VIEWPOINTS

Viewpoints: Treatment Options for Strabismus

It is well-documented that cosmetic and functional cure of strabismus is frequently obtained with optometric vision therapy without the need for surgical intervention. In cases with a poor prognosis for success with vision therapy alone, functional and cosmetic cure rates and stability are increased when vision therapy to equalize monocular skills and develop fusion is combined with surgery to reduce the deviation.

In this issue, the role of surgery in the optometric management of strabismus is explored by two authorities, Dr. Richard London and Dr. William Ludlam. Dr. London is affiliated with the Permanante Medical Group in Oakland, California, is an Associate Clinical Professor at the University of California-Berkeley School of Optometry, and is Editor-in-Chief of Mosby's Optometric Problem Solving series. He suggests that medical-surgical treatment provides a useful option in some cases of strabismus, serving as an adjunct to vision therapy in noncomitant and large-angle deviations. Dr. William Ludlam is a former Associate Professor at SUNY, State College of Optometry and is currently a Professor of Optometry at Pacific University College of Optometry. He emphasizes that strabismus surgery infrequently achieves functional binocularity, that cosmetic results are not always good and frequently deteriorate with time, that multiple operations are often required, and that surgery may be accompanied by adverse complications. He urges its use only in those cases where the angle of deviation is too great to secure alignment with vision therapy alone, or to achieve cosmetic results in those cases that do not respond to vision therapy.

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Surgical and Medical Options as Part of an Overall Treatment Approach for Strabismus

Richard London, O.D., M.A.

Abstract

Optometrists have many therapeutic options available in the treatment of strabismus. In some cases, the magnitude or cause of the ocular deviation make it unlikely that optometric intervention alone will provide complete functional and cosmetic cures. Surgery and medicine provide tools which may reduce the strabismic angle, and allow the optometrist to better employ uniquely optometric treatments aimed at functional improvement.

Key Words

strabismus, strabismus surgery, thyroid myopathy, myasthenia gravis, Duane's Retraction Syndrome, miotics, sensory fusion

Surgery and medicine provide viable treatment options in some cases of strabismus. They are tools to be used judiciously and in conjunction with other therapies. If our goal is a functional cure, we must consider using every tool available. I want to be very clear that I consider medical-surgical options as an adjunct to the total care of the patient. I am in no way advocating surgery *instead* of vision therapy, optical or prismatic prescription.

Any optometrist who has collaborated with a good strabismic surgeon has seen that medical-surgical intervention may be beneficial. Unfortunately, many optometrists are only exposed to poor surgical outcomes (as are some ophthalmologists to optometric cases)—dissatisfied patients who find their way to our offices. They do not experience the very satisfied patients who remain with their surgeons for follow-up care. In addition, most optometrists do not examine some of the severe cases of strabismus seen in ophthalmologic practice since the referral source for these patients is other physicians.

When reviewing surgical outcomes in strabismus (or carotid artery endarterectomy,[1] or open-heart surgery, etc.), remember that results often depend on the surgeon's (or surgical team's) skill more than on the surgical technique. Likewise, in vision therapy we all know some clinicians who are able to achieve impressive results in cases which other competent clinicians cannot. To hear these successful doctors present case reports and review the therapy step by step makes it sound simple. It is usually not so simple. During political battles, optometrists and ophthalmologists will cite the least favorable results from each profession. Truth, as usual, does not live at either extreme.

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VIEWPOINTS

The Role of Surgery in the Optometric Management of Strabismus

William M. Ludlam, M.S., O.D.

Abstract

Although strabismus surgery is often useful in reducing large angle strabismus deviation, it is approximate, does not usually result in functional binocularity, often requires more than one procedure to achieve even acceptable cosmetic results, has some very serious side effects (including death) and has a poor record in producing a long-term cure of the strabismus. Vision therapy, on the other hand, is a very conservative method and produces a high rate of functional and cosmetic cures, with a good long-term cure rate, and resolution of the sensory-perceptual problems: amblyopia, ARC, and suppression, which often accompany strabismus. Its only drawback is that it requires sustained effort of the parent and child in a therapy program over an average six- to 12-month period.

Key Words

strabismus, surgery, vision therapy, non-surgical strabismus interventions

Surgery for strabismus is but one of four approaches available for its treatment and rehabilitation. The others are *refraction,* including spherical, cylindrical and prismatic correction; *vision therapy,* including amblyopia therapy, fusion training and occlusion; and *pharmacologic,* including atropinization, penalization therapy, cholinesterase inhibitors, and botulinum injection to weaken the action of a given extraocular muscle, e.g., medial rectus in esotropia.

Non-surgical Approaches

Refraction and vision therapy have been in use for strabismus therapy for well over 100 years. Reports showing roughly 75% functional and cosmetic success rates in unselected populations have been presented by Javal,[1] Cantonnet and Filliozat,[2] Smith,[3] Ludlam,[4] Ludlam and Kleinman,[5] Etting,[6] Hoffman et al.,[7] and Flax and Duckman.[8]

The method takes time to achieve cosmetic and functional cures, but has shown almost no harmful side effects and produces high rates of long-term cure. The strabismus characteristics which have proven most difficult to treat non-surgically are:

1. Very large angles of deviation (greater than 50).[4,5]
2. Deeply embedded anomalous correspondence (ARC) in esotropia.[4,5,9]
3. Noncomitant strabismus in which an injury to either the 3rd, 4th or 6th cranial nerve, or to one or more of the corresponding extraocular muscles, has occurred.

Pharmacological

Of the pharmacologic treatments, long-term atropinization and long-term use of cholinesterase inhibitors have been largely discontinued because of serious side effects, including cataracts and death when additional muscle relaxants were prescribed.[10,11] Only monocular atropinization (instead of patching) for amblyopia therapy and botulinum injections are currently used. These, too, present problems.[12,13]

Surgery

Of the cases of strabismus listed above as being difficult to treat non-surgically, surgery is useful in producing improved results in only those with a very large angle of deviation. It has not proven useful with ARC and noncomitant strabismus.

The basic problem in concomitant strabismus is not a long or short, or strong or weak, extraocular muscle, but a supra-nuclear innervational problem of the oculomotor system.[14] Costenbader and Albert,[14] two eminent strabismus surgeons, state that extraocular muscle surgery in strabismus is a treatment which does not directly deal with the cause:

> The basic factors in the etiology of strabismus are rarely in the muscles themselves and thus the surgical procedures are not directed at the primary pathogenesis.

In fact, obtaining functional binocularity as well as cosmetic alignment surgically is so elusive that most studies of the results of strabismus surgery evaluate success solely in terms of the residual deviation. Cosmetic success or cosmetic cure is usually defined as $\pm 10^{\Delta}$ of residual deviation.[15,16]

Flax and Selenow[17] reviewed the literature of surgical results for intermittent exotropia over a 35-year period. They found only 22 papers which contained pre-surgical and post-surgical comparisons using stated criteria for success. These reports comprised a total of 1,490 patients. Flax and Selenow conclude:

> One-third of the reported cases did not achieve even the minimum benefit of being left with a low angle deviation, with the indication that one in six derived absolutely no bene-

fit at all or were harmed by the surgery. Two-thirds did achieve straight eyes, but only one-third attained normal binocular function along with alignment.

Scheiman and Ciner[18] reviewed the literature on esotropia surgery. Studies that reported on a total of 1,473 surgical procedures indicate a 15% functional cure rate and a cosmetic cure ($\pm 10^{\Delta}$ of residual deviation) rate of 43% when patients are evaluated shortly after the surgery. Long-term results, even when judged only by cosmetic criteria, are still poorer.[15,16,18-21]

The situation is summarized by surgeons Kennedy and McCarthy,[21] who report:

> With surgery one can only attempt to align the eyes; it does not of itself purport to keep them where they are placed. Where they eventually stabilize depends on the particular idiosyncrasies of each individual patient, and one cannot always forecast the long-term outcome.

Multiple Surgery

The need for multiple surgical procedures is dealt with in depth in the ophthalmological literature on strabismus. In reviewing the subject of multiple surgery on infantile esotropia, Ludlam[15] reports that the greater the time period allowed for follow-up, the greater the percentage of second and third surgeries required, so that an average of 30%-35% reoperations are reported for two-year follow-ups, with 50%-60% needed for five-year and longer follow-ups in order to maintain satisfactory cosmetic alignment.

Surgical Complications

Not only do the short- and long-term results with respect to alignment leave much to be desired, but a considerable number of serious side effects may occur as well. Several studies22-26 report perforation of the globe during strabismus surgery occurring in 8% to 15.5% of cases, frequently causing chorioretinal scars and ocular fundus anomalies. Other sequellae of globe perforation include: posterior chamber hemorrhage,27 hyphema,28 iris ischemia,29 anterior segment ischemia,30 retinal detachment,31 and endophthalmitis.32

Other untoward sequellae of strabismus surgery include:

1. subconjunctival cysts and corneal ulcers,[33,34]
2. pigmented episcleral mass from argyrosis,[35]
3. muscle inclusion cysts,[36]
4. amputation neuromas of eye muscles.[37]

In some cases, traction on the extraocular muscles during the surgical procedure, particularly with the patient under general anesthesia, causes activation of the oculocardiac reflex. This slows down the heart beat, sometimes causing irreversible cardiac arrest, which in turn can cause death or brain damage. There are various estimates of this scenario. Kirsch, Samet et al.[38] report it having occurred in 3,500 children undergoing strabismus surgery. Knoblock and Lorenz[39] report 60 deaths in 300,000 (one in 5,000) patients from strabismus surgery. Fifty-six had general anesthesia. Bietti[40] reports one death of an 8-year-old child from oculocardiac reflex during surgery for esotropia "out of nearly 2,200 cases operated on for strabismus" over a 10-year period. Bietti[41] feels the problem is so pressing that he recommends:

1. preoperative electrocardiography, both with and without a test for oculocardiac reflex (OCR),
2. the presence of an electric defibrillator and a pacemaker in the operating room during every ocular surgical procedure "involving either a pulling or a stimulation exerted on the ocular muscles."

Summary and Conclusions

Currently there exist opposing pharmacosurgical and refractive/visual therapeutic approaches to strabismus. The surgical technique produces immediate acceptable cosmetic results in some cases. In others, failure to achieve acceptable alignment necessitates additional surgical intervention, which causes further scarring of the extraocular muscles and additional exposure to the untoward side effects previously discussed. With the surgical approach, there is little expectation of functional binocularity, and the literature indicates that very little occurs. Published studies also report that the surgical approach affords poor long-term prognosis for continued alignment throughout life. Thus, in summary, the surgical approach in fact offers an immediate, abrupt change in deviation, hopefully toward alignment, with little chance of functional binocularity, poor long-term prognosis, and the chance of severe, harmful side effects, including death.

Vision therapy, combined with proper use of lenses and prisms, on the other hand, is a conservative, safe alternative, with good immediate and long-term prognosis for functional binocularity and alignment. Vision therapy has the single drawback of requiring sustained effort by the patient and parent over an extended period, generally six to 12 months, and continued compliance with the requirements of the therapy program.

In those relatively few strabismics where the angle of deviation is too large to secure alignment non-surgically, but where sensory fusion is achieved by prior vision therapy, surgery should be used as an adjunct to vision therapy to secure alignment. In those additional few patients where the individual does not respond to vision therapy because of temperament or anomalous neurological hook-up in the oculomotor system, surgery can be utilized as a cosmetic procedure to attempt alignment.

Although a definitive evaluation of cost effectiveness has not been performed, vision therapy has certainly not been shown to be more expensive than a surgical alternative for any given strabismic. Thus, it seems clear that the optometrist with the best interest of his/her patient in mind will utilize all available non- surgical techniques with the strabismus patient to achieve functional binocularity combined with ocular alignment. This provides, in a high proportion of treated strabismics, functional binocularity with long-term cure of the strabismus.

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Clarification of outcome goals for strabismic patients allows us to judge when surgery may be useful. Certainly we want to obtain the best possible visual acuity in each eye, establish sensory fusion, develop motor fusion across the field of view, and have the patient (and parents) satisfied with the cosmetic outcome. Thus, a full treatment plan may incorporate occlusion, active vision therapy, including amblyopia therapy, ocular motility and sensory fusion enhancement. Eventually motor fusion therapy is required to help align the eyes. Failure to obtain alignment will compromise sensory fusion, visual acuity improvement may regress, and the patient is likely to be disappointed with the final cosmesis. Prism prescription can certainly help, but if patients do not look good cosmetically they are not going to be totally satisfied regardless of the number of stereo rings they can perceive.

Surgery provides a treatment option aimed at functional or cosmetic outcomes. As a brief summary, surgery may: (1) permit better alignment of the eyes so that single, clear binocular vision may develop, (2) provide alignment in a critical position(s) of gaze in cases of incomitant deviations such as palsies, (3) permit better alignment in cases with compromised extraocular muscles, such as in thyroid myopathy or fibrosis, muscle contracture, or blowout fractures, (4) improve alignment and cosmesis in cases where the good eye must be held in check to "match the error" of the poor eye, such as in Duane's Retraction Syndrome. Medical treatment options are directed towards: (1) establishing better alignment of the eyes (miotics, Botox, and anticholinesterase drugs such as Mestinon), (2) preventing contractures in cases of recent palsy (Botox), (3) determining optical correction or encouraging plus acceptance (cycloplegics), (4) acting as an adjunct to amblyopia therapy (cycloplegic penalization). All of these options are examined below.

Surgical Treatment

Better Ocular Alignment

Optometrists often criticize surgery when it directly competes for cases with uniquely optometric treatment options. These include cases of divergence excess, intermittent esotropia, and perhaps congenital esotropia.[2] In fact, the two therapeutic approaches are not incompatible. Often surgery allows the eyes to obtain an alignment close enough to bifixation so that optometrists can intervene more effectively. Optometrists working with patients who have large strabismic angles may diligently attempt to establish sensory fusion, only to find that there is a severe limitation of techniques, especially home vision therapy, available for these large deviations. Prisms may help, but they also create distortions when the magnitude is over six to eight prism diopters per eye.[3] Mechanical reduction of the strabismus angle through surgery may permit the patient to use all the equipment and techniques available for visual efficiency (skills) cases. This allows more variety, which increases patient interest and makes it possible for patients to use home training equipment. Practical vision therapy must be viable over a diverse range of clinical skills as well as in diverse settings. Reducing a large angle strabismic to a small angle intermittent tropia or visual efficiency case makes it possible for more optometrists to treat and more patients to benefit. This is especially important for patients who must travel far distances to the optometric office, for practitioners in practices whose schedules do not permit regular, frequent follow-up visits, and for the working public who are unable to take off work often enough to comply with

in-office therapy. Imagine attempting to do divergence training on a constant esotrope of 40-60 prism diopters who is unable to keep office visits more frequently than once every two weeks. A surgical reduction of the angle of deviation to 10 prism diopters offers many more options for successful home therapy. If the angle is surgically reduced and sensory fusion is not attained, the patient still looks better than without surgery, and straighter eyes, in itself, may prevent social and psychological problems in the future.

Patients are sometimes warned that surgical overcorrection of the deviation resulting in a "consecutive" strabismus is a potential outcome of surgery. Overcorrection is probably less common with the use of adjustable sutures.[4] Nevertheless, it may not be such a fearful outcome in many cases. For example, young children with early onset esotropia may be easier to treat with vision therapy if slightly overcorrected, converting them into small angle or intermittent exotropes.

Palsies and Contractures

Vision therapy is most effective when the neural input to the extraocular muscles as well as the patency of the muscle tissue itself is intact. If the muscle is not getting neural innervation, or the muscle tissue has been modified by contracture or fibrosis, training becomes much more difficult. In other words, we do best when the problems are functional and are more likely to meet with difficulty when they are structural. Thus, patients with long-standing palsy, multiple sclerosis, myasthenia gravis, thyroid myopathy, progressive external ophthalmoplegia, etc. are not very good candidates for vision therapy alone. However, complete scope of therapy, including lenses, prism, surgery, and occlusion, certainly can help the patient obtain improved function and comfort.

Palsied muscles resulting in incomitant deviations which are still present six months following onset are very difficult to overcome through vision therapy alone. Recent superior oblique palsy is a good example. Patients will have particular difficulty in one horizontal position of gaze and in downgaze. When the deviation is above 10 prism diopters even prism prescription is ineffective, as the torsional effect of the muscles becomes very significant. The patient may develop a head tilt to help maintain fusion. Surgery can reduce the angle of deviation to an amount manageable by traditional optometric intervention.

Extraocular muscles compromised by fibrosis or contracture do not lend themselves to modification by standard vision therapy techniques. What can we do to help affected patients overcome their frequently constant and incomitant diplopia? If the goal is to establish binocularity in the patient's most important position of gaze, our options are prism or surgery. Depending on the amount of deviation, prism may be either very acceptable, or a source of distortions and poor cosmesis. If the effort to establish binocularity in one position of gaze is not successful, we may try to eliminate one image by occlusion or monovision correction. I have found surgery aimed at good alignment in primary gaze, combined with monovision correction, to be well accepted by patients for cosmesis and function.[5] The mechanical (i.e., surgical) correction allows fusion in primary gaze, and the monovision correction gives the patient one clear, easily identified image to attend to in the still diplopic positions of gaze.

Matching the Error

An unusual but effective use of surgery is to "match the error" in special cases of strabismus. For example, a patient with a left Duane's Retraction Syndrome may be fused in right gaze, orthophoric or slightly strabismic in primary gaze, and extremely strabismic on left gaze. This type of patient will often adopt a head turn to the left to maximize the area of fusion. One option to effectively extend this area and improve cosmesis is to surgically reduce the adduction of the right eye. This has the effect of forcing the patient to turn his head as he looks to the left, but both eyes will be aligned—and hopefully fused.

Medical Treatment

Miotics

Pharmaceuticals may assist in obtaining alignment of the eyes. Topical use of miotics such as diisopropylflurophosphate (DFP) or ecothiophate (phospholine iodide, PI) chemically alters the AC/A ratio by enhancing the ciliary body's response to small amounts of innervation. This permits an increase in accommodation without marked increase in convergence, thus yielding a lower AC/A ratio. Although I find bifocals to provide a larger reduction of the angles in cases of convergence excess with a high AC/A,[6] use of these drugs is an option with patients who do not tolerate spectacles well; when there is no need for correction at distance; and for use during summer breaks from school when children are playing outside and glasses, especially bifocals, often get in the way.

Systemic Anticholinesterase

Myasthenia Gravis presents a special challenge for the clinician interested in binocular vision. It is a condition that can mimic virtually any binocular anomaly. In addition, it is quite variable—especially in its early stages. It is desirable to stabilize the deviation prior to any definitive treatment. This may be attempted with pyridostigmine bromide (Mestinon), a systemic anticholinesterase drug. Unfortunately it is more effective with the systemic signs of myasthenia than with the ocular signs.[7] Nevertheless, it is a worthwhile attempt if the patient shows a positive response to intravenous edrophonium (Tensilon).

Botox (Oculinum, Botulinum Toxin)

Botox is a very dilute solution of botulinum toxin. It affects muscles by blocking cholinergic transmission and interfering with calcium metabolism.[8] Botox is injected into the stronger antagonist muscle in a strabismic eye, i.e., the medial rectus in an esotropic eye. Initially the eye will drift well out—into an exotropia. As the drug wears off the eye will come back in. As the visual axes become aligned, sensory fusion "glue" will help lock the eyes in alignment. This, of course, can be supplemented with sensory fusion techniques provided by the optometrist. The benefits of Botox injection are avoidance of general anesthesia, and no cutting of the muscles—thereby avoiding any transposition error when suturing. On the downside, it is an anxiety-producing procedure for the patient, and many do not like to endure weeks of exotropia and diplopia post therapy. Recently, Botox has been used to relax the antagonist muscle in recent palsy, thus avoiding the significant problem of secondary contracture.[9] Unlike patching schedules that depend on patients' understanding, memory (both often compromised by the same event which led to the paresis) and compliance, Botox is a compliance-free technique.

Cycloplegics

The use of cycloplegics to determine refractive error is particularly important with esotropes who may benefit significantly from plus lens prescriptions. Although some optometrists once feared that cycloplegia might "freeze" the ciliary muscle in place (thus not revealing true refractive error) rather than relax it, I do not think this concern exists any longer. However, after receiving the full cycloplegic prescription some patients cannot initially fully relax the ciliary muscle. Since full relaxation is imperative for clear distance vision with lenses and to take advantage of the AC/A ratio, this presents a management problem for the clinician. Allowing time for adaptation to the new prescription is usually the first step. Accommodative facility techniques can promote plus acceptance. Alternatively, the patient may be cyclopleged with atropine while wearing the new spectacles. If needed, a Fresnel press-on add may be used during treatment. As the atropine wears off, the patient has hopefully learned to stop fighting the effect of the plus lens correction and is able to respond positively.

Some cases of amblyopia are challenging simply because obtaining compliance with the occlusion or therapy schedules is so difficult. An option is to cycloplege the eye with better acuity (penalization) to force use of the amblyopic eye at near. An add may help the amblyopic eye if the initial near acuity is very poor.

Other Issues

Strabismus surgery has often been criticized for endangering life and vision (anterior segment ischemia). Much of the danger inherent in strabismus surgery was a result of the anesthesia. Recent improvement in monitoring blood gases during general anesthesia has further reduced this risk (which was very low to begin with—less than death resulting from dental extraction).[10] Anterior segment ischemia from strabismus surgery does not appear to affect children. In my experience it is a predictable sequelae to disinsertion of more than two adjacent recti muscles during a surgical procedure,[11] it is, therefore, avoidable.

Strabismus surgery also is criticized for being based on inexact approximations. Surgery performed on muscles under the effect of anesthesia is of necessity more art than science since the muscular tonus is very different from that in the conscious patient. The development of adjustable suture technique has reduced the impact of this problem.[4] This procedure, useful with older children and adults, completes the tie down of sutures the same day or the day following surgery. By adjusting the suture and checking the result with an alternate cover test (or subjective elimination of diplopia), the surgeon can establish alignment where he or she chooses on a conscious patient.

The recent, multi-centered prism adaptation test study has clearly demonstrated that presence of sensory fusion prior to surgery can greatly increase the likelihood of a positive long- term outcome.[12] While aware of the implications of this study, many strabismus surgeons seem unaware that sensory fusion "glue" can be enhanced in patients who do not respond on their own to the trial wearing of prism. Here, again, I believe the two professions have an opportunity to work together for the best possible results.

Many reviews of the usefulness of strabismus surgery take on a palpable "us versus them" tone. While there are clearly (and unfortunately) economic and political considerations (both conscious and subconscious) when evaluating available treatment options, an open-minded approach with the patient's best interest in mind may lead to advances in both healthcare disciplines. We must not be trapped into evaluating what strabismus surgery can offer in isolation, nor what vision therapy can do in isolation, but, rather, when appropriate, how much can each discipline provide for total patient care.

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