MANAGEMENT OPTION OF CHILDHOOD EXOSEVIATION

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ABSTRACT

Intermittent exotropia is the most common form of divergent strabismus. Treatment is indicated with increasing tropia phase to preserve or restore binocular function and restore/reconstruct normal ocular alignment. While medical treatment is sometimes helpful for temporary relief, surgical therapy is the preferred definitive treatment modality by most pediatric ophthalmologists and astrobiologists. Congenital exotropia is rarely associated with amblyopia. The treatment of choice in this condition is also surgical. Sensory exotropia is most often acquired after monocular visual loss. The preferred treatment is surgical recession/resection on the impaired eye. Convergence insufficiency is usually not diagnosed until the teenage years or later, and it is best approached non-surgically with convergence exercises. In this article, we review the current literature and practice on the diagnosis and management of exotropia with emphasis on intermittent exotropia.

KEY WORDS: Exodeviation, Exophoria, Exotropia, Amblyopia.

INTRODUCTION

Childhood exodeviation (outward deviation) is a form of strabismus (eye misalignment) in which one or both of the eyes turn outward, characterized by out ward deviation of the visual axis forming a divergent angle, it usually begins as exophoria shifts later to exotropia. An exophoria is a tendency for exodeviation of the eyes that only appears when binocular viewing is broken, when one of the eyes is covered. The two eyes are no longer looking at the same object and does not have the same visual stimulation. In such cases the covered eye is the one that will drift outward. The misalignment of the eyes starts to appear when a person is tired, therefore it is not present all of the time. An exotropia is a misalignment of the eyes that is present when the both eyes are open. A tropia is the resting position that your eyes go to when covered or when deviation is visible, even without breaking fusion, or by repetitively alternately covering each eye [1**]**.

Classification of childhood exodevation:

A-constant exotropia.

B-intermittent exotropia.

A-Constant exotropia:

This occurs when the eye turn is present all of the time, at all direction. This type occurs less frequently than the intermittent type. Constant exotropia may be of following types:

1- Congenital exotropia.

2- Decompensated intermittent exotropia

3- Sensory deprivation exotropia.

4- Consecutive exotropia.

1-Congenital exotropia:

Congenital exotropia is rare and present at birth and may be associated with neurological abnormalities like cerebral palsy, midline defects or craniofacial

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syndromes. Infantile exotropia manifests during the first year of life, it is characterized by large and constant angle of deviation.
- Since infant uses left eye in left gaze and right eye in right gaze (uncrossed homonymous fixation), development of amblyopia is rare. In some, if one eye is preferred for vision, then other eye may develop amblyopia.
- Infant has normal refraction.
- Adduction is not restricted.
- No lid involvement or pupillary abnormalities distinguishing it from oculomotor nerve palsy (third cranial nerve).
Treatment: usually surgical, bilateral lateral rectus recessions, after 6 months of age [2].

2-Decompensated intermittent exotropia:
In some patients, exophoria progresses to intermittent exotropia that eventually may lead to constant exotropia. Deviations usually occur first for distance and later appear for near fixation. However, there are exceptions. The deviation remains constant or rarely may decrease. Manifest intermittent exotropia may increase with time and progress to constant exotropia.
Treatment: usually surgical, bilateral lateral rectus recessions, after 6 months of age.

3-Sensory deprivation exotropia:
Sensory deprivation exotropia is due to disruption of binocular reflexes (monocular poor vision) by acquired conditions like opaque media due to a disease or cataract, a blind eye drifts outward into exodeviation (position of rest). Sensory deprivation usually occurs in older children or adults.
Treatment: involves elimination of treatable causes like removal of catract. lateral rectus recession of the affected eye gives temporary cosmetic relief for some time.

4-Consecutive exotropia:
Consecutive exotropia may develop following surgical overcorrection of esotropia (inward deviation of eyes), especially in an eye which is amblyopic. Occasionally, a deeply amblyopic convergent eye may become divergent (acquire resting position of eye). Also, can occur in case of high hypermetropia, and poor preoperative evaluation of patient [3].
Treatment: reduce hypermetropic correction re-operation if exotropia persists, lateral rectus recession or medial rectus advancement.

B-Intermittent exotropia:
Intermittent exotropia is an outward deviation of the eye that is not constant and is intermittently controlled by fusional mechanisms. Unlike a pure phoria, intermittent exotropia breaks down spontaneously into a manifest exotropia [4]. It is the commonest presentation of childhood exodeviation, usually present around the age of two years, varies from 6 months to 6 years. About 48–92% of the exotropic patients have intermittent exotropia. It Can occur in normal newborn up to 6-8 months because development of binocular vision still not complete. If it persists beyond 6-8 months, it is considered clinically significant. A child with intermittent exotropia does not develop diplopia (double vision) due to bitemporal suppression, unlike acquired manifest exotropia in adults.
With progressive suppression, constant exotropia may develop. Development of amblyopia (functional suppression of retina) is very rare because the deviation is more frequently latent or intermittent.

In many subtypes of intermittent exotropia, the deviation is more obvious on distant vision, which is the reason why it is also referred to as intermittent distance exotropia 5,6]. Since the deviation usually manifests when the person is tired, sick, it is also known as periodic or inattention exotropia [6].

The etiology of intermittent exotropia is not clear, but many factors, including neuro-muscular abnormality, insufficient fusion, refractive errors, a high accommodative- convergence accommodation ratio (AC / A) [7,8]

Manifest exotropia may be precipitated by factors such as fatigue, light glare, ill-health or visual distraction [9].

Patients with intermittent exotropia rarely have any complaints due to well-developed suppression mechanism. Patient may have symptoms like eyestrain, headache, blurring of vision or difficulty in prolonged reading. However, these symptoms are quickly controlled by development of sensory adaptation.

However, studies have shown that not all intermittent exotropia deviations are progressive as some of them remain constant for years and a lower percentage of patients improve over time [10,11].

Exotropia should be distinguished from conditions like oculomotor nerve palsy or pseudoexotropia. In pseudoexotropia, visual axis of both eyes is straight, but the eyes appear divergent [12].

Classification:

According to distance-near relationships, intermittent exodeviations may be subdivided as:

1-Convergence insufficiency exotropia: Due to convergence insufficiency, exotropia is worse for near vision, near deviation greater than distance.

2-Divergence excess: Due to divergence excess, exotropia is worse for distance vision, distance deviation greater than near.

3-Basic exotropia: Exotropia is equal for both near and distance vision.

Symptoms:

Patient with childhood intermittent exotropia may present with: -

- Patient may experience eyestrain following prolonged near work.
- Running together of words or missing of the word being read, due to divergence of eyes.
- Some patients may be aware of divergence and are able to control it voluntarily.
- Voluntary control of exodeviation may lead to accommodative convergence which makes letter appear small in size [12].
- Some patients have panoramic view i.e., increase in temporal visual field.
- A child may close one eye (eye which diverges) in bright light.

Diagnosis:

- Careful history, refraction and visual acuity. A complete eye examination is conducted including record of ocular motility.
- Measuring Ocular deviation for gaze at near (33 centimeters), distance (6 meters) and far distance (beyond 6 meters) is recorded. Assessment of the control of deviation is noted which helps in monitoring progression of intermittent exotropia. This deviation may be noted by the parents or is detected on eye examination. The degree of deviation may be different in primary (straight gaze) and lateral (side) gaze positions. Test for straight of binocular vision –relative accommodation and dynamic retinoscopy. This is important to record from surgical point of view, to avoid postoperative diplopia in lateral gaze [13].

Management:

Management decisions in intermittent exotropia patients still present a challenge to many clinicians. intermittent exotropia is treated when binocular vision is impaired, or the patient is symp tomatic. The objective of treatment is to reduce the symptoms and the frequency of manifest deviation by decreasing the angle of deviation or enhancing the ability to control it. There are different surgical and non-surgical management strategies. Non-surgical methods are non-invasive and can reduce the symptoms and postpone surgical interventions [14,15]

A- Non-surgical management option of intermittent exotropia:

Non-surgical management include, observation, correction of refractive error, overcorrecting minus lens therapy, patch/occlusion therapy, orthoptics/binocular vision therapy, and prism therapy, botulism toxin.

Management decisions in intermittent exotropia patients are presently guided by both the angle of deviation and the ability to control it. Although non-surgical management options are very effective for the treatment of intermittent exotropia, they are rarely associated with adverse outcomes. These options are believed to be more appropriate in many patients with a small angle of deviation (< 20 prism diopter). Moreover, these methods may be preferred in very young children who may become amblyopic or lose their binocular fixation due to the possibility of overcorrection in surgical procedures [16,17].

1-Correction of refractive errors:

One of the etiologies of intermittent exotropia is high hyperopia and /or low-to-moderate myopia, which can cause deviation through decreased accommodative convergence [18].

Anisometropia can also impair sensory fusion and thus result in deviation due to the relationship between sensory and motor fusion [19]. Therefore, correction of these types of refractive errors may resolve the problem by improving the sensory and subsequently, the motor fusion and

enhancing the ability to control the deviation [20]. According to a recent study by Han et al. uncorrected refractive errors impair stereo-acuity, and since impaired stereo-acuity may worsen intermittent exotropia in the

long-term, its treatment may improve the patient's status over time [21].

In myopic cases, full correction is suggested due to its effect on accommodative convergence. In hyperopic patients, although some clinicians believe that hypermetropia will increase both the frequency and the angle of deviation, this is not always correct. Many patients with high hyperopia and intermittent exotropia improve after optical correction [22]. Age, degree of hyperopia and amount of AC/A should be considered in prescribing glasses for this type of refractive error.

In general, mild-to-moderate hyperopia is not usually corrected due to the possibility of accommodative convergence relaxation and worsening of the angle of deviation. However, in hyperopia >4 D or anisometropia >1.5 D, refractive error correction usually improves the deviation control. Moreover, in these cases, due to the lack of accommodative effort, the retinal image is blurry and deviation becomes manifest; hence, correction of hyperopia can enhance the retinal image and improve deviation control [22,23].

By evaluating the changes in the angle of deviation after refractive correction

in intermittent exotropia patients, reported a mild increase in exodeviation following hyperopic correction (more than 10 Prism diopter in one-third of the hyperopic cases). By contrast, myopic cases, the angle of deviation remained constant or decreased following wearing spectacles for at least 6 months. The authors concluded that optical correction in cases with exotropia was useful before strabismus surgery, and considering a new angle of deviation after spectacle correction should produce better results [24].

Although findings related to the effect of refractive error correction on intermittent exotropia are controversial, it is widely accepted that correcting even insignificant amounts of refractive error (especially astigmatism and myopia) may result in a better deviation control and should be considered prior to surgical intervention.

2- Overcorrecting minus lens therapy: - In this method, the patient wears over-minus spectacles compared to their cycloplegic refraction at all times. The aim of this method is to stimulate accommodation and hence stimulate convergences and reduce the angle of deviation.

This temporary method is also used when children are not old enough to start orthoptic therapies. Minus lens therapy is usually used in preschool children [25]. However, there are reports of its efficacy in older ages [26]. It was previously believed that the minus lens was effective only in patients with a high AC/A ratio; however, more recent evidence suggests otherwise [26,27]. On the other hand, recent studies found that some high AC/A ratios was not detrimental to a good outcome, and it appears that even children with low and normal AC/A ratios may respond well to overcorrecting minus lens therapy [28,29]. It is reported that even a slight accommodative effort can trigger a surprisingly large vergence response that can be enough to permit the child to keep the exodeviation latent by using large fusional convergence amplitudes [28].

A widely held mechanism for the efficacy of a minus lens is that it stimulates accommodation, resulting in the stimulation of accommodative convergence and intermittent exotropia reduction [30].

Another theory that accounts well for the efficacy of minus lens therapy is the fusional convergence mechanism [31]. The use of "over-minus" lenses in intermittent exotropia could eliminate blur that is secondary to excess accommodation resulting from disparity-driven convergence and thereby could promote fusional convergence at near [32].

Two methods are used to determine the required overcorrection:

1- using a constant amount of over-minus lens according to the refractive error and age.

2- using a primary lens and applying gradual additions until intermittent exotropia control is achieved [33].

The overcorrecting minus power is − 0.5 D to − 5.00 D in most studies (more routinely − 1.00 to − 3.00) [34]. Indeed, the minimum minus power that a child can tolerate and does not compromise the child's vision while providing the best control over the deviation at the same time is usually considered. The first minus lenses are placed in a trial frame starting at − 0.50 D strength and working up in − 0.50 D steps until the patient controls his/her deviation at near and distance fixation [29]. A very

recent study of the effectiveness of minus therapy in intermittent exotropia in 2018 showed that over-minus correction improved intermittent exotropia control and decreased the angle of deviation at distance and near [35]. However, it should be noted that the majority of these studies assessed the results of short-term minus therapy, and there is no sufficient information regarding the long-term effects of this treatment method after discontinuing the use of the minus lens. Moreover, because excessive accommodation has
been implicated as a cause of myopia, there is a theoretical concern that overcorrecting minus lens therapy for exotropia may cause myopia. However, despite this concern, many previous studies found that this treatment did not seem to cause myopia [29,36]. Moreover, over-minus therapy can cause esotropia in near. Hence, after beginning
the therapy, the first examination should be performed 3 – 4 weeks later to evaluate alignment in near [28,37].

3- Prism therapy:

Prism therapy is another intervention that can be applied either alone to neutralize the deviation or in combination with orthoptic therapy [38]. Prism therapy is mainly used in patients with a deviation < 20 – 25 Prism diopters. Higher prisms are required in higher deviations, resulting in increased weight of the spectacles and distortion [39]. There are different strategies for prism prescription, including a neutralizing prism (a prism is prescribed according to the deviation magnitude to neutralize its angle), and an exercising prism (used to
induce diplopia and stimulate fusional convergence).

Some authors recently found the effectiveness of exercising prism addition, for improving Newcastle Scoring System in patients with basic intermittent exotropia.

Inverse prisms are used to increase fusional vergence ability since they increase the demand for controlling fusional vergence. Theoretically, base-out prisms increase the demand by shifting the image to where the eye is supposed to look. This shift will provoke diplopia, stimulate fusion, and encourage the visual system to improve sensory perception and motor control [40].

4- Patch therapy:

There are different methods for occlusion therapy, also known as anti-sup
pression therapy, including monocular or alternate occlusion and part-time versus full-time occlusion. Monocular occlusion (patching the dominant eye) is applied if there is a dominancy relationship between the eyes, and alternate occlusion is done when there is no dominant eye [41].

The aim of this approach is to eliminate visual adaptation (suppression induce
diplopia), and consequently motivate motor fusion.

Nonetheless, diplopia may not be induced in all patients with strabismus [42]. It is also suggested that occlusion may reduce the amount of deviation and change the deviation from exotropia to exophoria [43].

Some researchers believe that this method is useful in younger children and in patients who wish to postpone surgical treatment [42,43].

The effect of alternate occlusion for 3 hours daily versus observation on the deterioration of deviation (worsening of stereopsis or deviation becoming more constant) in children aged 3–10 years who were not previously treated. The cases were randomly allocated to obser-

vation (no treatment) or intermittent occlusion. After 6 months, a low rate of deterioration was observed in both groups (6.1% in the observation group and 0.6% in the occlusion group). Although the deterioration rate was lower in the occlusion group. In general, part-time occlusion for 2–6 hours were recommended in previous studies. However, we could not find a clear rationale for the duration of patching recommended in these studies or sufficient evidence to support the optimum patch time for intermittent exotropia treatment in different age groups of children. An important point to remember when using this method is that since fusion control improvement resulting from occlusion therapy is temporary, it should not be used alone for intermittent exotropia treatment. In fact, this method is mostly used to postpone surgical treatment [23,24].

5- Orthoptics / binocular vision therapy:

The main objective of orthoptics / binocular vision therapy is to eliminate suppression, stimulate diplopia awareness, improve sensory fusion and fusional reserve, and restore binocular Vision [44]. A combination of anti-suppression therapy and accommodation and vergence therapy is recommended. Vision training should initially address vergence skills by modifying fusional vergence amplitudes and vergence facility at near fixation for converges insufficiency and at distance, intermediate, and near for diverges excess and basic exotropia [45].

Researchers usually categorize vision therapies into some general categories for comparison, including home-based computer therapy, office-based therapy, and pencil push-up treatment. It has been shown that a combination of in-office and home-based therapy produces better results than does home-based therapy alone [46-48].

In general, there are three opinions about binocular vision therapy: some studies suggest that it has no effect on the deviation [49], some have shown that orthoptics / binocular vision therapy alone can reduce deviation and some others have found that it should be accompanied by surgery [50].

It can be concluded that orthoptics / binocular vision therapy is very effective in children or even in adults who are cooperative and willing to do the therapies.

6- Botulinum toxin:

The therapeutic principle of botulinum toxin in intermittent exotropia is to paralyze the injected muscle (lateral rectus muscle). It is at least as effective as surgery irrespective of the initial strabismus angle [51-53]. Significant improvements in fusion control and improved stereovision have been reported 6 months after botulinum toxin injection to the lateral rectus muscles [51].

Botulinum toxin chemo-denervation may be difficult to achieve long term alignment in cases of secondary exotropia [53]. Botulinum toxin injection in extraocular muscles may be used in patients with constant exotropia who are at risk of postoperative diplopia and also who have undergone multiple operations. Preoperatively, if after botulinum injection, a patient develops intractable diplopia, this is usually the case after surgical correction. Reviewed the studies related to different intermittent exotropia treatment modalities. The overall pooled success rate of different treatment modalities was 28% for prism therapy, 28% for over-minus therapy, 59% for vision therapy, and 37% for occlusion therapy. It was suggested that a combination of relieving prism and orthoptic therapy may be more successful than most of the above strategies.

B- Surgical management options of exotropia:

Surgical management may be indicated in patients with:

- Poor control of intermittent exotropia: When manifest intermittent exotropia is present at least half of the time during the day.
- Progressive deterioration of control of intermittent exotropia: Patients with increase in size of deviation, loss of control and a progressive inability to refuse images after manifest deviation.
- Severe eyestrain.
- Troublesome diplopia.
- Surgery may also be indicated to restore binocularity and also for cosmetic reasons.

Surgical results are better for intermittent exotropia as compared to constant exotropia.

The best time for surgery is when the child is between 4-5 years of age [54].

Surgical management options include: lateral rectus muscle recession, lateral rectus muscle recession with ipsilateral medial rectus muscle resection and bilateral medial rectus muscle resection.

Choice of operation:

- Each form of exotropia requires different treatment. The pattern of deviation tells which muscles to be operate on.
- Amount of deviation indicate how much surgery is needed.
- Divergence excess: recession of both lateral recti.
- Basic exodeviation: recession-resection, followed by operation on fellow eye when needed.

Surgery with preoperative conservative orthoptic or occlusion therapy gives the highest success rate [55].

Management Option of exophoria:

Although most normal individuals have at least some degree of phoria, the vast majority are asymptomatic.

If a deviation is uncovered on routine testing in an asymptomatic patient, no treatment is necessary. If a patient is complaining of asthenopia and a deviation is discovered during testing, one must first ensure that there are no coexisting issues prior to attributing the symptoms to a phoria. There are a number of causes for asthenopia symptoms that should be ruled out. To be sure, some of the refractive causes of asthenopia are via induced phorias [56].

Once other causes for symptoms are ruled out, the cause of phorias should be investigated, refractive error is a common cause. In certain cases, many cases benefit from base-out exercising prisms. A minimal adapted prism is applied in the trial frame for 15 to 20 minutes in the clinic. Otherwise, if there is no a significant adaptation to the prisms, the patient's symptoms will probably not be improving. Then a base-in relieving prisms are tried. Through, with the use of prisms and other therapies the symptoms persist, surgery should be considered.

Again, careful refraction helps the management of many cases of exophoria. With refractive correction in place, if any, cover tests should be performed, and accommodation should be evaluated by push-up measurement of accommodative amplitude or dynamic retinoscopy. Divergence excess (in contrast to convergence insufficiency) manifests as an increased angle of exophoria in the distance. In exophoria, correcting both myopia and hyperopia can help improve symptoms, but additional cautions should be taken when correcting hyperopia, as full correction of hyperopia may worsen the symptoms. A several-minute test with hyperopic correction should be attempted, if exophoric symptoms improve because

of clearer imagery or worsen by relaxing accommodation. If they worsen, prescribe the largest correction possible to treat the hyperopia while avoiding exophoric symptoms. A good starting point is one-third of the spherical error. Just as plus lenses can be helpful for esophoria, decreased-power plus lenses or even minus lenses can improve exophoria [56].

Base-in prisms may also be helpful for the treatment of exophoria. As with esophoria, the least amount of prism that eliminates exophoric symptoms should be used. The cover test should provide an estimate of the power of the prism to be used. Additionally, treating a minor hyperphoria with vertical prisms can allow the patient to compensate for exophoria with no need for horizontal prisms. If divergence excess is found to be the cause of the exophoria, prisms should be avoided. In this situation, base-in prism can cause esophoria at near, which patients do not tolerate well.

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