Intermittent Exotropia: Perspectives on Management

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Summary: The Orthoptist’s Role

For the treatment of intermittent exotropia, conventional thinking assigns sensory treatment to the orthoptist and motor treatment to the ophthalmologist. However, upon close examination of this line of thinking as discussed in this paper, one can see that the margins are blurred. It is the orthoptist that provides the measurements that are used by the surgeon to determine the surgical procedure and the amount of surgery. Clearly, the orthoptist is involved in the surgical decision-making process. We therefore have not only an opportunity but an obligation to our patients to expand our involvement by applying measurement strategies that provide the surgeon with the best information about the size of deviation and to do more research in this area. We also must expand our thinking to investigate features that might help predict response to standard surgery, enabling the surgeon to augment the procedure or the amount of surgery to improve surgical outcome when particular features are present pre-operatively. We cannot assume that it is random chance that the exact same surgical procedure for patients with the exact same measurements can result in a cure for some but to overcorrection or under-correction in others. Orthoptic research can help identify reasons for this variation in response to surgery.

Orthoptists have been tremendously successful in devising methods for non-surgical management of intermittent exotropia that include techniques for breaking suppression and building fusional convergence. Our role should not be limited, however, to treatment of the sensory anomalies of intermittent exotropia, but rather should include application of our knowledge, skills and research abilities to surgical planning for treatment of the motor component of intermittent exotropia. By expanding the involvement of orthoptists, the orthoptist/ophthalmologist team will be better able to cure intermittent exotropia. In the meantime, however, as we strive toward improving cure rates, we can be encouraged by the knowledge that treatment "failure" does not necessarily equate with patient dissatisfaction. In a review of charts of 69 consecutive patients in my practice that had surgery for intermittent exotropia, some also treated with orthoptics, 39 were not cured and were considered treatment failures at their most recent visit with failure defined as intermittent or constant tropia of any size or a phoria greater than 8’ at distance or near. In spite of being categorized as treatment failures, sixty-two percent of those 39 patients were happy with their outcome, unaware of any manifest deviation and asymptomatic. Therefore, obtaining a cure is not necessarily a requirement for patient satisfaction. As orthoptists we can have pride in our past
clinical and research accomplishments that have brought us methods for eliminating symptoms associated with intermittent exotropia. We must, however, continue to move forward, continue to learn, continue to explore. We can improve surgical success, we can render more of our patients asymptomatic and we can cure intermittent exotropia.

**Introduction**

The orthoptist's role in strabismus evaluation and management primarily involves sensory and motor assessment along with orthoptic therapy when indicated. The patient with intermittent exotropia is probably the most ideal patient for successful orthoptic therapy. Orthoptists have developed a wide armamentarium of treatment modalities aimed at building convergence amplitudes and breaking suppression. Studies in this area support the success of orthoptic therapy in improving the control of exodeviations and reducing symptoms. Those same studies also indicate, however, that orthoptic therapy does not cure the exodeviation, with rare exceptions. That is, although orthoptic therapy helps control the exodeviation, it does not eliminate the exodeviation. Therefore, for a variety of reasons, many patients with intermittent exotropia eventually undergo muscle surgery for their intermittent exotropia, including some who at one time were well managed with orthoptic therapy. In contrast to the abundance of literature on orthoptic therapy for intermittent exotropia, the orthoptist's role in surgical planning for intermittent exotropia has received little attention. Perhaps, in part, this is because orthoptists feel that their therapy has failed and that their role in the management of the patient is finished when the decision is made to have surgery. It is important to recognize, however, that surgery and orthoptics treat different aspects of the pathology in intermittent exotropia. Orthoptic therapy involves breaking suppression and building fusional convergence. Surgery involves correcting the misalignment. Surgery alone or orthoptics alone does not treat the entire problem. Either treatment alone does not cure the condition. Therefore, it is perhaps more accurate to view surgical treatment as another step in attempting to cure the condition by eliminating the motor component of the problem (the misalignment) rather than as a treatment offered when orthoptic treatment "fails". But does surgery eliminate the motor component of the condition? This paper will discuss surgical failure and possible reasons for surgical failure with emphasis on the need for greater involvement in surgical planning by the orthoptist.

**Background**

Success rates for one surgical procedure for intermittent exotropia have remained at a disappointingly low level throughout the past several decades (Table 1). The definition of "success" varies from one study to another with most definitions falling short of what most orthoptists would consider a cure. If strict criteria were applied to define "success", the success rates in many of the studies in table 1 would be even lower than reported. Studies that examine the results of surgery combined with orthoptic therapy show that orthoptic therapy can improve success rates, however they do not demonstrate that combining the therapies ensures treatment success and prevents recurrence of the deviation. The majority of surgical
treatment failures are due to under-correction or recurrence of the deviation. As seen in table 1, ophthalmologists have examined approaches to surgery for intermittent exotropia for over 50 years without significant improvement in surgical success rates. Orthoptists, however, have not been very involved in research in this area. In the period from 1993 to 2003, the 11 issues of American Orthoptic Journal contained only 3 papers for which orthoptists were the first author with strabismus surgery as the main topic. Over the same 11 year period, orthoptists were the first author of only 8 articles with strabismus surgery as the main topic published in the Japanese Orthoptic Journal. Perhaps if orthoptists get more involved in the surgical aspect of treatment, new insights will emerge that will have a positive impact on the outcome of surgical treatment of our patients. What is the role of the orthoptist in improving surgical success rates? To address that question, we must examine possible causes for surgical under-correction / recurrence and explore ways in which the orthoptist can participate in surgical planning with the goal of improving surgical success rates.

**Possible causes for surgical under-correction / recurrence of intermittent exotropia**

Surgical under-correction and post-operative recurrence of intermittent exotropia could be a result of a combination of factors. It is possible that the surgeon is not operating for the appropriate target angle, is doing an inappropriate procedure, or is not doing enough millimeters of surgery. It is also possible that etiologic factors are involved. We will take a look at each of these possibilities along with the role of the orthoptist in helping the surgeon identify the best surgical approach.

**Etiologic factors**

It is necessary to consider that it is simply the nature of the condition for the eyes to continue to diverge is spite of appropriate surgery and orthoptic treatment. It has been suggested that an increase in electrical activity of the lateral recti is associated with the exotropic drift when fusion is interrupted. Research by Jampolsky provided strong evidence for subcortical innervation favoring the lateral recti in normal subjects. Perhaps in

<table>
<thead>
<tr>
<th>Author</th>
<th>Year published &amp; Success rate</th>
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<tbody>
<tr>
<td>Cooper</td>
<td>1955 71%</td>
</tr>
<tr>
<td>Folk</td>
<td>1956 55%</td>
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<tr>
<td>Dunlap &amp; Gaffney</td>
<td>1963 25%</td>
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<tr>
<td>Burian &amp; Spivey</td>
<td>1965 55%</td>
</tr>
<tr>
<td>Pratt-Johnson et al</td>
<td>1977 41%</td>
</tr>
<tr>
<td>Hardesty et al</td>
<td>1978 57%</td>
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<tr>
<td>Richard &amp; Parks</td>
<td>1983 56%</td>
</tr>
<tr>
<td>Stoller et al</td>
<td>1994 58%</td>
</tr>
<tr>
<td>Ing et al</td>
<td>1999 62%</td>
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subjects with intermittent exotropia, there is a (genetic) predisposition for tonic innervation to the lateral recti beyond that of normal subjects. This tonic innervation could cause recurrence of the exodeviation. Other investigators have demonstrated that there is an adjustment in ocular alignment that occurs after birth. In a study by Nixon that included 1,219 newborns, it was found that fewer than half were aligned.15 Perhaps the mechanism responsible for bringing the eyes to an aligned position involves an "adjustment" in tonic innervation to the extraocular muscles. If the mechanism for this adjustment is faulty in subjects with intermittent exotropia, this could result in chronic increased innervation to lateral recti and lead to intermittent exotropia and be responsible for recurrence of the exodeviation post-operatively. Guyton recently presented a discussion of control mechanisms involving vergence tonus, disparity feedback and muscle length adaptation and their possible role in the cause and progression of strabismus.16 Perhaps his theory can explain why recurrence of exodeviations occurs in so many patients.

If indeed there is a continuous force driving the eyes to a divergent position in intermittent exotropia, it clearly does not render the condition beyond repair as evidenced by those patients who are successfully treated. However, further basic science research by orthoptists could give us a better understanding of the etiology of intermittent exotropia and lead us to new treatment schemes.

Inappropriate target angle

The likelihood of surgical under-correction is greater if the pre-operative measurements do not reflect the full angle of deviation. In many practices, it is the role of the orthoptist to obtain pre-operative measurements. Standard alternate prism and cover test measurements can fail to elicit the full exodeviation, especially if the patient has recently undergone a period of occlusion for anti-suppression or has had rigorous convergence training. Studies have shown that standard measurement techniques might not bring out the full deviation.

Dadeya advocates using prism adaptation to determine the surgical angle.17 In his study, he had an 89% surgical success rate in a group of intermittent exotropes operated for their prism adapted angle compared to a 54% surgical success rate in intermittent exotropes operated for the original angle obtained by standard cover testing.

Kushner conducted a study in which distance measurements were obtained three ways: with a fixation target at 6 meters; with the patient viewing out a window; and at 6 meters following 1 hour of monocular occlusion.18 The subjects were randomly assigned to have surgery for the angle measured at 6 meters (without a period of monocular occlusion) or for the higher angle obtained with the other methods. He reported an 86% success rate with surgery for larger angle (obtained by viewing out a window or following monocular occlusion) and a 63% success rate with surgery for original angle obtained at 6 meters without monocular occlusion.

The measured size of the deviation can also be influenced by the target used. A study by Mehta et al found that Snellen letters are superior to video cartoons or animal toys.19 As orthoptists, we need to be aware of the importance of measuring the full exodeviation as we evaluate these patients pre-operatively and consider utilizing strategies that research suggests has superior results. As researchers, we should explore additional measurement
techniques, their applications, and of course, the impact of new measurement techniques on surgical results.

**Inappropriate surgical procedure**

In most practices, the orthoptist does not make the decision regarding surgical procedure. However, the orthoptist is indirectly involved in the determination of surgical procedure. There is a relationship between target angle and procedure. The surgeon bases his/her decision regarding surgical procedure on the size of the deviation (how many millimeters of surgery) and on the classification of the deviation (distance vs. near; "A" or "V" pattern, lateral incomitance). Therefore, the orthoptist’s role in the determination of surgical procedure lies in the measurements that he/she obtains for the surgeon. Here again, standard measurement techniques might not be sufficient.

A study by Neikter demonstrated that dissociated vertical deviations, A and V patterns, vertical deviations and oblique dysfunction can be more easily diagnosed following Marlow occlusion. Veronneau-Troutman found that prism adaptation could produce significant changes in "A" and "V" patterns. The effect of prism adaptation on distance-near disparity was also investigated by Ohtsuki. Patients with distance-near disparity wore press-on prisms to neutralize the smaller deviation. Prism was added at 15-30 minute intervals until the angle stopped building in size. Of 128 subjects in the study, 24 changed classification after prism adaptation (Table 2). The study also showed that surgical success was significantly better for those subjects that changed classification than for those who remained in the original corresponding class (Table 3). Therefore, not only can prism adaptation influence the size of the target angle as described earlier in the study by Dadeya, but it also can influence the surgeon’s choice of procedure by clarifying the classification.

Kushner also studied methods of clarifying the classification of exodeviations. The focus of his work was on pseudo-divergence excess. Typically the diagnosis of pseudo-divergence excess is made when the distance measurement is greater than the near measurement but the near measurement equals the distance when measured with +3.00 lenses. Kushner advocates applying one hour of monocular occlusion prior to assessment of the AC/A ratio. If distance

<table>
<thead>
<tr>
<th>Number of subjects</th>
<th>Classification before prism adaptation</th>
<th>Classification after prism adaptation</th>
<th>New classification</th>
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<tbody>
<tr>
<td>14</td>
<td>basic</td>
<td>Convergence insufficiency</td>
<td>“pseudo-basic”</td>
</tr>
<tr>
<td>10</td>
<td>Convergence insufficiency</td>
<td>basic</td>
<td>“pseudo-convergence insufficiency”</td>
</tr>
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</table>
equals near after occlusion, his diagnosis is "pseudo-divergence excess with tenacious proximal fusion (TPF)". When this diagnosis is made, Kushner advocates surgery. However, when the patient has a true high AC/A ratio (distance remains greater than near after occlusion but distance equals near with +3.00 lenses), Kushner recommends treatment with minus lenses until the AC/A ratio normalizes. If the family wishes to proceed with surgery in the presence of a high AC/A ratio, Kushner recommends that the family be advised that bifocals will be needed post-operatively. According to another study by Kushner, patients with pseudo-divergence excess with TPF have good results with bilateral lateral rectus recessions, but patients with true basic X(T) do poorly and have better results with a unilateral recess/resect procedure. Kushner's work, therefore provides another example of how measurement technique can influence the surgeon's choice of procedures as well as the timing of surgery.

As one can see from the above studies, standard measurement techniques are not always sufficient for determining the best surgical procedure for intermittent exotropia. Orthoptists should be aware of the impact that the measurements can have on the determination of the surgical procedure and consider utilization of additional strategies when appropriate. The above studies show the importance of clearly identifying the classification of the exodeviation as well as distinguishing between pseudo-divergence excess based on tenacious proximal fusion from that based on a true high AC/A ratio.

It is interesting to note that all of the studies referenced in the last two sections involved measurement of strabismus, a task typically undertaken by orthoptists, yet all of the studies, except that by Neikter, were conducted by ophthalmologists. Clearly, orthoptists have not given enough attention to this area in their research. Not only can orthoptists contribute to surgical success by incorporating the techniques described above into their evaluation process when indicated, but though our own research, we can gain a better understanding of the relationship between choice of surgical procedure, classification of intermittent exotropia, measurement technique and surgical result.

Not enough millimeters of surgery

For some patients, surgical under-correction might simply be a result of insufficient amounts of recession or resection of the extraocular muscles. Most ophthalmologists have a surgical table that is used to determine the amount of surgery for specific sizes of deviations. The creation of these tables is based on surgical experience, reflecting amounts of surgery that, in

<table>
<thead>
<tr>
<th>Classification</th>
<th>Surgical success</th>
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<tbody>
<tr>
<td>True basic</td>
<td>24%</td>
</tr>
<tr>
<td>“pseudo-basic”</td>
<td>67%</td>
</tr>
<tr>
<td>True convergence insufficiency</td>
<td>9%</td>
</tr>
<tr>
<td>“pseudo-convergence insufficiency”</td>
<td>57%</td>
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</table>

Table 3 Surgical success by classification
general, have resulted in surgical success for most patients. Perhaps those patients that are under-corrected or have a recurrence of their deviation require more millimeters of surgery than does the average patient. The challenge here is to identify, pre-operatively, those patients requiring more aggressive surgery and similarly, identify patients who are likely to have a persistent overcorrection postoperatively and should have fewer millimeters of surgery than the average patient. Some studies have addressed that question. As seen in table 1, however, in spite of many studies of surgical outcomes spanning decades, there does not seem to be a trend for improved surgical success. Therefore, it appears that investigators have not had much success in identifying subjects requiring more aggressive or less aggressive surgery in terms of amount of recession or resection. This area is wide open for research by orthoptists. During the course of orthoptic therapy and orthoptic follow-up, orthoptists have a unique opportunity to become very familiar with subtle patient characteristics that could easily be overlooked by the ophthalmologist who spends less time evaluating the patient. Perhaps these subtle characteristics hold the key to predicting response to standard surgery. For example, it was my clinical impression that surgical under-correction was common for adult patients with a remote near point of convergence. To evaluate the validity of this observation, the following study was conducted.

**Convergence ability as a factor in surgical outcome**

**Methods:**

This was a retrospective study that included patients aged 13 years or older that had surgery for an exodeviation. Patients with a history of prior surgery for an exodeviation were included. Patients were included only if they demonstrated fusion with stereopsis before or after muscle surgery. Chart documentation of the pre-operative near point of convergence was required for inclusion in the study. Patients were excluded if they had Duane's syndrome, nystagmus, acuity worse than 20/60 in one or both eyes, more than 3 lines difference between eyes, or central nervous system dysfunction affecting their oculomotor system.

Subjects were categorized in one of two groups depending upon their pre-operative convergence ability. Group 1 consisted of those with good/fair convergence. These were subjects with fusion at near and with a near point of convergence to 25cm or better Subjects with constant exotropia at near who were able to bring their eyes from a divergent to a convergent position when instructed (encouraged) to look at their thumb at 7-8cm. were included in Group 1. Group 2 consisted of those with poor/no convergence. These were subjects with fusion at near and with a near point of convergence beyond 25cm and subjects with constant exotropia at near and unable to bring their eyes from a divergent to a convergent position when instructed (encouraged) to look at their thumb at 7-8cm.

Surgical outcome was assessed in the early post-operative period (within 6 weeks postoperatively) and at the final post-operative visit. Recurrence/undercorrection defined as an intermittent exotropia of any size at distance or near or an exophoria greater than 15 prism diopters at distance or near.

**Results:**

Thirty-five subjects were included in the study. Twenty subjects were in Group 1 (good
convergence) and fifteen subjects were in Group 2 (poor convergence). Most of the patients in
Group 2 had been treated with but failed to respond to or did not comply with home
convergence exercises aimed at improving their near point of convergence. Mean post-
operative follow-up was 7 months (range: 1 wk - 35 mos).

In the early post-operative period, 5 subjects in group 1 (26%) had recurrence/under-
correction of their exodeviation and 9 subjects in Group 2 (64%) had recurrence/under-
correction. Eighteen subjects in group 1 and 14 subjects in group 2 had follow-up beyond 6
weeks postoperatively. At the final visit, 6 of the 18 subjects in group 1 (35%) had
recurrence/under-correction and 13 of the 14 subjects in group 2 (93%) had recurrence/under-
correction (Table 5). This difference was statistically significant (p=.0068). There was no
statistically significant difference in outcome with regard to the presence/absence of distance-
near disparity, age at time of surgery, history of previous surgery, mean pre-operative
deviation or visual acuity. For Group 2, final exodeviations ranged from 7° to 45° with a mean
of 19° at distance and 25° at near.

Discussion:

Although additional research is needed to confirm the findings of this study, the results
suggest that patients with poor convergence should have more aggressive amounts of
surgery. Additional research can also help identify other groups of patients needing more
aggressive surgery, as well as those patients at greater risk of persistent overcorrection needing
smaller amounts of surgery. The rather disappointing surgical results reported in the studies
in table 1 as well as many other studies not listed in the table indicate that a significant
percentage of patients do not respond to surgery as would be predicted by standard surgical
principles. Often it is the orthoptist that follows patients carefully before surgery is considered.
During the course of follow-up, we have a unique opportunity to note subtle features of our
patients' motor and sensory condition. We must increase our awareness of such features and
study the possible impact of these features on surgical outcome.

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