating the full deviation (manifest plus latent) either by light reflex or just guessing at the magnitude of the downward movement of the eye as an occluder is moved from it to the fellow eye. Let's say you correctly estimate there to be a 20 Δ right DVD. You then put a 20 Δ prism base-down in front of the right eye and hold the occluder in front of the prism (hence the name *prism under cover*). Move the occluder to the left eye. Because you accurately neutralized the deviation, there will be no movement of the right eye. Had you underestimated the deviation and used a 15 Δ prism, there would be a residual downward movement of the right eye of 5 Δ when you shift the occluder. Had you overestimated and used a 25 Δ prism, there would be a 5 Δ upward movement of the right eye as you shift the occluder (see Fig. 8.5). Although the PUCT is the gold standard for quantifying DVD, you will not always get a stable endpoint with no movement as the occluder is shifted. In one prospective study of 141 cooperative patients with DVD, only 33% had a stable endpoint with the PUCT (unpublished data, collected as part of a published study [5]). The remaining 67% either had redress, which requires the examiner to estimate the size of the redress movement, or were variable. If redress or variability is present, several maneuvers may improve the accuracy of your measurement.

Only 33% of patients with DVD had a stable endpoint with the PUCT.

### Pearl

Redress and variability with prism and cover testing, including the PUCT, can be minimized by having the patient slowly read optotypes on the distance chart. If that does not eliminate redress, repeat the cover test while allowing the patient to have several

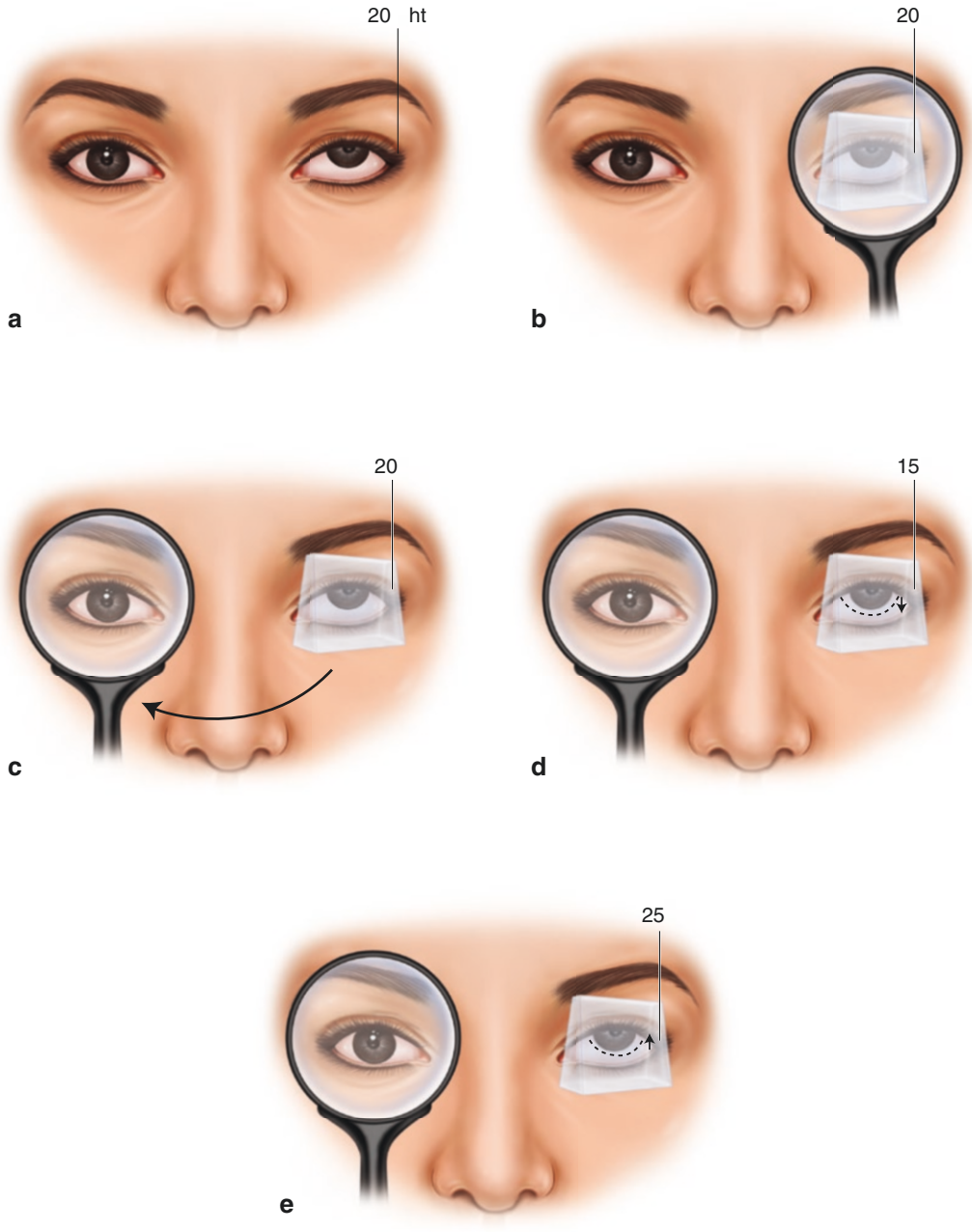



**Fig. 8.5** Prism under cover test for right dissociated vertical divergence. (**a**) Estimate the deviation in the right eye, in this case 20 Δ; (**b**) put a 20 Δ prism base-down OD and hold the occluder in front of the prism, and (**c**) switch the occluder to the left eye. There is no movement of the right eye. (**d**) If you incorrectly estimated the deviation to be 15 Δ, there would be a downward movement of the right eye of 5 Δ as you move the occluder to the left eye**.** (**e**) If you incorrectly estimated the deviation to be 25 Δ, there would be an upward movement of the right eye of 5 Δ as you move the occluder to the left eye

seconds of binocular viewing before switching the occluder from one eye to the other.

2. **Small hypo with contralateral large HT:** Consider a patient who fixes right eye (*oculus dexter*, OD) and manifests a 5 Δ left HYPO. But with left eye (*oculus sinister*, OS) fixation there is a large RHT of 20 Δ. There is a history of prior strabismus surgery. This could be a case of DVD in the right eye and a manifest L HYPO. Alternatively, it might be a L HYPO in an eye with somewhat limited elevation, either due to SR weakness or restriction inferiorly OS; the large RHT is a secondary deviation. Differentiating these two possibilities is important. In the former scenario, surgery should address a right DVD. In the latter, the large RHT will be corrected if the L HYPO is corrected and upgaze improved. In order to tell what is going on, you need to neutralize the L HYPO with prisms and see if the RHT persists. To do this, you put 5 Δ base-up OS and do an alternate prism and cover test. If you exactly neutralize the L HYPO with prism, or perhaps overcorrect it a miniscule amount, there will be no RHT if you were dealing with fixation duress. But if the RHT was due to DVD, it will still be present.
3. **Torsion:** Objective torsion should be assessed with indirect fundoscopy, and subjective torsion with the double-Maddox rods. Neither test replaces the other. Both are necessary and they provide complementary information. See Chap. 2 for a detailed discussion.

## Advanced Information

*Some "Soft Signs" Can Steer You in the Right Direction for Making a Diagnosis*

1. If the incomitance is greater vertically, e.g., bigger change in the HT from downgaze to upgaze than from right gaze to left gaze, consider a restriction or a vertical rectus muscle problem. If the incomitance is greater horizontally, e.g., right gaze to left gaze has a bigger change than downgaze to upgaze, consider an oblique muscle problem.
2. If the forced head tilt difference is large, e.g., large change in the hypertropia between tilt left and right, consider an oblique muscle problem or DVD. If the forced head tilt difference is small, consider a rectus muscle problem or another cause of vertical strabismus.

## Pearl

Whenever a patient has unequal visual acuity, consider the possibility that they may be fixing with the involved or paretic eye. I have seen patients with TED, SOP, monocular elevation deficiency, Brown syndrome, and Duane syndrome who fixated with the affected eye. In some patients this led to incorrect prior diagnosis and surgery.

## Basic Information

*Summary of Important Diagnostic Steps*

1. Determine if history can help with the diagnosis. If there was trauma, consider SOP or orbital fracture. If the onset was in infancy, consider DVD as a possibility. If there was a prior retrobulbar or peribulbar injection, evaluate for signs of myotoxicity. The presence of associated medical conditions might help pinpoint the diagnosis such as TED.
2. Observe if the patient has DVD (cover test shows HT that does not follow Hering's law vertically).
3. Consider that the patient might be fixing with the involved eye, especially if she has unequal vision.
4. Evaluate the incomitance pattern to see if it helps with the diagnosis.

5. If the three-step test is positive for a specific diagnosis, always consider the multiple causes of errors in the three-step test.

## Treatment

### Basic Information

*General Principles*

Below are four general principles that can guide your treatment decisions.

1. The primary position is most important, and the near downgaze reading position is second most important. Do not sacrifice good alignment in these two gaze fields in order to achieve alignment in eccentric gazes.
2. The size of the primary position deviation determines the surgical "dose," both with respect to number of muscles and millimeters of surgery. The pattern determines which muscles should be operated.
3. In patients with diplopia awareness, take torsion into account when formulating a surgical plan.
4. Consider weakening the yoke of a paretic muscle, unless the limitation of rotation is −3 or −4.

See Chap. 21, Case 21.30 on page 333, for a representative example of these principles.

### Basic Information

*Surgical Management of Unilateral SOP*

In keeping with the principles above, the primary position deviation determines the number of muscles I operate upon—above all I do not want to overcorrect the patient in the primary position. In general, 15 Δ is my threshold above which I will operate on a second muscle. In some cases, if the patient is cooperative enough for adjustable sutures, I may lower my threshold for operating on a second muscle at 10–12 Δ, depending on the pattern. Once I determine the number of muscles to operate, I let the pattern guide me as to muscle choice.

### Basic Information

*Surgical Choices Based on Pattern if Determined to Be a One-Muscle Operation (Generally <15 Δ in Primary)*

1. **Biggest deviation in the field of the IO**

| | | |
|---|---|---|
| | | +++ |
| | < 15 ΔRHT | ++ |
| | | + |

*Surgical choice*: weaken ipsilateral IO

2. **Biggest deviation across adducted field (in the field of IO and SO)**

| | | |
|---|---|---|
| | | +++ |
| | < 15 ΔRHT | ++ |
| | | + |

*Surgical choice*: weaken ipsilateral IO**.**

### Important Point

In the aforementioned scenario the SO will often show an increase in function after the "overacting"/contractured IO is weakened.

3. **Biggest deviation in the field of the SO:**

| | | |
|---|---|---|
| | | + |
| | < 15 ΔRHT | ++ |
| | | +++ |

*Surgical choice*: Depends on some variables:

(a) If exaggerated forced ductions show only moderate IO tightness, tucking the ipsilateral SO is a good option.

(b) If you do not do much SO surgery and are not comfortable performing an SO tuck, a good alternative is to recess the contralateral IR (on an adjustable suture if possible) for the HT, and nasally transpose the ipsilateral IR to correct the excyclotropia [28]. In most cases tucking the SO can be avoided, if you so wish. (See Chap. 21, Case 21.32 on page 335, for a representative example of management of iatrogenic Brown syndrome after a SO tuck.)

(c) An advantage in doing an SO tuck over the bilateral IR procedure is that it involves operating on only one eye, is faster, and directly addresses the problem of an underacting SO. An advantage in the bilateral IR procedure is that it is more forgiving than an SO tuck, can more easily be done with adjustable sutures, and does not require as much experience. Both are acceptable, and the choice is a matter of weighing trade-offs. The biggest factor in making the decision should be the surgeon's experience and comfort with SO surgery.

(d) If exaggerated forced ductions show a markedly tight ipsilateral IO, this might be the *inverted Brown syndrome* [29]. If so, weakening the IO will result in improved SO function once the restriction is released, and will give good correction in downgaze.

See Chap. 21, Case 21.33 on page 335, for a representative example of the aforementioned principles.

### Question

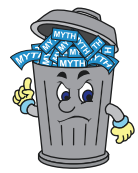
How does SO tendon laxity influence your decision to tuck the SO versus perform some other procedure?


### Reply

Tendon laxity influences the size of the tuck (I do larger tucks on lax tendons) but plays no role in the decision whether or not to tuck. I realize this flies in the face of an ever-growing dogma that dictates if the SO tendon is lax, a tuck is indicated, and if it is not lax, a tuck is contraindicated. The rationale for this dogma is predicated on the belief that the primary abnormality is a redundant and lax tendon. The reasoning continues with the hypothesis that if the redundancy is eliminated with a tuck, the muscle and tendon will function normally. Although there are good data that congenital SO palsies often have lax tendons, there are no data confirming this dogma with respect to treatment. In fact, a prospective study of patients with SOP who underwent orbital MRI showed that lax tendons were almost always associated with attenuated SOs [30]. So it appears that the lax tendon is secondary to muscle attenuation and laxity, and tucking a lax tendon would, by necessity, be putting the orbital portion of the SO on stretch. I think mandating that a lax tendon should be tucked is like saying a sixth nerve paralysis with a flaccid elongated LR should undergo a recess/resect procedure, and if the muscle is not flaccid, a resection is contraindicated. In fact, we approach sixth nerve palsy in the opposite manner, reserving resection for those cases with some LR function, and doing a transposition for those that have none.

> Saying a lax tendon must be tucked is as sensible as saying a flaccid dead LR must be resected.

> Creativity is dangerous. We cannot open ourselves to new insight.
> without endangering the security of our prior assumptions.
> —Robert Grudin, *The Grace of Great Things*




### Myth

A lax SO tendon must be tucked and a tendon that is not lax must not be tucked.

### Fact

There are no data to support this dogma, and intuition suggests otherwise. Tendon laxity should

influence the size of the tuck, but not the decision whether or not to tuck the SO.

> My karma ran over your dogma.
> —Source unknown (bumper sticker)

4. **Biggest deviation across the inferior gaze fields**

| | | |
|---|---|---|
| | | + |
| | < 15 ΔRHT | ++ |
| +++ | +++ | +++ |

*Surgical choice*: Pay careful attention to forced ductions for ipsilateral SR tightness. I have only seen this pattern when combined with a small primary position deviation and in the context of ipsilateral SR OA/contracture. If SR contracture is present, an ipsilateral SR recession for the HT is appropriate; however this will worsen the torsion. I would combine that with an ipsilateral Harada-Ito procedure. If you are not comfortable with SO surgery, you can address the torsion by nasally transposing the ipsilateral IR. If there is no evidence of SR contracture, I would treat it the same as in scenario number 3, above, where the deviation is greatest in the SO field.

5. **Biggest deviation across the inferior gaze fields and adducted fields**

| | | |
|---|---|---|
| | | + |
| | < 15 ΔRHT | ++ |
| +++ | +++ | +++ |

*Surgical choice*: This is also a very unusual pattern in the presence of a small deviation in primary (I am not sure I have seen it). I would recommend the same treatment options in scenario number 3, biggest deviation in the field of the SO.

## Basic Information

*Surgical Choices Based on Pattern if You Determined a Two-Muscle Operation Is Indicated (Generally, >15 Δ in Primary)*

For almost all patients in whom I am doing a two-muscle procedure, e.g., primary position deviation is bigger than 15 Δ, I recess the ipsilateral IO and the contralateral IR. Almost all patients with a large primary deviation have enough of a deviation in the IO field, as well as in downgaze, that this approach seems tailor-made. Although one can describe hypothetical patients who do not adhere to this, they are quite uncommon. However, if faced with one, I would individualize my muscle choice based on the general principles outlined at the beginning of this chapter. I do not like to simultaneously tuck the SO and weaken the IO in the same eye in the same sitting. In my hands, doing so invariably produces a post-op Brown syndrome that does not resolve as well as after an isolated SO tuck. There is one exception. Jampolsky pointed out that an acute SO paralysis will not have more than 2–3 Δ of HT in the ipsilateral abducted field, and if there is 10 Δ or more, there is probably some degree of SR OA/contracture [9, 31]. If that is the case, I will do a small ipsilateral SR weakening procedure combined with an ipsilateral IO weakening procedure.

## Important Point

When simultaneously weakening the SR and IO in the same eye in the same sitting, do a very small SR recession. Weakening both elevators is very powerful and can easily cripple upgaze. This is one of the few situations in which I do 2 or 2.5 mm recessions. Alternatively, a disinsertion of the nasal three-fourths of the SR will be equivalent to a small SR recession. This also has the advantage of effectively transposing the SR a small amount temporally, which will help reduce the excyclo.

## Important Point

In many patients with SR OA/contracture, forced ductions are normal. The muscle behaves more like it is "overacting" than contractured. The proposed mechanism for this is complex but has been described previously [32]. There are multiple mechanisms that can lead to a muscle

being strengthened, but contracture requires either chronic shortening with sarcomere loss or in some cases myotoxicity. SR OA/contracture most commonly occurs secondary to ipsilateral SOP. In this setting, chronic shortening is not present and, hence, the SR is not contractured.

### Pearl

If there is more than 10 Δ of HT in the ipsilateral abducted field, there is probably an element of SR OA/contracture. Consider adding a small ipsilateral SR weakening procedure to the surgical plan.

### Pearl

If you have what looks like a classic SOP, but there is no objective or subjective torsion, consider there being an element of ipsilateral SR OA/contracture. The excyclo that would be caused by the weak SO is counteracted by the intorsional force of the overacting SR. Usually in this setting there is a large forced head tilt difference.

### Question

I have a patient with a fourth cranial nerve palsy, a large deviation in primary, and marked IO OA. Would you recommend an IO anterior transposition?


### Reply

I personally never like to perform IO anterior transposition in patients with bifoveal fusion and diplopia awareness. If you consider how this procedure alters the force vector of the IO, it is not surprising that it has a complex effect on motility and comitance. Although I am aware some people advocate this for severe SOP, I have not seen any data that show the effect of this approach with respect to the size of the single binocular field. Furthermore, IO anterior transposition does not predictably decrease extorsion [18], and, when done unilaterally, often causes cosmetically noticeable lid fissure asymmetry [33] and can cause a restrictive HYPO in upgaze. Furthermore, the AES can result, and the incidence of its occurrence increases with time after surgery [17]. The fact that case series with short follow-up do not report this complication does not mean it will not occur; it will. For any size and pattern of unilateral SOP there are more predictable and less complication-prone surgical options.

### Basic Information

*Diagnosis of Bilateral SOP*

When a patient shows bilateral SO UA, bilateral IO OA, and alternating HTs (RHT on L gaze and LHT on R gaze), the diagnosis of bilateral involvement is obvious. But when there is asymmetric involvement, it can be easy to overlook the diagnosis in the lesser affected eye. There are some useful diagnostic signs that can alert you to a bilateral problem [13].

They are:

1. Diplopia starting after head trauma.
2. A subjective excyclotropia of 10° or more (total between the two eyes): Keep in mind that subjectively all the torsion may be localized to the nondominant eye.
3. Bilateral fundus excyclotorsion.
4. A relatively small Bielschowsky forced head tilt difference [8, 13].
5. A V-pattern with 10 Δ or more of eso-shift between the primary position and downgaze.
6. Chin down to fuse rather than a head tilt.
7. In spite of a large subjective excyclotropia (10° or more), a relatively small degree of IO OA.
8. A very small HT in primary with a large HT in adduction.
9. A reversal of the HT in the field of the IO or SO of lesser affected eye, or on head tilt toward the lesser affected eye.

## Basic Information

*Surgical Management of Bilateral SOP*

Although there can be substantial heterogeneity to the presentation of bilateral SOP, there are some consistent principles that guide my treatment choices. First, you need to determine if you will do a symmetric or an asymmetric procedure. I base this on whether or not there is a HT in primary gaze that I need to collapse. If so, I do an asymmetric procedure. If not, I do a symmetric operation, regardless of the presence of asymmetric oblique dysfunction on versions or asymmetric HTs in side gaze! This is based on the rationale that an asymmetric procedure in this setting might induce a HT in primary gaze, which I want to avoid.

## Important Point

Do a symmetric procedure if there is no HT in primary, regardless of the presence of asymmetric oblique dysfunction.

## Basic Information

*Surgical Management of Symmetric Bilateral SOP*

If there is no, or only a trivial HT in primary, I do symmetric IO weakening or SO tightening according to the following principles:

**If there is substantially more IO OA than SO UA and less than 15 Δ of esotropia in downgaze:**

1. I will symmetrically weaken the IOs. My preferred operation is a recession without anterior transposition. This is a relatively rare clinical presentation.

**If there is substantially more IO OA than SO UA and 15 Δ or more of esotropia in downgaze:**

2. I will symmetrically weaken the IOs and simultaneously recess both IRs 5–6 mm without horizontal transposition [34].

**If there is more SO UA than IO OA, and/or 15 Δ or more of esotropia in downgaze, I will do one of several things depending on the following:**

3. If there are significant HTs in right and left gaze, I will do bilateral SO tucks. Although I typically determine the size of a tuck for unilateral cases based on "feel" and forced ductions after the tuck is temporarily sutured, for bilateral surgery I do the first side by "feel," measuring the size of the tuck I have performed, and do the exact same amount on the other side.
4. If there are small HTs in right and left gaze I will do bilateral Harada-Ito procedures. If the esotropia in downgaze is more than 20 Δ, I will add simultaneous IR recessions [34]. (Note: I do not add the IR recessions if I am doing SO tucks, as they will correct more downgaze esotropia than Harada-Ito procedures.)

## Basic Information

*Surgical Management of Asymmetric Bilateral SOP*

If there is a HT in primary that is more than trivial, I will do asymmetric IO weakening or SO tightening, or address the primary position vertical with vertical rectus surgery, according to the following principles:

**If there is substantially more IO OA than SO UA, up to 10 Δ of HT in the primary position, and less than 15 Δ of esotropia in downgaze:**

1. I will recess the IOs asymmetrically. If the OA in the more affected eye is +1 or +2, I recess the IO 10 mm, and if it is +3 or +4, I recess 12 mm. In the less affected eye I recess the IO 6 mm.

**If there is substantially more IO OA than SO UA, 10 Δ or more of HT in the primary**

**position, and less than 15 Δ of esotropia in downgaze:**

2. I do symmetric IO recessions without anterior transpositions, and either recess the SR in the higher eye or the IR in the lower eye by an amount to correct the primary position HT.

**If there is substantially more IO OA than SO UA, up to 10 Δ of HT in the primary position, and 15 Δ or more in downgaze:**

3. I do symmetric IO recessions and bilateral, asymmetric IR recessions. I do a 6 mm recession of the IR in the lower eye, and use the size of the HT in the primary position to determine how much less I recess the IR in the higher eye.

**If there is more SO UA than IO OA, up to 10 Δ of HT in primary, and/or 15 Δ or more of esotropia in downgaze, I do one of several things depending on the following:**

4. If there are significant HTs in right and left gazes, I do bilateral SO tucks. Although I typically determine the size of a tuck for unilateral cases based on "feel" and forced ductions after the tuck is temporarily sutured, for bilateral surgery I do the more affected (HT) side by "feel," measure the size of the tuck I have performed, and do a smaller tuck on the other side. I determine the size on the lesser affected eye by measurement rather than feel with the difference in the tuck sizes based on the size of the primary position HT.
5. If there are small HTs in right and left gazes, I do bilateral asymmetric Harada-Ito procedures. Asymmetry can be achieved by transposing a larger portion of the tendon in the more affected eye, e.g., one-fourth in one eye and one-half in the other, and one-half in one and three-fourths in the other; or by resecting some of the anterior tendon in the more affected eye; or by transposing the tendon further in the more affected eye. If the esotropia in downgaze is 20 Δ or more I will add simultaneous IR recessions [34]. Alternatively, you can do symmetric Harada-Ito procedures bilaterally and asymmetric IR recessions. This is a good option if you are not comfortable attempting asymmetric Harada-Ito procedures. Note that the Harada-Ito procedure does not correct as much esotropia in downgaze as SO tucks, which is why I do not add the IR recessions if I am doing SO tucks.

**If there is more SO UA than IO OA, 10 Δ or more of HT in primary, and/or 15 Δ or more of esotropia in downgaze:**

6. Either do symmetric SO tucks or symmetric Harada-Ito procedures based on the principles above for cases in which there is no HT in primary, but add a vertical rectus recession in one eye to address the primary position hypertropia.

See Chap. 21, Case 21.18 on page 324, for a representative example of these principles.

Tables 8.3 and 8.4 summarize my treatment recommendations for symmetric and asymmetric bilateral SOP.

**Table 8.3** Treatment of bilateral symmetric superior oblique palsy

| Versions | Esotropia in downgaze | Treatment |
|---|---|---|
| IO OA greater than SO UA | <15 Δ | Symmetric IO weakening without anterior transposition |
| IO OA greater than SO UA | ≥15 Δ | Bilateral symmetric IO weakening without anterior transposition plus bilateral IR recession 5–6 mm |
| More SO UA than IO OA and significant side gaze HTs | ≥15 Δ | Bilateral symmetric SO tucks |
| More SO UA than IO OA and small side gaze HTs | ≥15 ΔΔ | Bilateral symmetric Harada-Ito procedures |
| More SO UA than IO OA and small side gaze HTs | >20 Δ | Bilateral symmetric Harada-Ito procedures plus bilateral IR recession 5–6 mm |

*IO* inferior oblique muscle, *OA* overaction, *SO* superior oblique muscle, *UA* underaction, *IR* inferior rectus muscle, *HTs* hypertropias, *Δ* prism diopters

**Table 8.4** Treatment of bilateral asymmetric superior oblique palsy

| Versions | Hypertropia in primary (Δ) | Esotropia in downgaze (Δ) | Treatment |
|---|---|---|---|
| IO OA greater than SO UA | <10 | <15 | Recess the IO in the more affected eye 10 mm for +1 or +2 OA, or 12 mm for +3 or +4. In the lesser affected eye recess the IO 6 mm |
| IO OA greater than SO UA | ≥10 | <15 | Bilateral symmetric IO weakening without anterior transposition plus IR recess in lower eye *or* SR recess in higher eye |
| IO OA greater than SO UA | <10 | ≥15 | Bilateral symmetric IO weakening without anterior transposition plus bilateral IR recession of 6 mm in the lower eye and less in the higher eye according to the primary position HT |
| More SO UA than IO OA, significant side gaze HTs | <10 | ≥15 | Bilateral asymmetric SO tucks |
| More SO UA than IO OA and small side gaze HTs | <10 | ≥15 | Bilateral asymmetric Harada-Ito procedures |
| More SO UA than IO OA and small side gaze HTs | <10 | ≥20 | Bilateral asymmetric Harada-Ito procedures plus bilateral symmetric IR recessions. Alternatively, do bilaterally symmetric Harada-Ito procedures and bilateral asymmetric IR recessions |
| More SO UA than IO OA and small side gaze HTs | ≥10 | ≥15 | Bilateral symmetric Harada-Ito procedures plus one vertical rectus recession for the HT |
| More SO UA than IO OA and significant side gaze HTs | ≥10 | ≥15 | Bilateral symmetric SO tucks plus one vertical rectus recession for the HT |

*IO* inferior oblique muscle, *OA* overaction, *SO* superior oblique muscle, *UA* underaction, *Δ* prism diopters, *IR* inferior rectus muscle, *SR* superior rectus muscle, *HT* hypertropia

## Question

This 38-year-old man has traumatic bilateral SO palsies. He has 8 Δ ET in primary, which increases to 18 Δ in downgaze and is ortho in upgaze. There is no HT in any midline position. In primary he has 15° of excyclotropia, +2 IO OA, and −3 SO UA. He prefers a chin-down head posture with which he fuses. Diplopia fields show him to have single binocular vision from 5° below primary to about 20° above primary. I am considering (1) bilateral IO recession (safe in my hands), (2) bilateral SO tucks (I have concern about not getting it exactly symmetric), or (3) bilateral IR recessions. What do you recommend?


## Reply

It is really unusual for a patient to be able to fuse 15° of cyclotropia, so I wonder if he can really fuse in primary as you say. If this is based on the diplopia field test, keep in mind that if done on the Goldmann perimeter with a light for fixation, a patient may not report diplopia if torsion is present, whereas they would be diplopic viewing a letter. Two round lights, superimposed, will appear as one light even if one is rotated. I would be surprised if he could fuse a letter in primary if he indeed has 15° of torsion. Specifically: I personally have never done the bilateral IR recession as a sole primary procedure for bilateral SOP, but have reserved it for those

patients needing an enhancement to address downgaze issues after prior surgery, or in addition to other surgery at the time of the first operations. Regardless, I do not think that procedure will correct 15° of excyclotropia. IO recession alone is not a good option, as that will mainly help upgaze, and he needs help in downgaze. Bilateral SO tucks are an option but there are good alternatives.

If he were my patient, given the information provided, I would do bilateral Harada-Ito procedures, as I think nothing less than SO surgery will help this much torsion. I would combine that with simultaneous IR recession of about 5–6 mm. If you wanted to do it in two stages, I would start with the Harada-Ito procedures and then recess the IRs.

## Question

I care for a 58-year-old man with a history of vertical diplopia of several years' duration. His motility looks somewhat like an RSO palsy measuring 20 Δ in primary and increasing a bit in left gaze and decreasing a bit on right gaze. On tilt right it is 25 Δ and on tilt left it is 12 Δ. In downgaze it decreases to 10 Δ. What concerns me is that he has no objective or subjective torsion. I know that recessing the LIR will address the horizontal incomitance, but I am concerned this will overcorrect him in downgaze. I am thinking of a RIO recession and RSR adjustable recession, but am nervous doing the RIO if he does not have an RSO palsy.


## Reply

I think what you are seeing is the SR OA/contracture syndrome that can accompany SOP. Sometimes it takes the form of SR contracture, but sometimes appears to have a true SR OA, with the HT greatest in upgaze in the field of the SR. In either scenario, the overacting SR can have an intorting force that can negate the excyclotropia that should come from the SOP so there is no net torsion. Recessing the RIO and RSR is what I would recommend here. However, the RSR recession should be very small, as upgaze will easily be crippled with an average-sized recession.

## Question

Are you ever concerned if the HT in primary is very small in a patient with fourth nerve palsy? In one of my patients this issue is extreme. A 68-year-old man with a presumed right fourth cranial nerve palsy is now a year post-onset. He has shown some improvement and has no HT in primary. On gaze left he measures 18 Δ RHT. It is worse up and left as there is +2 RIO OA. He is very symptomatic with diplopia starting only 5° into left gaze, and he would like something done. I did a patch test for 2 h, and I was able to bring out only an RHT of 2 Δ in primary. I was hoping to bring out at least 5 Δ to feel good about doing an RIO recession. Do you think I am OK doing an 8–10 mm recession of the RIO?


## Reply

I think it is good to worry about this, as there is a high chance of overcorrecting with an IO recession. I also would have done the patch test for the reasons you give. This usually brings out enough HT that you would be safe treating with an IO rec. But that did not happen in this man. Whenever I see a really small or no HT in primary in a patient that appears to have an SOP, I look extra hard for bilateral masked SOP. Look for torsion in the other eye, the presence

of a V-pattern, and a reversal of the HT in the field of the contralateral IO or SO. If it is not a bilateral masked SOP, you need to do something to prevent a post-op R HYPO in primary. I have treated a few patients like this in whom I did the IO recession and put the ipsilateral IR on an adjustable suture with a small recession. The few patients I treated this way ended up needing about 2 mm of recession (after any adjustment), so I assume I would have overcorrected them in primary had I not done the ipsilateral IR recession.

### Question

A 25-year-old man was hit by a bus and had closed head injury resulting in bilateral fourth cranial nerve palsies. Over 7 months, it appeared the right fourth resolved but left fourth persisted. However, no torsion is reported nor can it be elicited on the synoptophore. There is just the vertical deviation. He does not assume a head tilt. His main complaint is vertical diplopia on downgaze and down-and-right gaze. His measurements are as follows:

| | | |
|---|---|---|
| === | 2 Δ LHT | === |
| 12 Δ LHT | 2 Δ LHT | 2 Δ LHT |
| 15 Δ LHT | 10 Δ LHT | 8 Δ LHT |

This is the first time I'd seen a bilateral traumatic fourth cranial nerve palsy settle to a unilateral palsy, and also the first time I'd seen a traumatic fourth with no torsion. Have you seen this and how would you manage him? Would Alan Scott's "adjustable faden on the right IR" be the way to go? Also, is it okay to tuck an SO with an acquired palsy when there is no excyclotorsion to start with? Is there any risk of inducing incyclotorsion as a new problem?


### Reply

I definitely have seen bilateral acquired SOPs settle down to a unilateral palsy. But I am not 100% sure your patient has this condition. I would want to know at least qualitatively if the HT reverses in gaze up and right, and also on head tilt to the right. I would want those assessments before making a diagnosis. Can he fuse with his head tilted? With only 2 Δ of LHT in primary, if this was just a left fourth cranial nerve palsy the HT should go away on tilt R and he would do that to fuse. If not, I suspect that there is still some bilateral involvement. You say there is no torsion on the synoptophore. But I would also check with the double-Maddox rods and look for fundus torsion; that is also important. If indeed there is no torsion subjectively or objectively, you would likely cause torsional problems by tucking the SO. If the answers to all the above confirm a unilateral SOP, doing the RIR adjustable faden (recess and resect on the same muscle) as described by Scott is a good plan (see Chap. 10 for detailed discussion).

## Dissociated Vertical Divergence

### Basic Information

*DVD—Overview*

DVD gets its name from the observation that the movement of the two eyes appears to be dissociated from one another and do not follow Hering's law. If one eye is manifesting a DVD and is higher than the fixing eye, and the fixing eye is then covered, the hypertropic eye will move down to pick up fixation. Hering's law would mandate that the now-covered eye would infraduct an equal amount. With DVD, the covered eye does not infraduct and, in fact, may supraduct if the DVD is bilateral (which it usually is). Hence the term "dissociated." However, some elegant work by Guyton suggests that DVD does follow Hering's law if one takes the torsional component into account [35, 36].

### Advanced Information

*How DVD Actually Does Follow Hering's Law*

Using some sophisticated eye movement tracings that recorded vertical, horizontal, and torsional eye position, Guyton elucidated a mechanism for the vertical movement we call DVD [35, 36]. He observed that the initial event is that the fixing eye makes an intorsional movement to damp nystagmus, which is invariably present in patients with DVD, however subtle. Following Hering's law the non-fixing eye extorts. The intorsion in the fixing eye is mediated by the SO, which also causes the eye to depress slightly, and the extorsion in the non-fixing eye is mediated by the IO, which also causes that eye to supraduct slightly. Then the fixing eye must make a small supraduction to pick up fixation from its infraducted position. Again following Hering's law, the non-fixing eye supraducts an equal amount. The two small supraduction movements of the non-fixing eye are additive and are what we see as a manifest DVD. A very similar theory was presented by Brodsky, who approached the issue from an evolutionary perspective [37]. Although these theories are the best put forward to explain DVD, there are two clinical observations that they do not explain.

DVD does follow Hering's law if you take torsion into account.

1. Sometimes manifest DVD can be quite large—25 Δ or more. I do not see how this large deviation can be explained by these theories. The initial hypotropic shift of the fixing eye should only be a few prism diopters, and the same would be the case for the initial hypertropic shift of the non-fixing eye. Adding these values should still only account for a small vertical separation, and it is the sum of these two movements that should equal the size of the DVD. I do not see how it can account for the large deviations we sometimes see in patients with DVD.
2. DVD is typically intermittently manifest. These theories suggest that it is only manifest when the patient is damping nystagmus by intorting the fixing eye. It is observed that one can do very large SR recessions in patients with DVD yet almost never get an overcorrection [38]. Consider a hypothetical patient with an intermittently manifest right DVD of 25 Δ, and in whom you do a 7 mm RSR recession. After surgery, during those moments when the patient is not damping nystagmus by intorting the left eye, one should expect a large R HYPO after such a large SR recession. Yet this does not occur. This observation is not explained by the aforementioned theories.

I do not offer solutions. I only point out the problem.
—(Charles Schulz, *Peanuts*)
(Lucy talking to Charlie Brown)

## Basic Information

*Surgical Treatment of DVD*

If a patient has amblyopia in one eye, he or she will always fixate with the better eye. Even if he or she has bilateral DVD, it will only be manifest in the amblyopic eye; it will be latent in the better eye. For patients with approximately equal vision, the most common presentation of DVD is for it to be intermittently manifest in the nondominant eye, and latent in the dominant eye. Typically, the magnitude of the manifest DVD is greater than that of the latent DVD. Less commonly, a patient may freely alternate and either eye will intermittently have a manifest DVD. I dichotomize my treatment of DVD based on whether the patient has the potential to switch fixation after surgery, which typically means the acuity is better than 20/40 in the nondominant eye.

## Basic Information

*Unilateral Surgical Treatment of DVD in Patients with Unequal Vision*

In almost all circumstances I treat these patients with unilateral SR recessions. My surgical formula for unilateral SR recession for DVD is presented in Table 8.5. I prefer to do the surgery

**Table 8.5** Unilateral SR recession for DVD

| Size of deviation (Δ) | Size of unilateral SR recession (mm) |
|---|---|
| <10 | 5 |
| 10–14 | 6 |
| 15–19 | 7 |
| 20–24 | 8 |
| 25 and up | 9 |

*SR* superior rectus muscle, *DVD* dissociated vertical divergence, *Δ* prism diopters

with a fixed scleral suture rather than a suspension technique (also known as hang-back). Although placing fixed scleral sutures is technically harder than a suspension technique, especially in small children needing large recessions, I feel they are more precise, and the muscle is less likely to creep forward. Although some advocate adding a posterior fixation suture to the recession, and perhaps recessing a smaller amount, there are no data to suggest that this is preferable to simple recessions. And adding posterior fixation sutures definitely makes the procedure more difficult and increases the likelihood of complications, e.g., perforation and incarcerating the SO tendon [27].

## Question

What is your thought about unilateral IO anterior transposition in this patient group?


## Reply

In general, I do not like to do unilateral IO anterior transposition because of its effect on the palpebral fissure and versions. It does predictably cause a narrowing of the palpebral fissure, which can be a cosmetic problem in and of itself [33]. Also, if it results in good alignment in primary gaze, it often causes a noticeable limitation of elevation, which can also be a cosmetic issue [17].

## Basic Information

*Surgical Treatment of DVD in Patients with Approximately Equal Vision*

As stated, the majority of these patients have a strong fixation preference despite equal vision, and only one eye has a manifest DVD. There are two schools of thought regarding this common group of patients. One approach is to treat the DVD only in the nondominant eye with ipsilateral SR surgery. The advantage in this is that you get more effect from your surgery if you are not also weakening the SR in the fixing eye. Any SR recession in the fixing eye would tend to negate the surgery done in the nondominant eye. The disadvantage with unilateral surgery is that a substantial number of these patients will switch fixation post-op and present with a manifest DVD in the formerly fixing eye. Schwartz and coworkers reported that this occurred in about 10% of patients of this type undergoing unilateral surgery [39]. However, they considered only patients in whom this post-op DVD was more than 14 Δ. A manifest DVD of 10 Δ or a bit less can be a cosmetic issue. In a large series headed by von Noorden [38], about one-third of such patients shifted fixation and had a manifest DVD in the unoperated eye (Gunter K. van Noorden, MD, personal communication regarding data not included in published paper, 10 Nov 2015).

An alternative approach is to always perform bilateral surgery if the patients have the potential to switch fixation, to obviate causing the latent DVD in the fixing eye to become manifest. If this approach is taken, an enhanced surgical formula is needed as compared with unilateral surgery, for the reasons stated above. This approach is my personal preference.

I dichotomize treatment in this group of patients based on whether or not there is IO OA. To confirm the presence of IO OA you need to look for the presence of a V-pattern, bilateral fundus extorsion, and most importantly a HYPO of the adducting eye with alternate cover test in side gaze.

## Basic Information

*Surgical Treatment of DVD with Approximately Equal Vision and no IO OA*

I treat these patients with bilateral SR recessions. If there is a moderate difference in the size of the DVD (about 10 Δ difference) I recess the SRs 9 mm and 7 mm in the eye with the greater and lesser deviation, respectively. If the deviations are approximately the same magnitude, I will recess both SRs 7 mm for smaller deviations (up to approximately 15 Δ) and 9 mm if larger.

I never place the IO anterior to the IR insertion.

## Basic Information

*Surgical Treatment of DVD with Approximately Equal Vision with IO OA*

Bilateral IO anterior transposition is my procedure of choice in this group. I typically do it symmetrically unless there is a very large asymmetry to the deviation. If so, I still never place the IO anterior to the IR insertion, as doing so increases the risk of the AES [17]. Instead I may resect a small amount of the IO in the eye with the greater DVD. See Chap. 13 (Fig. 13.7) for my preferred technique for minimizing the occurrence of AES.

## Advanced Information

*Surgical Management of Recurrent DVD After Prior SR Recession*

Here I dichotomize my approach based on how large the prior recession was. If it was 7 mm or less, I re-recess the SR to 9–11 from the original insertion. If it was greater than 7 mm, I anteriorly transpose the IO. In this setting I would think carefully about doing this unilaterally, or bilaterally asymmetrically, based on the aforementioned admonitions about doing this procedure unilaterally. An alternative might be to resect the IR. I know of no large published series in which this approach was utilized, although it was Marshall Parks' preferred approach. I tend to avoid IR resections for DVD unless as a last resort, because IR resections narrow the palpebral fissure noticeably, and because isolated resections tend to stretch out with time.

## Advanced Information

*Surgical Management of Recurrent DVD After Prior IO Surgery*

In this setting, I am influenced by what was done previously to the IOs.

1. If they were recessed, they can be converted to anterior transpositions, which would be optimal.
2. If the IOs had previously undergone anterior transposition, I avoid moving them further anterior or resecting them, as this often causes AES and narrows the palpebral fissure. I would opt for SR recession. See Chap. 21, Case 21.27 on page 331 and Case 21.31 on page 334, for representative examples of these principles.

## Pearl

Recessing the SR in an eye that has undergone prior IO anterior transposition is very powerful and can cripple upgaze. Scale back on the surgical formula [33].

If the prior surgery was IO myectomy, the options are more limited for further IO surgery. SR recession would be the preferred option.

## Pearl

Even if you prefer IO myectomy to recession for routine IO weakening, consider doing recessions in patients with infantile esotropia. This will not close the door on subsequent anterior transposition.

## Advanced Information

*Surgical Management of Recurrent DVD After Prior IO and SR Surgery*

Treatment decisions here are predicated on what had been done previously to the IO or SR.

1. Prior small SR recession and prior IO recession: I recommend either IO anterior transposition or SR re-recession.
2. Prior small SR recession and prior IO anterior transposition: I recommend SR re-recession.
3. Prior small SR recession and prior IO myectomy: I recommend SR re-recession.
4. Prior large SR recession and prior IO recession: I recommend IO anterior transposition.
5. Prior large SR recession and prior IO anterior transposition: My recommendations are a matter of weighing trade-offs:
   (a) Re-recession of SR is technically difficult.
   (b) Resect or advance anteriorly transposed IO is technically harder.
   (c) Resecting the IR is the easiest option and least effective.
6. Prior large SR recession and prior IO myectomy: recommendations are a matter of weighing trade-offs.
   (a) Re-recession of the SR is technically difficult.
   (b) Resecting the IR is an option but less effective.

## Question

My patient has DVD and IO OA. I usually prefer IO myectomy to recession. Will IO myectomy help the DVD?


## Reply

Standard IO weakening in the form of myectomy or recession without anterior transposition will decrease some of the elevation in adduction, but will have little or no effect on the DVD in the primary position.

## Question

I have a patient with poor vision OS secondary to optic nerve hypoplasia. She manifests about 12 Δ of variable L HYPO, but also has DVD in that eye in that, at times, it supraducts manifesting about 15 of L DVD. There are vertical oscillations suggestive of the Heimann-Bielschowsky phenomenon [25]. The patient and her family only note the LHT. How can I address the LHT without making the manifest L DVD worse?


## Reply

A manifest hypo that intermittently changes to a manifest DVD tends to occur in one of the two settings. Sometimes it occurs after prior ipsilateral SR recession for DVD, resulting in a cosmetically noticeable overcorrection, but the DVD persists. Given how often large unilateral SR recessions are done for DVD, it is surprising that overcorrections of this type are fairly uncommon. The other scenario is accompanying the Heimann-Bielschowsky phenomenon, as in your patient. This phenomenon is characterized by monocular, coarse, pendular, vertical oscillations in an amaurotic eye. Although the amplitudes are usually small, they can be as large as 40–50 Δ. This is a difficult scenario to fix, but it can be improved upon. I have had satisfactory results by recessing the ipsilateral IR for the manifest HYPO, and placing a posterior fixation

suture on the SR. Jampolsky feels that the Heimann-Bielschowsky phenomenon is a manifestation of bilateral DVD in patients with one amaurotic eye. He advocates large bilateral SR recessions (Arthur Jampolsky, personal communication, 21 Oct 2016). This approach, of course, would have you doing aggressive surgery on the "good eye" of patients with an almost blind fellow eye. I have no experience with this approach.

## *Lagniappe* on Vertical Strabismus

### Question

I did a left IO myectomy on a girl at 2 years of age for what looked like a classic LSO palsy. She had 20 Δ of LHT in primary with a positive three-step test, a 25° head tilt to the right, and LIO "OA." She did well for about a year when she presented with a 25° head tilt to the left, an RHT of 25 Δ in primary, a positive three-step test for an RSO palsy, and slight RIO "OA." I assumed that this was a bilateral masked SOP. Is the time span of a year for it to "unmask" consistent with that diagnosis? Would you agree with my plan to weaken the RIO despite its only being slightly "overactive?"


### Reply

I do not fully understand why, but bilateral masked can "unmask" shortly after surgery or years later. This could be a bilateral masked SOP; however, it also could be a simple overcorrection, which can have a positive three-step test [14, 15], perhaps facilitated by late scarring or contracture that could explain the time course. An important differentiating finding is fundus torsion. If your patient has fundus extorsion in the right eye, I would think that this is a bilateral masked SOP and would weaken the RIO. If not, and especially if there is fundus incyclo OS, it may be a simple overcorrection. Had you recessed the LIO you could now reverse that. However, given that you did a myectomy, a RIR recession would be my suggested approach. Furthermore, the presence of fundus excylo suggests that the RIO myectomy scarred down in an anterior position, and you may be having some anti-elevation effect. Forced ductions are typically normal in that setting. If you go to the RIR, explore carefully for an anterior slip of the IO.

### Question

I recessed the LRs 8 mm OU using a suspension (hang-back) technique in a 10-year-old girl to treat a large intermittent exotropia. She was overcorrected and had UA of the right LR, and I suspected it has slipped. Several months later I explored the right LR and found that it had not only slipped and attached by a pseudotendon, but also the IO was scarred into the pseudotendon. I freed up the IO and let it go free, and advanced the RLR. One week after the re-op she had a 15 Δ R HYPO with marked underelevation of the right eye in adduction. I would not expect this degree of UA from an RIO palsy and suspect scarring and adhesions. Have you seen this?


### Reply

I agree that this degree of UA cannot be explained by a paralysis of the RIO. However, the time course is a bit early for actual scarring to cause this problem. I suspect that the IO has stuck down in an anomalous anterior position, altering its force vector and causing anti-elevation. You might even find that it was dragged forward with the LR causing the IO adherence syndrome [16]. The treatment is to reposition the IO back where it belongs.

### Question

I did an LIO recession on a 20-year-old man for a presumed congenital SOP. He is now 1 year post-op and is undercorrected. He has only 6 Δ of RHT in primary but 25 Δ RHT in adduc-

tion and 30 Δ RHT in the RSO field. He also has 8–9° of subjective excyclotropia. I think he needs the torsion corrected, so I am planning an RIO tuck and LIR recession. What do you advise?


## Reply

I agree that the 9° of torsion needs to be addressed, but I am concerned that with only 6 Δ of RHT in primary, a two-muscle procedure will overcorrect him. If you do a 1-h patch test and uncover a bigger HT in primary, your plan might be good. However, if not, I would suggest either a Harada-Ito procedure OD combined with an LIR recession or an LIR recession combined with a 7 mm nasal transposition of the RIR to correct the torsion. Either of these plans will minimize the risk of overcorrection in primary, address the fact that the HT is biggest down and left, and address the excyclotropia. Alternatively, a small RSO tuck is an option, but do less than usual or he may be overcorrected.

## Question

I care for a 5-year-old boy who had bilateral medial rectus recessions and IO anterior transpositions for infantile esotropia with DVD and IO OA. The IOs were placed 1 mm anterior to the IR insertions. He now has equal vision and alternates fixation. He manifests a constant 5 Δ L HYPO on simultaneous prism and cover test and an intermittently manifest 15 Δ R DVD and 5 Δ latent L DVD with the prism under cover test. The cosmetic issue is the R DVD of 15 Δ. I am considering (1) moving the RIO further anterior (I think this may be unpredictable), (2) RSR recession (I am concerned this will cause the L DVD to get worse), or (3) bilateral SR recessions. Your advice please.


## Reply

I am suspicious he may have AES in the left eye because the IOs were placed anterior to the IR insertions. The important thing here is to determine if the large R HT is in fact DVD versus a secondary deviation due to fixation duress from AES. To sort this out, do an alternate prism and cover test with 5 Δ base up OS. If, as you switch the cover from OD to OS, there is still a large RHT, you are dealing with residual DVD OD. If neutralizing the L hypo results in minimal downward shift OD as you switch the cover from OD to OS, you have determined that it is fixation duress in the left driving the bigger RHT. If you find the latter, carefully check forced ductions OS. The AES does not have a significant restriction on forced ductions as the anti-elevating vector is "innervational, e.g., it kicks in only on attempted elevation. If you find minimal restriction, convert the IO anterior transposition to a simple recession, which is the treatment for AES. If forced ductions are positive, there is a mechanical restriction that will need to be freed up, and usually the IR needs to be recessed. But if the prism testing does not confirm this to be a secondary deviation, you are dealing with residual DVD. I would then suggest bilateral asymmetric SR recessions. See Chap. 21, Case 21.2 on page 312 and Case 21.26 on page 330, for representative examples of these principles.

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# Diplopia

# 9

*Seeing double is not twice as good as seeing single, it's not even half as good*
*—A patient*

## Basic Information

*Overview*

In many cases the treatment of diplopia is straightforward. Correct the ocular misalignment and the diplopia resolves. In some cases, it is *not* that straightforward. Furthermore, in formulating a surgical plan in a patient with diplopia there can be special considerations that would not be issues if the patients suppressed. This chapter focuses on these latter situations.

## Basic Information

*Monocular Diplopia*

It is surprising to me how many patients with diplopia are referred to me as a strabismologist, when their diplopia is monocular. In most cases, the referring doctor correctly asked if the double vision went away if the patient closed one eye—and it did. However, if a patient has monocular diplopia, they will habitually close the affected eye when asked that question and report, "Yes, closing an eye eliminates the double vision." You have to then ask, "What happens if you close the *other* eye?" to determine that the diplopia is monocular. Patients usually do not understand the nature of binocular vision and diplopia, and do not intuit why we are asking them to close *an* eye.

## Pearl

If a patient says diplopia resolves if they close one eye, ask then what happens if they close the *other* eye.

## Basic Information

*Test for Fusion*

Before operating on any patient for diplopia, test to see if they are diplopia free if the deviation is offset with prism. It is most useful to do this in free space with handheld prisms while the patient fixates an accommodative target. Fixation on a light may fail to detect torsion or aniseikonia as an obstacle to fusion, and using red lenses or a Maddox Rod oriented vertically or horizontally is dissociating and does not simulate the real visual world. If a patient cannot fuse, consider several reasons.

1. Unexpected torsion can simulate a central disruption of fusion. I have seen torsion, when I did not specifically expect to, in cases of prior scleral buckling, prior penetrating keratoplasty, severe corneal scarring, monocular aphakia, long-standing strabismus, or prior vertical offsets of the horizontal rectus muscles [1]. Most were initially thought to have disruption of fusion because diplopia could not be eliminated with prisms. Always evaluate for torsion using the double-Maddox rod test. If you just ask the


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patient if the second image is tilted or at an angle, they will often answer affirmatively if there is a vertical and horizontal disparity, which they may describe as being "at an angle." I will ask them to look at a vertical line like the edge of a door or corner of the room, and ask if both lines run straight up and down like telephone poles.

2. Aniseikonia can also prevent fusion. When there is significant anisometropia the presence of aniseikonia may be obvious. In the absence thereof, the most common cause of unexpected aniseikonia is an epiretinal membrane [2]. I personally like the Awaya Aniseikonia test to screen for this issue (Fig. 9.1).
3. Horror fusionis or central disruption of fusion can be anticipated if there is a history of prior head trauma, long-standing strabismus, or a prolonged period of sensory deprivation like a monocular cataract [3]. With horror fusionis, the two images tend to repel one another similar to poles of magnets.
4. In the first situation the diplopia can be resolved if the torsion and other strabismus are corrected. Aniseikonia can be very hard to treat optically in this setting, as the magnitude of the disparity is often quite large. I know of no effective treatment for horror fusionis or central disruption of fusion.

**Pearl**

If a patient cannot fuse with their deviation offset with prisms, test for torsion and aniseikonia.

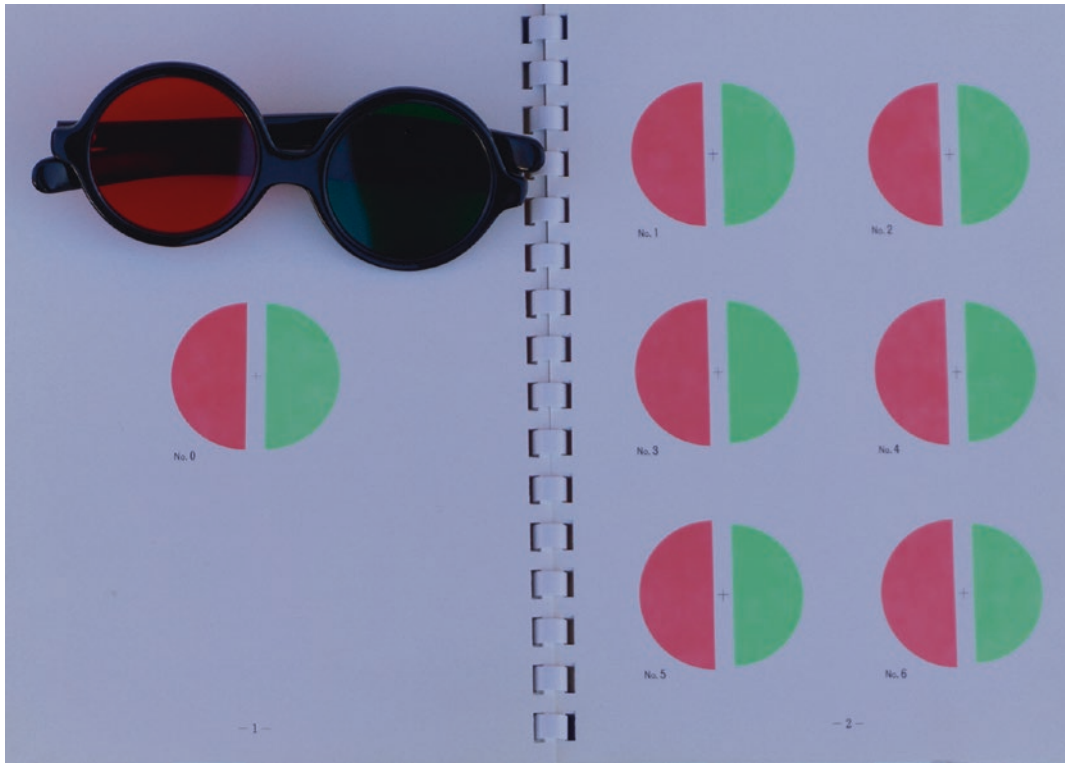


**Fig. 9.1** The Awaya Aniseikonia Test consists of pairs of *red* and *green semicircles*. The pairs progress with either the *red* or the *green* being larger than the other in increments of 1%. The patient wears *red-green* glasses and indicates which pair appears to be the same size

## Basic Information

*If the Diplopia Is Intermittent, Investigate the Pattern to the Intermittency*

Typically, the intermittency is mediated by gaze angle, e.g., downgaze, side gaze, and near fixation, or by activity, e.g., with prolonged reading or degree of fatigue.

1. If the intermittency relates to gaze angle, the patient usually has incomitant strabismus, and the treatment involves a surgical plan that addresses the incomitance. One particularly important scenario is if the diplopia is limited to downgaze. These patients may be asymptomatic until they become presbyopic and need to use a bifocal, which of course requires downgaze. In a series of 51 patients with diplopia limited to downgaze, causes included [4]:
   (a) An A-pattern (16%) or a V-pattern (16%) with orthophoria in primary
   (b) A hypertropia in downgaze due to a lag of one eye resulting from prior inferior rectus (IR) recession, typically for thyroid eye disease or orbital floor fracture (43%)
   (c) A lag of one eye on depression from other causes, typically partial third cranial nerve palsy (20%)

Of the 51 patients, 20 (39%) were successfully managed optically, 21 (41%) surgically, and 10 (20%) both surgically and optically. Optical management consisted of:

   (a) Fresnel prism over bifocal segment in (4 patients)
   (b) Slab-off or reverse slab-off prism in bifocal (2 patients)
   (c) Single-vision reading glasses (7 patients)
   (d) Switch from progressive to nonprogressive bifocal (3 patients)
   (e) Flat-top bifocal set higher than usual to use as "library glasses" (16 patients)

My decision as to which of the above modalities to try was based upon the patient's refractive error, visual needs, and size of the deviation in the reading position. For example, if the patient had a minimal refractive error and did not need glasses for most distance viewing activities, I recommended single-vision reading glasses. If the patient usually wore glasses and the deviation in the reading position was small—5 prism diopters (Δ) or less—I recommended slab-off or reverse slab-off prism in the bifocal. If the deviation in the reading position was greater than 5 Δ, I tried a Fresnel prism over the bifocal add. If the reading position deviation was 5 Δ or less, and the patient was using a progressive bifocal, I switched them to a flat-top bifocal or recommended "library glasses" as described above.

Sometimes this problem can result from a change in style of spectacle correction. Consider this patient:

**Case 9.1** This 55-year-old woman had a history of an orbital floor fracture repair in her right eye, which subsequently required a RIR recession. She had a small right hypertropia in far downgaze but was asymptomatic in her bifocals. She obtained a new pair of glasses with the identical prescription as her prior pair, but was diplopic for reading in the new glasses. A review of prior records showed that her prior pair of bifocals was a flat top D-seg. The new glasses were progressive lenses. As mentioned earlier, the progressive lenses required her to look further in downgaze to read with clarity, resulting in diplopia.

Surgical management of the patients with diplopia limited to downgaze consisted of [4]:

(a) Posterior fixation on contralateral IR (10 patients)
(b) Correction of A- or V-pattern (5 patients)
(c) Bilateral asymmetric IR recession (6 patients)

## Important Point

*Progressive bifocals can be problematic for patients with vertically incomitant strabismus.* They have a wide transition zone of as much as 16 mm between the distance and near optical center, and

require the patient to look significantly into downgaze to optimize near clarity. A "blended" bifocal is a no-line bifocal with a narrow transition zone of 2–3 mm, which works better for those patients who want to continue in a no-line bifocal.

2. If the diplopia is intermittent because it occurs only at distance or only at near fixation, the obvious causes are those in which there is a distance/near difference in alignment, e.g., convergence insufficiency and divergence insufficiency. However, if the patient fixes with one eye for distance and the other for near, you should always be on the lookout for fixation switch diplopia [5]. This occurs because some patients suppress when they fixate with their dominant eye, but do not transfer the suppression scotoma to the dominant eye when they fixate with their nondominant eye. In many cases, fixation switch as a cause of diplopia is obvious, but in some it can take a high index of suspicion to identify this problem.

   In a series of 16 patients with fixation switch diplopia [5], six had the problem because of iatrogenic monovision, and another six because they were prescribed an unbalanced refractive correction usually because their manifest (dry) refraction did not uncover the full amount of hyperopia in an amblyopic eye. In some patients the cause was insidious and not immediately obvious, such as in the following case:

**Case 9.2** This 21-year-old man had been diplopic for 2 years. As a child he was treated for amblyopia in his left eye. His uncorrected visual acuity was OD 20/50 and OS 20/25. His refraction and best corrected visual acuity was OD −0.75 sph 20/20 and OS +0.75 sph 20/25. He had a 10 Δ esotropia (ET). He was told he did not need glasses as he could use his left eye for distance and right eye for near. However, when he fixated with his formerly amblyopic left eye, he was diplopic due to fixation switch diplopia. Despite the seemingly trivial refractive error, he had to have his myopia in the right eye corrected with a spectacle lens in order to avoid switching fixation, and this eliminated his diplopia.

**Pearl**

*Amblyopic eyes have a decreased ability to discriminate lens changes.*

You may need to refract them objectively (retinoscopy or autorefractor) after cycloplegia to determine the amount of hyperopia present.

If diplopia is intermittent, determine if it is a motor vs. sensory intermittency.

3. If the intermittency is mediated by fatigue or state of alertness, it is helpful to determine if it is a motor or sensory intermittency [6]. A motor intermittency is characterized by the initially single image, separating into two that drift apart, and as fusion is regained the images will come together and blend into one. Conversely, a sensory intermittency is characterized by a sudden appearance of the second image in the periphery when the diplopia appears, which then will disappear from the same location in the visual periphery when the diplopia goes away. With a motor intermittency, it can be helpful to do a 1-h patch test to determine the full underlying deviation [7] and treat it accordingly. If the deviation is small, prisms may be a successful treatment option, and if it is larger than 10–15 Δ, surgery is typically needed. Usually these patients can be treated easily and successfully. A sensory intermittency is harder to treat. In this situation there is not an intermittent strabismus that is causing the problem. Instead, the sensory system switches from suppression to simultaneous perception without a change in motor alignment. The only modality I have found effective is to do a meticulous refraction and correct any changes in the refractive error, even if they seem trivial. This has proven successful in about one-third of these patients [6]. Hoyt informed me that he has treated some of these patients successfully with pilocarpine (Creig Hoyt, MD, personal communication, 30 Nov 2016). He feels that the miosis reduces (but does not entirely eliminate) the perception of the peripheral image. I have no experience with this treatment approach.

## Pearl

If you put a patient in prisms and they do not help, you must see him back with the prism glasses to determine if they were made incorrectly, or a Fresnel prism was placed with incorrect orientation, or the strength of the prism was not adequate. It surprises me how often such patients do not bring the prism glasses to the follow-up visit because they "did not work." Similarly, if a patient balks at trying prisms because they did not work in the past, you should get outside records to see what amount was prescribed. It may have simply been an inadequate dose.

## Question

I care for a 45-year-old man who has had a partly accommodative ET all his life. He was advised to have strabismus surgery as a child but his parents refused the offer. Over recent years his ET has been increasing, as has his need for additional plus correction. When I saw him for the first time at 44 years of age, he manifested 20 Δ of ET with his glasses, which undercorrected his hyperopia by about +1.50 diopters OU. He required almost all of that additional plus for visual comfort, resulting in a prescription of +5.25 sph OU. However, with the added plus, he has a manifest ET of 12 Δ with crossed diplopia. He also has crossed diplopia with the deviation offset with prisms and takes 8 Δ BI (base-in, of prism) to be diplopia free. Should I operate on him for his residual ET, and if I do, will he be diplopic?


## Reply

This patient probably had gone a number of years manifesting about 20 Δ of ET and had adapted to that angle with anomalous retinal correspondence (ARC).

In most cases problems like this can be corrected, but it may take a while. I would put him in whatever Fresnel prism is needed to make him diplopia free—in this case, 8 Δ BI. Then, I would try and wean him out of the prism by decreasing it a prism diopter or so every month or so. I have had some patients for whom this approach was successful in eliminating prism completely and others whom I was able to wean down to only a few prism diopters, which I had ground in their spectacles. If you operate on him now, there is a high likelihood of post-op diplopia.

## Advanced Information

*Recent Onset of Diplopia in Patients with Long-Standing Strabismus*

On occasion an adult patient with a history of childhood strabismus will present with diplopia that developed only recently. In my experience with 152 such patients, I was able to eliminate the diplopia in 132 (87%) following a systematized approach [6]. In most cases the onset of symptoms could be traced to:

1. A change in motor alignment, often with respect to directionality, e.g., a former esotrope who drifted exotropic
   *or*
2. A change in refractive management, often the intentional introduction of monovision [8]
   *or*
3. A change in the refractive needs: This most often was characterized by premature presbyopia causing a breakdown of an old accommodative ET, or a change in refraction management causing fixation switch diplopia as in the earlier example. Another common scenario was the patient with vertically incomitant strabismus beginning to need a bifocal.

A review of prior records in these patients can be most helpful in determining the motor state prior to the onset of the diplopia. The indicated treatment is to return the patient to that state.

## Advanced Information

If a patient is diplopic only through their bifocal segment, and if they have any significant amount of anisometropia, consider the possibility of induced prism from the anisometropia being the culprit [9].

To remedy this, you must do a prism and cover test measurement at near in downgaze with the distance prescription and add in place, and determine if there is a vertical misalignment. If so, this can be remedied with vertical prism in the bifocal via slab-off or reverse slab-off principles. To understand the difference, refer to Fig. 9.2. The slab-off technique removes a wedge of lens material, in effect, removing a base-down prism. Doing this the effect is, therefore, of adding base-up prism (it would take the removed base-down prism to neutralize it). Reverse slab-off does the opposite and induces a base-down effect.

## Pearl

Depending on the patient's refractive error, there are often technical considerations that make it more desirable to correct an anisometropia-induced prism with slab-off prism in one lens as opposed to reverse slab-off in the other, or vice versa. I typically write a prescription to give the optical lab a choice. A hypothetical example of how I write such a prescription is "… and 2 Δ base up in the seg OD via slab-off, or 2 Δ base down in the seg OS via reverse slab-off, at your discretion."

## Advanced Information

Diplopia after refractive surgery is particularly upsetting to the patient and physician. In a series of 28 such patients, Lionel Kowal and I identified five major causes for the problem [10]:

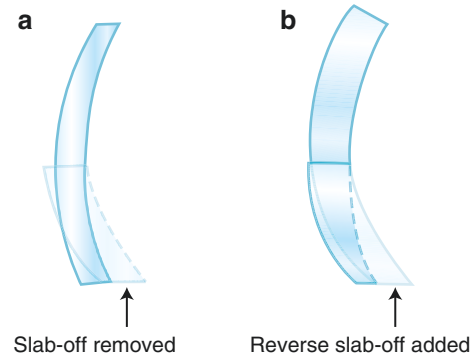


**Fig. 9.2** (**a**) Slab-off prism removes a wedge of base-down prism, thus creating a base-up effect. (**b**) Reverse slab-off does the converse, creating a base-down prism effect

1. Technical problems: these involved either scarring or having an optical zone smaller than the resting pupil size.
2. Aniseikonia induced by the refraction change.
3. Iatrogenic monovision.
4. Prior need of prism, usually unrecognized by the refractive surgeon.
5. Improper control of accommodation: This most often occurred in patients with a prior history of accommodative ET, in whom an overcorrection of myopia occurred with the refractive surgery, leaving the patient hyperopic.

## Question

My local refractive surgeon sends me patients with a history of strabismus to screen for potential post-laser in situ keratomileusis (LASIK) diplopia. What exactly should I do?


## Reply

The following screening procedures would have identified all of the patients pre-LASIK in the aforementioned series as being at risk, except those in whom technical problems were to blame.

1. Ask about the history of need of prisms, and check present glasses for prism. If prisms are in present glasses, do a pre-LASIK trial without prism or with Fresnel prism to neutralize existing prism.
2. Manifest (dry) refraction: For myopes surgery should target the least minus that gives threshold acuity. For hyperopes surgery should target the most plus that provides threshold acuity.
3. Cycloplegic refraction: If this uncovers a significant amount of latent hyperopia, there is potential for problems with esotropic patients.
4. In patients with a history or findings of diplopia, prism in glasses, intermittent strabismus, or a moderately sized phoria, check fusional convergence and divergence amplitudes to assess fusional reserve. If low, there is less margin for error with refractive surgery.
5. If monovision is the desired outcome of refractive surgery, do a pre-LASIK optical trial of monovision with spectacles or contact lenses in patients with a large phoria or any manifest strabismus.

## Question

I have a patient who needs both vertical and horizontal prism. I would like to use a Fresnel prism oriented obliquely over one eye so as not to induce blur OU. How can I determine the resultant power and orientation of the prism? Trial and error is frustrating, because as I change the orientation of the prism, the vertical and horizontal effective prism power keeps changing.


## Reply

There are two methods that work well.

Take the hypothetical example of a patient needing 7 Δ base-down and 10 Δ base-out. Draw a right triangle with the base 10 units (cm, inches, or whatever) long and the vertical side 7 units high. Draw and measure the hypotenuse, which will be, in this case, 12 units. You will need a 12 Δ prism. To determine its orientation, take a protractor and measure the angle between the hypotenuse and the base; it will be 35. You will need to orient the 12 Δ prism with its apex pointing toward 35°. If you do not have access to a protractor, you can cut out the triangle and hold it up to a trial frame (Fig. 9.3).

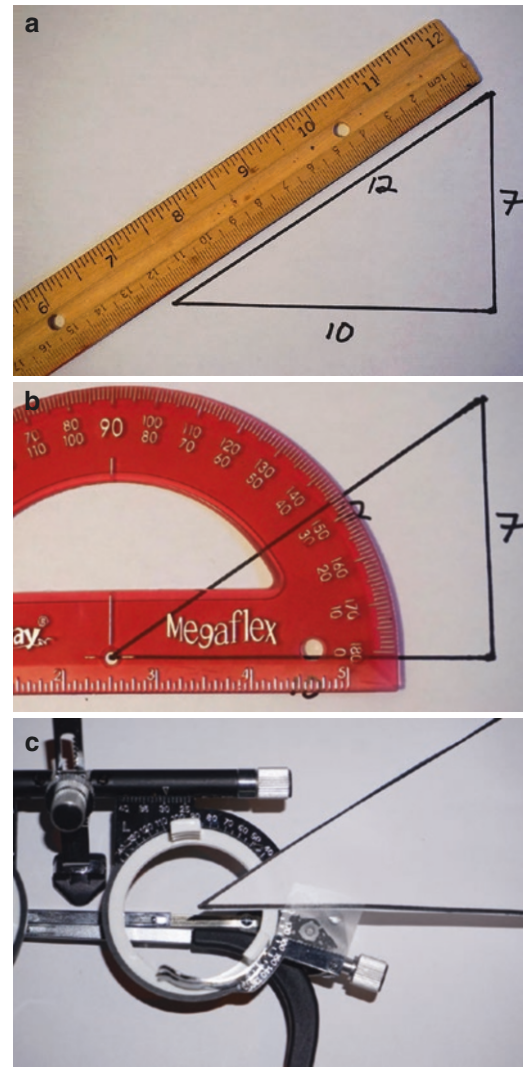


**Fig. 9.3** (**a**) To orient one prism to equal 10 Δ base-in and 7 Δ base-up, draw a right triangle with the base 10 units long and the vertical side 7 units long. Draw and measure the hypotenuse which will be 12 units, indicating that the power of the prism is 12 Δ. (**b**) Determine the angle between the hypotenuse and the base with a protractor, which in this case is 35°. The apex of the 12 Δ prism should point toward 35° on the lens. (**c**) An alternative to using the protractor is to orient the triangle on a trial frame to determine the angle

The Tanganelli vector method is another technique that involves less steps but requires a more attentive patient. Put a red lens in a trial frame over one of the patient's eyes, and a lens that will dissociate a light into a line such as a Bagolini lens. The patient will see a red light and a second white light with a line streaking through it. Then rotate the Bagolini lens until she sees the line connecting the red and white light. That tells you the orientation of the base of the final prism. Then put a horizontal prism bar oriented perpendicular to the axis you determined earlier, and run up base-out prism until the red and white lights merge. That determines the power of the final prism (Fig. 9.4). A Maddox rod will work as well as a Bagolini lens.

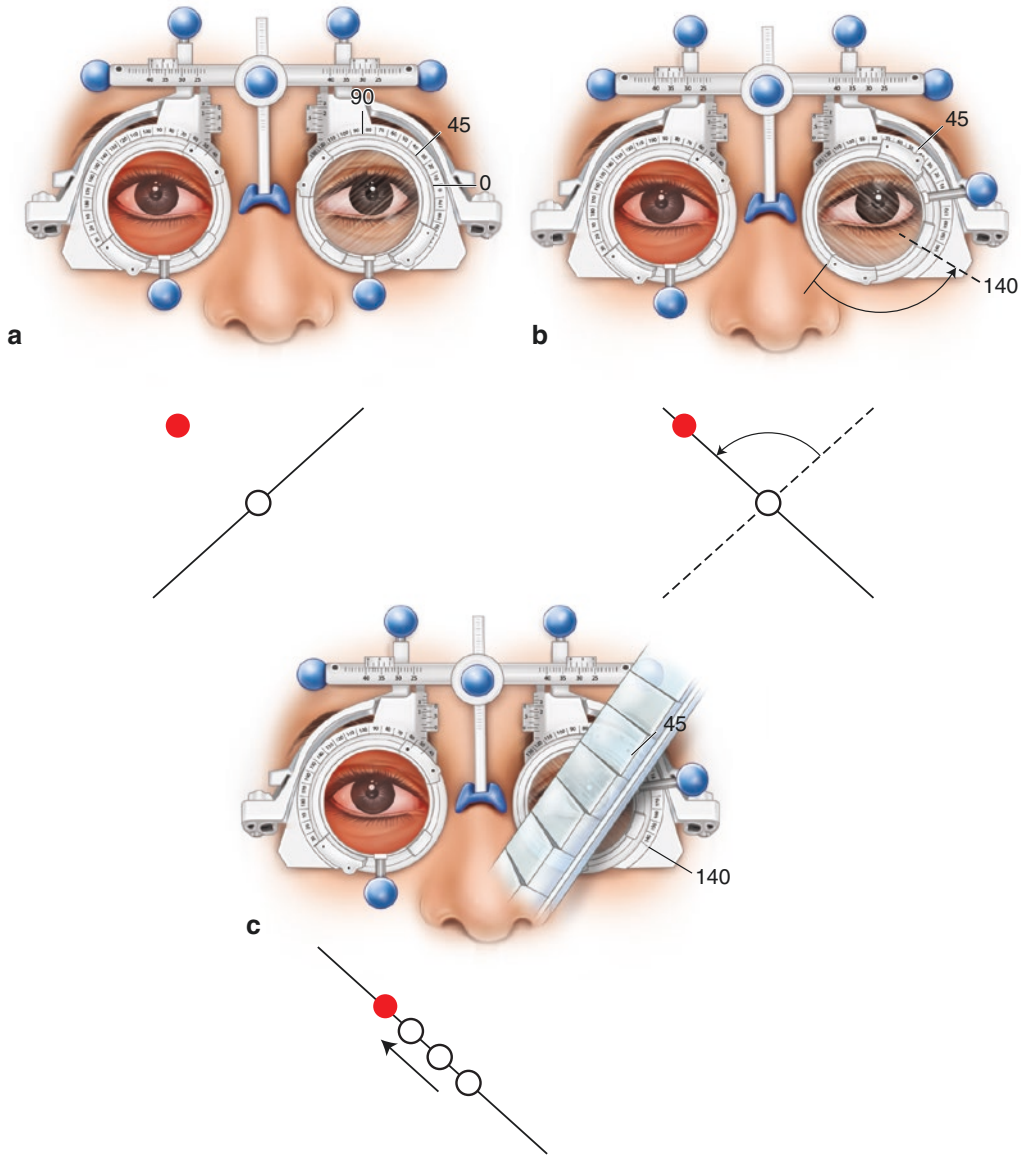


**Fig. 9.4** (**a**) For the Tanganelli vector test, a red lens is placed over one eye and a Bagolini lens over the other. The patient looks at a light and will see a *red* light and a *white* light, with a line streaking through the *white* light. (**b**) The Bagolini lens is rotated so the line connects the two lights. This is the axis at which the prism is to be oriented. (**c**) A *horizontal* prism bar is placed perpendicular to the determined axis of the prism, and the prism power increased until the two lights merge. This determines the power of the final prism

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# Strabismus Surgery

# 10

*Surgeons must be very careful*
*When they take the knife.*
*Underneath their fine incisions*
*Stirs the culprit,—life!*

*—Emily Dickinson*

## General Concepts

### Basic Information

We usually straighten eyes by shortening (recessing) muscles, which is thought to weaken them, or stretching (resecting, plicating, tucking, etc.) muscles, which is thought to tighten or strengthen them. When muscles are recessed, there are two different effects. An acutely shortened muscle should be weaker because of Starling's law on muscle contraction, which states that the more a muscle is stretched, the greater its force of contraction. With recession, slack is induced in the muscle, which shortens the length of each sarcomere, placing the actin and myosin filaments in a less advantageous relationship, thus decreasing both the contractile and elastic force of the muscle (Figs. 10.1 and 10.2) [1]. A muscle's total force is the sum of its elastic force, which rises with stretch, and its contractile force, which peaks at the muscle's normal resting length, but decreases if the muscle is either longer or shorter (see Fig. 10.2). Consequently, the total force is biphasic, decreasing with small amounts of stretch and then increasing with greater stretch. The muscle's torque is force times the lever arm. As long as the muscle is not recessed posterior to the equator, the reduction in torque is solely due to a decrease in its force. In theory, when a muscle is tightened a small amount via resection, plication, or tucking, there should be a small decrease

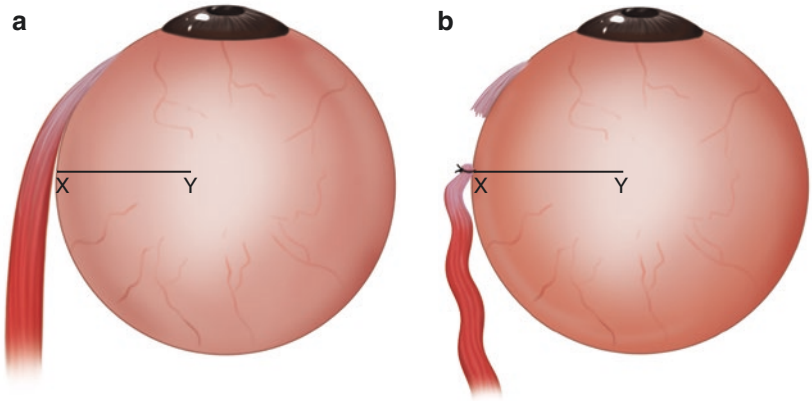



**Fig. 10.1** (**a**) Prior to surgery the muscle inserts anterior to the equator (*X*) which is the functional point of tangency. (**b**) If it is recessed to a point that is at or anterior to the equator, slack is created in the muscle shortening each sarcomere; however, the lever arm (*X*—*Y*) is not reduced


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_10

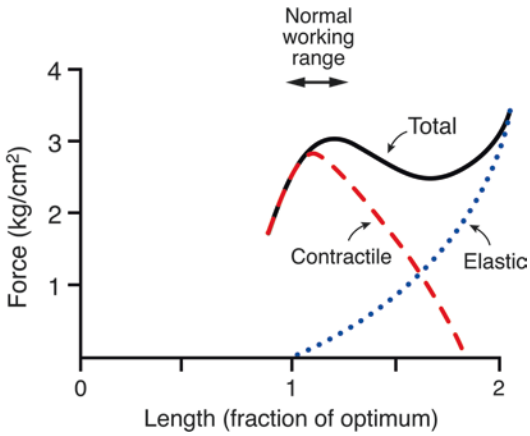
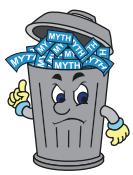



**Fig. 10.2** The length-tension curve for the elastic force of an extraocular muscle rises exponentially with stretch (*dotted blue line*). The length-tension curve for contractile force peaks at the resting length of an extraocular muscle and falls off at greater and shorter lengths (*dashed orange line*). The length-tension curve for total force of a muscle is the sum of the elastic and contractile force and is biphasic, peaking near the resting length (*solid black line*) (from Kushner [1], with permission. © 2006 American Medical Association. All rights reserved)

in total force. As the amount of tightening increases, the force increases primarily due to an increase in the elastic force; the contractile force decreases further. That is why these procedures serve more to "tighten" rather than "strengthen" muscles. But both effects, sarcomere shortening from recession or elongating due to stretching from tightening procedures, are probably temporary because sarcomere remodeling rapidly will restore the sarcomeres to their normal resting length [1, 2]. (For more information on this see below, Advanced Information: How Does Strabismus Surgery Work—Induced Effects.)

Resections serve more to tighten than strengthen muscles.




## Myth

Resections, plications, and tucks "strengthen" muscles.

## Fact

They actually increase the muscle's total force by "tightening" it via an increase in the elastic force, but not the contractile force. Consequently, the main effect of a tightening procedure is often in the opposite gaze field; for example, resecting a medial rectus muscle (MR) may give more correction in *abduction* than on *adduction*.

## Basic Information

*Should Recessions Be Graded from the Insertion of Limbus?*

Although older texts describe the extraocular muscles (EOMs) as each inserting a uniform distance from the limbus, it has been shown that there is variability for each of the EOMs [3]. This realization gave rise to a surgical formula based on recessing muscles a graded distance from the limbus, rather than from the insertion [4]. The reasoning went as follows: Consider two hypothetical eyes in which the MR inserted 3.5 mm from the limbus in one and 5.5 mm in the other, but in both the equator was 10.5 mm from the limbus. If you recessed the MR 5 mm in each, you would presumably be doing a different procedure in the two patients. In one you would be putting the muscle 2 mm anterior to the equator, and in the other at the equator. Conversely, it was argued that if you place the muscles a similar distance from the limbus—let's say 10.5 mm—you would also do a different operation in the two patients. In one you would be shortening the muscles 7 mm, and in the other only 5 mm (Fig. 10.3) [5]. To decide if recessions should be graded from the insertion or the limbus, it is important to know if the effect of a recession relates to the amount of shortening of the sarcomeres versus where the muscle ends up referable to the equator. This can be a difficult question to answer via a statistical analysis, because in any given patient, the farther the muscle is put from the insertion, the farther it

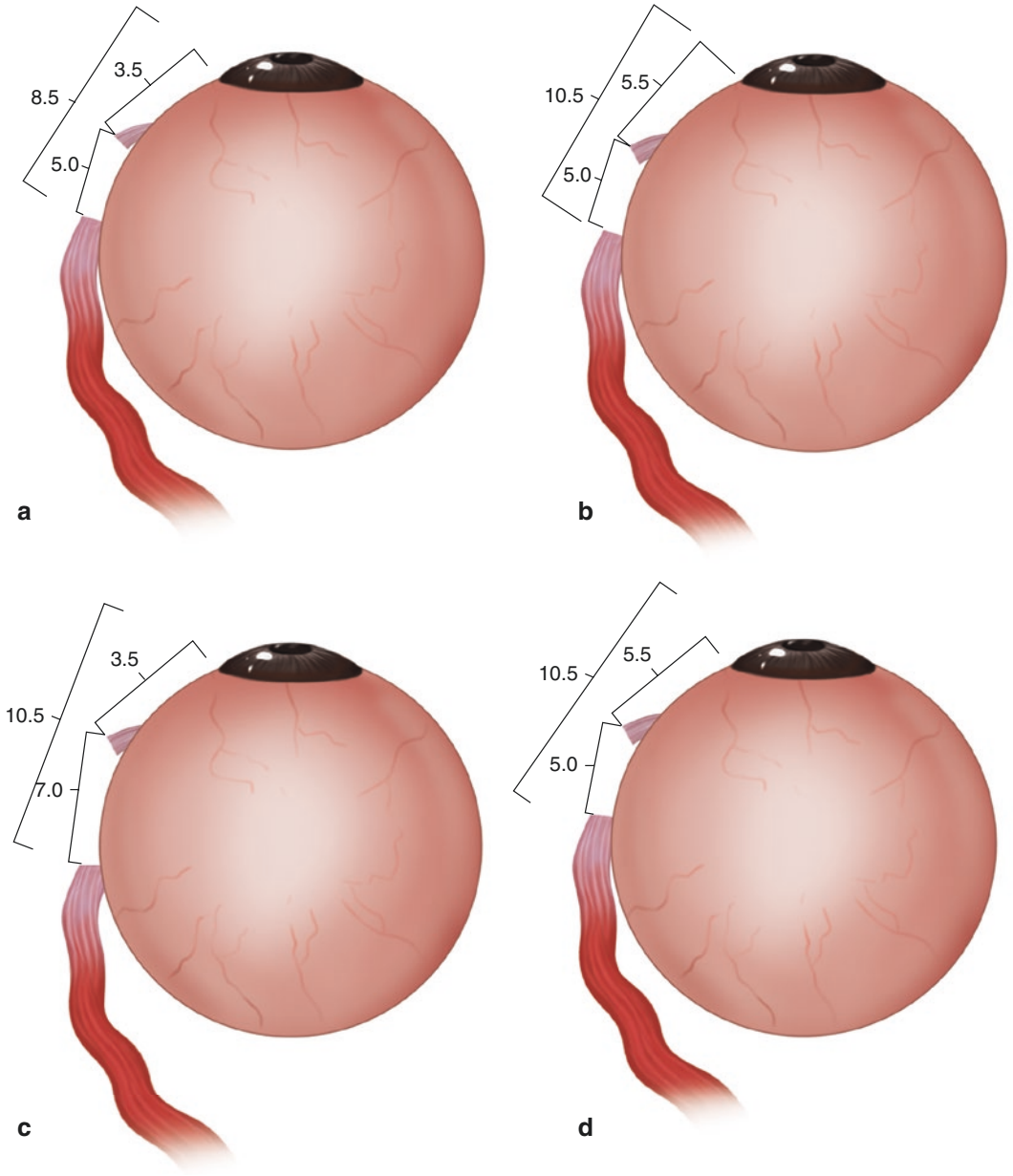



**Fig. 10.3** (**a**) If the equator is 10.5 mm from the limbus, and the medial rectus inserts 3.5 mm from the insertion, a recession of 5 mm will put the muscle 8.5 mm from the limbus and 2 mm anterior to the equator. (**b**) But if the original limbus insertion distance was 5.5 mm, a 5 mm recession will put the muscle 10.5 mm from the limbus and at the equator. (**c**) Recession of the muscle to the equator (10.5 mm from the limbus) would require a 7 mm recession if the muscle initially was 3.5 mm from the limbus, (**d**) but only 5 mm if the muscle had inserted 5.5 mm from the limbus

is also put from the limbus, and vice versa. However, using partial correlation coefficients, it was determined that the response to surgery correlated more strongly with the amount of recession from the insertion than from the limbus [5]. In other words, it is the actual amount of shortening of the muscle that is important. This was not surprising to me.

## Basic Information

*Should Recessions Actually Be Measured from the Insertion?*

Saying that your recession formula should be based on a given number of millimeters of recession from the insertion for a given size deviation is not the same as saying the actual measurement should be from the insertion. There are known artifacts that affect the accuracy of strabismus surgical measurements. After you disinsert a muscle and grasp the insertion with forceps to facilitate suturing, the entire insertion can move forward by as much as 1 mm due to temporary compression of the scleral fibers between the forceps and the insertion, depending on how much traction you are exerting [6]. In some cases there can be a lamellar tearing of the scleral fibers anterior to the insertion.

## Question

Given the aforementioned artifacts of measuring, should I measure from the insertion or from the limbus?


## Reply

If you believe that millimeters matter, and I personally do, the ideal thing is to measure the insertion to limbus distance before you grasp the insertion, and then again after you fixate it with forceps. If it has changed appreciably, you should measure from the limbus, but use a formula based on recession from the insertion. For example, if you want to do a 3.5 mm recession, and you measure the insertion-limbus distance initially as being 5.5 mm, you should measure your recession as being 9 mm from the limbus.

## Question

What about the artifact this introduces with respect to an arc versus cord measurement? Do I need a curved ruler if the recession is more than 10 mm?


## Reply

You cannot go wrong with a curved ruler, but it really is not necessary. The difference between an arc and cord measurement is based on the assumption that the globe maintains its almost spherical nature while you are measuring. In practice there is usually some flattening of the globe due to traction from the fixation forceps, and you can increase this effect by intentionally pushing down on the forceps to flatten the globe somewhat. Another simple alternative to a curved ruler is to cut a piece of suture material and use it as a measuring device as shown in Fig. 10.4. This has several advantages over a curved ruler. It will conform to different globe curvatures, which a curved ruler will not do. Also, if you wet the measuring suture after placing it on the globe, it stays in place while you suture, unlike a mark made by the end of the metal ruler, which can fade.

## Question

Should you measure from the insertion? Doesn't the insertion "move forward" after the muscle is disinserted?

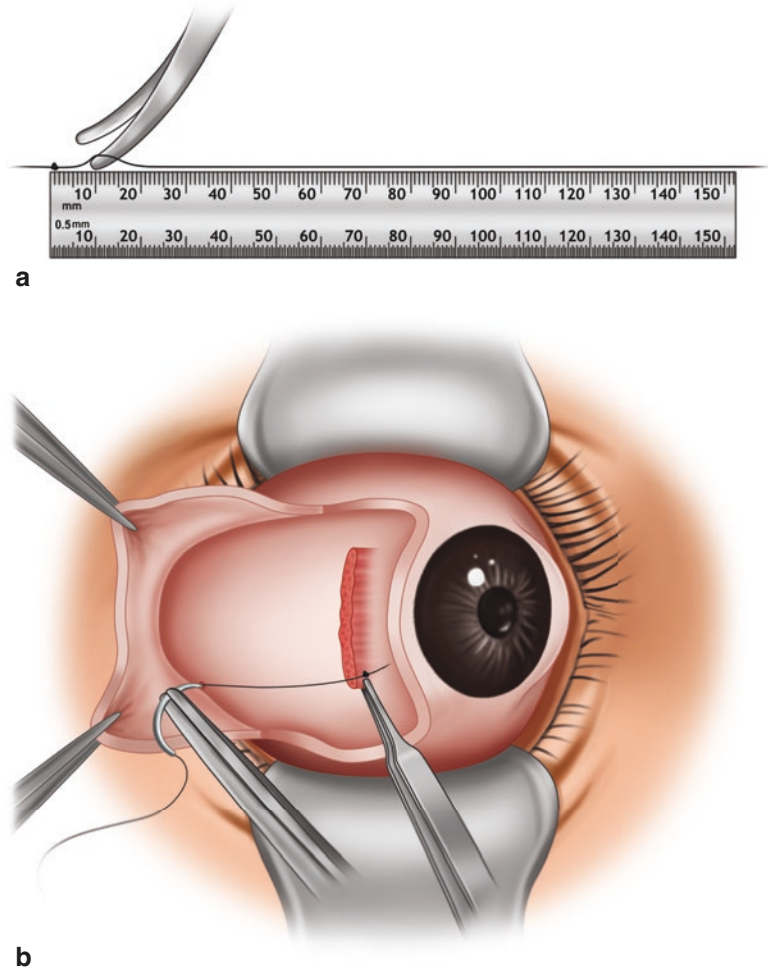



**Fig. 10.4** (**a**) A piece of suture material about 2 cm long has a knot tied several millimeters from one end. The knot is placed at the zero mark on a ruler and the suture is cut the desired length of the recession. (**b**) The piece of suture is placed on the globe with the knot at the insertion. The cut end of the suture marks the point at which the muscle should be sutured


## Reply

Yes and no. There is a common myth that the tension of the muscle pulls the insertion backwards, and that when the muscle is disinserted the insertion moves forward. That is not exactly true. If you measure from the limbus to the insertion prior to disinsertion, and then rotate the eye in the direction of the muscle, thus relaxing that backward pull of the muscle, the limbus-insertion distance is unchanged. This means it is not the pull of the muscle that is drawing the insertion posteriorly. But if you then disinsert the muscle and remeasure, the limbus-insertion distance appears less. This is actually an artifact related to the anatomy of the muscle's insertion as explained below.

## Try This Experiment

During surgery, hook the muscle and then put a mark (methylene blue or something similar) just anterior to but touching the front of the hooked muscle. This would denote what you would consider to be the anterior edge of the insertion. Place a second mark about 2 mm anterior to that, and a third at the

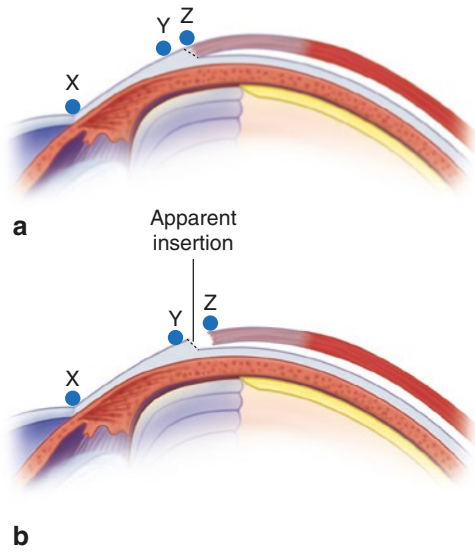



**Fig. 10.5** (**a**) Orientation of a rectus muscle insertion. The muscle is in a bed of sclera that is thinner behind the insertion. The muscle fibers insert running parallel to the scleral fibers. A *dot*, *Z*, is placed on the sclera just anterior and adjacent to the insertion. A *second dot*, *Y*, is several millimeters anterior to *Z* and a *third dot*, *X*, is at the limbus. Because the scissor blades are flat on the sclera during disinsertion, the transection plane is depicted as the *dotted line*. (**b**) Because of the configuration shown in (**a**), the *dot* just anterior to the insertion, *Z*, gets cut off with the muscle, and it appears as though the insertion migrated anteriorly (*arrow*). However the distance from the limbus, *X*, and the arbitrary *dot*, *Y*, does not change. This indicates that the sclera anterior to the insertion does not actually migrate anteriorly

limbus. Measure the distance from the limbus to the marked insertion, and from the limbus to the mark anterior to the insertion. Then disinsert the muscle. You will see that the mark, which you thought was just anterior to the muscle, gets cut off with the muscle. The anterior edge of the disinserted tissue will be somewhat anterior to the mark. This is because the muscle's insertional edge is perpendicular to the sclera, and the scissors for disinsertion are used with the blades parallel to the sclera, necessitating some of the sclera just anterior to the insertion to be cut off (Fig. 10.5). It is the anterior edge of this cut that appears to be the anterior edge of the insertion. However, the distance from the limbus to the mark that was anterior to the insertion does not change, indicating that the scleral fibers, and hence actual insertion, do not migrate anteriorly [6].

## Question

Do millimeters really matter? Don't recessions just come small, medium, and large, and these small differences in measurement can be ignored?


## Reply

I personally feel that these small differences are meaningful, at least in the short term with respect to the immediate response to surgery, and before sensory adaptations come into play. We can see at the time of postoperative suture adjustment that small changes in muscle position have meaningful changes in alignment. But my feelings are also based on a study I have conducted over the past 40 years. At the end of every recession I perform, I of course look at the muscle and decide if I feel it is positioned satisfactorily. If it is sagging or in some way not right, I fix it. But if it looks to me to be in good position, I then measure the actual amount of recession I obtained in the following masked manner. I have a caliper that does not have a millimeter scale on it, and have it set to some unknown distance by my operating room technician. Next I measure what the actual recession is with those calipers, and then hand them to the tech or fellow to read the actual distance by placing the calipers on a millimeter ruler. I have kept a record of the actual recession, the desired recession, and the deviation in a database for about 10,000 patients. If the actual recession deviated from the desired recession, I did not reposition the muscle if I had been satisfied with its position prior to the masked measurement. There were no muscles for which the masked measurement deviated by more than 1 mm from the desired measurements. There were some patients in whom the sum of the variance (right and left eyes) was 1.0 mm and a smaller number in whom the sum of the variance from that which was planned totaled 1.5 mm. After 40 years of collecting these data, I analyzed the response to surgery in two homogenous groups of patients—partly accommodative esotropia (ET)

with normal accommodative convergence/accommodation (AC/A) ratio and intermittent exotropia, in whom only bilateral symmetric MR recessions or lateral rectus muscle (LR) recessions were performed, respectively. None had adjustable sutures and none of the recessions were done using a suspension (also known as hang-back) technique. To avoid the confounding effect of extreme recessions (large or small), I limited my analysis to esotropes in whom the recessions ranged from 3.5 to 5.5 mm per muscle, and in intermittent exotropes from 4 to 8 mm. I compared the mean response for all patients who underwent a given amount of planned muscle recession and in whom the actual recession was equal the planned amount; for example planned 5 mm MR recession had a masked measurement of 5 mm, and compared that to the mean response for those patients who underwent a planned recession of the same amount, but for whom the actual recession deviated from what was planned. I found that the mean response at 1 week after surgery for esotropes was 2.8 prism diopters (Δ) more in patients in whom the actual measurements totaled 1.0 mm more than what was planned (sum of variance in right and left eyes—either 0.5 mm in each eye or 1 mm in one eye and none in the other), and 2.7 prism diopters (Δ) less if the actual measurements totaled 1.0 mm less than planned. For intermittent exotropes the results were similar with the increased response being 2.9 Δ if the masked measurements totaled 1.0 mm greater than what was planned, and 2.7 Δ less if the actual measurements were 1.0 mm less than what was planned. These differences were all significant ($P < 0.01$). For those patients in whom the actual measurements differed from the planned measurement by a total of 1.5 mm the discrepancy was 4.7 for esotropes and 5.1 Δ for intermittent exotropes. These differences were all significant ($P < 0.05$). I realize that an assessment made at 1 week after surgery does not foretell the final result, but it does reflect the immediate response to surgery before it is contaminated by sensory adaptations. But it means to me that it pays to be meticulous with respect to measurements.

Millimeters do matter.

## Basic Information

*Slipped Muscles and Stretched Scars*

Slipped muscle and stretched scars are distinctly different entities, which have in common the clinical picture of a muscle underaction after surgery. They both result from an improper bond forming between the muscle and sclera. If the muscle had been recessed, there is a progressive overcorrection. If the muscle had been resected, there is a progressive undercorrection. Although both slipped muscles and stretched scars share these clinical findings, they are different entities, have different causes, and—perhaps most importantly—should be managed differently.

1. A slipped muscle (or muscle slipped in the capsule) is caused by faulty suturing of the muscle, whereby the sutures incorporated only capsule and not a substantial amount of muscle tissue [7]. Usually a muscle that is slipped in the capsule will be evident within weeks or a few months after surgery. I think most cases can be prevented if attention is paid to a little known maneuver that I routinely practiced after muscle suturing. I would do the following. After I sutured the muscle and disinserted it, I reflected the cut end of the muscle back so that I could see its undersurface and if the central muscle fibers were secured with sutures, or if they were slipping posteriorly. It surprised me how often I would see that in the center of the muscle's cut end, the sutures were just in capsule and not securing actual muscle, despite my conscious attempt to be mid-thickness when I sutured the muscle. If I do see that the center of the muscle is not well secured, I pass one of the central suture arms through muscle tissue as shown in Fig. 10.6. I use a two-suture technique for recessions and resections, as I believe that is the only way to prevent the center of the muscle from sagging once the patient is awake and the muscle contracts. If you use a one-suture technique, which I realize is more popular than a two-suture technique, you can prevent suturing error by first making a small full-thickness (through and through) suture

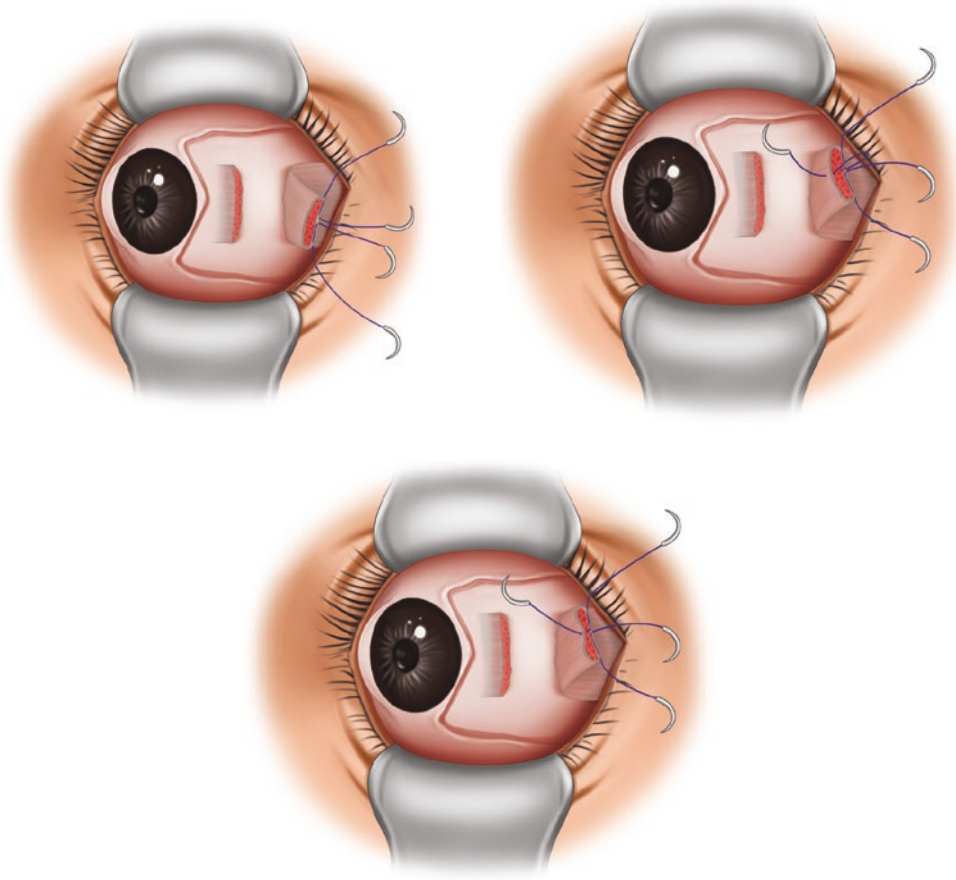


**Fig. 10.6** (**a**) Reflecting the muscle after disinsertion to expose the undersurface will reveal if the muscle is not secure. (**b**) If the muscle is not secure centrally, a suture pass is made to reinforce the center of the muscle. (**c**) When the suture is pulled up, the central muscle fibers are secured

pass at the center of the muscle, and securing that with a knot. Because slipped muscles are caused by faulty suturing technique, muscle slippage in the capsule is less likely to occur bilaterally (unless the same error was made on both sides). If a muscle had slipped in the capsule, you will find a thin, flaccid empty capsule attached to the sclera at surgery.

2. A stretched (or elongated) scar is not caused by faulty suturing. It presumably forms in patients who have an intrinsic wound-healing problem that predisposes them to developing a stretched scar [8]. This in turn causes a postoperative underaction with overcorrection after a recession or an undercorrection after a resection. Because it is due to an intrinsic healing problem, stretched scars often occur bilaterally. They typically do not appear clinically for months (and sometimes years) after surgery, unlike a slipped muscle, which more often occurs early after surgery. A stretched scar can be easy to overlook at surgery if one does not have a high index of suspicion that it may have occurred and

specifically look for it. The scar more closely resembles muscle tissue than does the empty capsule of a slipped muscle, but there is typically a distinct interface between the scar and actual muscle.

The treatment for both slipped muscles and stretched scars is to excise the abnormal tissue anterior to the edge of true muscle and suture viable muscle to the sclera. For a slipped muscle, you can use absorbable sutures, because with proper suturing the problem would not be expected to recur. However, with a stretched scar, nonabsorbable sutures are preferable, because recurrence is common due to the underlying healing problem. Consequently, it is important to know which of these two situations you are dealing with. Certainly, the history of onset, and bilaterality, can help in this regard. At surgery, distinguishing the two can be difficult. The gross appearance can be similar with both conditions. But if you run a muscle hook under and against the abnormal connective tissue, an empty capsule will have more translucency and is more flaccid than a stretched scar. In addition, in cases of stretched scar you will more often feel a "bump" as the hook passes by the interface of connective tissue and muscle.

When operating on a patient with either a stretched scar or a muscle slipped in the capsule, it is important to excise all of the connective tissue (scar or empty capsule) and be sure that you are suturing actual muscle to the sclera. It is wise to avoid a suspension (hang-back) technique in this setting or you run the risk of recurrent slippage.

## Question

Is there really a difference between slipped muscles and stretched scars? I thought one study suggested that they cannot be distinguished from one another and should be considered the same entity.


## Reply

After the initial description of stretched scars, I too was skeptical that this was just a variant of the slipped muscle. So while I was on the steep upslope of the learning curve for identifying these entities, I routinely sent the excised tissue to pathology for confirmation. Early on I was concerned that when I excised a lot of tissue (sometimes 15 mm or more), I wanted to be sure that I was not inadvertently resecting a large section of muscle. The histopathologic appearance of an empty capsule is different than that of a stretched scar, which contains much more fibrous connective tissue. As my experience (and confidence) grew, I was able to reliably assess at surgery whether I was facing a slipped muscle versus a stretched scar. However, this differentiation could not be made solely by the physical appearance. It was a confluence of history of onset, feel of the tissue, assessment of translucency, and sliding a hook along the surface to feel a "bump" at the interface of scar and muscle. The one study that attempted to differentiate these two entities relied solely on masked grading of operating room photographs [9]. I consider this inadequate for differentiating these two conditions.

## Basic Information

*Posterior Fixation Surgery*

The classic explanation for the effect of a posterior fixation suture is that it decreases the lever arm of the muscle increasingly as the eye moves into the muscle's field of action (Fig. 10.7) [10]. Scott has calculated that a posterior fixation on a MR 10 mm from the insertion will have essentially no effect on decreasing the torque until the eye is adducted 15° (about 30 Δ) [11]. Consequently, this procedure is most effective for balancing a duction deficit that causes incomitance, e.g., posterior fixation on the sound MR if there is an abduction lag in the contralateral eye.

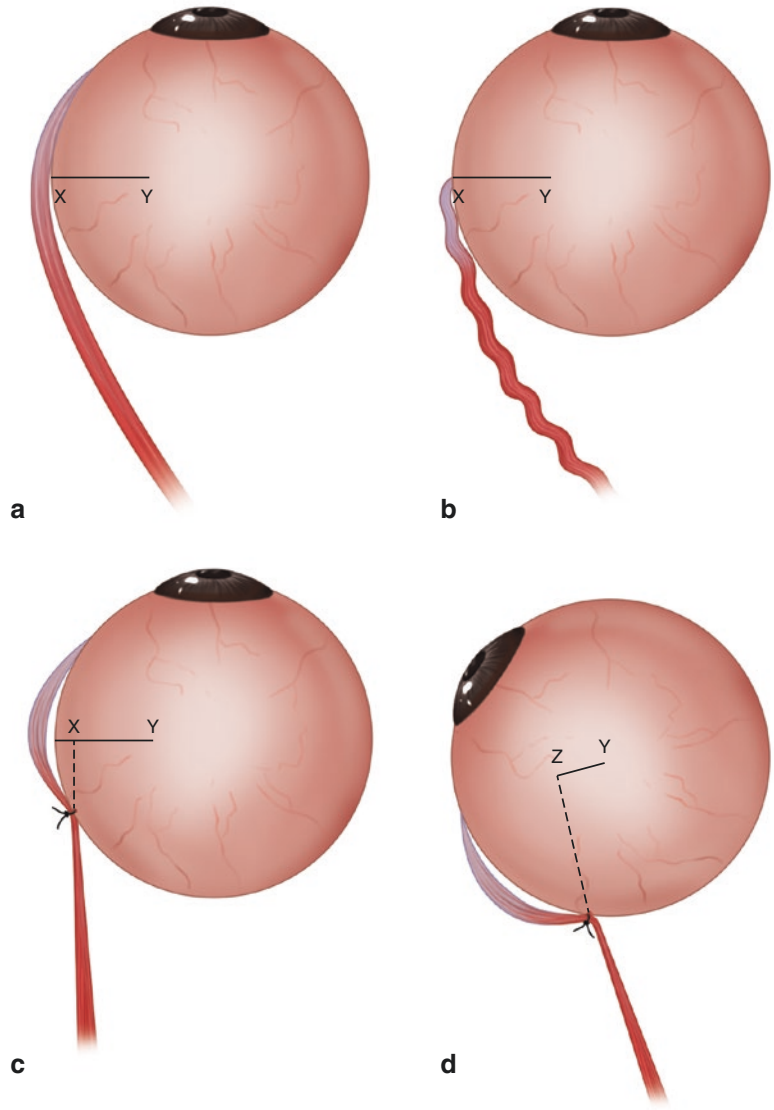



**Fig. 10.7** (**a**) The effective lever arm of a medial rectus muscle prior to recession runs from the center of rotation to the equator, lines *X–Y*. (**b**) If a recession is carried out, the lever arm does not change as long as the new insertion is not posterior to the equator. (**c**) With posterior fixation and the eye in the primary position, there is a negligible shortening of the lever arm. (**d**) After posterior fixation, as the eye moves into the field of action of the muscle, there is substantial shortening of the lever arm, lines *Y–Z*

## Advanced Information

*Posterior Fixation Surgery for High AC/A or Convergence Excess*

Based on the accepted concept of how posterior fixation surgery works, it is illogical to me to use it for treating a convergence excess pattern as discussed in Chap. 5. If the desired end result is good alignment in primary distance and near, the mechanism of action depicted in Fig. 10.7 should suggest that a posterior fixation suture will exert no effect if the eyes are aligned postoperatively. If a patient is orthophoric at distance, the amount they need to converge to be orthophoric at near is equal to their interpupillary distance (about 5–6 cm) times the dioptric

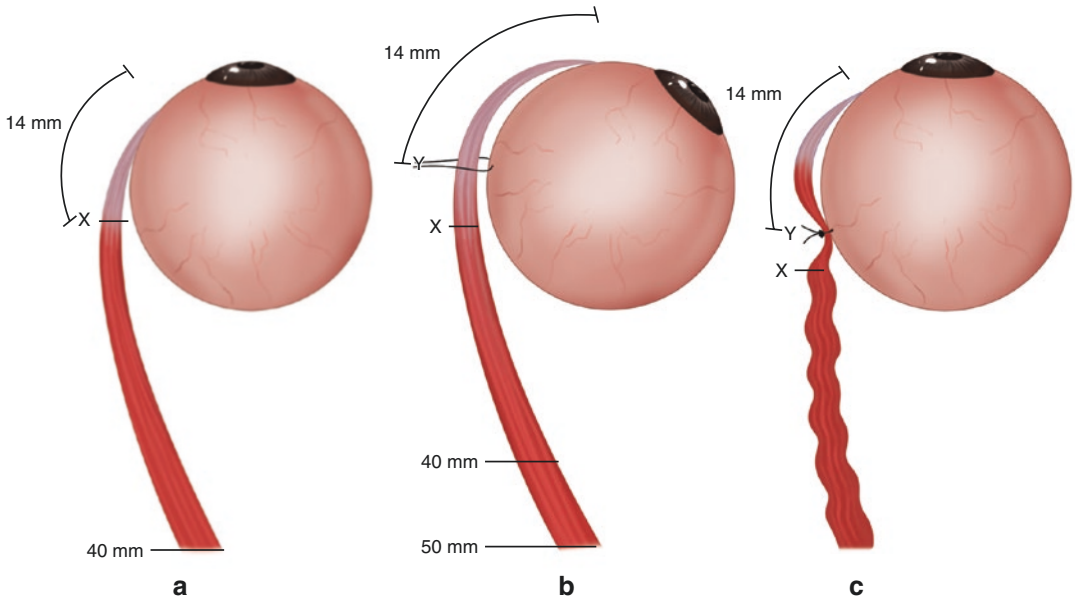



**Fig. 10.8** (**a**) If a medial rectus muscle (MR) is 40 mm in length, point *X*, which is 14 mm posterior to the insertion, is 14/40 times the length of the muscle posterior to the insertion. (**b**) In order to place the posterior fixation suture, the globe is rotated laterally, stretching the MR to 50 mm. Point *Y* is now 14 mm posterior to the insertion and is 14/50 times the length of the muscle posterior to the insertion. The muscle fibers at point *X* now lie posterior to point *Y*. (**c**) If the posterior fixation suture fixated point *Y* to the sclera 14 mm posterior to the insertion, slack is created in the muscle fibers posterior to the posterior fixation suture when the globe is rotated into the primary position

equivalent of the near testing distance (about 3) for a total of 15–18 Δ. Splitting this equally, it comes to about 7.5–9 Δ per eye or about 3.75–4.5°. Scott's calculations and logic would dictate that the posterior fixation suture is not contributing anything. Nevertheless, good results have been reported with posterior fixation surgery for convergence excess or high AC/A ET [10, 12]. Clearly, there must be another mechanism of action at play. One possible mechanism by which posterior fixation sutures can induce slack in a muscle, even when the eye is in the primary position, is depicted in Fig. 10.8 [10]. This mechanism probably accounts for the effect of posterior fixation sutures without recession on the primary position, and also explains why posterior fixation increases the effect of a simultaneous recession. In my experience, posterior fixation plus recession has more of an effect than standard formula recession for high AC/A ET, but is also more variable [13]. The mechanism shown in Fig. 10.8 not only explains why posterior fixation sutures increase the surgical effect, but it also explains why the response is variable. Even if the muscle is not recessed, the stretching of the muscle needed for exposure to place the posterior fixation suture results in selective shortening of the muscle posterior to the posterior fixation suture; the amount of stretch needed for exposure is a variable that is hard to control.

Posterior fixation will decrease a distance/near difference but unreliably.

## Advanced Information

*The "Adjustable Faden"*

Scott described an innovative modification of the traditional posterior fixation operation that permits postoperative adjustment [14]. By resecting a given amount of an extraocular muscle, and then recessing it an equal amount on an adjustable suture, one is in effect creating a new insertion posteriorly without shortening the sarcomeres in the remaining muscle (Fig. 10.9). This is akin to placing a posterior fixation suture without recessing the muscle. If one wants to have some recession effect because of a deviation in primary, one can recess more than the amount that is resected, with suggested ratios varying from recessing 50% more than the resection, to double that amount [14–16]. This procedure is very effective in balancing an eccentric gaze duction deficit, and for unexplained reasons (much to my surprise) does not predispose to muscle slippage when done on the inferior rectus muscle (IR) [15, 16]. However, there is something perplexing about this procedure that has not been adequately explained. It appears to be effective with recession amounts of as little as 4–6 mm. Yet we know that if a standard posterior fixation suture is placed only 4–6 mm posterior to the muscle's insertion, it has no effect. So I suspect that another mechanism is operating. Christiansen and coworkers reported that tenotomy alone of an extraocular muscle causes modulations in fiber remodeling and myosin expression [17]. Perhaps this contributes to the effect of the "adjustable faden" procedure. See Chap. 21, Case 21.9 on page 327 and Case 21.12 on page 319, for representative examples of these principles.

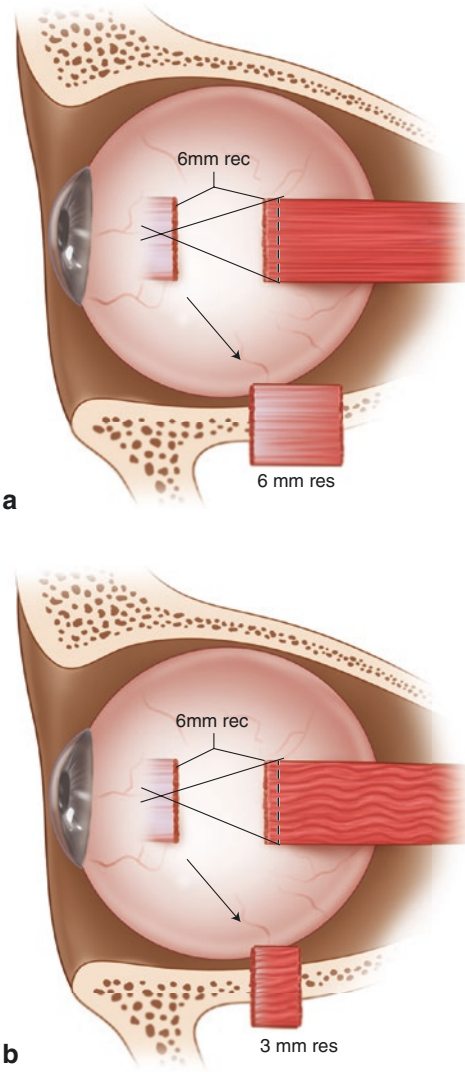



**Fig. 10.9** Scott's "adjustable faden" operation. (**a**) To have no effect on the primary position, the amount of recession equals the amount of resection (6 mm), and no shortening occurs to the sarcomeres in the muscle. (**b**) To get an effect on the primary position, the recession (6 mm) exceeds the resection (3 mm), thus creating some shortening of the sarcomeres in the muscle

## Advanced Information

*Importance of Eye Alignment Under Anesthesia*

Apt and Isenberg determined that patients with routine horizontal strabismus are approximately 30 Δ more exotropic or less esotropic under anesthesia [18]. Subsequently, some investigators have suggested that surgery should be modified if a patient deviates from these norms [19–22]. Romano proposed that if an exotropic patient is less exotropic than would be predicted by the Apt and Isenberg data, less than the routine amount of surgery

should be performed. If an esotropic patient has less of an exotropic shift under anesthesia than predicted, more than standard surgery should be performed. The converse reasoning would apply if a patient had a greater exotropic shift under anesthesia than was predicted. My own experience contradicts this approach. Over a period of about 25 years I routinely assessed and recorded eye position under anesthesia, but did not alter my surgery. For both routine esotropic and exotropic patients the dose–response curves I calculated in those patients who deviated from the Apt and Isenberg predictions, yet had standard surgery, did not differ from the dose–response curves for those patients who showed the predicted alignment under anesthesia.

### Important Point

The aforementioned discussion about the importance of alignment under anesthesia relates only to routine strabismus and does not relate to cases with restriction or paresis, where spring-back testing as described below and position under anesthesia may be important [23].

### Question

What intraoperative testing is important?


### Reply

For patients with normal ductions, our surgery tends to be formula driven, e.g., a given number of millimeters per prism diopter of deviation. Perhaps in the future our approach will take into account other factors such as muscle elasticity. But as for what is currently prime time, intraoperative testing will probably not alter our surgical approach in routine patients. However, if there is a limitation of rotation, intraoperative forced ductions are crucial, and they need to be repeated throughout the procedure at each critical juncture, e.g., after muscle disinsertion and after reattachment. These findings need to be coupled with findings from active force generation testing, which of course needs to be done with the patient in the awake state. In cases of restrictive strabismus, or simple reoperation, the spring-back test as described by Jampolsky is important [23]. To do this test, the eye is grasped at the limbus with forceps and rotated back and forth from adduction to abduction (for testing a horizontal muscle) 6–8 times, then held in either abduction or adduction, and released. Note where the eye comes to rest. Then repeat the maneuver, holding it in the opposite gaze field. The two resting points should be symmetrically and equally spread around primary when the restrictive forces are balanced. Note, however, that this test in isolation should not be used to determine the final amount of muscle recession or resection that is needed, because it does not take into account innervational factors. For example, if you do this test on a patient immediately after the onset of an acute sixth nerve palsy (before MR contracture occurs), the results will probably show normal centration. Yet, obviously, surgery would still be needed to restore alignment due to the severe innervational imbalance. Yet if done in that same patient months later (after the MR contractures), the results would be abnormal. So although this test is very useful for determining when restrictions are relieved, its findings are intended to be only part of the answer to the puzzle.

Guyton's exaggerated traction test is invaluable for assessing tightness of the oblique muscles [24]. To test for the inferior oblique muscle, my preferred technique is to grasp the eye at the 4:30 o'clock limbus (for the right eye) and push the forceps and limbus into the slightly inferior nasal quadrant. The forceps and limbus are then rolled toward the 6 o'clock position. The examiner can feel the globe rotate over the inferior oblique muscle and its tightness assessed. A mirror image approach is used superiorly for the superior oblique muscle (SO).

I find that torsional restrictions can be felt using rotary forced ductions. The eye is grasped at the limbus at 6 and 12 o'clock, and the eye rotated clockwise and counterclockwise until

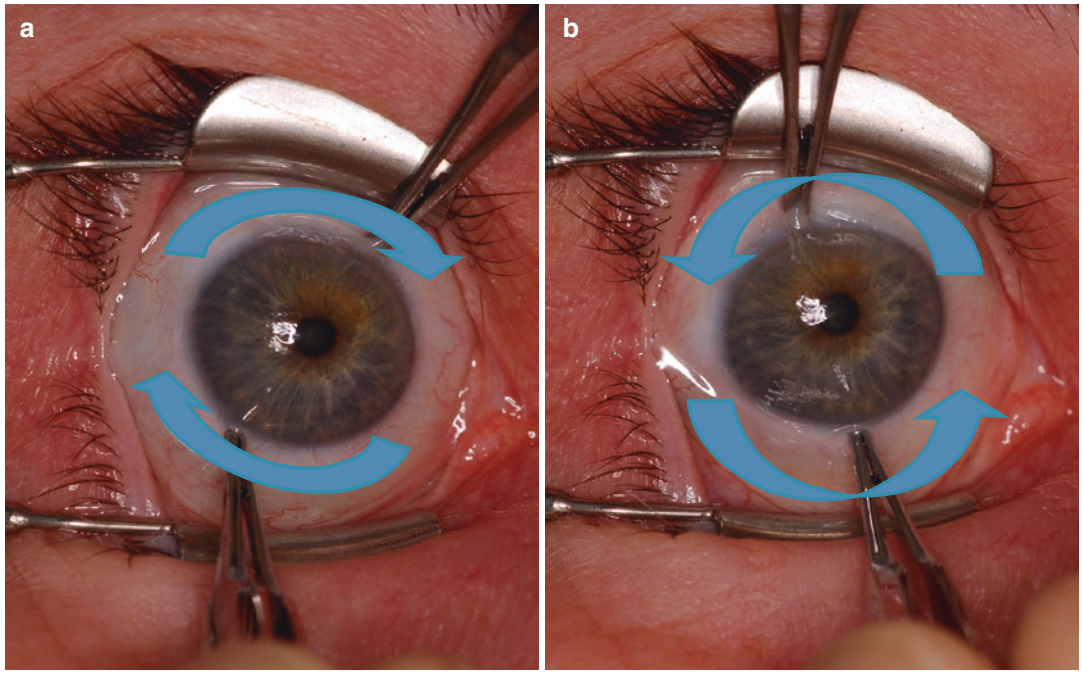



**Fig. 10.10** View of a right eye as viewed by the examiner facing the patient (superior limbus is near the top of the photograph). The photograph demonstrates the technique for rotary passive ductions to test for a torsional restriction. (**a**) The eye was initially grasped with forceps at the 12 and 6 o'clock positions and rotated clockwise (incyclorotation, *arrows*). The eye can be rotated approximately 40° clockwise until resistance is felt. (**b**) Without removing the forceps, the eye is then rotated counterclockwise (excyclorotation, *arrows*). It can be rotated only about 15° counterclockwise until the same amount of resistance is felt. This confirms a torsional restriction to excyclorotation

resistance is felt as shown in Fig. 10.10 [25]. Look for symmetry. I have not found it necessary to quantify the rotation with an actual scale graded in degrees. It is easy to assess symmetry by just looking. Alternatively, the forceps can be placed at the 3 and 9 o'clock position. See Chap. 21, Case 21.12 on page 319, for a representative example of these principles.

## Question

When do you worry about anterior segment ischemia (ASI), and how can it be prevented?


## Reply

The risk of ASI is inversely related to the patient's age and the status of his or her cardiovascular system. Although I am always fearful and hesitant, I think you can safely operate on three rectus muscles in the same eye, during the same sitting, in a healthy child. I have never done four rectus muscles at the same time, but I know some people have done so without problems. In adults, and certainly in smokers or those with compromised circulation, I do not operate on more than two rectus muscles in the same eye at one time. The risk increases when circulation through the muscles is compromised as in thyroid eye disease, and if one does two adjacent muscles, particularly the IR and MRs, which commonly needs to be done in thyroid eye disease patients. In theory, plication of a rectus muscle, instead of a resection, may spare some of the circulation. Also, there is evidence that a limbus incision, which disrupts the peri-limbal scleral blood supply, is a contributing factor to ASI. My preferred incision for the superior and inferior rectus muscles is to open conjunctiva directly over the muscle insertion, which is a modified Swan approach. This provides the best exposure and spares the peri-limbal circulation. I use it

routinely for vertical rectus surgery, regardless of whether I consider the patient at particular risk for ASI. Although I do not use that approach routinely for horizontal rectus muscles because it can leave a visible scar, with the vertical rectus muscles any scar is well hidden by the eyelids. Try it, you'll like it! If your surgical plan involves horizontal rectus muscle surgery in a patient you consider at risk for ASI, and if you are not accustomed to using a fornix incision, you can modify the Swan incision by placing it just anterior to (almost touching) the semilunar fold for the MR. For the LR, you can adduct the eye prior to incising, making the incision far enough laterally that it will be hidden by the lateral canthus when the eye is in primary. These will make for scars that are essentially invisible. Of course, you can always plan to use a vessel-sparing technique [26]. This procedure is discussed in detail later in this chapter.

> Doctor: "I will do the surgery one day, and then next day
> if the alignment is not what we want,
> I can do an adjustment."
> Patient: "I'm impressed. I never met a surgeon who believed in chiropractic."

## Basic Information

*Adjustable Sutures*

I use adjustable sutures routinely in almost all cases of adult strabismus involving rectus muscle surgery. In most routine cases, I will only put one horizontal and/or one vertical rectus muscle on an adjustable suture, to fine-tune the alignment. If more than one muscle is pathologic, e.g., involving thyroid eye disease, I may in select cases use adjustable sutures on more than one horizontal or vertical rectus muscle. I strongly prefer to put the adjustable suture on a recessed muscle, because I think resections are more variable by nature postoperatively, as they tend to stretch out. I have no hesitation about using adjustable sutures in both eyes, or on both a vertical and horizontal recuts muscle in the same eye. My personal preference is to do the adjustment on the morning after surgery in my office. I have not been drawn to adjusting in the operating room, because I strongly think that vision blur from the surgical manipulation and corneal surface issue make it difficult to control accommodation.

## Advanced Information

*Adjustable Suture Technique*

Of the two ways I use to secure the muscle on an adjustable suture (sliding noose and bow knot), I find pros and cons to each. Like so much in life, this is a matter of trade-offs. The sliding noose is easier to adjust but leaves a more bulky knot, which can predispose to a persistent lump over the insertion. The bow knot obviates the problem of the subconjunctival lump and looks better aesthetically, but it is more "fussy" to adjust than the sliding noose. In the end, I find myself defaulting to the bow knot unless I have a patient for whom I think the adjustment itself might be problematic with respect to cooperation. For those, I am more inclined to use the sliding noose.

Early in my experience with adjustable sutures, I left the arms of the adjustable suture long and taped them to the skin near the eye until I did the adjustment. More recently, I tuck the entire set of suture arms (several centimeters long) under the conjunctiva, leaving just a tiny loop that I can grasp and pull out at the time of adjustment. If I use an inferior fornix incision, I suture a long loop of vicryl through the top of the insertion that I can use to retract the conjunctiva superiorly to access the insertion at the time of adjustment (Fig. 10.11). If I use a limbus incision and recess conjunctiva, I make a point of recessing the conjunctiva to a point several millimeters anterior to the insertion so that the final knot can be tucked under the conjunctival edge (Fig. 10.12). When doing so, I find it important to include a small bite of episclera in the suture pass that connects the corner of the conjunctival flap to adjacent conjunctiva in order to prevent the conjunctiva from sagging posteriorly and exposing the insertion. If I use a limbus incision and do not recess conjunctiva, I put a long loop of vicryl in one corner of the flap to permit delayed closure after adjustment (Fig. 10.13).

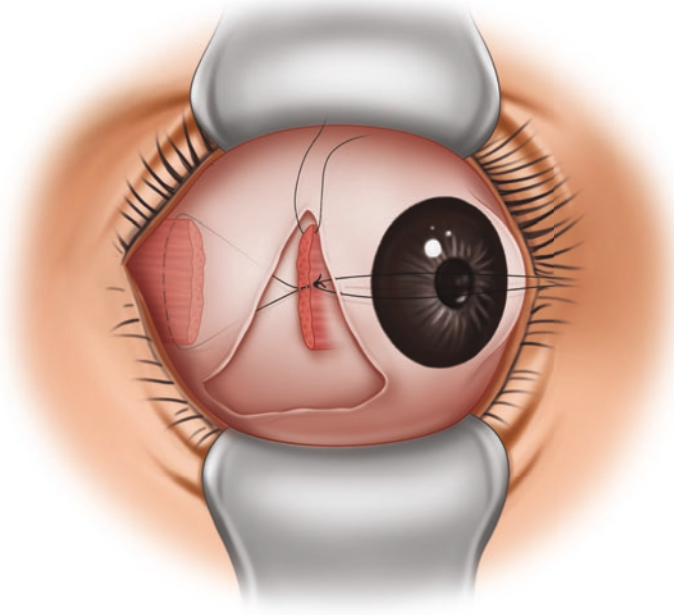


**Fig. 10.11** With an inferior fornix incision, a loop of 6–0 vicryl is sewn through the superior pole of the insertion. At the time of adjustment this can be used to retract the conjunctiva, exposing the sliding noose. After adjustment the readjustment suture is removed. The arms of suture holding the muscle are tucked under conjunctiva at the end of the procedure. Retracting the conjunctiva makes them readily accessible

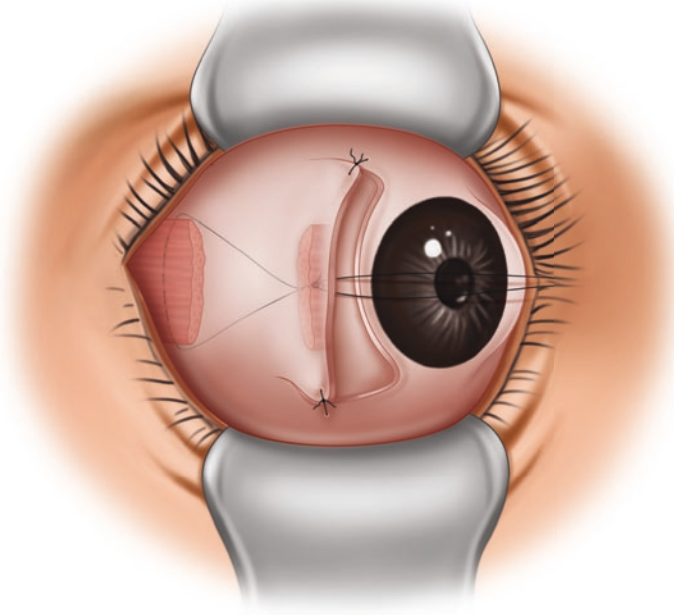


**Fig. 10.12** With a limbus incision and conjunctival recession, the conjunctiva is recessed just anterior to the insertion. This permits the knot to be covered after adjustment. It helps to stretch the anterior edge of conjunctiva taut vertically prior to suturing, and to pass the needle through episclera, to prevent sagging

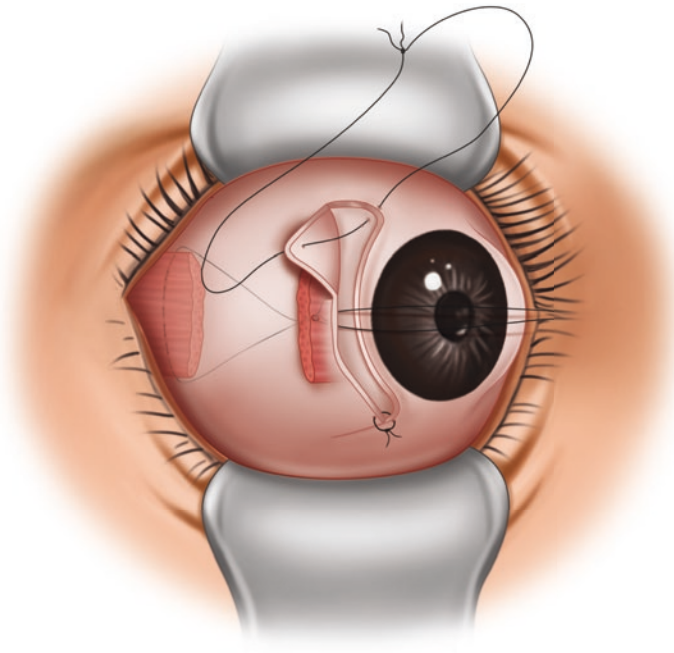


**Fig. 10.13** With a limbus incision and no conjunctival recession, one corner of the conjunctival flap is closed anatomically at the end of the procedure. The other is secured with a large preplaced loop to permit delayed closure

## Advanced Information

*Suspension (Or Hang-Back) Surgery Versus Fixed Scleral Sutures*

There are those who prefer to use a suspension technique for all strabismus surgery, including routine cases. The advantage of this is that it is safer, in that one is suturing to the insertion stump, and an inadvertent perforation so far anterior will not be through the retina. I prefer fixed scleral sutures to a suspension technique because they result in a more solid bond to the sclera, and in theory should decrease the incidence of muscle slippage or forward creep. Interestingly, the only histopathological study to look at this issue found that in 25% of 16 muscles recessed using a suspension technique, the muscle was found to adhere to the globe by a pseudotendon [27]. These findings were of sufficient concern to me that I am reluctant to use a suspension technique routinely. In cases in which I feel an adjustable suture would be helpful, I think the ability to adjust the alignment postoperatively outweighs the increased risk of muscle slippage. This is one reason, however, that I do not use adjustable sutures in routine cases of strabismus in children.

A suspension technique produces a less firm bond with the sclera.

## Pearl

If you use an adjustable suture and want to know where the muscle ends up after adjustment, the use of a reference knot can help. After the muscle is secured in surgery at the desired position on an adjustable suture, tie the two arms of the suture together some arbitrary distance from the cinch and measure that

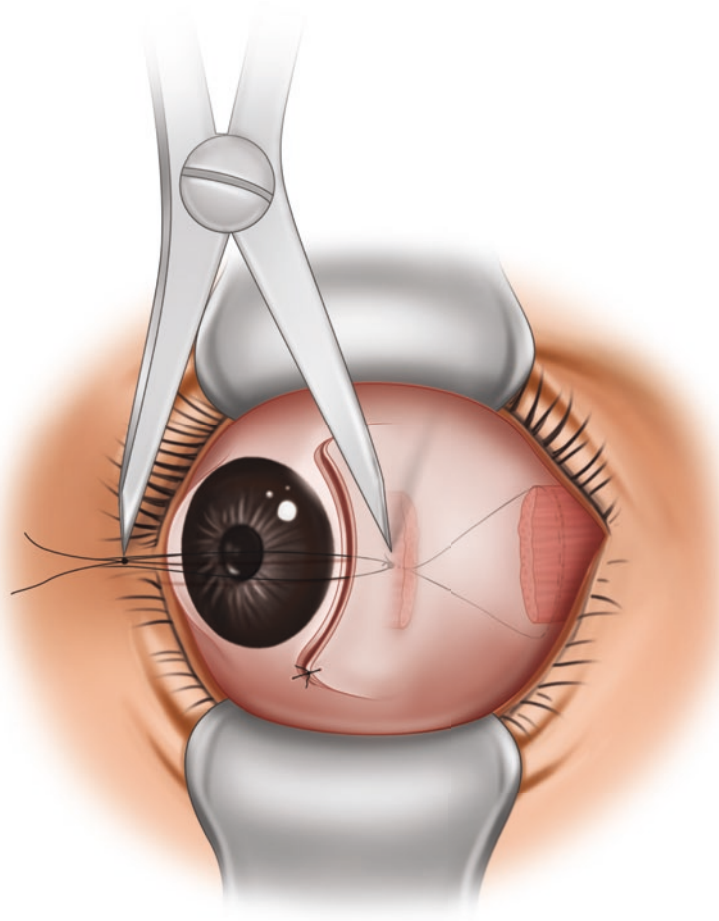


**Fig. 10.14** The two arms of the suture holding the muscle are tied together at an arbitrary distance from the cinch. The distance between the cinch and the knot is measured as "X." Comparing this initial distance to the cinch-knot distance after adjustment will allow for calculating the final position of the muscle

distance. At the time of postoperative adjustment, remeasure that distance. If the muscle did not slip, it will be the same just prior to adjustment as it was in the operating room. You can measure it again after adjustment. By subtracting the postoperative measured knot-cinch distance from the intraoperative cinch-knot distance, you can determine how much the muscle was moved at adjustment (Fig. 10.14). If you use a bow-knot technique, you can simply put a reference knot in one arm of the long suture holding the muscle and use it as a reference point for measuring.

## Basic Information

*The Semi-adjustable Suture*

A nice modification of the adjustable suture procedure is the semi-adjustable technique suggested by Spielmann [19, 28]. This procedure involves fixing the corners of the muscle directly to the sclera, and suspending the middle of the muscle on an adjustable suture (Fig. 10.15). It has the advantage of minimizing the problem of nonadherence, particularly with IR

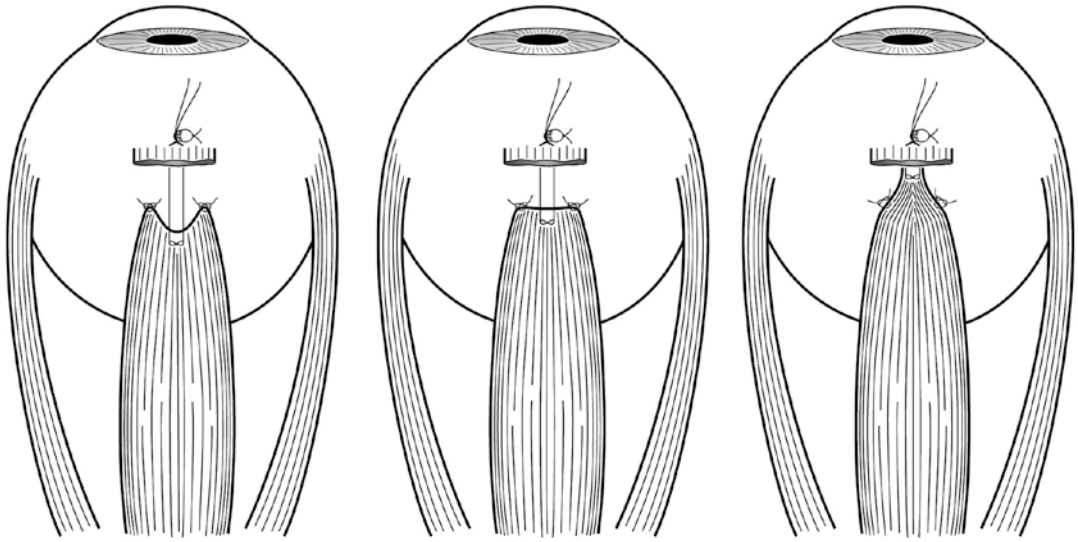


**Fig. 10.15** The semi-adjustable recession: (**a**) The corners of the muscle are sutured to the sclera so as to bunch the center somewhat, making advancement easier. (**b**) The center of the muscle is secured with a standard adjustable suture, and the center pulled up to eliminate any sag. (**c**) At the time of adjustment the center of the muscle can be advanced as far as to the original insertion

recessions, and it keeps the muscle from creeping forward. The main disadvantage is that, although you can essentially completely undo a recession and the time of adjustment, there is little ability to increase the recession. This can be obviated by increasing the amount of recession by a millimeter or two at the time of surgery, which minimizes the need for increasing the recession at the time of postoperative adjustment.

## Important Point

If you do a recession on a suspension, including use of a standard adjustable suture technique, and want to also transpose, e.g., to treat a pattern or create a small force vector in the direction of the transposition, you will not get the desired transposition effect. If the muscle is suspended from the insertion, it will "gravitate" toward the shortest path and return toward the non-transposed position (Fig. 10.16). This can be obviated, however, by using the semi-adjustable technique, which will keep the muscle in the transposed position [29].

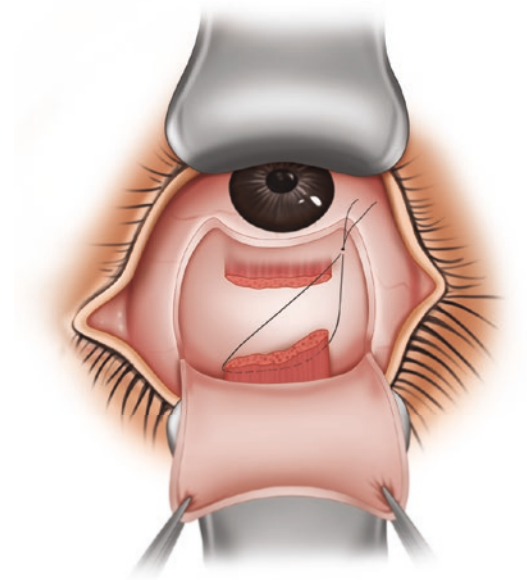



**Fig. 10.16** The right (R) inferior rectus muscle (IR) is recessed on a suspension. The sutures are passed through the nasal pole of the RIR insertion with the intent of nasally transposing the muscle. However, the muscle will take up slack by assuming the shortest wrap-around course on the globe, and will gravitate up to its original orientation. This will negate the intended infraplacement

## Basic Information

*Full Rectus Muscle Transposition to Treat a Duction Deficit*

As depicted in Fig. 7.5, transposing a rectus muscle creates a force vector in the direction the muscle is moved. Thus for an abduction deficit as seen in sixth cranial nerve palsy or *Duane syndrome*, the superior and inferior rectus muscles can be transposed to the LR to create a force vector for abduction. When performing this type of transposition, many people like to orient the new insertions of the vertical rectus muscles so that they are parallel to the fibers of the LR, as seen in Fig. 10.17a. This works well and for many years was my preferred approach. However, since the advent of lateral fixation augmentation sutures, as described by Foster [30], most people (myself included) orient the transposed muscles along the spiral of Tillaux, as shown in Fig. 10.17b, because that orientation allows for easier tying of the posterior augmentation sutures. Foster augmentation sutures involve tying the fibers of the vertical rectus muscles to the superior and inferior edge of

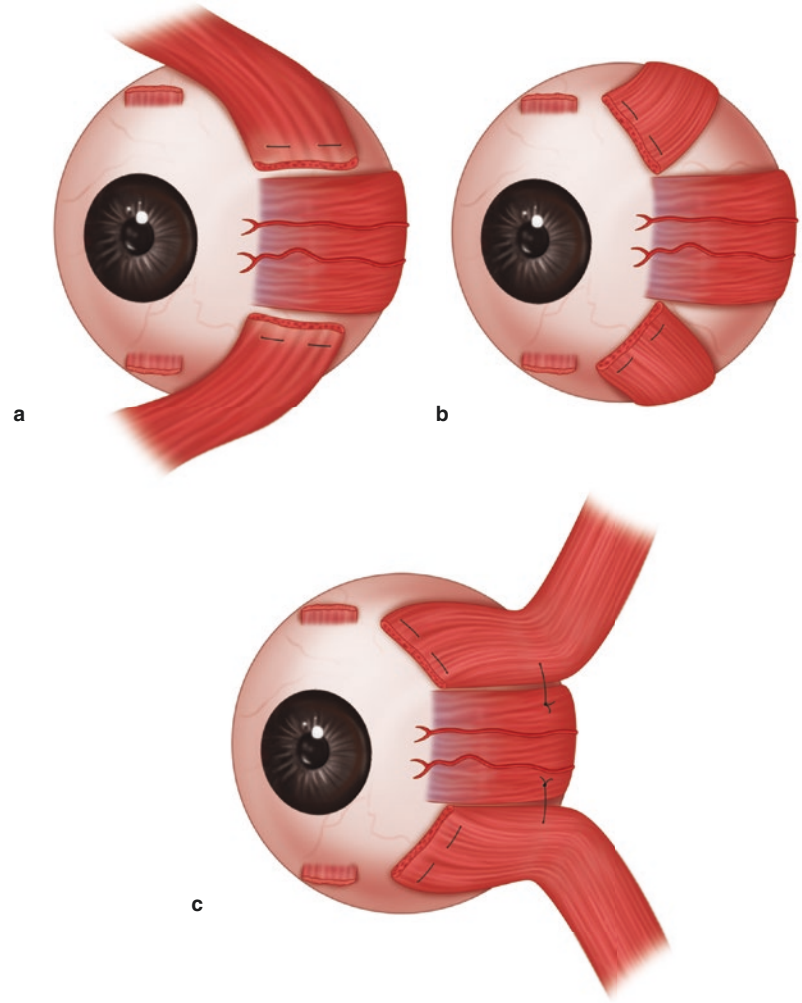



**Fig. 10.17** (**a**) Left eye showing vertical rectus transposition to the lateral rectus muscle (LR) with orientation of the new insertions of the vertical rectus muscles parallel to the LR. (**b**) Left eye showing orienting the new insertions of the transposed muscles along the spiral of Tillaux. (**c**) Left eye showing orienting the new insertions of the transposed muscles along the spiral of Tillaux vertical rectus transposition with lateral fixation augmentation

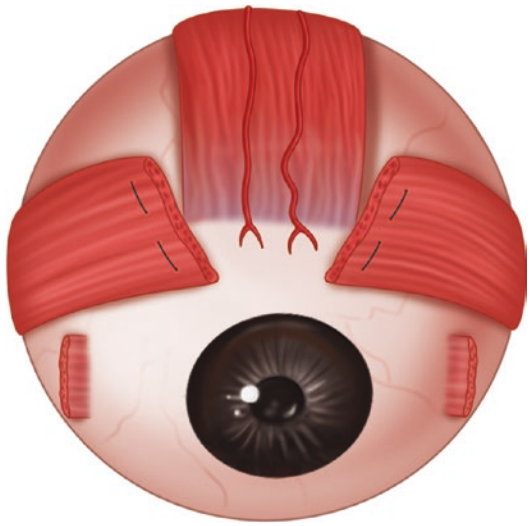


**Fig. 10.18** Orientation of the horizontal rectus muscles as originally described by Knapp for treating monocular elevation deficiency

the LR (about one-fourth width of the muscles), about 16 mm posterior to the limbus with a nonabsorbable suture (Fig. 10.17c). The suture is not put in the sclera. This is a powerful augmentation of the full tendon transposition procedure. Whenever you transpose the superior rectus muscle (SR), you should take care to separate the SO frenulum to prevent the SO from being dragged laterally [31].

The Knapp procedure utilizes similar principles of transposition for treating an SR palsy or monocular elevation deficiency by transposing the horizontal rectus muscles to the SR [32]. Fig. 10.18 depicts the way of orienting the transposed muscles as originally described by Knapp. Most people now either orient them with their new insertions parallel to the SR fibers, as is seen in Fig. 10.17a for a horizontal transposition, or along the spiral or Tillaux, as seen in Fig. 10.17b for a horizontal transposition. Although there are no data indicating that one technique is preferable to the others, orienting the muscles along the spiral of Tillaux as in Fig. 10.17b is optimum if Foster augmentation sutures are used. An inverse Knapp procedure (transposing the MR and LR to the IR) can be used to treat a depressor palsy.

## Advanced Information

*A Partial Rectus Muscle Transposition to Treat a Duction Deficit*

You can get quite a powerful effect by transposing one-half or three-fourths of opposing rectus muscles to treat a duction deficit, particularly if you add augmentation sutures. This is particularly useful when there is a high degree of concern about anterior segment ischemia. For example, to treat a sixth cranial nerve palsy you can transpose the temporal one-half or three-fourths of the vertical rectus muscles to the LR. You should take special care to identify the nasal anterior ciliary arteries in the transposed muscles and leave them undisturbed. Their location can guide you as to whether to transpose one-half or three-fourths of the muscle. When doing this, it is important to cut the check ligaments along the nasal border of the vertical rectus muscles, which will allow the intact nasal fibers to be dragged laterally.

> The idea seems absurd, but I can find no flaw in it.
> —Johannes Kepler

## Advanced Information

*Transposing Just the SR to Treat a Duction Deficit With or Without MR Recession*

At an American Association for Pediatric Ophthalmology and Strabismus Annual Scientific Meetings Earl Crouch presented a series of patients with Duane syndrome in whom he only transposed the SR to the LR with Foster augmentation sutures, and left the IR intact. Many people, myself included, were surprised at the good results he reported, because intuitively it seemed that this procedure should create a vertical imbalance as well as marked intorsion. Subsequently, I know many colleagues who have used it with good results, and large series have now been published confirming its efficacy and relative lack of

complications in patients with Duane syndrome and sixth cranial nerve palsy [33–35]. The advantages of this procedure, in addition to being easier and quicker than transposing both vertical rectus muscles, is that it spares anterior circulation even if the MR is simultaneously recessed. I currently consider this to be the procedure of choice for treating complete abduction deficits.

I consider SR transposition with or without ipsilateral MR recession to be the treatment of choice for complete abduction deficit.

## Question

How is it possible that full transposition of the SR with Foster augmentation does not cause a marked incyclotropia? After all, one can get a significant intorsional rotation by simply transposing the SR 7 mm temporally. Transposition to the LR with Foster augmentation should be more powerful in this regard.


## Reply

Everything we know about muscle forces and vectors suggests that this procedure should cause intorsion. But the facts are that it does not. I can, however, speculate on why it does not. In the largest series published on this procedure [33–35], only one patient developed intorsion. And that one patient was unique in that he was the only one in whom the surgeon secured the Foster augmentation suture to the sclera. I speculate that the transposed SR does in fact exert a force vector for intorsion (it must). If the muscle is fixated to the sclera, that force is transmitted to the globe resulting in intorsion. If the SR is merely connected to the LR by the augmentation suture, the intorsional force vector will tend to pull the LR toward the SR. That in turn will put the LR on stretch, creating an extorsional force vector. The two opposing torsional vectors might cancel one another.

## Advanced Information

*Jenson Procedure*

The Jenson procedure involves splitting the SR, IR, and LR longitudinally at the insertion, and then slinging the temporal halves of the vertical rectus muscles to the adjacent halves of the halves of the LR near the equator with a nonabsorbable suture without disinserting any of the muscles. Part of the original rationale behind this procedure was that it would maintain anterior segment circulation because the muscles were not disinserted, and, thus allow for simultaneous recession of the MR with less fear of anterior segment ischemia. Although it is unlikely that blood can flow through the transposed slips of muscle, as the sutures securing the muscle slips are under considerable tension, the procedure does involve leaving the nasal halves of the SR and IR undisturbed, and thus may preserve anterior segment circulation. Although there are people still describing good results with the Jenson procedure, I abandoned it many years ago for several reasons. I felt that the full muscle transposition with Foster augmentation was more effective. But moreover, reoperation, if needed, was far more difficult after a Jenson procedure. In many cases the transposed slips of muscle had atrophied and turned to scar tissue. Also, although at the end of the procedure the adjacent halves of the vertical rectus muscles and the respective LR were sitting in the superior and inferior temporal quadrant, if the LR was really flaccid, the configuration of the transposition changed over time. At reoperation I observed that the halves of the LRs stretched out and let the halves of the vertical rectus muscles simply drift back to their original orientation. In effect this ended up to be just a tightening procedure of the LR. This migration

can be prevented by securing the suture holding the muscle halves to the sclera in the inferior and superior temporal quadrant, although this modification is not part of the procedure as originally described.

## Advanced Information

*Small Horizontal Rectus Transpositions to Treat Small Vertical Deviations*

Utilizing the principle that a force vector is created in the direction a muscle is transposed, small vertical deviations accompanying horizontal strabismus can be treated with small offsets of the horizontal rectus muscles. For example, consider that there is a small R hypotropia accompanying an ET. If you plan to recess the MR OU, raising the right MR and lowering the left MR several millimeters can be effective in treating the hypertropia. If you are doing a recess/resect procedure, you could raise the MR and LR if you are operating OD, or lower both if operating OS. In general, I limit this approach to treating vertical deviations of less than about 8 Δ; if the hypertropia is greater, I usually add surgery on a vertical muscle instead of offsetting the horizontal rectus muscles. Although authors have published monograms as to how much effect you get per millimeter of surgery, I find them to be unhelpful. They assume that the affect is linear, e.g., *X* Δ per millimeter. This is misleading. If you imagine the change in the magnitude of the force vector as you supraplace or infraplace a horizontal rectus muscle, there will be essentially no vertical vector created for the first millimeter, a bit more for 2 mm, and it is not until you have transposed about 3 mm that the effect starts being meaningful. I typically transpose a minimum of 3 mm for up to 4–5 Δ and about 4 mm for deviations of 5–8 Δ.

Similar principles apply for horizontal transposition of the vertical rectus muscles to treat small horizontal deviations. However, vertical rectus muscle transposition will have a greater effect on torsion than horizontal rectus muscle transpositions vertically.

The vertical effect of vertical transposition of horizontal rectus muscles is not linear.

## Advanced Information

*A Potpourri of Lesser Known Surgical Procedures*

### Plication

In the years shortly prior to this publication, there has been a renewed interest in performing plications as an alternative to resections for tightening muscles. Studies have shown that plications are less likely to disrupt anterior segment circulation [36], and they can be performed using an adjustable technique [37]. Millimeter for millimeter, they are essentially interchangeable with resections. A variation of the plication procedure has been popular in Mexico since the early 1980s. This procedure, called a linear plication by Romero Apis, and affectionately called a "Mexican tuck" by Jampolsky (Arthur Jampolsky, personal communication, 24 Sep 2012), involves splitting the center muscle longitudinally with a hook at the insertion, placing a suture at the apex of the split, and then bringing the suture through the insertion. When the suture is tied, the center of the muscle is plicated (Fig. 10.19). It seems logical that a "Mexican tuck" preserves more of the anterior segment circulation than a standard plication, but may be more likely to result in a bulky mass under the conjunctiva; there are no data to support this speculation. I do not have experience with this procedure. It seems logical that this method of plication has more advantages with respect to preserving anterior segment blood supply than a traditional plication. More recently, Leenheer and Wright described a minor modification of this procedure by performing it as an office procedure under topical anesthesia, if limited to very small amounts of plication [38].

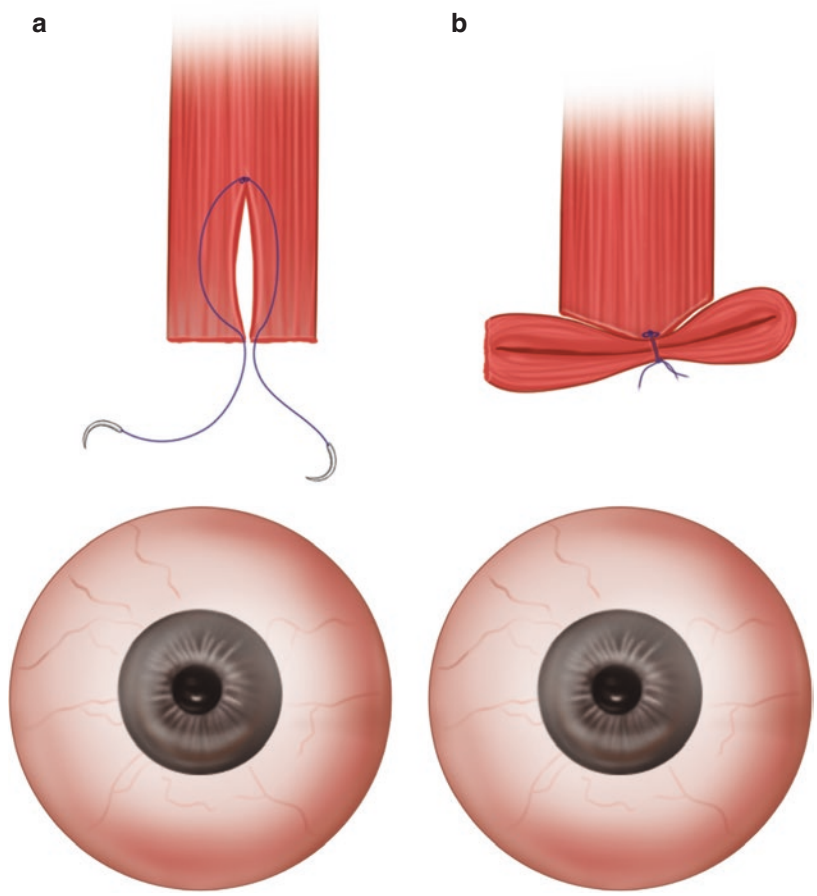



**Fig. 10.19** The "Mexican tuck." (**a**) The muscle is split centrally from the insertion to a distance posteriorly equal to the desired size of the plication. A suture is tied in the muscle at the apex of the split and the arms brought through the insertion. (**b**) The suture is tied, thus placating the muscle

## Pulley Fixation

Based on the hypothesis that the posterior fixation operation works by causing a mechanical restriction, rather than the traditional concept of decreasing the lever arm of the muscle, Clark proposed fixating the muscle belly to the pulley as an alternative to the classic posterior fixation procedure [39, 40]. This procedure has the advantage of being more likely to be reversible than the classic posterior fixation operation. I personally think it may work via a different mechanism than the classic posterior fixation, as evidenced by the fact that the aforementioned "adjustable faden" described by Scott has a similar effect as a standard posterior fixation, but does not create the restriction Clark suggested occurs with standard posterior fixation.

When planning vessel sparing surgery, always have a back-up plan in case the vessels are traumatized.

## Ciliary Vessel Sparing

McKeown described a technique for sparing the anterior ciliary vessels during recession or resection surgery [26]. This involves carefully

dissecting the vessels free from the muscle prior to disinsertion and taking care to keep them intact during disinsertion and suturing. It is a technically demanding procedure and, particularly if you do not have a lot of experience with it, is something you cannot always count on doing successfully. It is wise to always have a backup plan in case you inadvertently traumatize the vessels. In some cases of reoperation, it may not be technically feasible to spare the vessels due to prior scarring. I limit my use of vessel sparing to those patients at high risk for ASI, and in whom my surgical plan involves operating on a third rectus muscle in one or both eyes. I found the learning curve to be steep for this procedure, but after my first five cases (or so), I have not had to resort to my backup plan. I have not been successful in sparing vessels in previously operated muscles.

### Suturing Muscle to the Orbital Periosteum

When you want to completely disable a muscle's function, sewing it to the orbital periosteum is the most effective means [41, 42]. The most common situations in which this is useful are the disabling of the LR in Duane syndrome with severe retraction and in third cranial nerve palsy. Technically, this is not difficult, but when being done for the first time, it might have the strabismus surgeon outside of their usual comfort zone. You will need to cut down on the periosteum using a scalpel blade, or something similar, several millimeters posterior to the orbital rim and pass the suture firmly in periosteum. It is advisable to secure the muscle with a colored nonabsorbable suture such as blue prolene. This makes reversing the procedure easier should that be necessary. The one procedure I was involved with that involved reversing periosteal fixation was greatly facilitated by the visible presence of the blue prolene suture that the prior surgeon had the foresight to use.

### Periosteal Flap

When dealing with a complete muscle paralysis, and a lack of other active muscles that can be effectively transposed, there is a role for fixating the eye in the primary position using a periosteal flap [43].

A common scenario in which this might be useful would be the treatment of complete third cranial nerve palsy for which the LR could be sutured to the periosteum, as described above, and a nasal periosteal flap created to hold the eye in place. This procedure is probably best performed in conjunction with an oculoplastic surgeon.

### Orbitotomy for Retrieving Lost Muscles

Rosenbaum, Goldberg, and coworkers pioneered an approach for retrieving lost muscles by combining an anterior orbitotomy with a standard strabismus approach [44]. I believe that this is the most effective approach for lost muscles, with the highest success rate. This also should probably be done in conjunction with an oculoplastics surgeon. In cases of a long-standing lost muscle, or after trauma from endoscopic sinus surgery, it is useful to get a dynamic magnetic resonance image (MRI) to see if the posterior remaining portion of the MR is still contractile.

### Rectus Muscle Transpositions to Treat Torsion

As shown elsewhere (see Fig. 7.5) transposition of a rectus muscle creates a torsional force vector in the direction from which the muscle originally inserted [45]. Thus, temporal transposition of a SR or nasal transposition of an IR will intort the eye and correct extorsion. Movement in the opposite direction will extort the eye and correct intorsion. Similar reasoning applies to vertical transposition of the horizontal rectus muscles. But for reasons that are not clear, transposing the horizontal rectus muscles has a less powerful effect on torsion than the vertical muscles. I find that a 7 mm horizontal transposition of one vertical rectus muscle will correct about 7° of torsion [46].

### Scott's Graded Tenotomy for Small Deviations

For the treatment of very small vertical deviations (1–3 prism diopters), Scott described a procedure that involves disinserting the nasal or temporal fibers of the inferior or SRs [47, 48]. This is an office procedure done under topical anesthesia. The conjunctiva over the insertion is incised and the muscle identified. You start snipping away at the insertional fibers from one end (nasally or temporally) and repeatedly measure the deviation. There will start to be an effect when approximately three-fourths of the muscle is disinserted. The decision to approach from the nasal or temporal side can be influenced by the presence of torsion, if any, that you may want to correct. The procedure, in effect, acts like a small transposition of the insertion in the direction of the fibers left intact. Thus, if you perform this on an IR, and if it is a small excyclotropia that you want to reduce, you should disinsert from temporal to nasal, leaving the nasal fibers attached. This in effect would be like doing a small nasal transposition of the IR. Wright described a minor modification of Scott's procedure that involves disinserting the central fibers of the muscle [49]. Although I have no personal experience with either variation of this procedure, I know many people use it and like it. My lack of experience in part stems from the fact I have seen very few patients for whom this would be appropriate.

### Marginal Myotomy

The main effect for weakening a muscle occurs when there is shortening of the sarcomeres as occurs with a recession, or if the muscle itself is elongated but the insertion not changed. This latter can be accomplished with a myotomy, which will elongate the muscle without changing the insertion. Helveston showed that significant elongation of a muscle does not occur with small snips on one or both sides of the muscle. It does not occur until there are cuts in opposite directions that are long enough to overlap one another, and the elongation that occurs is equal to the amount of overlap (Fig. 10.20). Marginal Z-myotomies

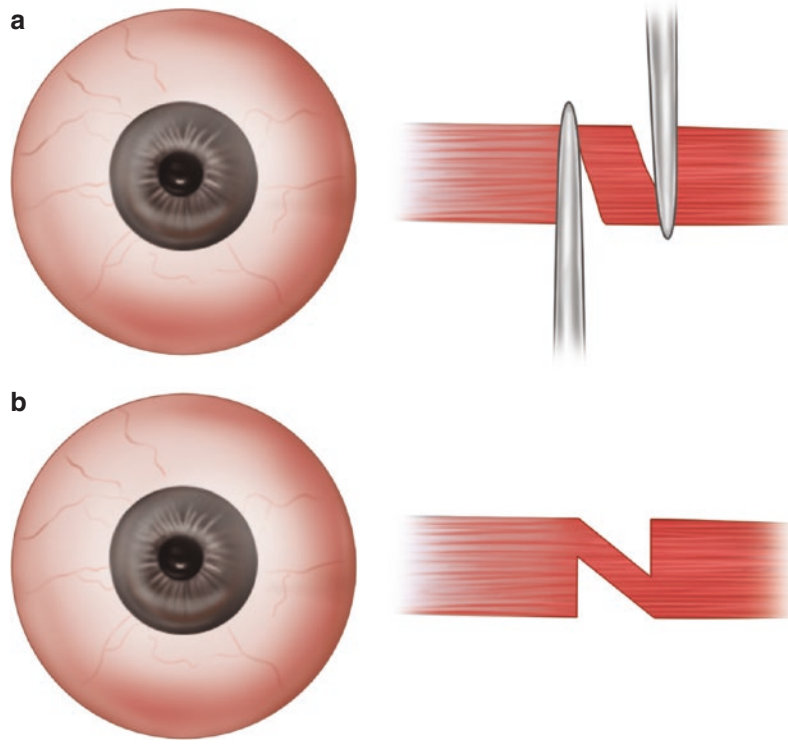



**Fig. 10.20** 75% Z-myotomy. (**a**) Hemostats are placed across the muscle. Cuts are made on the facing sides of the hemostats 75% the width of the muscle, going in opposite directions. (**b**) The hemostats are removed and the amount of lengthening is equal to the amount of overlap, in this case 50% of the muscle's width

can be useful when further weakening is needed, when a muscle is already in a significantly recessed position, and further recession risks causing an underaction. Technically, it is a relatively simple procedure, but extreme care must be taken to avoid accidentally transecting the muscle completely, which can easily happen if you are targeting myotomies that are 75% the muscle's width. The effect can be titrated, but only to a limited degree. The main disadvantage, which is substantial, is that it often results in considerable scarring, and reoperation after myotomy can be difficult. In addition to scarring, one often finds only a thin slip of intact muscle, which can easily pull in two. The width of the slip of muscle that connects the anterior and posterior muscle fibers after Z-myotomy is equal to the distance between the two cuts. The cuts should be at least 4–5 mm apart to avoid having too narrow a slip of connecting muscle.

### Lateral Rectus Equatorial Myopexy for Age-Related Distance ET

Demer and coworkers have identified the syndrome of age-related distance ET, which they also call the *sagging eye syndrome* [50–52]. It is characterized by an acquired (usually in adulthood) distance ET that exceeds the near deviation by 10 Δ and is associated with a degeneration of the connective band that connects the SR and LR. This results in inferior displacement of the LR which can be seen on coronal orbital imaging (Fig. 10.21) [53]. It can be treated with standard surgery in the form of MR recessions OU, LR resections OU, or LR equatorial myopexy [54]. This latter procedure aims to correct the cause of the problem by fixating the superior border of the sagging LR to the sclera with one nonabsorbable suture near the equator, elevating it to its normal horizontal position. Although that procedure is simpler than standard surgery, it is unclear if it is more successful. Although I have utilized this procedure in several patients with good results, my experience is insufficient to determine if LR equatorial myopexy is more effective than standard horizontal rectus recession or resection.

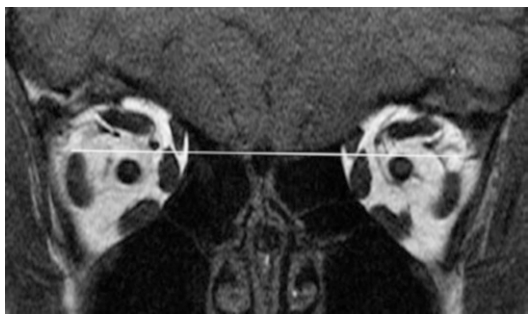


**Fig. 10.21** Coronal orbital MRI of a patient with pulley heterotopia. The left lateral rectus muscle (LR) is infraplaced with respect to the right LR. In some patients this can cause a divergence insufficiency pattern esotropia, and in others it may cause a hypotropia of the affected eye (from Kushner [53], with permission)

### Yokoyama Procedure for the Heavy Eye Syndrome (Also Known as Myopic Strabismus Fixus)

In 1997, Krzizoh and coauthors described a syndrome in which highly myopic eyes with increased axial length will develop an ET and hypotropia with limited abduction and elevation [55]. It occurs because the elongated globe herniates between the SR and LR, resulting in nasal displacement of the former and inferior displacement of the latter. Coronal orbital imaging is needed to diagnose this condition, which has been labeled the heavy eye syndrome or *myopic strabismus fixus* (Fig. 10.22). Yokoyama and colleagues are credited with describing a procedure to treat this condition [56]. It involves anastomosing the LR and SR to one another around the equator with a nonabsorbable suture. There is no suture placement in the sclera as was proposed by Krzizoh. See Chap. 21, Case 21.11 on page 319 and Case 21.23 on page 328, for representative examples of these principles.

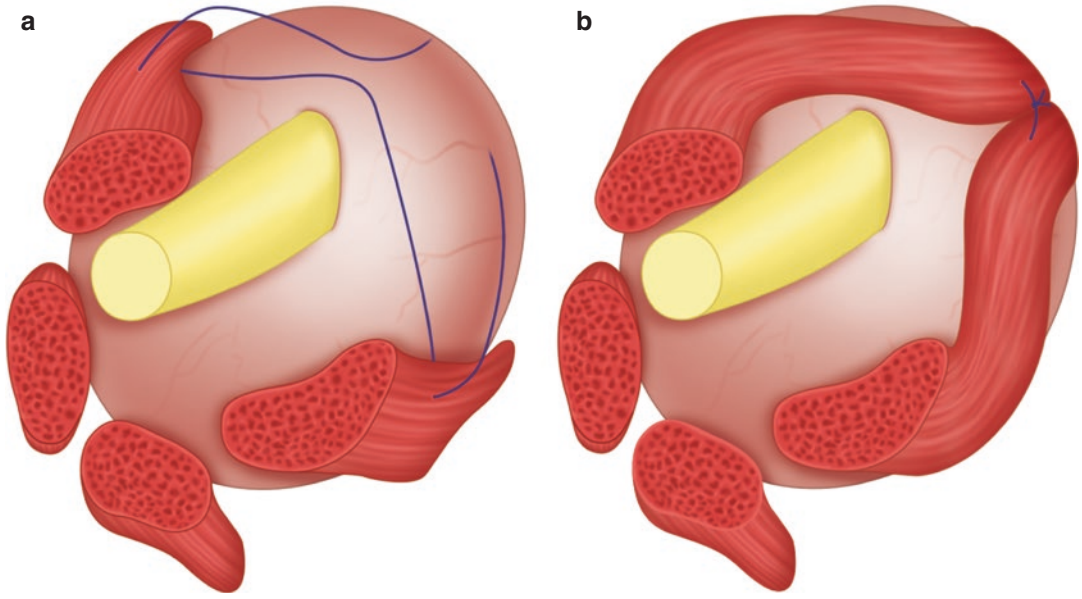



**Fig. 10.22** *Heavy eye syndrome*. (**a**) Artist's depiction of the heavy eye syndrome OD viewed from posterior. The lateral rectus muscle (LR) is inferiorly displaced and the superior rectus muscle (SR) nasally displaced. (**b**) For the Yokoyama procedure, sutures are passed from the edges of the LR and SR, which, when tied, approximate the muscles

### Important Point

*The heavy eye syndrome and the sagging eye syndrome are different entities and need to be differentiated* [51]. Standard surgery in the form of MR recessions or LR resections may work for the sagging eye syndrome. But LR resections may worsen the heavy eye syndrome. By tightening the LR, the pressure on the globe will increase and the herniation of the eye between the LR and SR will worsen.

### Pearl

If you treat a pseudophakic adult with acquired limitation of abduction and/or elevation, consider the heavy eye syndrome. Ask about a history of high myopia and, if in doubt, obtain an axial length.

### Pearl

Although orbital imaging is classically needed to diagnose the heavy eye syndrome, de Faber described an in-office test that he finds useful (Jan Tjeerd de Faber, MD, personal communication, 7 May 2017). In a patient in whom he suspects the heavy eye syndrome, he uses his index finger to retropulse the globe through the lower eyelid and then see if abduction improves. In the heavy eye syndrome, retropulsing the globe relieves some of the tension on the SR and LR, allowing them to return closer to their normal position, thus improving the ability of the eye to abduct. Given the cost of an orbital MRI, Dr. de Faber has been nicknamed Dr. Goldfinger. I have not had the opportunity to try this maneuver.

## Chemodenervation of Extraocular Muscles

The use of botulinum toxin (BTX) to pharmacologically denervate EOMS varies widely by

geographic region and individual practitioner preference. There are some who rarely use it and others who use it frequently in settings that others would opt for incisional surgery. My own experience with BTX is fairly limited, as I did not find it effective for large deviations; for smaller deviations I find many patients unhappy with the period of weeks to months after injection during which the injected muscles were pharmacologically paralyzed and there was a substantial overcorrection. They found it preferable to undergo surgery and get almost immediate improvement. In spite of this, there are several situations in which I find it quite useful.

1. The situation for which I find BTX the most useful is to treat an overcorrection shortly after surgery that involved a resection of an extraocular muscle, where ductions suggested that the resected muscle was too tight. BTX in the resected muscle a week or so after surgery would often avoid the need of a reoperation. Typically, I first try passive rotation exercises to stretch out the muscle, but if they are unsuccessful, chemodenervation with BTX is my next step.
2. I find BTX to be useful as an adjunct to incisional surgery, either to increase the effect of surgery without crippling a muscle or to avoid incisional surgery on a third or fourth rectus muscle with the intent of avoiding anterior segment ischemia. If the amount of MR recession that I feel is necessary to correct a deviation is so large that it would cripple the muscle, a standard recession of the muscle augmented with BTX is a viable option. Or, if I perform transposition of the SR and IR to the LR to treat a sixth cranial nerve palsy or Duane syndrome, but also need to weaken a tight ipsilateral MR, BTX to the MR is an alternative to recession and thus minimizes the risk of anterior segment ischemia.

I am confident that there are many more proper and effective uses for BTX for which I do not have much experience. I refer interested readers to the many publications on this subject [57–63].

### Pharmacologic Strengthening of Extraocular Muscles

As a corollary to chemodenervation of EOMS with BTX, there is exciting new work being done on injecting pharmacologic agents into EOMS to strengthen them. Most of this work has involved using bupivacaine [64], and is an outgrowth of the observation that inadvertent injection of an extraocular muscle with bupivacaine at the time of cataract surgery can lead to muscle hypertrophy and an overacting muscle [65].

### Special Surgical Procedures on the Oblique Muscles

These procedures will be discussed in detail in Chap. 12.

## Advanced Information

*Anticipate and Preempt*

We tend to choose our surgical plan foremost on the primary position deviation. There are circumstances in which your desired approach will predictably cause a problem in eccentric gaze. Consider a hypothetical patient with mild L sixth cranial nerve palsy, only slight limitation of abduction, and measurements of

Ortho ← 15 Δ LET → 25 Δ LET

You plan to approach this by recessing the left MR, which might be appropriate for a deviation of only 15 Δ in primary. This would, however, predictably cause an exotropia in right gaze where the patient is currently ortho. By anticipating this problem, you could obviate it by simultaneously doing a posterior fixation on the right LR. Another approach would be to not recess the left MR, and instead recess the right MR. This would give more correction in left gaze and have minimal effect in right gaze. See Chap. 21, Case 21.5 on page 314, for a representative example of these principles.

Anticipate adverse consequences of your surgical plan on eccentric gazes and preemptively prevent them.

## Pearl

*Surgical Drawing*

I think that I have learned more ways to improve my surgical technique by making a simple stylistic drawing of each strabismus surgical procedure at the end of the case. I use the template shown in Fig. 10.23, and have them printed on carbonless duplicate paper. One copy goes in the chart (if paper) or scanned into an electronic medical record, and the other is kept in a secure file in my clinic. On it I make notations about subtle findings that might be too esoteric to include in the dictated note. The drawing makes them easier to identify at a glance and is easier than reading a full op note. I can record the shape of the insertion, if perhaps I feel that there was a little sag to the muscle; if working with a resident or fellow, I could indicate who did which muscle, etc. The duplicate copy kept in my clinic was a defense against lost charts, computers being down, or even having to log in. I have made a drawing like this on every patient I have operated on since the middle 1970s, and have never regretted the few minutes it took to make it.

## Advanced Information

*How Does Strabismus Surgery Work—Induced Effects*

The torque a muscle generates is the force times its lever arm. Classic teaching is that recessing a muscle shortens the sarcomeres, thus decreasing the muscle's force as is depicted in Fig. 10.1. However, this does not take into account what I refer to as "induced effects" of surgery [66]. Consider an eye that is 35 Δ esotropic. If the eye straightens, any point on the nasal sclera will rotate forward in the orbit (Fig. 10.24). The amount of movement is a function of the degrees of rotation and the size of the globe. For a globe with a hypothetical diameter of 24 mm, a 35 Δ straightening of the eye will result in a 3.28 mm forward rotation of a point on the nasal sclera. As a result of this phenomenon, if you recess the MR 5 mm in an esotropic eye, and the eye straightens after surgery, the new insertion of the recessed muscle will move forward in the orbit 3.28 mm. The net shortening will only be 1.7 mm (Fig. 10.25). Moreover, the insertion of the unoperated LR will move back in the orbit 3.28 mm, thus shortening the lateral rectus muscle more than the operated MR (Fig. 10.26). When I first realized this, my initial reaction was "that's crazy." Of course for a 35 Δ ET, we would also be recessing the MR in the fixing eye, or resecting the LR in the eye that undergoes the MR recession. If you add a 7 mm resection of the LR in the deviating eye and the eye straightens, the resected LR will rotate posteriorly 3.28 mm, resulting in a net stretching of the LR of only 3.72 mm (Fig. 10.27). At first glance it would seem that you could avoid all these negating-induced effects by operating on the fixing eye. If it is already straight, it might seem that these induced effects would not occur. But the unoperated deviating eye will straighten, thus stretching the MR and shortening the LR. For any combination of surgery, e.g., symmetric recessions, recess/resect in either the fixing eye or the non-fixing eye, the net sum of muscle shortening or stretching is the same, and net amount of surgical shortening or stretching we actually get is much smaller than what we would assume necessary. How then does strabismus surgery work? Consider an eye that is deviated inward. For it to be at rest in that position, the torque of the MR minus any tissue resistance must equal the torque of the LR minus any tissue resistance (Fig. 10.28). If these net adducting forces did not equal the abducting forces, the eye would be in a different position. I think the actual effect of surgery is to change the orientation of the globe within the muscles while having a minimal effect of the length-tension of the muscles. In theory this would be the same as detaching the medial and LRs of our hypothetical esotropic patient, straightening the eye, and reattaching the muscles at whatever point on the globe does not change the muscles' sarcomere length. (Fig. 10.29). For a 35 Δ esotrope with a globe diameter of 24 mm the resultant recession and resection would each be 3.28 mm—obviously not enough. But this calculation does not take into

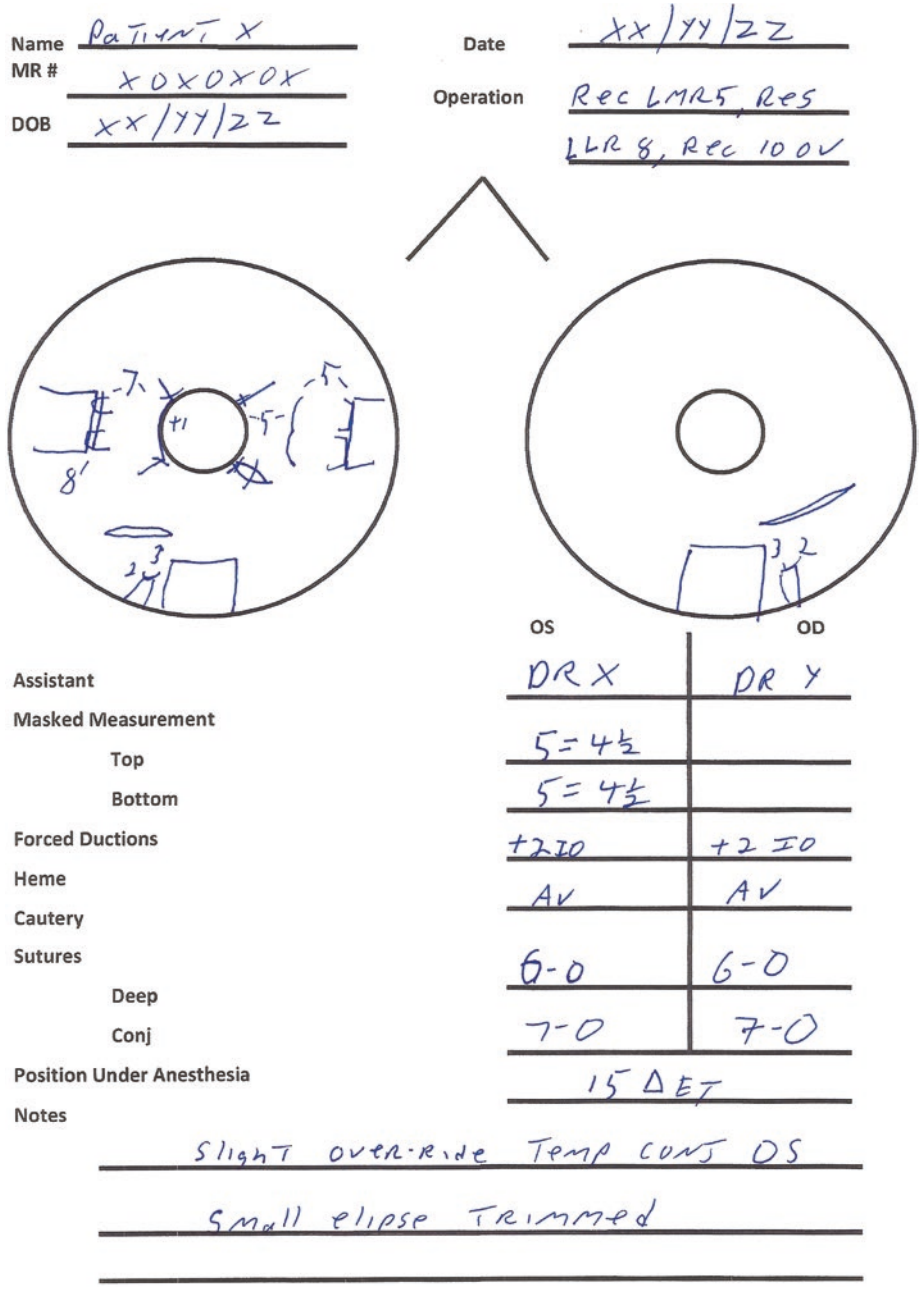



**Fig. 10.23** Template of drawing of muscle surgery filled out for a hypothetical patient who underwent an L medial rectus muscle (MR) recession of 5 mm, an L lateral rectus muscle (LR) resection of 8 mm through limbus incisions, and inferior oblique muscle (IO) recession OU to points 3 mm posterior and 2 mm temporal to the inferior rectus insertion through fornix incisions. The RMR initially inserted a 5 mm from the limbus with a scalloped insertion, and the RLR inserted at 7 mm from the limbus. A limbus incision was used, and I felt that there was slight overriding of the conjunctiva at the temporal limbus (+1). My assistants were Dr. X for the left eye and Dr. Y for the right. The initial position under anesthesia was 15 Δ ET. Exaggerated traction testing showed the IOs to be +2 tight. I used 6-vicryl on the muscles and closed conjunctiva with 7–0 vicryl. Bleeding was average. I felt that there was slight gapping of the superior-nasal relaxing incision, so I closed it with an extra suture. I trimmed a small ellipse of conjunctiva at the left temporal limbus due to the overriding. I did not close the fornix IO incisions

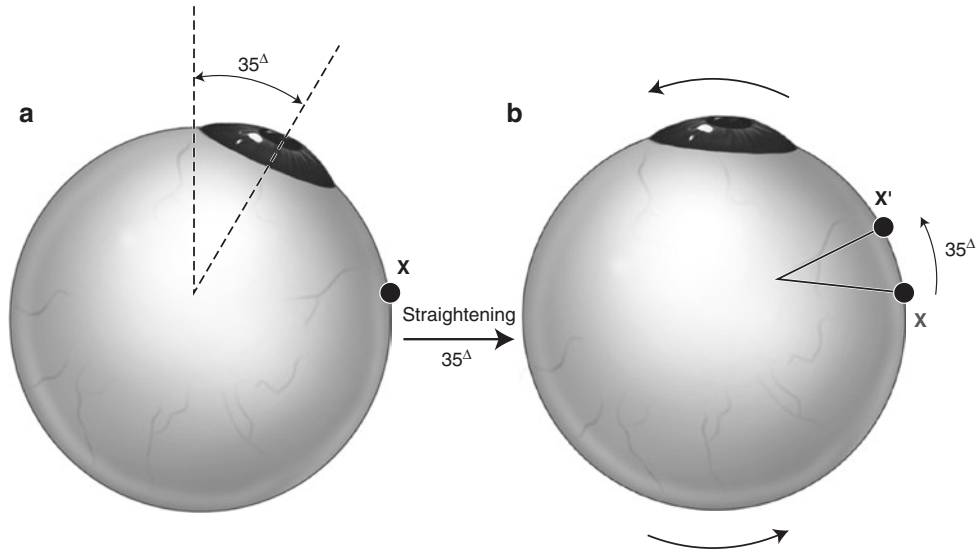



**Fig 10.24** (**a**) This eye is 35 Δ esotropic. (**b**) If the eye straightens, any arbitrary point on the nasal sclera (point *X*) will rotate forward in the orbit by 35 Δ or approximately 18.5°

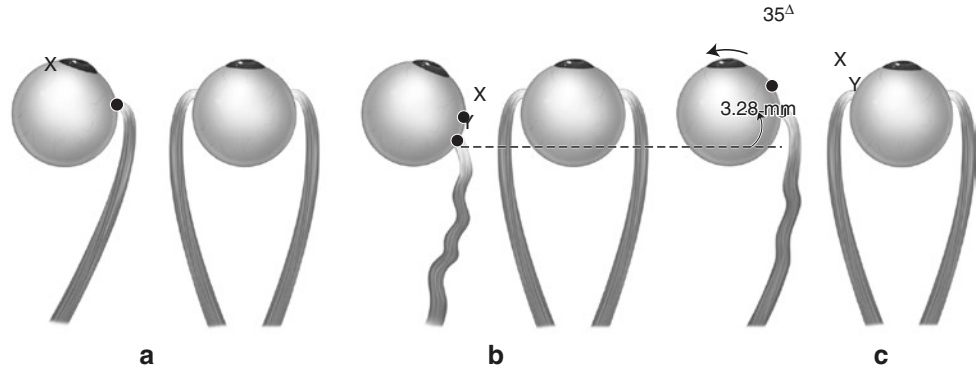



**Fig. 10.25** (**a**) The eye on the left is 35 Δ esotropic and has a horizontal diameter of 24 mm. The medial rectus muscle (MR) inserts at point *X*. (**b**) The MR is recessed 5 mm from point *X* to point *Y*. (**c**) If the eye straightens, the new MR insertion rotates forward in the orbit 3.28 mm for a net recession of only 1.72 mm

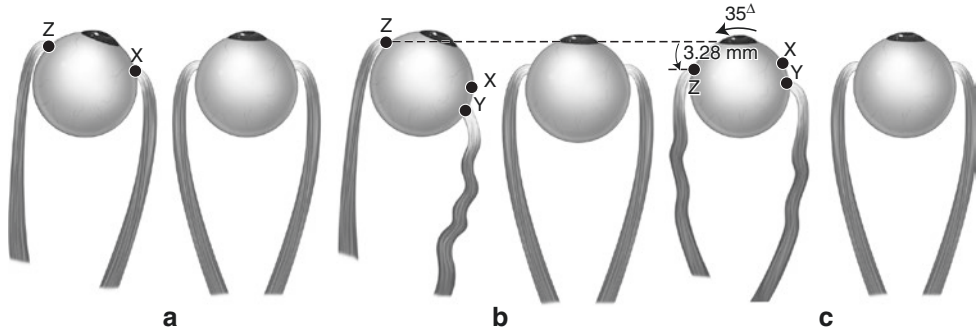



**Fig. 10.26** (**a**) The eye on the left is esotropic. The medial rectus muscle inserts at point *X* and the lateral rectus muscle at point *Z*. (**b**) The medial rectus muscle (MR) is recessed from point *X* to point *Y*. (**c**) If the eye was 35 Δ esotropic and has an axial length of 24 mm, the recessed MR will rotate forward 3.28 mm and the lateral rectus muscle (LR) will shorten 3.28 mm, if the eye straightens 35 Δ. This results in a net shortening of the MR of 1.7 mm and a shortening of the LR 3.28 mm

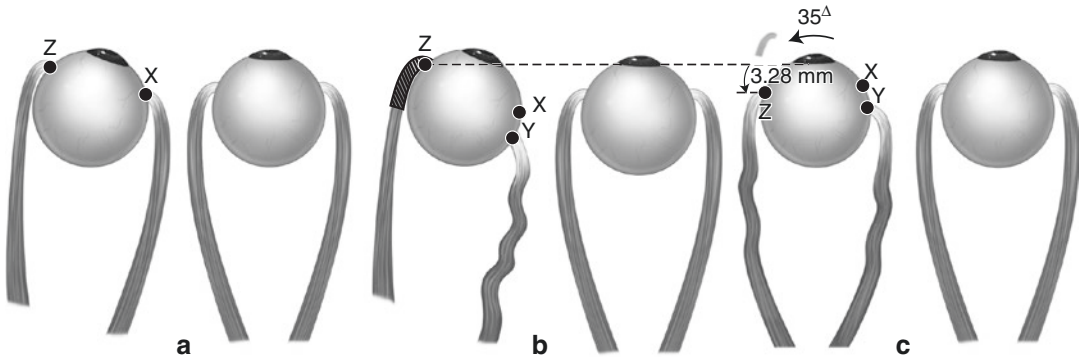



**Fig. 10.27** (**a**) The eye on the left is esotropic. The medial rectus muscle (MR) inserts at point *X* and the lateral rectus muscle (LR) at point *Z*. (**b**) The MR is recessed from point *X* to point *Y* and the LR resected 7 mm. (**c**) If the eye is 35 Δ esotropic and has an axial length of 24 mm, the recessed MR will rotate forward 3.28 mm and the LR will shorten 3.28 mm, resulting in only 3.72 mm of stretch in the resected LR

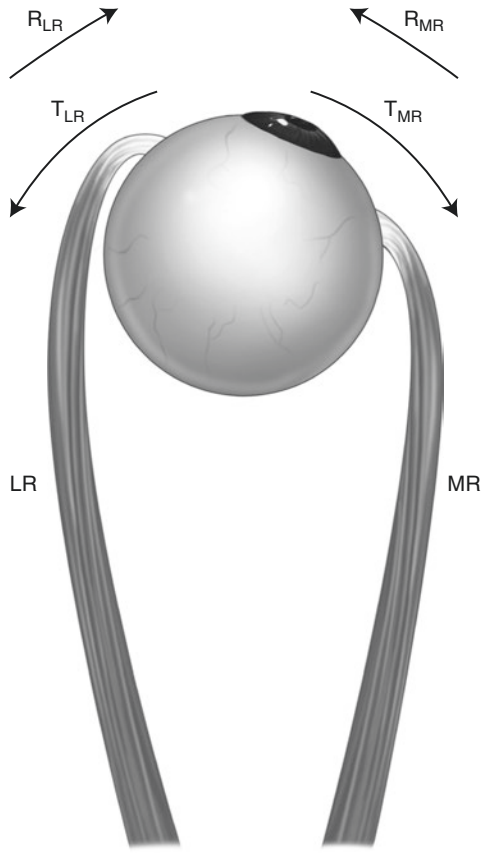



**Fig. 10.28** When this right eye is in the esotropic position, there must be an equilibrium between the torque generated by the medial rectus muscle ($T_{MR}$) and the lateral rectus muscle ($T_{LR}$) adjusted for whatever tissue resistance is present ($R_{LR}$ and $L_{LR}$)

account that there is tissue resistance that must be overcome, resulting in the need for somewhat larger amounts of surgery. This confusing subject has been discussed in more detail in a prior publication [66].

## Functional Benefits of Strabismus Surgery

### Basic Information

*Elimination of Diplopia*

It goes without saying that most patients with diplopia can be rid of that symptom with successful realignment of their eyes.

### Basic Information

*Improved Binocularity*

There is a common misperception—more on the part of comprehensive ophthalmologists, optometrists, and primary care physicians than strabismologists—that if strabismus surgery does not result in perfect binocularity and stereopsis, it is "merely cosmetic." This is particularly relevant in adults with long-standing strabismus, especially if amblyopia is present.

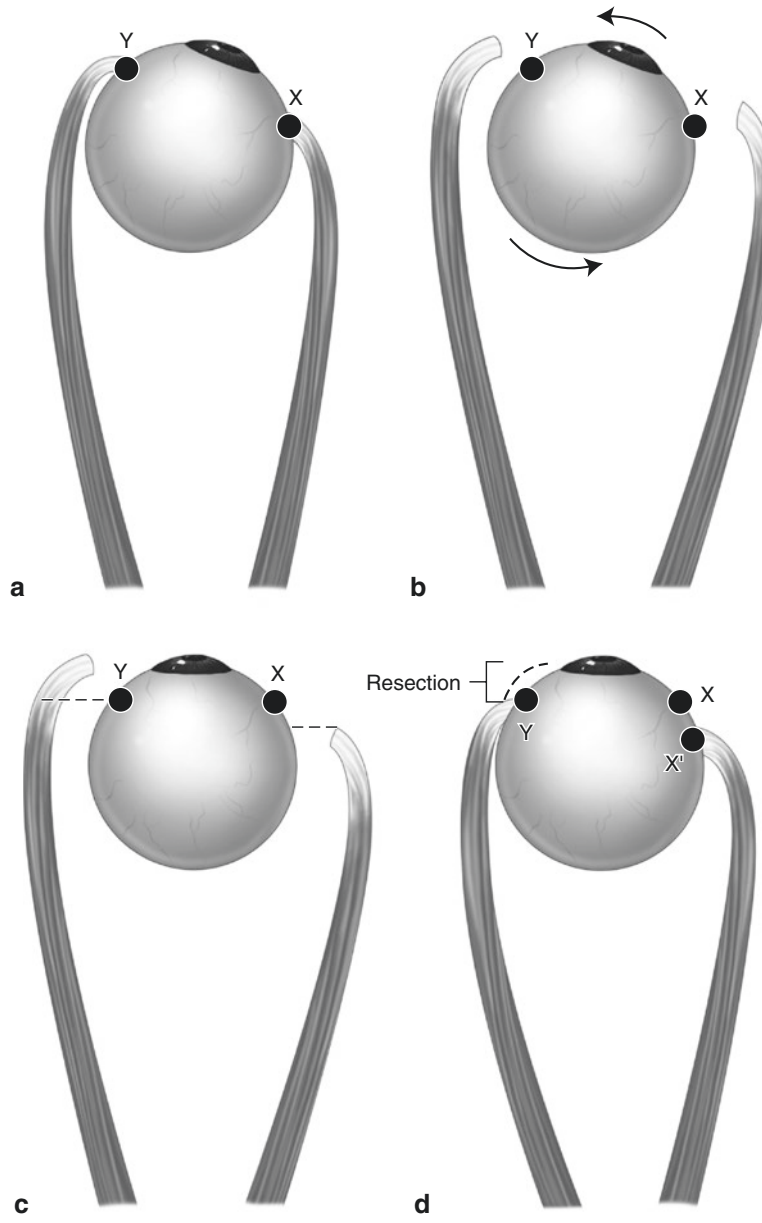



**Fig. 10.29** (**a**) In this esotropic right eye there must be a balance between all the adducting and abducting forces as depicted in Fig. 10.28. (**b**) To maintain the equilibrium, the "ideal surgical procedure" would be to detach the muscles, (**c**) rotate the eye to straight, and (**d**) reattach the muscles without altering their length. This would involve suturing the medial rectus to the point that is opposite its cut end (if the length of the muscle is not changed), suturing the lateral rectus to its insertion at the point on the muscle that is just opposite the insertion, and resecting what is anterior. This very hypothetical construct needs to be adjusted to account for tissue resistance

There are in fact many degrees of binocularity, with stereopsis being perhaps one of the highest levels. In one series of 359 adults who underwent surgery for long-standing constant strabismus, 86% of patients went from suppression to having a binocular response with the Bagolini lens test almost immediately after surgery [67]. Regardless of the type of deviation present preoperatively, the duration of strabismus, or the depth of amblyopia in the deviating eye (if present), the vast majority of patients developed binocularity after successful surgery. Moreover, the development of binocularity with the Bagolini lenses after surgery appeared to be related to the stability of the

postoperative ocular alignment. I feel the Bagolini lens test to be the best test to determine if a patient "sees with both eyes," as it most closely mimics everyday seeing circumstances.

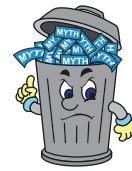
## Basic Information

*Binocular Field Expansion in Esotropes*

Patients with ET have a contracted binocular field of vision on the side of the deviating eye. Depending on the size of the deviation, this deficit can be up to 30°. In one study of 35 patients with ET 34 (97%) experienced an increase in their binocular visual field after strabismus surgery, consistent with the degree to which the eye was straightened [68]. There was no correlation with binocular field expansion after surgery and the type of ET (infantile vs. acquired), duration of the deviation, visual acuity in the deviating eye, or a history of satisfactory alignment in early childhood.

## Try This Experiment

With both eyes open fixate on the wall facing you. Now close one eye, and you can immediately appreciate the loss of visual field on the side of the closed eye. Then open the eye again to see the difference of the expanded visual field. I believe that expansion of binocular visual field is a major benefit of ET surgery. Reports indicate that visual field constriction is highly correlated with an increase in motor vehicle accidents [69–71]. In addition, dynamic visual fields typically improve after successful strabismus surgery, which is important for activities like driving a motor vehicle or navigating stairs [72]. I speculate that the developmental gains that are reported in infants undergoing surgery for infantile ET may be due to an expansion of their binocular field after surgery [68, 73].

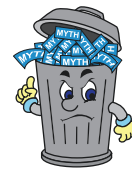



## Myth

If a patient does not develop high-grade stereopsis after strabismus surgery, there is no functional benefit.

## Fact

Stereopsis is only one form of binocularity. Most patients will show improvement in some forms of binocular function if properly aligned, even if they do not have stereopsis.




## Myth

If a patient does not have stereopsis, they have no depth perception.

## Fact

I think it is a mistake to tell a stereoblind patient that they lack depth perception, especially if they lacked stereopsis from infancy. Stereopsis is only one mechanism by which we judge depth, and such patients learn to use other cues. These include parallax, the fact that near objects overlap distance objects, and other contextual cues.

## Advanced Information

*Binocular Inhibition and Summation*

Binocular summation refers to the situation in which vision is better binocularly than in either eye separately; binocular inhibition refers to vision being poorer under binocular conditions than monocularly. Studies spearheaded by Pineles [74–78] show that strabismic patients have decreased binocular summation and increased binocular inhibition, both of which may become more normal after successful strabismus surgery.

## Basic Information

*Psychosocial Benefits of Strabismus Surgery*

There are serious and lasting effects of a psychosocial nature due to strabismus, according to numerous reports [79–84]. The majority of adults with strabismus feel that their ocular misalignment has substantial adverse effects on their quality of life, self-image, interpersonal relationships, ability to secure employment, social anxiety, and visual functioning. Also levels of distress higher than those of age- and gender-matched controls are reported in adults with strabismus [80]. Although these are subjective impressions on the part of strabismic adults, they have been confirmed objectively. Studies show that adults with strabismus are considered less intelligent, are viewed with a significant negative social prejudice, are less likely to be hired for a job, and are less likely to be promoted in the military [85–87]. According to dating agencies, aesthetically noticeable strabismus negatively influences the ability to find a partner [88]. These findings are extremely meaningful as reflected in the finding that a majority of adults with strabismus would trade a portion of their life expectancy in return for being rid of strabismus and its associated effects—this seems almost astonishing. A neurophysiologic basis for this bias was presented in a study by Berberat [89]. In that study, 31 volunteers underwent functional MRI while viewing images of strabismic and normal subjects. In 30 of the 31 volunteers, viewing strabismic images led to significant activation in the amygdala, hippocampus, and fusiform gyri, as compared to viewing images of normal subjects, indicating a negative emotional response. The authors concluded that "treatment of strabismus therefore changes the interpersonal dynamic for patients with strabismus on a demonstrable organic basis." Interestingly, the only one of the 31 volunteers who did not show activation in the aforementioned areas of the brain was an ophthalmologist who was involved in designing the study [90]. Studies show that adult strabismus surgery has a positive impact on health-related quality of life [91–93].

Strabismus has profound psychosocial consequences.

Although these psychosocial issues stem from having an abnormal appearance and abnormal eye contact, they are certainly real and go beyond being simply a cosmetic issue. Inability to get a job or to work in certain profession is as much a physical handicap as having an orthopedic or cardiovascular problem that prevents certain activities.

## Question

Is surgery in the adult with long-standing strabismus, no diplopia, and little or no fusion potential "only cosmetic"? After all, they may gain nothing other than an improvement in their appearance.


## Reply

No, it is not "only cosmetic." It demeans the significance of strabismus to refer to it as being only a cosmetic problem. A cosmetic problem is a feature that is perfectly normal, but with which a person is dissatisfied. Having misaligned eyes is not normal. Strabismus surgery corrects a pathologic condition and does not merely enhance a subjective sense of beauty. Consequently, it is more appropriately described as being restorative rather than cosmetic [80, 94, 95].

## Basic Information

*Outcome Criteria*

I think few things have hampered progress in strabismus research as much as

the lack of an agreed-upon criteria for a successful outcome. And I think few things have been as deleterious as the reliance on "within 10 Δ from straight" as a criterion for success [96]. Historically, I think this stems from Parks' thesis on the *monofixation syndrome*, in which he observed that it tends to be a stable and acceptable form of subnormal binocular vision, and that it can occur in both esotropes and exotropes of up to 10 Δ [97]. However, a patient who started with an ET, and after surgery has an XT of less than 10 Δ, is not as stable as a de novo monofixational exotrope [96]. Although for patients in whom surgery is planned only to correct aesthetics, and fusion or absence of diplopia is not an issue, a deviation of up to 10 Δ may be acceptable. But if diplopia is an issue, any overcorrection, however small, of an acquired vertical deviation is problematic. Similarly, any permanent overcorrection of an intermittent exotropia will result in diplopia. The range of diplopia-free vision is important. An intermittent exotrope who is perfectly ortho in primary after a recess/resect procedure may have diplopia beginning just a few degrees in side gaze to the side opposite the operated eye, which should not be considered a success. Being diplopia free in downgaze is also an important consideration for reading.

It is also problematic that many studies report only short-term outcomes, e.g., 6 weeks or 6 months. It can be equally problematic to evaluate a large series of patients as of their last exam, which may have considerable variation with respect to duration after surgery. I think of strabismus as a four-dimensional specialty—a metaphor I first heard from Harley Bicas, M.D. The first dimension is a point, which correlates to the primary position measurement. The second dimension is a plane, which is represented by our measurements taken in the cardinal fields of gaze. Depth is the third dimension, and, accordingly, we measure patients at distance and near. The fourth dimension is time. And that is what is missing from so many strabismus studies. We need to see how a patient fares not only in the short term, but also over a long period of time. I recognize how hard it is to obtain long-term follow-up on patients. But unless a study does so, its conclusions are limited. Collaborative work needs to be done to develop agreed-upon outcome criteria for success.

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# Complications of Eye Muscle Surgery

# 11

*I know a surgeon who always says a prayer at the start of each case, "God, these are your hands. Don't embarrass yourself."*

*—Thich Nhat Hahn*

## Basic Information

*Postoperative Irritation and Pain*

Although it is normal to have some degree of pain and irritation after strabismus surgery, one must always be attentive to the remote possibility of a cellulitis, which requires systemic antibiotics, or the even more remote possibility of endophthalmitis. Usually, swelling and discomfort are most prominent the morning after surgery and improve as the day progresses. However, if the symptoms are worsening, and particularly if the pain is of a deep nature, as opposed to surface irritation, the patient should be seen to rule out a serious problem. Common causes of surface pain include suture irritation (if conjunctival sutures were used), delle formation, or filamentary keratitis. For reasons that are not clear, preseptal cellulitis can occur and is more common than orbital cellulitis after strabismus surgery. One also must consider an allergic conjunctivitis to be the cause if postoperative topical antibiotics were used. If bilateral surgery was performed, and the irritation is unilateral, you are not dealing with an allergic problem. Treatment of allergic conjunctivitis includes discontinuing the offending medication, and if severe a short course of mild topical steroids.

## Basic Information

*Persistent Redness*

Some persistent redness that progressively subsides should be expected for several weeks after uncomplicated strabismus surgery. In cases of reoperations where there is substantial scarring, this may persist for several months. Some patients will have an increased sensitivity of the conjunctiva for as long as a year post-op. The eye may tend to redden more easily after swimming or exposure to irritants. This is also more likely to occur after reoperation procedures. Patients can be reassured that with time this problem should subside.

## Basic Information

*Wound Healing Problems*

These problems are minimized by use of the fornix approach; however, with care they can also be avoided with either limbus incisions or a modified Swan approach. Rectus muscle resections or plications can result in a redundancy of conjunctiva at the limbus as the tissues are dragged forward. If, at the time of closure, conjunctiva appears redundant at the


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_11

limbus, resecting a small ellipse of anterior conjunctiva will prevent postoperative conjunctival redundancy.

Sometimes the original insertion line remains visible after a rectus muscle recession. This is because the scleral bed just under the muscle behind original insertion is thinner, and the interface between and the thicker sclera at the insertion show as a dark line. Patients often think this is a scar. When this occurs, it usually does not change over time, but fortunately it is not a significant cosmetic issue.

## Basic Information

*External Infection*

In order of increasing severity, and decreasing frequency, postoperative external infections can take the form of conjunctivitis, preseptal cellulitis, or orbital cellulitis. Fortunately, all are relatively uncommon, with cellulitis having a reported incidence of 1 in 1900 cases [1]. Conjunctivitis will usually respond to topical antibiotics, whereas preseptal or orbital cellulitis requires systemic antibiotics—oral antibiotics may be adequate for a preseptal cellulitis, but orbital cellulitis usually requires intravenous treatment. Orbital cellulitis can be life threatening and vision threatening, usually presenting between 1 and 4 days after surgery. An orbital computed tomography (CT) scan may be needed to differentiate orbital from preseptal involvement.

## Basic Information

*Endophthalmitis*

This potentially devastating complication is quite rare following strabismus surgery with an estimated incidence of 1 in 30,000 cases [1]. It is generally felt that inadvertent scleral perforation at the time of surgery is the source of bacteria entering the globe; however, the incidence of scleral perforation greatly exceeds that of endophthalmitis [2].

## Basic Information

*Anterior Segment Ischemia (ASI)*

The blood supply to the anterior segment is via the anterior ciliary and long posterior ciliary arteries. When this is sufficiently compromised, an anterior segment inflammatory reaction results. Full-width rectus muscle disinsertion disrupts the anterior circulation to varying degrees, depending on which and how many muscles are operated upon, and various systemic risk factors. In general, disinsertion of the horizontal rectus muscles is less disruptive than the vertical rectus muscles, and the incidence of ASI goes up if both vertical rectus muscles or one vertical rectus and the adjacent medial rectus muscle (MR) are operated upon. The exact incidence of this complication is hard to determine, because most surgical procedures are limited to one or two horizontal rectus muscles in a given eye, circumstances in which ASI almost never occurs. Anything that is associated with poorer arterial circulation, especially increased age, increases the risk of ASI, which is extremely rare in healthy children. Other risk factors include vasculopathies, coagulopathies, and thyroid eye disease (TED). A limbus incision is more likely to result in ASI than a fornix incision, because it disrupts the proximal circulation, particularly if peri-limbus cautery is used.

McKeown described a technique for sparing the anterior ciliary arteries during rectus muscle recession or resection, which in theory should greatly decrease the incidence of ASI [3]. See Chap. 10, "Strabismus Surgery," for a detailed description.

Clinically, ASI presents as an iritis with or without iridoplegia, and corneal edema. In most cases sequelae are minimal, but severe cases can result in iris atrophy, a rigid pupil, cataract, loss of accommodation, and glaucoma. Treatment in the acute stage should include cycloplegia and topical corticosteroids.

## Question

How can I minimize the risk of my patient developing ASI?


## Reply

A conservative approach is to limit a surgical session to no more than two rectus muscles in each eye in adults, and to three rectus muscles in children. If a reoperation is needed, wait for at least 3–6 months before operating a fourth rectus muscle in a child, or a third or fourth in an adult. Err toward the higher number in patients who have the aforementioned risk factors for ASI. Although waiting decreases the likelihood of ASI developing, it is no guarantee that ASI will not occur. Studies have shown that iris perfusion defects can be permanent after rectus muscle disinsertion [4]. Consider an iris angiogram before reoperating. The absence of sectoral filling defects indicates that it is safe to proceed. Also, in patients who are at high risk, avoid a limbus incision or use of peri-limbus cautery. For the vertical rectus muscles, it is always safest to make the incision directly over the muscle insertion—a modified Swan approach.

## Advanced Information

*Subconjunctival Cysts*

Frequently subconjunctival cysts occur at the interface between the anterior cut end of the operated muscle and the sclera, at the level of the new insertion [5]. They are thought to result from epithelial cells being dragged into the scleral tunnel along with the suture. They can occur from weeks after surgery to as long as many years later. I have seen them appear clinically over 20 years after a rectus muscle recession. In theory, the most important step for preventing their occurrence is for the surgeon and surgical assistant to be careful to prevent the capsule of Tenon and surrounding connective tissue from being dragged into the scleral suture tunnel. Removal of these cysts can be tricky. Often, what is visible is only the tip of the iceberg, and the cyst turns out to be much larger than expected when it is dissected free (see Fig. 11.1) [5]. More importantly, when cysts form at the interface of the muscle and the new scleral insertion, the muscle ends up attached only to the posterior aspect of the cyst, attaching to the sclera only indirectly via the cyst (see Fig. 11.2). A pseudotendon usually forms between the intended new insertion site and the muscle posterior to the cyst. If you are not aware of this configuration, it is easy to end up with a lost muscle after you excise the cyst. You must look carefully for muscle attaching to the posterior edge of the cyst.

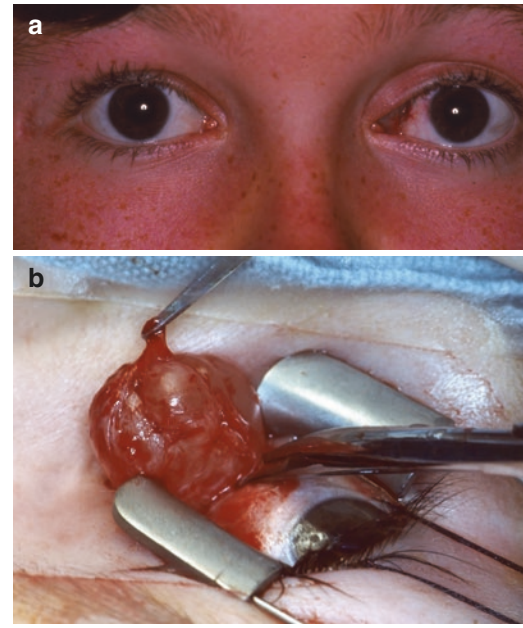



**Fig. 11.1** (**a**) What appears to be a small cyst appeared in the left medial canthal area 2 and a half years after bilateral medial rectus recessions (from Kushner [5], with permission. ©1992 American Medical Association. All rights reserved). (**b**) At the time of excision, it turned out to be much larger than was suspected on clinical exam

> Things are not always what they seem; the first appearance deceives many; the intelligence of a few perceives what has been carefully hidden.—Phaedrus, circa 370 BCE

## Important Point

*Removal of cysts can be tricky*. Look for the muscle attaching to the posterior aspect of the cyst instead of the sclera and be on the lookout for a pseudotendon, which can easily be mistaken for muscle.

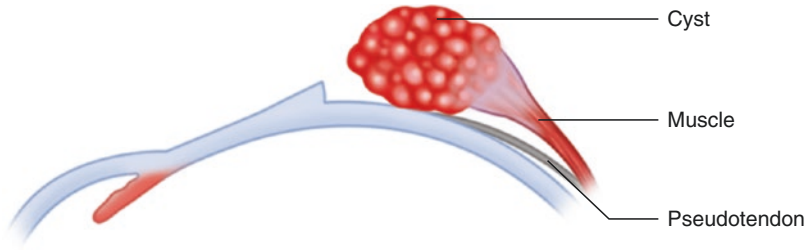



**Fig. 11.2** Artist's depiction of the common configuration found in patients with subconjunctival cysts resulting from strabismus surgery. The cyst is attached to the sclera at the site of the intended muscle insertion. From that site a pseudotendon runs to the undersurface of the muscle posterior to the cyst to the intended site of the recessed muscle's insertion. The muscle only inserts on the cyst

## Advanced Information

*Occurrence of Lost Muscles, Intraoperatively*

There are several main ways in which a muscle is lost intraoperatively. Defective or traumatized suture material may fray and break when being pulled through scleral tunnels or while being tied. If this happens and it leads to a lost muscle, the presence of remaining ends of suture material in the muscle may aid in retrieving the retracted muscle. Inadequate suturing, particularly of a tight muscle, may result in the sutures pulling out. For this reason, many surgeons use two sutures for resections, even if they use only one for recessions. That is wise. I prefer two sutures for both recessions and resections, and appreciate the added comfort that doing so provides. If you do use only one suture for any rectus muscle procedure, placing a suture bite and square knot in the middle of the muscle just posterior to the insertional end provides extra safety; if either arm of the suture pulls out or breaks, the muscle is still secured. The *pulled-in-two syndrome* is the situation in which a muscle snaps in two, typically during the cleaning up of surrounding connective tissue, but it may even occur with just presumably normal traction on the muscle with a hook. It is more likely to occur if there is an underlying myopathy such as TED or with a long-standing paretic muscle that has atrophied. Muscles can be lost during an attempted Z-myotomy if the cuts come too close to the opposite edge of the muscle. Finally, a muscle may become lost if a resection clamp slips off the muscle before it has been secured with sutures. In a series of 24 cases of lost muscles, Parks found that 19 of them were MRs, followed by three lateral rectus muscles (LRs), and only one each for the superior and inferior rectus muscles [6]. The MR was lost most frequently because it is more frequently operated on than the other muscles, has a shorter wraparound arc, and is not constrained by an adjacent oblique muscle.

## Advanced Information

*Treatment of Lost Muscles, Intraoperatively*

If you have the misfortune to lose a muscle during surgery, how you respond immediately can profoundly affect the outcome. Resist the instinct to rotate the eye out of the field of action of the lost muscle for the purpose of maximizing exposure, e.g., abducting the eye to provide exposure for a lost MR. Initially the muscle may not have retracted fully in the orbit and may be held somewhat anteriorly by tenuous remaining connective tissue attachments. Abducting the eye to find a lost MR may cause the muscle to break loose and retract further. Instead try to go after the muscle with as little rotation away from the muscle as possible. The use of a head light is wise, as is enlarging the conjunctival incision. Make as diligent a search as you can, because retrieving the lost muscle will always produce a better outcome than any other treatment. However, avoid making blind passes with forceps deep in the orbit, as that can result in substantial scarring. If the lost muscle is either of the vertical rectus muscles, you may be

helped by finding the adjacent oblique muscle or tendon. Sometimes the connective tissue attachments between the oblique muscle or tendon and the adjacent vertical rectus muscle may constrain a lost vertical rectus muscle from retracting further. If you cannot find the muscle, you should do some form of transposition procedure to give active force in the direction of the lost muscle.

## Pearl

If you do lose a muscle intraoperatively, resist the instinct to rotate the eye out of the field of action of the lost muscle for the purpose of maximizing exposure. Doing so may cause the lost muscle to slip back further.

## Advanced Information

*Surgical Treatment of Previously Lost Muscles*

Without doubt, the best time to treat a lost muscle is when it occurs. But often patients with lost muscles are referred to you after the fact. If this is within a day or so of the original surgical misadventure, immediate surgical exploration is wise. If several days have elapsed, it may be wiser to wait until things settle down, as there will be increased vascularity and bleeding that will compromise surgery if you intervene more than several days after the misadventure, yet before healing is complete. I like to wait until about 6 weeks after the initial procedure.

Orbital imaging can be invaluable in assessing these patients. I have seen patients who were thought to have lost muscles clinically, yet a CT scan or magnetic resonance image (MRI) showed the muscle attaching to the globe with what appeared to be a firm bond, in a somewhat posterior position. Such findings greatly facilitated retrieval. The boy in Fig. 11.3a had undergone a Kestenbaum–Anderson procedure for nystagmus with a face turn to his right. He underwent a large right (R) MR recession, RLR resection, left (L) LR recession, and last an attempt at an LMR resection by another ophthalmologist. The sutures were pulled out of the LMR and the muscle was declared "lost," and attempts to retrieve it were not successful. I saw him several months later when he was referred for anticipated transposition surgery. I found a large LXT with moderately decreased adduction OS. However, a CAT scan OS

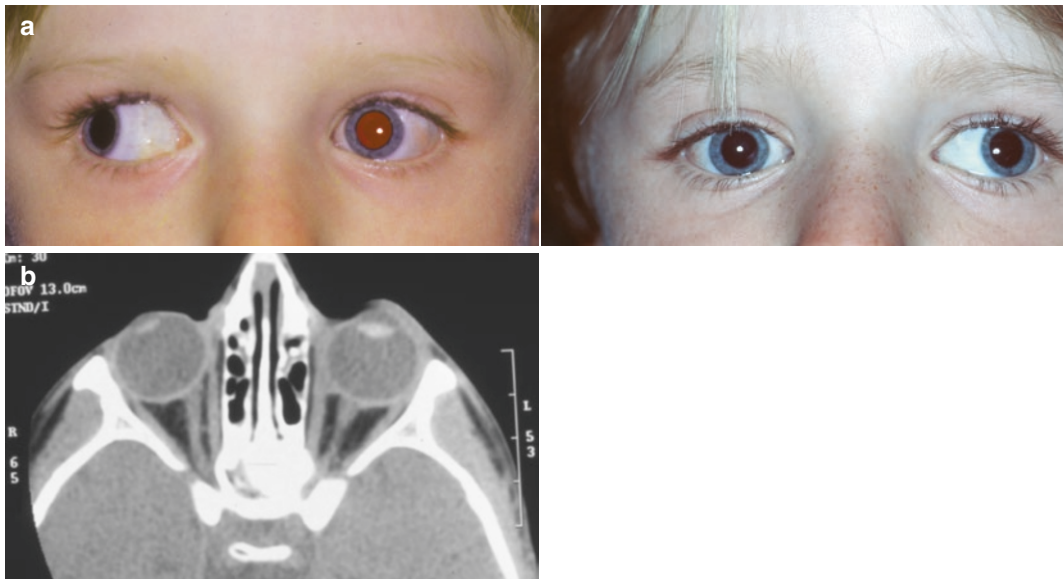



**Fig. 11.3** (**a**) A boy whose left medial rectus muscle was "lost" during a prior resection procedure. Adduction of the left eye is decreased. (**b**) Computed tomography scan of the left eye shows the medial rectus attached to the globe. The muscle appears to have normal integrity. (**c**) The boy in Fig. 11.3a seen 1 year after the left medial rectus muscle was retrieved and advanced. Adduction is good

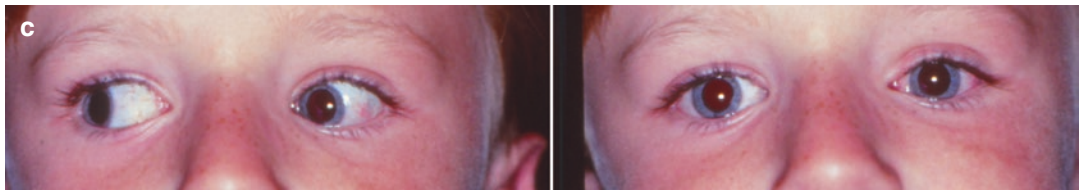



**Fig. 11.3** (continued)

showed the LMR attached to the globe with apparently good integrity (Fig. 11.3b). I was able to easily retrieve and advance the LMR, which improved the LXT and adduction OS (Fig. 11.3c).

A multipositional MRI (also known as *dynamic* MRI) cannot only help identify a muscle's location but also tell if the retracted muscle still has contractility. If so, retrieval of the muscle is worth attempting. If the muscle is not contractile, then retrieving that muscle may have no better outcome than a transposition procedure, which is usually less complicated. The following case is illustrative:

**Case 11.1.**

I first examined this woman at 21 years of age. At approximately 4 years of age she underwent what was planned to be a recess/resect procedure OD for an esotropia. The sutures were pulled out of the MR and the muscle could not be retrieved after extensive exploration. This left her with a large RXT, very limited adduction OD, and a hypertrophic conjunctival scar nasally (Fig. 11.4a). At 18 years of age she underwent surgery by a second ophthalmologist. At exploration, the RMR could not be found. Nasal transpositions of the R superior rectus muscle (SR) and R inferior rectus muscle (IR) were done, and the nasal conjunctival scar was revised. This decreased, but did not eliminate, the RXT, and she still had very limited adduction OD (Figs. 11.4b, c). After my initial examination I obtained a multipositional MRI of her orbits. It showed that the RMR was attached to the nasal orbital wall, and it showed good contractile function on attempted adduction (Fig. 11.5). Aided by that information I was able to retrieve the RMR via an orbitotomy approach and reattach it to the globe. This corrected the exotropia and improved her adduction; her motility has remained stable for the subsequent 14 years (Fig. 11.6).

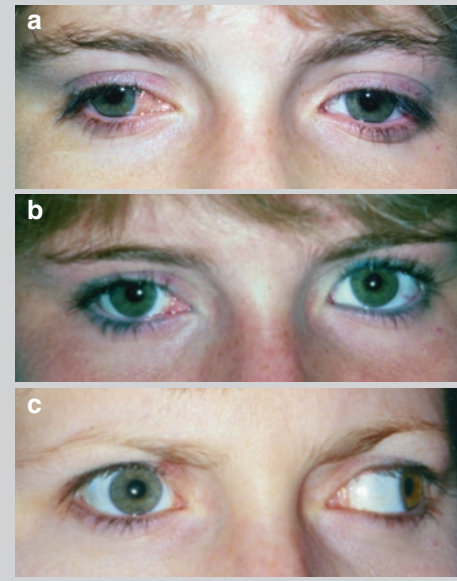



**Fig. 11.4** (**a**) 18-year-old woman with right exotropia (RXT) secondary to her right medial rectus muscle being "lost" during strabismus surgery 14 years earlier. (**b**) Three years after vertical rectus transpositions nasally OD and a nasal scar conjunctival scar revision, her RXT is less but still significant. (**c**) Her adduction OD is markedly deficient

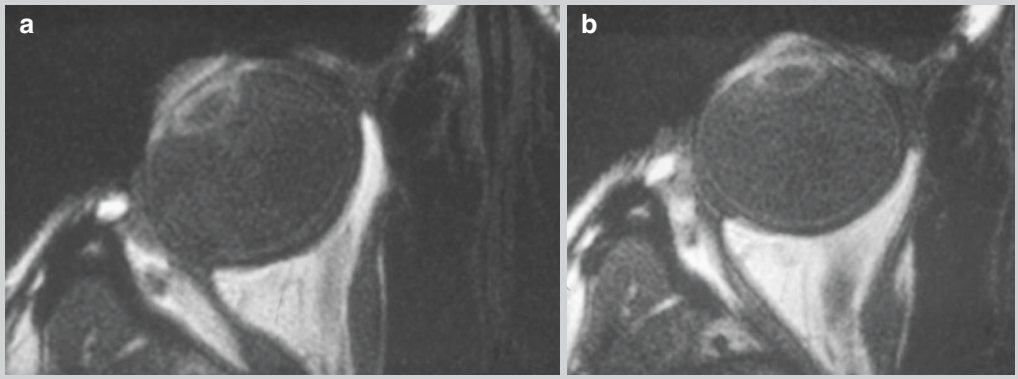



**Fig. 11.5** Multipositional MRI of the patient shown in Fig. 11.4 at 21 years of age. (**a**) Patient looking to her right. The right medial rectus is not attached to the globe and inserts on the medial orbital wall. (**b**) On attempted adduction OD, there is thickening of the right medial rectus muscle, indicating that despite 17 years having elapsed since it was lost, there is still good contractility.

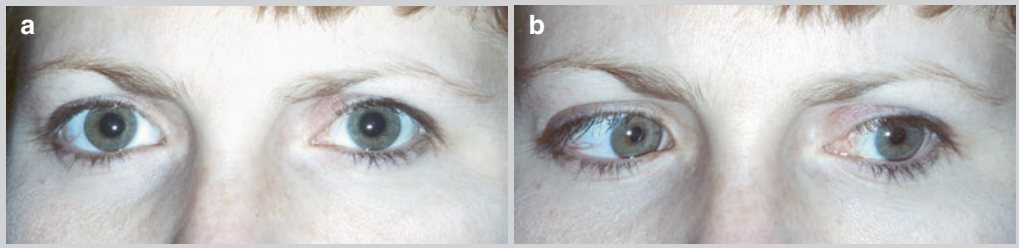



**Fig. 11.6** (**a**) Patient shown in Fig. 11.4, 14 years after the right medial rectus muscle was retrieved and resutured to the globe. There is good alignment in the primary position and (**b**) adduction is greatly improved

Orbital imaging, however, can also be misleading if you do not know what to look for. Early in my experience using orbital imaging to help plan surgery in cases of suspected lost muscles, I sometimes found that in patients in whom imaging suggested that the muscle was still attached to the globe by a strand of connective tissue, no such connection was found at surgery. I now realize that what may look like a thin attachment of the muscle to the globe may be an imaging artifact. If the muscle has completely retracted to the back of the orbit, the two opposing sides of orbital tissue that had been adjacent to the muscle belly come in contact with one another and create an interface that shows up on imaging (Fig. 11.7). What one is seeing is really the empty channel the muscle had occupied, which has collapsed. I call this the *collapsed empty channel sign*—a term I first heard from Lindell Gentry, MD. Even though it is empty of muscle or tendon and the channel has collapsed, it is still visible.

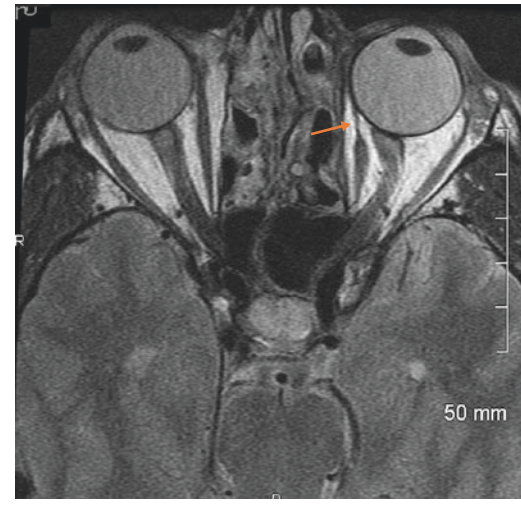



**Fig. 11.7** Orbital MRI of patient with a lost left medial rectus muscle secondary to a penetrating nasal orbital injury OD. Imaging suggests that the left medial rectus is still attached to the globe by a thin strip of connective tissue (*arrow*). In fact, at surgery no such connection was found. This is the *collapsed empty channel sign*

## Pearl

The appearance of a thin band connecting a presumed lost muscle to the globe on orbital imaging may be the *collapsed empty channel sign* artifact, and possibly there may not be a connection found at surgery between the lost muscle and the globe.

## Advanced Information

*Surgical Technique for Retrieving Lost Muscles*

If preoperative imaging shows that a lost muscle is in the posterior orbit, and multipositional images show that it is still contractile, the technique that I find most successful for retrieving it is the one described by the University of California, Los Angeles group [7]. This procedure is best done in conjunction with an oculoplastics specialist. It involves first locating the muscle via a medial wall approach done by an oculoplastics surgeon who then sutures the muscle and passes the sutures out under a conjunctival flap where the strabismologist takes over for the muscle positioning.

## Advanced Information

*Oblique Muscle Incarceration Syndromes*

See Chap. 13, "How to Perform Superior Surgery on the and Avoid Inferior Surgery on the Superior Oblique," for a detailed discussion.

## Advanced Information

*Surgeon's Disorientation Due to Torsion*

Keep in mind that in cases in which there is substantial oblique muscle dysfunction and/or fundus torsion present, the rectus muscle insertions will be rotated clockwise or counterclockwise from their expected position, and this can lead to surgical misadventures. I recall seeing a patient from a comprehensive ophthalmologist who was relatively new in practice. He thought he had performed bilateral MR recessions of 5 mm and IO recessions for a 35 Δ esotropia with a V-pattern. He got absolutely no effect. When I subsequently operated on the patient I discovered that instead of recessing the IOs, he had inadvertently recessed the LRs (as well as the MRs). The surgery on the antagonist LRs negated the effect of the MR recessions. Presumably due to the excyclorotation of the globe the LRs were situated inferior-temporally, and mistaken for the IOs. How the surgeon failed to get reoriented after seeing the direction the fibers ran in the muscle was never clear to me.

## Pearl

Substantial torsion will alter the location at which the rectus muscles will insert from the surgeon's viewpoint.

## Basic Information

*Incidence and Prevention of Scleral Perforation*

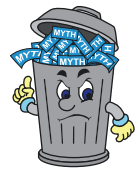
Published incidence figures for scleral perforation range from 1% per muscle operated to as high as 12%; it does appear that this figure is decreasing to perhaps below 1% [8, 9]. This is probably due to better instrumentation, surgical needle design, and surgeon training. In addition, the incidence probably varies greatly with suturing technique (separate from skill) and patient-surgeon positioning. The actual incidence of scleral perforation during strabismus surgery is probably unknown for a number of reasons. Most importantly, many cases are clinically unrecognized at the time of occurrence, and because fortunately there are

usually no sequelae, they remain unknown to the surgeon. Whenever I see a patient for the first time who has a history of prior strabismus surgery, I always take extra care to look in the retinal periphery under the rectus muscles' insertions after dilation. It is surprising how frequently I see evidence of prior scleral perforation in patients for whom prior clinic and operative notes gave no indication that perforation had occurred.

I consider the passing of scleral sutures to be a two-handed process. If the surgeon fixates the insertion with one hand and passes the suture with the other, she has far more control than if the assistant fixates the sclera. I also feel that there is more control if you always pass the suture toward the point of fixation. I like to sit opposite the muscle I am operating on, fixate the insertion with my nondominant left hand, and pass the suture toward the forceps, fixating the insertion with my right hand. So for operating on the left MR I would sit facing the patient's left ear. I know that most people who prefer fornix surgery also prefer the Parks "crossed swords" technique of suturing, where the assistant fixates the insertion. I recognize that there are some advantages with the fornix incision, but do not see any advantages with that method of suturing. I think it leads to an increased incidence of scleral perforations. At the very least, I would encourage those readers who do not want to abandon the Parks method of suturing (e.g., always sitting at the head, assistant fixating the insertion, and passing the sutures toward one another) to at least give up "crossing the swords." Parks' original reason for overlapping the suture passes stems from when catgut was used for strabismus surgery. That material was slippery, and Parks found that if he overlapped the suture passes (crossed the swords), the muscle was less apt to slip while he hand-tied the suture. With braided synthetic sutures, and a trend toward using forceps for tying knots (thank goodness), there is really no need to overlap the suture passes. "Underlapping" them will still allow you to use your usual technique with only this minor modification, and will decrease the incidence of perforation in my opinion.

When I use a fornix approach, I use my usual suturing technique, e.g., two double-armed sutures in the muscle with suture passes toward the insertion that I fixated. I find the use of a double-armed 0.5 fixation forceps useful. It not only provides excellent stability of the globe while sutures are being passed, but it also serves to retract the conjunctiva and keeps the field exposed.

> Even if you like the fornix incision and sit at the head of the table, there is no need to "cross the swords" when suturing.




### Myth

If you use the fornix approach, you also must use the crossed-swords suturing technique with the assistant fixating the globe.

### Fact

With minor modification you can use the fornix approach and a suturing technique with which the surgeon fixates the globe and passes sutures toward the forceps fixating the insertion.

### Basic Information

*Treatment of Recognized Scleral Perforation*

Most people agree that local treatment of a scleral perforation with cryotherapy is probably not necessary. The very low incidence of retinal detachment after strabismus surgery compared to the much higher incidence of scleral perforation, combined with the fact that many perforations are not recognized until discovered much later on routine fundoscopy, suggests that not treating is an acceptable course to follow. On the other hand, placing a spot of cryotherapy over the perforation site probably carries very little risk and doing so

could not be criticized, and I recommend doing so in patients at particularly high risk for retinal detachment. If you suspect that a possible perforation has occurred, you should inspect the retina with the indirect ophthalmoscope. The risk of endophthalmitis is higher than the risk of retinal detachment after scleral perforation. I recommend systemic antibiotics postoperatively in cases of scleral perforation recognized at the time of surgery.

### Important Point

If you suspect that a scleral perforation has occurred during strabismus surgery, confirm with indirect ophthalmoscopy. If it has indeed occurred, consider systemic antibiotics to prevent endophthalmitis. Cryotherapy may not be needed except in high-risk patients.

### Basic Information

*Disinserted Muscles, Slipped Muscles, and Elongated Scars*

These are three different scenarios that have in common the fact that the muscle did not form a proper bond at the expected scleral site. They each have different causes and somewhat differing clinical presentations.

## Disinserted Muscles

Parks used this term to describe muscles that form a healthy bond to the sclera, but in a somewhat posterior position to that which was intended [6]. Its causes are probably multifactorial, including imprecise measuring for a recession—in which case the muscle did not actually disinsert but may be thought to have done so, premature suture breakdown or suture breakage early after surgery, or possibly trauma. Clinically, a disinserted muscle will present with an overcorrection if it occurred on a recessed muscle, or undercorrection if on a resected muscle, which should be evident within several weeks after surgery. Depending on how far the muscle slipped, there may be only mild underaction of the affected muscle. At reoperation, the muscle and its attachment to the sclera may look normal, with the only anomaly being the location of the insertion site, which is posterior to what was expected. Orbital imaging typically does not show anything helpful other than ruling out the other problems discussed in this section. It is not helpful in identifying where the muscle is actually inserting, because usually it inserts anterior to the point the muscle starts wrapping around the globe on the orbital image. The actual insertion site is not discernable. Although this is often referred to as a slipped muscle, I like Parks' term "disinserted muscle" to distinguish it from a muscle slipped in its capsule.

## Slipped Muscles (Also Known as Slipped in the Capsule)

This phenomenon presumably occurs if the sutures are not adequately placed in muscle tendon itself, but are just threaded through the muscle capsule. See Chap. 10 for a detailed description. Dynamic orbital imaging shows a characteristic appearance that will distinguish a slipped muscle (Fig. 11.8) from a disinserted muscle. On attempted movement into the field of action of a slipped muscle the posterior aspect of the muscle shows a fusiform thickening indicating contractility; however, anteriorly the empty capsule does not thicken. This dynamic scan appearance is indistinguishable from an elongated scar and can also be similar to the appearance of the *collapsed empty channel sign* of a lost muscle (see Fig. 11.7). This distinction must be made clinically. A lost muscle will typically have a marked degree of underaction (−4 or −5), whereas slipped muscles and muscles with elongated scars will usually show some force.

Parks speculated that slipped muscles are occurring more frequently now as a result of finer suture material and needles [6]. He felt that

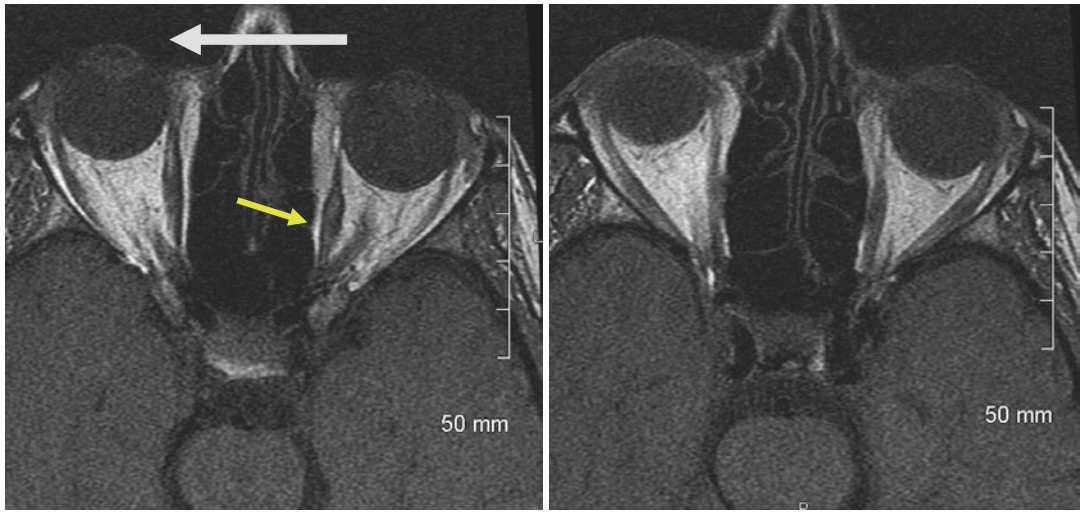



**Fig. 11.8** Dynamic MRI of a right medial rectus muscle (RMR) slipped in its capsule. In the image on the right, the patient is in right gaze. The RMR looks unremarkable. In the image on the left, the patient is attempting to look in left gaze. With this attempt at adduction OD, there is a fusiform swelling of the posterior belly of the RMR (*small yellow arrow*); however, anteriorly there is thin empty capsule which does not show contractility

it was harder to just pass a suture through capsule when the suture material and needles were larger than those available today. I believe that some steps can be taken to minimize the occurrence of this complication. If you use a one-suture technique, placing a small central full-thickness suture pass in the middle of the muscle at the insertion, and securing that with a square knot, will be the best preventative measure. If the bite is of full thickness, you can be sure that you have included tendon or muscle. Then be sure that the pole lock bites are wide enough to incorporate muscle and not just capsule. Finally, after the muscle is disinserted, reflect it back so that you can visualize the undersurface of the insertional end. If your sutures are not incorporating muscle, you will see the center of the muscle beginning to slip posteriorly (see Fig. 10.6). I do this routinely, and if I see slippage, I reinforce the center of the muscle as shown in the figure.

## Pearl

Slippage within the capsule can be prevented by placing a central full-thickness bite at the center of the muscle at the insertion, and by inspecting the undersurface of the muscle after it is disinserted. Reinforce the center if the muscle is not secure.

## Elongated Scars (Also Known as Stretched Scars)

Ludwig has described a wound healing issue in which a connective scar tissue bridge forms between the cut end of the muscle and the sclera, resulting in an indirect attachment of the muscle to the sclera that elongates over time [10]. She points out that scar tissue will elongate when under tension. See Chap. 10 for a detailed description.

## Basic Information

*Eyelid Changes from Strabismus Surgery*

Both the upper and lower eyelids can be affected by strabismus surgery on the SR or IR, respectively.

## Lower Eyelid Retraction

Because of the connection of lower eyelid retractors to the capsulopalpebral head, lower eyelid retraction can occur after IR recessions that exceed 4 mm. This can be a significant cosmetic problem, particularly if it occurs unilaterally. Complete dissection of the check ligaments and dissecting to and through Lockwood's ligament can minimize this complication. Intraoperatively, you can determine if the dissection has been adequate to prevent lower eyelid retraction with this simple maneuver. Grasp the eye with a fixation forceps above the superior limbus and rotate the eye into upgaze. Then perform a passive duction into extreme downgaze while observing any change in the position of the lower eyelid. It helps to use a piece of suture material held parallel to the lower eyelid as a reference point to sight for a change in eyelid position. If the connections have been adequately severed, there will be no change in the position of the lower eyelid between upgaze and downgaze and you should not get lower eyelid retraction postoperatively (Fig. 11.9). Care must be taken to avoid making this assessment while the curvature of the cornea alters the lower eyelid position. Depress the eye sufficiently that the superior limbus is below the lower lid margin (that is why you should grasp the eye well above the superior limbus). I call this the "string test." However, I have found that particularly in patients with TED, it is very hard to reach this endpoint of no lower eyelid movement on vertical passive duction. In cases of TED, or any case in which I am performing more than a 4 mm IR recession and cannot eliminate lower eyelid movement on passive duction, I will advance the capsulopalpebral head [11]. The procedure consists of dissecting the tissue overlying the IR back to the attachment of the capsulopalpebral head, which is typically about 15 mm posterior to the limbus, securing it with an absorbable suture, completing the dissection through Lockwood's ligament, recessing the IR, and then suturing the capsulopalpebral head to the IR at its original distance from the limbus (Fig. 11.10). This advances the capsulopalpebral head referable to its attachment to the IR. The procedure does not completely eliminate lower eyelid retraction, but was found to decrease its magnitude by about 50%.

The "string test" done intraoperatively can be useful in determining if you have adequately dissected the lower lid retractors

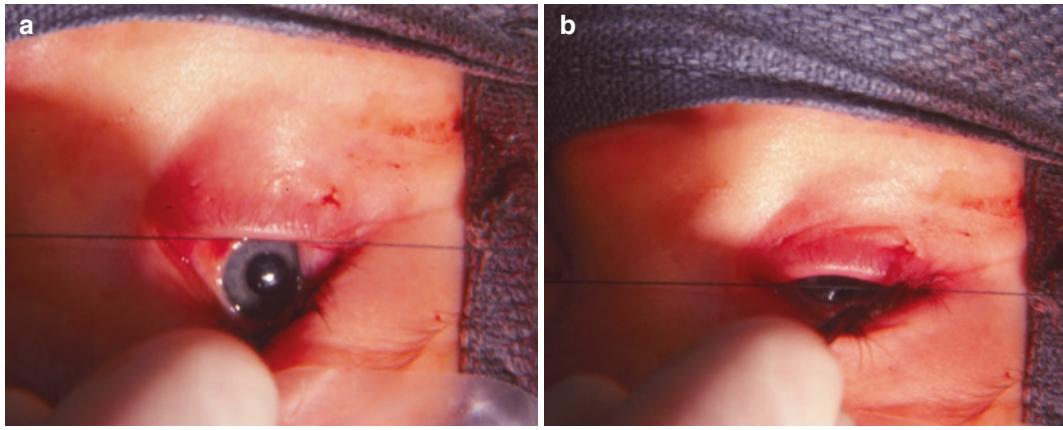



**Fig. 11.9** String test: (**a**) Surgeon's view of patient undergoing large IR recession for idiopathic inferior rectus muscle (IR) fibrosis. The eye is held in supraduction and a string or piece of suture material is positioned horizontally level with the lower eyelid margin. (**b**) The eye is infraducted with forceps. One can see that the lower eyelid has moved inferiorly several millimeters in reference to the string. This indicates that the dissection of the lower eyelid retractors was incomplete, and lower eyelid retraction would occur if a large IR recession is performed

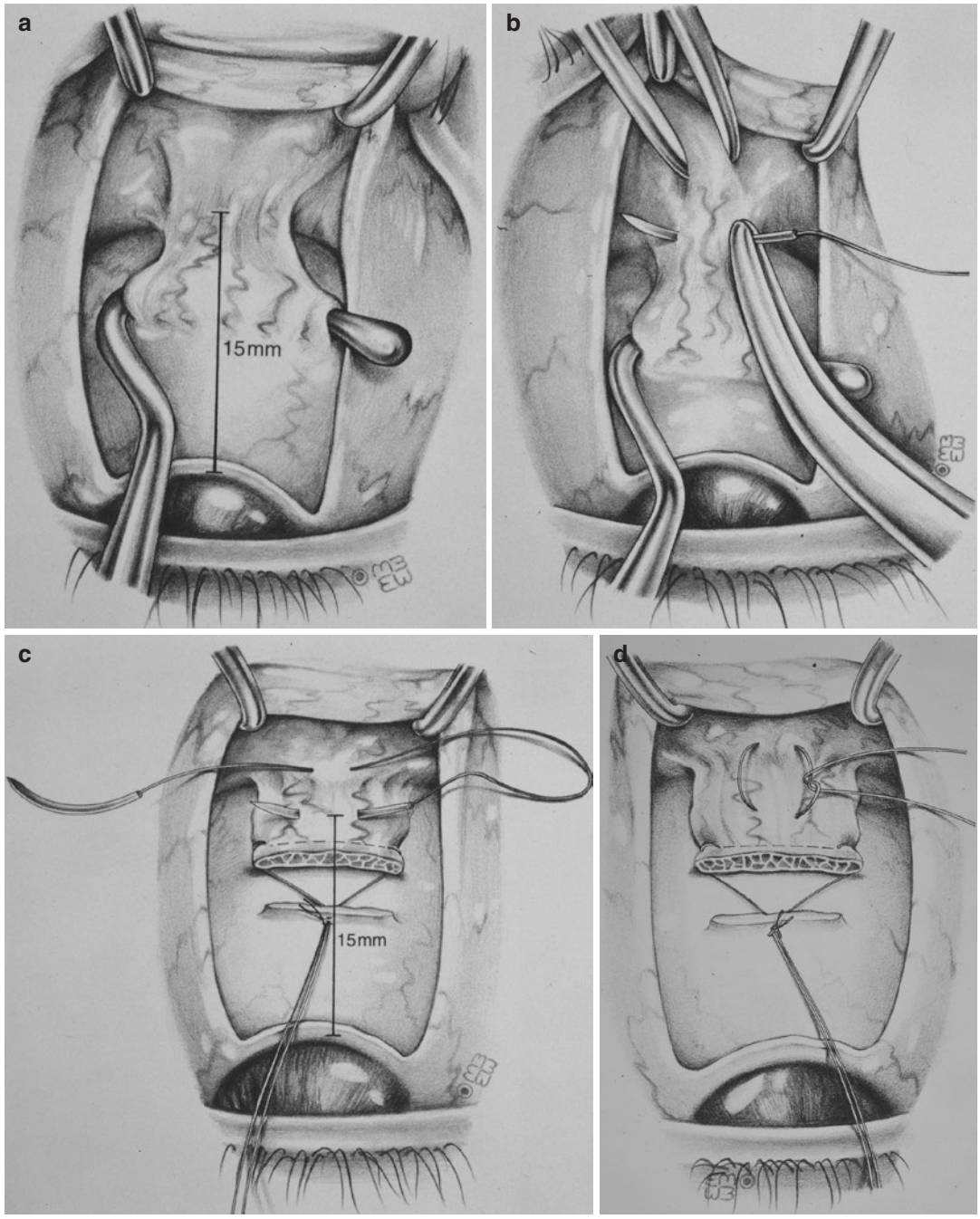



**Fig. 11.10** (**a**) Dissection of the IR showing the attachment of the capsulopalpebral head to the inferior rectus muscle (IR) approximately 15 mm posterior to the limbus. (**b**) A double-armed absorbable suture is placed through the capsulopalpebral head in horizontal mattress fashion. (**c**) The IR has been recessed, and one arm of the suture is then placed in horizontal mattress fashion though the IR approximately 15 mm posterior to the limbus. (**d**) The suture is tied uniting the IR to the capsulopalpebral head 15 mm from the limbus

### Lower Eyelid Elevation with Palpebral Narrowing

This can occur with IR resection and IO anterior transposition. It usually does not occur with IR resections that are less than 4 mm but typically does occur whenever the IO is transposed to the level of the IR insertion or further anterior [12]. In addition, IO anterior transposition will cause a characteristic bulge of the lower eyelid on upgaze and an alteration of the lower eyelid curvature. If these lid changes happen symmetrically, they are usually not a serious cosmetic issue. However, if unilateral, it can be quite unacceptable cosmetically. This is one reason I never perform IO anterior transposition unilaterally, except in unusual circumstances.

### Upper Eyelid Retraction

This can occur with large SR recessions, but can usually be avoided with careful severing of the connective tissue attachments around the SR, even with recessions that exceed 10 mm. However, I have seen significant upper eyelid retraction occur when large SR recessions for treating dissociated vertical divergence were followed by IO anterior transpositions for recurrent dissociated vertical divergence. After the IO procedure there was a marked limitation of elevation bilaterally. Upper eyelid retraction occurred due to fixation duress just to bring the eyes to the midline position [12].

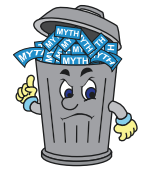
### Upper Eyelid Ptosis

This is the flip side of upper eyelid retraction from SR recession. Resection of the SR can result in ptosis if the connective tissue attachments around the SR are not adequately severed. It can usually be prevented with careful technique.

## Basic Information

*Intractable Postoperative Diplopia*

Certainly there are situations in which postoperative diplopia may be predictable and not necessarily intractable. For example, if an adult with exotropia is surgically overcorrected, you can expect them to be diplopic. Similarly, if a patient with a fourth cranial nerve palsy is overcorrected, diplopia is common. These are examples of surgery that did not successfully result in the targeted postoperative alignment, and the treatment is to realign the eyes either optically (with prisms) or surgically. But I am discussing here the patient who ends up with exactly the alignment you desire, but has a persistent diplopia that does not resolve. There is a common misperception on the part of optometrists and some primary care practitioners that this type of intractable diplopia frequently results from strabismus surgery in adults [13]. In fact I have documented it to be quite rare [14]. Specifically, it occurred in only 3 of 424 (0.8%) adults undergoing strabismus surgery.




### Myth

Intractable diplopia is common after surgery in adults with long-standing constant strabismus.

### Fact

Intractable diplopia is exceedingly rare in this setting. Moreover, there is testing that has 100% sensitivity and 100% negative predictive value for this adverse outcome. See Chap. 2, "The Examination," for details.

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# Thyroid Eye Disease

# 12

## General Concepts

### Basic Information

*Nomenclature*

When the extraocular muscles (EOMs) and orbital contents are affected in patients with clinical or subclinical thyroid disorders, a variety of terms have been used to describe the ocular condition. These include *Graves' disease, Graves' orbitopathy, endocrine ophthalmopathy, thyroid ophthalmopathy,* and *thyroid eye disease* (TED). It is also called thyroid-associated eye disease, because it can occur in patients who are euthyroid or have hypothyroid chronic autoimmune thyroiditis. I will use the term TED.

### Basic Information

*Pathophysiology*

In TED there is often an acute congestive phase characterized by edema in the orbit and EOMs. The muscle fibers are intact, but there is an accumulation of collagen and glycosaminoglycans with an inflammatory mononuclear cellular infiltration. This matrix is very hydrophilic, causing the muscle to swell greatly. This is followed by an inactive chronic phase in which the EOMs become fibrotic and inelastic. In a sense, the chronic inactive phase is that of a muscle compartment syndrome—edema leads to cell death and fibrosis. It is noteworthy that the EOMs are tight, but also weak. They have an increase in elastic force, but a decrease in contractile force.

### Basic Information

*Presentation and Systemic Evaluation*

Patients with TED typically present to the strabismologist in one of the three ways. On occasion they present with strabismus during the acute congestive phase, in which case the strabismus may resolve after the congestive phase has passed. A patient may present with restrictive strabismus along with obvious signs of Graves' disease, such as proptosis, lid lag, decreased blinking, and a history of clinical thyroid disease. In this case the diagnosis of TED is obvious. Finally, some patients present with restrictive strabismus with no other signs or symptoms of thyroid disease, in which case the diagnosis is less obvious. In these patients, orbital imaging is helpful by revealing the characteristic eye muscle hypertrophy that accompanies TED (Fig. 12.1). TSH receptor antibody is relatively sensitive and correlates with clinical features and prognosis. I also check


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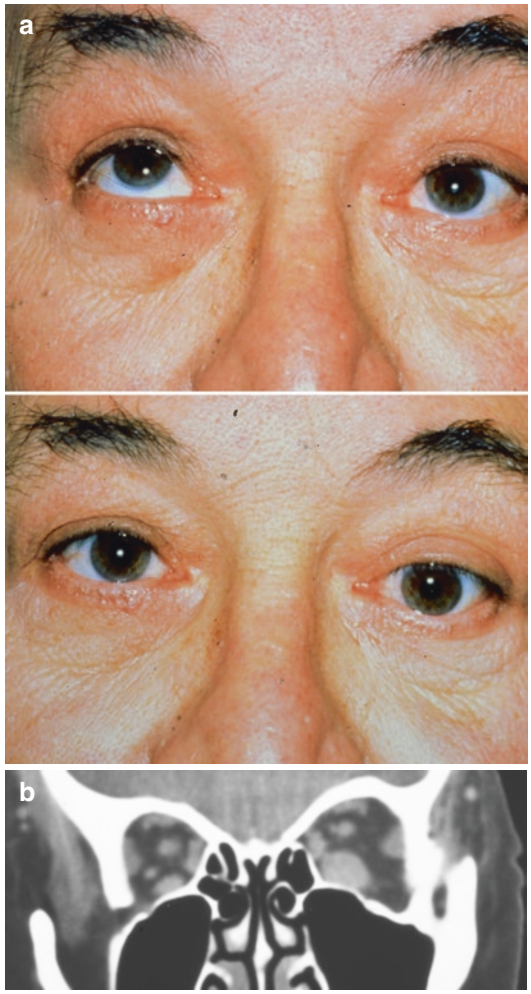


**Fig. 12.1** (**a**) Patient with a restrictive left hypotropia secondary to thyroid eye disease (TED). The patient has no other external eye signs to suggest TED. (**b**) Coronal imaging of the orbits of the patient shown in Fig. 12.1a shows hypertrophy of the left inferior rectus muscle characteristic of TED

thyroid function, with the awareness that TED can occur in patients who are hyperthyroid, hypothyroid, or euthyroid. I obtain T3, T4, and TSH, and if normal might obtain a TSH in response to thyrotropin-releasing hormone (TRH). Virtually all patients with TED have anti-thyrotropin-receptor antibodies, which suggests an autoimmune mechanism. The severity of the eye disease positively correlates with the antibody titer [1]. Cigarette smoking is the strongest modifiable risk factor.

Cigarette Smoking is the Strongest Modifiable Risk Factor.

## Basic Information

*Muscle Involvement*

A patient with TED may have involvement of one or multiple muscles in one or both eyes. Multiple muscle involvement and bilaterality are more likely to occur in patients who are clinically hyperthyroid than euthyroid. Prior orbital decompression predisposes to multiple muscle involvement, particularly if multiple orbital walls were decompressed. In one series of 53 patients [2], slightly more than half had bilateral involvement (Table 12.1). In that series, the inferior rectus (IR) muscle was by far the most commonly involved, followed by the medial rectus muscle (Table 12.2). Although the lateral rectus muscle is sometimes affected on orbital imaging, in my experience it is uncommon for it to be clinically an issue. Similarly, it is relatively uncommon for the oblique muscles to be clinically involved.

## Important Point

Although it is not uncommon for TED patients to have torsional symptoms, this is usually secondary to rectus

**Table 12.1** Number of muscles per individual patient in a series of patients with thyroid eye disease

| Muscles per individual patient (no.) | Patients $N = 53$ |
|---|---|
| *Unilateral* | |
| 1 muscle | 20 |
| >1 muscle | 5 |
| *Bilateral*[a] | |
| 2 muscles | 9 |
| > 2 muscles | 19 |

[a]Represented as total muscles for both eyes (i.e., one muscle each eye would be represented as 2 muscles total (data from Kushner [2])

**Table 12.2** Muscles clinically involved in a series of 53 patients with thyroid eye disease

| Muscle | Muscles (no.)[a] |
|---|---|
| Inferior rectus | 56 |
| Medial rectus | 31 |
| Superior rectus | 18 |
| Lateral rectus | 0 |

[a]Total is greater than 53 because some patients had multiple muscle involvement. A muscle was counted twice if it was affected in both eyes (data from Kushner [2])

muscle involvement. For example, a tight IR will not only cause a relatively small excyclotropia on its own, but it will also result in fixation duress to the inferior oblique muscle on attempted upgaze, resulting in more extorsion. In most cases releasing the rectus muscle restriction will correct the torsion.

## Pearl

If you suspect significant oblique muscle involvement, rotary forced ductions at the time of surgery can be helpful in making that diagnosis. Grasp the eye at the 3 and 9 o'clock positions (or 6 and 12 o'clock) and rotate the eye clockwise and counterclockwise. Look for symmetry of rotation (see Fig. 10.10).

## Basic Information

*Clinical Diagnosis of Strabismus*

The hallmark of strabismus with TED is restriction. There is typically a limitation of rotation(s). Forced ductions are crucial in determining that a restriction is present. If there is an acquired hypotropia (HYPO) with a limitation of elevation, the absence of Bell's phenomenon is helpful in diagnosing restriction in patients who are not cooperative for forced duction testing in the awake state. Active force generation, an underutilized yet easy-to-perform test, will help rule out paresis. My preferred technique for performing this test, which is a modification of original method, is shown in Fig. 2.5. I find that by having the patient first look in the direction of the gaze limitation, and then my pulling against the isometric contraction of the muscle in question, there is less likelihood of tearing conjunctiva than if I grasp the eye and then have the patient attempt a duction; this technique is also easier for the patient.

## Advanced Information

*Timing of Surgery*

Classic teaching is to wait for 6 months with no significant change in alignment before operating. Operating while the disease is still active can lead to not only an unstable outcome, but also a severe inflammatory reaction [3]. Waiting, however, can be extremely difficult for patients. In my experience, patients with TED are often referred to me for surgery by comprehensive ophthalmologists or optometrists without ever having had their deviation measured or their ductions quantified. The patient is only aware that they have had diplopia for many months, and are not able to comment on the stability of their motility. It is extremely disconcerting to them to have to wait another 6 months before having surgery, particularly since they may in fact have been stable. An unpublished pilot study I have done may provide a solution for this dilemma. T2-weighted magnetic resonance image (MRI) will show muscle edema, which may likely be a marker for active disease and instability (Fig. 12.2a). The study was prompted by a patient I saw in consultation from a pediatric ophthalmologist. Clinically, she had a tight left IR and left HYPO, but had been stable for 6 months (Fig. 12.2b). Orbital imaging as shown in Fig. 12.2a, however, showed that there was still edema in the muscles. Because of this I recommended deferring surgery; however,

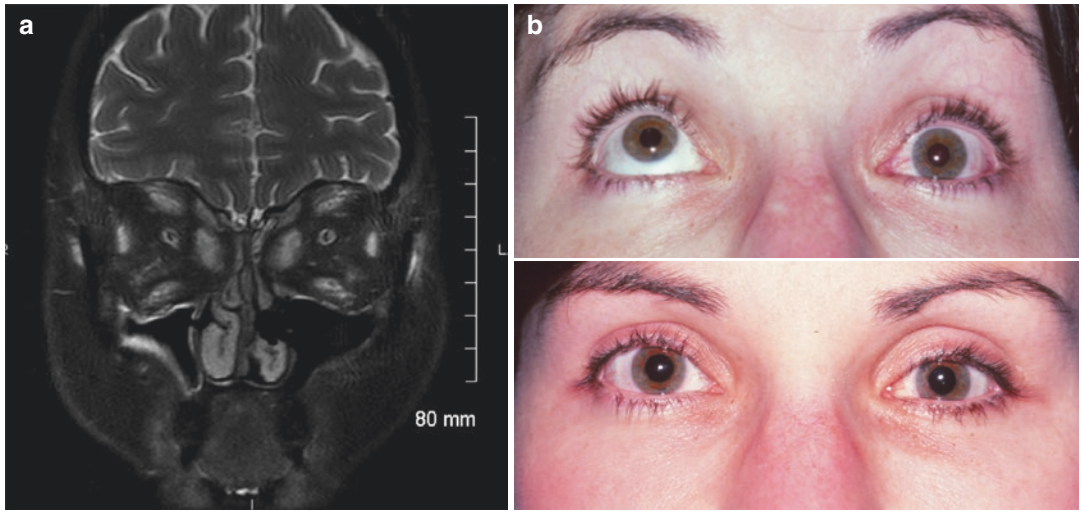


**Fig. 12.2** (**a**) T2-weighted MRI of the orbits of a patient with thyroid eye disease (TED). Edema shows up as a white coloration to both lateral rectus muscles and left inferior rectus muscle, suggesting an active phase to the TED. (**b**) Motility of patient whose MRI is shown in Fig. 12.1. It shows a left hypotropia with limitation of elevation

because she had been stable the referring doctor proceeded with a LIR recession. Six weeks after surgery her alignment was good (Fig. 12.3), but by 12 weeks her left HYPO had returned to its preoperative size (Fig. 12.4). Prompted by this patient, I began a retrospective pilot study in which I reviewed my patients with TED in whom I had obtained an) MRI and observed them for at least 6 months prior to operating. I wanted to see if the presence of edema on MRI was predictive of instability during the waiting period. Of the nine patients who met the criteria, five showed no edema on MRI, and all five remained stable. The remaining four patients showed edema on MRI, and all four had changing motility during the subsequent 6 months, characterized by either a change in their primary position deviation by 5 prism diopters (Δ) or more or a change in ductions by more than one unit (scale −4 to +4). This suggested to me that the absence of edema on T2 MRI may provide a green light for performing surgery; however, this hypothesis definitely needs to be tested in a larger series.

The absence of edema on T2-weighted MRI may be a green light that it is OK to perform surgery.

## Basic Information

*Surgical Planning—Choice of Muscles*

Above all, a successful outcome relies on a proper choice of muscles to operate upon, and in TED there are some common patterns that may make that choice unclear.

In most cases of TED, one eye is preferred for fixation and the other eye habitually

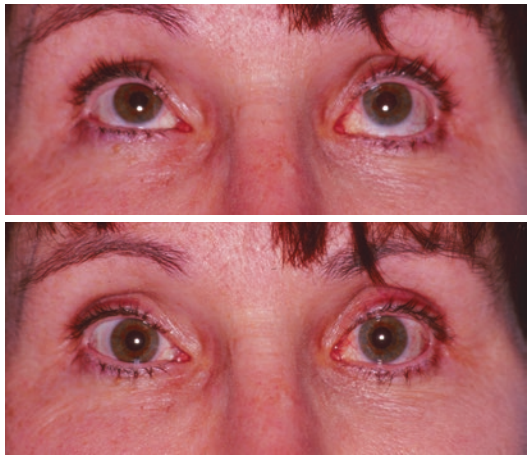

**Fig. 12.3** Motility of patient whose magnetic resonance image is in Fig. 12.2b, 6 weeks after undergoing a LIR recession. It shows elimination of the primary position L hypotropia and good elevation OS

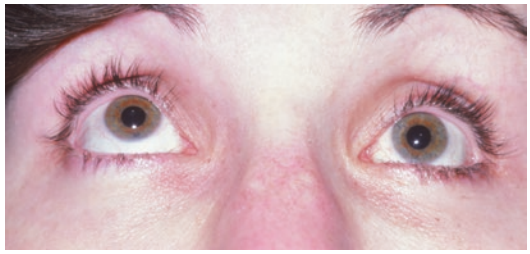

**Fig. 12.4** Motility of patient in Fig. 12.2b shown 12 weeks after the left inferior rectus recession, and 6 weeks after photographs in Fig. 12.3 were taken. Her left hypotropia has returned to its original level. A photo of her in the primary position is not available, but this figure shows that the limitation of elevation OS has returned to its preoperative degree

deviates. A common error is to overlook lesser degrees of involvement of muscles in the fixing eye, if careful assessment is not made of ocular rotations in all fields of gaze. This is particularly likely to occur if there is involvement of the opposite rectus muscle of the one that is primarily affected, in the other eye. For example, the patient in Fig. 12.5 has TED, fixes with the left eye, and manifests a right hypertropia (HT) with limited depression OD, secondary to a tight right superior rectus muscle (SR). The LIR is tight to a lesser degree than the RSR, resulting in limitation of elevation of the left eye. Failure to recognize the involvement of the LIR and not recessing it, recessing only the RSR, will predictably lead to a surgical overcorrection. Careful attention to ductions, versions, and forced ductions is crucial to avoid overlooking this pattern.

A second pattern that is easy to overlook is involvement in the fixing eye of the same muscle that is obviously involved in the non-fixing eye. The patient in Fig. 12.6 was referred to me for an opinion regarding a 5 Δ L HYPO secondary to a tight LIR associated with TED. The referring doctor planned to just recess the LIR. One and a half years prior to her presenting to me, she had a left HYPO of 25 Δ according to prior records. The patient and the referring ophthalmologist had assumed that the decrease she was experiencing in the size of the HYPO indicated improvement, and the deviation had remained at 5 Δ of L HYPO for over 6 months. In fact, what they thought was improvement was the result of progressive contracture of the IR in the right eye. The primary position HYPO had improved, but her upgaze had worsened bilaterally and she was developing an increasing chin-up head posture. When I examined her there was bilateral but asymmetric tightness of the IRs, which in part is why the HYPO was so small. Failure to recognize that the same muscle (IR) is involved in

A decrease in the size of the primary position hypotropia may be an indication of progressive bilateral involvement.

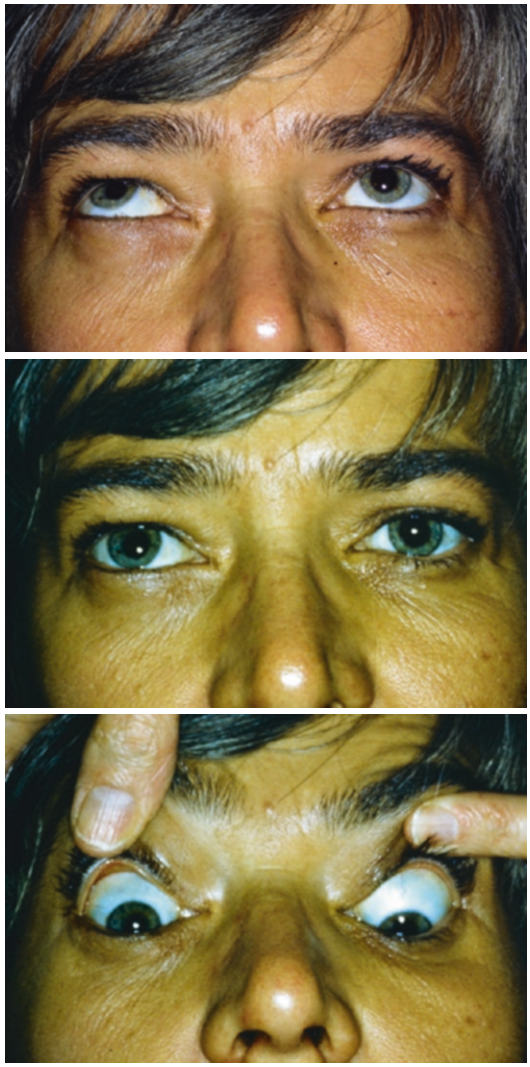

**Fig. 12.5** Patient with thyroid eye disease manifesting right hypertropia in primary gaze with limited depression of the right eye due to a tight right superior rectus muscle. There is also a lesser degree of restriction of the left inferior rectus muscle with limited elevation OS. Due to some degree of orbital asymmetry the gestalt from these photographs is that the left eye elevates further than the right. But assessment of the light reflexes confirms that there is a left hypotropia in upgaze, which is also found with cover testing (from Kushner [2], with permission)

both eyes, and omitting recessing *both* IRs, would invariably lead to a surgical overcorrection. She was treated with bilateral asymmetric IR recessions which eliminated her diplopia and chin-up head posture, and her upgaze improved (Fig. 12.7).

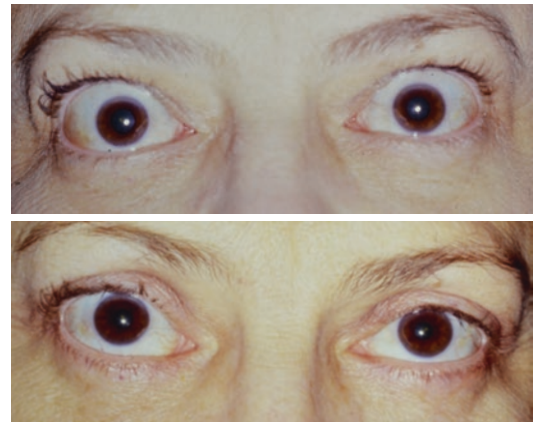

**Fig. 12.6** Patient with thyroid eye disease manifesting a 5 Δ L hypotropia secondary to a tight left inferior rectus muscle (*bottom*). However, there is limitation of elevation of both eyes, indicating that there is also a tight right inferior rectus muscle (*top*). The top photo shows her in maximum upgaze. Note the upper eyelid retraction on attempted upgaze, indicating maximum upgaze effort

A third pattern that is easy to overlook is the patient who habitually fixates with the affected eye. The patient in Fig. 12.8 has TED and a right HT. She was suspected of having a tight RSR. In fact, she has normal depression in the right eye. Her right eye has a best corrected acuity of 20/200 secondary to untreated amblyopia. She fixates with her 20/20 left eye, which has a tight IR.

## Important Point

Whenever a patient has unequal best corrected visual acuity, consider the possibility that they might be fixating with the involved eye.

## Basic Information

*Surgical Pointers*

The surgical treatment of TED usually involves recessing the restricted muscles. Traditionally, it has been taught that resections should categorically be avoided in

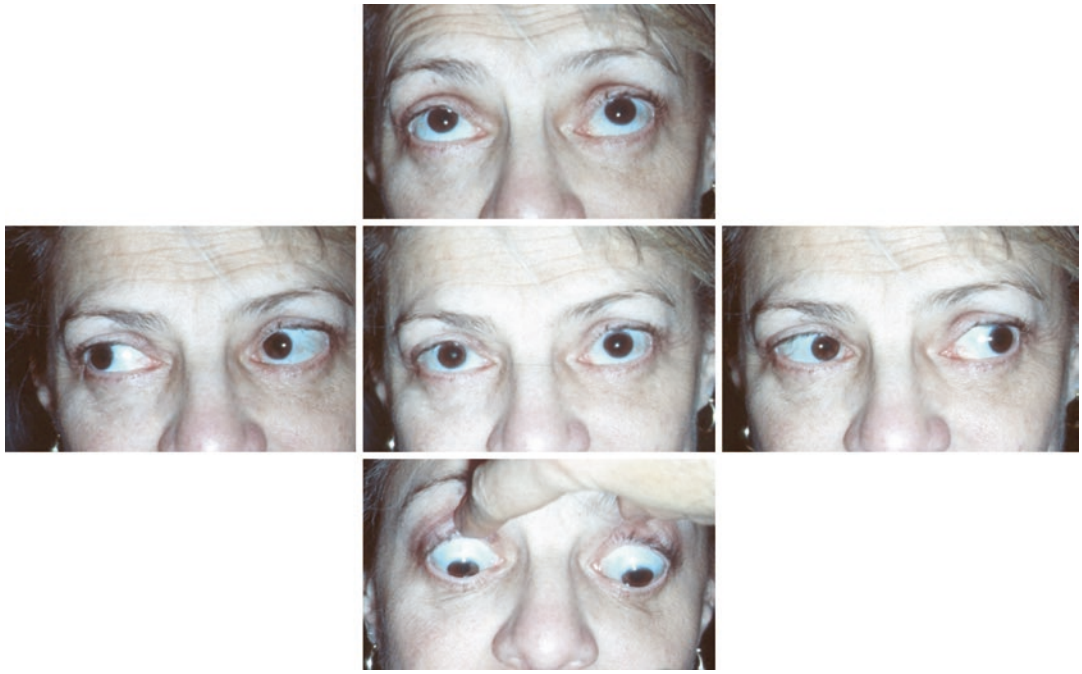

**Fig. 12.7** Patient in Fig. 12.6 after bilateral inferior rectus recessions, showing good alignment in primary gaze with improved elevation

TED, because there is a tendency for a marked inflammatory reaction after a muscle resection. More recently, some experts have been advocating small resections in select cases. I, however, do not feel that they are necessary and avoid resections.

The amount of recession needed is not formula driven in TED, but should be an amount needed to release the restriction. Determining how much that should be is a matter of experience. However, Traboulsi popularized a technique by which the amount of recession is determined by suturing the muscle to the globe where the disinserted muscle naturally fell when the eye is held in the primary position [4, 5]. A similar technique was described by Harper in 1978 [6]. Although I do not have experience with that technique, I have determined that in many cases that technique of positioning is very close to where I recessed the muscle relying on my own experience with muscles in TED. For someone without a lot of experience with TED, that technique would be a reasonable approach in my opinion.

## Advanced Information

*Adjustable Sutures*

I am enthusiastic about the use of adjustable sutures in TED, with several caveats. Muscles in TED are tight but not necessarily elastic. In some, the muscle retracts briskly after disinsertion, in which case adjustable sutures pose no special problem. In others, the inelastic muscle retracts only a few millimeters after disinsertion, and will not take up any more slack. In these cases, standard adjustable sutures may not work well, as the muscle will not retract if needed. In such patients, the use of the semi-adjustable suture as shown in Fig. 10.15 will obviate that problem. The scleral fixated corner sutures with that technique will insure that the muscle will stay in the desired recessed position.

EOMs in TED are tight but often inelastic. They may not take up slack when disinserted.

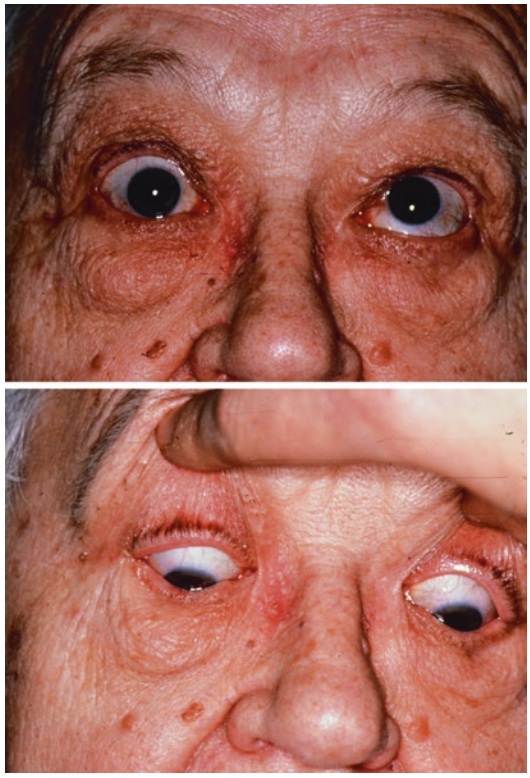

**Fig. 12.8** Patient with thyroid eye disease and a large left hypertropia, which was thought to be due to a tight left superior rectus muscle (*top*). She has normal depression in the left eye (*bottom*). Her right hypertropia is due to a tight right inferior rectus muscle, and the fact that she fixates with her right eye

## Advanced Information

*Muscle Slippage*

The IR is notorious for slipping after recession in patients with TED. Two factors contribute to this. Scott pointed out that there is more and thicker Tenon's layer on the undersurface of the IR than with other muscles. If it is not removed, it can prevent adherence of the muscle if a suspension or an adjustable suture technique is used (Alan B. Scott, MD, personal communication, 4 Feb 2016). The second major reason is that the IR has a much shorter arc of contact with the globe. Orbital imaging shows that after a modest IR recession, the muscle would lose apposition to the globe when the eye is in downgaze if it is not fixed to the sclera (Fig. 12.9) [7]. The use of a semi-adjustable suture technique will obviate both of these causes of muscle slippage and is my preferred technique. Others like to use nonabsorbable sutures and a standard adjustable suture technique. I am less partial to that approach, as the knot frequently erodes and causes irritation.

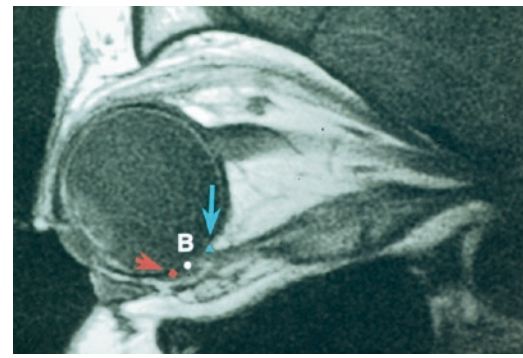


**Fig. 12.9** Sagittal MRI of the orbit of a patient with thyroid eye disease 1 day after a 5 mm inferior rectus recession using a semi-adjustable suture technique, taken with the eye in downgaze. The *orange arrow and diamond* indicate the point of tangency of the muscle prior to recession determined by where a line drawn from the origin of the muscle is tangent with the globe. Point B is the post-recession new insertion. The *turquoise triangle and arrow* denote where the global layer (post-recession) loses contact with the sclera. Both Point B and the *turquoise triangle* are well posterior to the point of tangency, and thus there is a complete lack of wraparound effect of the muscle with the globe. If the muscle had not been fixated to the sclera, it would lose apposition to the globe (from Chatzistefanou et al. [7], with permission)

## Basic Information

*Postoperative A-Pattern*

It is not uncommon for patients with TED to need to undergo large bilateral IR recessions. This can result in a postoperative A-pattern. This occurs because weakening both IRs causes fixation duress to the superior oblique muscles, resulting in their "overacting" and causing an A-pattern. Also, there is a postoperative loss of the adducting power of the IRs. Whenever you are doing bilateral IR recessions of 6 mm or

more, you should consider doing something prophylactically to prevent an A-pattern. Some people routinely nasally transpose the IRs to prevent the A-pattern. This, however, worsens the intorsion that accompanies this problem. Others temporally transpose the IRs to counteract the expected intorsion. This, however, worsens the A-pattern. I have seen patients that have had adverse outcomes after both of these approaches [8]. To my mind, the only approach that will prevent the A-pattern, yet not cause an incyclotropia, is to weaken the superior obliques, with posterior tenectomies being my procedure of choice in this setting. See Chap. 21, "Complicated Strabismus," Case 21.13 on page 320, for a representative example of these principles.

If recessing both IRs 6 mm or more, anticipate and act to prevent an "A"-pattern.

## Important Point

Nasal transposition of the IRs to prevent an A-pattern will worsen the intorsion, and temporal transposition to prevent intorsion will worsen the A-pattern.

## Advanced Information

*Upper Eyelid Retraction*

Many patients with TED have upper eyelid retraction due to direct involvement of the levator, but others have it due to fixation duress from bilaterally tight IRs. To tell if a patient will have postoperative upper eyelid retraction, observe what happens to the retraction preoperatively in downgaze. If the retraction is due to fixation duress from tight IRs, the upper eyelid retraction will disappear in downgaze (Fig. 12.10). On the other hand, if it persists in downgaze it will probably persist after IR recessions.

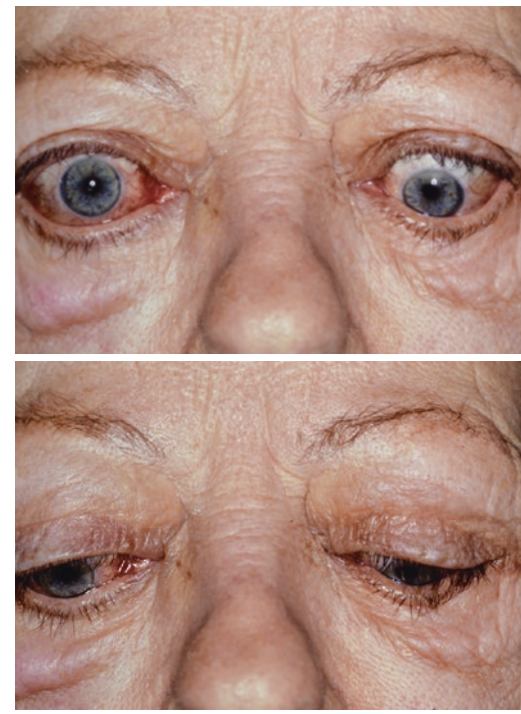

**Fig. 12.10** (*Top*) Patient with thyroid eye disease and bilateral upper eyelid retraction in primary gaze. (*Bottom*) The upper eyelid retraction is not present in downgaze, indicating that it was due to fixation duress

## Advanced Information

*Lower Eyelid Retraction*

Lower eyelid retraction is commonly present after large (greater than 4 mm) IR recession. In some cases, this is simply a reflection of proptosis and is not evident preoperatively when the eye is in downgaze. But in many cases, it is the result of the attachment of the capsule-palpebral head to the IR, which gets pulled back with recession [9]. Careful dissection to and through Lockwood's ligament is important. Finding a procedure to advance the capsule-palpebral head is helpful in

lessening, but not minimizing, postoperative lower eyelid retraction (see Fig. 11.11) [9].

### Pearl

Eyelid retraction is a hallmark of TED. There is an association between myasthenia gravis and TED. If ever a TED patient has ptosis, the patient should be worked up for myasthenia.

### Advanced Information

*Postoperative Diplopia*

A common scenario after surgery in TED is for there to be residual diplopia in downgaze when a unilateral IR recession was performed. The forces are no longer balanced for downgaze, with one IR being iatrogenically weakened [10]. Management of this scenario needs to be tailored to the visual and occupational needs of the patients. Solutions include slab-off prism in bifocals (see Fig. 9.2), single-vision reading glasses, Fresnel prism over the bifocal segment, occlusion of the bifocal unilaterally, or posterior fixation, with or without recession on the contralateral IR.

### Advanced Information

*Postoperative Instability*

A postoperative overcorrection that worsens with time suggests either a slipped muscle or a contracture of the antagonist of the recessed muscle. The usual tests for muscle weakness versus restrictions, e.g., forced ductions and active force generation, will be useful.

If ocular motility is satisfactory at 6 weeks after surgery, it is very uncommon for there to be a major change in motility unless there is a substantial change in the patient's thyroid status.

### Question

I have a patient with TED who has unusual motility. He fixes with his left eye and has an RHT of 75 Δ. What is odd is that his version seems pretty normal in the right eye and his left eye is limited to elevation. I am wondering if the RHT is due to fixation duress due to Hering's law. I am wondering if I should do a LIR recession combined with an RSR recession or RIR resection. Also, since he fixes with his left eye, would it be reasonable to just do a vertical recession and resection in the right eye?


### Reply

This does sound like classic fixation duress causing a big secondary deviation due to fixing with the affected eye. Here are some ways to sort it out:

1. Why does he fix OS? Is the acuity really poor OD? If he will continue to fix OS, incomitant strabismus can be hard or impossible to really fix. If there is a way to get him to fix OD (optically, cataract surgery, etc.), he will be better off.
2. Measure the deviation with him fixing OD and put all the prism base up over the left hypotropic eye. That will tell his real deviation, which is what you should target surgically. If the deviation is still really supersized, I strongly suspect some tightness of the RSR. Imaging should help confirm that. If it appears, after measuring in this manner, that you need to operate on two muscles, I opt for LIR and RSR recession, rather than a RIR resection.

### Question

My patient is a woman with TED. She has 25 Δ of left HYPO and limited upgaze OU, which I graded as −2 OD and −4 OS. With double-Maddox rod testing there is 5° of right excyclotropia and 15° of left

excyclotropia. There is also 4 Δ of esotropia (ET) in primary. She cannot fuse with the deviation offset with prism due to the torsion. An MRI showed an enlarged LIR. Can the torsion be due to the IR restriction, or do you think I need to do oblique muscle surgery? I was thinking of recessing the RIR 2 mm and the left 4 mm. Should I also address the ET with medial rectus muscle surgery?


## Reply

I have a few comments- Tight IRs in TED often cause excyclotropia, and in most cases releasing the IR restriction corrects the cyclotropia. However, if she had SO involvement from TED, the muscle would be tight and cause an *in*cyclotropia. I suppose if she had a nonrelated LSO palsy, that might contribute, or if she had TED involvement of the L inferior oblique. In any event, I would do rotary forced ductions in the operating room before and after you detach the IRs to test for a torsional restriction (see Fig. 10.10). Look for symmetry with incyclo- and excyclorotation. If really tight, you may want to look at the inferior oblique, but I have never had to do that in TED. Regarding the amount of IR recession, I do not think you should ever really go into a TED case with a predetermined number of millimeters to recess, as you need to rely on the forced ductions to determine the right amount of recession. But with 25 Δ in primary, I would anticipate needing to do about 3–4 mm more in the left than right, to collapse 25 Δ. Whether that ends up 5 mm and 2 mm, 6 mm and 3 mm, etc. will depend on what you find. Also, tight IRs will also cause restriction to abduction, and with only this small an ET, I would not do any medial rectus surgery.

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# How to Perform Superior Surgery on the Inferior Oblique and Avoid Inferior Surgery on the Superior Oblique

13

## Inferior Oblique

### Basic Information

*Errors in Diagnosis*

As stated in Chap. 8, "Vertical Deviations," not all causes of overelevation in adduction are a result of inferior oblique muscle (IO) "overaction" (OA). There are many causes of overelevation in adduction that are not caused by "OA" of the IO and will have an unsatisfactory response to surgical weakening of that muscle. These include pseudo-IO OA [1], *anti-elevation syndrome* (AES) [2], dissociated vertical divergence (DVD), *Duane syndrome*, pulley heterotopia [3, 4], inferior restrictions in the contralateral eye, and many cases with craniofacial syndromes.

### Basic Information

*Unintended Surgery*

Due to its anatomic proximity to the lateral rectus muscle (LR) insertion and the inferior rectus muscle (IR), the IO is particularly susceptible to being inadvertently damaged during surgical procedures on these other muscles.

### During LR Resection

When resecting the LR, special care must be taken to separate the LR from the IO insertion, or the IO can be dragged forward, resulting in the *IO adherence syndrome* [5]. The patient in Fig. 13.1 underwent a left LR resection as part of a surgical procedure to treat an esotropia. After surgery he had a left hypotropia (HYPO) with limitation of elevation in adduction and a left excyclotropia. Forced ductions were positive, and surgical exploration revealed the left IO to be scarred into the left LR near its insertion.

### Post-Scleral Buckle

If, at the time of a scleral buckling procedure, the surgeon inadvertently includes the IO on the muscle hook while hooking the IR, the encircling band may get passed between the reflected arms of the IO, resulting in the muscle being tucked (Fig. 13.2). The patient in Fig. 13.3 developed this restriction of elevation in adduction after a scleral buckling procedure. Subsequent surgery confirmed that the IO had been inadvertently tucked as depicted in Fig. 13.2. Obviously, the strabismus surgeon would not be the one responsible for creating this situation, but (h)she may be responsible for fixing it. It is important to be aware of this entity when operating on a patient with a restrictive HYPO and excyclotropia after scleral bucking surgery.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_13

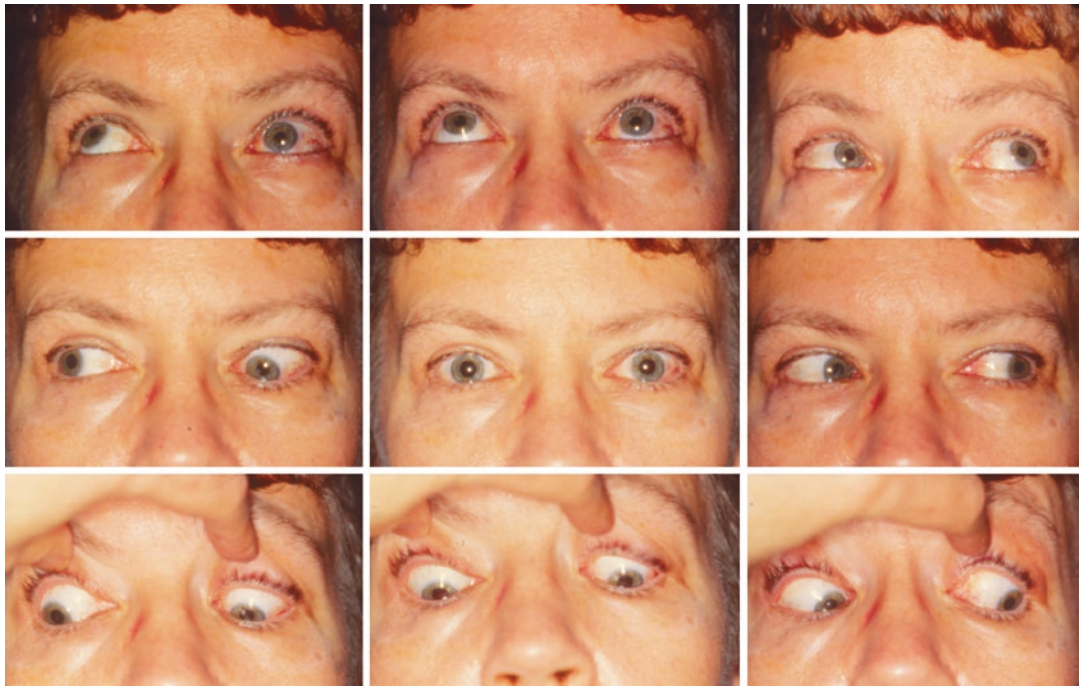

**Fig. 13.1** Left inferior oblique muscle incarceration syndrome after prior left lateral rectus muscle resection. There is a left hypotropia with limitation of elevation in adduction

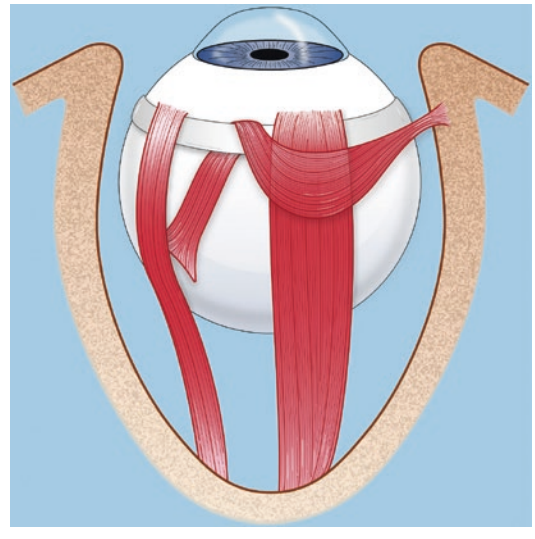

**Fig. 13.2** The manner by which the inferior oblique muscle can be inadvertently tucked by a scleral buckling encircling band (from Kushner [5], with permission, © 2007 American Medical Association. All rights reserved)

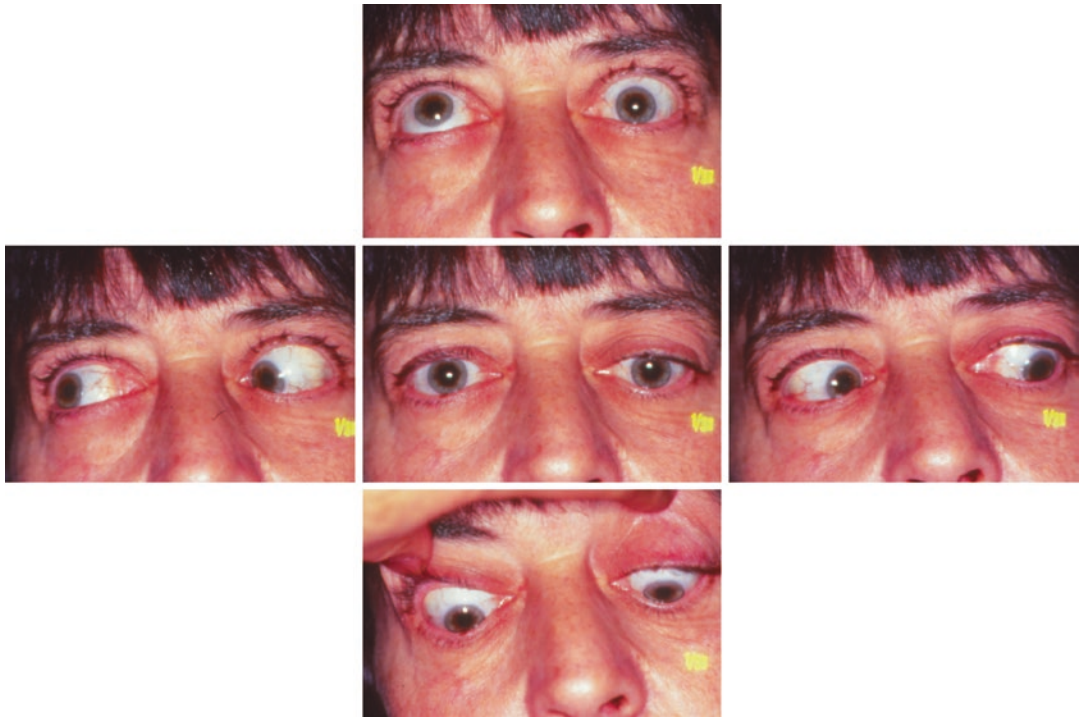

**Fig. 13.3** Patient who developed a limitation of elevation after prior scleral buckling in the left eye. Surgical exploration confirmed that the left inferior oblique muscle had been tucked by the encircling band as shown in Fig. 13.2

### After IR Resection

Because of the proximity of the IO to the IR via Lockwood ligament, the IO can be dragged forward if the IR is resected, much as can occur with a LR resection. Typically, this causes a restrictive HYPO and excyclotropia. Surgical exploration will reveal the configuration shown in Fig. 13.4. This can be prevented by taking special care to be sure that the IO stays back where it belongs when resecting the IR.

## Basic Information

*Technical Problems*

Although the IO is relatively forgiving with surgery, there are potential technical errors you should be aware of and avoid:

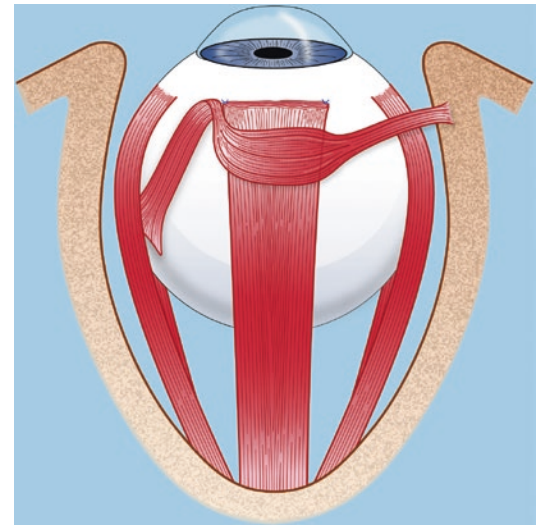

**Fig. 13.4** Depiction of typical configuration of the inferior oblique muscle being scarred into the inferior rectus muscle (IR) insertion after prior IR resection (from Kushner [5], with permission, © 2007 American Medical Association. All rights reserved)

### Incomplete Muscle Capture

Kraft and coworkers have reported that in as many as 8% of eyes the IO has a bifid insertion [6, 7]. This can lead to incomplete capture of the muscle when it is hooked. Incomplete capture can also occur with a normal IO if the tip of the hook is inadvertently passed through the muscle belly. There is usually a clear interface between the muscle itself and the orbital connective tissue. After passing the hook under the IO to capture it, tease the tissue back so that the tip of the hook is exactly at that interface (Fig. 13.5). Doing so will prevent violation of the fat pad. After the muscle has been captured and the tip of the hook bared, inspect the triangle between the two reflected arms of the IO (Fig. 13.6). Look carefully along the bare scleral triangle posteriorly to see if there are any missed muscle fibers. If there are, they will be seen running either horizontally along the sclera or in the reflected conjunctiva.

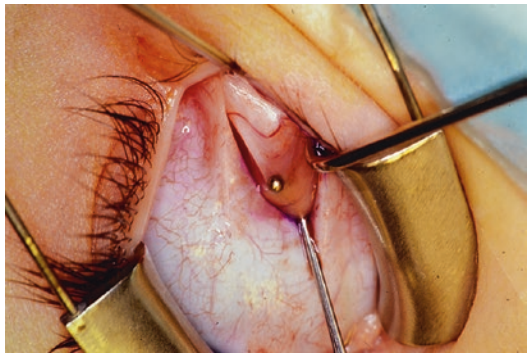

**Fig. 13.5** After the inferior oblique muscle is captured on the hook, tease the tissue so that the tip of the hook is exactly at the interface of muscle and connective tissue, as shown above, and then cut down on the tip of the hook or pop it through the interface to complete the muscle capture. This will prevent inadvertently violating the fat pad

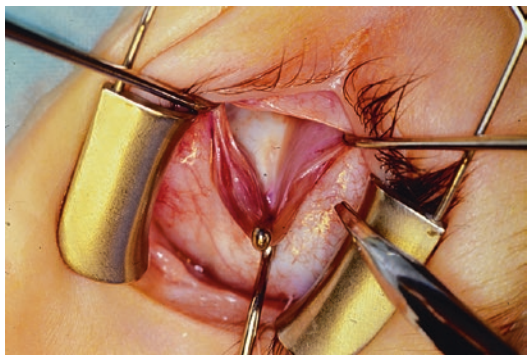

**Fig. 13.6** After the muscle has been captured and the tip of the hook bared, inspect the triangle between the two reflected arms of the inferior oblique muscle. Look carefully along the bare scleral triangle posteriorly to see if there are any missed muscle fibers. If there are, they will be seen running either horizontally along the sclera or in the reflected conjunctiva

#### Important Point

We typically operate on the IO if it is "overacting," and IO "OA" results in excyclotorsion. This can alter the anatomy, resulting in the LR being in the inferior temporal quadrant. I have seen patients who inadvertently had their LR operated on when the surgeon was intending to operate on the IO. Pay careful attention to the direction that the fibers are running. I have also seen globe extorsion result in the surgeon having difficulty locating the LR.

### Surrounding Tissue Damage

#### Basic Information

The five main complications of IO surgery in the form of surrounding tissue damage are (1) creating an adhesive syndrome, (2) bleeding, (3) LR damage, (4) iridoplegia, and (5) globe perforation.

### Adhesive Syndrome

Having let the hook pop through the connective tissue posterior to the muscle, rather than exactly at the interface between muscle and connective tissue (see Fig. 12.5), is the most common cause of a postoperative adhesive syndrome.

#### Pearl

*Visually Identify the Vortex Vein*

It is prudent to specifically seek out and locate the vortex vein in order to best avoid inadvertently traumatizing it. Sometimes it can get in the way of dissection of the connective tissue around the muscle, but more commonly it is close to the site at which an IO is to be sutured during a recession procedure. Bleeding from vortex vein trauma is the most common cause of excessive bleeding with IO surgery.

### Lateral Rectus Muscle Damage

The IO insertion is extremely close to the LR, and it is possible to sever the LR when disinserting the IO. This can be avoided by always identifying the LR and retracting it out of the way, either with muscle hooks or a traction suture, while disinserting the IO.

### Iridoplegia

Excessive traction on the IO can result in traction on the ciliary ganglion, resulting in an iridoplegia. This is more likely to occur after a denervation-extirpation of the IO, which usually requires more traction on the muscle. The best way to avoid this is to be cognizant that it can occur and to pull gently.

### Perforation of the Globe with Recession

The insertion of the IO is in a deep hole when approached surgically; it is close to the macula. When performing a recession, many surgeons prefer to place sutures in the muscle near the insertion prior to disinsertion—a technique popularized by Marshall Parks. I think it is safer to place a hemostat across the muscle near the insertion, and then disinsert it by cutting the insertional end flush with the sclera. The hemostat holding the cut end of the muscle can then be brought out of the hole and sutures safely placed in the cut end of muscle. This technique eliminates the risk of inadvertent perforation of the globe near the macula.

## Basic Information

*Choice of Weakening Procedures*

Traditionally, the IO is weakened by means of either recession or myectomy. Most surgeons have a strong bias for one procedure or the other. I am a strong advocate of recession. Whenever I have a choice between a reversible and a nonreversible surgical procedure, I will opt for the reversible procedure, all else being equal, and recessions can be reversed. They can be "undone" if too much effect is achieved, and easily converted to a more powerful procedure if the effect is insufficient. Whenever I reoperate on a previously myectomized IO, I feel it is a crapshoot; I never know what I will find. This is particularly problematic if I have to do bilateral surgery and want a symmetric effect, but only one IO has been operated previously via myectomy. On the other hand, the IO tends to be rather forgiving at surgery, and often you "get what you need." However, I find that surgery on the IOs is more "self-adjusting" when it comes to treating asymmetric OAs on side gaze, than it is for getting an asymmetric effect in the primary position to collapse a hypertropia (HT). If there is a HT in the primary position, bilateral asymmetric IO recessions are preferable to myectomies in my experience. In general, myectomy of a very large segment of the IO may be a bit more powerful than the largest recession. Table 13.1 lists the relative pros and cons of IO recession and myectomy. See Chap. 21, "Complicated Strabismus," Case 21.29 on page 333, for a representative example of these principles.

**Table 13.1** Pros and cons of inferior oblique muscle recession and myectomy

| Recession | Myectomy |
|---|---|
| Can be graded for asymmetric effect | Cannot be predictably graded |
| Reversible | Not reversible |
| Re-ops more predictable | Re-ops less predictable |
| Allows for subsequent anterior transposition | Does not allow for subsequent anterior transposition |
| Takes slightly longer | Quicker to perform |
| Has chance of globe perforation with needle | No chance of globe perforation with needle |
| Perhaps slightly less powerful | Perhaps slightly more powerful |
| Is usually effective | Is usually effective |

Symmetric IO myectomy may balance asymmetric IO OA on versions, but will usually not collapse a primary position hypertropia. For that you need bilateral asymmetric recessions.

## Important Point

As stated in Chap. 8, the most effective procedure for treating DVD with "IO OA" is anterior transposition of the

IO. This cannot be performed predictably after IO myectomy. For this reason, I suggest IO myectomy *never* be performed in patients with infantile esotropia. Recessions are preferred, as they can later be converted to anterior transposition if DVD develops.

### Question

How do you measure for an IO recession, particularly if you want to do it asymmetrically? What amount of recession do you choose?


### Reply

Parks popularized placing the IO 3 mm posterior and 2 mm lateral to the temporal end of the IR insertion, which he thought was an 8 mm recession [8, 9]. In a subsequent anatomic study, Apt and Call showed that the Parks recession was, in fact, a 10.4 mm recession with 1 mm anterior transposition [10]. They also gave guidelines as to where to place the IO for varying amounts of recession between 6 and 12 mm, which are reproduced in Table 13.2. If I need a standard (average) amount of weakening effect for either symmetric or asymmetric "IO OA," and if there is no HT in the primary position, I often default to using the Parks point, as I have had a long successful experience with that approach. If I really feel I need a maximum effect, typically in cases of superior oblique muscle (SO) palsy with a large deviation in primary, I do a 12 mm recession using Apt and Call's data for guidance. If I have asymmetric "IO OA" and a modest HT in primary, I do a 12 mm recession in the hypertropic eye, and a 6 or 8 mm recession in the other eye, depending on the size of the hypertropia.

**Table 13.2** Reinsertion point for graded recession of the inferior oblique muscle [10]

| Amount of recession | Millimeters inferior to temporal end of inferior rectus muscle insertion | Millimeters temporal to temporal end of inferior rectus muscle insertion |
|---|---|---|
| 6 | 5.0 | 6.4 |
| 7 | 5.0 | 5.4 |
| 8 | 4.0 | 4.4 |
| 9 | 4.0 | 3.4 |
| 10 | 4.0 | 2.4 |
| 11 | 4.0 | 1.4 |
| 12 | 4.0 | 0.4 |

## Special or Unusual Surgical Procedures on the IO

### Basic Information

*Anterior Transposition of the IO*

This procedure, first described by Elliott and Nankin, involves reinserting the IO anterior to the center of rotation, thus converting it from an elevator to an anti-elevator [11]. It is a very powerful operation that appears currently to be the best procedure for treating DVD with "IO OA." It does, however, create profound change in the force vector of the IO, and consequently, it has significant and perhaps unpredictable effects on comitance. Therefore, I *never* utilize it in a bifoveally fusing patient with diplopia awareness. I am aware that there are advocates of this procedure for unilateral superior oblique muscle palsy (SOP). However, I am unaware of any studies that have shown its effect in this population in eccentric gazes not on the range of single binocular vision field. Although this procedure has a profound effect on the vertical action of the IO, its effect on torsion is less profound. IO anterior transposition causes an incyclo-shift in the short term, but by 10 weeks after surgery the effect wears off [12]. In fact, if the procedure has an excessive effect in the form of the AES, there is an increase in the excyclotropia [13].

Anterior transposition of the IO has a noticeable effect on the palpebral fissure. It causes narrowing in straight-ahead gaze, and a bulge of the lower eyelid on upgaze, as well as a flatten-

ing of the lower eyelid curvature [14]. When done symmetrically, these are usually not a problem. But when done unilaterally, it can result in a noticeable cosmetic problem. This is yet another reason why I do not advocate this procedure for treating unilateral "IO OA," either idiopathic or associated with SOP. Furthermore, anterior transposition of the IO after prior superior rectus muscle (SR) recession for DVD, or subsequent SR recession, can profoundly cripple upgaze and cause upper eyelid retraction due to fixation duress [14]. If there has been prior SR recession, a less powerful degree of IO anterior transposition (AT) should be planned. Conversely, if there has already been prior IO AT, then subsequent SR recessions should be scaled back in size. Finally, IO anterior transposition can cause a noticeable restriction of elevation and ipsilateral HYPO (HYPO) in upgaze when performed unilaterally. See Chap. 21, Case 21.10 on page 318 and Case 21.20 on page 326 for representative examples of these principles.

IO AT combined with prior or subsequent SR recession can cripple upgaze resulting in upper eyelid retraction. Scale back the numbers.

## Question

How do you prevent the occurrence of the AES?


## Reply

There are presumably two factors that contribute to the AES. One is suturing the IO anterior to the IR insertion, and the other is spreading the posterior lateral corner of the IO out temporally [2]. However, I have found that the occurrence of this syndrome increases the longer one follows patients who have undergone the procedure. There are patients on whom I performed IO anterior transposition who did not have the AES at the time I described this entity, but developed it some years later. My preferred technique initially was to bunch the IO at the new insertion and to never place it anterior to the IR insertion, yet still some patients developed the AES. Over 10 years ago I switched to just suturing the anterior nasal corner of the IO to the temporal corner of the IR insertion, and letting the posterior lateral corner just fall posteriorly as shown in Fig. 13.7. I reasoned that because the AES appears to be caused by tautness of the lateral fibers of the transposed IO, having them less taut would help prevent AES. I have never had a case of AES in which I used that technique. Like so much in life, this is a matter of trade-offs. That approach is better with respect to not causing the AES. However, it is a less powerful procedure for treating DVD than my prior method.

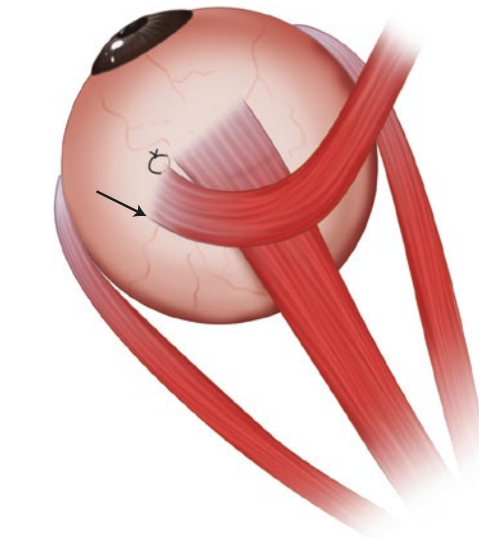

**Fig. 13.7** Preferred technique for suturing the inferior oblique muscle with anterior transposition. The anterior nasal corner is sutured to the temporal corner of the inferior rectus muscle insertion and the posterior corner (*arrow*) is allowed to drop back

## Pearl

To minimize the occurrence of the AES, suture the anterior nasal corner of the IO to the temporal corner of the IR insertion and let the posterior temporal corner hang back.

## Question

How do you treat the AES?


## Reply

Although I initially treated this syndrome with denervation-extirpation of the IOs, I rapidly realized that such an ablative procedure was not needed. Converting the anterior transposition to a simple recession by retroplacing the IO is quite successful in eliminating the AES. However, one must be prepared for a recurrence of DVD if it was present initially, and treat it accordingly. When faced with an apparent unilateral AES, it can be hard to tell if one is dealing with AES in one eye, versus contralateral "IO OA." This is a hugely important differential, because if there is contralateral IO OA, one should weaken the IO in the eye that over-elevates in adduction. But in the case of AES one should operate on the eye that does not elevate well in abduction. There are some signs that will help sort this out. If there is contralateral "IO OA," there should be fundus extorsion in that eye, and exaggerated forced ductions at the time of surgery will show a tight IO. If there is AES, the eye that does not elevate well in abduction may have marked fundus excyclotorsion, and the telltale bulge of the lower eyelid on attempted upgaze.

It can be tricky to differentiate unilateral AES from contralateral IO OA.

## Important Point

To differentiate between unilateral AES and contralateral IO OA, evaluate fundus torsion. With AES there is usually substantial fundus extorsion of the affected eye. With contralateral IO OA there is fundus extorsion in the contralateral eye.

## Important Point

*Forced Ductions are Typically Normal with the AES*

I think of the restriction to elevation in AES as being what I call an "innervational restriction," as opposed to a mechanical restriction. The anti-elevating force vector of the IO comes into being only in upgaze, and hence the IO is innervated. In the resting state, or under anesthesia, there is no mechanical restriction felt.

## Question

I operated on a woman for a 25 prism diopters (Δ) LHT and what seemed like a typical LSO palsy pattern. I compromised between a standard LIO recession and an anterior transposition by bunching it and placing it 1 mm posterior and 1 mm lateral to the temporal end of the LIR insertion, and I combined this with a 3 mm RIR recession. She is now 1 week post-op and is overcorrected. She not only has a 3 Δ L hypo, but has an L HYPO with diplopia across the entire upgaze fields. I think this is an unmasked bilateral SOP. How should I handle it?


## Reply

Although you typically should not get the AES from where you placed the LIO, I am suspicious that is what is going on. It would be unusual for a bilateral masked SOP to have diplopia across the entire upgaze fields so early on, as opposed to it only being, in this case,

in the field of the RIO. Look for torsion in the right eye. If it is not present, that would speak for AES or a restrictive cause of limitation of elevation OS.

**Advanced Information**

*Disinsertion or Z-Myotomy of the IO*

When a relatively small effect is needed, but you want both vertical and some torsional correction, a simple disinsertion or Z-myotomy at the insertion of the IO can be useful. Like myectomy, it tends to be somewhat self-adjusting, but is substantially less powerful than myectomy or recession.

**Advanced Information**

*When a Really Strong Procedure is Needed*

Denervation-extirpation is a very powerful operation for weakening the IO. I never find it necessary to perform this operation. If a complete myectomy is performed from the insertion to where the IO emerges from the IR, cutting the nerve to the IO should not add much effect. In other words, do an extirpation *without* the denervation. This also minimizes the risk of creating an iridoplegia, which can occur as a result of the denervation.

For recurrent or profound IO "OA," Stager et al. described a technique for doing a nasal myectomy [15], and Rosenbaum et al. described suturing the IO to the orbital wall [16]. Both of these procedures are effective, somewhat difficult technically, and in my experience rarely needed.

When one not only needs a profound weakening effect of the vertical force vector of the IO, but also a marked torsional effect, anterior and nasal transposition of the IO as described by Stager et al. can be useful [17]. The anterior transposition of the IO has an anti-elevating effect, and the nasal positioning changes it from an extorter to an intorter. This procedure is no more difficult than standard anterior transposition and in select (and unusual) cases can be useful.

**Advanced Information**

*Special IO Procedures with Respect to Torsion*

Because the IO has both a vertical and torsional effect, standard weakening procedures will affect both the vertical and torsional force vector. However, as with the SO, the anterior fibers have more of a torsional action and the posterior fibers more of a vertical action. One can selectively weaken either the anterior or the posterior fibers to affect just one of these two force vectors. To weaken the excyclo-vector of the IO, disinsert the anterior fibers of the IO as shown in Fig. 13.8. First put an absorbable suture in the anterior corner of the IO near the insertion. Then disinsert the anterior seven-eighths of the muscle. Finally, pass the suture around the IO and tie it so that it acts like a purse string and keeps the anterior fibers from scarring to the sclera; there are no suture passes made in the sclera. I find that this predictably gives about a 3–5° intorsional shift when performed alone, with no change in the vertical alignment. Conversely, one can weaken the posterior seven-eighths of the IO to get a small vertical correction without affecting torsion (Fig. 13.9). This will correct about 3–4 of vertical with no effect on torsion. See Chap. 21, Case 21.13 on page 320 and Case 21.30 on page 333, for representative examples of these principles.

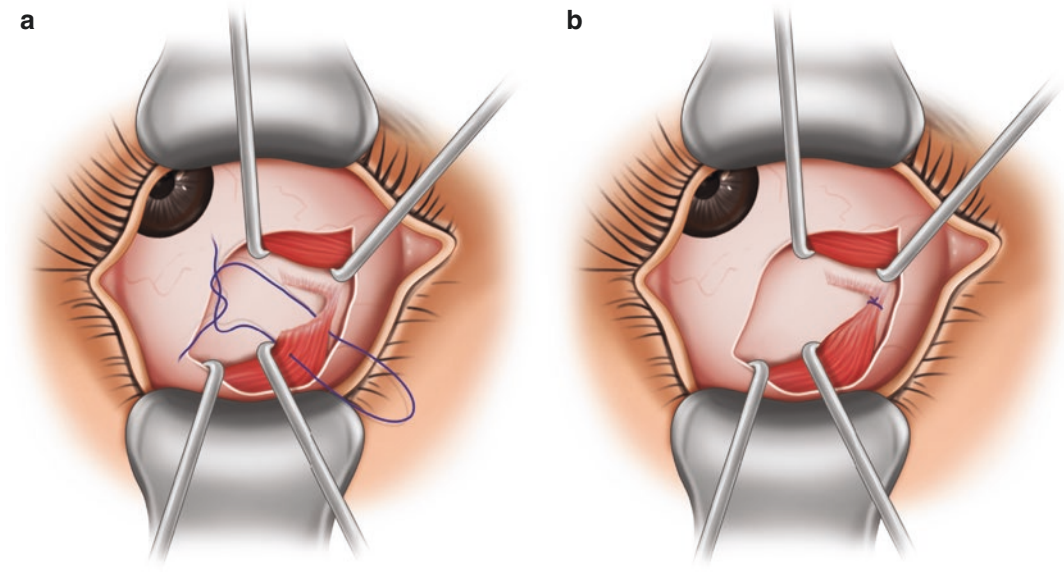


**Fig. 13.8** Anterior seven-eighths disinsertion of the inferior oblique muscle (IO). (**a**) A suture is securing the anterior corner of the IO near its insertion, and the anterior seven-eighths of the muscle is disinserted. The suture is passed around the IO in a purse-string fashion. (**b**) The suture is tied, bunching up the tissue to prevent the anterior fibers from sticking to the sclera

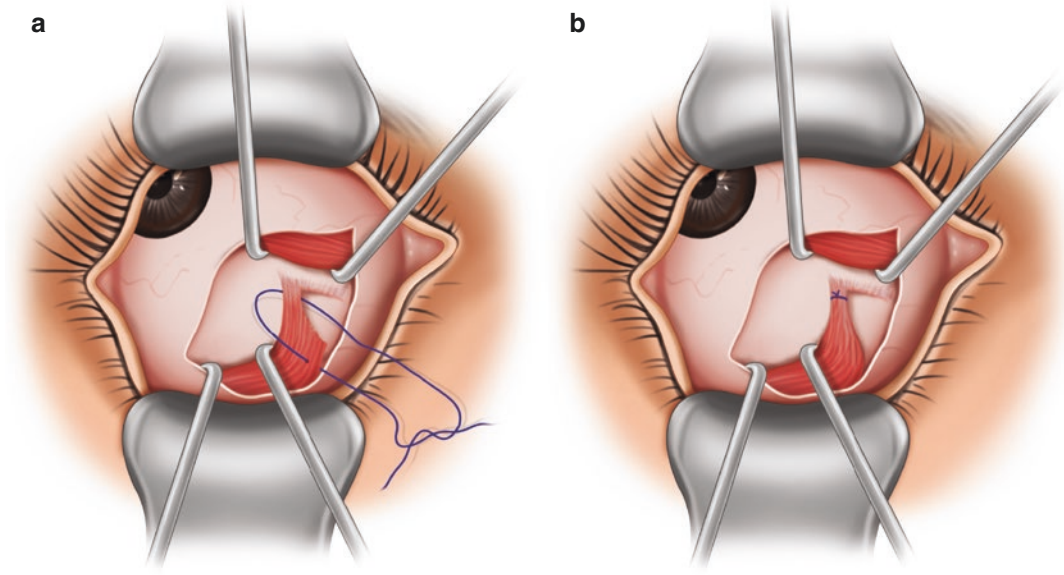


**Fig. 13.9** Posterior seven-eighths disinsertion of the inferior oblique muscle (IO). (**a**) A suture is securing the posterior corner of the IO near its insertion, and the posterior seven-eighths of the muscle is disinserted. The suture is passed around the IO in a purse-string fashion. (**b**) The suture is tied, bunching up the tissue to prevent the posterior fibers from sticking to the sclera

## Superior Oblique Muscle and Tendon

### Basic Information

*Anatomic Considerations—the Frenulum*

The anterior border of the SO tendon is about 4 mm posterior to the insertion of the SR when the eye is in the primary position. When the eye is rotated inferiorly, as is done to facilitate exposure at surgery, the SO tendon slides posteriorly so that its anterior border is about 8 mm posterior to the SR insertion. The SO tendon is loosely adherent to the undersurface of the SR by a connective tissue that has been described as a "frenulum' (Fig. 13.10) [18]. When the frenulum is left intact, the SO tendon will move with the SR. Consequently, if the SR is recessed and the frenulum is intact, the SO tendon will not only move posteriorly with the SR, but if the desired SR recession is more than 10 mm, the SO tendon (via the frenulum) will constrain the SR and prevent it from being suspended back the desired distance. Conversely, if the frenulum is left intact, the SO tendon will come forward during a SR resection. In addition, an intact frenulum will constrain the SO tendon from achieving maximum or perhaps even a desired amount of recession when the SO is weakened by a temporal approach as with disinsertion or suspension (hang-back) recession. It is important to note that the SO frenulum is easily stripped inadvertently when the SO tendon is hooked at the insertion and the tendon pulled out from under the SR for exposure. The degree to which one does this maneuver at surgery is often not paid sufficient attention, and inadvertently doing it asymmetrically can result in an unequal recession effect with a bilateral procedure (Fig. 13.11). Also, the SO tendon will be dragged laterally during an SR temporal transposition as may be done for treating sixth cranial nerve palsy, Duane syndrome, or for correcting excyclotropia, if the frenulum is not stripped. Failure to separate the SO tendon from the SR can cause the SO incarceration syndrome [19].

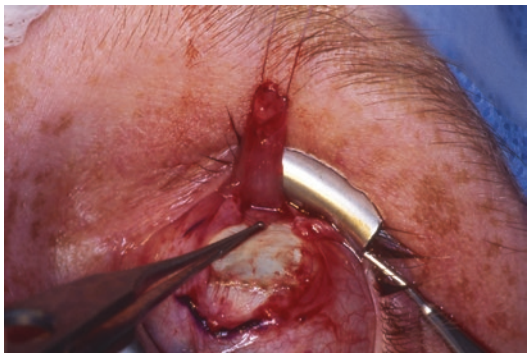

**Fig. 13.10** Surgical view from below. The superior rectus muscle (SR) was disinserted and is held up via preplaced sutures. The superior oblique tendon is held in forceps and seen to be attached to the SR undersurface via the frenulum

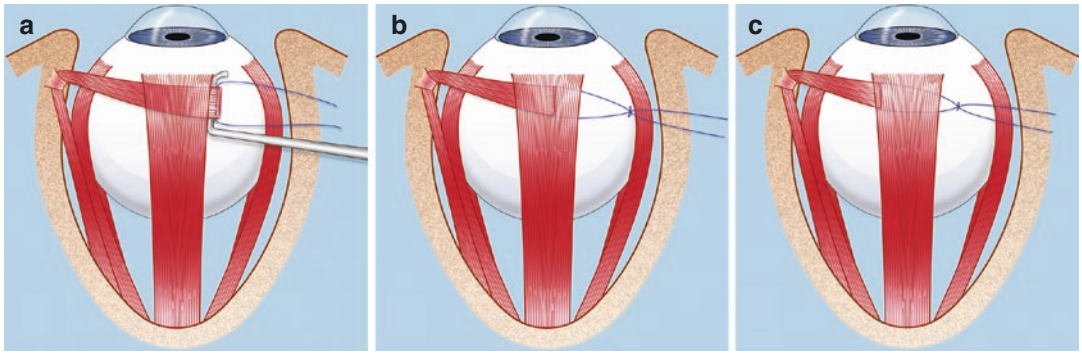


**Fig. 13.11** Disinsertion or suspension (hang-back) recession of the superior oblique muscle (SO). (**a**) The SO is hooked temporally and brought out into the surgical field. The amount of SO tendon that is exposed has a direct effect on how much the frenulum is stripped. (**b**) Recession with small amount of stripping of the frenulum. (**c**) Greater amount of recession than is seen in Fig. 13.11b due to greater amount of stripping of the frenulum (from Iizuka and Kushner [18], with permission)

## Question

Can you list specific guidelines (with reasons) as to how one should handle the frenulum when doing SR or SO surgery?


## Reply

These guidelines are the result of some specific in vivo studies conducted on the behavior of the SO tendon and SR with and without severing the frenulum [18].

1. For SR recessions of 10 mm or less, leave the frenulum intact. This will keep the SO tendon posterior to the new SR insertion and decrease the chance of the *SO incarceration syndrome* [19].
2. For SR recessions of more than 10 mm, sever the SO frenulum to permit the SR to achieve the desired amount of recession. This is particularly true if you plan to use a suspension technique. However, be aware that the new SR insertion may overlie the SO tendon, which can result in nonadherence of the SR. If you see that the recessed SR is overlying the SO tendon, you should either switch from a suspension technique to using fixed scleral sutures or use a nonabsorbable suture for the suspension.
3. Sever the SO frenulum for all SR resections to prevent the SO tendon from being incorporated in the new SR insertion, resulting in the SO incarceration syndrome.
4. For all bilateral temporal weakening procedures on the SO (e.g., disinsertion, suspension recession, tenectomy), be sure that you strip the frenulum symmetrically. This is accomplished by exposing an equal amount of tendon temporal to the SR, as shown in Fig. 13.11a. Failure to do so is the most common cause of an asymmetric response.
5. For all temporal SO weakening procedures, bilateral or unilateral, maximum effect requires maximum stripping of the frenulum as seen in Fig. 13.11. However, maximum stripping of the frenulum may result in the tendon sliding forward and adhering to the nasal corner of the SR insertion, causing the SO incarceration syndrome. Moreover, maximum effect is not as important as a predictable effect. This is one reason I opt for a nasal weakening procedure if a maximum effect is needed.
6. Separate the frenulum for all horizontal transpositions of the SR that are more than a few millimeters.

## Important Point

Failure to separate the SO frenulum is a major cause of unintended vertical deviations after horizontal transposition of the SR.

## Basic Information

*General Information About Weakening and Strengthening Procedures*

The SO has two major functions, depression (moreso in adduction) and intorsion (moreso in abduction), and, to a lesser degree, a third function of abduction. Any surgical procedures that involve the entire SO tendon (anterior and posterior fibers) will affect all three of these functions. If a selective effect is desired, surgery must be confined to the anterior fibers for torsion, or the posterior fibers for the vertical effect.

## Basic Information

*Strengthening (Tightening) to Affect all Three Force Vectors*

1. *Tucking*. The classic strengthening (tightening) operation on the SO is a tuck performed temporal to the SR. In my hands this procedure is not formula driven but done by "feel." I do not see the need for a tendon tucking

device. Instead, I pull the tendon up on a hook and use a hemostat (mosquito) to slide down the two reflected sides of the tendon and clamp at the desired spot. I then place a temporary nonabsorbable suture just inferior to the hemostat and do forced ductions. My desired endpoint is just to feel resistance as the inferior limbus is level with the horizontal canthi. If I have the right amount of tuck according to this criterion, I permanently tie the suture. There is a difference of opinion as to whether the apex of the tucked tissue should be sutured to the sclera. In theory you can get more torsional correction by suturing it somewhat anteriorly and temporally, and more vertical effect by suturing it more posteriorly and temporally. In practice, I do not feel this makes a real difference, and I often do not suture the apex of the tuck to the sclera.

2. *Resection*. A resection should have exactly the same effect millimeter for millimeter as a tuck
3. *Plication*. A plication should have exactly the same effect millimeter for millimeter as a tuck.
4. *Advancement*. In theory, advancing the SO tendon toward the lateral recuts muscle should have a somewhat more powerful effect as any of the aforementioned procedures. Millimeter for millimeter it will stretch the sarcomeres in the SO to an identical amount as a tuck and thus have the same increase in contractile force. But it will also increase the arc of contact and thus further increase torque.

## Question

I am used to strengthening the SO by tucking. If an advancement is more powerful, should I switch to that procedure?


## Reply

Not necessarily. The SO tends to be an unforgiving muscle, and experience with a procedure is important. You can always increase the effect you get with tucking the SO by doing a larger tuck. In addition, doing the more powerful procedure is not necessarily optimum. It is probably more important that you become confident and comfortable with one procedure—*either* one.

## Important Point

Although SO tucks are often described as being reversible, this is not always the case. In my hands, tucks are often difficult to reverse. Most of the time the two arms of the tuck scar and blend into one another, and at reoperation there is no interface at which you can separate them and undo the tuck. Similarly, an SO resection cannot be reversed. An SO advancement is probably the most reversible of the strengthening procedures.

## Basic Information

*Weakening Procedures to Affect All Three Force Vectors*

There is a hierarchy to weakening procedures, which also have respective pros and cons. The following are listed approximately in order from weakest to strongest:

1. *Disinsertion*. Disinserting the SO tendon is a simple mild weakening procedure that can be done when only a little effect is needed. On the downside, this procedure can be variable in its effect. I think the biggest cause of variability is that we typically do not pay enough attention to how far we pull the SO out from under the SR when we hook it, and this results in a variable effect on stripping of the frenulum. Minimal pulling out of the SO will result in a smaller effect with disinsertion; more pulling will have a greater effect, as the tendon will retract (recess) further.
2. *Recession on a suspension (hang-back)*. Of course the magnitude of effect with suspension-recession is in part a function of the magnitude of the recession. If a recession is done on a suspension, the same caveats hold

as with disinsertion with respect to stripping of the frenulum. If a big recession is desired but there is minimal stripping of the frenulum, the tendon may not retract the desired amount. On the other hand, if a large recession is desired and the frenulum is substantially stripped, there is an increased likelihood of the SO tendon adhering to the nasal corner of the SR and causing the SO adherence syndrome [20]. This is one reason I tend to shy away from this procedure.

3. *Recession with scleral fixation at desired point*. This is probably a more predictable procedure than a suspension-recession, as it eliminates the variability of stripping of the frenulum.

### Important Point

There is another issue which is a cause for concern with SO recessions whether done on a suspension or with fixed scleral sutures. If you recess the tendon following the normal course of the SO, even a modest recession will result in the new insertion lying anterior to the center of rotation, which will change the SO vertical force to that of elevation in abduction. This often results in a restriction to depression in abduction. It is, in a sense, the mirror image of the AES that occurs after IO anterior transposition. For this reason I avoid recession of the SO along its normal anatomic path, either with fixed scleral sutures or on a suspension. To prevent the complication of restriction of depression in abduction, Prieto-Diaz advocated recession of the SO to a point 3–4 mm nasal to the SR, but also 12–13 mm posterior to the limbus [21]. This is a powerful weakening procedure but does not have the advantage of being graded. For this reason, I prefer a nasal weakening procedure when a large effect is needed. I have no experience with the procedure advocated by Prieto-Diaz.

4. *Nasal tenotomy*. This is an effective weakening procedure when weakening of all three actions of the SO is desired. When done bilaterally, there can be an undesired asymmetry to the effect, as a simple tenotomy does not control how much the two cut ends of the tendon separate. For this reason, I like to use a piece of nonabsorbable suture to bridge the gap, what Knapp described as a "chicken suture." Use thereof does help insure a symmetric effect in bilateral cases.

### Important Point

The location at which a tenotomy is performed also affects the power of the procedure—the closer to the trochlea, the more powerful. So if you desire a symmetric bilateral effect, take care to do the tenotomy in the same place in each eye. For a long time, I wondered why the location of the tenotomy should make a difference. After all, if the tendon is completely severed, it would seem that no contractile force would be transmitted to the globe regardless of where the cut is made. However, Helveston showed that there are indirect connective tissue attachments all along the nasal aspect of the tendon, between the tendon and the globe [22]. The closer the cut is made to the SO insertion, the more of these attachments remain intact; the closer to the trochlea, the more of them are separated from the contractile SO.

5. *Tendon elongation procedures—silicone spacer and split tendon lengthening*. These two procedures have similar effect and efficacy, and have relative pros and cons. The silicone spacer, described by Wright [23], involves elongating the SO tendon nasal to the SR by inserting a strip of silicone with a length equal to the desired amount of elongation. The split tendon lengthening accomplishes the same thing by splitting the SO tendon longitudinally nasal to the SR, and then suturing the nasal end of one half of the split to the temporal end of the other half of the split as depicted in Fig. 13.12, using

a nonabsorbable suture [24]. Both of these procedures have the advantages of maintaining the original SO insertion, are less likely to cause the SO adherence syndrome than procedures done temporally, and—at least in theory—can be titrated for an asymmetric effect if done bilaterally. I find that both procedures can be titrated to treat bilateral asymmetric "SO OA" with respect to normalizing versions. However, I have had less success in eliminating a hyper in primary due to asymmetric "SO OA" with asymmetric silicone spacers than with asymmetric split tendon lengthening. The silicone spacer is technically easier because suturing into the full tendon is easier than suturing half of the tendon, which can be challenging. However, the silicone spacer requires that you maintain the integrity of the nasal intermuscular septum or you run the risk of the silicone sticking to the sclera. This can be challenging, as the septum can easily be inadvertently violated. It is probably less crucial to maintain the intermuscular septum with split tendon lengthening, because the only foreign body is the one knot in the suture, which is further from the SR than the temporal knot with a silicone spacer. As such it is further off the sclera and less likely to adhere. Weighing the pros and cons of both procedures I prefer the split tendon lengthening. The advantages of both procedures are listed in Table 13.3. See Chap. 21, Case 21.19 on page 325, for a representative example of these principles.

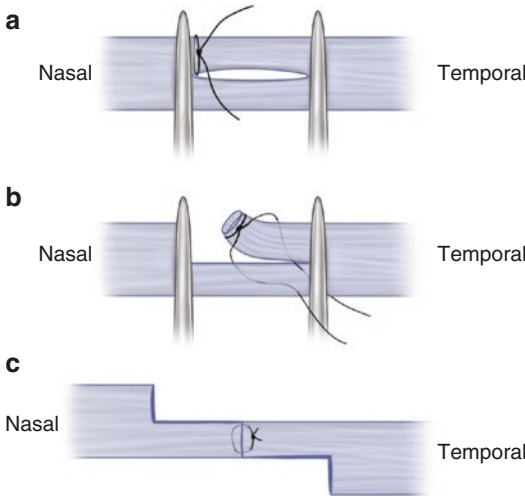


**Fig. 13.12** Spit tendon lengthening of the superior oblique muscle (SO) tendon. (**a**) Mosquito hemostats are placed across the SO tendon in the nasal quadrant. The distance between the hemostats equals the desired amount of lengthening. The tendon is split longitudinally between the hemostats with Stevens hooks. A double-armed nonabsorbable suture is passed through the nasal end of one half of the split tendon adjacent to the mosquito. (**b**) The sutured half is tenotomized just inside the mosquito, and the sutures are then passed through the temporal end of the other half of the split tendon inside the mosquito. (**c**) The remaining half of the tendon is transected at nasal edge of the nasal hemostat. The sutures are tied, connecting the nasal half tendon to the temporal half tendon. Note: Alternatively, the posterior half of the tendon can be sutured and transected at the temporal edge and the anterior half at the nasal edge

6. *Tenectomy*. This is the most powerful SO weakening procedure, which will always result in a SOP. I limit its use to the treatment of SO myokymia, where an ablation of the SO function is needed [25]. I always anticipate and treat the inevitable SOP by simultaneously doing a large myectomy of the IO.

**Table 13.3** Advantages of silicone spacer and split tendon lengthening

| Advantages of both procedures over other weakening procedures | Advantages of split tendon lengthening over silicone spacer | Advantages of silicone spacer over split tendon lengthening |
|---|---|---|
| Maintains normal insertion | Minimal foreign material (just the knot) | Possibly easier: no need to suture into half tendon slips |
| Maintains normal tendon path | No extrusion risk | |
| Potentially reversible | Less infection/inflammation risk | |
| Possibly can be titrated for asymmetric effect | Possibly easier: less need to preserve intermuscular septum | |

### Question

My patient has bilateral asymmetric "SO OA" (+2 in the right and +4 in the left) in conjunction with an intermittent A-pattern exotropia. Because of the asymmetric "SO OA," he has an 8 Δ RHT in primary. He fuses with a face turn to the right, as that takes him out of the more overacting LSO and the face turn neutralizes the hyper. How should I manage his SOs to eliminate the hyper?


### Reply

I have seen a number of patients with asymmetric "SO OA" who have a hyper in primary that is cancelled out with a face turn. For years I sought a predictable way to asymmetrically weaken the SOs to treat these patients. Early in my practice, I would do a disinsertion on the SO of the eye with the lesser degree of OA, and a nasal tenotomy on the more overacting side. However, I never felt I had sufficient ability to titrate the effect as much as I would like. My early experience with silicone spacers was disappointing with respect to eliminating the primary position hyper, although it was effective in balancing the asymmetric versions. I find that asymmetric split tendon lengthening works well in this setting. An alternative is to do a symmetric weakening of the SOs, and then recess a vertical rectus muscle to address the primary position hyper.

### Question

I have a patient with +2 SO OA and an A-pattern exotropia. I am thinking of 50% Z-tenotomies on the SOs to treat the OA and A-pattern. Do you agree with that plan?


### Reply

By saying a 50% Z-tenotomy, I presume you are talking about the type of procedure that involves merely making two cuts 50% of the width of the tendon, as shown in Fig. 10.20, one on the posterior edge and the other on the anterior edge. However, Helveston showed that there is no meaningful lengthening of the muscle or tendon with myotomy or tenotomy until the cuts overlap one another, e.g., are more than 50% of the width, and the amount of lengthening is equal to the amount of overlap. So a 50% Z-tenotomy will not have a significant effect. But more importantly, he also showed that there is very little interdigitation between parallel SO tendon fibers. If the cuts are made greater than 50% across, which would be necessary for any significant weakening effect, there would be no fibers that are completely continuous, resulting in a great risk of the tendon pulling apart after surgery, resulting in a free tenotomy. The split Z-tendon lengthening as described by Bardorf and Baker [24] is different than a simple Z-tenotomy, and would be effective in this setting. However, for SO OA of +2, you might also consider the posterior seven-eighths tenectomy [26], which is my personal preference here.

### Question

I have an adult with a 40 Δ L esotropia and bilateral +4 "SO OA." She is amblyopic in her left eye and also has an L HYPO of 10 Δ in primary. How should I handle the HYPO? Because I do not expect she will develop high-grade fusion, I plan to ignore the A-pattern.


## Reply

In cases of non-alternating strabismus with oblique dysfunction, there may appear to be a vertical deviation in primary that is due merely to the fact that the deviating eye is further in field of the overacting oblique. In your patient, the left eye is esotropic and that puts it in the field of the SO, which can cause the HYPO. If that indeed is what is causing the HYPO, it should disappear if the eye is horizontally straightened, since the eye would no longer be in the field of the SO. In order to test for this, you need to measure the horizontal deviation with the prism split equally between the two eyes. The vertical you then measure, if any, is approximately what you can expect to be present with horizontal surgery alone. That is what you need to treat. I suspect that in your patient it will be a clinically insignificant amount.

With non-alternating strabismus, a primary position vertical may reflect the eye's position relative to oblique dysfunction, and may be gone after horizontal surgery.

## Basic Information

*Surgery That is Selective for Weakening the Vertical Vector of the SO*

There are circumstances in which you want to weaken the vertical action of the SO but not affect the torsional vector. The most common example is the patient with bifoveal fusion potential, mild "SO OA," but no torsion. Weakening the entire SO runs the risk of inducing torsion. For this scenario, the posterior seven-eighths tenectomy as described by Prieto-Diaz is useful [26]. It involves disinserting the (roughly) posterior seven-eighths of the SO tendon, and then excising a trapezoid of tissue containing the entire posterior seven-eighths of the insertional fibers, extending about 7–8 mm nasally (Fig. 13.13). This is a relatively mild weakening procedure and is useful if the "SO OA" is +1 or +2.

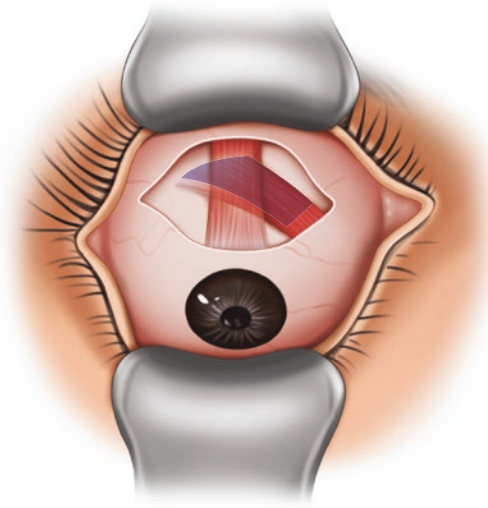

**Fig. 13.13** Posterior seven-eighths tenectomy of the superior oblique muscle. The *shaded trapezoid* depicts the tissue that is excised

## Basic Information

*Surgery That is Selective for Weakening the Torsional Vector of the SO*

More or less a mirror image of the posterior seven-eighths tenectomy, one can do an anterior seven-eighths tenectomy as shown in Fig. 13.14. This will correct about 5° of incyclotropia without significantly changing the vertical alignment.

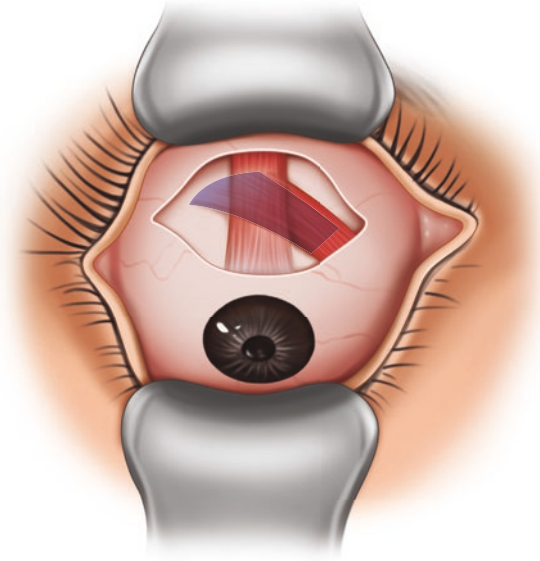

**Fig. 13.14** Anterior seven-eighths tenectomy of the superior oblique muscle. The *shaded trapezoid* depicts the tissue that is excised

## Basic Information

*Surgery That is Selective for Strengthening the Torsional Vector of the SO*

The Harada-Ito procedure strengthens the anterior fibers of the SO and increases its intorsional vector [27]. Most people use a modification of the original procedure and disinsert and transpose the anterior one-fourth to one-half of fibers of the SO to the superior border of the LR at 7–8 mm posterior to the LR insertion [28]. This predictably corrects up to 10° of excyclotropia. Traditionally, surgeons used nonabsorbable sutures for all SO tendon surgery, but in recent years good results have been reported with absorbable sutures [29]. It appears to me that a greater loss of effectiveness over time has been found in series that used absorbable sutures, and this may be important. An advantage, of course, with absorbable sutures is that there is no visible knot remaining. If I want a bilateral asymmetric effect, I accomplish this by either taking more of the tendon in the eye in which I need more correction or resecting a small amount of tendon. One can also obtain asymmetry by transposing less in the eye in which a small correction is needed, and putting the tendon several millimeters superior to the superior edge of the LR. Some people prefer to use an adjustable suture technique [29], and others like to titrate the amount of transposition in the operating room by directly observing the change in fundus torsion. I have shied away from using an adjustable suture for this procedure in part because it is typically self-adjusting (you get what you need) and because of the tendency for postoperative regression. Similarly, gauging the amount of tightening by observing fundus torsion changes in the operating room seems like unnecessary overkill to me. I am not impressed that it improves the final outcome. But, doing so may give a sense of security to some surgeons. I find the main indications for this procedure to be (1) residual torsion after the vertical deviation was corrected in patients with SOP, and (2) patients with bilateral SOP with a lot of extorsion and minimal vertical deviations in primary or side gaze.

## Pearl

Any surgery that affects the torsional action of the SO or IO will cause a rotation of the axis of astigmatism of between 5 and 10° because the globe has been

surgically rotated around the *y*-axis [30]. In an astigmatic patients with bifoveal fusion and diplopia awareness, they will report blurred vision with their old spectacles. However, in the patient with subnormal fusion, and perhaps treated amblyopia, the more astigmatic eye is typically the nondominant eye, and the patient may not notice the blurred vision. It is important to keep this in mind whenever oblique muscle surgery is done on a patient with high amounts of astigmatism in either operated eye. This change should be permanent by 6 weeks post-op.

### Important Point

Whenever you do oblique muscle surgery on a high astigmat, re-refract them 6 weeks after surgery with particular attention to a change in the astigmatic axis.

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# Abnormal Head Postures

# 14

*Parent: "My daughter's teacher says she always turns her head to the left while reading the overhead. She thinks Susie is just using her right eye."*
*Doctor: "In fact, if she turns her head to the left it is probably because that way she can used both eyes together better, not because she is wanting to use her right eye only. Actually, if she were only using one eye, a face turn to the left usually means she is using her LEFT eye."*

## Overview of Causes of Abnormal Head Postures (AHP)

### Basic Information

Most PCPs tend to think of orthomuscular causes for AHPs like muscular torticollis. All strabismologists know of children who were sent for physical therapy when in fact they had a fourth cranial nerve palsy. Conversely, strabismologists may tend to think that all patients with an AHP have an ocular cause. The truth lies somewhere in between. In one prospective multidisciplinary study of 63 children presenting to PCPs with an AHP [1], the cause of the AHP was orthopedic in 35 (56%), ocular in 25 (40%), and neurologic in 5 (8%). No specific cause could be found in the remaining eight (13%) patients (total is more than 100% as some patients had multiple etiologies). Congenital muscular torticollis was the most common orthopedic cause accounting for 31 patients (49%). The most common ocular cause was superior oblique muscle palsy, which accounted for 12 patients (19%). In two patients (3%) with fourth cranial nerve palsy there was secondary neck muscle contracture suggesting an orthopedic cause.

### Important Point

Not all AHPs in children have an ocular cause.

### Basic Information

*Ocular Causes of AHP*

In 1979 I published a series of 188 consecutive patients I had seen with an AHP due to ocular causes [2]. The breakdown of etiologies from that series is shown in Table 14.1, with some numbers updated after I subsequently learned the relationship between DVD and AHPs, which is really a form of incomitance [3]. In my initial report I referred to these patients as having an "ocular posture" as described by Lang. Although I identified seven different ocular causes, the vast majority were due to either incomitance (69%) or nystagmus (20.2%). Of the patients with an AHP due to incomitance, 46 (35% of the incomitance patients and 24% of the total 188 patients) had incomitance due to fourth cranial nerve palsy, and 31 (24% of the incomitance patients and 16% of the total 188 patients) had Duane syndrome. The ten patients who had an AHP to permit foveal fixation either could not


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_14

**Table 14.1** Ocular causes of abnormal head posture in 188 consecutive patients

| Etiology | Number (% of total) |
|---|---|
| Incomitance | 130 (69)[a] |
| Nystagmus | 38 (20.2) |
| To permit foveal fixation | 10 (5.3) |
| Cosmetic (voluntary) | 4 (2.1) |
| Oculomotor apraxia | 3 (1.6) |
| Spasmus nutans | 2 (1) |
| Astigmatism | 1 (0.5) |

[a]Data from Kushner [2], but also includes 12 patients with incomitant dissociated vertical divergence who were previously labeled as having an "ocular posture," as described by Lang

move their eyes to the primary position due to restriction (ocular fibrosis, Graves' orbitopathy, scarring, etc.) or had a dragged fovea in the fixing eye due to retinopathy of prematurity. Four adult patients voluntarily assumed an AHP either to hide a cosmetically unacceptable conjunctival scar or because their incomitance strabismus looked better with the AHP despite no improvement in binocular function.

## Basic Information

*General Approach to Patients with AHP*

As stated, the vast majority of patients with an AHP due to an ocular cause either have incomitant strabismus with some type of fusion—perhaps peripheral and/or subnormal—in their preferred head posture or have nystagmus that damps with the AHP. Less commonly, there are patients with hemifield defects who turn their face toward the defective field. Your first task in evaluating a patient with an AHP is to determine if the AHP indeed has an ocular cause.

### Nystagmus

In many cases it may be obvious that there is a manifest nystagmus that damps in the preferred AHP. In other cases the damping of the nystagmus may be too subtle to make that determination. If so, you should check visual acuity under binocular conditions with the patient's head straight and in the AHP. An improvement in acuity in the AHP is a strong indication that the AHP is driven by the nystagmus null point. However, the converse is not necessarily true. Sometimes the damping of nystagmus results in a visual benefit that does not translate to improved acuity. Patients may experience a prolonged foveation time resulting in improved "visual efficiency," e.g., reading speed and comfort, that is not detectable with routine office tests. Fortunately, in the majority of patients the damping of nystagmus in the AHP is obvious. Sometimes, comparing the amplitude of nystagmus in right and left gazes while viewing the fovea with a direct ophthalmoscope is useful.

### Important Point

The absence of an improvement in visual acuity when a patient with nystagmus assumes their preferred AHP does not rule out a nystagmus null point as the cause of the AHP.

### Incomitance

In many cases in which incomitance drives an AHP, the relationship between the AHP and strabismus may be obvious. A patient may change from experiencing diplopia or suppression to more normal fusion with the AHP. However, many patients with incomitance may assume an AHP if their angle of strabismus merely decreases to an amount that permits peripheral fusion—typically 10 Δ or less. This is particularly common in patients with "A"- or V"-pattern strabismus, and speaks to the hard-to-define subjective benefits patients perceive from peripheral fusion. Many patients with bifoveal fusion potential may assume an AHP because it merely decreases a manifest deviation to a size they can more easily control with fusional vergences. This is particularly common in patients with fourth cranial nerve palsy.

### Important Point

Incomitance may drive an AHP by merely decreasing, but not eliminating, a manifest strabismus.

There are several useful diagnostic steps that can help determine if incomitance is driving an AHP.

1. Observe the patient with each eye (separately) patched. If the AHP is gone with monocular occlusion, you can be sure that incomitance is driving the AHP. If observing this in the office setting is inconclusive, it is often helpful to have the patient (or parents) patch one eye at home for several hours or so, and another time or on another day to patch the other eye. The effect of monocular occlusion on the AHP should be noted. If the AHP is driven by improved fusion due to incomitance, it should be eliminated with occlusion of the nondominant eye. In most cases, it will also be eliminated with occlusion of the dominant eye. However, especially if the nondominant eye is amblyopic, there may be a persistence of an AHP when the dominant eye is occluded, just as some patients with amblyopia struggle to see with their amblyopic eye, and may turn or tilt their head. Contrary to common belief, the persistence of a head tilt when the dominant eye is patched is usually not caused by torsion in the nondominant eye. The head tilt that accompanies fourth cranial nerve palsy is driven by incomitance and a decrease in the hypertropia, and is not caused by torsion [4].

### Pearl

An incomitance-driven AHP may be eliminated by monocular occlusion of the nondominant, but may on occasion persist with occlusion of the dominant eye, especially if amblyopia is present.

### Important Point

A large manifest head tilt is not necessarily indicative of a large amount of torsion. Rather, it indicates that there is a decrease in the vertical deviation with a large head tilt.

2. When in doubt, and particularly if both nystagmus and incomitance are present, the diagnostic use of prisms can help sort the cause of an AHP. Consider a patient with a 25 Δ "A"-pattern esotropia that decreases to less than 10 Δ in downgaze, a manifest nystagmus, and a chin-up head posture. It appears that the nystagmus is less when the patient assumes their chin-up AHP. There are three potential causes for the AHP. It is important to sort this out because the treatment will be different for each scenario. Prisms can be used to determine the etiology for the AHP, either in the office with prism trial lenses or in the form of a short trial of Fresnel prisms at home.
   (a) The AHP may be driven by incomitance and the fact that there is some improvement in fusion in downgaze because the deviation is smaller. In this case, 25 Δ base-out prism over either eye should eliminate the AHP, but base-up prism over both eyes will not. In this case, surgery to eliminate the 25 Δ esotropia (ideally, but not necessarily) to address the "A"-pattern will eliminate the AHP.
   (b) Alternatively, the nystagmus may have a null point in downgaze. In this scenario, base-up prism in front of both eyes will eliminate the AHP, but base-out prism will not. Surgery to shift the null point in the form of a vertical Kestenbaum–Anderson procedure (recession IR OU and resection SR OU) will correct the AHP.
   (c) Finally, it is possible that there is a manifested latent nystagmus in primary gaze that converts to a latent nystagmus when the eyes are aligned in downgaze. In this

scenario, either 25 Δ base-out prism over either eye or base-up prism over both eyes will eliminate the AHP. In this case, surgery to eliminate the 25 Δ esotropia (ideally, but not necessarily) to address the "A" pattern will eliminate the AHP.

### Pearl

When there is both nystagmus and incomitant strabismus, prisms can be used diagnostically to sort out which of the two is driving the AHP, and can guide treatment.

### Pearl

Infrequently, a head tilt may decrease an esotropia. The mechanism for this head tilt-dependent esotropia is unclear, and the finding appears to be limited to patients with Down syndrome [5].

### Advanced Information

Some patients with craniosynostosis show an AHP that appears to relate to vertical strabismus, yet persists after successful surgical correction of the vertical strabismus. According to Linda Dagi, M.D., "some craniotomy patients may have an AHP that was initially ocular in nature, and is no longer. The causes may be multiple and might include asymmetric weight and shape of the head" (unpublished personal written communication, March 21, 2016). According to Ken Nischal, some of these patients have an abnormally tilted interface between the base of the skull and the first vertebrae (unpublished personal written communication, May 29, 2016). For these patients, it is useful to see if the AHP resolves with monocular occlusion. If it does not, it is probably not (or no longer) ocular in nature.

### Advanced Information

There is a syndrome in which children either born with severe unilateral amaurosis or who had an eye enucleated in infancy develop a manifest nystagmus in the seeing eye with a null point in adduction, resulting in an AHP [6]. This appears to be a unilateral manifested latent nystagmus and to occur disproportionately (but not exclusively) in patients with a positive family history for infantile strabismus. It has been theorized that these patients inherited a genetic predisposition to subnormal fusion which results in infantile strabismus. The enucleation or amaurosis acts like monocular occlusion in making what would have been latent nystagmus, manifest.

### Question

I have a patient who appears to have the syndrome of uniocular blindness, nystagmus, and an AHP. His face turn and tilt have persisted after two surgical procedures that should have addressed it. How can I tell if the AHP is visually driven?


### Reply

A useful way to sort this out was described by Lionel Kowal, M.D. (unpublished oral personal communication, April 9, 2016). Have the patient sit up and close her eyes. Move her head to an eccentric position and ask her to straighten her head. If it is straight with her eyes closed and tilted/turned when she opens them, the AHP is visually driven. But if the AHP persists with the eyes closed, it is a vestibular problem and not reliably fixed with muscle surgery.

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# Brown Syndrome

# 15

## Basic Information

When an eye is in adduction and the superior oblique muscle (SO) contracts, the eye depresses because the SO inserts posterior to the center of rotation. Conversely, when an eye with a normal SO elevates in adduction, the SO insertion moves posteriorly, pulling the SO tendon through the trochlea. If the SO is tight, it cannot pass through the trochlea due to swelling or anatomic variants or, possibly, if the insertion is anomalous the eye cannot elevate in adduction. This is the clinical manifestation of *Brown syndrome.*

## Basic Information

*Presentation*

Brown syndrome is usually congenital but can be acquired. Acquired cases are due to trauma affecting the trochlea-SO tendon complex, to orbital tumors affecting that area, to sinusitis, or to autoimmune inflammatory disease. Congenital cases may be asymptomatic and go unnoticed if there is no deviation in primary. Sometimes parents note what appears to be an overshoot of the unaffected eye in upgaze and think that is the eye with the problem. In mild cases the only imbalance seen is a hypotropia (HYPO) on attempted elevation in adduction. If the tendon is very tight, there may be a HYPO of the affected eye in primary gaze and/or a downshoot in adduction. If there is a HYPO in primary gaze, congenital cases typically assume a chin-up and/or face turn toward the unaffected eye to fuse. On version testing Brown syndrome might be confused with an inferior oblique muscle (IO) palsy. However, there are three important distinguishing features.

1. Because with Brown syndrome the eye cannot move up due to the restriction of the SO, there is a splaying out in upgaze, resulting in a V-pattern, whereas an IO palsy is accompanied by an A-pattern for the reasons outlined in Chap. 7, "A-Pattern, V-Pattern, and Other Alphabet Pattern Strabismus."
2. An IO palsy should have significant Bielschowsky head tilt difference because of the reasons presented in Chap. 8, "Vertical Deviations," but with Brown syndrome the head tilt difference is typically minimal.
3. Forced ductions are positive for elevation in adduction with Brown syndrome, and are normal with IO palsy.

Congenital Brown syndrome is usually unilateral but can occur bilaterally. It is usually sporadic, but there are patients for whom it appears to be hereditary.


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## Pearl

*A limitation of elevation in adduction in the presence of a V- or Y-pattern is probably Brown syndrome*. If an A-pattern is present, it is likely an IO palsy.

Acquired cases, in the absence of trauma or orbital tumor, are usually due to inflammatory disease. They are often associated with tenderness in the region of the trochlea. Some patients feel or hear a click on attempted elevation if there are inflammatory nodules on the tendon that get "stuck" passing through the trochlea.

## Basic Information

*Clinical Course*

Most cases of congenital Brown syndrome remain stable; however I have seen some that get worse over time, presumably due to the development of ipsilateral inferior rectus muscle contracture. There are also recent reports of cases of congenital Brown syndrome spontaneously resolving with one series reporting as high as a 75% resolution rate in a series of 32 patients [1]. This has not been my experience. I often just treat mild cases of Brown syndrome conservatively with observation, and I have only seen it resolve very rarely. My experience is similar to that reported from Iowa where they found that only 3% of patients with Brown syndrome resolved, 12% improved but did not resolve, 76% were unchanged, and 5% worsened [2]. Cases that are secondary to systemic autoimmune disease may exacerbate or improve as the systemic condition varies.

## Advanced Information

*Workup of Acquired Brown Syndrome*

The trochlea is akin to a synovial joint and as such can be subject to a tenosynovitis. This can result in an inflammatory Brown syndrome. When a patient with a known history of inflammatory disease presents with an inflammatory Brown syndrome, no further workup is needed. If there is no known history of autoimmune disease, a basic rheumatoid or autoimmune workup is indicated. However, in my experience, this almost never turns out positive. Similarly, the trochlea is a very difficult area to image, and I have rarely found orbital imaging helpful in these patients. Nevertheless, in select patients it may be worth doing, especially if there is suspicion of an orbital tumor.

## Advanced Information

*Treatment of Inflammatory Brown Syndrome*

There are reports of success with the use of oral nonsteroidal anti-inflammatory agents. Although I typically try them initially, I have been uniformly disappointed in the results. I have had considerable success with steroid injection in the region of the trochlea. Typically I inject 1 cc of either triamcinolone or Celestone Soluspan (Merck & Co, Whitehouse Station NJ, USA) through the upper lid, aiming into the anterior superior-nasal quadrant. I aim in the general region of the trochlea but do not make any attempt to be specifically in the trochlea (Fig. 15.1). Usually there is a

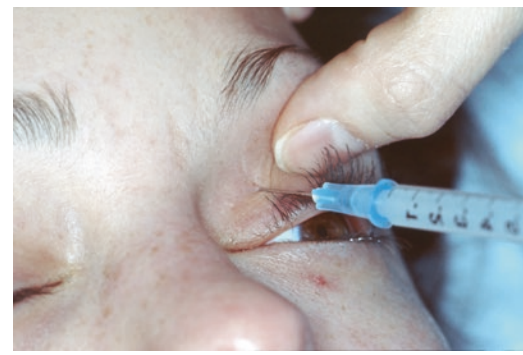

**Fig. 15.1** For treating inflammatory Brown syndrome, 1 cc of corticosteroid is injected through the upper eyelid into the anterior superior-nasal quadrant in the region of the trochlea

positive response within a day or so—often complete resolution. In cases that do not respond, I will repeat the injection in a month or so, and in some cases have had a good response after retreatment. In some cases, one injection has a complete and lasting effect. In others, the effect wears off after several months, in which case I will re-treat. Thus far I have never had to perform more than three injections on the same patient. For example, I injected the trochlear region in a 22-year-old man, and got complete resolution that lasted 2 months. A repeat injection provided complete resolution for the subsequent 6 months. His third injection also resulted in resolution, and he has been symptom free for the last 18 years.

I have the impression that steroid injection is more effective in the cryptogenic cases of inflammatory Brown syndrome, and less so in those associated with identifiable autoimmune disease. Some cases of Brown syndrome secondary to sinusitis may resolve after the sinusitis is treated.

## Pearl

Corticosteroid injection into the region of the trochlea is often (but not always) effective for treating inflammatory Brown syndrome.

## Basic Information

*Surgical Treatment of Congenital and Acquired Brown Syndrome*

I limit treatment of Brown syndrome to those patients who have a HYPO in primary gaze with either an associated abnormal head posture (congenital cases) or a diplopia (acquired cases). If the only abnormality is on gaze up and in, I do not recommend surgical intervention in part because of the reports of spontaneous resolution, but also because surgery for Brown syndrome has a higher complication rate than other strabismus conditions.

Initially, it was felt that Brown syndrome was caused by an abnormality of the sheath of the SO tendon, and that treatment should involve surgically stripping the sheath [3]. This was usually unsuccessful. It is now recognized that the tight SO tendon must be tenotomized or lengthened. Early reports of free tenotomy of the SO described the frequent occurrence of iatrogenic SO palsy in the affected eye, as one might expect. This led Parks to advocate simultaneous SO tenotomy and ipsilateral IO recession [4]. He found that approach effective both in treating the Brown syndrome and in avoiding an iatrogenic SO palsy. I have had good results with that two-muscle procedure, but switched to a split tendon lengthening procedure of the SO (see Fig. 13.12). I feel that equally good results can be obtained with a guarded tenotomy of the SO (chicken suture) or the use of a silicone expander. I prefer the split tendon lengthening for the reasons outlined in Chap. 13, "How to Perform Superior Surgery on the Inferior Oblique and Avoid Inferior Surgery on the Superior Oblique."

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# Duane Syndrome

# 16

*God and Satan are walking down the street and they see this brilliantly shiny object on the ground. God reaches down and picks it up and says "Ah, it's truth." And Satan says, "Oh, yes, give it to me. I'll organize and classify it."*

*—Tale told by Ram Dass*

*Duane Duane, go away … —Rephrasing of nursery rhyme from the late 1500s*

## Basic Information

*Overview*

*Duane syndrome* is the most common form of aberrant innervation. It is the result of a maldevelopment in which the sixth cranial nerve either does not form or forms only partially, resulting in absence or greatly diminished lateral rectus muscle (LR) recruitment on attempted abduction [1, 2]. In addition, branches of the third cranial nerve are redirected to aberrantly innervate the LR, resulting in anomalous recruitment of the LR, most typically, on attempted adduction. Generally, fibers destined for the medial rectus muscle (MR) get redirected to the LR. Depending on how many fibers normally destined for the MR get redirected in this manner, there may be essentially normal, somewhat subnormal, or markedly abnormal MR innervation and strength [3, 4]. The amount of aberrant innervation to the LR is, therefore, often inversely proportional to the amount of normal MR innervation. Duane syndrome, when isolated and not part of a syndrome, is sporadic about 90% of the time. Occasionally, it is familial. Duane syndrome is usually unilateral, but bilateral occurrence has been reported in between 10 and 20% of cases [5]. Based on some elegant work studying adults with thalidomide embryopathy, a population in which there is a high incidence of Duane syndrome, Miller determined that there is an early sensitive period at approximately days 21–26 after conception during which the insult resulted in Duane syndrome [6].

## Basic Information

*Clinical Findings*

The hallmark of most classes of Duane syndrome is complete or partial underaction of the LR on attempted abduction. On adduction, the LR co-contracts to a varying degree, depending on the amount of anomalous innervation there is to the LR. This may result in globe retraction, which in turn causes narrowing of the palpebral fissure, upshoots and/or downshoots of the eye, and some limitation of adduction because the LR is co-contracting against the MR. The degree to which these findings occur is a product of the amount of aberrant innervation to the LR, the anatomic path of the LR with respect to the crest of the globe, how easily the LR can slip up or down along the surface of the globe, and if the MR is underinnervated. Depending on the balance of MR innervation and LR anomalous innervation, there may be an esotropia (ET), exotropia (XT), or orthophoria in the primary position. The majority of cases are ET with the primary position deviation usually being under 35 Δ. In unilateral cases, there is usually a face turn to allow fusion, and if


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amblyopia is not present, there is typically normal fusion in the preferred head posture. In unilateral cases, patients who are ET will have a face turn toward the side of the affected eye, and if XT toward the unaffected eye. Most patients with Duane syndrome deny experiencing diplopia on a day-to-day basis, even when their eyes are in eccentric gaze. However, on sensory testing or diplopia field testing, a diplopic response is found in gazes in which the eyes are not aligned. This speaks to the fact that suppression versus diplopia awareness is not an either/or phenomenon, but there are varying depths and qualities to diplopia.

Most patients with Duane syndrome will give a diplopic response on diplopia tests, even though they rarely complain of diplopia.

## Basic Information

*Natural History*

The aberrant innervation in Duane syndrome is congenital and does not change over time. However, secondary changes can occur in the muscles that can cause a change in the clinical appearance. Jampolsky feels that XT Duane syndrome is more common in adults, because over time the LR undergoes contracture [3]. I have seen children with ET Duane syndrome who became more esotropic over time as they developed MR contracture.

## Basic Information

*Differing Manifestations of Anomalous LR Innervation*

The hallmark of Duane syndrome is deficient innervation of the LR from the sixth cranial nerve and its anomalous innervation from the third cranial nerve. If the amount of aberrant innervation to the LR is mild, there may be little or no visible globe retraction on adduction. The greater the aberrant innervation, the more one will see either globe retraction or what Jampolsky refers to as "escapes" [3]. The most common escape is an upshoot and/or downshoot of the globe on adduction. This is caused by the co-contracting LR slipping up or down on the globe as an "escape valve" for the opposing contracting forces that would otherwise result in retraction [7]. A second type of escape is a limitation of adduction, especially if the MR is underinnervated. Rather than opposing contracting forces causing globe retraction, there is less MR force and hence less adduction. Finally, if the co-contracting LR completely overpowers the MR on attempted adduction, the eye will actually abduct, resulting in what is called synergistic divergence, also called "the splits" [3, 8]. Souza-Dias has correctly pointed out that the term *synergistic divergence* is not satisfactory, because the term *divergence* by itself implies both eyes going temporally in opposite horizontal directions [4]. He prefers the term *synergistic abduction*, which I agree is preferable.

## Basic Information

*Examination of the Patient with Duane Syndrome*

In addition to the usual aspects of the strabismus exam, there are several additional parameters you should assess in patients with Duane syndrome that are important for creating a surgical plan.

1. What is the degree and quality of LR co-contraction? Does it manifest as globe retraction, upshoots and/or downshoots, or limited adduction, and to what degree? This can usually be assessed by simple observation. See section below on upshoots and downshoots for further discussion.
2. Is there any LR function on attempted abduction in the horizontal plane, and if so to what

degree? Is there any abduction movement in upgaze and/or downgaze? If so, and it exceeds that seen in the horizontal plane, there is added LR recruitment on elevation and/or depression. This should be considered in the surgical plan. The mere presence of an XT in primary is not evidence of abduction! It could be function of a tight LR that does not recruit on abduction. Instead, abduction is defined as a temporal movement of the eye as the contralateral eye shifts from primary gaze to adduction. Conversely, a tight MR can restrict abduction. The MR often develops some degree of contracture in Duane syndrome. This tightness, although often relatively mild, could be enough to mask mild degrees of LR recruitment on abduction. Active force generation testing is needed to sort this out.

3. How much of the limitation of abduction is due to MR tightness versus failure of the LR to recruit on abduction? Forced duction testing is needed to sort this out, which can be deferred to the operating room.
4. Is adduction limited, and if so how much and why? Is it due to an underacting MR, a tight LR, or LR co-contraction? To sort this out, I recommend the combination of forced duction and active force generation testing (see Fig. 2.6).
   (a) Do forced ductions for adduction. If positive, the LR is tight. If not, there is either an under-innervated MR or a co-contracting LR limiting adduction.
   (b) Do active force generation to see if there is any degree of recruitment of the LR on attempted abduction. This should be done even if clinically there does not appear to be any abduction, because a tight MR can mask it if mild.
   (c) Have the patient fixate with the non-Duane eye halfway between primary and abduction. With the patient in that gaze, fixate the Duane eye and pull gently into adduction. If the LR is co-contracting, you will feel its actively generated force. If this force is minimal but the limitation of adduction is not trivial, there is probably an underacting MR. Then without altering your purchase on the forceps, have the patient move the non-Duane eye into adduction. You will feel the force of the previously co-contracting LR degrade, provided that there was no active force in the Duane eye LR on attempted abduction. This is a modification of the force degeneration test described by Romero-Apis, which provides confirmation [9].
   (d) To confirm if the MR is underacting, do active force generation testing on the MR for adduction.
5. Note if there is a small ET versus small XT of the Duane eye in adduction. This is important in surgical planning. An XT in adduction can be a result of LR co-contraction or an underacting MR.
6. Assess the preferred head posture.
7. Is the patient fixing with the Duane eye? In unilateral cases this may give rise to very erratic and seemingly bizarre eye movements in the non-fixing eye. Particularly if the Duane eye would be having upshoots or downshoots were it not fixating, there will be exponentially larger vertical movements in the non-Duane eye due to fixation duress. In bilateral cases, by definition the patient will be fixing with an eye with Duane syndrome. This can greatly exaggerate the need for addressing vertical problems in the other eye, because they will appear larger due to fixation duress.
8. Are there upshoots or downshoots? If so, determine if they are probably due to the LR slipping above and/or below the crest, or some other cause (see section below on upshoots and downshoots).
9. Are there secondary changes in vertical muscles? In most cases vertical deviations in Duane syndrome are caused by the behavior of the LR. At times, however, there can be secondary contracture or overaction of one or more of the vertical muscles, most commonly the superior rectus muscle (SR). In my experience this usually manifests as hypertropia when the non-Duane eye is in the primary position. If the hypertropia only occurs on attempted adduction it is probably not caused by secondary vertical rectus muscle changes.

**Table 16.1** Examination of the patient with Duane syndrome

| Clinical parameter | How to assess | Comments |
|---|---|---|
| Degree and effect of LR co-contraction | Observation | Upshoots, downshoots, retraction? |
| Any degree of LR recruitment on abduction | Observation plus active force generation | An XT in primary does not mean abduction |
| How much does MR tightness contribute to limitation of abduction? | Forced duction | |
| Is adduction limited and why? | Combination of forced ductions and active force generation. See text for details | Could be tight LR, co-contracting LR, or weak MR |
| Alignment in adduction: ET or XT | Observation | |
| Preferred head posture | Observation | |
| Might he be fixing with the Duane eye? | Observation | |
| Upshoots or downshoots? Mechanism? | Observation | Usually LR slipping up and/or down |
| Any secondary changes in vertical muscles | Is there a hypertropia in primary gaze? | |
| Any other associated neurologic or developmental issues | Exam. History | Always ask specifically about hearing loss in infants |

*LR* lateral rectus muscle, *XT* exotropia, *MR* medial rectus muscle, *ET* esotropia

10. Are there associated neurologic or developmental issues that may require attention, e.g., hearing impairment and *Goldenhar syndrome*? When the patient is an infant, always specifically ask about possible hearing issues. I have treated a number of infants for whom my diagnosis of Duane syndrome led to the early discovery of unsuspected hearing issues.

The above clinical parameters are summarized in Table 16.1.

## Pearl

When seeing an infant with Duane syndrome, always ask about and/or test for hearing impairment that may not be known to the parents. Alert the primary care practitioner about possible hearing issues.

## Basic Information

*Upshoots and Downshoots*

The upshoots and downshoots that occur in Duane syndrome resemble inferior oblique and/or SO overaction; however, in my experience they are usually not caused by the oblique muscles. Jampolsky points out that they are escape mechanisms for the opposing forces of the MR contracting in adduction, and it being opposed by the co-contracting LR [3]. If, on attempted adduction, the co-contracting LR stays on the crest of the globe, there will just be globe retraction. But if the LR slips above the crest, the escape mechanism manifests as a knife-edge upshot of the eye; if it slips below the crest, there is a downshoot (Fig. 16.1). This bridle effect has been well described by Souza-Dias, Scott, and Jampolsky [3, 4, 10]. Figure 16.2 is a clinical example of these escape mechanisms.

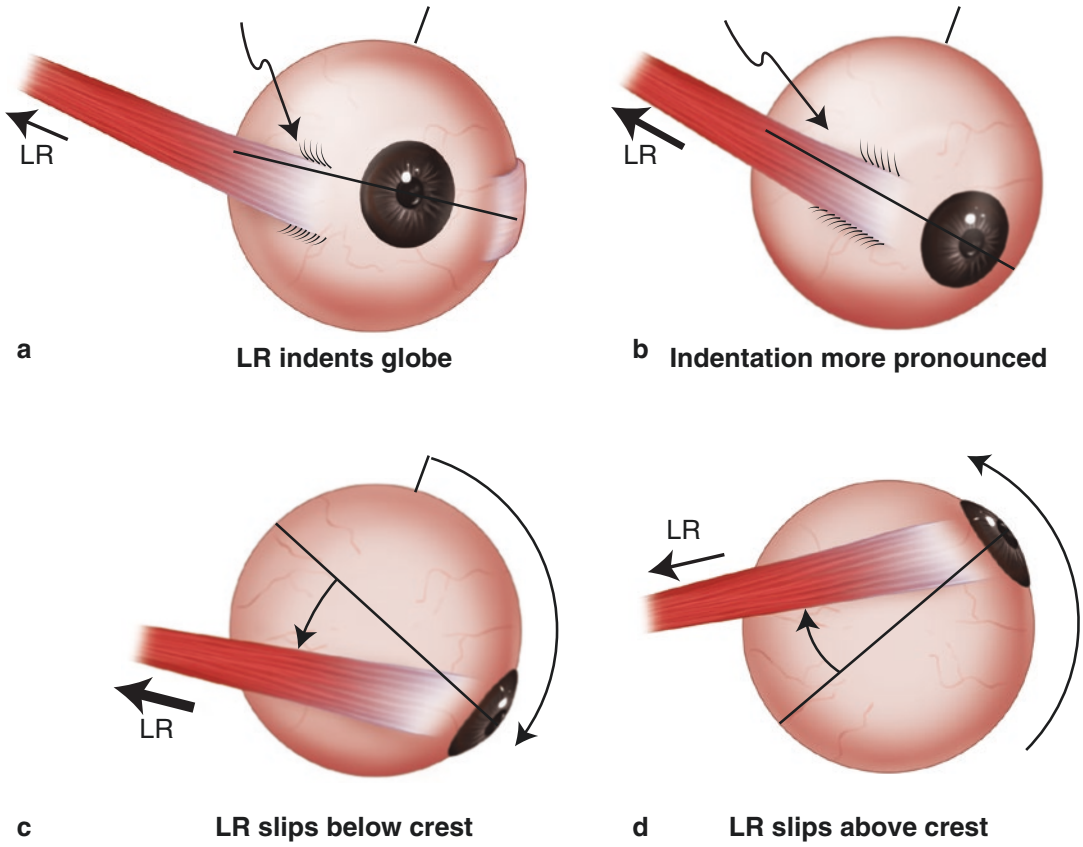


**Fig. 16.1** (**a**) If the co-contracting lateral rectus muscle (LR) stays on the crest of the globe, there is just globe retraction on attempted adduction. (**b**) If the LR co-contraction increases, there is more indentation of the globe by the LR and an increase in retraction. (**c**) If the LR slips below the crest, there is a downshoot on attempted adduction. (**d**) If the LR slips above the crest, there is an upshoot on attempted adduction

## Question

How can I tell if the upshoots and downshoots are due to this bridle effect vs. oblique muscle overaction or secondary vertical rectus muscle changes?


## Reply

If a patient shows an upshoot while the eye is taken into adduction while above the midline, and a downshoot while taken into adduction below the midline, you can be sure that the co-contracting LR is the cause due to the bridle effect mechanism. However, not all patients who exhibit the bridle effect have both upshoots and downshoots. Also, if a patient has an upshoot, and you have them adduct above the midline to bring out the upshoot, and then from the adducted position bring them into gaze down and in, the upshoot may not only persist but get worse, because the LR, once slipped upward, has nowhere to go except further up if from that position the eye is stimulated to depress. The converse holds for a downshoot getting worse on attempted elevation from the down and in position. This finding, if present, is diagnostic for the bridle effect, but the absence of the finding by no means rules out the bridle effect.

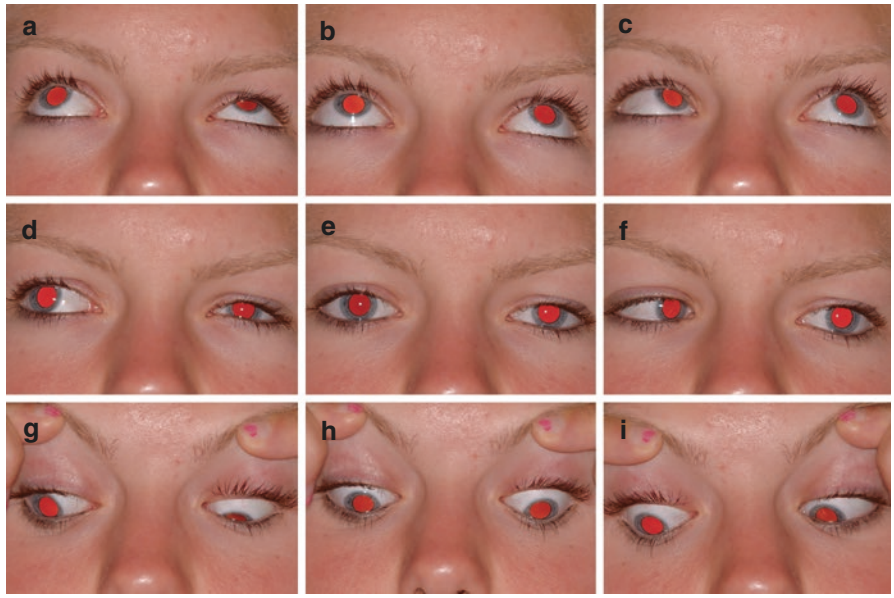


**Fig. 16.2** Motility of a patient with Duane syndrome OS with a lot of lateral rectus muscle co-contraction. (**a**) up right; (**b**) straight up; (**c**) up left; (**d**) middle right; (**e**) middle straight; (**f**) middle left; (**g**) down right; (**h**) straight down; (**i**) down left. On attempted adduction above the midline there is an upshoot, below the midline there is a downshoot, and on direct horizontal side gaze there is substantial globe retraction

In the absence of the aforementioned findings, you should consider the relatively unlikely possibility that there are other causes. True oblique muscle overaction is characterized by a more gradual elevation or depression in adduction than the "knife-edge" vertical movement caused by the bridle effect. This distinction, however, can be hard to make clinically; it can be subtle. I find the presence of significant fundus torsion to be helpful in tipping the scales toward true oblique muscle overaction; however, even that can be misleading. Many people feel that some cases of Duane syndrome have anomalous innervation to the vertical rectus muscles, which may co-contract on adduction. This is largely based on EMG studies showing the SR recruiting during an upshoot [11, 12]. Jampolsky strongly feels that there is never anomalous innervation to the vertical rectus muscles in Duane syndrome and points out that the EMG findings suggesting otherwise were probably recording passive changes in the muscle as the eye elevated or depressed for other reasons [3]. He feels that the vertical rectus muscles can secondarily become contractured and "overact" because of repeated elevation or depression of the eye due to the bridle effect mechanism. I have a hard time with that explanation. Most patients with Duane syndrome spend most of their days with the eyes in their position of fusion and tend to avoid repeatedly looking with the eye in adduction. So, it is not clear to me how contracture would occur. Also, if vertical rectus contracture did occur, I think the vertical deviation would be less in adduction than closer to primary; however, I realize that there could be the additive effect from the bridle mechanism. Nevertheless, there are some patients in whom there appears to be contracture/overaction of a vertical rectus muscle. As stated earlier, they tend to have a vertical deviation that is not limited to adduction.

## Important Point

Most upshoots and downshoots in Duane syndrome are not caused by oblique muscle overaction but are caused by the co-contracting LR.

## Question

I have a 2-year-old patient with a 30 prism diopters (Δ) ET in primary, and −4 for abduction of both eyes. There are no retractions and no upshoots or downshoots. He is too young for active force generation testing. How can I distinguish bilateral sixth nerve palsies from bilateral Duane syndrome?


## Reply

If the size of the ET is not what you would expect from the degree of LR underaction, you are probably not dealing with bilateral sixth nerve palsy. If there is −4 to abduction bilaterally caused by bilateral sixth nerve palsy, the ET in primary should be very large and in the range of 60 Δ or much more.

## Advanced Information

Traditionally, Duane syndrome is classified as being type 1, 2, or 3 based solely on whether there is ET or XT in primary gaze and if the main gaze limitation is abduction, adduction, or both. Some people have added types 4 and 5 to this classification, but there is no consensus as to what constitutes these two added classes. In general, classification systems are useful if they identify different underlying pathophysiologies, and perhaps more importantly indicate different treatment approaches (think of Knapp's classic classification of fourth cranial nerve palsy [13]). In this regard, I do not find any of the existing classifications of Duane syndrome to be useful. Proper treatment options include many variables that are not incorporated in the existing classification schemes, e.g., the degree of co-contraction of the LR, presence of upshoots or downshoots, presence of any LR function from the sixth nerve, size and direction of the deviation in adduction (ET vs. XT), as well as other factors listed above. Any useful classification scheme should take these factors into account. In fact, I view the traditional classes of Duane syndrome as having the same underlying pathophysiology, but just in varying degrees. Rather than considering ET Duane, orthophoric Duane, and XT Duane as different conditions, I see them as just points on a spectrum. An ET Duane is a milder form, in which the amount of LR co-contraction is mild enough that it does not kick in and halt adduction until the eye is inwardly deviated. An orthophoric Duane has more co-contraction of the LR, which puts a brake on adduction from MR tonus before the eye gets into adduction. An XT Duane has more severe co-contraction of the LR, which stops adduction even when the eye is in an abducted position referable to primary. Not surprisingly, patients with XT Duane have the worst retraction, upshoots, and downshoots. For ET Duane, the bigger the ET, the less one finds retraction, upshoots, and downshoots. So Duane types 1 and 3 represent different points on the spectrum of severity. Type 2 Duane is somewhat different. In this type there is at least a modest amount of normal innervation to the LR and also a good deal of aberrant innervation of the LR by fibers that normally go to the MR. Consequently, there is some abduction, but limited adduction due to a combination of the MR being under-innervated and the LR co-contracting on attempted adduction.

## Important Point

Paradoxically, the greater the ET, the less severe the Duane syndrome.

## Advanced Information

*Special Forms of Duane Syndrome*

As described in Chap. 7, "A-Pattern, V-Pattern, and Other Alphabet Pattern Strabismus," a Y-pattern associated with pseudo-inferior oblique overaction is a variant of Duane syndrome. The LRs co-contract with the SRs, causing a splaying out of the eyes in upgaze [14].

## Advanced Information

*Synergistic Abduction*

This condition was called synergistic divergence or "the splits" [15]. It occurs when the co-contracting LR so completely overpowers the ipsilateral MR that the eye abducts when a normal eye would adduct. Consider the hypothetical situation of synergistic abduction in the left eye. When the fixing right eye looks into right gaze (abducts), the left eye also abducts, hence the descriptive term "the splits." Synergistic abduction can occur idiopathically, in which case it is probably due to a marked amount of aberrant innervation to the LR and possibly a congenitally weak or under-innervated ipsilateral MR. It also can occur after prior surgery in the form of either excessive weakening of the MR in a Duane eye with a lot of aberrant innervation to the LR or ipsilateral LR resection.

## Basic Information

*Surgical Recommendations*

Surgical recommendations are influenced by the factors in Table 16.1. For some clinical scenarios I may give several treatment options, with there being relative trade-offs between them. In many of these cases, I believe that it is the patient's or parents' right to choose, given a full explanation of the relative pros, cons, and trade-offs of differing treatment options (assuming that the patient or parents have the intellectual capacity to make such a choice and/or wish to). For me this comes up with Duane syndrome more than any other condition. I feel that there are several patterns of Duane syndrome for which I might do bilateral MR recessions, or a vertical rectus transposition (perhaps with ipsilateral MR recession), and find either acceptable. The choice between the two is a matter of trade-offs. I think that transposition surgery gives the best outcome when it works well, but is more prone to adverse side effects like induced vertical deviations requiring enhancement. Bilateral MR recessions are safer, simpler, and less prone to adverse outcomes. But even in the best cases, they rarely expand the range of single binocular vision as well as transposition surgery. So if I feel that the patient or parents are appropriate for this discussion, I tell them that there is one approach that, when successful, gives the best outcome. But it is trickier and more likely to require secondary surgery. There is another approach that gives good results, but is less prone to causing complications, but never gives quite as good results. So if your bottom line is to get the best result possible, and you are willing to accept a higher chance of needing more surgery, I suggest the transposition procedure. But if your bottom line is to avoid needing an enhancement, and are willing to accept improvement, but not as much as with the transposition, go with the MR recession OU. Both are acceptable choices.

## Pearl

In many cases both bilateral MR recession or vertical rectus transposition are good choices, with different risk/benefit ratios. If the patient or parents are able to make an informed choice, the choice should be theirs.

## Basic Information

*Discussion of Different Surgical Procedures for Duane Syndrome*

### Ipsilateral MR Recession

This is the most commonly performed procedure for esotropic Duane syndrome, and in most cases it is not the best choice. It is popular because it is seductively appealing. It will almost always eliminate the face turn which is initially a main complaint; is safe, simple, and quick; and is relatively free of major initial complications. Parents are immediately pleased, as they see resolution of the ET and face turn. The problem is that if there is any significant co-contraction, it will further decrease adduction and narrow the range of single binocular vision.

> Although ipsilateral MR recession is safe and simple, and usually improves the AHP in Duane syndrome, it may not be the best choice.

### Bilateral Symmetric MR Recession (More in the Non-Duane Eye)

This is one of the two most commonly used treatment options in my schema for treating esotropic Duane syndrome. It allows me to limit my MR recession in the Duane eye to 4 or 5 mm, and not limit adduction as much as would occur if I tried to correct all the ET with a single MR recession. To get optimum effect, the MR recession in the non-Duane eye should be very large, 7 mm or more. Jampolsky has recommended 10 mm, and, in fact, I observed him do this in a 4-year-old girl, who showed full adduction of that eye on the day after surgery [3]. Because of the lack of abduction in the Duane eye, this will never cause an XT in the field of the LR of the Duane eye. Furthermore, causing fixation duress in the non-Duane eye as it moves to primary tends to inhibit the innervation to the MR of the Duane eye. This will allow for more ET correction from a smaller MR recession in the Duane eye, because the Duane eye MR is innervationally inhibited. This also will prevent MR contracture from occurring.

### Posterior Fixation of the MR in the Non-Duane Eye

The presumed rationale for this is similar to that for doing a large MR recession in the non-Duane eye—specifically, to cause fixation duress in the direction of the Duane LR. However, this is less effective in my opinion because the effect of posterior fixation on reducing torque of a muscle does not come into play until the eye is about 30 degrees into the field of action of the muscle; it has minimal effect in the primary position and hence would not have the benefits of a large recession when it comes to decreasing the ET in the Duane eye or inhibiting the MR in the Duane eye in primary gaze [16, 17]. In general, I do not recommend this procedure as it adds very little.

### Superior and Inferior Rectus Transposition to the LR with or Without Foster Augmentation Sutures

Transposition procedures can give the best results in several patterns of Duane syndrome [18, 19]. The trade-off is that the procedure is more complex than simple MR recession, and more can go wrong. One does have some risk of inducing a vertical deviation and/or getting superior oblique incarceration. A key to preventing adverse outcomes is to do forced ductions repeatedly during the procedure and adjust the position of the muscles accordingly. Transposing both vertical rectus muscles with augmentation sutures will overcome a small-to-moderate amount of MR contracture. But if the MR is fairly tight, it will need to be recessed. If so, transposition of the temporal halves or three-fourths of the vertical rectus muscles is an option that will spare the anterior segment circulation.

### Transposition of the SR to the LR with Augmentation Combined with MR Recession in the Duane Eye

This procedure is currently replacing transposition of both vertical rectus muscles [20, 21]. Although it seems intuitive that this should cause a vertical and torsional problem, it appears to work well the vast majority of the time. It has the advantage of allowing a recession of the MR with low risk of anterior segment ischemia. See Chap. 10, "Strabismus Surgery," for further discussion.

### LR Recession in the Duane Eye

If there is significant globe retraction, upshoots, or downshoots, a standard recession of the LR is usually inadequate. But in the absence of those findings, it is sometimes useful to recess the LR in the Duane eye if the co-contraction is resulting in an XT in adduction. If so, it should be combined with bilateral MR recessions.

### Recession with Y Splitting of the Duane LR

If uphoots and/or downshoots are moderate, this procedure is helpful. I find that it rarely eliminates the problem, but can provide substantial improvement [3, 22]. As described by Jampolsky, the LR is split longitudinally at the insertion as far back as possible. The superior half is supraplaced 10 mm and the inferior half infraplaced 10 mm, so there is 20 mm between them. Although some authors do not emphasize the need to also do a generous recession, Jampolsky feels that is important, and I agree [15]. I typically do 7–9 mm of recession of both halves, depending on forced ductions. This procedure gets the LR off the crest of the globe. Figure 16.3 shows how this will prevent the LR from slipping up or down when it co-contracts.

### Posterior Fixation of the Duane LR for Treating Upshoots and Downshoots

This procedure has been advocated as a means of preventing the LR from slipping up or down and hence preventing upshoots and downshoots [10]. I think that because upshoots and downshoots are "escape mechanisms" to prevent globe retraction, this procedure appears to decrease the upshoots and downshoots at the expense of increasing retraction. It is not part of my recommended armamentarium.

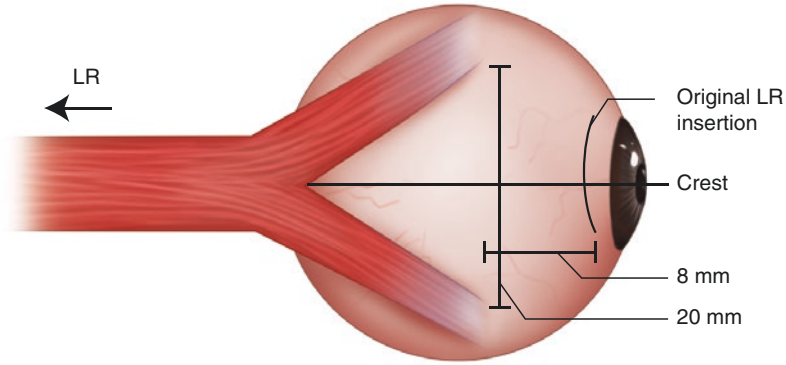


**Fig. 16.3** Depiction of Y-splitting of the lateral rectus muscle (LR) with 20 mm separation between the split halves and a recession of 8 mm. Prior to Y-splitting and recession the co-contracting LR causes an upshoot or a downshoot of the globe as depicted in Fig. 12.1. After Y-splitting and recession the two split halves will anchor the LR from slipping up or down

## Orbital Wall Fixation of the LR

When profound weakening is needed of the LR due to severe co-contraction, globe retraction, upshoots and/or downshoots, fixation of the LR to the lateral orbital wall periosteum as described in Chap. 10 are the most effective options [23].

### Vertical Rectus Recession

In those cases in which there is a vertical deviation in primary gaze that does not appear to be due to LR slippage, recession of the SR may be indicated, particularly if forced ductions show it to be tight. This occurs very infrequently.

### Supraplacement of the LRs

As mentioned in Chap. 7, the pseudo-inferior oblique overaction which is associated with a Y-pattern (a variant of Duane syndrome) may not need treatment if there is no deviation in primary. If there is, and surgery is needed, full tendon supraplacement of the LRs is effective in collapsing this pattern [14].

### Resection of the Duane LR

Traditional teaching is never to resect the LR in an eye with Duane syndrome, because doing so will worsen the co-contraction and globe retraction and run the risk of causing synergistic abduction. Although recently some authors have reported good results with LR resection in select cases of Duane syndrome, specifically those with minimal co-contraction, I have never found it necessary to do so.

*For specific recommendations of surgical procedures for different manifestations of unilateral Duane syndrome that have no normal LR force on attempted abduction, see* Table 16.2. For patients with unilateral Duane syndrome who have some normal LR force on attempted abduction, follow the guidelines in Table 16.2 with these caveats. Regardless of their being ET, orthophoric, or XT, and with any degree of LR co-contraction, scale back ET surgery or increase XT surgery accordingly. Do not create fixation duress to the Duane eye LR with a large recession of the contralateral MR.

**Table 16.2** Surgical recommendations for unilateral Duane syndrome with the absence of normal LR force on attempted abduction

| Primary position or pattern type | Lateral rectus muscle co-contraction | Caveats | Surgical recommendation |
|---|---|---|---|
| ET | Small | ET small (≤ 20) | Recess Ips MR 3–5 mm (to free forced ductions) |
| ET | Small | ET larger (> 20) and ET ≥ 10 in adduction | Recess Ips MR 6–7 |
| ET | Small | ET larger (> 20) and ET < 10 in adduction | Recess Ips MR 4–5 (to free forced ductions) & recess con MR 6–9 |
| ET | Moderate: Definite but mild retraction, and/or only mild upshoots and/or downshoots | ET small (≤ 20) and ET ≤ 10 in adduction | Recess Ips MR 3–5<br>Recess con MR 6-9[a]<br>***or***[b]<br>Transpose Ips SR to LR with augmentation and recess Ips MR amount pending forced ductions[a]<br>***or***[b]<br>Transpose Ips SR and IR to LR with augmentation ***if*** Ips MR is only mildly tight[a] |
| ET | Moderate: Definite but mild retraction, and/or only mild upshoots and/or downshoots | ET small (≤ 20) and ET < 10 in adduction | Recess Ips MR 3–5<br>Recess Ips LR 7–9<br>Recess con MR 7–10<br>***or***[b]<br>Transpose Ips SR to LR with augmentation and recess Ips MR amount pending forced ductions[a]<br>***or***[b]<br>Transpose Ips SR and IR to LR with augmentation **if** Ips MR is only mildly tight |
| ET | Marked: Significant retraction and/or upshoot and/or downshoot | ET in primary is almost always < 20 | Recess and Y-split Ips LR 7–9 mm, or for more effect fixate Ips LR to orbital wall<br>***And***<br>Transpose Ips SR to LR with augmentation[a] and recess Ips MR amount pending forced ductions<br>***or***[b]<br>Transpose one-half or three-fourths tendon of Ips SR and IR to LR with augmentation ***if*** Ips MR is only mildly tight |
| Orthophoric to XT ≤ 20 | Is always marked: Significant retraction and/or upshoot and/or downshoot | Forced duction and active force generation show poor Ips MR function | Recess and Y-split Ips LR 6–8 mm[c] |
| Orthophoric to XT > 20 | Is always marked: Significant retraction and/or upshoot and/or downshoot | Forced duction and active force generation show poor Ips MR function | Recess and Y-split Ips LR 7–9 mm & recess con LR<br>***or***[b]<br>For more effect fixate Ips LR to orbital wall and do large con LR recession[c] |
| Hypertropia or hypotropia in primary with any of the above patterns | | If vertical is not due to LR slippage but is due to vertical rectus activity IO OA (uncommon—look for fundus extorsion) | Recess Ips SR or IR an amount commensurate with deviation or weaken Ips IO, in addition to above surgical plans |

**Table 16.2** (continued)

| Primary position or pattern type | Lateral rectus muscle co-contraction | Caveats | Surgical recommendation |
|---|---|---|---|
| Synergistic abduction | | | Fixate Ips LR to orbital wall<br>***And***<br>Transpose Ips SR to LR with augmentation |
| Pseudo-IO OA with Y-pattern | Is always minimal | Surgery not needed if no deviation in primary | Usually recess LR OU if XT or recess MR OU if ET and supraplace LRs full tendon width |
| Any pattern, moreso with XT | Any pattern | | |
| ET, orthophoric, or XT | Any degree | Any degree of LR function on abduction | Follow above protocol but scale back ET surgery or increase XT surgery accordingly. Specifically do not create fixation duress to the Duane LR with large recession of the con MR |

*ET* esotropia; *Ips* Ipsilateral, eye *with* Duane syndrome; *MR* medial rectus muscle; *Con* contralateral, eye *without* Duane syndrome; *SR* superior rectus muscle; *LR* lateral rectus muscle; *IR* inferior rectus muscle; *XT* exotropia; *IO* inferior oblique muscle; *OA* overaction; *OU* both eyes

[a]Provided the retraction, upshoots or downshoots are insignificant to be ignored. Otherwise will need Ips LR recession

[b]Intelligent patient or parents to participate in choice of options (see text)

[c]Vertical rectus transposition is to be avoided if the Ips MR is weak, as it will usually cause persistent exotropia

## Important Point

The most important factor in determining how much ipsilateral MR recession is safe to do is the amount of aberrant innervation to the ipsilateral LR.

## Important Point

The presence of some degree of normal LR force on attempted abduction in the Duane eye mandates scaling back ET surgery or increasing XT surgery.

## Question

An 18-year-old woman has a unilateral Duane OD. She is orthophoric in primary with no head turn. But she is −4 to abduction OD and has marked narrowing of the palpebral fissure on adduction, upshoots, and downshoots. What can I do for her?


## Reply

This is an example of where perfect may be the enemy of good. One can make a good case for not operating. But if you feel the wish to intervene, you first need to determine what is bothering her. Is it the retraction, narrowing, upshoots and downshoots, or limited abduction? If the former, a Y-splitting and modest recession will help with little (but not zero) chance of making her worse. For more effect, you can do a larger LR recession with Y-splitting, and also recess the ipsilateral MR on an adjustable suture. If you feel that you need to both increase abduction and get maximum effect on the co-

contraction escapes, you can fixate the LR to the periosteum and transpose the ipsilateral SR to the LR insertion. This will also have the greatest risk of an adverse outcome.

## Question

My patient has a right ET Duane syndrome. She has 20 Δ of right ET in the forced primary position. She prefers a right face turn of 30 degrees and is orthophoric with this head posture. Twenty Δ is about 10 degrees, so I am perplexed why her face turn is more than that. I would think that with a face turn of 10 degrees her visual axes would line up and she would fuse.


## Reply

It is usually the case that the face turn with Duane syndrome is greater than the number of degrees of misalignment in the primary position for the following reason: Consider a hypothetical patient with a 10 degree right ET that is comitant, unlike the ET that accompanies Duane syndrome. If this hypothetical patient maintains distance fixation with the left eye and turns her face to the right, both eyes will move into left gaze by an amount equal in degrees to the face turn—that is the definition of comitant. The two visual axes will never line up in side gaze. On the other hand, if the right eye was frozen in the orbit and did not move at all as the face turned, the visual axes would line up with a face turn that exactly equaled 10 degrees. Duane syndrome is somewhere in between these two extreme examples. As your patient turns her face to the right, the left eye will move into left gaze an amount exactly equal to the face turn in degrees. The Duane right eye will also move into left gaze, but not as much as the left due to the co-contacting LR, which is a brake on adduction. The face turn must be greater than the primary position misalignment in degrees, before the abducting left eye matches up with the right eye, which is adducting to a lesser degree.

> … and never the Duane shall meet.—Rephrasing of Rudyard Kipling, *Back-room Ballads,* 1892(à propos of trying to balance alignment in bilateral Duane Syndrome)

## Advanced Information

*Bilateral Duane Syndrome*

This is one of the trickiest conditions the strabismus surgeon may be called upon to operate. Because there are abnormal eye movements in both eyes, any movement of either eye, if fixing, may exponentially compound movements in the other eye due to fixation duress. When planning surgery, it is important to take into account all relevant factors, including amount (if any) of normal LR innervation, amount of aberrant innervation to the LRs, and strength of the MR. I agree with Jampolsky that it is useful to dichotomize bilateral Duane syndrome into those patients with fusion and those without fusion [3].

1. *Bilateral Duane syndrome with fusion*. If a patient with bilateral Duane syndrome has fusion in or close to primary gaze, and does not have a significant head posture, it is often best to ignore mild-to-modest retraction and/or upshoots and downshoots. The potential for disrupting fusion with surgery is high and might not be worth the risk. As Jampolsky said, "The lesser the surgical indications, the greater the likelihood of post-operative complications" [3]. Use prisms whenever possible to correct an abnormal head posture. If, however, these "escape phenomena" are significant, one may consider surgery. The same principles apply as with unilateral Duane syndrome. However, you need to keep in mind the role of fixation duress. Any change in

force vectors to the fixing eye will be magnified in the fellow eye. For example, a patient may have an alphabet-pattern ET (A, V, Y, or λ) with ET in primary. If there is no normal innervation to the LRs, it may be safe to recess the MRs for the ET. However, if there is any normal innervation to the LRs, recessing the MRs a "standard" amount may open the door for a significant postoperative XT.

> The lesser the surgical indications, the greater the likelihood of post-operative complications.—Art Jampolsky, MD

2. *Bilateral Duane syndrome without fusion*. These patients tend to have large deviations (ET or XT) because the fixing eye has abnormal movements, resulting in fixation duress to the yoke muscles in the non-fixing eye. Again, similar principles apply as with treating unilateral Duane syndrome. However, you need to be aware of the leveraging effect that anything you do in the fixing eye will have profound ramifications in the non-fixing eye due to changes in fixation duress.

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# Miscellaneous Strabismus Syndromes

# 17

## Monocular Elevation Deficiency (MED)

### Basic Information

*Overview*

The original term for this condition was *double-elevator palsy*, because it was felt to be due to a paralysis of both the ipsilateral superior rectus muscle (SR) and inferior oblique muscle (IO). We now recognize that it can be caused by inferior rectus muscle (IR) restriction in isolation [1, 2], supranuclear causes [3, 4], or paralysis of the SR without ipsilateral IO weakness [2–4]. There also can be IR restriction secondary to ipsilateral SR paresis. It is characterized by an inability to elevate above the midline in all horizontal gaze fields.

### Basic Information

*Clinical Exam*

The different causes of MED can usually be sorted out by clinical exam alone.

In cases of SR paresis, or IR restriction plus SR paresis, there is usually a hypotropia in the primary position and, depending on fusion potential, a chin-up head posture. True ptosis is often present. Patients with a supranuclear etiology may be ortho in primary or have a hypotropia. They do not have ptosis but if hypotropic may have pseudoptosis. The presence of sensory fusion is typical, and the absence thereof speaks more for a paretic SR. Patients with isolated IR restriction are mixed with either a hypotropia or an orthophoria in primary gaze. They should have no ptosis if ortho in primary and have pseudoptosis if there is a hypotropia in primary. Forced ductions are crucial to make the proper diagnosis and they may need to be deferred to the operating room, depending on the age of the patient. Typically, they are markedly positive with isolated IR restriction due to fibrosis, and only slightly to moderately positive with IR restriction due to secondary contracture if there is also SR paresis. You should try to assess vertical saccades or test active force generation of the SR if possible. They will be normal from below midline to midline in cases of supranuclear MED and isolated IR restriction. They are reduced if there is SR paresis. Many patients with IR restriction have an exaggerated lower eyelid crease that increases on attempted upgaze [1].

### Pearl

True ptosis suggests SR weakness, pseudoptosis suggests IR restriction or a supranuclear etiology with a hypotropia in primary, and neither suggests a supranuclear cause with orthophoria in primary.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_17

### Pearl

The presence of an exaggerated lower eyelid crease that increases on attempted upgaze is strong evidence for IR restriction. It is more likely to be present with isolated IR restriction than IR restriction secondary to SR paresis.

### Pearl

Forced ductions are markedly positive with isolated IR restriction and more apt to be slightly or moderately positive if the IR restriction is secondary contracture due to SR paresis.

### Pearl

Congenital supranuclear MED is more likely to have sensory fusion. The absence of sensory fusion is more common with SR paresis.

### Basic Information

*Differential of Congenital MED*

Most other causes of congenital limitation of elevation are easy to distinguish from MED because of associated findings or motility factors. Brown syndrome shows more limitation of elevation in adduction than in abduction. Duane syndrome can, in rare circumstances, have a limitation of elevation that might be confused with MED; however, other signs of Duane syndrome should be evident. Craniofacial syndromes may have agenesis or attenuation of the SR, but in those cases the presence of the craniofacial syndrome should be obvious. Third cranial nerve palsy, if complete, will have other muscles involved and may have pupil involvement. Congenital cranial dysinnervation syndromes (formerly called congenital ocular muscle fibrosis syndrome) usually have other muscles and gaze directions involved.

### Basic Information

*Differential of Acquired MED*

This includes any acquired cause of IR restriction such as orbital fracture, thyroid eye disease (TED), orbital myositis, anesthetic toxicity [5, 6], orbital tumor, postscleral buckle or seton, or eye muscle surgery resulting in an oblique incarceration syndrome [7, 8]. It also includes any acquired cause of SR weakness such as neurologic cause of partial third cranial nerve palsy, chronic progressive external ophthalmoplegia, or myasthenia gravis. In addition, cerebral vascular accidents can result in supranuclear cases of MED.

### Basic Information

*Treatment*

Not all cases of MED require treatment. If there is little or no hypotropia in primary, there is no significant chin-up posture, and the patient is not symptomatic, observation is appropriate. If treatment is indicated, your choice of procedures should be dictated by the presence of IR restriction, degree of SR weakness, presence or absence of a Bell's reflex, and if the etiology is supranuclear.

If the Bell's reflex is normal, the etiology is probably supranuclear. If it is not, you need to determine the degree of IR restriction versus SR paresis.

If the IR is severely restricted, it should be recessed. The amount of recession should be determined more by intraoperative forced duction and spring-back testing after the muscle is recessed, rather than the size of the preoperative hypotropia. Recessions often need to be large ranging from 5 to 10 mm centering the eye about

5° below midline, and the conjunctiva may need to be recessed if it is tight. Metz found that 73% of patients with MED develop full elevation after recession of a restricted IR [2, 9].

In cases in which the IR tightness is secondary to SR paresis (mild-to-moderate restriction), there will be a residual hypotropia and/or limitation of elevation after IR recession alone. If you are able to make the diagnosis of concurrent SR paresis before surgery, it is wise to attempt to correct the problem in one procedure. Testing active force generation of the SR from downgaze toward primary is the definitive test, but this is hard to do in young patients. Other signs that may be helpful are the following:

1. The absence of an accentuated lower lid crease in the presence of IR restriction speaks for secondary IR restriction.
2. The absence of a hypotropia in primary in the presence of IR restriction rules out SR paresis. However, the converse is not true. The presence of a hypotropia in primary can be either isolated or secondary IR restriction.
3. Brisk saccades, perhaps with vertical optokinetic nystagmus testing, from downgaze toward primary, in the face of IR restriction will rule out SR paresis.
4. True ptosis is less likely to be present if there is no SR paresis, but pseudoptosis may be present with isolated IR restriction if there is a hypotropia in primary.

If you determine with a good degree of confidence preoperatively that you are dealing with secondary IR restriction and SR paresis, and if you wish to try to do things in one stage, you can combine IR recession with a partial Knapp procedure [10]. A full Knapp procedure not only runs the risk of anterior segment ischemia but often results in overcorrection if the ipsilateral IR is also recessed. In this setting I prefer to transpose only one-half of the medial rectus muscle (MR) and lateral rectus toward the SR and do not move them up all the way. Struck has observed that the Knapp procedure often limits downgaze; he prefers to do a contralateral SR recession of 10 mm with which he has had good results combined with ipsilateral IR recession of 5 mm (Michael C. Struck, MD, personal communication, 21 May 2015) [11]. If you are not confident that you can sort out preoperatively if there is concurrent IR restriction and SR paresis, it is best to plan a two-stage procedure if need be. Start with an IR recession. If there is a residual hypotropia and limitation of upgaze, either a contralateral SR recession of 10 mm or partial Knapp procedure can be done. Be very cautious in this situation about not doing a full Knapp procedure, as it is very likely to cause an overcorrection in this setting. I have seen good results with recessing the contralateral SR and prefer that approach.

A full Knapp procedure either combined with or following an ipsilateral IR rec has a high likelihood of overcorrection.

If there is SR paresis with little or no IR restriction, a full Knapp procedure is most commonly recommended. But here Dr. Struck's approach of a contralateral SR recession of 10 mm works well and is less likely to limit downgaze than the Knapp procedure. If there is no hypotropia in downgaze, the initial surgery should be limited to recessing the contralateral SR. However, if there is a hypotropia in downgaze, an ipsilateral IR recession should be added.

For supranuclear MED with no hypotropia in primary, surgery is often not needed. However, some patients have a hypotropia in primary and need surgery. If there is a good Bell's reflex, the IR will show minimal or no restriction on forced duction testing. A Knapp procedure has a high likelihood of causing an overcorrection if there is little or no hypotropia in primary. Here, a contralateral SR recession of 10 mm works well. Struck reports that about 20% show mild regression, in which case he recommends a 5 mm ipsilateral IR recession; 80% percent do not need an enhancement. If there is a large hypotropia in primary, a Knapp procedure is an effective operation but is more likely to limit downgaze than the aforementioned vertical rectus muscle procedures.

Table 17.1 summarizes these findings.

**Table 17.1** Monocular elevation deficiency—clinical findings and surgical recommendations

| Etiology | SR active force (saccades or active force generation) | Forced ductions for elevation | Bell's phenomenon | Primary position deviation | Recommended surgery |
|---|---|---|---|---|---|
| SR paresis | Decreased | Normal to mildly positive if some secondary IR contracture | Absent or decreased | HYPO | Knapp procedure<br>***Or***<br>Con SR recess 10 mm and Ips recess IR 5 mm only if HYPO in downgaze |
| Isolated IR restriction | Normal from downgaze to up | Markedly positive | Absent | Ortho or HYPO | Ips IR recess 5–10 mm |
| IR restriction secondary to SR paresis | Decreased | Slightly to moderately positive | Absent | HYPO | Ips IR recess 4–5 mm and partial Knapp procedure<br>***Or***<br>Ips IR recess 4–5 mm and con SR recess 10 mm |
| Supranuclear MED | Normal from downgaze to up | Normal | Present | Ortho or HYPO | Con SR recess 10 mm If ortho<br>***Or***<br>Ips IR recess 5 mm and con SR recess 10 mm if HYPO<br>***Or***<br>Ips Knapp procedure If HYPO[a] |

*SR* superior rectus muscle; *IR* inferior rectus muscle; *HYPO* hypotropic; *Con* contralateral, eye *without* MED; *Ips* ipsilateral; *Ortho* orthophoric; *MED* monocular elevation deficiency
[a]Greater chance of limiting downgaze with Knapp procedure

## Flap Tears

### Basic Information

*Overview*

Ludwig described this entity as occurring after trauma to the globe [12–14]. After blunt trauma, a rectus muscle, usually the IR, will undergo a traumatic separation of part of the orbital layer from the global layer. The avulsed flap of orbital layer muscle may get scarred into surrounding connective tissue. In most cases it presents clinically as a weak or underacting muscle, but in some cases it causes a restriction of movement in the opposite gaze field. Although Ludwig considers this to be a common occurrence, I have found it to be relatively uncommon. Very possibly I missed diagnosing this on occasion. However, my surgical approach for treating weak or restricted muscles after trauma has been sufficiently successful using standard procedures (weakening the antagonist or yoke muscles) that it may not be crucial to identify and treat most flap tears.

### Basic Information

*Diagnosis*

One needs to have a high index of suspicion that a flap tear has occurred, because the objective findings can be subtle. At surgery one will see either thinning or narrowing of the remaining global layer for part of the muscle, and the normal muscle capsule will not be evident. Although very-high-quality magnetic resonance imaging (MRI) may occasionally show a flap tear, it is usually not visible on orbital imaging.

### Basic Information

*Treatment*

If a flap tear is recognized, it makes sense to repair it. The flap should be carefully dissected free from the surrounding connective tissue and sutured back in its normal anatomic position, for which Ludwig advises the use of nonabsorbable sutures.

## Orbital Floor Fracture

### Basic Information

*Overview*

The classic strabismus presentation after orbital floor fracture is restrictive hypotropia due to the IR getting incarcerated in the fracture site. In many cases, there is both a limitation of elevation due to restriction and a limitation of depression simulating an IR weakness [15]. In the past this was thought to be due to damage to the nerve to the IR in the fracture site. I believe that in most cases this is caused by either a very posterior fracture altering the torque vector of the IR (Fig. 17.1) or a flap tear of the IR, as described above.

### Basic Information

*Management*

Often repair of the fracture corrects the strabismus, but sometimes strabismus persists. If the fracture were adequately repaired, strabismus surgery is needed. The most common scenario is a residual hypotropia, and what is needed is a recession of the restricted IR. The size of the recession should be dictated by intraoperative forced ductions and spring-back balance testing, rather than a set formula. If the recession needs to be more than 5 mm, there is often a lag of that eye on downgaze afterwards, resulting in a hypertropia. If this cannot be handled optically (e.g., slab-off prism as shown in Fig. 9.2, single-vision reading glasses) surgery on the contralateral IR in the form of recession or posterior fixation may be needed. If the amount of initial IR recession is very large, I think it makes sense to proactively anticipate a postoperative hypertropia, and do contralateral IR surgery at the time of the initial strabismus surgical procedure. See Chap. 21, "Complicated Strabismus," Case 21.3 on page 313 and Case 21.12 on page 319, for representative examples of these principles.

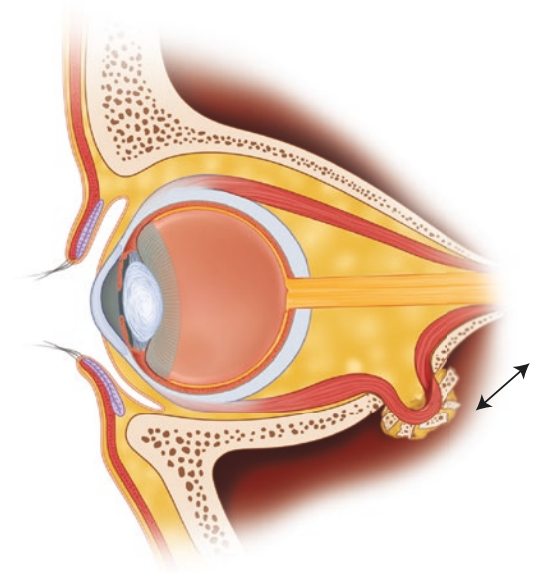

**Fig. 17.1** Posterior orbital floor fracture can not only restrict upgaze, but also alter the torque vector of the inferior rectus muscle, weakening its depressing action

### Question

My patient underwent repair of an orbital floor fracture 6 months ago. He has a hypot-

ropia and limitation of elevation of that eye. Imaging shows that the fracture is well repaired; however there are a lot of adhesions between the IR and the floor implant. Should I have oculoplastics revise the implant?


### Reply

In my experience, going back later is never successful in restoring motility. It is best to treat the strabismus as you would any other IR restriction case.

### Question

My patient underwent repair of an orbital floor fracture and now has the clinical picture of a restricted IR that is also paretic (or pseudo-paretic). There is a miniscule hypotropia in primary with −3 for elevation with a hypotropia in upgaze and −2 for depression with a hypertropia in downgaze. How should I manage him?


### Reply

I think you first need to determine if there is a flap tear causing the pseudo-paresis by surgical exploration. If so, the tear should be repaired and the tight IR recessed. If that does not correct the lag on downgaze, an enhancement involving posterior fixation of the contralateral IR would be the next step. If there is no flap tear, I have good success with recessing both the ipsilateral IR and SR [15]. However, I feel that I have improved on these published results by adding a posterior fixation procedure to the contralateral IR at the same time.

## Marcaine Myotoxicity

### Basic Information

*Overview*

After the index case of myotoxicity after periocular anesthetic was reported [5], strabismus surgeons began seeing this entity with rapidly increasing frequency [16–19]. Evidence suggests that inadvertent injection of anesthetic agents directly into an extraocular muscle (EOM) causes damage. Initially, there is a paresis followed by muscle fibrosis and hypertrophy. Marcaine is the agent most likely to cause this problem. With the growing trend away from retrobulbar or peribulbar anesthesia for cataract surgery, this entity is becoming less frequent.

### Basic Information

*Clinical Findings*

The clinical picture of EOM myotoxicity will have stabilized several weeks to months after the initial insult. There is a deviation in the direction of the affected muscle, e.g., a hypotropia if the IR is affected. Although the IR is most commonly affected, the SR can also be affected. Capo and Guyton showed how an injection given inferiorly can impale the SR near the apex of the orbit if the needle passes under and posterior to the globe [18] (Fig. 17.2). Interestingly, this condition sometimes presents as a restrictive strabismus in which the deviation increases in gaze opposite that of the affected muscle; for example, IR involvement causes a hypotropia that increases in upgaze. But in many cases there is the picture of a truly "overacting" muscle; for example, if the SR is involved there is a hypertropia that increases in upgaze. I have observed that cases in which the SR is involved are more likely to present as muscle overaction, and those involving the IR are

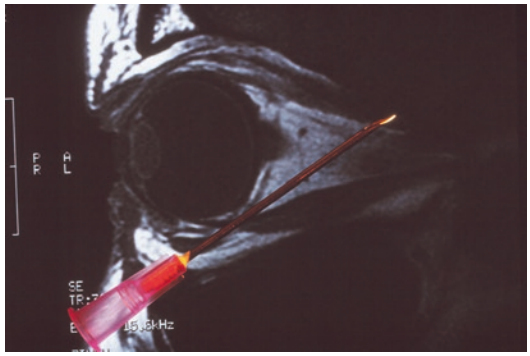

**Fig. 17.2** A 22-gauge needle superimposed on an orbital MRI. In order for the needle to clear the inferior orbital rim and the bottom of the globe, it can only impale the SR near the apex of the orbit, as has been shown by Capo and Guyton [18] (from Kushner [19], with permission)

more likely to present as restrictive strabismus [19]. I speculate, based on the observations of Capo and Guyton [18], that if the SR is involved the muscle involvement is more likely to be focal near the apex of the orbit, because the globe blocks a needle entering through the inferior eyelid from impaling the middle or anterior parts of the SR. Fibrosis limited to the posterior part of the SR puts the anterior part of the muscle on stretch, thus strengthening it. But if the IR is involved, a needle entering through the lower eyelid can impale the IR anywhere along its length. Consequently, the damage is more likely to be throughout the length of the muscle.

Regardless of whether this myotoxicity takes the form of muscle restriction or overaction, there is muscle hypertrophy, which can be seen on imaging studies, that is quite similar to the appearance of muscles in TED [5]. In both forms, forced ductions are abnormal; however they are moreso in the restrictive form.

## Basic Information

*Treatment*

In both forms of this condition—muscle restriction or overaction—the treatment is to recess the affected muscle. In cases that have muscle overaction, the results from recessing the affected muscle are usually excellent. Often motility is fully normal after surgery. In cases of restriction, results are similar to those found with TED. If a large recession is needed to free the restriction, the muscle often underacts after surgery with a lag in its field of action. Surgery may be needed on additional muscles to prevent or correct this.

# Strabismus After Scleral Buckling Surgery

## Basic Information

*Overview*

Transient strabismus and/or diplopia due to orbital or muscle edema, hemorrhage, or blurred vision is common after scleral buckling surgery and in most cases resolves. There is no documented incidence figure for strabismus and/or diplopia that persists after scleral buckling surgery, but it is probably between 1 and 10%. The majority of cases are restrictive due either to adhesions or to mass effect of the explant material. Less frequently there can be alterations of muscle path and torque vector if either of the oblique muscles is incarcerated or tucked by the buckle [7, 8]. This should always be suspected if there is a very large amount of torsion that increases in gaze opposite the deviation (e.g., hypertropia and incyclotropia that increase in downgaze, implying that the superior oblique muscle (SO) has been anteriorized or tucked). Permanent muscle damage from ischemia can also occur, leading to segmental atrophy. Finally, there can be a loss of fusion due to induced anisometropia, metamorphopsia, and macular issues. Large buckling elements, and in particular radial sponges or meridional elements placed under a muscle, substantially increase the risk of postoperative strabismus. See Chap. 21, Case 21.7 on page 315, for a representative example of these principles.

## Pearl

If you suspect a disruption of fusion in cases of diplopia after scleral buckling surgery, e.g., no fusion when the deviation is offset with prisms, always suspect torsion as the cause. Check for torsion even in the absence of a hypertropia [20]. See Chap. 2, "The Examination," for further discussion.

## Basic Information

*General Management Principles*

Certainly small deviations that are fairly comitant can be successfully managed with prisms. If the deviation is too large to manage with prisms, but is fairly comitant, strabismus surgery can be performed on the other eye. This has the advantage of being substantially easier, and does not run any risk of causing re-detachment of the retina. Unfortunately, most cases of strabismus after scleral buckle are quite incomitant and better treated with surgery on the affected eye. Also, many patients are reluctant to have surgery done on their "good" eye.

## Basic Information

*Overview of Surgical Management*

Proper surgical planning involves accurate forced duction and active force generation testing to determine the relative influence of restriction and paresis; in most cases it is primarily restriction. I always make a great effort to obtain an operative report from the retinal surgery, so I know exactly what explant material was used and where it was placed. I also like to know where the clip securing the encircling element was placed. If records are not available, some of the needed information can be determined by careful indirect ophthalmoscopy. With that in mind, I find that in most cases, strabismus can successfully be corrected without removing the buckle (see below); however, I always like to get the opinion of a retinal surgeon (ideally the one who placed the buckle) if it is safe to remove the buckle if found advantageous to the strabismus procedure.

## Question

I have a patient with a restrictive10 Δ LHT that increases in downgaze 1 year after a scleral buckling procedure in his left eye. He is diplopic. The retinal surgeon advises just removing the buckle. How likely is that to be successful?


## Reply

This is a very common question, because just removing the buckle is often the first line of treatment by retinal surgeons. However, in my experience this never corrects the strabismus sufficiently. If a deviation associated with limited rotations has been present for even a few months, there is always contracture of EOMs that has to be released.

## Basic Information

*Specifics of Surgical Management*

Strabismus surgery after scleral buckling surgery is one of the more technically challenging procedures the strabismologist will encounter. Decisions need to be made based on intraoperative findings, including repeated forced duction testing during surgery. The following principles may be helpful:

1. I generally prefer a limbus approach, as sometimes conjunctiva is shortened and needs to be recessed.

2. In some cases, the entire muscle that is over the buckle is completely atrophic, and it appears as though the muscle inserts at the posterior edge of the buckle. In others, there appears to be viable muscle over the buckle, but it is scarred to the buckle. Infrequently, the muscle overlying the buckle is viable, and there is a reasonably open plane between the muscle and buckle. In the first two situations, you will need to hook the muscle posterior to the buckle, and only in the last situation can you usually hook the muscle at the original insertion. Initially, you can try to hook in the usual manner, but if that is not possible, hook posteriorly.
3. If the entire muscle over the buckle is atrophic, treat the anterior edge of muscle just posterior to the buckle as though it was the muscle's insertion. Leave the atrophic tissue on the buckle. If there appears to be viable muscle scarred to the buckle, you have to make a decision as to whether that should be left as part of the muscle, and your sutures placed near the original muscle insertion, or if that tissue should be sacrificed as in the prior scenario. I like to start with placing sutures as though I will be sparing/utilizing that tissue, and then decide on whether to continue with this plan after disinserting it and assessing its integrity.
4. Once the decisions outlined above have been made, and the muscle sutured and disinserted, forced ductions need to be assessed with the muscle disinserted. If they are normal, there is usually no need to consider removing the buckle. If they are still restricted, dissection of adhesions should continue until forced ductions are normal. Keep in mind that adhesions can cause leash restrictions as well as reverse leash restrictions [21]. If an eye is restricted to downgaze, for example, the restriction may be inferior. In my practice, it is only in those cases in which I cannot free up forced ductions that I give consideration to removing part or all of the buckle.
5. If the spot to which you desire to recess a muscle is overlying the buckle, you need to make a decision about using nonabsorbable sutures. I use the following guidelines: If in the course of dissection you were able to leave intact the fibrous capsule that invariably surrounds the buckle, absorbable sutures will work satisfactorily; the muscle will bond to the capsule. This is true both with fixed sutures and with an adjustable suture (e.g., suspension or "hang-back"). But if the dissection necessitated opening the fibrous capsule and you are looking at bare silicone, the muscle will not form a secure bond. A nonabsorbable suture is needed.
6. I have found the use of adjustable sutures to be helpful in these cases. The need for a nonabsorbable suture in some cases is discussed above.

### Pearl

If you suspect a lot of scarring, and if you wish to avoid the difficult dissection of muscle from buckle, you can make the incision more posteriorly, hook the muscle posterior to all the buckling material and scarring, and proceed as though the muscle's insertion is at the posterior edge of the buckle. You basically sacrifice all muscle anterior to that point.

### Important Point

The muscle will not form a solid bond to bare silicone but will to the fibrous capsule that surrounds the buckle. If you are positioning a recessed muscle to the new insertion that will be overlying bare silicone, use a nonabsorbable suture.

### Question

I was taught that it is always safe to remove a buckle if it has been in place for about a year. Is this not the case? And if not, what about just dividing/cutting the buckle to relieve traction?


### Reply

In many cases a buckle can be removed after 1 year; however, this is not completely clear-cut, and you should always get the advice of a retina surgeon. In my experience, many younger retina surgeons do not have a lot of experience with scleral buckles, as they usually repair detachments with a vitrectomy approach. Consequently, their default position is to leave the buckle in place so as not to take a risk. So you should get the advice of a retinal surgeon experienced with scleral buckles. My experienced retina colleagues feel that, regardless of the length of time the buckle has been in place, it is the amount of proliferation, and traction at the vitreous base, combined with the thinness of the retina, that determines the risk of removing the buckle. In many cases, it may be safer to just cut the buckle and not remove it, as there will still be some buckling effect afterwards. The problem for the strabismus surgeon is that just cutting the buckle will usually do little to reduce any mass effect that is contributing to the restriction. I rarely find that useful. If you do have to remove a buckle, it is extremely helpful to know where the sleeve was placed. If you do know, you can cut the buckle next to the sleeve and pull on the sleeve, and the whole buckle will snake its way out. But if you cannot find and grasp the sleeve, you cannot remove it this way because the sleeve will prevent the buckle from just sliding out. In those cases in which you feel the buckle itself is contributing to the restriction, removing just a segment of the buckle may solve the problem; you may not need to remove the entire buckle. This is different than just cutting/dividing the buckle.

Even if a buckle has been in place for a long time, it may not be safe to remove. Consult a retinal surgeon experienced with scleral buckles.

## Strabismus After Aqueous Drainage Devices

### Basic Information

*Overview*

The incidence of strabismus after placement of an aqueous drainage device is not well defined. It is probably a function of the type of device used, its location, surgeon's skill, and the patient's underlying fusion potential, given that many of these patients have poor vision and peripheral visual field loss, which interferes with peripheral fusion.

### Basic Information

*Management*

In many ways the treatment of this disorder is similar to that of strabismus after scleral buckling surgery. This is because both conditions are usually restrictive in nature, often caused by the mass effect of the external hardware, may involve muscle weakness from ischemia or trauma, and are frequently associated with poor vision. A few principles can help guide management:

1. If there are either no significant limitation of rotations and hence no restriction, or if rotations are limited but forced duction and active force generation testing show paresis instead of restriction, resection of the muscle opposite the deviation (e.g., resect IR if a hypertropia is present), or yoke surgery on the other eye, is preferable to operating in the area of the drainage device. Surgery in that area runs a risk of compromising pressure control.
2. If rotations are limited, ask the glaucoma surgeon if she is willing to remove the drainage device and replace it with a smaller one in another area. Unlike removing a scleral

buckle, removal of an aqueous drainage device often corrects the strabismus. I believe that the reason for this is that the mass effect is often greater with drainage devices than with scleral buckles.

3. If the drainage device cannot be removed and there is restriction, you should do the surgery as far posterior as possible to avoid compromising the bleb. See Chap. 21, Case 21.16 on page 323, for a representative example of these principles.

## Strabismus After Sinus Surgery

### Basic Information

*Overview*

This can occur after endoscopic sinus surgery for polyps or after an ethmoidectomy; if the surgical instrument violates the lamina papyracea and enters the orbit, it can damage the MR and at times the orbital portion of the SO. This can be a devastating complication that can be difficult to repair, depending on the integrity of the remaining muscles.

### Basic Information

*Diagnosis*

The most typical presentation of muscle damage after sinus surgery is a very-large-angle XT with limited or no adduction. Active force generation shows decreased or absent MR function. There may also be significant elements of restriction both to abduction and adduction due to leash and/or reverse leash tethers. Dynamic orbital imaging is crucial for proper surgical planning. Imaging will usually show severe damage to the MR, which may be completely severed, and parts of which may be scarred into the iatrogenic defect in the medial orbital wall.

### Basic Information

*Treatment*

If imaging shows that there is an intact posterior aspect to the MR, and if dynamic imaging shows that it has contractility on attempted adduction, there is a good likelihood that you can get a satisfactory outcome with strabismus surgery (see Figs. 11.4 through 11.6). In this situation the best result can be obtained using the orbital wall approach to retrieve the posterior segment of the MR as described by Goldberg, Rosenbaum, and coauthors [22]. If imaging shows that there is no segment of the MR intact at the apex of the orbit, or if dynamic studies show that it is not contractile, the only remaining option is a nasal transposition of the vertical rectus muscles. This will give a less optimum outcome than if a contractile MR is salvaged and reattached. As an aside, I strongly recommend that you look at orbital images yourself and go over them in person with an experienced neuroradiologist. In my experience, many neuroradiologists have very limited formal training in evaluating the EOMs. It is by default that this particular subspecialty usually reads these scans. They are accustomed to reading images of the head and brain, and the orbits show up on those studies. The MRI shown in Fig. 11.7 was reported to show "normal EOMs" by an academic neuroradiologist at an excellent medical college. I have seen orbital scans read by board-certified neuroradiologists as having normal EOMs when a SO was almost absent. I recall another, also read as normal, in which the medial rectus muscle was essentially absent after a sinus surgical procedure resulted in serious violation of the orbit.

## Congenial Cranial Dysinnervation Syndrome (*Formerly Called* Congenital Fibrosis of the Extraocular Muscles)

### Basic Information

*Overview*

This is an autosomal dominant condition characterized by restrictive strabismus and ptosis [23, 24]. It most commonly affects the IR most severely, and consequently the eyes are fixed in downgaze with limited elevation. It has been shown to be associated with hypoplasia of the third cranial nerve [24]. Sporadic forms exist that are more likely to be unilateral, or involve only some of the EOMs.

### Basic Information

*Management*

As in all cases of restrictive strabismus, the key to success is to release the restriction. In this disorder it usually involves very large IR recessions. It is not uncommon to recess the IRs 8 mm or more in this condition. Even if doing so initially seems to give a good result, recurrence is common, as the recessed muscles take up slack and undergo further contracture, in part because their antagonists are weak.

### Question

I find doing such large recessions in these tight orbits technically difficult. I have now resorted to doing free tenotomies. But I did that recently and got essentially no effect. What happened?


### Reply

Because doing such large recessions in these tight orbits is technically difficult, many surgeons resort to either free tenotomy or recessions using a suspension (also known as hang-back) technique. I am not a fan of either in this setting. These muscles are very tight but also quite inelastic. They do not retract when disinserted. If a free tenotomy or suspension technique is used and the eye does immediately come into a more supraducted position, the muscle may stick down anteriorly and hardly be recessed at all. I recall operating on a child who had undergone four prior IR weakening procedures for unilateral IR fibrosis. The first three gave temporary improvement. The last consisted of bilateral free tenotomies of the IRs, and it provided no improvement. When I did the fifth operation, I found that after the free tenotomy the IR had bonded to the globe only 8 mm posterior to the limbus. If you are not comfortable placing fixed scleral sutures 8 or more mm posterior to the insertion in these patients, you can use a suspension (hang-back) technique, but also tie the eye into upgaze with traction sutures for about 1 week. That will keep the muscle from sticking down anteriorly. However, postoperative traction sutures are very uncomfortable for the patient. See Chap. 21, Case 21.4 on page 314 and Case 21.9 on page 317, for representative examples of these principles.

### Important Point

A free tenotomy of the IR may result in very little recession. If the eye stays in downgaze, the IR may bond to the sclera in an anterior position.

# Strabismus and Craniofacial Syndromes

## Basic Information

*Overview*

Craniofacial syndromes are a large and heterogeneous group of disorders that have a variety of ophthalmic manifestations. At one end of the spectrum is mild non-syndromic craniosynostosis, which can be subtle and have minimal eye findings. At the other end of the spectrum are the severely dysmorphic conditions like Crouzon and Apert syndromes, and others. Most commonly, strabismus associated with these syndromes include pattern strabismus (V-pattern far more frequent than "A"-pattern), often with severely abnormal ductions and versions.

## Basic Information

*Diagnosis*

The strabismus is diagnosed by the usual clinical means; however, the treatment plan should rely heavily on imaging studies of the EOMs. Common findings that contribute to strabismus include the following:

1. Abnormal positioning of the trochlea, either antero-placed or, more commonly, retro-placed: This can be congenital or the result of craniofacial surgery. Retro-placement of the trochlea can alter the torque vector or the SO and decrease its vertical force.
2. Absent or hypoplastic muscles, most frequently the SR: This can be a cause of apparent contralateral IO overaction due to fixation duress.

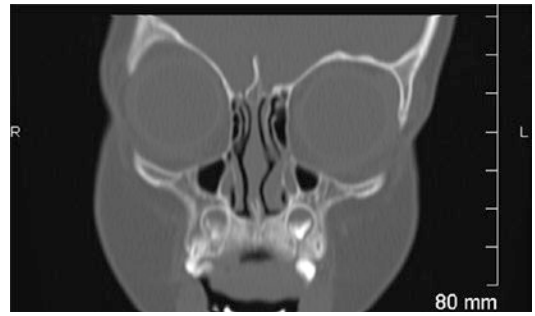


**Fig. 17.3** Harlequin-shaped R orbit in a child with craniosynostosis. The right superior oblique is also atrophic

3. Harlequin-shaped orbit (see Fig. 17.3).
4. Excyclorotation of all the EOMs: This will have the effect of causing a V-pattern, overelevation, and under-depression in adduction, as shown in Fig. 7.2.
5. Other abnormalities of the muscle paths, e.g., superior-lateral translation of the SR in unilateral craniosynostosis that can mimic ipsilateral IO OA [25].

## Basic Information

*Treatment*

Proper management is based on an understanding of the underlying mechanism of the strabismus in each individual patient. It is most important to realize that in most cases overelevation in adduction is not caused simply by an overacting IO. An approach to treating these patients that is based on the presentation by Dagi as part of the first Kushner Medal Webinar (Linda R. Dagi, MD, personal communication, 20 Jan 2016) is based on evaluating the following parameters:

1. Is fusion present? If not, correcting fundus torsion, or even choosing a surgical plan that may worsen it, may not be important.

2. If torticollis is present, is it ocular? A patch test is useful in sorting this out. Dagi said, "Some craniotomy patients may have an abnormal head posture that is no longer ocular in nature. The causes may be multiple and might include asymmetric weight and shape of the head" (Linda R. Dagi, MD, personal communication, 21 Mar 2016). Her point is that some patients may have started out with an ocular torticollis, lost fusion, yet retained the associated tilt. Nischal has indicated that many of these patients have an abnormally tilted union between the base of the skull and the first cervical vertebra. This results in a persistent non-ocular head tilt even after strabismus has been corrected (Ken K. Nischal, MD, personal communication, 29 May, 2016).
3. The severity, asymmetry, and nature of the orbital anomaly.
4. The amount of EOM excyclotropia as seen on imaging.
5. The degree of SO tendon laxity.
6. The amount of dysgenesis of any of the EOMs.
7. Is the V-pattern worse in upgaze or downgaze?
8. Is there migrated hardware from craniofacial surgery?

Treatment principles include the following:

1. Tucking the SOs if they are intact: Very lax tendons may be found and may need large tucks.
2. Excyclorotated EOMs can be repositioned to treat patterns, but doing so will worsen the torsion. It is OK to do this in non-fusing patients.
3. The surgical plan must account for muscle agenesis or hypoplasia.
4. Torticollis may be non-ocular.

See Chap. 21, Case 21.10 on page 318, for a representative example of these principles.

## Decompensated Esotropia Due to Early Presbyopia

### Basic Information

*Overview*

Guyton and coworkers observed that patients with a history of successfully treated esotropia have an increased incidence of recurrence as they approach presbyopia—specifically between 30 and 50 years of age [26]. Many esotropes have decreased accommodative amplitudes. This may have been the original cause of developing esotropia. Alternatively, it theoretically can be a result of wearing full hyperopic correction, and possibly a bifocal, as a child. Because accommodative convergence is a function of the effort to accommodate, these patients may be exerting more than the normal amount of accommodative effort to see clearly at near as they approach, but are not yet presbyopic. Further, in my experience many of these patients are being followed by comprehensive ophthalmologists or optometrists who often do not perform cycloplegic refractions in this age group, and often they have a lot of hyperopia that is not corrected by their spectacles.

### Basic Information

*Management*

The best management for this is prevention. I tell all my formerly esotropic patients in this age range to pay careful attention to the earliest sign of presbyopia, and to not defer bifocals until they are having major trouble reading (as many patients are prone to do). At least in theory, the early institution of a bifocal may prevent decompensation. Also, I always do both a manifest and

cycloplegic refraction and pay careful attention to the existence of any significant amount of latent hyperopia. If latent hyperopia is more than 0.50 D, I will try and push plus to get the patient to accept more hyperopic correction.

### Pearl

An adult with a history of treated esotropia should be put in a bifocal at the very earliest sign of presbyopia. Any significant latent hyperopia should be identified and treated optically; push plus as the patient tolerates doing so.

## Myasthenia Gravis

### Basic Information

*Overview*

Myasthenia is an autoimmune disorder that results in impaired transmission and reception of acetylcholine at the neuromuscular junction. It can result in a variable ptosis and/or weakness of any of the EOMs. In its generalized form it has systemic manifestations, but in the ocular form it may only clinically affect the EOMs and levator. The majority of patients with the generalized form present with ocular symptoms first. Conversely, the majority of patients presenting with just ocular findings will go on to develop generalized disease; however, if it is solely ocular for 3 years, systemic findings usually do not develop. Neonatal myasthenia occurs in about one in seven children of myasthenic mothers. It is transient and resolves within weeks. The more chronic form of myasthenia can present in children as well as adults. The juvenile form is more likely to go into remission than the adult form.

### Basic Information

*Diagnosis*

The diagnosis of myasthenia will often be suspected based on a history of a variable strabismus, which changes with respect to pattern, magnitude, and frequency that grow worse with fatigue. Often there will be variability during the course of the strabismus exam as the muscles fatigue. Ptosis is usually present, and the pupils are spared. The ptosis often worsens on prolonged upgaze as the levator fatigues. The presence of Cogan's lid twitch is a helpful diagnostic sign. The lid will overshoot as the eye goes from downgaze to primary, and then settles to its proper position. About 5–10% of patients with myasthenia have concurrent Graves' disease. Upper lid retraction is a hallmark of TED. If ever a patient with TED also has ptosis, myasthenia should be suspected. The ice test involves placing an ice pack over the eyes for 2 min and then reevaluating the patient [27]. It is probably more useful if ptosis is present, and the endpoint is decreased in the ptosis, than it is for a change in ocular motility. Antibody testing can be a helpful way to avoid the unpleasantness of pharmacologic testing, but antibody testing is only positive in less than 50% of myasthenics. I order acetylcholine receptor antibodies. If they are abnormal, the diagnosis is made. If they are normal, one still may be dealing with myasthenia. If myasthenia is suspected and cannot be diagnosed by the ice test or with immunologic testing, further testing is indicated. For years tensilon testing was the standard next step in evaluating a patient for myasthenia. Increasingly, colleagues are moving away from that because of rare but serious medical emergencies as a result of the test. They are moving to a trial of Mestinon (Valeant Pharmaceuticals, Bridgewate NJ, USA). Finally, single-fiber electromyography studies can be diagnostic in difficult cases. In a great many cases, the diagnosis can be made on clinical grounds

alone. These include history of reduced symptoms upon awakening, variability of ptosis and strabismus, and Cogan's sign. Hoyt reports that he finds that the orbicularis is almost always weak in myasthenics, so he also tests the strength of forced eyelid closure (Creig Hoyt, MD, personal communication, 6 Dec 2016).

### Pearl

If you ever see a patient with TED who has concurrent ptosis, you should suspect myasthenia.

### Important Point

Abnormal antibody testing can be diagnostic for myasthenia. But if antibodies are normal, you have not ruled out myasthenia.

### Basic Information

*Treatment*

The treatment of myasthenia is medical and carried out by a neurologist or neuro-ophthalmologist. Because of the variable nature of the disease, surgery is generally contraindicated. However, there are reports of satisfactory surgical intervention in patients who were stable for at least 12 months [28, 29]. I have, on rare occasion, operated on stable adults with myasthenia gravis with satisfactory results.

## Superior Oblique Myokymia

### Advanced Information

Superior oblique myokymia is an uncommon acquired eye movement disorder characterized by intermittent episodes of shimmering oscillopsia associated with vertical and torsional diplopia, due to microtremors of the SO [30]. It can be idiopathic, but in some cases is the sequelae of an acquired SO palsy. In some patients, episodes can be brought on by stress, caffeine, or nicotine. The oscillations can usually be seen best at the slit lamp and can often be brought on by stressing or relaxing the SO, e.g., tilting the head right or left, or infraducting or supraducting the eye in adduction. Patients typically find the symptoms very debilitating. In many patients, symptoms can be controlled medically, with Tegretol (Novartis, Hanover, NJ, USA), propranolol, or topical beta-blockers being most frequently used [30]. My experience with medical management of SO myokymia is limited, because most of the patients I care for are referred to me after medical management has failed. When medical treatment is unsuccessful, surgery should be considered. In order to eliminate the oscillopsia, the SO must be completely disabled from transmitting any force to the globe or to surrounding tissue that could indirectly transmit force to the globe. I recommend a large nasal tenectomy of the SO tendon, with removal of tendon from where it crosses the nasal edge of the SR to as close to the trochlea as is technically possible. This will effectively eliminate the oscillopsia, but will, of course, result in an ipsilateral SO palsy. Consequently, I simultaneously do a large ipsilateral IO myectomy, removing all of the IO temporal to the IR. In a series of 14 consecutive patients treated in this manner, I successfully eliminated symptoms of oscillopsia in all of them [30]. None of the patients had an iatrogenic postoperative hypertropia in the primary position, but 5 of the 14 (36%) had a lag of the operated eye in downgaze with diplopia. In all five patients, the diplopia was successfully treated with either optical management (prisms or single-vision reading glasses) or an enhancement to weaken the contralateral IR. Because the aforementioned surgical procedure results in iatrogenic ipsilateral SO and IO palsies, it is not surprising

that some patients ended up with incomitant strabismus. What is surprising is that the majority of patients did not. With an ipsilateral IO and SO palsy, one would expect there to be a hypertropia of the affected eye on ipsilateral head tilt, and a hypotropia on contralateral head tilt. Interestingly, many of these patients were orthophoric after surgery in all fields of gaze, including on head tilt to the right and left. I have speculated that this observation could possibly be explained by the complex interaction between the compensatory and anti-compensatory torsional movements that normally occur with forced head tilt, but my speculations are yet to be confirmed [31]. I find it interesting that, according to my patients, the debilitating effect of oscillopsia far exceeds that of diplopia, and those that developed diplopia in downgaze felt it was a reasonable trade-off for being free of oscillopsia. One patient said to me, "Double vision is just annoying. The shimmering drove me nuts."

I have seen patients who previously had undergone unsuccessful surgery for SO myokymia when the weakening of the SO was not sufficiently ablative. The use of a silicone spacer, a chicken suture, or doing a split tendon lengthening will not eliminate the force that the twitching SO transmits to the globe. Similarly, surgery to just weaken the anterior (torsional) fibers of the SO will not eliminate the vertical oscillopsia and diplopia that come from the posterior fibers.

### Important Point

When treating SO myokymia, surgery must eliminate all force from the SO being transmitted to the globe. A large tenectomy is needed.

Although good results have been reported with surgery, the surgical procedure I recommend is irreversible. Moreover, although I did not experience any adserse complications, the potential for them exists. As such, surgery should be undertaken only as a last resort after the failure of medical management.

### Important Point

Surgery to treat SO myokymia is irreversible and, in theory, carries a moderate risk of complications. As such, it should only be undertaken after an unsuccessful trial of medical therapy.

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# Nystagmus

# 18

*Since the mathematicians have invaded the theory of relativity (and tried to classify it), I do not understand it myself anymore.*

*—Albert Einstein.*

## Basic Information

*Overview*

Nystagmus has been defined as involuntary to-and-fro rhythmic and repeated eye movements. Nystagmus can be congenital due to either a sensory vision deficit or a congenital motor abnormality. One large study found a sensory cause in 91% of patients with congenital nystagmus, almost half of whom had albinism [1]. It also can be acquired due to a host of medical and neurologic disorders. This chapter addresses only the types of nystagmus of interest to the strabismologist.

## Basic Information

*How Nystagmus Is Characterized or Described*

The movements can be pendular or jerk, and vertical, horizontal, or rotary (torsional). If pendular, the two phases (right and left, or up and down) have the same velocity. If jerk, one phase is fast (saccadic speed) and the other slow. The same can be said of rotary nystagmus, which may have a fast and slow phase, or have both phases of equal velocity. The nystagmoid movement can be either manifest or latent, the latter being present when fusion is disrupted either with monocular occlusion or due to a manifest tropia. The slow phase is the abnormal component, which takes the fovea off the object of regard, and the fast component is corrective, bringing the fovea back to the object of regard. By convention, nystagmus is described by the direction of the fast phase, e.g., right beating and left beating. Visual acuity function is best when the fovea is on or very near the object of regard, and foveation time is defined as the interval where the eyes' velocities are less than 5°/s [2]. So a wave form in which the eye slowly drifts off the fovea will have longer foveation time than one in which the movement off the fovea is fast. The wave form of typical idiopathic infantile nystagmus is a jerk nystagmus that maximizes foveation time by having a slow phase with increasing velocity, e.g., starts out slow near foveation and then increases. This is one reason why this condition is compatible with good vision. This is in contrast to the wave form of latent nystagmus, which has a decreasing velocity (Fig. 18.1). Just after foveation it rapidly takes the fovea off the object of regard and then slows as it gets farther away from the object. Hence it is slowest just before the corrective saccade, which is very inefficient for vision. That is why the uncovering of latent nystagmus with occlusion degrades vision so severely. When this typical wave pattern of latent nystagmus occurs in the manifest state, it has been referred to by the term *manifest latent nystagmus*. The term is an oxymoron. According to Souza-Dias, it came into use by misquoting Lang's original term of *manifested latent nystagmus*, which is more appropriately descriptive. I prefer a term I heard from Souza-Dias, *manifest nystagmus of the latent*


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_18

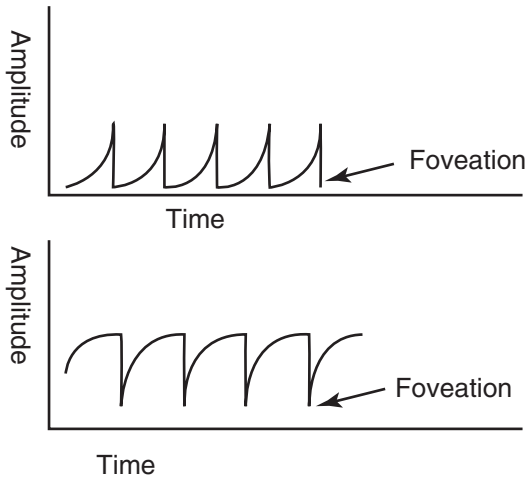


**Fig. 18.1** (**a**) Tracing of jerk nystagmus with increasing slow-phase velocity. The velocity is slowest just after foveation, which maximizes vision. (**b**) Tracing of jerk nystagmus with decreasing slow-phase velocity. The velocity is fastest just after foveation and slowest before the re-fixation saccade, making for inefficient vision

*type* (Carlos R. Souza-Dias, MD, personal communication, 17 Apr 2017).

Infantile nystagmus will have a direction of gaze in which the amplitude and velocity of the nystagmus decrease or disappear, which is called the null zone. If nystagmus is jerk, it will reverse directions on the other side of the null zone. If the gaze is not in the null zone, the phase takes the fovea toward the null zone, and the fast phase away from it. Thus it is *beating* away from the null zone.

## Basic Information

*Clinical Course*

Idiopathic infantile nystagmus usually does not appear until 2–3 months of age, and consequently it is more accurately called *infantile* than *congenital*. At first there may be wide-searching horizontal eye movements with little in the way of purposeful horizontal pursuits or saccades, and the child may be thought to be blind. However, vertical pursuit and saccadic movements are intact, which can be demonstrated with an optokinetic drum.

## Important Point

Many infants who develop idiopathic infantile nystagmus are thought to be blind in the first few months of life due to the absence of horizontal pursuit and saccadic movements before the nystagmus becomes evident.

## Pearl

The presence of vertical optokinetic nystagmus responses superimposed on the searching horizontal movements in a child beginning to manifest infantile nystagmus is an important prognostic sign with respect to vision.

According to Reinecke, at about 6–8 months a primarily pendular nystagmus appears that evolves into a jerk nystagmus with a null zone at 18–24 months of age [2]. At about this time a compensatory head posture may develop to allow the child to use their null zone. Patients with idiopathic infantile nystagmus may also develop head shaking. These head oscillations are not compensatory for the nystagmus; for example, they are not of equal but opposite velocity and direction to stabilize the image of regard on the fovea. Their purpose is not fully understood. Perhaps they are purposeless and just part of the pathology.

For unknown reasons, visual acuity develops slowly in children with idiopathic infantile nystagmus. Reinecke stated that it is not uncommon for a child to be 20/200 at 6 years of age and improve to 20/40 by 8 years of age [2]. I have not seen such dramatic changes in vision after age 6, but I do agree that vision development is delayed in these children.

## Basic Information

*Examination*

In addition to the usual components of a complete eye exam, there are several

items that need specific added attention in patients with nystagmus.

1. *Pupils:* Check for paradoxical pupil constriction to darkness. Normally, when the room lights are abruptly dimmed, the pupils dilate. The paradoxical response consists of an initial constriction of the pupils when the lights are turned off, followed by a slow dilation. This abnormality, which I initially observed in a family with congenital stationary night blindness [3], has subsequently been seen in patients with achromatopsia, optic nerve disease, and a variety of retinal dystrophies [4]. Its presence rules out idiopathic nystagmus. At times it can be hard to test for this in young children. I use a muscle light on a dim setting to provide the minimum necessary illumination to see the pupils as I abruptly turn off the room lights. You must be sure that the child continues to fixate in the distance. There is a tendency for the child to immediately look at the muscle light when the room lights are turned off, and the near reflex may result in miosis. Alternatively, you can have a dim desk lamp or the light on the slit lamp/phoropter pole to provide background lighting when the overhead lights are turned off. Generally sluggish pupils suggest retinal or optic nerve disease.

**Pearl**

The presence of paradoxical pupillary constriction to darkness in a patient with nystagmus is a strong indication that there is a sensory deficit.

2. *Visual acuity:* Monocular occlusion may bring out a latent component to the nystagmus that can seriously degrade vision. The ideal way to check vision in each eye is with some type of vectograph target that, when viewed with polarized glasses, allows each eye to see different letters. This, however, is not readily available in most clinics. The next best way to test vision is to fog one eye with a +3.50 D to +5 D lens. Reinecke has pointed out that using a greater amount of plus power for fogging may bring out the latent component much like occlusion [2]. You also need to assess visual acuity under binocular conditions and compare that to the acuity in each eye separately. You also should check near vision binocularly, which is often better than distance vision.
3. *Head position:* An abnormal head position is often the main indication for surgical intervention in patients with nystagmus. It tends to be maximum with visual effort, so you should assess it while the patient is reading small optotypes at distance. Ideally, you should assess it in casual settings. Increasingly, I ask parents to make a video of their child while he or she is watching television, with the camera mounted just above the screen. When assessing a head position, you should separately note three components: face turn right or left, chin up or down, and tilt right or left. The three components of a head posture should be quantified in degrees as best you can. I like to use either a goniometer or the cervical range-of-motion device [5] for quantifying head postures.

**Pearl**

It is useful to have parents make a video of their child's head posture while he or she is watching television.

**Important Point**

The three components of a head position include face turn, chin up or down, and tilt.

4. Assess the pattern of the nystagmus. Determine if it is vertical, horizontal, or torsional. If it remains horizontal in all fields of gaze, it is probably idiopathic infantile nystagmus or a

cone dystrophy. If jerk, determine if it is right beating or left beating, and if the directionality changes with right or left eye fixing. If it does, it is probably a manifested latent nystagmus. Be sure to look at the pattern multiple times over several minutes to be sure that you are not dealing with periodic alternating nystagmus.

5. Check carefully for a sensory cause. Most commonly there is a form of albinism. Look at the slit lamp with coaxial illumination for iris transillumination. Look carefully for retinal or optic nerve disorders. If you suspect a retinal problem, an electroretinogram is indicated.
6. If the patient is fusing, check fusional convergence amplitudes to see if he or she is candidate for artificial divergence surgery (see below).

## Basic Information

*Treatment of Amblyopia in Children with Nystagmus*

Amblyopia often develops in children with nystagmus, in part because they tend to have significant refractive errors. The principles for treating amblyopia are essentially the same in children with nystagmus as those without nystagmus with one caveat. Occlusion may bring out a latent component that may further degrade vision. If that happens, you may wish to rely more on pharmacologic penalization than occlusion.

## Basic Information

*Timing of Surgery to Treat a Head Posture*

Reinecke observed that significant changes occur as nystagmus develops in children up to 24 months of age [2], and I have observed not only changes in the amplitude of nystagmus, but also a change in directionality and location of the null zone up to 3 years of age. So it is my preference to defer surgery until about age 3 and a half or 4.

## Basic Information

*Surgical Treatment to Shift the Null Zone*

The most common surgical procedures for treating a face turn, or chin elevation or depression, are based on the principle of Kestenbaum and Anderson to shift the null zone [6, 7]. Put in reductionist terms, if a patient has a null zone 35 prism diopters (Δ) to the right, if you do strabismus surgery to shift both eyes 35 Δ to the left, they will then be centered when the patient innervates them for right gaze, thus centering the null zone. The Kestenbaum procedure does this via a recess/resect procedure in each eye, and the Anderson procedure does it by recession only. In both cases you are doing esotropia (ET) surgery in one eye and exotropia (XT) surgery in the other. It was learned early on that one had to do more surgery than one would normally do to shift the eyes the predicted amount. Initially, Parks recommended doing what he then, in 1973, considered to be the maximum safe amounts of surgery which were 5 mm for the medial rectus muscle (MR) recessions, 6 mm for the MR resection, 7 mm for the lateral rectus muscle (LR) recession, and 8 mm for the LR resection (his "5–6–7–8" rule) [8]. Because this resulted in many undercorrections, other authorities have augmented this formula by either arbitrarily adding 1 mm to each muscle's dose or incrementally adding 20, 40, or 60% to the 5–6–7–8 formula, depending on the size of the face turn. In actuality, one needs to create a gaze limitation in the opposite direction for this to be effective. It is interesting that even years after a Kestenbaum procedure, the patient's eyes will be markedly deviated to the side they were surgically rotated if you view them when the patient is asleep, by raising their lids. My preferred dosage is in Table 18.1 for correction of a horizontal face turn and in Table 18.2 for correction of a chin-up or down-head posture. Note that for a chin-down head posture I recommend anterior transposition of the inferior oblique muscles (IOs) instead of resecting the inferior rectus muscles (IRs) as described by Roberts et al. [9].

**Table 18.1** Recommended surgery for horizontal compensatory head posture for nystagmus

| Muscle | Up to 20° face turn 20 % augmentation[a] (mm) | 21–40° face turn 40 % augmentation[a] (mm) | >40° face turn 60 % augmentation[a] (mm) |
|---|---|---|---|
| *Right face turn (move eyes right)* | | | |
| RMR recession | 6.0 | 7.0 | 8.0 |
| RLR resection | 9.5 | 11.25 | 12.5 |
| LMR resection | 7.25 | 8.5 | 9.5 |
| LLR recession | 8.5 | 9.75 | 11.25 |
| *Left face turn (move eyes left)* | | | |
| LMR recession | 6.0 | 7.0 | 8.0 |
| LLR resection | 9.5 | 11.25 | 12.5 |
| RMR resection | 7.25 | 8.5 | 9.5 |
| RLR recession | 8.5 | 9.75 | 11.25 |

*RMR* right medial rectus muscle, *RLR* right lateral rectus muscle, *LMR* left medial rectus muscle, *LLR* left lateral rectus muscle

[a]Augmentation above the 5–6–7–8 rule recommendation of Parks [8]

**Table 18.2** Recommended surgery for vertical compensatory head posture for nystagmus

| | Up to 30° Chin up (mm) | >30° Chin up (mm) |
|---|---|---|
| Chin up (move eyes *up*) muscle | | |
| IR recession OU | 5.0 | 8.0 |
| SR resection OU | 5.0 | 8.0 |
| Chin down (move eyes *down*) | | |
| SR recession OU | 5 | 8 |
| IO AT OU | Level with IR insertion | Level with IR insertion |

*IR* inferior rectus muscle, *OU* both eyes, *SR* superior rectus muscle, *IO* inferior oblique muscle, *AT* anterior transposition

## Important Point

The large amounts of surgery on the vertical rectus muscles needed to correct large degrees of head posture will often create an A- or V-pattern. Large IR recession and superior rectus muscle (SR) resections will cause an A-pattern and conversely large SR recessions may cause a V-pattern, moreso if the IRs are resected than if the IOs are anteriorly transposed. If the patient does not have fusion, these adverse effects can be minimized by horizontally transposing the muscles in the appropriate direction. However, in fusing patients, the adverse effect this will have on torsion may cause symptoms.

## Question

I operated on a girl with a face turn left secondary to nystagmus using a 40% augmented Kestenbaum approach. Her null zone was in right gaze. After surgery she reversed her face turn and now has a null zone in left gaze. Did the surgery unmask a second null zone? What should I do now?


## Reply

Most patients in whom this occurs have periodic alternating nystagmus (PAN) that was not recognized before surgery. However, I have definitely seen this in patients for whom I carefully studied pre-op videos, and they did not have PAN. I assume that they had two null zones in opposite gazes, and one was so far in the periphery that they never used it. Surgery unmasked this. I treat them by recessing the previously resected muscles, converting them to a four-muscle recession

status. The several I treated thus had good outcomes with significant decrease in any abnormal head posture.

## Advanced Information

*Surgical Treatment for a Head Tilt*

In some ways the principles for correcting a head tilt are similar to those for a face turn, or chin elevation or depression, and in some ways they are quite different. The similarity is that in all cases the eyes are shifted in the direction of the head position. Thus for a face right they are surgically moved to the right, for a chin up they are surgically moved up, and for a tilt right they are rotated in the direction of the tilt—right eye excyclorotated and left eye incyclorotated. However, the underlying principle is totally different. In the case of a face turn right, the eyes are shifted so the fovea is now pointing far to the right of the object of regard, and there has to be subsequent left gaze innervation to bring the fovea back to the object. In the case of surgery to correct a tilt, rotations are around the fovea so the object of regard is never off the fovea. Something very different must be happening. Consider a patient who fixes with his left eye and has a 30° right head tilt. That person "likes" having the perceived vertical meridian as running from approximately 1 o'clock retina and 7 o'clock retina. Surgery to incyclorotate that eye will put 1 o'clock retina at the 12 o'clock position, and 7 o'clock retina at the 6 o'clock position. Afterwards, if the patient orients vertical lines on the meridian between what was originally 1 and 7 o'clock retina, the head will be straight (Fig. 18.2).

One thing that can make torsional nystagmus surgery tricky is that many surgical procedures to tort the eyes will also create a vertical deviation. If you wish to excyclorotate a right eye and incyclorotate a left eye, you might consider weakening the right (R) SO and tucking the left (L) SO, if you do not think this through carefully. Although this would have the desired torsional effect, it would cause an R hypertropia and/or an L hypotropia, thus inducing a large vertical deviation. Procedures must be chosen that tort the eyes without having significant vertical action. Depending on the amount of correction needed and what other muscles may need to be done for a turn or chin position, the following options can be considered in those patients in whom you do not want any vertical shift:

1. To incyclorotate an eye:
   (a) Harada-Ito procedure
   (b) Temporal transposition of SR 7 mm and/or nasal transposition of IR 7 mm
   (c) Disinsertion of nasal three-fourths to seven-eighths of the SR and/or disinsertion of the temporal three-fourths to seven-eighths of the IR
   (d) Anterior seven-eighths disinsertion of IO (see Fig. 13.8)

2. To excyclorotate an eye:
   (a) Anterior seven-eighths tenectomy SO
   (b) Nasal transposition of SR 7 mm and/or temporal transposition of IR 7 mm
   (c) Disinsertion of the temporal three-fourths to seven-eighths of the SR and/or disinsertion of the nasal three-fourths to seven-eighths of the IR

## Important Point

It is crucial to understand that only surgery done in the fixing eye will address a head posture. Then surgery should be done in the non-fixing eye to match what was done in the fixing eye, and/or adjust for any strabismus that may be present.

## Advanced Information

*Treatment of a Multiplanar Head Posture*

It is not uncommon to see patients with a null zone that results in a face turn, a chin up or down, and a tilt. Surgical planning can be difficult in these patients because one

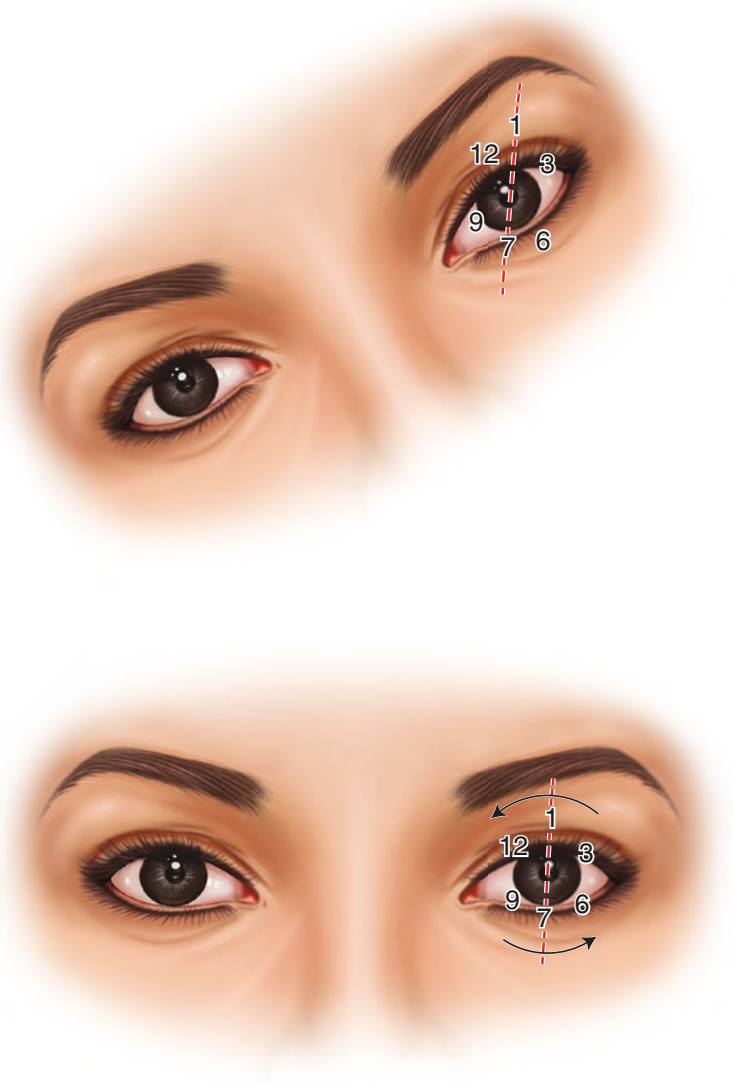


**Fig. 18.2** (**a**) Patient with head tilt right who fixes OS vertical lines will orient between 1 and 7 o'clock retina in the left eye. (**b**) After surgery to intort the fixing left eye, the prior 1 and 7 o'clock retinal meridians are now oriented vertically. If the patient "likes" to orient vertical lines between those two meridians, the head will be straight

may be limited as to how many muscles can be operated on in one sitting due to concern about anterior segment ischemia. Some principles are the following:

1. Identify which component is largest and make a plan for treating it. Most often it is the face turn, but not always.
2. See if you can correct the second most important component with muscle transpositions. For example, if the biggest component was a face turn and you are planning a horizontal Kestenbaum, treat the chin up or down by transposing the horizontal rectus muscle.
3. Consider doing the Anderson approach of recessions only, thus sparing the muscles that would be resected with a Kestenbaum procedure.
4. If the tilt is the least significant, you might ignore it initially. I find sometimes a tilt decreases if the turn and chin up/down element are corrected.
5. When possible, use oblique muscles for the tilt to spare rectus muscles.

## Question

I have a 6-year-old patient with optic nerve hypoplasia OD and acuity of 20/200 OD. Her left eye is normal, sees 20/30, but has nystagmus. She has a face turn left of 30° and a right head tilt of 30°. There is a variable right ET of about 20 Δ. How should I manage her?


## Reply

Upon my recommendation, this patient underwent a 7 mm LMR recession and 9 mm LLR resection, to address the turn. At the same time the LMR was raised 1 tendon width and the LLR lowered three-fourths of a tendon width to give some incyclorotation for the tilt; the lateral was lowered more than the medial was raised because it was resected and hence tighter. In addition, a left Harada-Ito procedure was done and an anterior seven-eighths disinsertion of the LIO. After surgery, all components of the head posture were corrected.

## Question

I have a patient with idiopathic infantile nystagmus who fixes with her left eye in adduction and has a 30° left face turn. She also has a 30 Δ RET. I plan to do a 40% augmentation Kestenbaum in her left eye. What should I do in the right eye?


## Reply

In this situation, the amount of surgery you do OS will be much more than what one would do to correct a 30 Δ ET. You can expect that procedure alone will make the right eye exotropic a modest amount. I would plan to do a medium-sized recession and resection for XT in the right eye. It can be very hard to predict exactly how much surgery to do in these situations. You take a best guess, with plans to do an enhancement if you do not estimate it correctly. Or you cannot be faulted for planning it as a to-stage correction. I usually opt for the former approach. By doing so, I have a reasonable chance of getting the job done with one operation, and with the other approach you are committing to two procedures. There are numerous scenarios that are similar to this one which you may encounter:

1. Needing to do ET surgery in the fixing eye when the fellow eye is already exotropic. A large amount of surgery will be needed for the XT.
2. Needing to do XT surgery in the fixing eye (rare situation where the patient fixes in abduction) and the fellow eye is esotropic. A large amount of surgery will be needed for the ET.
3. Correction of an iatrogenic XT in the nondominant eye after a Kestenbaum procedure in a formerly orthophoric patient. This nondominant eye had undergone XT surgery as part of the Kestenbaum procedure and it was not enough to match the fellow eye. Further LR recession and/or MR resection is needed.
4. Correction of an iatrogenic ET in the nondominant eye after a Kestenbaum procedure in a formerly orthophoric patient. This nondominant eye had undergone ET surgery as part of the Kestenbaum procedure and it was too much to match the fellow eye. Partial reversal of the prior LR recession and/or MR resection is needed.

## Important Point

When I edited the Grand Rounds section of *Binocular Vision & Strabismus Quarterly*, I had cases with each of these aforementioned scenarios, each of which was discussed by a group of about five experts. It was shocking to me how disparate the

recommendations of these experts were. I recall one case which was similar to the third scenario above, for which one expert recommended a 1–1.5 mm re-recession of the previously recessed muscle, and another recommended 5 mm additional recession. The bottom line is that there is no solid formula to refer to in these situations, and experience is invaluable. When in doubt, plan to do it in two stages.

## Advanced Information

*Four Muscle Retroequatorial Recession*

This procedure was proposed by Bietti in 1957 [10]. It involves recessing all four horizontal rectus muscles posterior to the equator, presumably to decrease their force. I have had limited and mixed success with this procedure. I do not use it if there is a consistent abnormal head posture, but do it in some patients with nystagmus associated with albinism or idiopathic infantile nystagmus. I typically recess the MRs 10 mm and the LRs 12 mm from their insertions. I have some patients in whom this appeared to decrease the amplitude of the nystagmus and some who felt the "efficiency" of their vision improved, despite no objective improvement in Snellen acuity; I realize this is subjective and cannot be quantified. I have found this procedure useful for patients with alternating head turns due to periodic alternating nystagmus. Surprisingly, ductions are only mildly reduced after this procedure.

## Advanced Information

*Artificial Divergence Surgery*

This surgery is limited to fusing patients with good fusional convergence amplitudes. It was proposed by Cüppers in 1971 [11, 12]. The rationale is that, because nystagmus damps with convergence, bilateral medial rectus recessions in a fusing patient will create an XT. If the patient has good convergence amplitudes, they will overcome the XT with fusional convergence and damp the nystagmus. In a large published series of this technique, Spielmann advocated recessions of both MRs of up to 12 mm [12]. I have had success with this procedure in a small number of patients, but I usually limit my recession to 5–7 mm. Using base-out prisms can be useful in determining who might respond well to this surgery, and in some cases their use can be effective long term as an alternative to surgery. This is described in more detail later in this chapter.

## Advanced Information

*Posterior Fixation*

Various authors have added posterior fixation sutures to the recessions when doing Kestenbaum–Anderson type of surgery. I do not think this is logical. Posterior fixation sutures are effective for decreasing a duction in eccentric gaze starting about 30° into the field of action of a muscle. They would not be expected to contribute to the alignment in primary gaze.

## Advanced Information

*Tenotomy and Reinsertion*

Based on some initial work on an achiasmic dog with nystagmus, Dell'Osso and Hertle have recommended simple tenotomy at the insertion and reattachment of the horizontal rectus muscles as a means of damping nystagmus [13]. There is controversy as to the efficacy of this procedure; a proposed mechanism of action is unclear, and I have no personal experience with it. It is thought that this procedure might interrupt the proprioceptive feedback loop from the palisade endings in the anterior tendon, which in turn damps the nystagmus. It is unclear why this should be more

effective than the four-muscle retroequatorial recession, which I find to have limited benefit. Furthermore, if the goal is to interrupt the proprioceptive feedback loop, it would make more sense to me to resect the anterior 3–5 mm of the tendon, thereby eliminating the palisade endings, and then recessing an amount exactly equal to the resection, thus not altering the sarcomere length in the muscle itself.

## Advanced Information

*Manifested Latent Nystagmus*

As stated earlier, when the wave pattern for latent nystagmus becomes manifested, vision is seriously degraded. Having a manifest strabismus will make nystagmus manifest in a patient who otherwise would have latent nystagmus. It is crucial to recognize this, because in such a patient correcting the strabismus can have a dramatic effect on reducing nystagmus (it will become latent) and improving acuity [14]. The most important diagnostic sign is that the nystagmus will change direction as either eye is occluded (right beating to left beating and vice versa). See Chap. 21, "Complicated Strabismus," Case 21.22 on page 328, for a representative example of these principles.

## Pearl

Correcting manifest strabismus in a patient with manifested latent nystagmus will convert the nystagmus to latent and improve visual acuity.

## Advanced Information

*Nystagmus Compensation Syndrome*

This condition (initially called the *nystagmus blockage syndrome*) is characterized by a variable and large-angle infantile ET in which convergence is used to damp nystagmus [11]. There is an abduction deficit with a manifest nystagmus that increases on attempted abduction. The ET is greater in side gaze as the non-fixing eye converges far into adduction to damp the nystagmus. In my experience, this is frequently over-diagnosed, and the label applied to all infantile esotropes with nystagmus or infants with the *Ciancia syndrome.* I think the nystagmus compensation syndrome is a relatively rare condition, but, nevertheless, a real one. Initially, Cüppers recommended MR recession with posterior fixation [11]; however, most experts feel that results are a good with just large MR recessions; with that I agree.

## Advanced Information

*Nystagmus with Uniocular Blindness*

There is a syndrome in which children born with one amaurotic eye, or who develop unilateral blindness or undergo enucleation, have a manifested latent nystagmus in their seeing eye, fixate in adduction, and have a face turn toward the seeing eye [15]. Kestenbaum-type surgery in the seeing eye is effective in treating the head posture.

## Advanced Information

*Periodic Alternating Nystagmus*

This is characterized by a conjugated horizontal jerk nystagmus that manifests a change in null zone and the directionality of the nystagmus according to some regular cycle, typically several minutes. The patient may note an alternating face turn. Although it is most commonly acquired secondary to a variety of neurologic disorders, it can occur in infants and has a high prevalence in patients with albinism. I find the four-muscle retroequatorial recession procedure the most effective in treating the alternating head turns.

## Basic Information

*The Use of Prisms to Treat Nystagmus*

"Yoked prisms" is the term to describe orienting prisms with the base in the same direction over each eye, e.g., base left OU or base right OU. In theory the prisms can simulate the effect of a Kestenbaum–Anderson procedure. I have found very little use for yoked prisms in managing nystagmus patients long term. If you consider that 1° is 2 Δ, a 30 Δ prism will only shift the image about 15°. A 30 Δ is rather large—too large to grind in glasses, and a Fresnel prism of that amount induces a lot of blur. Yet, a 15° head turn is about the smallest one would treat. On the other hand, prisms can be used to predict who may benefit from the artificial divergence procedure. An approach I learned from Kowal is to incorporate 6–7 Δ of base-out prism in each spectacle lens (ground in) combined with 1 D of over-minus (Lionel Kowal, MD, personal communication, 20 Nov 2016). Given a normal accommodative convergence/accommodation (AC/A) ratio, the addition of the 1 D of over-minus correction should add an additional 4–5 Δ of accommodative convergence, resulting in a total of about 16–19 Δ of convergence. By having some of this come from accommodative convergence, there is not as much strain on the fusional convergence system. If the patient can wear these spectacles comfortably, and if they correct the abnormal head posture, the patient should do well with the artificial divergence procedure. Interestingly, Kowal reports that some patients do well with these spectacles long term as an alternative to surgery. I do not have experience with their long-term use.

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# Paralytic Strabismus: Third and Sixth Cranial Nerves

19

This chapter addresses the surgical treatment of third and sixth cranial nerve palsy. The treatment of fourth cranial nerve palsy is discussed in Chap. 8, "Vertical Deviations."

## Basic Information

*General Principles*

When dealing with a limitation of rotation, it is crucial to distinguish if the problem is due to restriction, paresis, or a combination of both. Thus not only are forced ductions necessary, but you must also assess the strength of the muscle in question. Active force generation testing is the most practical way to do this in cooperative older patients. For younger or uncooperative patients, you can gain a lot of relevant information regarding rectus muscle strength by visually assessing the speed of saccades as the patient rapidly shifts gaze, or by observing the saccadic phase of optokinetic nystagmus. This is not useful for superior oblique muscle (SO) palsy, because intact vertical rectus muscles are sufficient for generating saccades [1]. Quantitative saccadic velocity testing is probably the most accurate test for assessing rectus muscle contractile force. However, I recognize that most clinicians do not have ready access to that test. I feel in most cases you can make good surgical judgements without saccadic velocity testing.

## Important Point

The presence of positive forced ductions does not rule out the coexistence of paresis. Positive forced ductions could be the result of secondary contracture of a muscle that was shortened because the antagonist was paretic.

The following general principles can guide you in treating paretic strabismus:

1. If there is a restriction as determined by forced duction testing, it must be relieved.
2. If there is −4 to rotation in any direction, and forced ductions are essentially normal, the limitation is due to paresis. A transposition procedure is indicated.
3. If a limited duction is due to a combination of restriction and paresis, a recession of the restricted muscle and resection of the paretic one, or a recession of the restricted muscle and recession of the yoke of the paretic muscle, is indicated.
4. Completely dead muscles cannot be "strengthened" by resections, but paretic muscles can be effectively resected.


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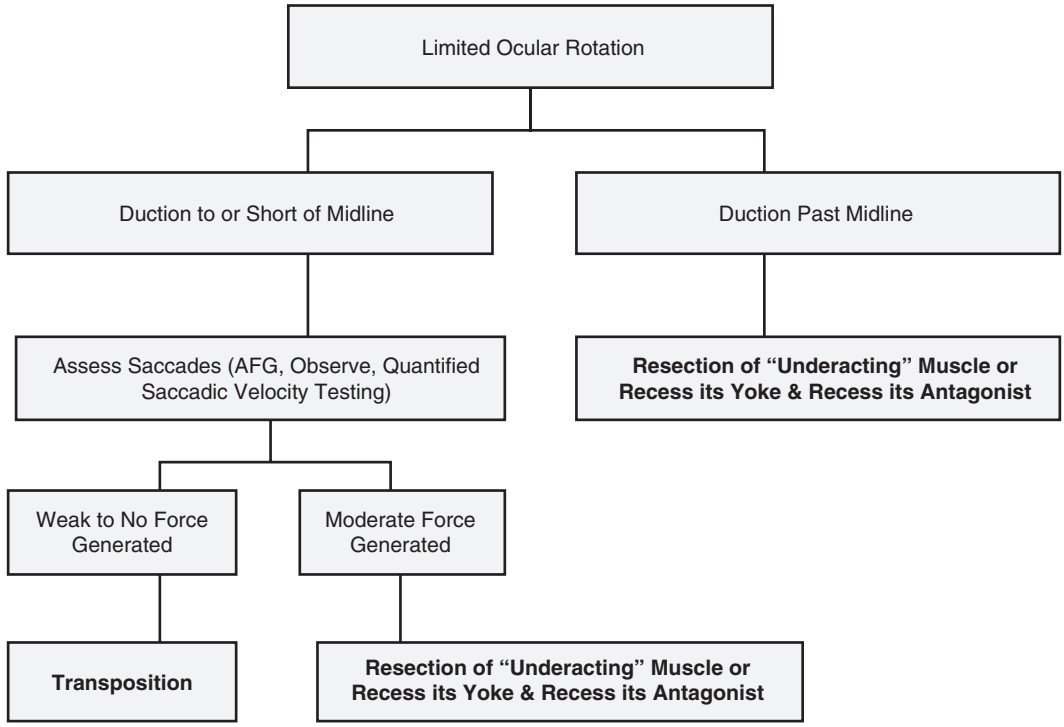


**Fig. 19.1** Algorithm for treating limitations of rotation

Rosenbaum and Santiago developed an algorithm for determining the surgical approach for patients with a limited duction [2]. Figure 19.1 is my modification of that algorithm, which relies less on quantitative saccadic velocity testing and incorporates the option of recessing the yoke of a mild to moderately paretic muscle.

## Basic Information

*Timing of Surgical Intervention*

When paralytic strabismus is acquired, you should wait for at least 6 months from the onset before surgically intervening, to allow for possible resolution or improvement that may require less surgery. If there has been no improvement by 6 months, resolution is very unlikely and surgical intervention is appropriate. However, if at 6 months the problem is still improving, you should wait until about 12 months from onset or until there is no further improvement. With acute sixth cranial nerve palsy, chemodenervation of the ipsilateral MR with botulinum toxin (BTX) may prevent secondary contracture and allow the patient to have some degree of single binocular vision while you are waiting for resolution. However, it does not improve (nor hinder) the likelihood of resolution [3]. Any benefit to using BTX in this situation relates to possible earlier visual rehabilitation.

If acquired paretic strabismus has improved but not resolved by 6 months, further resolution may occur.

# Sixth Cranial Nerve Weakness

## Basic Information

*Paresis Versus Palsy*

It is most important to distinguish between a partial sixth cranial nerve weakness (paresis) and a complete weakness (palsy),

because the treatments are different. Certainly, if abduction occurs past the midline, you are dealing with a paresis. However, the converse is not true. A paresis with substantial secondary MR contracture may not have abduction past the midline. You will need to rely on the aforementioned tests (forced ductions, active force generation, assessing saccades) to make the distinction.

### Important Point

You must distinguish between paresis and paralysis. Forced duction testing alone is insufficient for making that distinction.

### Basic Information

*Surgical Treatment of Sixth Cranial Nerve Paresis*

If you can establish that you are dealing with a paresis, a recess/resect procedure gives the best results. I find the use of adjustable sutures helpful in cooperative patients. If the deviation in primary is so large that you feel a recess/resect procedure is not adequate, add a recession to the contralateral MR.

### Basic Information

*Surgical Treatment of Sixth Cranial Nerve Paralysis*

Treatment of sixth cranial nerve paralysis should be dichotomized according to whether the ipsilateral MR is contractured to a degree that it needs to be released (recessed). In many cases if there is only mild MR contracture and very slightly positive forced ductions, a transposition procedure will be sufficient to overcome the restriction. But if the restriction is more substantial, some form of MR weakening is needed. Once that is decided, the specific type of transposition procedure recommended is a function of the size of the deviation in primary and the risk of anterior segment ischemia (e.g., had other muscles already been operated upon).

## Lateral Rectus Muscle (LR) Paralysis and Medial Rectus Muscle (MR) Is Not Contractured

This calls for a transposition procedure that does not require ipsilateral MR weakening. Listed in order of decreasing power the ones I utilize are the following:

1. Full tendon transposition of the superior rectus muscle (SR) and the inferior rectus muscle (IR) to the LR with lateral fixation augmentation with BTX to the ipsilateral MR: The vertical rectus muscles are oriented along the spiral of Tillaux. This is extremely powerful but rarely needed.
2. Full tendon transposition of the SR and IR to the LR with lateral fixation augmentation [4]: The vertical rectus muscles are oriented along the spiral of Tillaux (see Fig. 10.17). This is the same as in the first procedure above but without the BTX injection of the MR.
3. Full tendon transposition of the SR and IR to the LR without lateral fixation augmentation with BTX to the ipsilateral MR [5]: The vertical rectus muscles are oriented parallel to the LR (see Fig. 10.17).
4. Full tendon transposition of the SR and IR to the LR without lateral fixation augmentation and without BTX to the ipsilateral MR: The vertical rectus muscles are oriented parallel to the LR. This is the same as in the third procedure above but without the BTX injection of the MR.
5. Full tendon transposition of just the SR to the LR with lateral fixation augmentation plus recession of ipsilateral MR [6]: The SR is oriented along the spiral of Tillaux.
6. Full tendon transposition of just the SR to the LR with lateral fixation augmentation: The SR is oriented along the spiral of Tillaux. This is the same as in the fifth procedure above except that the ipsilateral MR is not recessed (see Fig. 10.17).

### Important Point

Orientation of transposed muscles so that the new insertions are parallel to the LR as shown in Fig. 10.17a creates a more powerful abduction vector than if they are oriented on the spiral of Tillaux as also shown in Fig. 10.17b. However, addition of lateral fixation sutures, which requires orientation on the spiral of Tillaux, is more powerful than orientation parallel to the LR.

### Question

I plan to do one of the vertical rectus muscle transposition procedures combined with BTX to the ipsilateral MR. Should I inject BTX prior to the surgery, during the surgery, or after it?


### Reply

Like so many things with which we deal, this is a matter of trade-offs. It is certainly easier and requires less expertise to inject the BTX under direct visualization at the time of the vertical rectus transposition. The downside however is significant. If the patient has an unwanted significant hypertropia (HT) immediately after surgery, you will not know if it is due to the transposition or spillover from the BTX injection. If the former, you would want to intervene immediately and reposition the muscles. If the latter, you need to wait until the BTX wears off. If you inject several days to a week before surgery, and do induce a vertical deviation, you are left with a dilemma as to whether or not to postpone surgery. You would not want to operate in the face of the iatrogenic vertical, because you will not be able to judge if the transposition induced a vertical just after surgery. I find injecting prior to surgery is more comfortable for the patient than injecting immediately after; the eye is not sore. Injecting several days to a week after surgery is probably best with respect to motility, but is least desired with respect to patient comfort. Assuming a cooperative patient, I inject after surgery. In a child or an uncooperative patient, I might inject during surgery, or choose one of the surgical options that does not call for BTX.

Some other transposition procedures worth mentioning include the following:

1. Transposition of the temporal halves of the SR and IR to the LR with or without lateral fixation augmentation, and with or without ipsilateral MR recession: Transposing half of the vertical rectus muscles is nearly as powerful as transposing the entire muscles, but somewhat less so. I utilized this approach for many years when I was concerned about anterior segment ischemia, either because the MR needed to be recessed or had already been recessed earlier. I now opt for full tendon transposition of just the SR.
2. Jenson procedure [7]: I think there are better alternatives to the Jenson procedure and it is no longer on my list of recommended procedures. This stems more from my experience in having to reoperate and try to modify or take down a Jenson procedure, rather than having bad outcomes. The concept behind the Jenson procedure was that anterior circulation would be spared because the muscles are not disinserted. I doubt circulation is maintained in the transposed halves of muscles, as they are under considerable tension; a case of anterior segment ischemia has been reported after a Jenson procedure [8]. Classically, the muscle halves are just approximated in the superior and inferior temporal quadrant by a loop of suture, which is not fixated in the sclera. At reoperation I find that the transposed halves of the flaccid LR have stretched out, allowing the halves of the SR and IR to return to their original untransposed orientation. This can be prevented by modifying the procedure by fixating the suture loops to the sclera in the superior and inferior temporal quadrants. In other cases, I have found that the transposed halves of the SR, IR, and LR have atrophied into tight fibrotic bands that are adherent to the sclera

throughout their length. This makes it impossible to modify or take down the transposition.

3. Adjustable transpositions: Carlson and Jampolsky described an adjustable technique for a half tendon transposition procedure [9], and Laby and Rosenbaum described an adjustable full tendon transposition [10]. I have no personal experience with either of these procedures, because I am happy with the transposition procedures outlined above; I have not felt the need to switch. However, I have heard from colleagues whom I know and respect that they are effective.

### Question

I would like advice on a 67-year-old patient with a complete unilateral traumatic right sixth cranial nerve palsy. He has had two BTX injections to the right (R) MR with no lasting benefit. He has a 25 prism diopters (Δ) esotropia in primary and −4 abduction in that eye. He is now 10 months post-trauma. What would be your preferred approach: full tendon transposition of the SR and LR with lateral fixation augmentation, or half tendon transposition (with or without augmentation) plus ipsilateral MR recession?


### Reply

This is not straightforward. Usually, if there is a complete sixth cranial nerve paralysis and −4 to abduction, the esotropia is more than 25 Δ in primary gaze. In your patient you need to be sure that this is not a case in which there has been moderate LR recovery and MR contracture is preventing abduction. You need to do active force generation or in some way assess LR saccades. If there is some LR function but MR restriction, a recess/resect procedure would probably work well. Or you could do half tendon transfers of the vertical rectus muscles combined with an ipsilateral MR recession. I would not use lateral fixation augmentation on the transposed halves, given the small deviation in primary. But if the LR is dead, I would do full tendon transfers of the vertical rectus muscles with lateral fixation augmentation, but not recess the MR.

## Third Cranial Nerve Weakness

### Basic Information

*Overview*

The third cranial nerve innervates four of the six extraocular muscles plus the levator and pupil. As such, there can be isolated weakness (complete or partial) of any one or several of the muscles it innervates, or the weakness can affect all of the muscles it innervates. In addition, acquired third cranial nerve palsies often will recover with aberrant innervation to any of the muscles, including the levator. The general principles for treating paretic muscles listed at the beginning of this chapter apply.

### Basic Information

*Isolated SR Weakness*

This was covered in the section on monocular elevation deficiency in Chap. 17, "Miscellaneous Strabismus Syndromes." Isolated SR weakness is probably the most common manifestation of partial third cranial nerve weakness.

### Basic Information

*Isolated IR Weakness*

If there is a complete palsy of the IR, an inverse Knapp procedure [11] is the most effective option. It there is a paresis, treatment consists of an ipsilateral SR recession and IR resection, and perhaps a contralateral IR recession, all depending on the size of the deviation and degree of paresis.

## Basic Information

*Isolated MR Weakness*

If there is a complete palsy of the MR, some form of transposition of the SR and IR nasally is in order. The relative effectiveness of the different options for transposition, e.g., full tendon, half tendon, and orientation on the spiral versus parallel to the MR, follows the same hierarchy as I outlined for temporal transposition for an abduction deficit.

## Advanced Information

*Paralysis of the Inferior Division of the Third Cranial Nerve*

The inferior division of the third cranial nerve innervates the IO, MR, IR, and pupil. Isolated paresis or paralysis of this nerve branch is relatively uncommon, with reported etiologies including local orbital disease, trauma, viral, ophthalmoplegic migraine, aneurism, vasculitis, demyelinating disease, and unknown causes [12]. The clinical findings include exotropia (XT) and HT of the affected eye with limited adduction and elevation, as well as iridoplegia. I have good success in treating this condition with a procedure first described by Knapp [12]. It consists of transposition of the ipsilateral IR to the MR, the ipsilateral LR to the IR, and doing an ipsilateral SO tenectomy (Fig. 19.2).

## Basic Information

*Complete Third Cranial Nerve Paresis*

If there is some active force found in the affected muscles, your best results will come from a recess/resect procedure, perhaps adding a recession of the contralateral normal LR to create fixation duress for adduction in the affected eye.

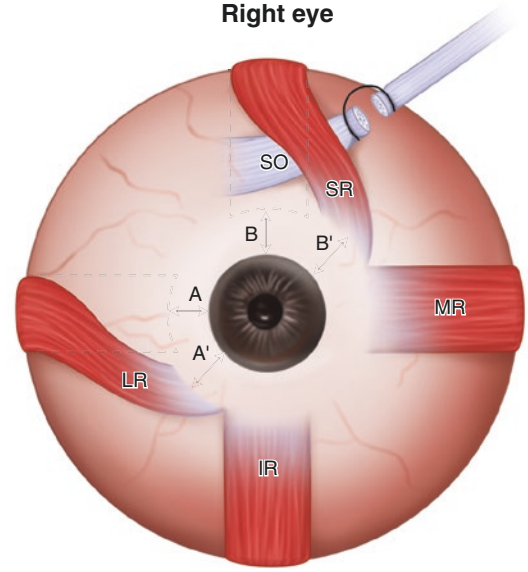


**Fig. 19.2** Procedure for treating palsy of the inferior division of the third cranial nerve. The superior rectus muscle (SR) is transposed to the medial rectus muscle, the lateral rectus muscle (LR) to the inferior rectus muscle, and superior oblique muscle (SO) tenotomy is performed. The original insertion to limbus for the LR (**a**) and the SR (**b**) is kept the same. The SO is tenectomized utilizing a chicken suture

## Basic Information

*Complete Third Cranial Nerve Palsy*

This is a very difficult condition to treat because there is so little remaining with function—just the LR and SO. My first line of treatment is a large recession of the LR and resection of the MR in the paretic eye, combined with anterior nasal transposition of the SO, as described by Scott [13]. This involves transecting the SO tendon at its border with the nasal edge of the SR, resecting several millimeters, and suturing the SO to the sclera 2 mm anterior and immediately nasal to the nasal corner of the SR insertion. Forced ductions should then be done; the desired endpoint is a mild restriction to abduction. If this procedure is unsuccessful, as a possible alternative the eye can be stabilized in the primary position with a nasal periosteal flap as described in Chap. 10, "Strabismus Surgery" [14].

## Advanced Information

*Aberrant Innervation*

This can take many forms and have varied presentations. There are circumstances in which you can use it to your advantage. I cared for a woman who had a traumatic complete left third cranial nerve palsy. She regained some MR function so that her primary position XT was 30 Δ and adduction was −2. She had complete ptosis, but had aberrant innervation to the levator. On right gaze the left lid would suddenly pop open when the right eye was about 30° into abduction. The surgical plan included a large RLR recession to cause fixation duress to right gaze. This not only cured her XT but eliminated the ptosis as well.

## Question

I care for an 18-year-old boy who had a traumatic left third cranial nerve palsy at age 3. He is amblyopic OS and suppresses. There is partial recovery aberrant innervation. He has 25 Δ left (L) HT in primary, which decreases to nil in left gaze and increases to 35 Δ in right gaze. There is residual ptosis, which decreases in upgaze where the left eye is a bit hypotropic. There is only −1 to adduction and there is no horizontal deviation in primary. I am thinking of infraplacing the LLR.


## Reply

Often with complex strabismus we have to choose between one treatment that is safer and simpler, but in the best of circumstances gives a less satisfactory outcome than one that is more complex and risky, but if it works gives far better results. This is the case with your patient. I think any procedure to just bring the left eye down will make a bigger hypotropia in left gaze, where there is no vertical now. I agree this must be aberrant regeneration of a left third nerve palsy. I think that aberrant regeneration, in which there is no vertical in eccentric gaze, or a reversal of the hyper in some eccentric gaze (as you have here) can be one of the most challenging cases we confront. Any "obvious" approach to reduce the hyper in primary will worsen it in left gaze and upgaze in your patient. The only effective approach, I find, is to see if there is a way to utilize the aberrant regeneration to your advantage. You can in this case, but it requires boldness and an understanding and willing patient, as it does not always work.

Here you have a big LHT in primary that goes away with innervation to left gaze. You can take advantage of this by moving the right eye outward, so when left gaze is innervated to bring the operated right eye to primary, you will get the decrease in LHT we now see in left gaze. This is similar to the principles of nystagmus surgery. But you must do enough surgery in the right eye to cause fixation duress to bring it to primary. Best would be a recess/resect procedure, as you would do for a nystagmus null point, like a RMR rec of about 6.5 or 7 and a RLR res of 10. Then you need to do horizontal surgery in the left eye to compensate for the XT this causes, like an LLR rec of about 11 and a LMR res of 8.

That said, there are other problems with just transposing the LR down. Doing so makes a vector for depression, but also weakens its abducting vector. I think you will end up with an esotropia in primary. If you want to try to fix this with a transposition, you could transpose both the MR and LR down, which will balance the eso and exo shift and hopefully cause no horizontal deviation.

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# Re-operations

# 20

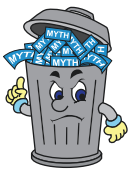
*View every failure as a substantial discovery in a mission of trial and error.*
*—Robert Grudin*

## Overview

### Basic Information

*Cooper's Dictum Revisited*

Every discussion about strabismus reoperations usually begins with a mention of *Cooper's dictum* [1]. Cooper is quoted as stating that then when doing a reoperation, you should approach it as a fresh case; for example, do not make allowances for what was done previously. In my opinion this is bad advice in many situations. However, in Cooper's defense, his words are really taken out of context. Cooper made his pronouncement as part of a discussion of a narrowly defined scenario. At the time a common precept was that if a patient has a consecutive exotropia (XT), one should always undo the prior operation (e.g., advance the medial rectus muscles [MRs]) and do a greater amount of surgery than had previously been done. Cooper, in fact, said, "Whenever an overcorrection results from extraocular muscle surgery, the case should be re-evaluated based on the diagnostic findings determined following the last operation. It is inadvisable to assume the attitude that one should undo some of the surgery which has previously been done. .. The idea that that surgery for secondary XT should *always* (emphasis added) undo what was previously done should be rejected." On the other hand, Cooper specifically advised that, if a consecutive XT has a convergence insufficiency pattern, the MRs should be advanced. It is only if there is a divergence excess pattern that the lateral rectus muscles (LRs) should be recessed. It is noteworthy that Cooper published these guidelines in 1961, when strabismus surgical formulae recommended much smaller recessions that they do now. In fact, Cooper advised that the MRs should never be recessed more than 3.5–4 mm. I imagine that if he were treating patients with consecutive XT who had undergone MR recessions of 6.5 or 7 mm, as is often done currently, he would have more frequently recommended advancing the previously recessed muscles. How Cooper's dictum became misconstrued to the reductionist idea that all reoperations should be treated as fresh cases is inexplicable to me. I imagine Cooper would have been greatly distressed to know that his words would become so misconstrued by so many people for so many years. There are countless scenarios in which treating a reoperation as a fresh case would be unwise. Certainly, if there are limitations of rotation, they must be addressed.




### Myth

Cooper's dictum states, "when doing a re-operation, approach it as a fresh case."

### Fact

Cooper advised against a formulaic approach and suggested the surgical plan be formulated based on all findings after the prior surgery, as well as what had been done before.


B. Kushner, *Strabismus*, DOI 10.1007/978-3-319-63019-9_20

## Basic Information

*Surgery on Previously Operated Muscles Versus Previously Operated Patients (But Unoperated Muscles)*

I feel any discussion of reoperations should be dichotomized based on whether you are operating on muscles (or an area) that has been previously operated or if you will be operating on previously unoperated muscles.

## Basic Information

*The First Decision You Must Make Is Whether You Should Operate Fresh Muscles, or Reoperate on Those Previously Operated*

The answer to this question may be different, depending on whether there is an overcorrection or an undercorrection. The following guidelines may be useful:

1. If there is a limitation of rotation (restriction or paresis), you must operate on the previously operated muscles.
2. There are soft signs for muscle underaction that should be considered:
   (a) If a consecutive XT has a near deviation that exceeds the distance by even a small amount, there is probably subclinical MR under action (UA); they should be explored and advanced or tightened [2].
   (b) If a consecutive esotropia (ET) has a distance deviation that exceeds the distance by even a small amount, there is probably subclinical LR UA; they should be explored and advanced or tightened.
3. If you are treating an undercorrection and the prior surgery was close to the "standard maximum"—4.5–5 mm or more for the MR or 6–7 mm for the LR—consider tightening the antagonist muscles of those previously recessed. Re-recessing when a large prior recession had been done is walking a fine tightrope between not enough correction and crippling the muscle. Careful judgement is needed to juxtapose the size of the prior recession and the amount of correction needed.

If you are treating an undercorrected XT, and the residual XT is now smaller in both R and L gaze compared to the primary position, you run the risk of overcorrection in side gaze if you re-recess the LRs, even if they only underwent modest recessions. Consider tightening the MRs. See Chap. 6, "Exotropia: Cultural and Social Implications," page , for a detailed discussion of the significance and management of lateral incomitance in surgically undercorrected intermittent XT.

1. If you are now dealing with a vertical problem after prior horizontal surgery, you can treat it like a fresh case as long as there is no limitation of rotation. If there is, consider an iatrogenic cause such as inadvertent incarceration of an oblique muscle.
2. Fig. 20.1 depicts a flow chart for decision making about operating on fresh or previously operated muscles.

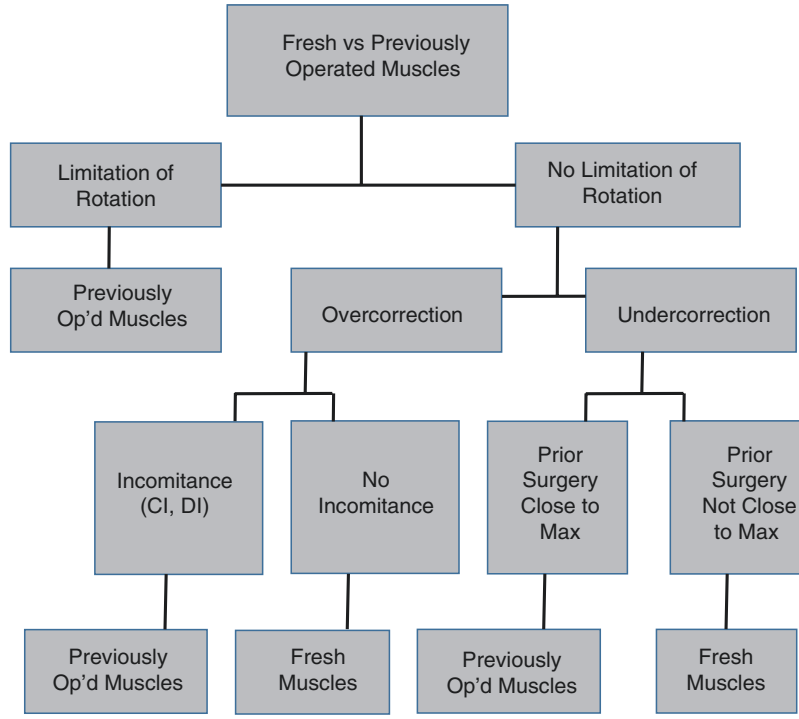



**Fig. 20.1** Flow chart for decision making about operating fresh or previously operated muscles. *Black* denotes clinical findings; *red* denotes treatment recommendations. Fresh = previously unoperated muscles. Previously Op'd muscles = previously operated muscles. CI = convergence insufficiency. DI = divergence insufficiency

## Operating on Fresh Muscles in Patients Who Previously Had Strabismus Surgery

### Basic Information

*When to Modify Surgery on Fresh Muscles*

In many cases, if the above criteria are met for operating previously unoperated muscles, you should not modify your surgery because of prior surgery. However, an exception might be when there were prior large recessions. As stated above, when Cooper practiced, surgical formulae had much lower doses than they do now. The idea of even a 5 mm recession of the MRs was new and almost considered radical. If a patient had very large prior recessions, e.g., MR recession of 6–7 mm, and I am operating for an undercorrection, I scale back my resections of the LRs by about 1 mm.

### Basic Information

*Evaluation*

I find it invaluable to obtain records of prior surgery, and do so whenever I can. This not only allows me to better plan in advance, but there are also some situations in which surgical exploration may not

be fully revealing. If I know that an inferior oblique muscle has undergone myectomy, I will not plan to do an inferior oblique muscle anterior transposition. If a posterior fixation suture had previously been done on a rectus muscle that I was planning to recess, and I did not know the prior surgical history, my recession may turn out to be ineffective. I might not identify the previously placed PF suture, as dissection for a recession is typically not that far posterior. If I cannot obtain the prior op note, I at least try to determine if the surgery was for ET or XT, and if one or both eyes were operated. Usually old photographs will help answer this first question.

In addition to the usual measurements of the misalignment, care must be taken to assess incomitance patterns and limitations of rotations. It is crucial to determine if any limitation of rotation is due to a paresis or restriction. Knowing the nature of prior surgery is helpful in this regard. If there is a limitation of adduction and the patient was previously esotropic, there is probably UA of one or both MRs. If a limitation of adduction follows XT surgery, there is probably a restriction laterally.

See Chap. 21, "Complicated Strabismus," for many representative examples of these principles.

### Basic Information

*Timing*

In most cases, reoperations should not be done prior to 6–8 weeks after surgery. Exceptions include a suspected slipped or lost muscle, a very large overcorrection after a superior oblique muscle tuck, or an HT of more than 5 prism diopters (Δ) on the day after a vertical rectus transposition. These are all best treated within 1–2 days after surgery. If you do need to intervene shortly after surgery, it is best to do so in the first week (the earlier, the better), as edema and vascularity will be less than if you wait until a week after surgery. These subside around 5–6 weeks after surgery. If you cannot intervene within the first week, it is best to wait until things settle down.

In most cases reoperations should be at least 6–8 weeks after surgery.

### Pearl

If you have to re-intervene early after surgery, it is best to do so in the first few days and no later than 1 week post-op. If you cannot, it may be best to wait until 6–8 weeks post-op.

## Operating on Previously Operated Muscles or Areas

### Important Point

When doing reoperations on previously operated muscles or areas, you must be prepared to alter your plan based on what you find. It is never formulaic.

If you want to make god laugh tell him about your plans—Woody Allen

## Technical Aspects of Operating on Previously Operated Muscles or Areas

### Basic Information

*Position of the Eyes*

As described in Chap. 10, "Strabismus Surgery," eyes are typically about 30 Δ more XT or less ET under deep anesthesia. If the alignment deviates from that norm, there may be a restrictive component. Jampolsky's spring-back balance test can be helpful in this assessment [3] (see Chap. 10).

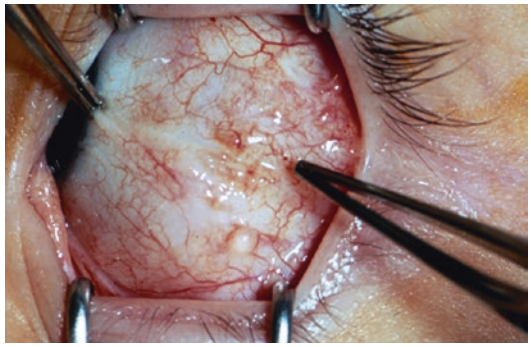


**Fig. 20.2** Conjunctiva causing restriction due to scarring from prior surgery. Note stringlike indentation of conjunctiva with forced duction into field of restriction. The forceps on the left is grasping the eye at the limbus. The forceps on the right is pointing to the stringlike indentation of the globe (photo courtesy of Art Jampolsky, MD)

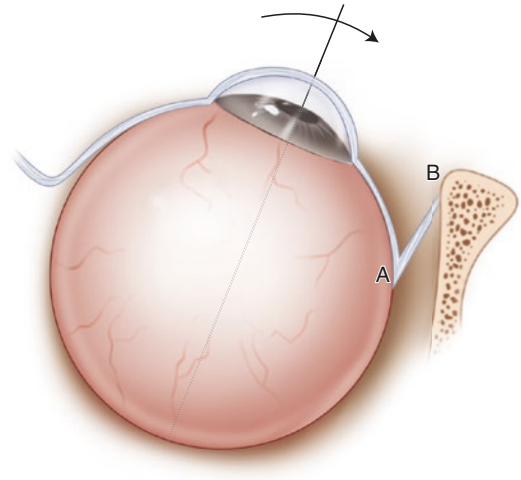



**Fig. 20.3** There is a reverse leash restriction extending from a point on the nasal sclera **(a)** anteriorly to a point on the nasal orbital wall **(b)** in this right eye. This will cause a mechanical restriction to adduction

## Basic Information

*Conjunctival Opening*

If the area of prior surgery does not look heavily scarred, and if there is no significant limitation of rotation, reoperations can be done using a fornix approach if that is your preference. However, if there is a limitation of rotation, you must consider the possibility that conjunctival scarring is a contributing factor and that conjunctiva may need to be recessed. If so, or if the conjunctival area looks heavily scarred, a limbus opening may be preferable. You should do forced ductions at the beginning of the case and observe conjunctiva as you rotate the eye into the field of limitation, looking for Jampolsky's string sign [4]. If the conjunctiva is causing a restriction, you may see a stringlike indentation of the conjunctiva as the shortened conjunctiva is put on stretch (Fig. 20.2).

## Basic Information

*Restrictions and Reverse Leashes*

When performing reoperation surgery on eyes with limited rotation, intraoperative forced duction testing is crucial. In most cases restrictions will be found opposite the gaze limitation. For example, if forced ductions are positive for adduction, the restriction will be temporal. But if you cannot free forced ductions after clearing the suspected area of adhesions, consider the possibility of a reverse leash restriction (Fig. 20.3). Look for it in the quadrant of the gaze limitation, e.g., nasally if the forced ductions are positive for adduction.

## Basic Information

*Hooking the Muscle*

In some cases of reoperation, the muscle hook will slide unimpeded under the muscle. In others there may be scarring between the muscle and sclera blocking the path of the muscle hook. In the latter case, you need to use a blunt scissors to open the path under the muscle, and then pass the hook from the opposite side, while keeping

the open scissor blade in place to act as a guide for the hook. If scarring is moderate, the hook may pass under the muscle but not come out cleanly on the other side. It will have connective tissue on its tip, which you can cut down on directly.

## Advanced Information

*Pseudotendon*

When a muscle is recessed, there is often a sheet of connective tissue that forms from the new insertion of the muscle to the original insertion, even if the muscle has formed a firm bond with the sclera at the recession site. This pseudotendon can look much like muscle if it is hooked, rather than the actual muscle, which is further posterior. On more than one occasion I was tempted to suture that tissue, thinking it was muscle, only to find the actual muscle bypassing a second hook further posteriorly. You should suspect this if you find what you think is muscle substantially anterior to where you expected to find it. This pseudotendon plays no role in the muscle's function. It should be excised, because it can get in the way of a muscle advancement, and just adds bulk to the overlying conjunctiva even if the muscle is re-recessed. This is different than the pseudotendon that forms when a recession is done on a suspension (hang-back), where the muscle itself is not bonded directly to the sclera [5].

## Important Point

Always suspect that you might have hooked a pseudotendon if what you think is muscle is found anterior to where you thought it should be.

## Advanced Information: Slipped Muscles and Elongated Scars

See Chap. 11, "Complications of Eye Muscle Surgery," for detailed discussion.

## Basic Information

*Closing Conjunctiva*

In cases where there is a limitation of rotation, you need to consider recessing conjunctiva. After the muscles have been repositioned, approximate conjunctiva at the limbus, feeling carefully for the amount of tension it is under. If it is restrictive, you should recess the conjunctiva. I typically recess it to the muscle's original insertion, passing my sutures directly into the insertion site. The bare scleral area will epithelialize and look quite natural. However, should further surgery be needed, you will not be able to raise a flap at the limbus. You will need to do so at the anterior edge of the recessed conjunctiva.

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# Complicated Strabismus

# 21

## Advanced Information: Overview

This chapter includes an eclectic collection of redacted versions of e-mail consultations I have received about complex strabismus cases. I consider this entire chapter advanced information, so I will not be repeatedly using that icon. For the most part I have avoided giving "outcomes" for these case presentations, and I am anticipating many readers will be troubled by this and may consider it an unfortunate omission. Over the years, I have received many e-mail consultations that are strikingly similar to one another. For presentation here, I often combined the salient points from multiple patients to end up with one conflated (perhaps hypothetical) patient presentation. In that case an "outcome" would also be fictitious. In addition, it is unfortunately the rule, rather than the exception, that I do not get follow-up from the ophthalmologist requesting advice. I am often unsure if they even followed my advice. In addition, my recommendations only represent one of many possible acceptable approaches—there are many roads to orthophoria. I have often felt that my making a recommendation, and then writing that it worked well, conveys the inappropriate idea that my approach was the one right one, and all others are incorrect. My intention in presenting these cases is to outline a thought process of how one may approach complex strabismus. For each one I will list one or more take-home "pearls" that the case was chosen to highlight.

There are many roads to orthophoria.

## Case 21.1 An Escape from Prism

### Question

I have a patient who underwent a left (L) inferior rectus muscle (IR) recession of 5 (elsewhere) for what was felt to be a right (R) fourth cranial nerve palsy. His vertical deviation is overcorrected and he now has an L hypertropia (HT) (manifested as an R hypotropia (HYPO) greater in left gaze with slight LIR under action (UA)). He also has an intermittent exotropia X(T) and is bothered by diplopia. He has equal vision but fixes OS. His measurements (prism diopters [Δ]) are as follows:

| | | |
|---|---|---|
| | 30 Δ RX(T)<br>10 Δ R(Hypo) | |
| 35 Δ RX(T)<br>6 Δ R(Hypo) | 35 Δ RX(T)<br>12 Δ R(Hypo) | 35 Δ RX(T)<br>20 Δ R(Hypo) |
| | 35 Δ RX(T)<br>20 Δ R(Hypo) | |


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My concern is that, although he can intermittently fuse in free space and fusion is easier with a small amount of prism, he cannot fuse if I offset the XT with 30 or 35 Δ base-in; he has vertical diplopia. I am wondering if that is because when he is fusing, both eyes are in primary, and his HT is smaller in primary than in left gaze. But with prisms to offset the deviation, his eye is exotropic, and the HT is bigger in side gaze. Does this mean I should not fully correct the XT?


### Reply

Your thought about the eye being in an exotropic position with prisms might make sense, depending on which eye the prism is held in front of. You are correct that when fusing without prism, both eyes, by definition, are in primary where the R HYPO is only 12 Δ. If you put the prism over the habitually deviating right eye, the left eye is still in primary, and that should not make it harder for him to fuse because of incomitance. But if you put the prism over the left eye, it then would be in left gaze where the HYPO is bigger, and fusion would be harder. I suspect that the inability to fuse with fully correcting prisms relates to the blur and image distortion that comes with viewing through a 35 Δ prism. I think that in this patient you should target 35 Δ for the XT correction, and advance the LIR, which may have slipped.

### Pearl

Keep in mind that when prisms are in place to neutralize a horizontal deviation, the eye behind the prism is not in primary if the eye without the prism is in primary. This can be important when assessing what happens to a vertical deviation that is horizontally incomitant, when sensory testing is done with prism in place.

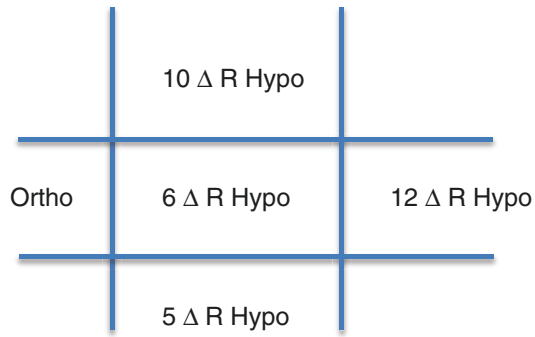
### Pearl

Although prisms are necessary to neutralize a deviation, large amounts of prism (rigid plastic, but Fresnel even moreso) induce blur, which can be an obstacle to fusion.

## Case 21.2 What Goes Up Must Come Down

### Question

I have a 3-year-old girl scheduled for surgery who underwent an RIO anterior transposition (AT) by another ophthalmologist. He placed the IO 2 mm posterior to the IR insertion. Review of prior records shows that pre-op she had an RHT but limited elevation OS. The girl was felt to have a monocular elevation deficiency OS. In spite of that the surgeon did what he called an IO AT OD. Now the girl has anti-elevation OD with −3 limitation of elevation in adduction. Elevation OS seems normal. Measurements are as follows:




She now has a face turn to the L of 20°. I assume she started as an R fourth cranial nerve palsy. Would you recommend converting the RIO AT to a standard recession, or doing an RIO nasal myectomy as described by Stager [1]?


### Reply

There is something fishy here for making the diagnosis of *anti-elevation syndrome* (AES). That entity does not occur with the IO placed 2 mm posterior to the IR. I suspect something like a fat adherence syndrome or a mechanical restriction. That said, if forced ductions are normal (they are normal in the AES) and if the IO is at the IR insertion, I am happy with just moving the IO straight back about 4–5 mm. I do not see the need for nasal myectomy. If forced ductions are abnormal, which I think is what you will find, you must free up the restriction and probably do a small RIR recession.

### Pearl

Forced ductions are normal with AES.

### Pearl

The AES is caused by a change in the torque vector of the IO, and should not occur with placing the IO several mm posterior to the corner of the IR insertion. If there is a limitation of elevation after such a procedure, it is probably caused by a restriction and not by AES.

## Case 21.3 When Depression Would be a Good Thing

### Question

This 35-year-old man had an orbital floor fracture OS that was repaired 1 year ago. He now has a vertically incomitant RHT, but his forced ductions seem normal. His midline measurements are as follows:

10 Δ LHT

18 LHT

35 Δ LHT

What is going on and how should I treat it? Should I recommend that the fracture repair be revised?


### Reply

Although this looks like a LIR palsy, in most cases it is a pseudo-palsy. If the fracture is posterior in the orbit and/or the muscle is adherent to the plate posteriorly, the force of the IR is transmitted to the adherence site, greatly decreasing its force vector for depression. In this case, forced ductions would be normal, but active force generation (AFG) would be decreased. In my experience, revising the fracture repair this long after the injury never results in resolution of the problem. You will need to treat this like an IR palsy, with either an inverse Knapp procedure or a contralateral IR weakening (recession or posterior fixation [PF]). Another possibility is a flap tear of the IR. You should look carefully for that, and if found, it should be repaired. Depending on how large it is, fixing the flap tear might correct the problem (unlikely) or you may have to combine that with other vertical muscle surgery.

### Pearl

Limited depression after orbital floor fracture is usually not due to nerve damage to the IR. It is a pseudo-paresis of the IR due to a force vector change caused by entrapment of the IR posteriorly, or a flap tear of the IR.

## Case 21.4 When Depression Is a Bad Thing

### Question

I recently saw a 50-year-old school teacher who had been physically assaulted by a student, resulting in extensive orbital fractures OD, which were repaired; a ruptured globe (also repaired); a R HYPO; and subsequent vision loss (best corrected 20/200). I performed a 5 mm RIR recession, followed later by an additional 3 mm of RIR recession for an undercorrection. This left him with 35 Δ of right HYPO with limited elevation, and he still had positive forced ductions to elevation. I then performed a free tenotomy of the RIR, which accomplished nothing. He still has 35 Δ of R HYPO. What should I do next?


### Reply

I did the next enhancement on this patient, and found the RIR attaching to the sclera 8 mm from the limbus. Forced ductions were positive and remained positive after I disinserted the RIR and until I freed up inferior restrictions. It is not completely surprising that I found the RIR essentially at the insertion after a free tenotomy. If a free tenotomy is done, and the eye stays depressed due to other restrictions, the muscle may bond where it inserted, or, in this case, anterior to the recessed insertion point. Some contractured muscles are very elastic and will retract further when disinserted. Others are like inelastic tethers. They may restrict gaze but do not retract when disinserted. This is sometimes the case in thyroid eye disease. You must use either fixed scleral sutures at the desired recession point (a suspension, also known as hang-back, will not suffice as the muscle will not take up slack) or pullover traction sutures post-op to rotate the eye away from the recessed muscle, thus forcing it to stay in the recessed position. For this patient I recessed the RIR 10 mm and used pullover sutures for 7 days post-op. After surgery he had less than 5 Δ of R HYPO.

### Pearl

A free tenotomy may result in the muscle adhering fairly anteriorly if the muscle does not take up slack, or if the eye stays deviated in the direction of the operated muscle. I never do free tenotomies.

## Case 21.5 A Preemptive Strike

### Question

My patient is a 57-year-old man who developed an R third nerve palsy after a retrobulbar block for a retinal procedure. He has ptosis OD and an incomitant XT and HYPO with diplopia. An orbital magnetic resonance image (MRI) was unremarkable with respect to the extraocular muscles. He is 20/20 OU, and has −3 to adduction OD and slightly limited elevation, with normal forced ductions. AFG is decreased for the RMR and equivocal for elevation OD. His measurements are as follows:

| | | |
|---|---|---|
| 10 Δ RXT<br>14 Δ R Hypo | 35 Δ RXT<br>20 R Δ Hypo | 50 Δ RXT<br>16 Δ R Hypo |
| 8 Δ RXT<br>12 Δ R Hypo | 30 Δ RXT<br>10 R Hypo | 50 Δ RXT<br>16 Δ R Hypo |
| 10 Δ R XT<br>2 Δ R Hypo | 25 Δ RXT<br>6 Δ R Hypo | 45 Δ RXT<br>6 Δ R Hypo |

Head tilt R: 35 Δ RXT and 12 Δ R HYPO. Head tilt left: 30 Δ RXT and 8 Δ R HYPO.

He describes intorsion, but I have not measured it due to the large horizontal deviation. I doubt it is large, as he can fuse with the vertical and horizontal offset with prism. I think he has enough RMR function for a recess/resect procedure (R&R) to work, but I am afraid I will overcorrect R gaze. I could do a smaller R&R and add a PF to the LLR

for added correction in left gaze. Or should I do a LLR res? What are my options for the vertical?


### Reply

A RLR recession and RMR res should be good for the XT; however, it will predictably overcorrect the smaller deviation in R gaze. This is an example of when you want to anticipate iatrogenic problems from your surgical plan, and preemptively act to prevent them. Adding a PF to the LMR would be perfect in this situation. Regarding the HYPO and intorsion, normally one thinks of weakening the R superior oblique muscle (SO), which often needs to be done in third nerve palsy. But here you have very little R to L incomitance, and oblique surgery has a large incomitance effect. So weakening the RSO will give a lot more vertical correction in left gaze than in right gaze, which you do not want here. I would recess the L superior rectus muscle (SR). Although it also will have more effect in L gaze than R gaze, the difference would be smaller. It will give less torsional correction than weakening the RSO, but you do not need a lot. Finally, it will give more correction in upgaze than downgaze, which is what is needed.

### Pearl

If your surgical plan for correction in primary gaze can be expected to cause an iatrogenic problem in an eccentric gaze, anticipate it and operate preemptively to prevent it.

## Case 21.6 Target Practice

### Question

I will be operating on a 9-year-old girl for an intermittent 25 Δ RXT. She had retinopathy of prematurity as an infant, and although she corrects to 20/20 OU, she has a moderate positive angle kappa OD. When she fixes OD, her eye appears to be about 20Δ exotropic. Should I target 25 Δ for the surgery, or should I operate for about 45 Δ to correct for the angle kappa?


### Reply

The important factor here is that this patient has good vision, an intermittent deviation, and, I assume, bifoveal fusion potential. As such, she will be able to fuse only when bifoveally fixating, and hence you want the right eye to be in its apparent exotropic position. Thus you should target 25 Δ. The situation would be completely different if there was poor vision and no potential for meaningful fusion. In that case it will not matter if she is not bifoveal and it would be fine to operate for 45 Δ, eliminating the cosmetic blemish of the positive angle kappa.

### Pearl

If a patient has fusion potential, you should ignore a positive or negative angle kappa and operate for the prism and alternate cover measurements. If a procedure is mainly cosmetic, you should take angle kappa into account and operate for the light reflex measurement. Krimsky is far preferable to Hirschberg.

## Case 21.7 Jail Break (Freeing from Incarceration)

### Question

I am caring for a 62-year-old man whose main complaint is torsional diplopia. He had a scleral buckle OS followed by four vitrectomy procedures OS for proliferative vitreoretinopathy. He is 20/20 OD and 20/100 OS and has been stable for 6 months. He has a 15 Δ LXT which is comitant, and with a Fresnel prism cannot fuse because of 15° of L excyclotropia. Interestingly, he only has 1–2 Δ of LHT, which is

also comitant. I know what to do for the XT but am unsure about the torsion. I assume that the SO is incarcerated in the buckle. Advice please.


### Reply

Although it is always wise to consider that an oblique is incarcerated in the buckle in cases of torsional diplopia after scleral buckling, I do not think that is what you are dealing with. First, if the SO were incarcerated in the buckle there is usually an incyclotropia, and here you have an excyclotropia [2]. If the IO was stuck to or tucked by the buckle, you would then expect an excyclotropia [3]. However, that syndrome should have a restrictive HYPO, which is not present in your patient. I suspect that one of the two things is going on. The most likely is that this is a manifestation of what Guyton described as a free-wheeling torsional drift that occurs in patients if fusion is disrupted [4], in this case due to the poor vision OS and the retinal procedures. I have seen this occur after scleral bucking procedures where torsion was not expected or tested for, due to the absence of a vertical deviation, and the patient was felt to have disruption of fusion [5]. The other possibility is that the retinal surgeon cut or disinserted the SO. This is less likely, as one would expect a vertical deviation if that occurred. As for treatment, I would first test the patient on a synoptophore with the torsion offset. That is the only way to tell if there is a chance for fusion if you correct the torsion surgically. Assuming that there is fusion potential, you should start the surgery with rotary forced ductions to test for a torsional restriction as shown in Fig. 10.10. I doubt in this case that you will find a restriction, but if you do, you should address it. If not, explore the LSO area. If it had not been cut and is intact, a Harada-Ito procedure would be a good choice. If it is not intact, or if the anatomy is too distorted from the retinal surgery to permit a Harada-Ito procedure, I would recommend a 7 mm temporal transposition of the LSR. That procedure could also be an alternative to a Harada-Ito procedure, even if the SO is intact.

### Pearl

Testing on the synoptophore is the only way to tell if a patient with significant torsion will be able to fuse if the torsion is corrected.

### Pearl

Not all torsional problems after scleral buckle are mechanical. A torsional drift from fusion disruption (Guyton's "sensory torsion" [4]) can occur. Rotary forced ductions can be helpful in sorting this out.

## Case 21.8 When "OD" Stands for Overdose

### Question

I am planning to operate on a 12-year-old girl with a consecutive XT that measures 30 Δ at distance and 35 Δ at near. There is mild MR UA which is probably secondary to the fact she underwent 6.5 mm MR recession OU at 6 months of age. Assuming that there is no slippage or stretched scar and I find the muscles in the expected position, should I advance them to the original insertions? How do you dose the amount to advance previously recessed muscles? Do you use a standard surgical dose table?


### Reply

Although many cases of previously unoperated strabismus may lend themselves to a surgical formula (e.g., *X* mm of surgery corrects *Y* Δ of strabismus), this does not work well with previously operated muscles. A recessed muscle shortens (loses sarcomeres end to end), which is clinically apparent as "stiffness."

A stiffer muscle will have more effect for the same amount of surgery than a less stiff muscle. Any surgical decisions should be made intraoperatively where the muscle stiffness can be assessed. This is a situation for repeated intraoperative forced duction testing and spring-back balance testing.

In short, there is no simple answer. I feel it is folly to apply formulas in this setting, as there are many variables. I think this is one area where experience counts for a lot. But here are some general principles:

1. If the muscle is found in a place that was not too far posterior to the equator (did not slip), you can "sort of" use the same formula used for fresh cases, thinking of each mm of advancement as being equal to a mm of resection, but with two important caveats. Often muscles in recessed positions become contractured and stiff. If so, scale back the surgery, as you will get more effect than normal for each mm of resection or advancement. Secondly, if you advance instead of resecting, you are increasing the wraparound effect, which slightly increases effect. That is why I said "sort of." Let's say I found a MR 10.5 mm from the limbus, and the deviation was something for which I would resect 6 mm if it was a fresh case. I might advance it 5 mm if not tight (not the full 6 mm due to increased wraparound effect) and maybe 4 mm if the muscle was a bit tight. This needs to be determined by feel intraoperatively.
2. If the muscle has slipped and is further back than had been initially planned, you just have to use judgement juxtaposing how far back the muscle is and the deviation size. But in this setting, just advancing the muscle anterior to the equator will increase its strength a lot.
3. Always look carefully for a stretched scar. If you find that, depending on the length of the scar and deviation size, you can often get by with just excising the scar and reattaching the muscle at the point the scar inserted on the globe.

### Pearl

When it comes to advancing previously recessed muscles for the treatment of overcorrections, there are too many variables to rely on a fixed formula of $X\ \Delta$ effect per mm of surgery.

## Case 21.9 Pullover to the Side, Please

### Question

I saw a 30-year-old man who had a lipodermoid removed supero-temporally OD. He developed some scarring, which restricted adduction. His oculoplastic surgeon reoperated, found positive forced ductions to adduction, removed conjunctiva and scar tissue in that area, and used amniotic membrane to cover the defect, followed by mitomycin C. He is now 6 months post-op and feels he is unchanged. There is −1 adduction of the right eye with positive forced ductions. A band of tissue runs from the temporal bulbar conjunctiva to the lateral canthus with a positive *string sign*. He is orthophoric in primary but becomes exotropic and diplopic at about 10° into adduction. I think I need to remove the scar, again try amniotic membrane, and, if any restriction remains, do a small LR recession OS. But I wonder about the role of postoperative traction (pullover) sutures.


### Reply

From your description, it does not sound like this should be too hard to fix. One important question is this: Was he initially better post-op, and *then* it recurred, or did the problem *never* improve? If the surgery never helped, even for a short time, the LR may be contractured despite the surgeon saying forced ductions were normal. (Sorry, but I am never completely confident if a non-strabismologist says that.) I have addressed similar cases recessing the

conjunctiva and scar tissue far back, after thinning it out. More recently, I use (and like) amniotic membrane and on rare occasions added mitomycin C. You may need to put the LR on a small adjustable recession. Keep in mind that he has a large diplopia-free field to the opposite side, and you want to be sure not to make him worse. Consequently, this sounds like a case in which Alan Scott's adjustable faden (R&R on same muscle) [6] done on the LLR would balance this mild duction deficit, especially because he is only −1 adduction. I think I personally would opt for one more attempt on the left eye, saving the LLR procedure for an enhancement, if needed. If he reports that it had been good initially after surgery and then recurred, I would consider postoperative pullover sutures. I leave them in 1 week. They are definitely uncomfortable but work well. Hopefully, they are not needed here.

### Pearl

Postoperative pullover sutures can be helpful in cases that recurrently scar down to an abnormal position.

## Case 21.10 When the Inferior Oblique Muscle (IO) May Be an Inferior Choice

### Question

I have been plagued with what to do with syndromic craniofacial patients with "IO over action (OA)" and SO UA" and big V-patterns. I question whether IO AT would be better than IO myectomy or other standard weakening procedures. But I am concerned about the AES, especially because these patients typically have significant fundus excyclotorsion, and IO AT does not reduce that. It seems that the procedure is fraught with problems with respect to fusion.


### Reply

I believe that there is not a "one size fits all" for these patients, and decisions should be based on orbital imaging. I do not routinely image all complex strabismus, but in these patients you need to know what is going on with respect to the muscles. Many have already had head computed tomography scans that may show the muscles. It is good to alert your cranial surgeon to always order at least one orbital view when imaging the child so that you do not need to repeat the scan. Most of these patterns are caused by orbital/muscle rotation, and the best fix is to reposition the rotated muscles. So if the MRs are up and the LRs are down, you may need to operate on them and move them in the right direction. Usually operating on the IO does not help a lot, unless it is tight on exaggerated forced ductions, in which case you should add that to the horizontal surgery. The hardest patients are the ones with absent or really atrophic SRs. I think the apparent IO OA in these patients is fixation duress due to the atrophic or absent contralateral SRs. Regarding your specific question about IO AT, I limit my use of it more than many colleagues, but feel comfortable with my indications. I mainly use it for dissociated vertical divergence (DVD) with IO OA, and it is my procedure of choice in that setting. I almost never do it unilaterally (there are rare exceptions), and I never do it in bifoveal fusing patients, as it causes a lot of incomitance.

### Pearl

Orbital imaging can be most helpful in planning surgery in patients with craniofacial syndromes. There is great variability to the muscle and muscle path anomalies that occur in this population, and knowing the anatomy in advance can be crucial to surgical planning.

## Case 21.11 "ET, Phone Home …"

I have a patient for whom I would like advice. He is a 58-year-old who is myopic (−16 D OU) with a long history of successfully wearing prism for a 15 Δ ET. He recently underwent MR recession OU of 3.5 mm by another surgeon, and immediately thereafter it increased to 25 Δ ET. He doesn't have a large HYPO of a classic heavy eye syndrome, but I wonder if he has a variant of that syndrome brought out by the bilateral MR recession. He does have a 5 Δ HYPO of the abducting eye in both R and L gaze, and moderate limitation of elevation. Imaging shows a moderate degree of bilateral SR nasal displacement and LR inferior displacement. It seems to me that connecting the muscle bellies of the SR and LR at or behind the equator as described for classic heavy eye might be too much for him and would overcorrect his deviation in primary, which is not too large. I am not sure if that can be done unilaterally here. He does have good vision and can alternate fixation. So I thought about resecting the laterals and displacing the insertion superiorly by 1 tendon width. I know that usually wouldn't be enough to correct a classic heavy eye syndrome, although maybe in this milder case it would be. I also wondered about putting a faden-type suture posteriorly on the LR to keep it from slipping inferiorly but am concerned about how thin the sclera will be in this patient.


### Reply

I agree that the full SR-to-LR union would overcorrect him in primary gaze. There are several options you might consider. In general, people feel that resecting the LRs is bad in these cases, as tightening the LR causes the globe to herniate out more. I have heard of one other patient who got worse after MR recession, and the only thing I can postulate is that weakening the MR let the LRs pull harder and caused the aforementioned effect.

I would do one of the two things for this patient. One would be a three-fourths or full tendon transfer of the LRs up without resection, and more MR recession on adjustable sutures. The other would be to use Dacron or a similar suture material in the muscles near the equator to create a sling to pull the SR and LR toward each other, but far short of having them approximate each other, as classically done for the heavy eye syndrome. Just get the muscles closer to the normal position. I am aware of this working well in several patients, but have not had the opportunity to try it myself.

### Pearl

If a lot of correction is needed for a duction deficit, but the primary position deviation is not very large, you can modify transposition procedures either by not transposing the entire muscle or by transposing the entire muscle part way.

## Case 21.12 It Was Traumatic to Be Blunt

### Question

This 66-year-old man had blunt trauma OS. He developed a traumatic cataract and is now aphakic. The orbital repair of an extensive inferior floor fracture and medial wall fracture was not immediate. He complains of diplopia that is vertical and torsional. He was fit with a contact but doesn't wear it due to diplopia. He has a left HYPO that is worse in upgaze, and a left HT in gaze down and left. His versions are limited for upgaze with elevation in adduction

worse than elevation in abduction. With double-Maddox rods he measures 6° of L incyclotropia. With prism offset he has torsional diplopia. Measurements were obtained with him wearing a trial contact lens. They were as follows:

| | | |
|---|---|---|
| 12 Δ L Hypo | 20 Δ L Hypo | 14 Δ L Hypo |
| 8 Δ L Hypo | 12 Δ L Hypo | 12 Δ L Hypo |
| 5 Δ L Hypo | Ortho | 6 Δ LHT |

Tilt R – 4 Δ LHT Tilt L = 5 L Hypo

I am thinking about exploring the LIR, getting rid of adhesions, and doing a LIR resection plus a recession on a semi-adjustable suture, with a temporal offset for the torsion. My reasoning is that the LIR resection would strengthen it for the downgaze lag, and the recession would ease the restriction.


### Reply

I do not think that combining a resection and recession on the same muscle has the effect of both strengthening and releasing restriction. If you think of the effect of a resection, you need to stretch the sarcomeres. If the recession is greater than the resection, you are still shortening the sarcomeres. If the recession and resection are the same, you are not changing sarcomere length. And if the resection is greater than the recession, you are stretching the sarcomeres, which will worsen he restriction. The procedure of doing both on the same muscle, as described by Scott [6], is a completely different concept, which in simple terms has the effect of doing an adjustable faden. In this case you do not want a faden effect on the LIR, as that would cause more limitation in downgaze. Prior to surgery, I would review his post-repair orbital images (if there are any) to see if the orbital portion of the SO is entrapped nasally (see comments below). I would check carefully at surgery to see if the LHT in downgaze is due to superior restriction. If so, recess the LIR with offset for torsion, and also recess the LSR a lesser amount. It is not exactly the same, but a similar principle has been described to treat IR paresis and restriction after orbital fracture [7]. You should also do rotary forced ductions. If restricted, the belly of the SO muscle may be entrapped nasally. If so, you will need to weaken the SO. If forced ductions are normal to downgaze, this is a paresis or pseudoparesis of the LIR. If so, recess the LIR with temporal offset for torsion, and also put a posterior fixation or do an adjustable recession of the RIR.

### Pearl

Rotary forced ductions can be helpful in sorting out limitations of rotation after orbital fracture.

## Case 21.13 It's Twisted

### Question

I have a really interesting patient for whom I would like advice. He is a 28-year-old man who underwent MR recession OU for a childhood-onset esotropia. Immediately thereafter he had a residual ET and developed an LHT. He then saw a second ophthalmologist, who found 12 Δ of LHT and 4 Δ XT at distance but 20 Δ ET at near. He recommended supraplacement of the RMR with PF, and PF on the LMR, which was subsequently performed. Afterwards he developed a small consecutive XT, a residual but smaller LHT, and torsional diplopia. He is now 8 years after the first operation. Adduction OS is limited (−2) and OD (−1). There is bilateral SO OA (+2). He has 10° of incyclotropia OD with double-Maddox rod testing; the right fundus is intorted and the left shows no torsion. If the deviation is neutralized with prism, he reports torsional diplopia. His measurements are as follows:

| | | |
|---|---|---|
| | 25 Δ LXT<br>6 Δ LHT | |
| 35 Δ LXT<br>5 Δ LHT | 25 Δ LXT<br>5 Δ LHT | 25 Δ LXT<br>5 Δ LHT |
| | 30 Δ LXT<br>5 Δ LHT | |

At near he has 35 Δ LXT' (measurement at near) and 8 Δ LHT'.

Head tilt right = 30 Δ LXT and R Δ LHT. Head tilt left = 30 Δ LXT and 10 Δ LHT.

After a 1-h patch test his near XT did not change.

## Questions

My questions are as follows:

1. Is supraplacement or infraplacement of the horizontal rectus muscles an appropriate way to treat a vertical deviation?
2. Would the supraplacement of the RMR be the cause of the torsion?
3. What explains the reversal of the HT on head tilt?
4. How should he now be managed?


## Reply

*Reply to question 1*. I do not recommend horizontal rectus muscle offsets for vertical deviations as large as he has. I employ them when a vertical of up to about 5–6 Δ accompanies a horizontal deviation that needs surgery, but if greater than 5–6 Δ, I add a vertical rectus muscle to the surgical plan. Although there have been attempts to associate a formula for the effect of offsets, e.g., 1 Δ per mm of offset [8], I find the effect to be nonlinear. This makes sense. As you raise or lower a horizontal rectus muscle, the vertical vector you create is a trigonometric function of the arc degrees the muscle is shifted. Thus, you get negligible effect for a 1 or 2 mm offset, and progressively more per mm of offset as the number increases. But when you get to large offsets (5 mm or more), you similarly start decreasing the horizontal vector. This can have unpredictable effects, and for that reason it is better to add a vertical rectus muscle procedure when the vertical is more than 5–6 Δ.

*Reply to question 2*. I think here the torsion is a result of the combination of supraplacement of the MR combined with PF. Vertical offsets of the horizontal rectus muscles will induce a torsional change, but in general it is less than with horizontal offsets of the vertical rectus muscles [9]. The addition of the PF suture will result in the posterior belly of the MR being transposed higher than if it were allowed to follow the shortest path, as it would if there were no PF suture. This will greatly increase the torsional vector, and I have seen this in several patients.

*Reply to question 3*. The head tilt test can be meaningless if prior surgery on the vertical rectus muscle, or in this case that affected the vertical force vector of horizontal rectus muscles, was done. We do not know the cause of the original LHT, so it is impossible to determine the impact of the subsequent surgery.

*Reply to question 4*. The findings indicate that the MRs are underacting, and based on that, I think the PFs should be taken down. This is not always easy, and one does not know what one will find going back on a PF. Nevertheless, I think that because there is a limitation of adduction, especially in the left eye, recessing the LRs will not give long-term success. As an aside, this is one of the reasons I do PF "when I have to," and not "whenever I can." It can be hard to reverse. The torsion is tricky. First, I would test him with prisms that overcorrect the XT and make him optically ET. It is unusual for a childhood ET to have this type of diplopia, and I would hope that making him a small ET again would have him suppressing. If so, just make him a small-angle ET. If not, I would not do a full weakening procedure of the RSO because he has equal and symmetric SO OA. Doing so would leave him incomitant. I would then do a tenectomy of the anterior seven-eighths of the RSO, or transpose the RSR temporally 7 mm combined with a small semi-adjustable recession.

### Pearl

Vertical offsets of horizontal rectus muscles can treat small vertical deviations. The dose response of these vertical offsets is not linear.

### Pearl

Raising or lowering a horizontal rectus muscle not only creates a vertical force vector, but it also creates torsional vector which may be accentuated if PF sutures are used.

### Pearl

If you cannot get a diplopic patient to fuse, see if you can find a scotoma in which they suppress. For surgical overcorrections this often requires replicating the direction of the initial deviation.

## Case 21.14 Buckling Up for Safety

### Question

I saw a 15-month-old with a history of infantile ET for which he underwent an R&R in the right eye. Subsequently, he was found to have a hereditary form of retinoschisis with large areas of schisis bilaterally. Our retina surgeon also thinks he may have a retinal detachment in the right left eye and has him scheduled for an exam under anesthesia with plans for a possible scleral buckling procedure at a later date if needed. By light reflex he has about 30 Δ of ET while wearing his cycloplegic refraction of +5.00 OU. I am considering an R&R for 30 Δ in the left eye. Should I do it at the time of the exam under anesthesia, to spare him one anesthetic, and spare me having to operate after the scleral buckling procedure?


### Reply

I have several comments:

1. If he has retinoschesis, it is quite probable that there is macular involvement and he may be fixing eccentrically. If he is 30 Δ ET now, and if that is by light reflex testing, he may actually be aligned with respect to his retinal fixation points, or at least less than 30 Δ ET. If that is the case, surgery to get rid of a cosmetic "ET" may not be the best functional answer. I grant you that in a 15-month-old, this can be hard to sort out.
2. Regardless of the above comment, I would not do strabismus surgery prior to retinal surgery. I understand the temptation, as it is easier, but retinal surgery can alter motility. If that were to happen, it would undo your good result. Then you are faced with an enhancement that is post-buckle and post-strabismus surgery. Not fun!
3. No rush to align him. At 15 months of age, and with schisis and an ET, there is no real chance of good fusion. So no reason not to delay.
4. If, at the time of exam under anesthesia, he does not and probably will not need a scleral buckle, one could make a case for straightening him while he is asleep. I would do so.

### Pearl

If retinal detachment surgery will be needed, do not do strabismus surgery prior to it, as scleral buckling can alter motility.

## Case 21.15 An "Inferior" Oblique

### Question

I saw a 24-year-old man who had a benign orbital tumor OD that caused diplopia. The oculoplastic specialist who resected it indicated that it was contiguous with the RIO temporally, and extended above the RLR superior

border. His diplopia worsened, and when I saw him he looked like he had Brown syndrome OD. He had a large R HYPO that increased on L gaze and R head tilt along with 8° of R incyclotropia. I assumed that he either had a fat adherence or some type of restriction, versus a RIO palsy. When I operated, I found completely normal forced ductions, so I did an RSO tenectomy with a chicken suture. Afterwards, his primary position HYPO improved from 10 Δ to 5 Δ, but he is still −4 to elevation in adduction, and −2 to elevation in the midline, and is diplopic everywhere except in right gaze or with a right head tilt. I have seen IO palsy after orbital surgery but never with this degree of limitation of elevation in adduction; usually it is only −1 or −2. I feel my options are limited due to the incomitance and the fact that I already weakened the RSO. What next?


### Reply

I agree it is perplexing that an IO palsy would cause −4 to elevation in adduction. This scenario really suggests restriction, but with normal forced ductions, one must think out of the box. My first thought is LR displacement or instability (call it pulley slippage), which can cause a pseudo-Brown picture [10]. I have seen −4 to elevation in adduction caused by the LR slipping down on attempted elevation or elevation in adduction. With the tumor against the IO and LR, it could have disrupted the LR attachments to the SR. I would get a good-quality orbital MRI to see if anything is out of line. Do a dynamic scan to see what happens to the RLR on attempted elevation, specifically looking for inferior slippage, and also to see the contractile change in the IO. If the LR does slip down, the fix is to fixate its belly around the equator to the sclera in a slightly elevated position.

For the sake of discussion, let's say everything looks good on the scan. Then you are limited as to how good you can get him. I would do a larger RSO weakening procedure (longer chicken suture or tenectomy) and recess the LSR. I realize here you are trading off primary for side gaze. It will undercorrect him in adduction and probably overcorrect in abduction, but could get him good in primary with a reasonable range of simple binocular vision. See the section on monocular elevation deficiency in Chap. 17, "Miscellaneous Strabismus Syndromes." If your patient is only −2 to elevation in the midline, it should help. *Postscript*: This patient did, in fact, have pulley slippage with the RLR sliding far inferiorly on attempted upgaze. The aforementioned stabilization procedure corrected the problem.

### Pearl

A complete paralysis of the IO will not cause −4 to elevation in adduction. If versions in the IO field are that limited, consider a restriction or LR pulley laxity.

## Case 21.16 Tort Reform

### Question

My patient had an Ahmed drainage device implanted superotemporally in her left eye, and now has 15 Δ of LHT and 10° of excyclotorsion OS. I presume that there has been disruption of the LSO. There is some limitation of adduction and elevation, so I cannot really tell if there is "IO OA." I would prefer not to go through all the scar tissue to do a Harada-Ito procedure. How much torsional correction would I get from an LIO recession?


### Reply

I do not think you will get as much as 10° of torsional correction. You will probably get about 5°, which may be enough. But that is provided that the torsion is not due to a restriction. You will need to do rotary forced ductions to see if there is a bias in one direction of rotation compared to the other. If so, there is no option but

to free it, or try and fix the torsion by matching it in the other eye. In other words, incyclorotate the right eye to make the torsional misalignments parallel. I have done this on several occasions, and know of others who have so with good results.

### Pearl

If there is a torsional restriction as determined by rotary forced ductions, you will get less than the usual torsional correction by operating on other cyclorotary muscles, if you do not relieve the restriction.

### Pearl

If there is a torsional problem you cannot correct, you can eliminate symptoms by creating a parallel torsional misalignment in the other eye, e.g., create incyclotropia in the fellow eye of a patient with a non-fixable excyclotropia in the affected eye.

## Case 21.17 An Unsolved Abduction Case

### Question

I care for a 30-year-old man who is mildly amblyopic (20/40) in his left eye and has a stable traumatic sixth cranial nerve palsy in his preferred right eye. With right eye fixing he has 50 Δ LET and with left eye fixing he has 25 Δ RET. He is −2 to abduction in his right eye. Should I target 25 or 50 for my surgical correction?


### Reply

The important factor here is that he is amblyopic and will not shift fixation after surgery. The other important factor would be whether you operate the right or left eye. With only −2 to abduction, an R&R would probably suffice. If he only lets you operate on OS, or if that is your preference, you need to operate for 50 Δ. He will continue to be fixating under duress if the right is not operated, and that will be driving the larger secondary deviation. If you operate the right eye, you eliminate the fixation duress. So you need to operate only for the deviation he shows when not fixating under duress, which in this case is 25 Δ.

### Pearl

When there is a secondary deviation due to fixation duress (paresis or restriction in one eye) you should operate for the smaller deviation if the surgical plan will eliminate the fixation duress, or the secondary deviation if there will still be fixation duress after surgery.

## Case 21.18 Inferior Superior Obliques

### Question

I treat a 5-year-old girl with a V-pattern XT with an incomitant HT, which I think is due to bilateral superior oblique palsy (SOP) because there is significant reversal on head tilt. She has bilateral IO OA (+2) and bilateral fundus excyclotropia. She has equal vision and a negligible refractive error. Is it OK to do 8 mm IO recession OU and 6 mm LR recession OU, or can I get away with just doing the LRs and supraplacing them? Her measurements are as follows:

| | | |
|---|---|---|
| 35 Δ X(T)<br>8 Δ RH(T) | 35 Δ X(T)<br>8 Δ RH(T) | 35 Δ X(T)<br>8 Δ RH(T) |
| 25 Δ X(T)<br>8 Δ RH(T) | 25 Δ X(T)<br>10 Δ RH(T) | 25 Δ X(T)<br>10 Δ RH(T) |
| 20 Δ X(T)<br>5 Δ RH(T) | 20 Δ X(T)<br>8 Δ RH(T) | 20 Δ X(T)<br>8 Δ RH(T) |

At near she is 20 X.

Head tilt right: 20 Δ XT and 25 Δ RHT. Head tilt left: 20 Δ XT and 10 LHT.


### Reply

Neither of the procedures you describe will adequately address the HT in primary. There are several options as I see it:

1. Recess the LRs 25Δ plus IO recession OU, recessing 12–14 mm in the R eye and 6 mm in the left. However, in my experience this will not quite collapse 10 Δ of HT, which is what you need.
2. Do the above, but also infraplace the RLR 3–4 mm, which should add enough vertical correction.

or

3. The most predictable approach would be to recess the LROU for 25 Δ, recess IO OU 8 mm, and also recess one vertical rectus for 10 Δ, either the RSR or the LIR. This latter approach is a bit more complex but most predictable.

I would choose the third option, but the second is fine.

### Pearl

You do symmetric vertical surgery and expect to collapse a significant HT in primary.

## Case 21.19 Out of Balance

### Question

I examined a 7-year-old boy with bilateral asymmetric SO OA, resulting in a HT in primary, and in X(T) that is horizontally incomitant. He has a face turn to the right. Both his HT and X(T) are better in left gaze. How should he be approached surgically? His measurements are as follows:

| | | |
|---|---|---|
| | 15 Δ X(T)<br>4 Δ L (Hypo) | |
| 25 Δ X(T)<br>12 Δ L(Hypo) | 20 Δ X(T)<br>6 Δ L (Hypo) | 10 Δ X(T)<br>2 Δ LH(T) |
| | 30 Δ X(T)<br>8 Δ L (Hypo) | |

At near he is 10Δ X' and 4 Δ L HYPO.

Versions show +3 RSO OA and +1 LSO OA with bilateral fundus intorsion. He fixates with about 25° of face turn right, and that is the main complaint. How can I sort this out?


### Reply

I have seen about 8–10 patients with bilateral asymmetric SO OA resulting in a HT of the lesser affected eye in primary, who had a face turn toward the lesser affected eye. This puts the more affected eye into abduction and out of the field of the SO, thus neutralizing the vertical deviation. That may be driving the turn in your patient. I have treated them by doing an asymmetric SO weakening procedure. Initially, I did a disinsertion in the lesser affected eye and a nasal tenotomy in the other. Although my results were acceptable, I longed for a procedure that would allow for better titration. When I started doing silicone expanders, I found I could titrate the effect for asymmetric versions, but it was unsuccessful in collapsing the HT in primary. The split tendon lengthening procedure seems more effective than the expander for this situation [11]. However, your patient has an incomitant X(T), which also decreases significantly in the preferred head position, and this also could be driving the face turn. If the former, you must do asymmetric

SO weakening. If the latter, you need to tailor the exotropia surgery to address the incomitance. In this setting, I like to use prisms to sort it out. Have a trial with 6 Δ of vertical to correct the HT, either in the office in a trial frame or with a Fresnel, and observe if the face turn resolves. Then do the same thing with horizontal prism to correct 20 Δ of XT. This will tell you which problem must be addressed; however, there is really no reason not to address both. I would do bilateral split tendon lengthening of the SOs, about 4 mm more in the right than left. For the XT, you could do an R&R in right eye, which would address the incomitance. If your preference is for lateral rectus recessions, you can do that instead, but do about twice as much in the right eye as the left, with the total amount of recession equaling what you normally do for 20 Δ.

### Pearl

When it is not clear if a head posture is driven by a vertical or horizontal incomitance (or in other cases, nystagmus), you can use prisms diagnostically to sort this out.

## Case 21.20 Double Trouble

### Question

How would you approach this 36-year-old woman whom I had treated for an RSO palsy 6 years ago. Because her deviation was significant in primary, worse in upgaze and left gaze, and she only had a minimal RH in downgaze, I recessed and anteriorly placed the RIO level with and at the temporal edge of the RIR insertion. I thought a standard recession would not correct the primary position deviation, and I did not want to add a contralateral IR recession for fear of overcorrecting her in downgaze. She did well for several years but now has a symptomatic LHT and is diplopic. Her measurements are as follows:

| | | |
|---|---|---|
| 16 Δ LHT | 14 Δ LHT | 12 Δ LHT |
| 16 Δ LHT | 12 Δ LHT | 6 Δ LHT |
| 6 Δ LHT | 4 Δ LHT | 6 Δ LHT |

Head tilt right: 10 Δ LHT. Head tilt left: 10 Δ LHT.

Versions show +2 LIO "OA", +1 LSO "OA" and −2 RSR "UA."

I assume that she was a bilateral masked SOP. Should I treat her as such?


### Reply

I am really suspicious that this is the AES in the right causing fixation duress appearing like LIO overaction. It can be an art to sort out if it is that, versus a bilateral masked SOP, or a simple overcorrection, which can mimic a bilateral masked problem. Some objective signs may help point the way. You need to look for fundus torsion. If there is none in the left and excyclo in the right, it is most likely AES. The converse is not always as meaningful. The absence of fundus excyclo OD does not rule out AES. The presence of excyclo OS would speak toward a bilateral masked SOP. The absence of a positive Bielschowsky head tilt test strongly speaks to AES and against either bilateral masked SOP or a simple overcorrection. However, again, the converse is not as helpful. Sometimes AES will have a positive Bielschowsky and sometimes not. The main way to tell is by observing the dynamics of version testing. With AES you get the sense the OD is not elevating well, as opposed to the LIO overacting. A characteristic finding is the lower lid elevating on upgaze. Look for a bulging of the lower lid OD on attempted upgaze. Note that forced ductions are normal in AES, as the limitation of elevation occurs only when the IO is recruited. Finally, the timing here is typical for AES, which occurs more frequently as time goes by. This is one of the reasons I do not like to do IO AT in bifoveally fusing

patients. A simple overcorrection should have occurred sooner. For some reason, bilateral masked SOPs often do not unmask until some years pass. Assuming that this is AES, the treatment is to retroplace the RIO to a point about 4–5 mm posterior to where you placed it. If this is a bilateral masked SOP or a simple overcorrection (both unlikely), I would recess the LIO.

In the end, you need to commit to LIO OA or AES OD. If the latter, put the IO back along the IR border about 4–5 mm post to the insertion. If not AES, recess the LIO.

As an aside, I *never* do IO anterior transposition in fusing patients unilaterally for this reason. I think the AES comes on with time. The longer you follow them, the more patients develop it.

### Pearl

It can be hard to tell AES from a contralateral unmasked bilateral SOP or simple overcorrection. It is crucial to make the differentiation, as the treatment of AES is completely different than the other entities.

## Case 21.21 Double Vision Is Not Twice as Good as Single Vision, It's Actually Not Half as Good

### Question

I did an LLR recession of 9 mm and LMR res of 5 mm a 14-year-old girl for an almost constant 40 Δ X(T) at distance and near. She was 20/20 OU with her cycloplegic correction of −3.00 D OU. She had neither fusion nor stereopsis. A week after surgery she was 8 Δ ET and 14 Δ ET' with diplopia. By 3 weeks post-op she was orthophoric and 14 Δ ET' with small bilateral latent DVD. She still has uncrossed diplopia and I cannot get her to fuse with prism. Two weeks of alternate occlusion did not help. Although she is able to function well in school and home, she sometimes complains of visual confusion when viewing at the blackboard in school; she sees two different objects superimposed on one another in the straight ahead visual direction.

I have four specific questions:

1. What should be the next step in managing this patient?
2. Does she require a second surgical procedure to correct the consecutive ET?
3. Should I aim for orthotropia to eliminate her diplopia, or should I aim for a small XT?
4. Why is she having diplopia at distance in spite of having no horizontal deviation? What would be the best surgical plan for her and when should that be done?


### Reply

These can be tough and hard to sort out, but here is an approach I have found successful.

The fact that she has DVD means she has the sensory backdrop of infantile strabismus and not the usual for X(T). The goal is not to get her to fuse but find what it takes to get her to suppress. Start with the prism bar over the eyes to optically put her back at her original angle. For example, if she is now orthophoric at distance, and pre-op was 40 Δ XT, use 40 Δ base-out. She should suppress and see single at that angle. Then decrease the prism slowly to find the point at which she is out of her scotoma and sees double. Usually this will be a fairly small angle of XT. Then put her in a Fresnel prism for the least amount she needs to be single. Usually over a period of months you can progressively decrease the prism and ultimately (sometimes) get her out of prism entirely. If not, you need to either keep her in prism, or operate to put her at the angle at which she suppresses.

### Pearl

The goal in some patients with seemingly intractable diplopia is not to get them to fuse, but to find out how to get them to suppress.

## Case 21.22 Whole Lotta Shakin' Goin' On

### Question

I did bilateral MR recession of 6 mm in a 1-year-old for a large-angle ET. She also had a manifest jerk nystagmus and alternated fixation with fixation in adduction, and hence had alternating face turns. She now has a 25 Δ XT with mild MR UA OU (−1), but still fixates in adduction with alternating face turns, making the XT even bigger due to fixation duress. I am concerned that advancing the MRs will worsen her face turn, as will recessing the LRs. Advice please.


### Reply

This is an uncommon situation but I have a few thoughts. In nystagmus patients with alternating turns I always want to be sure that it is not manifested latent nystagmus, which would damp if fusion occurred. In order to tell, I do a trial of prism in the office, or better, with a Fresnel to offset the deviation and see what happens. If the turn is improved, then things are easy. Just fix the exo, get her fusing, and the turn should improve. But of course the strabismus gods are not always shining on us, and that may not be her response. I think here your best option is one I do not like in general, but invoke here. Specifically, do posterior fixation on the MR OU and recess the LR OU.

### Pearl

Whenever there is an alternating head turn with nystagmus that changes with a change of fixation (as opposed to periodic alternating nystagmus), consider the possibility of manifested latent nystagmus.

## Case 21.23 Divergent Opinions

### Question

I have a 69-year-old patient with a 10-year history of diplopia at distance. He has a relatively comitant 25 Δ ET with diplopia at distance but at near is only 6–7Δ of E without diplopia. He is about −5.00 OU with 20/20. I have seen a lot of "divergence insufficiency" ET but not to this extent. I assume I should resect the LRs but am afraid of making him diplopic at near. What do you recommend?


### Reply

I agree this is a fairly extreme degree of divergence insufficiency, and in fact as much as can occur. Recall that if eyes have the same alignment at distance and near, they change by an amount equal to the interpupillary distance (about 6 in an adult) times the dioptric equivalent of the near target distance (about 3 D) for a total of about 18 Δ. Thus converging from distance to near normally results in 18 Δ of convergence, and going from near to distance about 18 Δ of divergence. Given that this patient is about 18 Δ moreso at distance, he is not demonstrating any divergence at all. Although many advocate treating divergence insufficiency ET with LR res OU, there are several clinical scenarios that you should rule out before using that approach; it is not one size fits all.

1. Tight MRs can cause the motility pattern. I have seen it in subclinical thyroid eye disease affecting the MRs. I have also seen unexpected tight MRs in patients with chronic sinusitis. The lamina papyracea is paper thin (hence the name) and the MRs run adjacent to it. I believe that chronic sinusitis can cause fibrosis in the adjacent MRs. Forced ductions with attention to MR tightness is crucial here. If tight, you will want to recess the MRs.

2. The *sagging eye syndrome* is an aging change, in which the connective band between the SR and LR dehisces and the LRs sag. Reports are mixed. Clark advocates elevation and fixation of the LR belly to the sclera at the equator, whereas others report good results with LR resection [12, 13].
3. You can only diagnose this with good coronal orbital images. Also, if there is a history of sinusitis, I would recommend orbital imaging. If not, then resect the LRs. In most cases (but sadly not all), doing that in this setting does not make them XT at near.

**Pearl**

Divergence insufficiency can be caused by tight MRs or the sagging eye syndrome, as well as just weak LRs.

## Case 21.24 A Superior Inferior Oblique

**Question**

How worried should I be about the small size of the primary position deviation in this patient? After closed head trauma he has what looks like a RSO palsy. However, there is no deviation in primary. He has 18 Δ of RHT in left gaze, and more up and left; there is +2 RIO OA. He has 20° of excyclotropia, and his diplopia starts just 20° into left gaze. I did a 1-h patch test and only got 2 Δ of RHT. I am concerned that an RIO recession will overcorrect primarily.


**Reply**

I think it is right to worry about this, as there is a good chance of overcorrecting with an IO recession. You were wise to do a patch test, which usually brings out enough HT that you would not worry about weakening the RIO, but that did not happen here. Whenever I see a really small or no HT in primary in this setting, I look extra hard for bilateral masked SOP. Look for torsion in the other eye, the presence of a V-pattern, very small changes on head tilt, and a reversal of the HT in the field of the contralateral IO or SO. All of these point toward a bilateral problem, and if found, you should consider bilateral surgery [14]. If it is not a bilateral masked SOP, you need to do something to prevent an overcorrection in primary. In this situation, I like to recess the ipsilateral IO and put the ipsilateral IR on an adjustable suture with a small recession. The patients in whom I did this ended up typically needing about 2 mm of recession (after any adjustment), so I assume I would have overcorrected them had I not done this second muscle.

**Pearl**

A very small HT in primary in a patient with an SOP, and a large deviation in adduction, is a soft sign for a bilateral masked SOP. The same is true if there is a very small forced head tilt difference.

**Pearl**

If treatment of a deviation in an eccentric gaze threatens to compromise good alignment in primary, anticipate the problem and preemptively act to prevent that.

## Case 21.25 Over and Under

**Question**

I would like advice on this 18-year-old girl. She initially had a LSO palsy which only measured 2 Δ LHT in primary, and 12 Δ in right gaze, and was orthophoric in left gaze. There was no torsion. I recessed the

LIO placing it 3 mm posterior to the temporal corner of the LIR insertion. She is now overcorrected, having stabilized at 6 Δ RHT in primary, 14 Δ RHT in right gaze, and 2 Δ RHT in left gaze. There is −2 to elevation in adduction OS and a subjective L incyclotropia of 5°. She is diplopic but, despite the torsion, she can fuse with vertical prism. I was thinking of recessing the RSR but am wondering about weakening the LSO (which was paretic to start with).


### Reply

There are a few odd things here. I am not 100% sure that this was initially a LSO palsy. I do not know if "no torsion" means subjective only, or if there was none on fundoscopy. It would not be uncommon for a congenital SOP to have none subjectively, but it should be present on fundoscopy. If there was, and if the head tilt was positive, that would convince me this was a SOP. With an acquired SOP, there should be both objective and subjective torsion. Also, although overcorrections can and do occur after IO weakening, it is really uncommon for there to be a significant underaction of the IO post-op, unless there is some adhesion or restriction.

Putting the IO 3 mm back but not at all lateral is closest to a 12 mm recession based on Apt's Tables [15]. That is a generous recession for only 2 Δ of HT in primary. So, possibly is just an overcorrection.

I would approach her as follows: First, check fundus torsion and see if that matches the subjective. Is there an incyclotropia OS? If so, this is probably an overcorrection or a restriction. I would choose a procedure now that addresses the torsion as well as the vertical pattern. But if there is no torsion on fundoscopy, you don't have to.

At surgery, carefully do forced ductions for a restriction inferiorly OS. If positive, free it up. Depending on how severe (e.g., can it account for the whole limitation of elevation?), you may want to convert the IO recess to a smaller recession. Changing it to a 6 mm recession would have you put it 5 mm posterior and 6.5 mm lateral to the temporal corner of the IR insertion. Do that if the restriction is not enough to explain the limited elevation. If no restriction is found, you could still convert the IO recession to a 6 mm, or weaken the LSO. I would do a small or modest procedure such as a post seven-eighths tenectomy, or a small lengthening (spacer, split tendon lengthening, or tenotomy with a short chicken suture). If there is no fundus incyclotorsion OS, you could just weaken the RSR.

### Pearl

If you think you have a simple overcorrection after IO weakening, and if there is more than −1 limitation of elevation in adduction, there is probably a restriction limiting elevation.

## Case 21.26 The Masked Marauder

### Question

Three months ago I operated on a 7-year-old boy who showed signs of a right fourth nerve palsy. Prior to surgery, he was orthophoric in right gaze, 10 Δ RH(T) in primary, and 35 Δ RHT in left gaze with +3 to +4 overaction of the RIO, normal action of the LIO, and normal action of the RSO. I don't recall much of a head tilt. I performed an uneventful right inferior oblique myectomy via an inferotemporal fornix incision.

Over the 3 months since surgery, he has developed signs of a left fourth nerve palsy versus an iatrogenic AES of the right eye. He lifts his chin and tilts his head to the right to avoid diplopia. He has a 10 Δ LH(T) in right gaze and 3 Δ LH(T) in primary and is orthophoric in left gaze. He has 4 Δ LH(T) in upgaze and is orthophoric in downgaze. He is orthophoric on right head tilt and has 6 Δ LH(T) in left tilt. He has a 6 Δ LH(T) with near fixation. His rotations show a – ½ underaction of the RIO, +3 overaction of the LIO versus

pseudo-overaction from RIR restriction, and minimal if any underaction of the RSR.

I can think of only two possibilities here. Either I have unmasked a left fourth nerve palsy in a child who preoperatively had bilateral fourth nerve palsies unbeknownst to me, or, alternatively, he has an anti-elevation syndrome of the right eye due to restriction in the area where I performed the RIO myectomy. I am more familiar with this happening after an inferior oblique anterior transposition than after a simple inferior oblique myectomy.

I plan to do forced ductions, and if I feel the right eye is restricted inferiorly when rotating into elevation, I would probably explore the inferotemporal and inferior quadrants and look for the RIO scarred to the RIR, which I would likely clean up and/or recess the RIR 2–3 mm if there is still restriction after freeing things up. If, on the other hand, there is no initial restriction inferiorly when rotating into elevation, I plan to only perform an LIO myectomy and leave the RIO alone.

I welcome your thoughts on the above, i.e., the etiology, what I am apt to find at surgery, your surgical recommendations including any pearls from your own personal experience in this setting.


### Reply

A lot here points to bilateral masked SOP. The fact there was no compensatory head tilt pre-op despite what appeared to be a classic SOP points toward it being a bilateral problem, and the timing certainly goes along with that. I do not think this is what I call AES, which is different than an inferior restriction [16]. In that syndrome, the anti-elevating force of the operated IO only kicks in on attempted elevation, e.g., recruitment of the IO. There are normal forced ductions. It is caused by the change in the force vector of the IO and cannot happen after a myectomy, except in the highly unusual situation where the proximal cut end of the myectomized IO sticks down anteriorly (that would be unusual). However, a simple adherence syndrome could cause this and would have positive forced ductions. In addition, neither the AES nor an IO adherence would be expected to have a positive head tilt test for a contralateral SOP. In your patient you have a mildly positive test, so that may be of equivocal help in sorting this out. Two are important things to look for: First, is there fundus excyclo OS? If yes, it strongly speaks toward a bilateral masked SOP. Second, look carefully for the lower lid changes that have been described on upgaze of the operated eye [17]. If present, you are dealing the AES or possibly IO adherence. Finally, keep in mind that a simple overcorrection of an SOP can mimic an unmasking of a bilateral problem, as has been detailed by Ellis [18]. He pointed out that if there is an overcorrection, and the initial horizontal incomitance pattern remains after surgery, the three-step test will be positive for an SOP in the contralateral eye.

Do the following:

1. Careful forced ductions. If positive, you have your answer. Release the restriction and possibly do a small RIR recession.
2. If forced ductions are normal, and if there is fundus torsion OS and none of the lid changes OD, weaken the LIO.
3. If forced ductions are normal and there is no fundus torsion OS or lower eyelid changes OD, explore the RIO to see if it is stuck down anteriorly. If so, fix that. If not, recess LIO.

### Pearl

Forced ductions are normal in the AES. A simple overcorrection of an SOP can mimic a bilateral masked problem.

## Case 21.27 Adding to My DVD Collection

### Question

I care for a 7-year-old boy who underwent bilateral LR recession and IO myectomies by another ophthalmologist at 3 years of age for a V-pattern XT. He developed a consecutive ET and at 5 years of age I recessed

his MROU. His horizontal alignment is good but he has a manifest L DVD of 12 Δ and a latent DVD in his right eye of a few PD. He has +2 LIO "OA" in that there is a HYPO of the right eye with cover testing in right gaze, but it is not as large as the LHT. The L DVD is what is bothersome. He has good and equal vision and does alternate fixation. When I operate, I may find that the LIO attached somewhere. I fear that if I reattach it in an anterior position, it will cause problems because some of it has been resected. Should I myectomize it if I find any fibers reattached? Should I do SR OU recess either way and if so should it be symmetric or asymmetric? How much should I recess each SR, knowing that he has had IO myectomies and I will be creating double-elevator palsies? I was thinking I would myectomize any attached LIO I find and do asymmetric SROU recessions of something like 5 OD and 6 or 7 OS.


### Reply

This patient is exactly why I think IO recessions should always be done instead of myectomies. If he had recessions, you could anteriorize the IOs, or at least more predictably address the whole picture. Doing a myectomy closes the door on that option. But here, recessions were not done and you have the scenario you are now dealing with. I completely agree with your final plan. Myectomize any attached LIO, or you could recess it depending on where found, and do about 5 mm on the RSR and 7 on the LSR.

### Pearl

Doing an IO myectomy closes the door on later being able to do an IO AT. Although the majority of patients for whom this may be needed start with infantile esotropia, there are some XT patients and acquired ETs who develop symptomatic DVD and may benefit from IO AT.

## Case 21.28 Seeing Through the Fog

### Question

A patient of mine is a 6-year-old girl who had medial rectus recessions at 6 months of age for infantile ET. She now has good horizontal alignment wearing her +3.00 sph OU, an intermittent manifest R DVD of 15 Δ, and a latent L DVD of 4 Δ. She has equal vision and alternates fixation. Can I just fog her right eye to maintain fixation OS? If so, what do you recommend?


### Reply

I often hear colleagues say that they use ½ D or 1 D of fogging correction to solve problems like this, but it is not that simple. If you fog using a small amount of plus correction, you will be successful at distance. However, that will facilitate fixation with the fogged eye at near where the problem will be more frequently manifest. If you use minus power to fog, it may facilitate fixation at distance with the fogged eye, as children often like seeing with less than full plus. The only way to ensure that you fog at both distance and near is to use more than 3 D of plus or minus, depending on the patient's accommodative facility. If the patient had a significant astigmatism, you could be successful by removing the cylinder from the eye you want to fog. Patients tolerate that well. Oddly, if you try and accomplish the same effect by putting significant cylinder in front of the eye you want to fog, when there is no astigmatic error, patients often complain of asthenopia.

### Pearl

A small amount of plus or minus power to fog the eye for the purpose of switching fixation is usually not adequate, because it will facilitate fixation at either distance or near while inhibiting it at the other viewing distance.

## Case 21.29 An Oblique View

### Question

In a young patient with a partly accommodative ET who has unilateral or very asymmetric IO "OA" with a V-pattern, but no HT in primary gaze, I have been reluctant to do a unilateral IO recess for fear of inducing a HYPO in the operated eye post-op. I've also been reluctant to do bilateral IO surgery if the less affected eye shows no or minimal IO overaction. How should I handle these cases?


### Reply

Studies show that in a large number of these patients the unoperated IO becomes clinically overactive if you do unilateral surgery. If there is no HT in primary now, but definitely some contralateral IO "OA," weakening both IOs is the way to go. The effect on versions seems self-adjusting—you get a lot in the eye you need a lot, and less where you need less. The only time I do something asymmetrically in these bilateral asymmetric IO "OA" cases is if there is a HT in primary. Then I do asymmetric IO recessions.

### Pearl

In cases of bilateral asymmetric IO "OA," symmetric weakening surgery is self-adjusting.

## Case 21.30 High Eye in the Big Sky (Case from Montana)

### Question

This is a case of an older gentleman with a left HT for many years. He had an inadequate amount of prism in his glasses and essentially tolerated diplopia much of the time. Previous attempts by his optometrist to use additional prism were reportedly difficult for him to adjust to. His LHT is greatest in primary gaze (11 Δ). No matter how much prism or monocular covering I do, I cannot get him past 9 Δ LHT in right or left gaze. He also measures 9–10 Δ in up- and downgazes. When I put prism in front of the eye to help him fuse, he tolerates less in side and downgazes than he does in primary. I keep thinking he's just masking a larger HT somewhere other than primary, but I can't bring it out, and am concerned that if I correct the entire HT in primary, he will be overcorrected elsewhere. Added to that, his double-Maddox rod findings show 6° of excyclotropia. It is apparently long-standing enough that it doesn't prevent him from fusing in clinic when he has the proper amount of prism. I've already forewarned him that prism, a lesser amount than he currently needs, will probably still be needed post-op, but am wondering what the best muscle is to operate on. Recessing the RIR will, I think, likely overcorrect him in reading gaze where he currently is able to intermittently fuse with his smaller amount of prism. Recessing the LSR is probably what I'll do but it will, of course, worsen the excyclo. I want to do an LIO recession but am quite sure that he would have an overcorrection in right gaze afterwards. LIR resection seems to have the same problems as LSR recession. Do you have any other thoughts?


### Reply

You are concerned that an RIR recession will overcorrect him in downgaze. But he measures 11 in primary and 9–10 down. So undercorrecting primary deviation a touch should be a good outcome for him. I would do that. I understand the temptation to do the RSR, but I too would worry about the 6° excyclotropia. Six is an upper limit of what people can fuse, and if you worsen that, he may have trouble. If you want to do the RSR, I would combine it with an anterior disinsertion of the RIO to minimize the torsion (see Fig. 13.8).

### Pearl

When deciding what to do when incomitance raises concerns that correction in primary gaze may overcorrect in an important eccentric gaze, consider mildly undercorrecting primary.

## Case 21.31 A Divergent Path

### Question

Help! I operated on a 30-year-old musician who had a symptomatic R DVD for years. She is plano OU and 20/20. She had eye strain and discomfort for reading or when following the conductor. She preferred L eye fixation and manifested an intermittent R DVD of 7 Δ, and a trivial ET, but absolutely no DVD in the left eye despite prolonged and slow cover testing. I did an RSR recession of 7 mm and did not operate on the left because I could not see any DVD. After surgery she manifests 4 of R HYPO when fixing with the left eye, but an L DVD of 7 Δ. If the R HYPO is neutralized with 4 Δ base up, the total L deviation is 10 Δ of HT (sum of DVD and true). There is also a slight limitation of elevation OD. I have always done bilateral surgery if the patient had a bilateral DVD and the potential to shift fixation after surgery (not amblyopic). What should I do next?


### Reply

Years ago a very prominent colleague reported results of unilateral SR recession for patients of this type, and in the article reported a negligible overcorrection rate—close to zero. I wrote him for clarification as to what is meant by overcorrection. Did he mean a HYPO of the operated eye, or did he consider the other eye going up an overcorrection. He replied that he meant the former. When I then asked how many acted as your patient did with the other eye manifesting a DVD, he replied between 30 and 50%. It was interesting that the article did not mention that observation, although it was apparently known to the author.

Deciding what to do with a patient like this for the first round is a matter of trade-offs. More than one-half or close to two-thirds of patients will do fine with unilateral surgery, but a large number (one-third to one-half) will reverse fixation and need an enhancement. My approach is to always operate on both eyes if I feel there is potential to shift fixation, which I arbitrarily cut off at about 20/40 in the nondominant eye. When I do bilateral DVD surgery, I up the dose for the recession, as the surgery done in the fixing eye tends to negate that done in the other eye. But if I do unilateral surgery, I use a more modest table. For this patient's initial deviation of 7 Δ of DVD, a 7 mm unilateral SR recession would be a bit excessive, and not so, if done bilaterally. Here you have an overcorrection, albeit small, since there is a manifest HYPO of the operated eye. That is uncommon with DVD surgery. There is also a modest limitation of upgaze. So the dilemma is whether to advance the RSR or recession the LSR. I think, given the size of the new L DVD, I would do the LSR, as you would have to advance the R a fair amount to treat the L DVD, and that could get you back to where you started. Finally, I must comment that 7 Δ of DVD is barely noticeable cosmetically. I agree that some patients with DVD do get asthenopia and related symptoms when the DVD becomes manifest. You must be very careful to be certain that the DVD is the cause of the symptoms.

### Pearl

Of patients with unilateral DVD who undergo unilateral surgery, between one-third and one-half of them will manifest a DVD of the unoperated eye postoperatively if they have approximately equal vision and the potential to shift fixation.

### Pearl

DVD is not just cosmetic. Some patients with DVD have asthenopia from trying to control the DVD.

## Case 21.32 Doubling Down

### Question

A patient of mine is a 34-year-old professional violinist who had a traumatic L SOP for which I did a small LSO tuck (6 mm total—3 mm on each side of the tuck). I chose an SO tuck because he had minimal LIO "OA" and the deviation was greatest in the LSO field. After surgery he is perfect in primary and downgaze, but has an iatrogenic Brown syndrome with −3 to elevation in adduction with a 12 Δ L HYPO in upgaze with diplopia. He has been stable for 8 months (the tuck has not loosened,) and he is very bothered by this, as he needs to have his eyes in upgaze to read music while playing the violin. I thought a posterior fixation of the RSR would help, but he has hardly any range of movement of the left eye into upgaze, so the PF probably would not increase his range of single binocular vision enough. I have heard some experts address this problem by taking the tuck down and not doing anything else. In one of these patients, this corrected the Brown syndrome and somehow the alignment in other gazes remained good. Have you done that? Do you have any experience with this situation or recommendations?


### Reply

I understand the dilemma. This is complicated by the fact that he is good in primary. I agree that you have to try to take down the tuck. I have seen that work. The problem is that tucks cannot always be "taken down." Sometimes both sides of the tuck fuse together, and you cannot neatly separate them. I am suspicious you may find that in your patient, as it is odd to get a persistent Brown after such a small tuck, unless there is odd scarring. But you should try. Still you need a plan B. If you cannot take down the tuck, you could try a small SO weakening nasally: either a tenotomy with a short chicken suture, a very small expander, or very small split tendon lengthening (my preferred option—whichever weakening procedure you are most comfortable with and accustomed to doing). Another option might be to take the whole tucked SO complex and do a small recession from the insertion. For all these forced ductions will have to guide you as to how much to do. None of these surgical procedures are easy, but I think you need to address the SO.

### Pearl

SO tucks cannot always be taken down. When planning to do so, you need a plan B.

## Case 21.33 Tuck It?

### Question

I did an LIO recession on a 24-year-old man for a LSO palsy. One year after surgery he has a symptomatic residual deviation with 8° of L excyclotropia, 2 Δ of LHT in primary, but 12 Δ in downgaze, and a bit more in downgaze in adduction. I am considering a LSO tuck. Do you agree?


### Reply

Although a LSO tuck will address the incomitance pattern, the HT, and the torsion, I have concern that it will overcorrect the primary position deviation of only 2 Δ. I expect even a small tuck would cause 6–8 Δ of vertical change in primary. A better choice would be to do a Harada-Ito in the left eye, and a small RIR recession or posterior fixation.

### Pearl

Although surgery should be tailored to address the incomitance pattern, the size of the deviation in primary is of paramount importance.

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