Ophthalmology® ROUNDS

SEPTEMBER/OCTOBER 2007
Volume 5, Issue 5

AS PRESENTED IN THE ROUNDS OF THE DEPARTMENT OF OPHTHALMOLOGY AND VISION SCIENCES, FACULTY OF MEDICINE, UNIVERSITY OF TORONTO

Adult Strabismus Part 1: Myths and Reality

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References to adult strabismus go back to ancient times. Egyptian statues dating from 2750 BCE and 1850 BCE reveal pharaohs with horizontal strabismus,[1] while Egyptian papyri from the 18th century BCE describe a misaligned eye and an eye with amblyopia.[1] Despite the long-documented history of adult eye muscle disorders and their treatment, several misconceptions are still prevalent among eyecare professionals. In addition, potentially helpful therapy for adults with strabismus, particularly surgery, may not be offered or may be delayed for an unnecessarily long time due to a lack of knowledge or awareness of its benefits on the part of the primary eyecare practitioner or the patient.[2]

This article is divided into two parts: Part 1, in this issue of *Ophthalmology Rounds* presents the prevalent myths about adult strabismus and the reality while, Part 2, in the next issue, will concentrate on treatment aspects. For the purposes of discussion, adult strabismus refers to strabismus in patients who are beyond the age of visual maturity, which is generally considered to be age 8 to 9 years.[3-6] This issue will hopefully help to "realign" the thinking and knowledge base of many practitioners and lay to rest any myths and misconceptions.

The myths

The following are commonly-held misconceptions about adult strabismus:

- It is almost always a condition that originates in childhood.
- Amblyopia is irreversible in adults.
- Adult strabismus is a cosmetic condition and, therefore, surgery and other treatments provide no functional benefit.
- The functional benefits of eye muscle surgery are not possible if amblyopia is present.
- Surgery for longstanding strabismus carries a high risk of double vision after the eye is realigned.
- The success rate for regaining stable alignment after surgery is lower in adults than in children.
- There have been few advances in the diagnosis and treatment of adult strabismus when compared to other fields in ophthalmology.
- Strabismus surgery in adults is beyond the skill set of general ophthalmologists and should only be undertaken by surgeons specializing in the field.

The reality

Does adult strabismus always originate in childhood?

Strabismus that manifests beyond the age of visual maturity can have its origin in infancy or childhood, but it can arise for the first time in older patients. In many cases, the misalignment was treated in the early years by nonsurgical or surgical means, but the eye turn either recurred or was not fully corrected. Surgery in childhood can yield a satisfactory result for many years, but control can deteriorate as a result of any number of factors, including changes in refraction, other eye procedures (eg, cataract or refractive surgery), temporary loss of vision in one eye, systemic illnesses, and trauma. On the other hand, patients from disadvantaged backgrounds or from countries where resources for eye surgery are limited, may not have had the opportunity to have their condition treated during childhood and may present for the first time as adults requesting correction of their eye turn.

Strabismus acquired in adulthood may be due to many causes, which can be subdivided into innervational and mechanical etiologies. Innervational disorders include cranial nerve palsies,

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both infranuclear and supranuclear, due to a host of causes, including strokes, head trauma, and systemic neurologic conditions such as myasthenia. Mechanical etiologies encompass any disorders in the orbit that restrict the eye muscles. These can be subdivided into abnormalities of the bone, muscles, soft tissues, and the eye itself. Bony causes include fractures and deformities associated with craniofacial syndromes. Examples of muscular etiologies are thyroid orbitopathy and scarring from prior strabismus surgery. The soft tissues can be disturbed by intraconal or extraconal space-occupying lesions or by diffuse conditions such as idiopathic inflammatory pseudotumour. Intrinsic eye conditions include very high myopia that can displace the globe within the orbit. Finally, there are iatrogenic causes of strabismus such as scleral buckle surgery and placement of glaucoma drainage implants.

Adults with strabismus that has persisted from childhood usually do not have subjective complaints. Suppression to eliminate diplopia that develops in childhood continues to act in the years after visual maturity is attained. In contrast, eye misalignments that recur after years of stability or arise *de novo* in adults often produce one or more of several symptoms, including:

- diplopia (the perception of the same image in two different visual directions)
- visual confusion (the perception of two different images in the same visual direction)
- asthenopia (various symptoms related to "eye strain" such as headaches, focussing difficulties, and impaired tolerance for near work).

Adults may adopt a compensatory head posture in order to maintain binocular fusion if the strabismus is incomitant and there is a region of binocular single vision (BSV) within their binocular field.

Is amblyopia reversible in visually-mature patients?

For decades, it was assumed that once a person reached visual maturity, the window of opportunity to reverse amblyopia was over. As a result, until recent years, it was not standard practice to offer traditional treatments such as patching and penalization to children older than 9 or 10 years or to teenagers who had vision loss resulting from refractive or strabismus causes. However, the approach to these "older amblyopes" completely changed after several studies published over the past 50 years documented significant recovery of vision in previously amblyopic eyes of older children or adults who were either treated for their amblyopia or lost vision in their better-seeing eye.[7] In one study, children aged >8 years gained as much improvement in vision as those aged <8 years, as long as they complied with therapy.[8] Another group demonstrated that children aged 9-15 years could attain marked improvements in vision with conventional therapy, including patching and wearing of glasses, even if they had not been previously treated for amblyopia.[9]

The most comprehensive study of older amblyopes was conducted in 2001-2004 by a large group of pediatric ophthalmologists in several North American centres under the acronym PEDIG (Pediatric Eye Disorders Investigative Group). They treated >500 patients, ranging in age from 7 to 17 years, with anisometropic and strabismic amblyopia with penalization or patching, along with daily periods of near work for the amblyopic eye. They found that 35% had improvement of ≥2 lines of best-corrected vision, with the same time course to recovery as children treated with these regimens.[7,10]

The cumulative evidence from these studies supports the premise that the adult visual system retains some neuroplasticity well into the third and fourth decades of life. Since one cannot predict which patients will respond to therapy, it is no longer acceptable to deny treatment to an amblyope of any age who may be motivated to improve the vision in their poorer eye. These studies also suggest that if any improvement is to occur, it will generally be evident within 4 to 6 weeks after starting treatment. Therefore, if no change is seen within this timeframe, the eyecare professional could consider discontinuing the therapy.

The discussion of this topic would not be complete without mentioning various oral medications reported to augment the effect of patching in reversing amblyopia in both children and young adults. Published studies have demonstrated that dopaminergic medications (eg, the combination of levodopa and carbidopa) may lead to improvements in vision in patients who do not respond well to therapeutic levels of patching or have very limited gains in vision.[11] European studies have touted the virtues of other compounds, such as citrulline, in improving visual performance in amblyopic eyes; however, other investigators have not found these agents to be as effective. At this time, there is no consensus on the indications for using these medications in the treatment of amblyopia in children or adults.

Is adult strabismus correction cosmetic or functional?

There is abundant literature to support the fact that realignment of the eyes in older children and adults is not cosmetic; rather, that it generates several functional benefits. However, the misconception that such treatment is purely a cosmetic exercise continues to pervade eyecare specialties, as well as the general medical community, and has also led to obstacles to reimbursement for surgeons performing adult strabismus surgery in some jurisdictions in North America.

The first step in acknowledging the virtues of adult strabismus therapy is a semantic one. The term "cosmetic treatment" implies a change from a *normal* situation to a *different,* but *still normal* one.[12] However, an eye turn is an *abnormal* anatomical and physiological state, and realigning the eyes *restores* the binocular situation to a normal or less abnormal one. The appropriate term for this correction is "restorative" or "reconstructive" therapy.[12,13] There is no doubt that realigning a misaligned eye generates a cosmetic benefit for the patient in terms of appearance but, the fact is, this change converts an *abnormal* situation to a more *normal* or natural one.

Surgery is often needed to correct strabismus in the visually-mature patient and the remainder of this section addresses the functional benefits of eye muscle surgery. However, it should be noted that there are several nonsurgical methods for restoring binocular function and good alignment. Both nonsurgical and surgical options will be reviewed in Part 2 of this article, in the next issue of

Ophthalmology Rounds. The benefits of "restorative" strabismus surgery can be analyzed according to 6 categories: eliminating symptoms; regaining binocular vision (fusion and stereopsis); restoring the static field of binocular vision; restoring the dynamic field of BSV; improving psychosocial functioning; and calculating the cost-effectiveness of adult strabismus surgery.

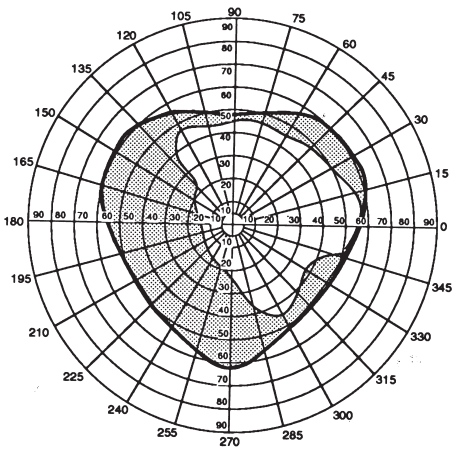
Eliminating symptoms: Two of the most troublesome symptoms experienced by adult patients with strabismus are diplopia and torticollis. The success rates of strabismus surgery in eliminating diplopia have ranged from 55% to 94%, with a mean of 71%.[14] Compensatory head postures can be caused by a variety of strabismus conditions and successful realignment surgery can eliminate torticollis in >80% of cases.[15] A Canadian study of 222 adults revealed a 76% rate of resolution of preoperative symptoms of diplopia, asthenopia, and ocular torticollis after surgery.[5] A study of 299 adult strabismus patients revealed that surgery led to significant improvements in nonwork-related tasks of daily living (walking, driving, reading, and leisure activities), job-related problems, and eye-related symptoms (including diplopia, eyestrain, and vision-related headaches).[16]

Regaining binocular vision (fusion): A large body of literature has confirmed that patients beyond the age of visual maturity have a high rate (in the order of 67%) of regaining binocular sensory fusion.[14] These studies utilized various tests of binocular function, including the Worth 4-dot test, Bagolini striated lenses, and Titmus or Lang stereotests.[3,6,17-19] Another important finding is that regaining fusion is not limited to patients who developed strabismus after the age of visual maturity. In fact, 50% of patients with childhood-onset strabismus regained sensory binocular vision following surgery during their adult years, whether or not they had undergone previous surgery during childhood.[3,18]

Many adult patients who undergo surgery and do not show sensory fusion on the conventional fusion or stereopsis tests postoperatively still demonstrate a long-term, small, stable motor angle result.[4] This suggests that motor fusion may play a role in stabilizing the result, even in the absence of demonstrable sensory fusion. In one study, motor alignment success was higher in patients who had strabismus onset after visual maturity than in those whose misalignment started in childhood (81% versus 63%).[14]

Restoring the static field of binocular vision: One of the under-recognized benefits of adult strabismus surgery is normalizing the static binocular field of vision or the "panorama" of the binocular field. This field can be measured using conventional instrumentation, including the Goldmann or arc perimeter, or computerized visual field testing devices. This benefit is especially dramatic for patients with esotropia. Several studies have confirmed that the horizontal extent of the binocular field is truncated by up to 30% on one or the other side as a result of crossing the eyes.[20] Losses of the static binocular field have been correlated with a higher risk of motor vehicle accidents.[21] Successful surgery for esotropia restored the panorama of binocular field to normal in >90% of patients in 2 series.[22,23] Expansion of the field in the vertical dimension is also possible after corrective eye muscle surgery on the vertical muscles.[24]

Figure 1: Plot of the dynamic field of binocular single vision (BSV) in a patient with a right fourth nerve paresis showing the region where the patient notes diplopia (shaded) and single vision (unshaded).



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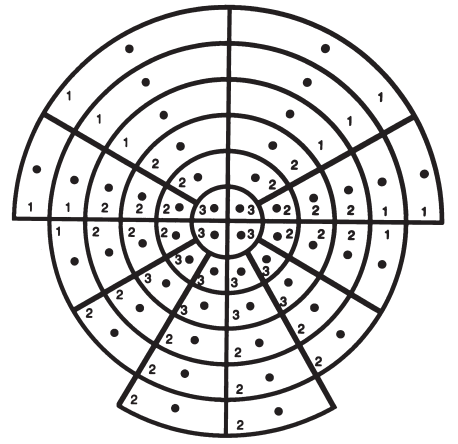
Restoring the dynamic field of binocular single vision: One of the most useful tests of disability due to strabismus is the dynamic field of BSV. The patient's field of fusion is plotted on a Goldmann or arc perimeter using either a spot or fusion target as the patient moves the eyes along meridians from the primary position into the periphery.[25,26] The plot provides a pictorial depiction of the area of single vision and the region of the field, in which the patient experiences diplopia (Figure 1). The field can be scored using a template that is weighted to the most important functional areas, namely the area within 30° of primary position and also the downgaze field (Figure 2).[25] Strabismus surgery that is optimally designed and performed can

Figure 2: Weighted scoring template for use with plot of the field of binocular single vision (BSV). The total number of points is 100. When the template is placed over the field in Figure 1, the BSV segments total 53 points, which is converted into a weighted single vision (fusion) score of 53%.



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lead to significant expansion of the range of the BSV field and improvement in the weighted BSV field score.[12,27,28] Expansion of a patient's field of BSV correlates with subjective feelings of improvement in activities of daily living.[25]

Improving psychosocial functioning: Many studies have addressed the social and subjective disabilities of adults with strabismus. Amblyopia and strabismus impact negatively on an adult's subjective visual function, well-being, self-image, interpersonal relationships, school work, and enjoyment of sports.[29,30] Up to two-thirds of adults surveyed with a history of eye turns reported adverse consequences at some time in their lives due to their eye condition.[30] Strabismus creates a serious barrier to gaining employment, with the impact being more severe for female than for male applicants, and worse for esotropia than exotropia.[31,32]

Successful alignment surgery can reverse many of these negative social and psychologic stresses.[30,33] In one study, patients whose eyes were straight after surgery had marked reductions in severity of several critical indicators of disability when compared to those who had noticeable residual deviations.[16] In another report of 31 adults who underwent surgery for longstanding horizontal strabismus, there were major improvements in several psychosocial indicators.[34]

Quantitating the cost-effectiveness of adult strabismus surgery: Several well-designed studies have applied cost-utility analyses to support the premise that adult strabismus surgery is cost-effective. A large prospective study indicated that there was a mean value gain of 2.61 quality-adjusted life years (QALYs) after strabismus surgery in adults and the cost-utility of the surgery is US$1600/QALY, both representing "very cost-effective" values.[35] Another group utilizing the standard visual function-14 (VF-14) questionnaire to assess the quality-of-life impact of several eye conditions found significant negative subjective and quantitative effects in adults with strabismus and amblyopia.[29] Finally, treatment for amblyopia has been shown to be very cost-effective when subjected to cost-utility analysis.[36]

Does amblyopia limit the functional benefits of adult strabismus surgery?

Although it may seem counterintuitive, the presence of amblyopia is not an impediment to gaining many of the benefits of strabismus correction. Restoring a static binocular field of vision is successful whether amblyopia is present or not.[2,23] In a large study of adults who underwent corrective surgery, amblyopia did not limit the potential of regaining peripheral fusion.[18]

Is there a high risk of diplopia after surgery?

There is a pervading myth among eyecare practitioners that there is a high risk of diplopia after realigning an adult eye. For this reason, many patients are unnecessarily denied the chance for surgery or alternative treatments to restore normal alignment.[2] The fact is, it is uncommon for patients to suffer intractable and prolonged diplopia after successful surgery. This premise holds true irrespective of the duration or the age-of-onset of the eye misalignment.[3]

A retrospective study in >800 adult patients undergoing surgery revealed that among those whose eyes were aligned to within a few prism diopters, the incidence of chronic diplopia in primary position was only 1.4%.[3] Two studies analyzed the risk of postoperative diplopia according to whether the patients did or did not appreciate diplopia when a prism was used to offset the ocular deviation at the last visit before the surgery. Among those who reported no diplopia with the prism, the risk of intractable diplopia after surgery was 0% to 3%. Even among patients who reported diplopia with the prism, the risk of long-term double-vision was still low, from 1% to 8%.[37,38] Other studies have confirmed a low rate of chronic diplopia after corrective surgery, ranging from 1% to 7% among patients who did not report diplopia preoperatively.[14] Although the risk of postoperative diplopia must be discussed with adult patients before surgery, it should not be overstressed when a patient is deciding whether to go ahead with a procedure that has many potential benefits.

Is the success rate of surgery in adults lower than that in children?

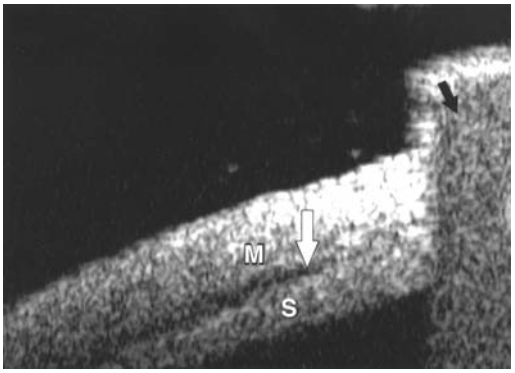
In adult strabismus surgery, the reported rates of success in reducing the angle of deviation to a few prism diopters, ranges from 70% to 92%, with follow-up of up to 10 years after surgery.[3,6,14,18,39] These rates are comparable to those for surgery in children. A recent prospective study demonstrated that the rate of successful realignment in patients above the age of visual maturity was similar to the rate for those below that age.[16] Furthermore, several studies have confirmed that the duration of strabismus does not limit potential improvements in eye alignment, panorama of vision, or regaining of fusion.[18,22,23]

What advances have occurred in adult strabismus?

There have been many developments in the diagnosis and treatment of adult strabismus in recent decades. These advances can be listed under the categories of basic science, diagnostic modalities, medical therapy, and surgery. Space limits this discussion to a few highlights under each heading; nevertheless, advances have been numerous and frequent in this field.

Advances in basic sciences: Recent anatomic and imaging work has defined and characterized the soft tissue pulleys of the extraocular muscles. This knowledge has led to new insights into the cause of various syndromes such as A and V patterns and Duane retraction syndrome.[40] As noted earlier, prospective studies of strabismic and anisometropic amblyopia demonstrated the potential to reverse vision loss in a substantial minority of patients who are above the age of visual maturity.[7] Finally, an enhanced understanding of the contributions of afferent proprioceptive signals from the extraocular muscles has led to advances in the treatment of nystagmus and other conditions.[41]

Figure 3: Ultrasound biomicroscopy (UBM) image of a previously recessed lateral rectus muscle. The shadow at the right (black arrow) is one arm of a caliper that helps to measure the insertional distance from the limbus. The muscle insertion is identified by the anterior end of the potential space (white arrow) separating muscle (M) from sclera (S). It was measured to lie 9.0 mm. from the limbus.



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Progress in diagnostic modalities: Advances in imaging the eye muscles and orbits have helped to elucidate the etiologies of many conditions. For example, dynamic magnetic resonance imaging (MRI) allows visualization of abnormal eye movements in disorders such as Duane syndrome.[42] Ultrasound biomicroscopy (UBM), a technology that is useful for anterior segment disorders, has been shown to localize eye muscles, including those that were previously repositioned (Figure 3).[43] Major strides have also been made in the technology of eye movement recording that allow better classification of nystagmus cases and improved accuracy in diagnosing various neurologic disorders that affect eye movements.[44] Finally, telehealth initiatives using wide bandwidth technology, have the potential to facilitate remote diagnosis of eye muscle conditions.[45]

Medical therapy advances: In the late 1970s, studies of botulinum A toxin led to its widespread availability as an alternative to surgery for the treatment of strabismus in both children and adults.[46] Recent, large, prospective studies confirm that pharmacologic penalization for amblyopia was as effective as patching in reversing vision loss in the first 4 months after starting treatment.[10] As noted earlier, oral medications such as L-dopa and citrulline may enhance the success of amblyopia reversal in some refractory cases.[11] Finally, the introduction of Fresnel prisms in North America in the 1960s allowed control of diplopia in many cases that could not be controlled by conventional glasses modifications, thus sparing patients the unsightly use of an eye patch.[47] Some of these alternatives will be discussed in more detail in Part 2 of this article.

Surgical advances: Many new surgical procedures have developed following new insights into eye muscle anatomy and physiology in recent years, including approaches to treating A and V patterns, cranial nerve palsies, and complex strabismus such as Duane's retraction syndrome.[47] New methods for treating ocular cyclotorsion have led to improved success rates for treating fourth nerve palsies and strabismus following scleral buckle and macular translocation procedures. Adjustable sutures have revolutionized the approach to both routine and complex strabismus correction in adults.[48] These advances and others will be discussed in Part 2.

Is strabismus surgery in adults out of the realm of the general ophthalmologist?

All ophthalmology programs in North America include basic minimum training in strabismus surgery as a prerequisite for graduation. Residents have to become proficient in the management of basic horizontal muscle procedures, and many programs include exposure to vertical muscle surgery. Once these skills are mastered, there is a very short learning curve to attaining the same proficiency in adult strabismus cases. It is also a small step to include adjustable sutures as a routine skill set of the surgeon, although the surgeon has to allot time later the same day or the next day to adjust the sutures. Surgery for complex strabismus and reoperations on previously operated muscles will likely continue to be done by strabismus specialists.

Conclusion

In the field of adult strabismus, the reality is that there have been many advances in recent years in basic science, diagnosis, and treatment, including confirmation that amblyopia can be reversed in many patients over the age of 8 or 9 years. The treatment of adults with strabismus includes several nonsurgical and surgical options that yield numerous functional benefits. Surgery for adults is restorative and highly cost-effective. Surgery is very successful in restoring satisfactory alignment, and a majority of patients have resolution of their preoperative symptoms with a low risk of complications, including diplopia.

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***Acknowledgment:** The author would like to thank Mrs. Frances Kraft and Drs. David Smith, Brian Arthur, and Michael Flanders for reviewing the manuscript and providing helpful suggestions. The author would also like to thank Carmelina Trimboli for technical assistance with the figures.*

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Upcoming events

Nov. 10-13, 2007 AAO, New Orleans, Louisiana (see below)

Nov. 23-24, 2007 Department Walter Wright Program
The Revealing Retina
The Old Mill, Toronto
Course director: Dr. David Chow

February 16, 2008 Toronto Cataract Course –
The Old Mill, Toronto
E-mail: help-OPT0801@cmetoronto.ca
Conf. website: http://www.cme.utoronto.ca

April 18, 2008 Pearls in Surgical Pediatric Ophthalmology
Hospital for Sick Children, Toronto
E-mail: help-OPT0803@cmetoronto.ca
Conf. website: http://www.cme.utoronto.ca

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Upcoming meeting

10-13 November 2007
American Academy of Ophthalmology
111th Annual Meeting
New Orleans, Louisianna
Contact: www.aao.org

Disclosure Statement: Dr. Kraft has no disclosures to announce in association with the contents of this issue.

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