Review Article

Intermittent exotropia: Surgical treatment strategies

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Surgical management of intermittent exotropias (IXTs) is ambiguous, with techniques of management varying widely between institutions. This review aims to examine available literature on the surgical management of IXT. A literature search was performed using PubMed, Web of Knowledge, LILACS, and the University of Liverpool Orthoptic Journals and Conference Transactions Database. All English-language papers published between 1958 and the present day were considered.

Key words: Intermittent exotropia, strabismus surgery, success rate of intermittent exotropias

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The management of intermittent exotropia (IXT) is controversial. This review will examine surgical aspects of IXT.

IXT is a common form of childhood exotropia, accounting for about 50–90% of all the exotropia and affecting about 1% of the general population.[1-4] There is a lack of longitudinal prospective study about this, and only a few retrospective studies[5,6] of untreated IXT are published.

The age of onset of IXT coincides with the age of visual maturation for children, which is between 3 and 6 years. The binocular function is often disturbed by the exotropia.[7] As a rule, during the phoric phase of IXT, the eyes are well aligned, and the patient may have a bifoveal fixation with excellent stereopsis ranging between 40 and 60 arc second. During the tropic phase, most patients will show large regional suppression of the temporal retina and anomalous retinal correspondence. A minority of patients with IXT may have the monofixation syndrome and do not develop normal bifoveal fixation with a high grade of stereopsis.[8] Some patients may even have a significant amblyopia.[9]

Clinical Presentation

IXT is an exodeviation controlled by fusional mechanisms. Exotropia is usually preceded by a phase of exophoria, and the deviation is most noticeable when children are tired, sick, inattentive or after long time near reading task. Adult patients may manifest exodeviation after imbibing alcoholic beverages or taking sedatives.[10] Patients may exhibit normal retinal correspondence when both eyes are aligned, but abnormal retinal correspondence when one eye is deviated.

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Patients with late onset exotropia, that is, after the ages of 6–7 years may experience diplopia because the exotropia occurs after the loss of visual plasticity. One symptom that deserves a special comment is the closure of one eye in bright sunlight. Bright sunlight dazzles the retina so that fusion is disrupted, causing the deviation to become manifest, and resulting in the closure of one eye. Monocular eye closure in sunlight is a mechanism used to reduce photophobia and is not related to avoidance of diplopia.[11,12]

IXT could progress to constant exotropia or remain stable.[13] In some cases, an exophoria can progress to an IXT that eventually becomes constant exotropia. Such deviation usually occurs first at a distance, but later at near fixation as well.[14]

Dissociated horizontal deviation is defined as a change in horizontal ocular alignment, unrelated to accommodation, that is, brought about solely by a change in the balance of visual input from the two eyes. It usually manifests as a spontaneous unilateral exodeviation or an exodeviation of greater magnitude in one eye during prism and alternate cover testing. Unlike in other forms of IXT, the observed exodeviation is slow, variable, and asymmetrical in the two eyes. In some instances, fixation with one eye evokes an esodeviation of the other eye during prism and alternate cover testing.

Etiological Factors

There are many etiological factors for exotropia. Abnormal anatomy of the extra ocular muscles, loss of fusional control

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and/or convergence insufficiency is the most accepted explanations. It is generally considered that the lens therapy-glasses or contact lenses alone is insufficient for myopia with IXT.[15-17] However, a study reports that implantable collamer lens restores ocular alignment and improves binocularity.[18] Strabismus affects stereoacuity development considerably, as its onset period is just around the key phase of bifixation development. In a study of 104 patients, it was found that only about half patients with IXT could acquire and keep static distance stereoacuity after surgery over a long period.[19] Timely surgical intervention can help restore and enhance binocular function.[20]

Indications for Surgery

Surgery is thought to be an effective method for the treatment of IXT.[3] Popular intervention criteria include a reduction in or loss of stereoacuity (at near or distance), deteriorating fusional control, a large angle of deviation, or a combination; but potential thresholds remain poorly defined.[21] The goals of the surgery for IXT are to restore alignment and preserve or restore binocular function. However, the optimal time for surgical intervention remains unclear. The critical point is the postoperative stereoacuity establishment. The Newcastle Control Score is a useful measure of the clinical severity of exotropia, can be used to serially assess improvement or deterioration and is a useful tool for the management of these patients.[22]

Nonsurgical treatment has been advocated, as has a single lateral rectus recession for small angle exotropia, but what constitutes a "small" angle and whether or not and how it should be treated remain unclear.[21] According to a study, the presence of the monofixation syndrome after surgery for presumed IXT most likely reflects the fact that it was present preoperatively, and the term monofixational exotropia is more appropriately descriptive for these cases.[23]

Time of Intervention and Goals of Surgery

Currently, most surgeons believe that early surgery for children is indicated to prevent progression to constant exotropia and restoration of bifixation, whereas in most adult patients surgery can only help to achieve orthotropia, but not bifoveal fixation.[24]

Researchers have proposed that patients may achieve a superior sensory outcome with motor realignment before age 7 or < 5 years of strabismus duration.[1,6] Others are of the view that the surgery needs to be postponed for several years because IXT patients can still keep IXT and hence bifixation can be obtained even if operated late and not all IXTs are progressive.[10] In some cases, the deviation may remain stable for many years whereas, in a few cases, it may even improve. Nevertheless, some researchers thought that patients might easily achieve binocular function if operated after age 7 and over 5 years of strabismus duration.[5,25,26] Favorable results after a surgical intervention before 2 years of age have also been reported by some authors.[27]

In IXT, one important indication for therapeutic intervention is the increasing tropia phase, as this indicates a deteriorating fusional control. The increasing frequency or duration of the tropia phase indicates the diminished fusional control and potential binocular function loss.[28] Progression should be monitored by documenting the size of the deviation, duration of manifest deviation and ease of regaining fusion after dissociation from the cover-uncover test.[15]

Researchers have proposed that distance stereoacuity as an objective measurement and indicated the time to intervene surgically.[29] In order to avoid diplopia and other symptoms, patients with IXT may develop anomalous retinal correspondence and/or suppression. Postoperatively, positive correcting exercises should be administered to restore normal retinal correspondence and enhance binocular function, such as the synoptophore exercise.

Classification and Choice of Procedure

Burian,[30-34] classified IXT based on distance/near differences and recommended different surgical procedures based on this classification. They defined the condition of patients in whom the distance deviation equaled the near deviation as a basic type of exotropia and recommended it be treated with a unilateral recess/resect surgery. A study by Kushner also suggested the same.[35] One study included in the Cochrane review, conducted by a single surgeon in the USA, compared surgery on one eye to surgery on both eyes in 36 children with the basic type of exotropia. Success was defined as no exotropia (or other strabismus) 1 year following surgery. The study found that surgery on one eye was more effective (82% success) than surgery on both eyes (52% success).[21]

Patients in whom the distance deviation exceeded the near deviation were considered to have a divergence excess type of exotropia; for such patients Burian recommended symmetric lateral rectus recessions. They based these recommendations on the assumption that bilateral lateral rectus recessions would affect the distance deviation more than the near deviation, and that a recess/resect procedure would affect the distance and near deviation equally. Scobee[36] observed that many patients with an apparent divergence excess type of exotropia would manifest an increase in their near deviation after 24 h of monocular occlusion. Subsequently, it was determined that approximately 1 h of occlusion was sufficient to elicit this increase.[33,34] And surgery should be performed for the largest angle measured.[37] Burian[30-34] defined the condition of these patients as a simulated divergence excess type of exotropia. Kushner further subdivided Burian's classification concerning the divergence excess pattern caused by an excess of divergence or by excessive accommodative convergence.[38] Because they believed that these patients really had a near deviation that equaled the distance deviation, but that it was masked by fusional vergences at near, they recommended that they be treated as if the patients had a basic type of exotropia and underwent recess/resect surgery. Another study suggested that patients with basic type IXT should be treated with recess/resect procedures, but patients with simulated divergence excess do well with lateral rectus recessions.[35]

Subsequently, other authors have described good results in treating patients with all 3 of these patterns of IXT with symmetric lateral rectus recessions.[39,40]

In the 18th annual Scobee Lecture, November 10, 1987, Dallas, Texas,[34] reported that patients with a simulated divergence excess exotropia (according to Burian's classification)[32] did as well as patients with a true divergence excess pattern if they were treated with bilateral lateral rectus recessions. However,

patients with a basic type of exotropia did not do as well if treated with bilateral lateral rectus recessions as patients with either true divergence excess or simulated divergence excess exotropia. A long-term (2 years) survival analysis of bilateral lateral rectus recession versus unilateral recess-resect for IXT concluded that although the surgical outcomes were comparable in both groups, the final outcomes were better in the bilateral recession group than in the recess-resect group. This may be caused by the difference of recurrence rate over time: Continuous recurrence of exotropia in the recess-resect group while recurrence was low after 6 months of bilateral lateral rectus recession.[41]

Bilateral lateral rectus recession has also been advocated for cases of so-called bilateral dissociated horizontal deviation or for unilateral dissociated horizontal deviation combined with exotropia.[42]

Weakening procedures on all four obliques which are overacting in large angle exotropia had been advocated, but later it was found that such overaction of all oblique muscles is often only an apparent one that may disappear after surgery of horizontal recuts muscles.[43]

Surgical Outcome

Due to lack of a standard definition for a successful outcome, variability in classification systems, multiple treatment approaches, and a paucity of long-term data it is difficult to determine the true outcome of currently available treatments for IXT. The success rate of IXT is dependent on the length of postoperative follow-up. Longer the follow-up higher the incidence of recurrence. In recent studies, the reported success rate in all types of IXT has been about 60–70%.[44-47]

Undercorrection is common after an initial bilateral lateral rectus recession with 21–31% requiring a second procedure.[39,48] Use of base in prisms of power greater than residual deviation has been advocated to provoke convergence and thus lessen the exodeviation.[49-51]

Surgical overcorrection varies according to different authors (6%,[39] 8%,[52] 10%,[30] 11%,[53] 17%[54] and 20%[55]) small degree of esotropia a watchful waiting is advisable. The second operation should not be performed until at least 6 months have elapsed, except when there are significant limitations of ductions that cause incomitance in lateral gaze.

A recent study[56] analyses the relation between age and surgical outcome in IXT. According to it, in younger patients (<7 years) in whom surgical dose was reduced, there was no significant change in success rate (77%), compared with those who had surgery using standard tables (75%). In older patients (>12 years) in whom surgical dose was increased, there was a statistically significant increase in success rate (80% vs. 41%). Thus, modifying the surgical dose according to age can improve the success in patients with IXT.

Conclusion

The preoperative deviation is one of the strongest predictors for the favorable surgical outcome.

Surgery with preoperative orthoptic/occlusion therapy had the highest success rates. Surgery with orthoptic/occlusion therapy was more effective in reducing exodeviation (prism diopters per millimeter of horizontal rectus surgery), compared with surgery alone.

Strabismus surgery can help to preserve or restore the binocular vision in IXT. Children receiving the surgery at young ages might develop better postoperative binocular vision, and the postoperative synoptophore exercise/amblyopia therapy can help to restore the binocular vision.

Short-term studies with 6 months to 1-year follow-up reports a success rates of approximately 80%, whereas studies with 2–5-year follow-up have shown a 50–60% success rate with one surgery. In most of these reports, success was defined as alignment within 10 prism diopters of orthophoria, and mean follow-up was no >4.5 years.

However, literature review suggests that more studies will help us understand the pathogenesis of the IXT and hence understand the behavioral pattern of the disease, which in turn will help us treat it better. Surgical intervention based on the stereopsis, Newcastle scores, and the fusion amplitudes give good results in terms of long-term success of the surgery. Early detection of abnormal stereoacuity for distance and near fusional vergence amplitudes may help to decide the proper timing of surgery in IXT. Thus, there is a need for robust clinical trials to improve the evidence base for the management of this condition.

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Conflicts of interest

There are no conflicts of interest.

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