Surgical Considerations in the Treatment of Intermittent Exotropia: A Review

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ABSTRACT

The surgical management of intermittent exotropia has been subject to wide discussion and debate in the literature. In general, recommendations for best practice guidelines for the surgical management of this condition are not possible due to the conflicting finding of various studies and a lack of high quality evidence. The main issues concerning surgical intervention relate to the timing, type and amount of surgery. This paper presents a review of the current literature with a particular focus on these three issues.

Keywords: intermittent exotropia, divergence excess, surgery, treatment

INTRODUCTION

Exotropia (XT) is a manifest deviation with a temporal or outward misalignment of the visual axes. Intermittent XT has a manifest phase at which time the deviated eye is said to be suppressed or the individual experiences diplopia; whilst during the 'controlled' phase, the eyes are straight with binocular single vision (BSV) being present.[1] Although intermittent XT can be used to describe any XT that is not constant, it commonly refers to one that is manifest at distance fixation with or without a manifest phase for near.[2] Intermittent XT is the most prevalent exodeviation[3-6] with a higher rate in females.[4, 7]

Management of intermittent XT can be surgical and or non-surgical. Various non-surgical management options include minus lens therapy, prisms, fusion exercises, alternate patching and observation.[8, 9] The surgical options include bilateral lateral rectus recessions or unilateral medial rectus resection and lateral rectus recession. Management of intermittent XT is aimed at reducing the manifest deviation whilst aiding sensory fusion so that normal binocular alignment and BSV can be achieved.[10]

Although the natural history of intermittent XT is relatively unclear[3] and with few reports of patients with smaller angle deviations benefiting from non-surgical management,[3, 8, 9, 11] many view it as primarily a surgical condition when treatment is required. Chia, Seenyen and Long[8] recently reported that half of those who present with intermittent XT eventually have surgery. However, the indications for surgery are not well defined or generally agreed upon. Despite this, surgical intervention is usually or perhaps often warranted when the deviation is manifest for over 50% of the time or an increase in size is noted with concerns that this may lead to the disruption of binocularity[2]. Parental concerns regarding the cosmetic appearance also play an important role in the decision leading to surgery.[12]

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SURGICAL TREATMENT

There is widespread opinion as to the most appropriate surgical intervention for intermittent XT, with the literature reflecting various conflicting findings. The three main issues concerning surgical intervention are: (i) the age at which surgery should take place, (ii) the type of surgical procedure and (iii) the amount of correction.

A significant shortcoming in the literature is the inconsistent definition of a successful outcome between studies. A successful outcome can range from a result within 10 prism dioptres (pd) of the ortho-position, or within 8 pd of ortho to merely no manifest deviation. Some studies also include post-operative sensory status in determining the success of surgery. Adding to this are the different lengths of follow up.

This is of particular concern given that intermittent XT often has a tendency to recur with time.[13, 14] Further, some studies have also included patients with a constant XT[15, 16] or patients with A/V patterns[14, 15, 17] and lateral incomitance[12]. Constant XT, although similar to intermittent XT, is a different type of exodeviation as there is a total loss of BSV. In addition, the effects of A/V patterns and lateral incomitance on final horizontal alignment are not well established[18] with some studies suggesting that small vertical imbalances decrease chances of a successful result.[17]

EARLY VERSUS LATE SURGERY

The age at which surgery should be performed remains controversial. Whilst there are studies demonstrating the advantages of operating at a younger age,[15, 17] there are those in favour of delaying surgery till later.[1, 12] Conversely, other studies suggest that age has no effect on final outcomes.[13, 14, 19-23] Further adding to the uncertainty is that there is no agreement on the age range that qualifies as early or late surgery.

Advocates of early surgery believe it is important to 'break the habit of the deviation' so it is not reinforced over a longer period of time and to prevent intractable sensory anomalies (i.e. temporal hemi-retinal suppression).[1, 15] However, as stated earlier, the cut-off age distinguishing between early and late surgery differs between studies making it difficult to draw direct comparisons.

In a retrospective study, Pratt-Johnson, Barlow and Tillson[17] reported that individuals who underwent surgery before 4 years of age had a better chance of a successful outcome after a 1 year follow up. Abroms, Mohney, Rush, Parks and Tong[15] also advocated early surgery reporting better sensory outcomes in individuals who received surgery before 7 years of age and before 5 years onset of the intermittent XT. Though often ignored, onset may also be an important factor to consider when investigating differences between early and late surgery. Age alone does not relate to the duration of the XT and the deterioration of control over time.

Asjes-Tydeman, Groenwoud and van der Wilt[16] similarly found that patients who received surgery before the age of 7 had a better chance of a successful result in terms of ocular alignment and sensory outcome. However, in this study patients operated on after 7 years demonstrated constant deviations and on average larger deviations. In addition, the majority of patients in the older group received unilateral surgery, whilst the majority of those in the younger group received bilateral surgery.

The relative risk of operating on a younger child is namely the possibility of developing amblyopia and monofixation syndrome owing to sensory adaptations through cortical plasticity.[1, 17] Pratt-Johnson et al,[17] for instance, reported that individuals who underwent surgery prior to 4 years of age were more likely to develop monofixation and amblyopia. Edelman, Brown, Murphee and Wright[24] followed patients with consecutive esotropia and also found that those under 6 years of age were more likely to lose stereopsis and develop amblyopia. This has often been considered a reasonable argument for delaying surgery.

On the other hand, Richardson[12] found that individuals receiving surgery after the age of 6 had a better chance for a successful outcome with BSV at near and distance. Richardson[12] believed that this was owing to the more precise measurements possible in older children, which subsequently reduced the risk of consecutive esotropia. In support of this rationale, Chia et al[8] reported that children under 5 years of age were much more variable in their measurements between visits.

Pre-operative orthoptic treatment[8, 10] is also better utilised in older patients[12] and is viewed as another reason to delay surgery.[1, 12] For instance, Figueira and Hing[10] reported that patients in their study who had orthoptic treatment combined with surgery had a better chance of a successful outcome compared to those who received surgery alone.

Adding to the conflicting findings of these studies are the several papers that have suggested that age is not a factor for a successful outcome.[13, 14, 19-23] Theses studies have reported that there is no correlation between age at surgery and final outcome. It is also noteworthy that Stoller et al[13] found age at surgery is not predictive of success when using a retrospective 'survival analysis' where 'survival time' was considered to be the time from surgery to recurrence.

Although this paper has focused age at surgery, it is worth noting that many have suggested that control rather than age may play a greater role in success.[13, 15, 25, 26] In most studies, loss of control is defined as deterioration in magnitude or frequency as perceived or observed by the parents or the examiners. Stoller et al[13] and Abroms et al[15] found better outcomes when patients were operated on whilst their deviation was partially controlled. Similarly Jeoung, Lee and Hwang[25] reported that all cases of overcorrection had poor pre-operative stereopsis; stereopsis being the measure of control.[26] Indeed in all of the studies, surgery was only indicated on the basis of either the parents' or examiners' concern of the deterioration of the intermittent XT in either deviation size or frequency.

Current studies investigating the effect of age on surgical outcome are based on retrospective studies which have inherent bias or flaws. As indicated by Stoller[13], a major draw back of using a retrospective analysis is the inclusion of cases that are lost to follow up. Outcome is judged by the last visit and therefore late failures could be counted as successes and vice versa. In addition, no study has yet randomised the age of surgery. However, a randomised controlled trial (RCT) for early versus late surgery may be difficult given that ethical issues play a large role in the decision on when

Table 1. Studies using bilateral lateral rectus recession surgery

Study	Study Size	Follow Up	Success
Pratt-Johnson et al 1977	100	1 year	81% good cosmetic result,
			41% good sensory result
Stoller et al 1994	57	At least 1 year	58% good cosmetic result,
			no sensory criteria
Ruttum 1997	60	6 months	63% good cosmetic result
			no sensory criteria
Ing et al 1999	52	6 months	61.5% good cosmetic result
			no sensory criteria
Abroms et al 2001	76	6 weeks	67% good sensory result

to operate. As an example, an examiner may not be able to justify delaying surgery in a 2 year old patient who has a large and frequently manifesting intermittent XT and is at threat of losing BSV. Overall, the evidence for the influence of age on surgical outcomes is, at best, still inconclusive. With conflicting findings from retrospective studies and the lack of RCTs, no recommendations as to the most appropriate age for surgery can be safely made at the present time.

TYPE OF SURGERY

Several studies have compared the effectiveness of different types of surgery for intermittent XT. In many studies bilateral lateral rectus recessions (BLRR) has been the procedure of choice for intermittent XT.[13, 15, 17, 18, 20, 22, 27] However, this procedure has produced varying success rates, as detailed in Table 1. There is also the suggestion that the type of intermittent XT[1, 28] should influence the chosen surgical procedure. For example, some authors believe that true distance intermittent XT (distance angle is largest) should receive BLRR whilst a simulated distance XT (distance angle largest due to a high AC/A ratio) or a basic XT (deviation same for all distances) should receive unilateral surgery.[1]

Chia, Seenyen and Long[29] found a higher success rate for unilateral surgery compared to bilateral surgery at 1 year follow up. Their study group contained a large proportion of divergence excess intermittent XT patients where the type of surgery received was not determined by the type of intermittent XT. However, at the 3 year follow up, a greater proportion of patients with a successful outcome in the unilateral surgery group demonstrated an exotropic drift at distance as compared to those who received bilateral surgery. Despite this, the unilateral group still had a slightly higher success rate at 3 year follow up. The few prospective randomised control trials in this area[25, 30] have also reported greater success with unilateral surgery. Kushner[30] for instance compared bilateral and unilateral surgery in a group of patients who demonstrated a basic intermittent XT and found that the unilateral surgery group had a higher success rate. Similarly, Jeoung et al[25] found a significantly higher success rate with unilateral surgery for basic intermittent XT.

In contrast, Lee and Lee[31] reported no significant difference between the two surgical procedures for either basic or simulated distance intermittent XT. Similarly Maruo et al[14] found no difference in success rates between bilateral and unilateral surgery at 1 month. However, in concordance with Chia, Seenyen and Long[29], at 4 years, Maruo et al found that the success rate of the in the unilateral surgery group dropped considerably more than that of the bilateral surgical group.

While the majority of studies have compared BLLR with unilateral surgery, there have also been reports of unilateral lateral rectus recession or single muscle surgery being effective in a small number of patients with moderate size intermittent XT with no reports of post operative lateral incomitance.[32-34]

Agreement for best practice recommendations for the surgical correction of intermittent XT is still lacking. Many retrospective studies investigating the effectiveness of bilateral rectus recessions on intermittent XT have produced varying success rates[13, 15, 18, 20, 22, 27, 35] and the prospective studies[25, 30] have focused primarily on basic intermittent XT. A trial randomising surgical procedure for all types of intermittent XT is needed to further investigate this issue.

FULL CORRECTION VERSUS OVERCORRECTION

The postoperative outcome of surgery with regards to the amount of planned correction has also been widely debated. Many have suggested that overcorrection is necessary because of the postoperative exotropic shift that patients with intermittent XT display.[21-23, 27, 36-38] Even though these studies agree that initial overcorrection is needed, they vary in the amount they recommend. This is highlighted and summarised in Table 2.

There have been a number of retrospective studies investigating the effect of the initial postoperative correction

Table 2. Studies recommending overcorrection

Study	Study Size	Follow Up	Amount of overcorrection	Success rate
Clarke and Noel 1981	78	No minimum	10 to 15 pd	42%
Beneish and Flanders 1994	67	6 months	Any amount of esodeviation	60%
Ruttum 1997	60	6 months	Less than 10pd	63%
Kim et al 2005	68	6 months	20pd	71%
Oh et al 2006	365	No minimum	Over 10 pd	60.3%
Koo et al 2006	199	1 year	Success group had larger overcorrection	60%

on long term alignment. Beneish and Flanders[36] reported that patients with a consecutive esodeviation at 1 week or 1 month had better final outcomes, similar to Lee and Lee's[30] who also reported that initial overcorrection produces better long term outcomes. Likewise, Koo et al[21] found that a greater proportion of patients in their successful group had an initial overcorrection – 86% of patients in the success group showed initial esodeviations compared to 62% in the fail group.

Souza-Dias and Uesgui[38] also reported that an esodeviation of 10pd was the most desirable postoperative immediate outcome. Any lesser overcorrection would increase the chances of recurrence and any more would increase the chances of a longstanding consecutive esotropia. Clarke and Noel[37], also in favour of overcorrection, found that individuals who were overcorrected by 10–15pd had better results than those who demonstrated orthophoria, exophoria or small angle esophoria immediately after surgery. Similarly, Oh and Hwang[23] found that those with the highest chance for success were those who were initially overcorrected by over 10pd. Ruttum[22] reported that those initially corrected from orthophoria to 9pd of esotropia had the best success rates. In contrast Kim, Kim and Hwang[27] reported that 71% of patients with an immediate post-operative overcorrection of at least 20pd demonstrated successful results of within 10pd of orthophoria on follow-up visits.

In the aforementioned studies it is not clear whether the overcorrections were planned. Exact correlations between the immediate post-operative alignment and final long term alignment have also yet to be established, even in the studies that recommend initial overcorrection.[21-23, 27, 36, 37] The other consideration is the risk of monofixation syndrome and amblyopia[17, 24] which can result from persistent consecutive esotropia. This issue was raised by Kim et al[27] and by Edelman et al[24] who had a re-operation rates for consecutive esotropia of 5.9% and 11% respectively.

Another issue concerning the amount of planned correction relates to the variable size of the exodeviation during different testing conditions. As suggested by some, a testing distance of beyond 6 metres or measurement after monocular occlusion may reveal a greater angle of deviation in patients with X(T).[1, 18, 35] In an RCT, Kushner[18] found that many patients exhibit larger deviations when they fixate beyond 6 metres or after 1 hour of monocular occlusion. In this study he randomised patients to either receive surgery for the traditional 6 metre distance or the larger angle found. The group receiving the surgery for the larger angle had a success rate of 86% compared to the 6 metre group of 62.5%. Similarly, Kim and Hwang[35] found this approach to be effective and safe with only 9% of patients who had a surgical correction for their 'largest angle' requiring a postoperative re-operation. These findings may explain the exotropic drift seen when surgery is planned for the smaller 6 metre angle. According to Kim and Hwang[35] and Kushner's[18] findings an undercorrection is potentially achieved when aiming for full correction of the XT as measured at the 6 metre distance.

No randomised trials comparing full correction and overcorrection are yet available. With little still known about the nature of exotropic shift, it is difficult to suggest the amount of overcorrection necessary, if indeed necessary at all. Until there is stronger evidence available overcorrection or persistent consecutive esotropia are still an issue.

CONCLUSION

With conflicting reports and the majority of evidence based on retrospective studies, the most appropriate surgical management for intermittent XT remains somewhat elusive. Lacking at present are prospective RCTs that would provide the stronger evidence required. Agreement on the definition of early surgery and of a successful outcome is also needed, so that future studies can have a common aim and be comparable. Further, these future studies need to include appropriate follow up periods to effectively analyse long term postoperative stability. With reports of increasing numbers of intermittent XT in certain populations[4] and the possible consequence of monofixation syndrome and amblyopia with unsuccessful treatment, it is important that more investigation on surgical treatment take place so that the best practice guidelines can be developed for the management of intermittent XT.

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