A Patient & Parent Guide to Strabismus Surgery

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Part I:

Background Information

Chapter 1:

Basic Anatomy and Actions of the Extra-ocular Muscles

The muscles that move the eye are called the extra-ocular muscles. There are six of them on each eye. They work together in pairs—complementary (or yoke) muscles pulling the eyes in the same direction(s), and opposites (or antagonists) pulling the eyes in opposite directions. Below are some basic movements for each of the pairs.

Horizontal Rectus Muscles
The medial rectus, or nose-side, muscles move the eyes inwardly; when working simultaneously, they converge, or cross, the eyes. The lateral rectus, or temple-side, muscles move the eyes outwardly; when they work simultaneously, they diverge, or splay apart, the eyes. When moving the eyes from side to side—their principal function, they work in teams, so that the normal movements are smooth and coordinated. For example, to gaze to the right, the nose side (medial rectus) muscle of the left eye, and the outside (lateral rectus) muscle of the right contract; their opposites (or antagonists)—the left lateral and right medial recti—relax.

Vertical Rectus Muscles
The vertical rectus muscles—superior moving the eyes upwardly, and inferior, moving them downwardly—are teamed in a similar fashion to the above-described horizontal recti. Because of the angle of the muscles on the eye, these muscles also have an effect on horizontal and torsion (or rotation) movements of the eyes. These secondary and tertiary actions are generally only of importance in complicated strabismus.

Oblique Muscles
The most complicated muscles—both in anatomy and action—are the oblique muscles, superior and inferior. These muscles, like the rectus muscles, also work in antagonist pairs; the inferiors contract while the superiors relax, and vice versa. The inferior oblique muscles turn the eyes upwardly when the eye is looking inwardly, and rotates outwardly (extorts) the eye when looking outwardly. The superior oblique muscle turns the eye downwardly when the eye is turned inwardly, and rotates the eye inwardly when the eye is directed outwardly.

Cranial Nerves Innervate The Extraocular Muscles
The muscles move in response to nerve impulses carried from the base of the brain (brain stem) by three cranial nerves, named the oculomotor (or III nerve), the trochlear (or IV nerve) nerve, and the abducens (or VI nerve). The trochlear nerve enervates the superior

oblique muscle, the abducens nerve enervates the lateral rectus muscle, and the oculomotor nerve enervates the remaining four, plus the levator (or lifting) muscle of the upper eye lid.

Chapter 2:

What is Strabismus?

Strabismus refers to eyes that are out of alignment. The eyes maybe converged (crossed), diverged (outwardly deviated), vertically (one eye higher than the other) or torsionally misaligned (one or both eyes rotated inwardly or outwardly). These planes of alignment (and misalignment) are like the types of movement of boats and airplanes—sometimes called yaw, pitch and roll.

- Crossing (or deviation of one or both eyes toward the nose) is called **esotropia**.
- Outward (toward the ear) deviation of one or both eyes is called **exotropia**.
- Vertical deviation or divergence of the eyes is called **hypertropia** (higher) or **hypotropia** (lower).
- Torsional (rotational or tilting) misalignment is called **cyclotropia**.

Misalignment of the eyes may be constantly manifest—called a tropia—such as above, esotropia, esotropia, etc. Or the deviation may be intermittent, called intermittent esotropia, etc. Surgery may be appropriate for either constant or intermittent deviations, depending on a number of factors, including magnitude of the deviation, constancy or frequency of the misalignment, and the presence of other signs and symptoms.

One relatively simple way to think about strabismus is the concept of the **position of rest** of the eyes. Therefore, the problem of strabismus is not necessarily that of an abnormal eye. The problem is the angle of deviation between the two. Consequently, the eye that "drifts" is not necessarily abnormal; quite simply, the dominant or preferred eye is "straight"—that is, directed (pointed) toward the object being viewed—while the other assumes the position of rest. The importance of this concept is to explain that to correct the problem requires eliminating this angle of deviation between the eyes. For treatment, muscles on one or both eyes may be repositioned to eliminate or diminish this angle and eliminate the strabismus.

Chapter 3:

What Causes Strabismus?

There are many medical conditions that are associated with strabismus, and a few of these associations are mentioned in this communication. In general, however, it can be said that the "causes" fall into the following categories: hereditary influences, neurological problems, selected medical conditions, and by far the largest category, unknown. The prevalence of strabismus in the general population is about 4 %. Please remember, not all strabismus requires treatment and a minority of strabismus requires surgery.

Hereditary Influences
Certain genetic syndromes—Down's Syndrome being the most common—have a high incidence of strabismus. In some of these conditions, it may be so characteristic as to be expected in most cases. When there is a family history of strabismus (without a known genetic syndrome), with or without amblyopia, the prevalence in existing and subsequent family members can be as high as 25%. Please note two aspects about this number: the overall chances are actually against a child having strabismus if parents have it; and importantly, ***statistics only have meaning for populations and not individuals***. And the hereditary patterns of strabismus are quite variable. In some families and for certain conditions, only a few are affected; sometimes called sporadic or recessive with low genetic expression (penetrance). In others, many family members are affected; and the pattern is more dominant, and/or with higher penetrance of genetic expression.

Neurological Problems
Neurological problems encompass a diverse group of conditions that includes developmental problems and delays such as prematurity, cerebral palsy, and head trauma. Depending on the condition, the incidence of strabismus may be as high as 50% of cases. Please be sure to share with your doctor any concerns you may have about your child's or your condition that may relate to neurological and developmental issues. Please make particular note of any instances of head trauma or fractures of head and face bones, even if in the distant past. There are many medical conditions that may affect the alignment of the eyes. Examples include:

- thyroid disease (also called Grave's disease),
- myasthenia gravis,
- circulatory problems (including stroke)
- and diabetes.

Again, please share all you know about your medical condition and its treatment, including any allergies you have. Please include allergies to foods, medicines and other substances, such as latex. Please make special note of any problems with anesthesia experienced by you ore related family members.

Unknown

Finally, strabismus is, in most cases, unrelated to any of the above. It just happens to some and not others. Therefore, most of the time, there is no known cause or association with other medical conditions. While the prevalence is low—less than 4 per cent—the population is large, making this by far the largest category.

Chapter 4:

What are the Signs and Symptoms of Strabismus?

Signs of strabismus are those aspects that may be observed by the affected individual, parents or others. These include the misalignment itself (crossing, drifting, etc.), squinting of one eye closed, sometimes rubbing of one or both eyes, and a compensatory head posture.

Symptoms are the feelings or subjective observations of the affected individual. There may be no symptoms whatsoever, especially in young children or in persons whose strabismus is long standing. If there are symptoms, these may include double vision (diplopia, or two images seen for one object) or "split" vision (like seeing 1+1/2 images), unstable images, eyestrain or fatigue, headache and an awareness that an eye is moving about; it may feel as if one is "crossing" the eyes, yet the preferred eye feels fine. Importantly, there can be other sensations that are unpleasant relating to the affected persons sensitivity to their condition. These include the awareness that they are different and that others treat them differently as a consequence. This may affect one's self-image and confidence. Some experience difficulties in a variety of areas including activities of daily living, such as reading and driving; work-related activities, including effectiveness, hiring and advancement; social interactions, including ability to communicate; and personal relationships and interactions, including the ability to maintain eye contact that may lead to embarrassment.

In certain forms of strabismus, it is possible to control deviation of the eyes by positioning the head, called a compensatory head posture. The head may be turned from side to side, chin up or down, or tilted to right or left. Such head postures may be also effected to control nystagmus (shaking or dancing eyes), or to compensate for large refractive errors (that is, the need for eye glasses). Long standing abnormal head positions may lead to arthritic and other changes in the bones and muscles of the neck and spine.

Only the persons affected and/or their families, in consultation with their doctor(s), can determine the degree to which such signs and symptoms are sufficient to consider strabismus surgery. Doctors can help with information, the perspectives of existing knowledge, experience and then provide recommendations. The remainder of this booklet will provide additional information about the experience of strabismus surgery, so that children and adults with strabismus—and their families—will have additional information to make decisions about care.

Chapter 5:

Why is Strabismus Surgery Performed?

General Comments About Strabismus Treatment

Many forms of strabismus can be managed with non-surgical treatment, including eyeglasses, prisms in eyeglasses, patching, and in certain circumstances, exercises. In general, each of these approaches has limitations. Eyeglasses may completely control strabismus, as in accommodative esotropia. In other cases, eye glasses may have no effect at all. Prisms are useful for relatively small, stable angles of deviation (strabismus); yet they may need to be made progressively stronger, are expensive, and there are practical limits to the power that may be applied (strong prisms turn light into rainbows!). In children, patching of a dominant eye to improve vision will sometimes, in conjunction with other treatment such as eyeglasses, facilitate improved eye alignment. In certain forms of early childhood strabismus, patching will improve alignment, although sometimes only for a period of time. Exercises may completely control some forms of strabismus and have no effect on others. Your doctor will try all appropriate non-surgical methods of treatment prior to recommending surgery. And many mild forms of strabismus—those with no or very mild symptoms—may require no treatment at all. Overall, only about 25% of strabismus warrants surgery.

Some forms of strabismus require surgery, and non-surgical methods may only temporize and delay a recommendation for surgery. In other cases, non-surgical means may work for a period of time (even for several years), and then cease to be effective. Generally, deviations of a significant degree (usually 5 degrees of deviation or more, with or without symptomes) and smaller deviations (where symptoms cannot be controlled otherwise) are appropriate to consider surgery. Of course, all factors that are appropriate to a given individual's needs should be considered in the context of the patient's desires and best interests. A decision to proceed with surgery should make sense to all parties to the decision!

Indications for Surgery

Surgery is recommended when strabismus and its effects are "clinically significant", meaning the angle of deviation is large enough, the condition is amenable, and the adaptations or consequences significant enough to promise improvements in alignment and function. These potential benefits are correlated to the particulars of a given person's circumstances, be they a child or adult.

A person's visual system develops and functions in a complex interplay between and among the vision in each eye, alignment of the eyes, the field of vision, fusion of the images from each eye, plus the affected person's experience and perception of strabismus and its consequences. Early visual development occurs rapidly, and when strabismus occurs, adaptations—including decreased vision (amblyopia) and loss of binocular

function (stereopsis or depth perception)—occur equally swiftly. The keys to successful treatment are: prevention if possible, early detection and prompt treatment.

In some individuals, strabismus can be overcome with a compensatory head posture, such as tilting or turning the head. In some of these cases, there may be long-term adverse consequences in the bones and muscles of the head and neck (please see following sections and appendix for examples of when surgery may be appropriate to align the head position).

In adults, the conditions either are "held over" from childhood strabismus, or acquired later in life, generally spoken of as onset "after visual maturation" (generally after nine years of age). In either children or adults, the benefits of strabismus treatment may be multiple, depending on the type, severity and individual effects of the condition. Treatment is therefore promptly directed to:

- ***Improve Vision***—The earliest adaptation to strabismus in a child is often the development of amblyopia, or decreased vision secondary to suppression of the image from the misaligned eye. Glasses and patching are often required to treat amblyopia. Elimination of the misalignment (strabismus) often will make the task of visual rehabilitation easier. Amblyopia does not occur in adults.
- ***Align the Eyes***—Good alignment facilitates both good vision and binocular vision. As above, eyes that are out of alignment present a significant risk to the vision of young children and their developing sight. Moreover, misalignment precludes the development or maintenance of good binocular vision, including stereovision; persistent misalignment, for as short as three continuous months, may also cause adults to lose (irretrievably) their stereopsis (three dimensional vision). Establishing or reestablishing good alignment can improve binocular visual outcomes; in certain circumstances (not always predictable) binocular vision is completely normalized.
- ***Align the Head***—Abnormal or compensatory head positions may occur with certain strabismus syndromes (for example, Duane's syndrome, IV cranial nerve palsy, thyroid eye disease) or nystagmus. The long-term consequences may be orthopedic (arthritis, etc.), appearance (asymmetry of the face), social challenges (e.g., teasing), or practical (e.g., wearing glasses effectively). Such problems may be avoided by straightening the head—by moving the eye muscles—and in some instances of nystagmus, vision may be slightly improved.
- ***Improve Binocular Vision***—Binocular vision refers to simultaneous teaming of the eyes and includes the functions of stereovision and depth perception. These functions occur in the brain; having good alignment is necessary (although not necessarily sufficient) for improving the quality of binocular vision. In general, the younger the child, both the risk and the opportunity for binocularity are increased.
- ***Eliminate Diplopia When Present***—Diplopia, or double vision, occurs when eyes have previously been aligned and good binocular function obtained. When the eyes become subsequently misaligned, double vision occurs. This is relatively

uncommon in very young children, but may be present in older children. It is common in acquired strabismus in adults.

- ***Improve the Field of Vision***—Crossed eyes, technically called esotropia, will diminish the total field of vision by decreasing side or temporal vision in amounts that are directly related to the degree or amount of crossing of the eyes. Elimination of the misalignment will therefore increase the field of vision for these persons (and will decrease in exotropia).
- ***Improve Self Image***—Persons with misaligned eyes of almost any age above three to four years will develop a sense of difference related to the significance of their strabismus. This awareness will often affect a child's self-concept (image) and confidence, and may be reflected in shyness or withdrawal.
- ***Improve Social Interaction***—Strabismus may affect the ways in which a family, friends and others will interact with a child, and thereby affect all of their relationships. Older children and adults may experience difficulty in communication related to difficulties in maintaining eye contact. Others may be distracted and behave differently towards persons with strabismus, and this may be hurtful to them.
- ***Improve Employment Opportunities***—Children with strabismus may experience, perhaps in part related to the above, some perceived and actual limits to career options and advancement. Strabismus is a disqualifier for certain occupations. Other persons will occasionally and quite inappropriately question the intellectual capacity of those with strabismus.

Strabismus at any age has multiple and complex effects. The sooner strabismus and its associated problems are identified, the longer and more profoundly the benefits may be enjoyed!

In Appendix A., there are expanded discussions of selected specific conditions in strabismus. You may wish to read the portion(s) that apply to your or your child's diagnosis.

Part II:

Making a Decision About Surgery

Chapter 6.

What are the Options in Strabismus Treatment?

As with any non-life threatening condition, the options range from no treatment—through optical, prismatic, certain medical, and occasional exercise therapies—to surgery. In leading up to a recommendation for surgery, you and your doctor have either tried or excluded for lack of benefit, non-surgical options. Hence, the choice will likely narrow to continuing with the condition and its current therapy (if any) or eye muscle surgery. Unfortunately, the only party that can make this decision is the person who bears the condition or that person's family, particularly parents and caregivers.

Your doctor will give you his or her best recommendation, and you should feel comfortable that you understand the risks, potential benefits, limitations and alternatives to surgery. If you do not have this comfort, you should discuss the matter with your doctor or seek a second (or more) opinion(s) from additional specialists. Your doctor should be willing and able to help you seek additional opinions about you or your child's care.

Chapter 7:

The Preoperative Consultation

Prior to recommending and performing surgery, all appropriate evaluations should be completed. First and foremost, the measurement of the deviation of the eyes must be reliable and consistent with a known or working diagnosis. Sometimes these measurements may require more than one evaluation to assure consistency of the presence and degree of the strabismus. Second, the sensory status—how the eyes see and work together—of the eye should be evaluated as possible, and any problems with vision addressed. In particular, refractive errors requiring eyeglasses and patching for decreased vision (amblyopia) should be initiated or complete. Third, the general medical status should be known and stable, to assure that there will be little to no risk to anesthesia. Finally, your doctor will discuss with you the potential risks, the benefits to be derived, the limitations of surgical correction of strabismus and the alternatives to surgery and its timing. Importantly, you should have a clear picture of the problem, how it currently affects (and may change in the future) your or your child's life, what is being proposed, what the experience is likely to be, and what the most likely outcomes will be.

Chapter 8:

Choosing Your Surgeon

To the extent possible, you should feel comfortable about the surgeon you have selected. There are some ways in which your choices may be limited, and over which you have little to no control. Your health plan may restrict whom you may see with your current coverage. You may be isolated geographically, which limits your practical choices. Or you may have limited other resources that place limits on seeking additional opinions or options. Only you can determine the correct course of action for yourself and your child. The following may help you in this effort.

There is general agreement in medicine that a good surgeon is more than steady hands, as important as that is. Good surgeons also have: intelligent minds, honed with continuing study and inquiry; good hearts, taking the best interests of their patients as their habit of practice; courage and coolness under fire, to do what needs to be done particularly in difficult or unexpected circumstances; and experience, which leads to good judgment and the ability to avoid complications whenever possible. Good surgeons tend to operate expeditiously, not because they hurry, but because they do what needs to be done and nothing else (meaning no unnecessary or "complicating" moves). Strabismus surgery is technically demanding, and preoperative testing and surgical plan creation is challenging. Nonetheless, in the hands of skilled and experienced surgeons, it can (and for most cases should) be routine.

If you need advice about a surgeon to entrust you or your child's care, the most informed sources include the people with whom they work professionally. These include referring physicians, operating room nurses, anesthesiologists and other strabismus surgeons. The latter will likely be somewhat cautious in their appraisals, not for lack of knowledge but for concerns about the propriety of commenting. Health plans, despite their "credentialing" process, are not likely to have included physicians on their plans based primarily on their clinical skills. Patients who have had care or surgery provided can be a valuable source of information about how they were treated and give important information about what they have learned from their experiences, broadly speaking. General information about a surgeon's credentials can be gleaned from reference sources, and can be of some benefit. The Internet has a wealth of information, yet it may be difficult to interpret it in light of the specific context of an individual's care. An experienced strabismus surgeon in whom you have confidence is your best source of perspective as to what should be done and when. Remember: if in doubt, it is reasonable to seek a second opinion.

Chapter 9:

Risks, Benefits, Limitations and Alternatives to Surgery

Risks:

The most common risk to strabismus surgery is partial or complete failure to correct the condition and/or its effects, including associated symptoms such as double vision. In general, the more complicated the strabismus and its associated conditions, the more difficult it is to completely control the deviation and its effects. Because the visual system is complex, involving much of the brain, repositioning the extraocular muscles cannot be expected to resolve all problems associated with strabismus. In particular, the potential for binocular teaming of the eyes is known to have a significant effect; that is, if binocular vision can be achieved or recaptured, the likelihood of additional surgery diminishes. Decisions as to what and how much to do are based on experience—that experience is on a large number of patients, the effects and benefits of which cannot be known looking forward. How a given patient will respond to the experience of those before is unknowable; and there is a statistical bell shaped curve of effects for the population. In essence, therefore, some patients will be overcorrected, and some under corrected. The net effect is that additional surgery is required in a significant number of cases; the likelihood of more than one surgery, depending on a number of factors, can range from as low as 10% to 50% or more, with an average of between 20 and 25 percent. Most other complications, including anesthetic problems, infection and potential loss of vision are very rare. Working together, the patient, family, doctors and nurses can, and will take steps to assure that these potential complications are minimized. For example, post-operative antibiotics taken by mouth are often used to prevent infection, and careful attention to preoperative instructions, meticulous operating room procedures and prompt notification of any problems will be likely to prevent adverse consequences to these rare events.

Benefits:

The benefits of surgery are aligned with the goals of surgery, and may vary from person to person. Alignment of the eyes may make amblyopia therapy less intense. Proper alignment may eliminate a range of symptoms such as double vision, eye strain or fatigue, and restore the normal relationships between the eyes themselves and other facial structures. Only the patient and his family, with the advice of their doctor can determine if these benefits warrant undergoing strabismus surgery.

Limitations:

Because of the complexity of the system, strabismus surgery may solve only a portion of a complex problem. Some forms of strabismus respond better than others. And some problems are not amenable to surgery. Your doctor will be able to explain the application of these generalities to your condition.

Alternatives:

Non-surgical options in the treatment of strabismus include patching (occlusion) prisms, botulinum toxin injection, monocular occlusion or fogging, and in some cases eye exercises.

Occlusion of one eye (or patching) may be useful in selected circumstances. Its principal use is in the treatment of amblyopia, or decreased vision associated with strabismus or other conditions. It is also somewhat useful in children with intermittent outward deviation of the eyes in infancy (typically under two years of age), called intermittent exotropia.

The use of prisms in adult strabismus is commonly applied. Indications for use are broad and flexible based on the individual patient's circumstances. Small angle strabismus with diplopia is the most common condition where prisms are effective. For example, a patient with a vertical deviation of five to six diopters (the unit of measurement of angles of deviation; one diopter equals about ½ degree of angle), which is comitant (meaning the same in all directions of gaze), would certainly be a candidate for prisms. Prisms may be useful in those patients that show an early over correction following strabismus surgery, and may be effective in helping to maintain good binocularity. Prisms have also been used in helping the surgeon decide how much surgery to do. The Prism Adaptation Trial showed some effectiveness in the preoperative evaluation in esotropia. Those patients who responded to the prism (binocularly) and whose angle increased showed a greater surgical success. However, it should be mentioned that prisms are not without disadvantages. Primarily, prisms are limited by the fact that it is impractical to correct large deviations due to the thickness and weight of the prism. In addition, those patients whose deviation is incomitant or changes from one gaze position to another may continue to manifest diplopia. In addition, patients who normally do not wear glasses may find difficulty in adjusting to glasses with prisms. When using prisms, there are two options. Ground in (to eye glass lenses) prisms are useful when the patient will be wearing the prisms for an extended period of time. Fresnel or stick-on prisms are useful when the prisms will be temporary, although many note degradation in visual acuity with and the unusual appearance of Fresnel prisms.

Botulinum toxin has gained significant popular appeal particularly with its cosmetic indications such as removing unwanted wrinkles. Interestingly, botulinum toxin's role in medical therapy was discovered by a strabismus surgeon 25 years ago in California. Botulinum toxin is one of the most deadly toxins known to man. However, in microscopic quantities it is effective in temporarily weakening specific muscles for six to twelve weeks. The potential advantages of this option, most commonly used in adults, are: 1) it can be injected in the office without requiring general anesthesia; 2) it can be employed as a temporary treatment, e.g., in patients with acute cranial nerve palsies, and this option may be highly effective in the short term while the surgeon waits to see if the palsy will resolve; and 3) this may be an effective treatment in patients who show early over corrections following strabismus surgery. The chief disadvantage of botulinum toxin is that its effect maybe variable and unpredictable. In addition, it is not useful as a

permanent form of treatment as its effect wears off after one to two months. In addition, risks include ptosis which maybe very troubling for the patient.

In patients who manifest intractable diplopia or who are poor surgical candidates occlusion or fogging may be an alternative treatment to avoid diplopia. In some patients who suffer mid-brain injury or disease, motor fusion may be permanently impaired. These individuals may never be able to fuse normally. Patching one eye or fogging one lens with a filter may be the only treatment that relieves them of their diplopic symptoms.

The use of eye exercises in strabismus is only occasionally beneficial in the long term. The best example of their potential usefulness is convergence exercises in persons with convergence insufficiency—a form of exodeviation where there is a tendency of the eyes to drift outwardly apart for near visual tasks. However, exercises may be useful: 1) for selected individuals and mild conditions; 2) when signs and symptoms are mild; 3) when affected persons are motivated, and 4) when expected benefits may be sustained through time.

The Informed Consent Process and Document(s)

Completing the documentation of your informed consent for strabismus surgery is dual—medical information combined with certain legal requirements. You have both legal and moral rights to know what the diagnosis for your condition is, what treatment is being proposed (including surgery), and what are the most likely risks, benefits, limitations and alternatives to the treatment proposed. This booklet, along with your discussions with your doctor, can help with these elements.

The documents that you will be required to sign, consenting to the surgery, are legal documents. They are constructed to fully inform you of the worst thing(s) that could happen, and sometimes the effect is to add to your anxiety about the procedure. While such added anxiety is unfortunate, there is no option to their required use. Please read them with this understanding in mind. Do not consent to the surgery if you have unanswered questions or concerns. Surgery is best deferred until you have the information you need to make an informed decision about your or your child's care.

Please also see Chapters 15 & 16, which describe what to expect and watch for after surgery. These matters should be considered in the context of the risks and consequences of surgery, as well as understanding what may be of concern.

Chapter 10:

How is Strabismus Surgery Performed?

What is done in strabismus surgery should make sense to you, and reasonably address the condition as you understand it. For example, if an eye is turned inward, one can logically conceive that the muscles that pull it inward are either overacting or too tight. Therefore one should do something to weaken the pulling power, or loosen, that muscle. There are a number of methods to weaken a muscle's pulling power, including moving it (generally called a recession) or, in certain instances, severing all or a portion of it. Strengthening a muscle's action may be performed by tightening it (generally called a resection) or by "borrowing" muscle power from and adjacent muscle. Transposing all or portions of muscles may be of benefit when there are certain patterns to strabismus, or when a muscle is absent or neurologically weakened. The following is a summary of commonly performed operations for some common conditions. Be aware that there is more than one method for effectively treating strabismus surgically; and your or your child's condition may warrant something varying from these examples.

For esotropia:

Recession of the medial rectus muscle of each eye

Recession of one medial rectus and resection of one lateral rectus

For very large angles, combinations of the above

For exotropia:

For small angles of deviation, recession of one lateral rectus muscle
Recession of the lateral rectus muscle of each eye
Recession of one lateral rectus muscle and resection of one medial rectus muscle
For very large angles, combinations of the above

For hypertropia:

Recession of one or more vertical rectus (superior and inferior) muscle(s)
Weakening of one or more oblique (superior and inferior) muscle(s)

For inferior obliques, common operations include recession and myectomy

For superior obliques, common operations include tenotomy, tenectomy, lengthening with spacer(s), and recession

Particular circumstances may require:

Transposition of a muscle and its insertion
Detaching a muscle
An assistant or co-surgeon

Additional Methods for Consideration

Exercises: Eye exercises, sometimes called orthoptics or vision training, have proven to be of some value in strabismus care. In some instances, exercises may be used as a temporizing or preparatory strategy prior to surgery. However, when the amount of the strabismus is substantial, the deviation is not likely to be overcome with exercises.

Botulinum toxin: Botulinum toxin may sometimes be used as part of or an adjunct to surgery. For additional perspectives, please see the section on alternatives to surgery.

Adjustable sutures: Some surgeons will use adjustable suture techniques in adults and older children routinely, some in selected circumstances, and others rarely if at all. Reasonable and experienced surgeons continue to debate the relative merits of these procedures. The potential merits seem intuitive, i.e., if the correction is not proper, it may be adjusted immediately after the surgery, hopefully to avoid additional procedures.

There remain unanswered questions. The adjustment is generally accompanied by significant discomfort; there may be some imprecision in where the muscle actually reattaches to the globe, and some changes in eye position continue to occur during the healing process. Hence, it is not definitively known the degree to which (and in what ways) adjustments may be of benefit, or the necessary and sufficient criteria for their application. This dilemma would benefit from a prospective controlled trial of comparison in patients with similar conditions performed by the same surgeons. Your surgeon will share with you her or his recommended choice of technique.

Chapter 11:

Timing of Surgery

Strabismus surgery is rarely an emergency procedure. Accordingly, there is time, and the time should be taken to fully evaluate the condition. On the other hand, it should be remembered that strabismus rarely goes away spontaneously. If anything, there is a general tendency for strabismus to worsen with time—in degree and/or frequency of deviation of the eyes, as well as complications such as loss of vision and binocular capacity. Furthermore, in children, there is a general principle that the younger the child, the more flexible and adaptable is their neurology, including the entire visual system. This can work for both good and bad. The following general approach is therefore recommended. Once the condition has been fully evaluated, the indication for surgery is established and the choice to proceed made, there is no particular advantage to waiting. In certain circumstances, waiting (or delay) may decrease the likelihood of a positive outcome. A second advantage to proceeding with surgery when warranted in children is that older children often worry more and are more anxious about the surgical experience.

Part III:

What to Expect Around the Time of Surgery

Chapter 12:

Before Surgery

Preparing for Surgery

Administrative and insurance approvals

Most strabismus is covered by health insurance. Some policies have restrictions on so-called "pre-existing" conditions, who may perform your surgery, where it may be performed, what will be "covered", copayments and deductibles, what will be paid and to whom, and other limitations. The insurance company or health plan, as part of the contract between and among employers, beneficiaries and others, generally imposes these restrictions. Similar restrictions are placed on providers, including surgeons under contract to the insurance company or health plan. Those contracts require all parties to "play by the rules". There are often significant frustrations created by these rules and rulings of the managed care or insurance carriers. While your surgeon's staff will assist you in knowing how your policy(ies) will affect reimbursement for your care, the ultimate responsibility for understanding your policy, and what it will provide, is yours. Your surgeon's office responsibility is to provide you, and where contractually required, your insurance or managed care company with information about your diagnosis and proposed treatment. A fact of life is that these matters are often complicated and take time and resources to sort out. You may rest assured that any frustration you may experience is at least matched by your surgeon and his or her staff.

Scheduling

Once it has been determined what options are available, scheduling is a matter of matching your (or your child's) medical needs with an appropriate facility at which your surgeon has obtained privileges and the combined schedules of the necessary parties. Apart from acute trauma, timing of most strabismus surgery is generally not medically critical. Nonetheless, once the decision has been made, it is generally advantageous—gaining the benefits of correction and avoiding long term fretting about surgery—to you or your child to proceed at an early convenient time.

Medical clearance

It is reasonable and prudent to gain prior advice from doctors that provide ongoing care—such as pediatricians, internists and family practitioners—prior to proceeding with surgery. This will maximize the safety to you or your child through communication. There are also general guidelines about what preoperative testing, such a blood and urine tests, chest X-rays, and EKG that may be important to your anesthesiologist and health facility in determining the advisability and safest method of

administering an anesthetic and performing surgery. Please be certain share any ongoing medical problems and issues with your surgeon and anesthesiologist prior to surgery. In particular, inform them about any allergies you may have to medicines, any bleeding tendencies you may have, and any past or family history of problems with anesthesia.

Since strabismus surgery is almost always elective in the sense of timing, every effort is taken to avoid taking unnecessary risk. Therefore, if a person is experiencing a temporary illness—for example, upper respiratory congestion, significant cough/sore throat, vomiting and/or diarrhea, and especially fever—it is often wise to postpone surgery. Please inform your surgeon if you are concerned about your or your child's health before surgery, so that arrangements may be changed if necessary before coming to surgery.

Fasting instructions

It is unsafe to perform surgery soon after eating solid food. Therefore, a very important aspect of the hours preceding surgery is the restriction of the intake of solid foods and liquid drinks. The very important concern is that as anesthesia is being administered, food—indeed, any stomach contents—may be vomited and aspirated into the lungs, causing inability to breathe or pneumonia; in the worst case, this may be life threatening. The risk of aspiration of stomach contents in to the lungs increases with both volume of material in the stomach and its pH (acidity), and both increase with any food and drink. Your surgeon and anesthesiologist will provide you with detailed instructions about when to stop all intakes of liquids and food. In general, if surgery is to be performed in the morning, nothing should be taken by mouth after midnight the preceding evening. The following is a table of advice for children, based upon age and time of surgery. Please be certain to follow these instructions or those of your anesthesiologist. BEWARE: hungry children and adoring parents will often assume "just one bite" or "sip" will be acceptable; it will not be and will likely require delay or rescheduling of the surgery. Children will need constant supervision to assure they are in compliance with these requirements. One last note: clear liquids, such as water or apple juice, means you can see through them; orange or grapefruit juice and especially milk, for example, are not clear liquids.

Table for Fasting Instructions:

For Infants and Children Under Thirteen Years of Age:

- **8 HOURS**: Solid high fat food is allowed until 8 hours prior to the scheduled procedure (Meat, cheese, fried food)
- **6 HOURS:** Solid low-fat food is allowed until 6 hours prior to the scheduled procedure (Formula, milk, cereals, breads, fruit juice with pulp)
- **4 HOURS:** Breast mild is allowed until 4 hours prior to the scheduled procedure
- **2 HOURS:** Clear liquids are allowed until 2 hours prior to the scheduled prodedure (Water, Pedialyte, apple juice, Popsicles, clear Jell-O, Kool-Aid or Gatorade)

For Adolescents and Adults:

- **Nothing to eat or drink after midnight** prior to surgery; or at least eight hours prior to surgery.

Medications:

- Should be taken as usual with a sip of water up to 2 hours prior to the procedure

Inhalers:

- For asthma may be given at any time

Chapter 13:

The Day of Surgery

On arrival

Most hospitals or surgical facilities will ask you to arrive one to three hours before surgery. Although details and timing will vary, the purpose of this time is to perform administrative check-in procedures, acclimate and educate you or your child to and about the facility and procedure, and perform a check of vital signs such as height, weight, blood pressure, temperature and blood oxygen (with a monitor on a finger). Some of these aspects may be completed on a day before the surgery. All necessary paper work, including medical record documentation and review of informed consent forms will be organized and completed as necessary.

Anesthesia preparation

Your anesthesiologist and the operating room nurse will visit with you prior to going to surgery. Children, although often not infants, generally receive a premedication such as Versed, whose purpose is to relieve anxiety, calm them, decrease separation anxiety from family, and reduce unpleasant memories of the experience. This medicine may be given either as an oral liquid or nasal spray. Adults may have an IV started and receive premedication via this route.

During Surgery:

Staffing

Several people will be in the operating room, including anesthesia, nursing, and surgical staff. The anesthesiologist/ anesthetist, circulating nurse, surgical scrub nurse/technician and surgeon will be in constant attendance. Additional personnel may include and assistant surgeon or surgical assistant of your and your surgeon's choosing. In other specified circumstances, personnel in training, such as residents and fellows may be present.

Induction of Anesthesia

In the operating room, anesthetic gases are generally administered to children through a (scented) mask. After the children are asleep, a breathing tube is placed in the throat, either in or over the laryngeal opening to the lungs. By this means, breathing can be monitored and, as necessary, controlled throughout the procedure. IV's are placed in children after they are asleep. In adults with IV's in place, anesthetic agents may be administered as oxygen is breathed by mask. After asleep, a breathing tube is placed as above.

Monitoring

There will be continuous monitoring of key functions during surgery, under constant observation of anesthesia staff. These functions include breathing and respiratory functions, blood oxygen, temperature, blood pressure, and heart beats (EKG). Most strabismus surgery lasts less than one hour.

Chapter 14:

After Surgery:

This and the following (15 & 16) chapters are written by time sequence, and describe events—expected and possible—after surgery; but it should not be read that way. In other words, understanding these matters prior to surgery should be helpful in knowing what to expect or what may be of concern. Please consider them in the context of understanding the consequences and risks of the procedure (please also see Chapter 10).

Recovery process

You, your child or family member will go directly from the operating room to recovery area, where constant monitoring continues under the observation of anesthesia and nursing staff until patients awake. Parents or family (generally one member) may invited to be present in the recovery room, although usually are reunited in the so-called "step down" recovery area. Breathing tubes are removed either in the operating room or recovery room; either is appropriate and safe. Thereafter, patients are moved to a "step-down" area where family join in the continuing awakening and recovery process. IV's are generally removed in this area after liquids are being taken by mouth. Clear liquids are offered at this time. Popsicles are a popular option for children. There should be no excessive concern about rubbing the eyes. Doing so will be uncomfortable, and even children will not harm themselves or what was done in the surgery, once they have completely emerged from anesthesia. A cool, moist washcloth over the eyes is generally soothing.

Appearance: Short and Long Term

Immediately after surgery, tears on operated eyes will be blood tinged; this is normal and related to the fact that the surface of the eye is moist and very vascular (many vessels). This usually clears in a few hours, and has not significance to the outcome. They may be safely wiped away with a moist cloth. The first few days after the surgery, there is also an excess secretion of mucus in response to the surgery. This presents itself as moist or dried secretions that accumulate on the eyelashes, and may "stick" the eye lids together. Some children will resist wiping these secretions away with a moist cloth, and that will cause no harm. Eventually, within a matter of a few days, they will dislodge and no harm will ensue.

Bruising is unavoidable. On the surface of the eye, this appears bright (or blood) red. This is so because the blood lies beneath a clear membrane (the conjunctiva). The amount of bruising will vary from person to person, and even from eye to eye. While this observation is the most dramatic after surgery, it is probably the least meaningful, in that it will all go away within about two weeks. In children and on first muscle operations, the redness may last only 7 to 10 days. If a resection or reoperation of a muscle has been performed, there is more likely to be swelling on the surface—this may look like a blistering or ballooning of the surface membrane or conjunctiva, causing it to protrude between the lids—called chemosis. This may take longer to resolve, lasting three weeks

or more. After the bruise is gone, it will take several weeks for healing to be complete, and redness may gradually diminish over several months.

Sometimes there is bruising of the lids (a "shiner") as well. This is more common in older adults with fragile blood vessels, persons who have been on blood thinners such as aspirin and Coumadin, persons undergoing reoperations, and persons having surgery on the oblique (superior and especially the inferior) muscles.

After all healing is complete, there are subtle and unavoidable evidence that surgery has been performed. Ophthalmologists, observant patients and families, and occasionally others routinely make these observations. They generally derive from the anatomy of eye muscles as they attach to the globe, and the body's normal healing responses. With careful technique, they can be minimized, but not entirely avoided. Four examples of these are: 1) evidence of incision on the surface (scar), 2) bluish discoloration of the white of the eye underlying a recessed muscle, 3) a ridge on the white of the eye where the muscle previously attached, and 4) persistent thickening and redness of the white of the eye in the region of muscles undergoing complicated or reoperation procedures. The conjunctival scar may be minimized with careful technique; many surgeons will place the incision above or below the normal eye lid position (called a cul-do-sac incision), so that it may only be observed by pulling the lids up or down. Bluish discoloration of the sclera (or white) of the eye relates to the thinness of sclera under a normally positioned muscle. When muscles are recessed, this thinner sclera will sometimes appear as an oval shaped bluish discoloration (blue for the same reason the sky is blue—scattering of light). This is quite variable, and not seen in all persons. When a muscle is recessed, it is detached; where it was previously attached, the sclera is thicker and a low ridge or elevation is seen. The ridge is generally more prominent in adult patients. Multiple or complicated operations may lead to extended inflammation and scarring, and may be seen as raised and red tissue on the white of the eye. This scar (and redness) can often be surgically improved with removal of the scar (called "debulking") and repositioning of the conjunctiva; it may recur, although generally to a lesser degree.

Pain

The experience of pain seems to vary widely after strabismus surgery. The typical experience, especially for first-time operations, is moderate pain that responds to Tylenol or Motrin. The duration of pain varies from a few hours to several days. There is surface irritation associated with the preparation and incision; and there is aching soreness, associated in particular with movement of the eyes. The former generally lasts up to 48 hours, and the latter typically up to one week. Please bear in mind that individual circumstances vary widely. Adults often appear to experience more discomfort than children. The day of surgery is generally the most uncomfortable. However, especially for children, a nap in their own bed at home seems to be the best medicine. After this nap, children will sometimes awake as if ready to go at full speed with normal activities. Some adults will have minimal pain, others significantly more. Prior to surgery, please inform your surgeon about previous experiences with and tolerance to pain, plus medications known to be effective for your. In general, the more muscles requiring surgery and the performance of reoperations (previous eye or strabismus surgery) will increase the degree of discomfort. In some instances,

particularly older children and adults, eye drops may help to decrease inflammation and assist in pain control.

Activities and Ability to Function

Returning to normal activities after surgery is rapid. Most persons, even children, will choose and return quite rapidly to their normal lives. While there is some variation in ability to function following surgery, most persons will be able to do basic things within hours to a few days following surgery. General rules of thumb include:

- If the activity is not painful, it is likely to be acceptable
- One should avoid potential contamination of the eyes with irritants, such as soaps and shampoo, for two to three days
- Swimming (head submerged) should be avoided for several days.
- Driving should be a matter of individual confidence; some may drive as early as the day following surgery

Alignment, Double Vision and Head Position

Alignment of the eyes should be improved immediately after surgery. This may be somewhat obscured by bruising and swelling. Alignment may, and likely will change as healing occurs. Therefore, no final conclusions about the effectiveness of the procedure can be rendered in the first few days after surgery. Experience has shown, however, that certain patterns may be discerned. It is encouraging if eyes were crossed before surgery and completely straight following, and if double vision present before and absent immediately after. However, sometimes double vision will take a few days to weeks to resolve, even with successful surgery. If double vision was not present before surgery, it may even be an encouraging sign; it is after all the brain perceiving images simultaneously. With time, hopefully, the brain will "lock in" and fuse to receive the images together as binocular vision.

In the instance of intermittent or manifest exotropia, it is generally beneficial to initially overcorrect somewhat, and this may lead to temporary crossed eyes and double vision. As the muscles (typically the lateral recti) heal, they tend to pull the eyes outwardly and predictably. Occasionally, patching of one eye or prisms may be useful in reestablishing binocular vision while muscles heal. These methods are occasionally useful in early apparent over corrections of esotropia and hypertropia as well.

When surgery is performed to correct abnormal head positioning, the effect is usually immediate; in fact, in some instances it may be slightly overcorrected, only to return to a straighter position. In general, no final conclusions about the effectiveness of surgery can be made in the first week following surgery. By six to eight weeks after surgery, healing is nearing completion and more accurate assessments may be made. Even after this time or with apparent success there can be changes, particularly in cases where there is no binocular visual function or evolving medical conditions such as thyroid eye disease.

Wearing glasses and contact lenses

Glasses may be worn immediately following surgery. The surgery does not change the prescription of glasses to any appreciable degree. However, if glasses have

prism in them prior to surgery, then glasses without prism should be acquired for use immediately after. Contact lenses are generally not comfortable for approximately two weeks following the procedures.

Chapter 15:

What are the Potential Complications of Surgery?

Unsatisfactory alignment

Unsatisfactory alignment is usually apparent to all parties. Over and under corrections may be apparent to observers, or to the patient. This may be known by observation of the alignment proper, or by symptoms such as double vision. Remember that early post-operative alignment (within the first week following surgery) does not always indicate final alignment. Generally, the results of surgery can be reliably assessed at six to eight weeks following surgery.

Double vision

In children, double vision is uncommon or quite transient. The child's brain is able to adapt to the new alignment of the eyes very rapidly. Yet even if double vision does occur, it may be a positive indication! For example, if a child has been suppressing the vision in one eye before surgery, the appreciation of two images may indicate that they are being received simultaneously. One way for such children to resolve the double vision is to fuse, or process the images binocularly; such fusion can be beneficial long term in the development of vision and maintenance of straight eyes.

Infection

Infection in strabismus surgery is very uncommon. Three strategies have proven effective in its control. First, careful preparation with sterilizing agents at the time of surgery limits the number of bacteria in the area. Second, some surgeons will routinely use prophylactic antibiotics taken by mouth after the surgery. And third, prompt attention to the possible signs of infection can permit early institution of antibiotic therapy and prevent complications. The signs of infection are usually not subtle! The combination of large amounts of swelling and redness of the lids plus fever and unremitting and worsening pain suggest the possibility of infection. Call you doctor if you are concerned!

Loss of vision

Loss of vision can occur, yet is extremely rare. The usual cause of loss of vision is infection that spreads to the inside of the eye. The coat of the eye is thin, and an aggressive infection can spread along suture tracks. The key to successful treatment is early detection. Please note the signs of infection as above.

Anesthesia related problems

Serious problems with anesthesia are so rare as to be difficult to measure in the population of persons undergoing strabismus surgery. There has been a quiet revolution in anesthetic procedures and safety within the past 20 years. Modern medicines and monitoring techniques, coupled with highly skilled medical personnel, have made a general anesthetic extraordinarily safe. In healthy persons, the risk of serious

complications is probably on the order of one in one million. Please be certain to share with your doctors, especially your surgeon and anesthesiologist, all of your current medicines, allergies and medical problems. Make special mention of neurological problems, diseases involving muscles, and any problems with anesthesia family members may have had in the past.

Chapter 16:

Myths About Strabismus Surgery:

This booklet is meant to explain many aspects of strabismus and its surgical treatment. Hopefully, it will dispel some common misunderstandings about how the procedures are performed. The following are some common misapprehensions.

Myth # 1: Eye Position During Surgery

It is not necessary to remove the eye to perform strabismus surgery. The muscles attach to the sides of the eye and the surgery is performed with the eyes simply turned to the side, much as one turns one's eyes to look in normal directions of gaze.

Myth # 2: Use of Lasers

Lasers are not required (or appropriate) for strabismus surgery. Lasers generally are used to destroy tissue to mold or remove it. In strabismus surgery, the goal is to move or modify muscles (and tendons) to weaken or strengthen their actions, thereby to change the angle between the eyes. Hence, removing or destroying tissue would be counter productive.

Myth # 3: Anesthesia Risks

Anesthesia is no longer very risky. Modern anesthesia techniques and agents have made it very safe. This is not to say that there is no risk, simply that it is so small as to be difficult to measure. Anesthesia in healthy persons, including children of all ages, carries a probable major risk on the order of one in a million or less.

Myth #4: Surgery in the Office

Surgery cannot be performed in the office. Because anesthesia must be administered, surgery must be performed in a hospital or outpatient surgical facility. Generally a general anesthesia is required and appropriate.

Myth #5: Patches or Bandages After Surgery

Bandaging the eye(s) is not necessary following surgery. No patches need be applied; however, if comfort is increased, one or both eyes may be covered.

Myth #6: Trouble Seeing and Wearing Glasses

You will be able to see following surgery. Since the muscles are attached to the outside of the eye, there is little effect on seeing. Blurring of the vision is common following surgery, and may be associated with preparation before surgery (including eye drops that dilate the pupil and sterilizing irrigating solutions), and tearing and mucus secreting reactions after surgery. These effects are temporary, generally lasting a few to 48 hours. Surgery generally changes refractive error (strength of glasses) very little, and so glasses will not need to be changed. Glasses (minus prisms!) may be worn immediately after surgery.

Myth #7: Returning to Activities

You will be able to return to school or work within a very few days following surgery. While there will be bruising and some soreness, most persons are able to be up and about soon enough to return to school or work in two to five days.

Part IV:

Additional Things to Consider

Chapter 17:

About Children and Strabismus Surgery

It can be difficult to know how much a child can understand about strabismus surgery and what they should be told. Experience has shown that children, even children as young as three or four years of age, have a remarkable general understanding of what their problem is and what is being proposed. As the years go by, both that understanding and the consequent anxieties and concerns tend to become more significant and complex. When it comes to sharing the details of the surgical plan and experience, a person can be trusted to tell the truth about what they wish to know and in what detail. Your doctor may ask, or you may volunteer on behalf of yourself or your child, how much detail you wish to have presented, both before and after the surgery. Parents will have the best idea for their children, as individuals, what should be shared. One potential strategy is to simply say and ask as follows: "The doctor is going to fix your eyes. Do you have any questions?" Or, "What would you like to know about that?" Please share with your doctor what you would like to know and in what detail. Hopefully, what comes before this will have helped!

Parents may have a number of reactions to strabismus in their child. Please remember that a substantial part of a parent's job description is to worry and even second-guess themselves. Strabismus is a problem that fits into the category of "it is what it is", and we happily have a good range of treatments for it. There is certainly no need for blame or guilt, in that there are no circumstances where something was done wrongly by a parent or where something could have been done to prevent the condition. The impressive experience of seeing children bounce back very quickly from surgery is up lifting; most parents will note that the experience was harder on them than the child.

Chapter 18:

About Adults and Strabismus Surgery

Strabismus—and surgery to correct it—is similar to surgery in children, with some significant differences. Adults may have had strabismus their entire conscious lives, and are more likely to have had previous surgery. Strabismus may have also been acquired and be secondary to a wide range of associated medical conditions—traumatic, medical and neurological. Adults also have longstanding, and often more complex feelings about their strabismus. They know about their conditions, may have worried about the effects on their futures, and have accumulated (of ten negative) experience—their own and the reactions of others. Technically, the procedures can be more challenging, particularly if previous eye surgery has been extensive and/or the conditions more complex. Adult, especially elderly, tissues are more delicate to manipulation and slower to heal than children. All of this, when successfully overcome, makes successful correction of strabismus in adults gratifying to all.

Chapter 19:

Why It May Be Important to a Person to Have Strabismus Surgery (and How Much)

Persons who have endured strabismus know why it is important. And doctors who specialize in strabismus surgery have come to appreciate how beneficial it can be to have well aligned eyes. The potential benefits have been previously discussed (please see the goals section). It can be difficult to quantify those benefits, particularly as they are likely to vary from one person to another. And it is difficult to objectively measure benefits in children. Nonetheless, how such benefits compare and relate to other medical and life circumstance conditions are beginning to emerge. Such studies have been done and are being validated in adults. The results are striking. If we accept that the benefits in adults would be (at least) equal to those in children, we may extrapolate the results of studies in adults to children, hence all persons with strabismus. Using a method called utility analysis—utilities are measures of the quality of life associated with a health status—one can relate how important, cost effective and beneficial strabismus surgery is.
While the data is preliminary, one can say that to have strabismus (and experience its effects) is roughly equal to moderate vision loss in one eye or experiencing a small stroke! When asked whether they would surrender a portion of their remaining life, on average persons with strabismus responded that they would give up approximately 10% of their remaining life to be rid of strabismus! Strong testimony indeed.

Appendix

Common Types and Patterns of Strabismus for Which Surgery is Indicated?

Congenital or Infantile Esotropia.

This form of esotropia is present within the first six months of life. Generally, the amount of crossing, or angle of deviation, is large, measuring 15 degrees or more. The classic pattern is one of cross fixation, where objects to the right side are viewed with the left eye, and vice versa. This condition is a syndrome of likely associated conditions. The crossing of the eyes is the earliest manifest and the most obvious. Other features tend to present somewhat later, and include over or underaction of oblique muscles. The most common of these—present in about 75% of children with infantile esotropia—is over action of the inferior oblique muscle, and is recognized in side gaze. The eye looking toward the nose elevates higher than the opposite eye. When present bilaterally, a "V" pattern is present, meaning the eyes are more crossed in down gaze than up. Less commonly, overaction of the superior obliques—with over depression of the eye looking toward the nose—will result in an "A" pattern, meaning the eyes are more crossed in up gaze than down. In addition to oblique muscle dysfunctions, there is a tendency for one eye to drift upwardly and sometimes outwardly, a condition known as Dissociated Vertical Deviation (or DVD). These movements tend to occur if amblyopia (decreased vision in one eye) is present, and when affected persons are tired, ill or inattentive. These additional portions of the syndrome may require additional surgery, after correction of the initial crossing. Other features of the infantile esotropia syndrome are more variable and include amblyopia, for which patching may be required, and latent nystagmus (rhythmic or irregular, dancing or shaking movements of the eyes), for which no treatment is likely required or possible. Generally the first indicated operation for infantile esotropia is recession of the medial rectus muscle of each eye.

Partially Accommodative Esotropia

Accommodative esotropia is acquired somewhat later in childhood, typically age two to five years, and is treated with eyeglasses. Many children with this condition will also require patching of the dominant eye to improve vision in the crossing (or amblyopic) eye. In certain individuals, eyeglasses, even eyeglasses with bifocals, will not fully correct the crossing. In these cases, surgery is generally performed to correct the crossing that is not corrected by wearing glasses. Attempts to correct all crossing, including that corrected by glasses, may be misguided in young children, because to the degree that these children will outgrow their farsightedness, they will partially outgrow the need for their glasses. In effect, doing enough surgery to correct all crossing may increase the likelihood that children will need further surgery for over correction, or outward

deviation of the eyes (consecutive exotropia). Generally, the first indicated operation for partially accommodative esotropia is recession of one or more medial rectus muscle(s).

Intermittent Exotropia

Exotropia refers to eyes that drift apart, one eye staying centered, while the other drifts to the outside or toward the temple. Affected persons will often squint one eye closed (generally the eye that drifts), especially in bright light or when tired or performing prolonged near visual tasks. The usual pattern, no matter the age of onset, is intermittency. One eye will drift apart at times of fatigue, if the person is ill, "under the influence", or even daydreaming. It is generally more noticed when looking at the distance, rather than near. It will typically self-correct if attention is called to the deviation, or if a near object is viewed. Progression is typically manifested by an increase in the frequency and duration of the deviation. The total angle of deviation usually is an individual matter, and does not increase over time. The principal functional risk is the loss of binocular vision. The condition becomes symptomatic (increased eye strain and fatigue) with age. Double vision is not usually a prominent symptom, although it can be intermittently bothersome while driving, especially at night. Think of this problem as essentially being a problem of an abnormal, divergent position of rest of the eyes; that is, the eyes will tend to drift apart when effort is not being or able to be made to control the divergent angle. This is not a condition that typically self-corrects. While patching in very young children (under the age of two), eyeglasses with prisms (for small deviations in older persons who wear glasses) and convergence exercises (for older children and adults with small deviations who wish to try and will be compliant) may have some benefit, that benefit is often temporary and typically does not fundamentally change the position of rest. Surgery is the one modality that will change that position. Although conditions may vary, surgery is indicated when the condition is progressive and seen every day. While there is no emergency, there is typically no particular benefit to deferring surgery, once the indication for it is secure. However, delay will generally not cause an adverse outcome unless the deviation has become constant. Typically, the initial procedure performed is recession of one or both lateral recti.

Exotropia associated with poor vision and/or other abnormalities

In general, poor vision in infancy causes the eyes to converge; thereafter, if poor vision occurs in one eye, that eye will drift outwardly. This is known as sensory exotropia. Sometimes surgery performed for crossed eyes (esotropia) overcorrects the condition and the eyes drift outwardly. This is known as consecutive exotropia. One special form of this condition occurs when there is slippage of a muscle, typically the previously operated nose side, or medial rectus muscle. The muscle may not adequately attach to the eye and therefore be disinserted; alternatively, the muscle may gradually slip within its capsule (or sleeve) or gradually stretch/elongate with time, called a slipped or stretched muscle. When this occurs the opposing muscle, typically the lateral rectus muscle, contracts and becomes overly tight. For these conditions, surgery is often performed only on the affected eye and may consist of a recession of the lateral (or temple side) muscle, with or without a resection (tightening) of the medial (nose side) rectus muscle. When slipping,

stretching or disinsertion of a muscle is suspected, that muscle generally explored and reattached at the appropriate place. The "appropriate place" is generally dictated by findings at the time of surgery and guided by the judgment of an experienced strabismus surgeon. If a muscle is detached and "lost" (unable to be retrieved and typically a medial rectus muscle), pulling force may be borrowed from other muscles in the form of a transposition of all or some of the vertical (superior and inferior) rectus muscles. Sometimes such decisions must be made at the time of surgery, and your surgeon will explain and ask for your prior permission to do the appropriate thing based on findings at surgery. Your doctor will share, and you should understand the range of possibilities ahead of time; in part this is to permit your surgeon the latitude to do the proper procedure.

Hypertropia

Vertical misalignment of the eyes may be present and equal in all fields of gaze, called a comitant deviation. More frequently, vertical misalignment ia greater in one or a few fields of gaze, called an incomitant deviation. Three variations are described below.

Over or underaction of the oblique muscle(s): These conditions may be effectively treated in a number of ways. One common approach is to recess the muscle; and the recession may be graded in response to the degree of overaction. Despite some potential limitations, some surgeons prefer to sever the muscle (myotomy) or remove a portion of the belly of the muscle (myectomy). If dissociated vertical deviation is present, a recession of the muscle may be modified to transpose the insertion of the muscle forward (called and anterior transposition); this operation serves the dual purpose of weakening the overacting muscle while holding the eye down against the tendency to drift upward. It is often preferred as the primary inferior oblique procedure in the context of infantile esotropia. If over action of the inferior oblique is persistent, despite anterior transposition of the muscle, a nasal myectomy of the muscle may be performed. In very rare circumstances—where there is absence of the opposing superior oblique, in selected instances of extreme over action of the inferior oblique and under action of the superior oblique, and large torsional deviations—anterior and nasal transposition of the inferior oblique may be performed. A few surgeons will remove the muscle entirely for extreme over action of the muscle; however, this is an operation of last resort because it forecloses any other surgical options on the oblique (as above). When the muscle is under acting or weak, its antagonist (superior oblique) may be weakened to balance the forces (please see below).

Over action of the superior oblique has lead to a number of techniques to weaken the superior oblique that mimic similar operations on the inferior oblique. The anatomy differs in that the surgery is performed only on the reflected tendon and not muscle. The most commonly performed weakening procedure is either severing the tendon (tenotomy) or removing a segment of the tendon (tenectomy). When good binocular vision is present with stereopsis, a posterior partial tenectomy may be performed. In other cases, to preserve binocular function in the face of marked over action or tightness (for example, Brown's Syndrome), the tendon may be lengthened by placement of a silicone spacer. Some recommend a recession of the superior oblique tendon.

When weakness, or under action, of the superior oblique is present, the tendon may be tucked (or imbricated); alternatively, the antagonist muscle (inferior oblique) may be weakened as above.

All strabismus surgery outcomes are improved with good technique; and this is particularly meaningful in the context of oblique muscle surgery, particularly the superior oblique. It is the most challenging type of surgery to perform properly.

Vertical Rectus Muscle(s): When there is over or under action of the vertical rectus muscles (superior and inferior), a vertical imbalance occurs. These muscles may be tight, such as in inferior rectus fibrosis, thyroid eye disease involving muscles or fibrosis following injection with local anesthetic during cataract or other surgery under local anesthesia. In these cases the muscles act like a leash, mechanically restricting full motion of the eyes. Treatment is directed at releasing the leash or restrictive effect, generally by recession of the affected "tight" muscle.

Less commonly, as in conditions such as Dissociated Vertical Deviation, the muscles may apparently overact. In this condition, one may consider either recession of the superior rectus (to weaken vertical movement) or resection of the inferior rectus (to hold the eye down).

Fourth or Trochlear Nerve Palsy: One relatively common vertical muscle misalignment is caused by weakness of the trochlear (or fourth cranial nerve). In this case the muscle is receiving inadequate enervation from its brain stem nerve. The condition may be congenital (meaning present at or around birth, whether or not it is hereditary) or acquired, particularly as a consequence of closed head trauma. The most common strategy is to weaken the antagonist inferior oblique muscle, as above. In severe cases, particularly congenital fourth nerve palsy with a large head tilt, the superior oblique muscle may be tucked.

Compensatory (or abnormal) head posture (tilting or turning)

If a person assumes an abnormal position of the head, it may be secondary to a number of ocular conditions. There are a few eye problems that cause abnormal head positions. Strabismus may cause tilting of the head toward one shoulder, turning the face to one side (to turn the eyes in the opposite horizontal direction, e.g., face right for eyes left), and/or elevating or depressing the chin (to turn the eyes in the opposite vertical direction, e.g., cgin up for eyes down). In the above conditions, one or more of these compensatory positions may be assumed.

Duane's Syndrome comes in three types, and if abnormal head positions are assumed, the usual direction is a horizontal turn of the face; there also may be associated vertical up and down shoots of the eyes observed. To change the head position, depending on the type, either recession of the medial (usually) or lateral rectus (less commonly) muscle is appropriate.

In Brown's syndrome, the malpositioning of the head usually involves a chin up position, in an effort to avoid pulling against a tight superior oblique tendon. Tendon lengthening with silicone spacer is a commonly performed procedure.

In fourth nerve palsy, the head position is more complicated, with head tilt, turn and chin depression combined; a long standing head position can lead to developmental changes in the face, with hypertrophy (overgrowth, or enlargement) f one side of the face. As noted above, a common procedure is to recess the antagonist inferior oblique muscle; in large or long-standing deviations, recession of one or more vertical rectus muscle(s) on either eye may be added or substituted for the foregoing.

Nystagmus

Nystagmus is a special case. Some children will have calming of the motions of the eyes in one position, called the null point. The null point is a neurologically driven phenomenon, and the head positioning is a strategy to achieve the best possible vision. Improved vision is a powerful driver of a head position in these children. Nystagmus surgery is designed to move the head by moving the neurological null point to a place closer to or at straight ahead. Generally, large amounts of surgery on multiple muscles are required to move the eyes—and their null point—to a different position. A common procedure performed for this condition is the so-called Kestenbaum procedure, where simultaneous recession and resection procedures of large amounts are performed on the appropriate medial and lateral rectus muscles. The purpose is to move the eyes in the direction of the face turn, and away from the preoperative gaze (or eye) position.

Thyroid disease affecting eye muscles

Thyroid eye disease typically has three components: protrusion or bulging of the eyes, retraction of the eyelids (making the protrusion appear more prominent), and strabismus. The strabismus is the result of low grade inflammation of the extraocular muscles. Imaging studies may show thickening of the muscles. As a consequence of the inflammation there is stiffening, restriction and retraction of the muscles. With retraction, the eyes are pulled out of alignment. The most commonly involved muscles are the medial and inferior rectus muscles, although any may be involved. The most common operation performed is recession of one or more of the affected muscles. Eye muscle surgery may make retraction and bulging of the eyes more noticeable. Thyroid eye disease can be difficult to manage, as the condition may progress or change, and with it the alignment of the eyes; consequently additional operations are frequently required on eye muscles and eye lids.

Injury to eye muscles or orbital bones and tissues

Eye muscles may be injured in a variety of ways. Examples include:

- injuries that sever or lead to swelling of and hemorrhage into the muscle,
- injection of local anesthetics such as those used in cataract surgery,

- injuries to muscles during other surgery such as sinus drainage and plastic surgical procedures, and
- so-called "blow out" fractures of the orbit (bony socket in which the eye sits).

The first challenge in trauma is to carefully define the problem, its extent and effects. Imaging studies, such as computed tomography (CT) and magnetic resonance imaging (MRI), may be appropriate.

If strabismus occurs, particularly with diplopia, initial steps should involve primary repair (if appropriate and possible) of the original injury. After swelling has diminished and healing of the injury has occurred, strabismus surgery may be indicated. In general, the effects of injury cause scarring and restriction of eye movements, and therefore surgery is directed at releasing the restriction to permit alignment. Like some other complicated strabismus problems, the surgery is intended to improve the condition, but may, and likely will not completely reverse the effects of the original insult. Consequently, "perfect" outcomes are rare, and multiple surgeries may be required.

Under or overcorrected prior eye muscle surgery

The most common "complication" of eye muscle surgery is that the first procedure did not effectively align the eyes, either short or long term. There are reasons for this. First, the "problem" of primary (no obvious cause) strabismus is complex, involving multiple areas of the brain. Simplistically, one may say that it is "caused" by and abnormal pattern of nerve impulses sent from eye movement centers at the base of the brain. Current knowledge cannot contemplate "rearranging" these brain centers, and thus treatment is directed to the end organs, or eye muscles. In effect, we have a mechanical solution to a complex neuromuscular interaction. There is bound to be (and there is) significant imprecision in achieving desired effects. Second, the outcome of surgery is substantially related to the potential for binocular vision. If it can be established, or retrieved, long-term motor (or physical) alignment is more likely to be achieved and sustained.; and not vice versa. And thirdly, anticipated effects and improvements are derived from experience. That experience is derived from performing similar procedures on prior patients with the same condition of the same degree. How a given person will respond to that experience has a statistical variation that is only partially related to technique. Much of the variation is carried as a fact of nature in the human specie. For some, multiple procedures are required to achieve desired alignment. Only general statements may be made about the likelihood of needing additional surgery: complicated strabismus, neurological problems, and absence of binocular potential are risk factors for additional procedures.

The bottom line message is that reoperations are commonly necessary, either for over/ under corrections or for new problems that may arise. Bear in mind that, for most conditions, additional surgery is generally available and effective (within the above bounds). For most individuals, odds accumulated in their favor; meaning that (complicating factors notwithstanding) additional surgery carries a positive expectation of outcome.

Malformation of the position, or absence of, one or more eye muscles

Congenital malformations involving abnormalities in the formation or position of eye muscles are at once uncommon and challenging. Often, these problems accompany others, such as craniofacial malformations. Two examples are rotation of the eye muscles (so that they do not pull in the anticipated directions), and absence of the superior oblique tendon/ muscle. Imaging studies such as a computed tomographic (CT) scan can assist your surgeon is designing an appropriate approach.

New knowledge about the anatomy and function of orbital tissues, including muscles, is emerging. Aided by detailed magnetic resonance imaging (MRI), specialized elements of muscles and their surrounding attachments (called "pulleys") are gaining better understanding. Early basic information is beginning to be clinically applied to an emerging understanding of the potential role of these microstructures in clinical strabismus. This information holds significant promise for the future, particularly in complicated forms of adult strabismus.

About the Authors

Doctors Beauchamp and Mitchell are pediatric ophthalmologists and strabismologists (quite a mouthful!), practicing respectively in Dallas/ Fort Worth, Texas and Hartford, Connecticut. They are members of the American Ophthalmological Society and were trained in the specialty at Children's National Medical Center in Washington, DC. Both are actively involved in clinical practice, teaching and research.

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