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CONGENITAL EXOTROPIA: A RARE CASE SERIES

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Abstract

Background: Congenital exotropia (CXT) is a rare form of strabismus, characterized by divergent strabismus, where the eyes deviate outward. It typically becomes apparent in infancy, generally before six months of age, and persists beyond this stage.

Objective: This case series aims to explore the surgical management of congenital exotropia (CXT) in five children, focusing on the outcomes of the monocular recess-resect procedure and bilateral lateral rectus recessions.

Methods: Five children with congenital exotropia were included in the case series. Surgical interventions included monocular recess-resect and bilateral lateral rectus recessions.

Results: All patients underwent successful surgical correction of large-angle exotropia. In the case of a 1.6- year-old child, a left-eye recess-resect procedure was performed, and residual exotropia of 15 degrees was noted on the first postoperative day. Additional surgery was taken up in the other eye after 3 months which resulted in a good postoperative alignment.

Conclusion: This case series demonstrates the effectiveness of the recess-resect and bilateral lateral rectus recession procedures for managing congenital exotropia, with favorable postoperative outcomes, even in cases with delayed surgery.

Keywords: Congenital exotropia, motor and sensory outcomes, amblyopia

Introduction

Congenital exotropia (CXT), also known as Infantile exotropia is defined by idiopathic, large angle constant exotropia appearing the first six to twelve months of life. It typically becomes apparent in infancy, generally before six months of age, and persists beyond this stage. It is classified as primary infantile exotropia, where patients are healthy with no evidence of systemic or ocular disease or it can be secondary to various systemic and ocular causes like craniofacial syndromes, mid-line defects, cerebral palsy, genetic syndromes, ocular albinism etc. Some authors classify early onset intermittent exotropia also in this sub classification. True infantile exotropia occurs sporadically, but maybe familial as autosomal dominant with variable penetrance, as was suggested in one case report [1].It is characterized by large constant angle of deviation that goes beyond thirty prism diopters (PD) and may reach upto ninety PD in severe cases. The occurrence of this condition happens more rarely than infantile esotropia since it affects one in 30,000 births. The rarity of congenital exotropia presents a significant challenge for appropriate treatment because there is little research available from clinical trials. Universal treatment guidelines become difficult to establish

due to the insufficient availability of data. Although congenital exotropia happens infrequently it requires individualized management strategies because of its important medical implications in each affected patient. The condition leads to substantial effects on binocular vision that could hinder visual development without proper management approaches [2]. CXT displays certain unique natural characteristics when comparing with other kinds of strabismus. The outward eye deviation reveals itself to external observation shortly after birth and becomes more evident as the child grows older [3]. Even then, there are certain challenges to diagnosis with respect to vision evaluation and amblyopia detection in these preverbal children, identifying age of onset of strabismus and diffrentiating it from early onset intermittent exotropia. Vision evaluation in preverbal children can be done by assessing for fixation preference. Reviewing child's old photographs, history from parents and paediatrician's health records may help in identifying the age of onset.
It may be associated with dissociated vertical deviation (DVD) combined with inferior oblique over action (IOOA) in about 50-60% of patients, superior oblique over action (SOOA) and rarely latent nystagmus. Pattern deviations in the form of V, A and X patterns may be seen. As with any other type of strabismus, any refractive error should be corrected and amblyopia, if detected should be addressed with occlusion /penalization therapy before planning surgery. This helps in stabilizing the degree of deviation, but is unlikely to lead to resolution of the exotropia. The patients are reevaluated after atleast two weeks of spectacle wear and amblyopia therapy. The distribution of refractive errors in infantile exotropia is similar to that of normal infant population [4]. Strabismic amblyopia is seen in a minority of patients but is responsive to traditional therapy [4].

The primary method for treating congenital exotropia consists of surgical procedures especially when patients have persistent and large deviations. The correction of eye misalignment together with improvement in binocular vision serves as the main objective when performing surgical procedures. There are multiple surgical options with several studies reporting no clear advantage of one procedure over the other. The various surgical options include unilateral(U/L) or bilateral(B/L) lateral rectus recessions, U/L or B/L medial rectus resection, lateral rectus recession and medial rectus resection of the affected eye, additional strabismus surgeries to correct any associated DVD, oblique muscle dysfunction and residual strabismus after large angle exotropia surgery which may require 3 or 4 horizontal muscle surgery. Surgeons have varying preference, with different permutations all reported in the literature. The most popular surgical option for treating this condition include bilateral lateral rectus (B/L LR) recessions and monocular recess- resect. The recess-resect surgical procedure serves as the preferred method for treating congenital exotropia among most surgical professionals currently. The surgical method involves making the lateral rectus muscle weaker through recessing then making the medial rectus muscle stronger through resecting. Despite its many benefits, the recess-resect operation has some potential risks that may occur. The major side effects from this surgery consist of strabismus overcorrection or under correction and additional surgical procedures for large angle exotopia. Leuder GT et al evaluated long term outcome of surgery in infantile exotropia and concluded that the need for more than one surgery is higher in infantile exotropia when compared to other forms of childhood strabismus, but most children achieved good alignment with one or two surgeries [5]. They also assessed the need for neurologic evaluation in these patients and stated that developmental delay is common in patients with infantile exotropia , but this was usually recognized at the time of initial evaluation and that routine neurologic screening or imaging of these otherwise developmentally normal infants was not required [5].Na KH et al conducted a study on early v/s late surgery in infantile exotropia and concluded that older age at surgery was associated with a risk of recurrence, but it was not correlated with surgical outcomes for patients with intermittent exotropia in this study [6]. Good outcomes from strabismus research will probably help develop improved protocols and enhance knowledge about congenital exotropia throughout ongoing investigations. The aim of this study is to discuss the case series of five children with CXT, focusing on the clinical presentation and outcomes of the recess-resect procedure and bilateral lateral rectus recessions.

Case 1: A 1.6-Year-Old Child with Congenital Exotropia

A 1.6-year-old child presented with a history of squinting since early childhood. The parents noticed the outward deviation of the eyes during infancy, but the child's general condition had been poor, delaying surgical intervention until the age of 1.6 years. The child showed large alternating angles of exotropia with Hirschberg measurement at 40-45 degrees of exotropia. Child had equal fixation preference thus indicating absence of amblyopia with normal extraocular movements that was unrestricted in all directions. Results from cycloplegic refraction indicated normal results as per child's age. Testing with Krimsky test,the prism bar cover test (PBCT) showed 90 PD of exotropia. The delayed presentation of the condition resulted from the child experiencing poor overall health until reaching his 18-month milestone. The child underwent left eye recess-resect (LE R/R) procedure which achieved a successful outcome. Residual exotropia of 15 degrees was seen on the first postoperative day. Patient was advised to follow up one week and one month afterward for further assessment. Additional surgery in the form of right eye R/R was planned three months after first surgery. Patient had stable alignment following the second surgery. The case shows that assessment of general child health status before performing strabismus surgery is mandatory and that two staged strabismus surgery may be required in large angle exotropia to achieve a successful outcome.

Figure 1: Congenital exotropia : Preoperative and postoperative pictures after LE R/R

Case 2: A 1.3-Year-Old Child with Large Angle Exotropia

A 1.3-year-old child presented to the clinic with a noticeable outward deviation of both eyes, first identified at six months of age. The child had undergone general health improvement after initial delays, which allowed for surgical correction at the age of 2 years. On clinical examination, Hirschberg testing revealed 35 degrees of exotropia. Extraocular movements were within normal limits, and there were no signs of restrictive strabismus. Refraction results were within the normal range. PBCT was performed using the Krimsky test, which neutralized with 70 PD exotropia. The child demonstrated equal fixation preference with no signs of amblyopia. The surgical approach chosen was a monocular recess-resect procedure. Postoperatively, the child's alignment improved significantly, with a reduction in the angle of exotropia. The child's binocular vision remained unaffected, and the alignment was stable during follow-up.

Case 3: A 2.6 -Year-Old Child with Amblyopia and Exotropia

A 2.6-year-old child with congenital exotropia presented with an outward deviation noticed since birth. Unlike other cases, this child had a significant fixation preference, indicating early development of amblyopia in the deviating eye. Upon examination, the child demonstrated 35-40

PD exotropia with full extraocular movements. However, the child's fixation preference was clearly towards the non-deviating eye. A Krimsky test confirmed the deviation of approximately 80 PD, and cycloplegic refraction was found to be normal. The child was diagnosed with amblyopia in the deviating eye, and a monocular recess-resect surgery was planned. Post-surgery, visual rehabilitation with patching therapy was initiated. The child showed significant improvement in both ocular alignment and visual function. Amblyopia was also addressed successfully with patching therapy, and by the age of 3, the child demonstrated near-normal visual acuity and no fixation preference.

Case 4: A 1.8-Year-Old Child with Constant Exotropia and Full Ocular Movements

A 1.8-year-old child was referred to the clinic due to constant exotropia since the age of 6 months. Despite the early onset, the child did not present with any complications such as amblyopia. Examination showed 40 degrees of exotropia with full, normal extraocular movements. No associated ocular motility restrictions were noted. Krimsky testing revealed around 85 PD of exotropia. The child exhibited normal fixation preferences in both eyes, with no amblyopia. Surgical correction was carried out using a bilateral lateral rectus recession technique. Postoperative follow-up indicated a significant improvement in alignment and visual performance, and the child maintained good alignment with no recurrence of the exotropia.

Case 5: A 2.1-Year-Old Child with Asymmetric Exotropia

A 2.1-year-old child presented with a history of large-angle exotropia, which was first detected at 6 months of age. Examination revealed 30 degree of exotropia on Hirschberg testing and the patient showed free movement through all directions of gaze. Krimsky testing showed 60 PD of exotropia. The child fixed his gaze in a typical manner without developing amblyopia. The eye's refractive ability aligned with typical development for the child's age. Monocular recess-resect procedure on the left eye was done. Postoperative examination revealed marked improvement in the patient's alignment as the exotropia decreased. The child maintained stable fixation preferences while avoiding amblyopia development and showed outstanding aesthetic along with functio nal outcomes.

Figure 2: Krimsky test to measure the amount of exotropia

Postoperative Follow-up and Residual Strabismus

Although the overall outcomes were positive, it is important to note that one child presented with residual strabismus postoperatively. Among the children studied there was a 1.6-year-old patient whose alignment revealed a persisting 15-degree of exotropia after the first day of recovery from

surgery for large-angle strabismus. This required additional horizontal muscle on the other eye which was planned three months after first surgery. Detailed preoperative evaluation, meticulous surgical methods and close postoperative monitoring is essential to achieve optimal patient results.

DISCUSSION

Congenital exotropia (CX) is a rare but significant condition, typically presenting in infancy, with symptoms often noticed before six months of age. The eyes exhibit a large constant outward deviation in this condition thus affecting the coordination of both eyes and their ability to perceive depth. Children typically display this condition in early infancy but delayed treatment usually happens because of poor overall health status which makes early identification also difficult. Early surgical intervention before age two years affords the best chance of attaining good alignment. Reasonable sensorimotor results are obtained in healthy infants aligned before 2 yrs of age [4,7,8,9]. However, with slight overcorrection in early surgeries, there is a risk of developing monofixation esotropia with foveal suppression, but this situation is rare. A much more common result is undercorrection. Hence more experts suggest a purposeful targeting for a slight overcorrection in the immediate postoperative period [4]. The study tracks five exotropic subject patients who received monocular recess-resection procedure or bilateral lateral rectus recession surgical procedures for their exodeviation based on their individual manifestation patterns. The surgical procedures performed on patients showed positive results with improved eye position and they maintained stable fixation in every case even though patients started treatment at different ages. Developmental status and health factors serve as essential considerations for early intervention since the surgeries were conducted at different developmental phase of the children. The recess-resect surgery represents an established solution to treat exotropia angles of considerable magnitude especially when treatment focuses on a single eye especially in the presence of amblyopia. The procedure uses muscle weakness in the lateral rectus to combine with muscle strengthening in the medial rectus which yields successful eye realignment. To achieve optimal results in the treatment of congenital exotropia, medical professionals must address any associated dissociated vertical deviation (DVD), inferior oblique overaction (IOOA) and superior oblique overaction (SOOA) when performing the primary surgery. Such preventative measures lead to balanced eye realignment while decreasing the necessity for future surgeries thus enhancing functional and cosmetic outcomes [10]. None of our patients had any associated DVD or oblique muscle dysfunction at the time of presentation.

Age of onset of less than 6 mths was associated with worse sensory prognosis but not with a difference in motor outcomes [10]. The level of binocularity determines how successfully strabismus surgery has worked in the eyes of patients. Medical research conducted by Hunter DG et al. demonstrated that half of infantile exotropia patients commonly present with intermittent exotropia as their presentation but surgery effectively corrects their misalignment in most such cases. More than fifty percent of patients developed stereopsis after surgery but bifixation remained impossible for all cases [11]. Strabismus surgeons face considerable difficulty when trying to restore complete binocular function in patients who have congenital exotropia and severe cases that have existed for a long time.Surgery within 2 years of birth has been reported to yield an 80 % success rate, while it decreases to 40 % thereafter [12]. In a recent study conducted by Mohan K et al on long-term Motor and Sensory Outcomes After Unilateral Lateral Rectus Recession–Medial Rectus Resection for Infantile Constant Exotropia,60% of patients achieved a successful long-term motor outcome and 25% achieved peripheral binocular single vision after unilateral recession-resection for infantile constant exotropia. Stereopsis outcome was nil. Age at surgery and duration of strabismus had no effect on motor and sensory outcomes [13].

Conclusion

Surgery is required almost universally in cases of true infantile XT. Once diagnosis is made, and refractive error ruled out and amblyopia therapy has commenced, if required, after only a few weeks of follow up to establish repeatable, reproducible angles of deviation, surgery should be performed

with minimal delay. In the absence of amblyopia or eye preference, alternate occlusion therapy can be instituted during the waiting period till the child becomes fit for surgery to improve the surgical outcome and postoperative stereopsis. Radiographic imaging is indicated only if neurological signs and /0r craniofacial anomalies are present. Comprehensive surgical intervention for congenital exotropia, which includes addressing both horizontal and oblique muscles during the primary surgery, results in less postoperative drift and better alignment outcomes. Surgery should be performed within six months of life or at least within two years of life to maximize the potential for attaining single binocular vision with stereopsis.

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