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Adult Strabismus Part 2: Therapeutic Options

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The last issue of *Ophthalmology Rounds* presented the prevalent misconceptions and realities associated with adult strabismus. It discussed why adult strabismus is a disabling condition in most patients and explained why treatment for adults with eye muscle conditions should be considered restorative or reconstructive and not cosmetic. Part 2, in this issue, discusses strabismus in the context of cataract and refractive surgery and presents therapeutic alternatives for adult patients with strabismus. Dieffenbach is credited with performing the first eye muscle operation in 1839 on a patient with esotropia;[1] however, anecdotal accounts suggest that a form of strabismus surgery was done by itinerant physicians as far back as the 1730s.[1] The first reference to the use of adjustable sutures in eye muscle surgery was exactly 100 years ago and is attributed to Bielschowsky,[1] although the modern use of this technique was popularized by Jampolsky in the 1960s.[2] Fresnel prisms, originally designed in the 1820s, became a widely available nonsurgical option for diplopia management in the 1960s.[3,4] Pharmacologic weakening of the eye muscles using botulinum toxin became an alternative or adjunct to surgery in the early 1980s.[5] Despite the long history of adult eye muscle disorders, potentially helpful therapy for adults with strabismus, particularly surgery, may not be offered or delayed for an unnecessarily long time due to a lack of knowledge or an awareness of its benefits by the primary eyecare practitioner or the patient.[6] This issue of *Ophthalmology Rounds* provides an update to help practitioners refer patients for treatment in a timely and appropriate manner. As in the last issue, "adult strabismus" refers to strabismus in patients who are beyond the accepted age of visual maturity, which is generally considered to be age 8 to 9 years.[7,8]

Strabismus and cataract surgery

There are several scenarios in which strabismus can complicate a technically uneventful cataract surgery and lead to a variety of symptoms, including diplopia, headaches, and asthenopia. The etiologies can be divided into the 4 categories described below. It is important at the outset to differentiate true binocular diplopia from monocular causes of diplopia.

Pre-existing disorders: Strabismus conditions that precede the onset of cataracts can lead to troublesome diplopia after vision is restored. These disorders include thyroid orbitopathy, childhood strabismus, isolated cranial nerve palsies, and myasthenia gravis.[9-13] It is important to document any preexisting conditions so that patients can be warned about the risks of postoperative diplopia.

Strabismus precipitated by prolonged vision deprivation: A cataract that compromises vision for a long time can disrupt fusional vergences and lead to decompensation of a previously undiagnosed heterophoria, usually a vertical phoria.[10,11] In rare instances, it can also lead to permanent loss of fusion ability, a condition termed "central fusion disruption," which is refractory to traditional therapy, including prisms and surgery.[9,11] Before cataract surgery, patients should be advised about the risks of diplopia, including the possibility of a rare debilitating diplopia that may only be controllable with an occlusive patch, filter, or contact lens.

Local trauma: Retrobulbar and peribulbar injections and superior rectus bridle sutures have been documented to cause restrictive muscle conditions that can lead to eye misalignments.[9,11,12] The most common forms are vertical imbalances caused by damage to the inferior rectus muscle.[9,13,14]

Optical aberrations: Binocular diplopia and asthenopia can be caused by a variety of optical factors, including decentered intraocular lenses or corneal irregularities, that may also cause monocular diplopia.[10,13] These symptoms can also result from an asymmetric quality of the images in the two eyes that can be induced by anisometropia or aniseikonia.[9,13] Finally, switching fixation from the dominant to the nondominant eye can create asthenopia and, occasionally, diplopia in adults.[9]

Strabismus and refractive surgery

Refractive surgery can be beneficial in eliminating certain forms of strabismus related to either symmetric or asymmetric refractive errors. However, several precautions should be taken in the

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preoperative evaluation of patients. It is imperative that all patients considering refractive surgery have their eye muscle balance documented by an eyecare practitioner or an orthoptist. A careful history should also be taken to inquire about prior strabismus treatment to avoid these unexpected postoperative binocular vision problems.

Refractive surgery to treat strabismus

Refractive surgery can be a useful alternative when treating some forms of strabismus. There are small case series in adults that have documented the successful treatment of accommodative esotropia by eliminating the hyperopic refractive error.[15-20] Refractive surgery can eliminate the accommodative component of a mixed accommodative and nonaccommodative form of esotropia[21] and it has been reported to have eliminated exotropia in 2 patients with myopic anisometropia when the refractive errors were corrected in both eyes.[18] Small vertical deviations have responded to laser for correction of refractive errors.[20]

On the other hand, refractive surgery can lead to unexpected or unwanted results. In one report, a minority of patients with longstanding accommodative esotropia had no changes in their esotropia despite successful correction of hyperopia. These patients could not be identified by sensorimotor testing prior to laser surgery.[17] Another study by Snir et al describes a series of myopic patients whose exodeviations significantly worsened after refractive surgery.[22]

Binocular vision problems after refractive surgery

Strabismus-related symptoms can occasionally complicate uneventful refractive surgery. The most common complaints are diplopia and asthenopia. The causes include technical problems; a pre-existing need for prisms; aniseikonia and anisometropia; induced monovision; improper control of accommodation; and prior strabismus surgery:[23,24]

- Although not common with progressively improving laser technology, optical aberrations or a decentered ablation can lead to either monocular or binocular diplopia,
- Corrections for diplopia (eg, prisms in glasses) must be documented prior to surgery. Failure to do so can lead to unexpected diplopia once the refractive error is eliminated.[23,24] The preoperative alignment may also be influenced by prisms induced in high-powered spectacles and should be taken into account preoperatively.
- In some cases, asymmetric corrections can lead to aniseikonia that precludes binocular fusion.[23-25]
- Induced monovision can cause a change in ocular dominance or lead to decompensation of a marginally-controlled horizontal or vertical deviation.[24-26]
- Prior to surgery, binocular alignment must be checked with accommodation-controlling targets and appropriate accommodative correction to determine the basic deviation prior to refractive surgery. Otherwise, an asymptomatic strabismus can decompensate after surgery.[23] The surgeon must confirm that the original refractive correction was not under- or overcorrecting the true refractive error; otherwise, latent heterophorias may manifest after surgery.
- Care must be exercised in patients who have had prior strabismus surgery, since their suppression mechanism may be altered by a change in alignment resulting from correction of their refractive errors.[24,25]

Nonsurgical options for adult strabismus

"Tincture of time"

Not every adult strabismus problem demands practitioner intervention. Some conditions may resolve on their own or do not require active treatment.

One example is a cranial nerve paresis from a microvascular cause (eg, diabetes), although temporary treatment for diplopia may be needed in some patients. In the absence of any other complicating factors, the paresis typically resolves over a 3- or 4-month interval. Another example is diplopia after surgery for exotropia. This is an expected symptom since surgical dosages are often chosen to create a temporary overcorrection of the primary position angle of deviation. In most cases, the esotropia gradually resolves over the first 6 to 8 weeks after surgery. A third scenario where treatment is not required is a misalignment that manifests only in certain gaze positions that does not affect the patient's daily functioning; eg, an orbit floor fracture that limits upgaze in one eye. If the patient does not have diplopia in primary position and the field of binocular single vision (BSV) shows a good range in the other directions of gaze, then the range may spontaneously expand further into upgaze over time. Even if it does not improve, it may not require treatment if it does not compromise the patient's occupational needs or other activities.

Finally, a small heterophoria that occasionally dissociates under stress due to illness or fatigue does not require intervention if it does not affect the patient's daily activities or enjoyment of life.

Eye exercises

In the realm of ophthalmology, there are very few disorders where eye exercises are felt to be of proven benefit or used to any great extent.[27]

Convergence insufficiency: Eye exercises are unquestionably beneficial in convergence insufficiency, which causes eyestrain (asthenopia) and, occasionally, diplopia. There are 2 forms of this condition.

The "typical form" generally occurs in younger adults who exhibit a symptomatic exophoria in the near position, along with reduced fusional convergence amplitudes. The condition must be confirmed by quantitative assessment of the amplitudes using prisms. Therapy consists of convergence training with prescribed stereoscopic and fusion tasks; this leads to resolution of symptoms in >70% of cases.[28] Some patients regress over time and require periodic retraining of convergence. In severe cases, fusional amplitudes can be improved with prism exercises ("prism jump therapy"), while more extreme cases may require the constant use of prisms for near work (see next section on prisms). Convergence insufficiency after head trauma can be refractory to exercise and may require prisms or surgery.[28] The assessment and nonsurgical treatment of convergence insufficiency is greatly facilitated by an orthoptist, an allied health practitioner, trained in the assessment of strabismus and administration of nonsurgical treatments.

A second form of convergence insufficiency is typically observed in adults who have a combination of poor convergence and poor accommodative amplitudes.[28] In any case of convergence insufficiency, it is important to accurately measure the accommodative amplitudes.[29] If they are weak, the first step is a plus correction to improve the comfort in performing tasks in the near-fixation position. Although there may be concerns that the lenses risk increasing near exophoria and, therefore, will worsen the condition, the improved vision comfort at near often promotes improved fusional amplitudes, allowing patients to control the heterophoria. If the near correction does not relieve symptoms, a trial of convergence exercises is prescribed while the patient wears the near correction.[28] This combined approach often controls the condition; otherwise, further options can be offered, including prisms or surgery.

Other exercises: One useful exercise is the purposeful duction effort to "unwind" a tightened muscle after strabismus surgery to prevent its contracture during the short-term postoperative course. The regular voluntary movement of the eye into the gaze field opposite the tight muscle can improve the comitance of the eye movements into that field over a number of weeks. Anti-suppression exercises to increase diplopia awareness are rarely performed, except by some orthoptists in treating convergence insufficiency.

Prisms

Prisms are used in 3 contexts in the management of adult strabismus: to regain or maintain fusion; orthoptics; and diagnostic testing.[30]

Prisms to regain fusion: The most common reason for prescribing a prism is to control diplopia, the most troublesome symptom for older children or adults with misaligned eyes. Prisms can correct diplopia due to a horizontal or vertical heterotropia or a combined disorder (causing oblique separation of images). They are also helpful for maintaining fusion in symptomatic heterophorias, such as cases of exophorias in the near position causing asthenopia (see previous section). Prisms can be used to train vergences over a number of weeks for various conditions, including convergence insufficiency and decompensated vertical heterophorias ("prism jump therapy").[3,28]

Determining the appropriate amount and orientation of a prism to correct a patient's problem often requires some trial-and-error work with the patient. It is very helpful to have an orthoptist make these determinations. There are several useful rules of thumb that can help the practitioner. First, if the patient has some fusion reserves, indicating that the strabismus was likely longstanding and probably regressed slowly over time, then one can try a prism that corrects approximately half the total angle. This principle applies to both horizontal and vertical deviations. If the patient can manage comfortably for a period of time with this prism in place, it is not necessary to offset the entire deviation. On the other hand, some adults who present with an acquired strabismus and who have very poor fusion reserves may require a prism that almost completely offsets the deviation. This scenario is frequently encountered in conditions such as thyroid strabismus or in strabismus following head trauma.

Patients who have an oblique deviation can be managed with a horizontal correction for one eye and a vertical prism for the other eye. Alternatively, a single prism can be prescribed that is oriented at an angle that optimizes both vectors. There are books with tables that give the recommended single prism (power and orientation) equivalent to the result when the two components are determined separately.[3] The practitioner can also make a reasonable guess of the required power and angle by using the principle of the right-angle triangle.

Prism orthoptics: Prisms can be used to modify the basic alignment of both eyes to provide information that is useful in managing various conditions. For example, a patient with nystagmus and straight eyes may benefit from surgery to create an artificial divergence, thus forcing the patient to use convergence power that can dampen the nystagmus and improve the binocular visual acuity. To determine if this approach would be beneficial, the surgeon can prescribe a temporary base-out prism and assess the patient's response to the induced divergence stress.[3,31] If the patient returns with improved vision and less intensity of nystagmus, then surgery may be successful. Another example in nystagmus management is the use of prisms to conjugately deviate the eyes to correct a head turn due to nystagmus. Prisms are placed base-out before the adducted eye and base-in before the abducted eye to shift the images in the same direction in the two eyes.[3]

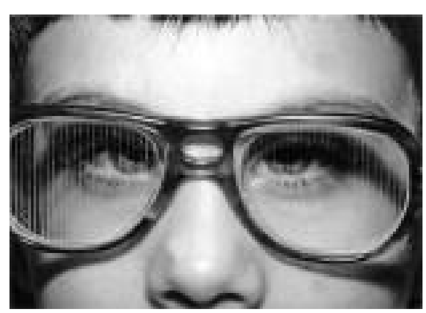
Prisms can be used preoperatively to assess the relative significance of a vertical and horizontal deviation when both are present, eg. if a patient with exotropia has a small vertical deviation as well, the total horizontal angle can be offset with a prism to see if the patient can control the vertical deviation. If the vertical component does not preclude fusion when the horizontal deviation is corrected, then the surgeon does not have to be concerned with correcting the vertical heterotropia at the same time. Similarly, if a patient has a cyclotorsional problem along with either a vertical or horizontal component, a prism can be used to offset the latter vector to see if the patient's cyclofusional amplitudes can allow control of the torsional component.

Finally, prisms can be used postoperatively to "rescue" the fusion state of an under- or overcorrected angle in order to regain fusion. They can also be used to slightly overcorrect the residual angle to create diplopia with the goal of building up vergence reserves.

Diagnostic tests with prisms: Prisms can be helpful in the preoperative assessment of convergence excess esotropia, which is characterized by an esotropia that is larger at near than at distance. Prisms can be used to totally offset the esotropia in the near position to see if the patient can control the induced exotropia at distance. If the patient shows no tendency to an overcorrection at distance with the prisms, the surgeon can be fairly confident that surgery to correct the entire near angle will have a low risk of overcorrection at distance.[32,33] Prisms have also been used to determine the target angle for exotropia when the deviation measurement is different at distance and near.[3] In addition, prisms have also been used preoperatively to try to eliminate suppression and build up fusional vergences.[30]

Forms of prisms: Three types of prisms are used in strabismus therapy: ground-in prisms, temporary (applied) prisms, and prism bars. Ground-in prisms are part of a glasses or contact lens prescription that is, by definition, a long-term treatment. Adhesive prisms are usually of the Fresnel variety and used on a temporary basis (Figure 1). A prism ground into a spectacle lens becomes cumbersome when it is >8 prism diopters (PDs); therefore, ground-in prisms are not practical for long-term use when the total prism correction exceeds 15 or 16 PDs. For larger angles, a Fresnel prism is a reasonable alternative. The limit for prism power in a contact lens is 2 to 3 PDs, and these are used almost exclusively for treating vertical diplopia. The advantages of Fresnel prisms over the ground-in type are their lower cost, lighter weight, the opportunity to easily change

Figure 1: **Fresnel prisms applied to spectacles worn by a patient with esotropia**



powers, and the higher range of possible powers (up to 40 PDs). The ground-in form has the advantage of clarity, since the Fresnel prism creates a slight blur due to the plastic. Also, Fresnel prisms discolour or fray over many months, and require replacing.

The decision regarding the type of prism to use is dictated by cost considerations, the size of the angle, whether the eye muscle condition is expected to be temporary or not, and whether there will be changes over a short period. For example, diplopia in the post-operative period that is expected to resolve with time would be best treated with a Fresnel prism that can be tapered over several weeks. On the other hand, a patient with a stable small-angle strabismus causing diplopia would do well with a prism ground into glasses or a contact lens. Finally, prism bars that are used to measure ocular deviations can also be used in a prescribed program of eye exercises to build up vergence reserves ("prism jump therapy").

Occlusion

Cases of troublesome diplopia or monocular diplopia that cannot be solved by surgical or nonsurgical methods may require blocking the vision of the deviated eye. The simplest solution is an eye patch. However, there are now alternatives that are more cosmetically appealing, one of which is the Bangerter filter (or foil). The Bangerter filter comes in a range of densities in 0.1 log unit steps, and one can apply the lowest density that alleviates the patient's symptoms (Figure 2). Alternatively, one can order a frosted lens in a spectacle frame (such as a Min lens) or use translucent surgical tape to accomplish the same result. In general, the deviated eye is occluded.

Pharmacologic options

Topical agents: The use of cycloplegic agents, such as atropine,was discussed in Part 1 in the context of therapy for amblyopia. Miotic agents are occasionally used to treat children with some forms of esotropia, but these are not generally used when treating adult forms of strabismus.

Oral medications: Oral medications, including dopaminergic agents used to treat amblyopia in older children and adults were also discussed in Part 1. Oral agents, both steroidal and nonsteroidal, may be required in some acquired inflammatory cases of strabismus due to orbital diseases (eg, idiopathic inflammatory pseudotumour or orbital myositis) or those resulting from systemic diseases (eg, giant cell arteritis). Finally, medications used to treat the ocular signs of myasthenia can correct the eye muscle problems and ptosis caused by that condition.

Figure 2: A Bangerter foil applied to one spectacle lens of a patient with intractable diplopia

Injected agents: Three agents are used in the diagnosis and treatment of eye muscle conditions: anti-inflammatory drugs, local anaesthetics and botulinum toxin. Some forms of adult strabismus, such as acquired Brown syndrome, are caused by local inflammation and can be treated by injection of steroids into the orbit. Local anesthetics such as lidocaine can be injected into an eye muscle under electromyographic (EMG) control to temporarily weaken that muscle. This can be used either as a therapeutic trial to help assess the potential surgical result of surgery on the muscle or as a diagnostic test to see the contribution of a muscle in a complex ocular muscle condition such as Duane syndrome.[34,35]

Botulinum A toxin injections into extraocular muscles have been used as an alternative to strabismus surgery since the late 1970s and came into widespread use in the 1980s. This agent has been used to treat a wide variety of eye muscle problems.[5,36] A minute quantity of the toxin is injected into the eye muscle, either under EMG guidance in an out-patient setting or under direct observation at surgery. Experience in adults has confirmed that it is less successful than surgery for the long-term alignment of eye turns;[37] However, success has been higher for small strabismus angles (<25 PDs) than for larger deviations, and it is slightly more successful for esotropia than exotropia.[37] One prospective study demonstrated that patients undergoing adjustable suture strabismus surgery had a much higher rate of long-term success than those who had botulinum toxin injections.[38]

Botulinum toxin injections were touted in the past as a useful option for treating acute sixth nerve paresis by temporarily relaxing the medial rectus muscle and allowing recovery of the weak lateral rectus. However, experience has not shown that botulinum toxin leads to better recovery of lateral rectus function than would occur with natural healing.[39,40] It is still a useful adjunct to surgery in treating large-angle strabismus, especially large-angle exotropia and total third nerve and sixth nerve palsies.

Surgery for adult strabismus

As discussed in Part 1, strabismus surgery in adults is "restorative," which is highly successful in eliminating a patient's symptoms, as well as gaining several functional benefits.[41] A high percentage of adults regain satisfactory alignment, usually defined as a reduction of the preoperative angle to a few PDs.[7,42,43] Surgery in adults can be done using local anesthesia, although recent advances in general anesthesic agents and laryngeal airway management have improved case turnover times.

New developments in surgery: In recent years, there has been significant progress in strabismus surgery, both in terms of technical quality and in the variety of available procedures. Very few strabismus conditions are now considered untreatable, due to the myriad of available tools. One paradigm that has shifted in recent years is the design of the strabismus procedure, which is now individualized for each patient. It is no longer acceptable to simply plan a correction for the primary position alignment. Planning for strabismus surgery should attempt to maximize the field of BSV (the field of fusion) so that the patient gains the widest possible range, whereby the eyes move in tandem. Ideally, single vision should be gained within a range of 30º from the primary position.[41] This may require shifting the traditional planning of surgery in terms of the choice of muscles and the dosage of surgery.

In recent years, one area where surgical advances have been made is in correction of the various orientations of head postures due to nystagmus.[44] Another area is in eye muscle transpositions to treat a variety of conditions, including sixth nerve palsy and dissociated vertical deviation (DVD).[45] Current understanding of the functions of the ocular muscle pulleys has led to novel approaches to treating A and V syndromes, ocular muscle slippage, and the eye movement anomalies in Duane syndrome.[46] Finally, new methods for treating cyclotorsion have improved success rates for treating fourth nerve palsies, as well as strabismus following macular translocation and scleral buckle surgery.

Adjustable suture surgery: One of the major advances in strabismus surgery has been the use of adjustable sutures, popularized by Jampolsky in the 1960s.[2] This option allows the surgeon to place the eyes in the most desired position in the immediate postoperative period to gain the best chance for successful long-term alignment. Conversely, it affords the surgeon the chance to avoid leaving the eye in an undesired position.[47] One study showed that adjustable suture surgery had a higher success rate than conventional surgery, although it was particularly advantageous in cases of repeat strabismus surgery and in correcting complex horizontal and vertical misalignments.[48] There are two basic methods for adjustable sutures on eye muscles: One involves a bow-tie knot that can be untied and retied as needed. The other method uses a suture handle that can be moved up and down the muscle sutures to change the muscle position[49] (Figures 3 and 4). These sutures are generally adjusted on the same day as surgery or up to 24 hours later, although use of various scleral lubricating compounds at surgery may allow adjustment up to 3 or 4 days later. Scleral stay sutures brought out onto the skin can be used to augment a recession in severe restrictive strabismus, and these can be adjusted as needed for up to 10 days after surgery.[49]

Figure 3: Diagram of a bow-tie adjustable suture method for recessing a rectus muscle. A hang-back suture is used for the muscle and a temporary knot is fashioned in the muscle sutures that can be undone when the muscle position is adjusted later in the day. Note the temporary scleral loop suture near the limbus, which allows the assistant to fixate the globe while the surgeon adjusts the muscle position.

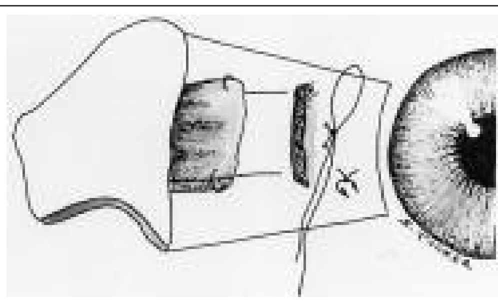


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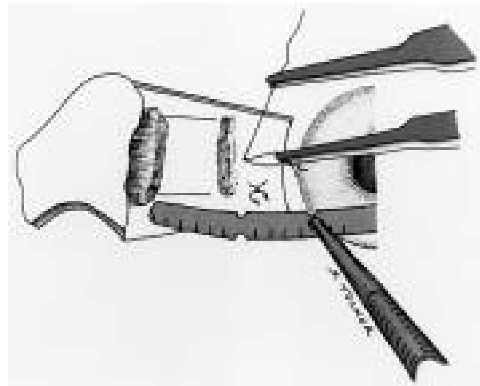
Figure 4: Diagram of slip-knot adjustable suture method for recessing a rectus muscle. A suture "handle" is tied around the muscle sutures and is moved up or down the muscle sutures as needed to change the muscle position at the time of adjustment.



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Studies of adjustable sutures in horizontal eye muscle surgery have shown rates of successful long-term alignment ranging from 83% to 92%.[7,42,43,47,48] These results are higher than for traditional (nonadjustable) surgery, where the reported success rates are about 75% to 80% overall.[48] One study revealed that the success rate for esotropia and exotropia repairs was equivalent to primary and repeat surgeries.[47] Their success rates of >85% for all groups were contingent on achieving a tight range of postoperative alignments immediately after surgery of within 3 PDs of "0" for esotropia and between 3 and 7 PDs of overcorrection for exotropia. Success rates for other forms of strabismus vary by condition but, overall, a majority of patients can benefit from a significant improvement in their alignment and an overwhelming majority are pleased with their postoperative results.

Complications of strabismus surgery: Complications of strabismus surgery are, fortunately, uncommon.[50] The rate of infections after surgery is low with the general use of postoperative topical antibiotics. The rate of endophthalmitis is extremely rare and estimated at 1 in 20,000 to 30,000 cases. Suture reactions are uncommon since most sutures are self-dissolving and made of synthetic materials. These reactions are usually controlled by topical steroids. Dellen can occur if the conjunctiva is edematous and they respond to the use of lubricants. Conjunctival or subconjunctival cysts may have to be removed if unsightly or irritating to the patient. Anterior segment ischemia can arise in vascular-compromised patients who have surgery involving >2 rectus muscles in one sitting. Other unusual complications include retrobulbar hemorrhages, myositis, scleritis, and scleral perforations.

Conclusion

The treatment of adults with strabismus includes several nonsurgical and surgical options that yield numerous functional benefits. Many patients do not require surgery and can be managed by other means that can be safe and effective. Surgery for adults is restorative and highly cost-effective. Surgery is very successful in restoring satisfactory alignment and a majority of patients have a resolution of their preoperative symptoms with a low risk of complications, including diplopia.

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