Case Report

Multistage Surgical Correction of Monocular Elevation Deficit with Large Angle Exotropia in a Pediatric Patient: A Case Report

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ABSTRACT

Monocular elevation deficit (MED) is characterized by restricted upward gaze, often associated with hypotropia and horizontal deviation. We report the case of a young patient with right eye MED and large-angle exotropia. A negative forced duction test ruled out mechanical restriction. Surgical management was performed in three steps; partial Knapp operation combined with horizontal rectus recession and plication in the RE, followed by inferior rectus recession in the RE and horizontal rectus surgery in the contra lateral eye, and finally ptosis correction. A multistage approach provided precise correction of ocular alignment and improved cosmesis. This case highlights the value of individualized, stepwise surgical planning for complex strabismus associated with MED and large horizontal deviations. Follow-up continued for 6 months after the final procedure, during which ocular alignment and eyelid position remained stable.

Keywords: Monocular Elevation Deficit, Exotropia, Hypotropia, Ptosis.

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INTRODUCTION

Monocular elevation deficit (MED) is a condition where the limitation of upward gaze with hypotropia is similar in adduction and abduction.[1,2] Signs of MED include hypotropia and ptosis. Horizontal strabismus is often associated with MED, and exotropia is generally common.[3]Horizontal deviation is generally between 15-25PD BI,[3] but also reaches up to 50 PD BI.[4] Surgical techniques vary and are typically based on forced duction test (FDT) results.[5] The available studies reported different surgical techniques with various outcomes.[3,4,6] However, the surgical management of congenital MED with large exotropia, as presented in this case, has not been previously reported.

Case presentation

A 2-year-old male presented with congenital strabismus and no significant medical or family history of strabismus, trauma, or eye disease. Birth and developmental milestones were normal. Examination showed right-eye hypotropia with large-angle exotropia (>95 PD) on Krimsky test. The fundus examination was normal. Elevation of the right eye was limited to −4 in all gazes, while the left eye was normal. Cycloplegic refraction showed +1.00DS in the right eye (OD) and +1.50 DS in the left eye (OS). Pseudoptosis and chin-up posture resolved with right eye fixation. Bell's phenomenon and the prism cover test could not be performed due to poor cooperation on part of the patient. The patient was diagnosed with

right MED associated with large exotropia.

The patient underwent strabismus surgery under general anaesthesia. Intraoperatively, the Forced Duction Test (FDT) was negative, excluding Inferior Rectus (IR) restriction in the right eye. A partial Knapp procedure was performed: transposing the superior halves of the medial and lateral rectus tendons superiorly and suturing approximately 8mm from the original insertion, adjacent to one third tendon superior rectus using 5-0 Mersilene® non-absorbable sutures. The inferior halves were modified by 6 mm plication of the medial rectus and 9 mm recession of the lateral rectus, using 6-0 polyglactin (Vicryl®)double-armed sutures. Mild under-correction persisted, though accurate measurement was limited due to the patient's poor cooperation.The interval between the first and second surgeries was three years; the initial procedure was performed at age two, and the second was undertaken once the child became more cooperative.

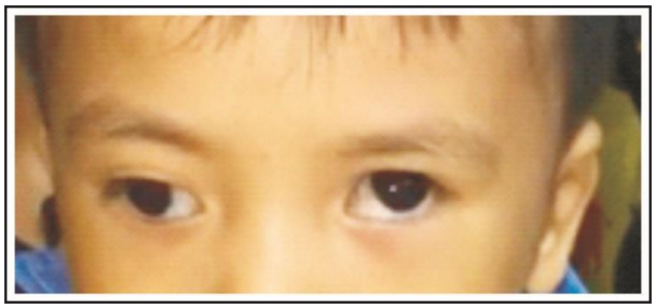

Figure 1: Patient at the age of 2 years with right pseudoptosis, left hypertropia, and large exotropia.

The patient returned at 5years of age and was more cooperative for ophthalmological examination. There was deviation of 65PD BI and 25 PD BU in OD with left eye fixating. Bell's phenomenon was positive.A second strabismus surgery was performed, involving a 5 mm inferior rectus (IR) recession in the right eye and, in the contra lateral eye, a 10 mm lateral rectus (LR) recession with a 7 mm medial rectus (MR) plication, using 6-0 polyglactin (Vicryl®) sutures. Postoperatively, residual deviation was 10 PD BU in OD and 20 PD BI with left eye fixating. Although the eyeball position was still slightly under-corrected, the parents were satisfied, and ptosis correction was performed two months later (Figure 2).

The initial surgical plan for ptosis correction was levator resection, as the levator function was more than 4 mm. However, intraoperative assessment suggested that levator resection alone would be insufficient, so a combined procedure with frontalis suspension was performed. The exact levator resection length was not recorded; instead, lid height was titrated intraoperatively by matching the operated eyelid to the contra lateral side in primary gaze, given the coexistence of levator dysfunction and long-standing pseudo ptosis secondary to hypotropia. This combined approach achieved satisfactory eyelid elevation and stable ocular alignment. Postoperatively, visual acuity was 1.0 with alternating suppression on the Worth Four Dot Test. At the final follow-up, six months after ptosis surgery, both alignment and eyelid position remained stable without complications.

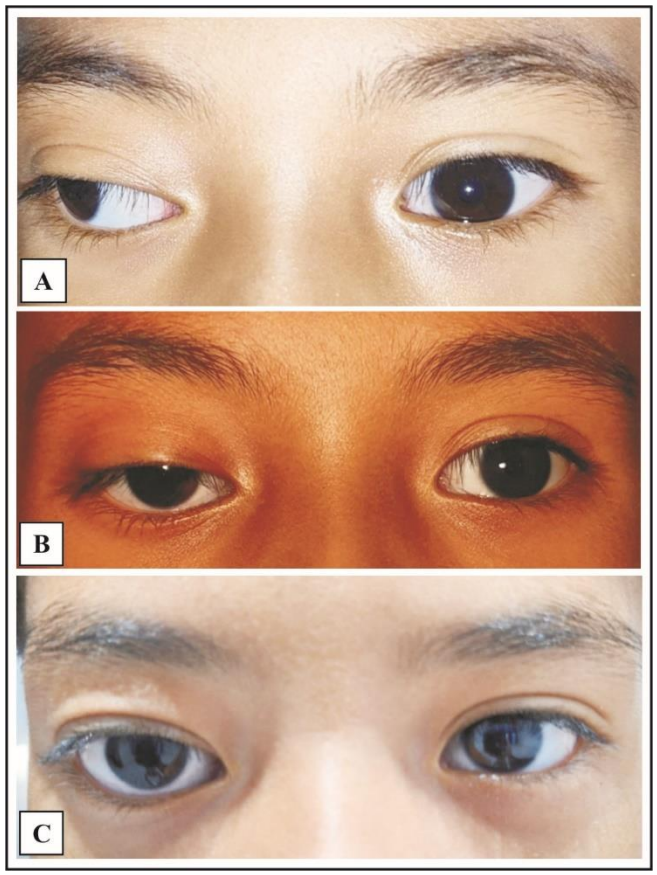

Figure 2: Top: Child at 5 years of age, before second strabismus surgery. Mid: Two months post-second surgery with mild residual hypotropia and XT. Bottom: Final postoperative appearance after strabismus and ptosis surgery.

DISCUSSION

MED with large exotropia is rare. Despite limited cooperation, marked vertical and horizontal deviations were noted. The patient was diagnosed with right-eye MED with hypotropia and ptosis when left eye was fixating. It was assumed that involvement of the right

eye was presumed congenital, with the left eye considered secondary to it.[3] The ptosis appears likely to have resulted from attachments between the superior levator palpebrae and superior rectus muscles. MED, an incomitant strabismus, shows greater secondary than primary deviation due to increased innervation effort in the affected eye.

The main goal of surgery is to improve ocular alignment in the primary gaze and widen the binocular visual field. Techniques include Knapp procedure, partial Knapp (vertical Hummelsheim), augmented/ modified Knapp, IR recession, and vertical muscle recession/resection.[5]

The choice of surgical technique depends on the Forced Duction Test (FDT), which evaluates IR restriction.[3] Studies report IR restriction in 70–82% of MED cases.[3] In this patient, a partial Knapp procedure was performed. Although combining the partial Knapp procedure with IR recession may be performed in conditions of large hypotropia, it increases the risk of anterior segment ischemia.[5] The partial Knapp typically corrects <10 PD up to 25 PD following IR recession. This differs from the full tendon Knapp procedure, which can correct hypotropia of 20-55 PD.[3] Single horizontal muscle transposition combined with IR recession is reported to correct more than 48 PD and reduce the risk of anterior segment ischemia.[8,9.]

To correct the residual hypotropia, a 5 mm IR recession was performed.[10] It improves elevation by weakening the restrictive IR muscle; in contrast, the Knapp procedure improves elevation by altering the muscle's mechanical vector through transposition. A5mmrecession was selected to minimize the risk of downgaze hypertropia and lower eyelid retraction.[3] In this case, however, although with mild residual deviation (10 PD BU), the family was satisfied with the cosmetic outcome.

This patient exhibited large-angle exotropia(< 95 PD), consistent with prior reports showing 22-70% exotropia among MED cases, with deviations ranging from 15-50 PD.[3,4] In this case, simultaneous correction of exotropia and hypertropia was performed using partial horizontal muscle transposition and recess-plication, without complication of anterior segment ischemia. Other studies also showed successful outcomes following simultaneous correction, although some studies are against combining Knapp and horizontal surgeries due to residual risk, which depends on deviation magnitude.[2,4,8.]

The patient underwent three surgeries: two for correction of misalignment and one for ptosis repair. Pseudoptosis commonly occurs in patients with marked hypotropia and resolves after extraocular muscle surgery, whereas true ptosis is present in approximately 50–60% of MED cases.[5] In our patient, persistent ptosis was attributable to levator muscle dysfunction for which a frontalis sling suspension was performed. Binocular vision was not achieved, likely due to residual exotropia (20 PD BI) and the long-standing congenital deviation, consistent with prior reports in which only 18% of MED patients attained binocular vision.[5] Alignment and eyelid position remained stable over 6 months of follow-up after the final stage, supporting the durability of a multistage approach in MED with large-angle exotropia.

CONCLUSION

A tailored, multistage surgical approach achieved optimal ocular alignment and cosmetic outcome in this complex case of MED with large-angle exotropia. Individualized, stepwise planning proved essential for stable long-term results.

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Conflict of Interest: Authors declared no conflict of interest.

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