A Promising Outcome of the Augmented Modified Hummelsheim Procedure in a Challenging Case of Inferior Rectus Hypoplasia

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Dear Editor,

Inferior rectus (IR) hypoplasia/aplasia is a rare abnormality. In Asia, cumulative data from the Japanese population from 1930-2009 recorded only 16 cases of IR aplasia.[1] This condition presents with various clinical signs, including abnormal head posture (AHP) with head tilt, incomitant hypertropia, limitation of infraduction, incyclotorsion on retinal imaging, and a forced duction test showing complete laxity for upward deviation and variable tightness for downward deviation. The pathogenesis is believed to result from either an aberrant insertion or a failure in the condensation of the common inferior mesoderm complex.[2] Misdiagnosis is common in IR hypoplasia due to its similarity to other more prevalent causes of IR underaction. These include disorders affecting the oculomotor nucleus, nerve, myoneural junction, or extraocular muscles (e.g., congenital anomalous bands).

Orbital imaging, such as computed tomography or magnetic resonance imaging (MRI), is the definitive non-invasive diagnostic modality. However, surgery remains the gold standard for both invasive diagnosis and treatment of IR hypoplasia.[3] The main goal of surgical treatment is to achieve ocular alignment in primary position, followed by improving ocular movement and addressing cosmetic concerns. The choice of surgical approach depends on the degree of ocular deviation, ocular motility limitation, and severity of muscle dysgenesis. With various surgical techniques available, surgeons must adapt and select the most suitable technique based on intraoperative findings.[4] In this case report, we present the successful management of a challenging case of IR hypoplasia using the augmented modified Hummelsheim procedure.

A 25-year-old female, who provided written informed consent for publication, presented with esotropia and hypertropia in the right eye without reported diplopia. She had a right head tilt and slight face turn since early childhood, with no history of trauma (Figure 1A). Visual acuity in the right eye was 6/6 with correction of 0.75 sphere and 0.75 cylinder (160° axis). The left eye exhibited 6/6 emmetropia. The prism alternate cover test showed 12 prism diopter (PD) esotropia and 15 PD hypertropia of the right eye in the primary position, with vertical deviation worsening in downward/outward gaze (Figure 1B). A three-step test revealed weakness of the right IR muscle, while fundus photography showed 11° incyclotorsion (Figure 1C). Further investigation with MRI suggested that the muscle belly was located far retroglobally (Figure 1D). No abnormalities were noted in the left eye.

The initial diagnosis was congenital IR palsy and atrophy, and the patient was planned for transposition surgery. A forced duction test was performed intraoperatively and yielded negative results. Subsequently, an exploration of the four rectus muscles was conducted. The lateral and medial rectus were identified. However, upon exploring the inferior sector to a distance of up to 15 mm from the limbus, only two ciliary arteries were found on Tenon's capsule or the sheath, with no identifiable IR muscle. This finding led to a revised diagnosis of congenital IR hypoplasia/aplasia (Figure 1E). The

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patient underwent a modified Hummelsheim procedure, with the muscle halves positioned as if the IR muscle were still intact, along with scleral augmentation sutures 5 mm behind the original insertion (Figure 1F). Postoperatively, there were significant improvements in ocular alignment, AHP, and funduscopic incyclotorsion, as well as partial improvement in infraduction ocular movement from -4 to -2 (Figure 1G, H, I).

In rare cases, establishing a definitive diagnosis can be particularly challenging. In congenital IR hypoplasia, careful interpretation of orbital imaging and intraoperative findings is crucial. Our case demonstrated a distinct difference between the anterior and posterior segments of the muscle belly on sagittal MRI, with intraoperative identification of ciliary arteries leading to a revised diagnosis of hypoplasia. In this case, the surgeon performed a modified procedure involving lateral and medial rectus muscle/tendon splitting to simulate the presence of an IR. This was followed by augmentation with Mersilene sutures on the sclera (augmented modified Hummelsheim) to correct vertical misalignment, along with medial rectus recession of the contralateral eye to address esotropia.

The muscle-splitting technique was specifically designed to preserve the anterior ciliary vessels of each muscle.[4,5] The technique was further developed with the addition of an equatorial fixation suture (augmented suture) by Scott Foster to enhance the verticalization of the transposed muscle

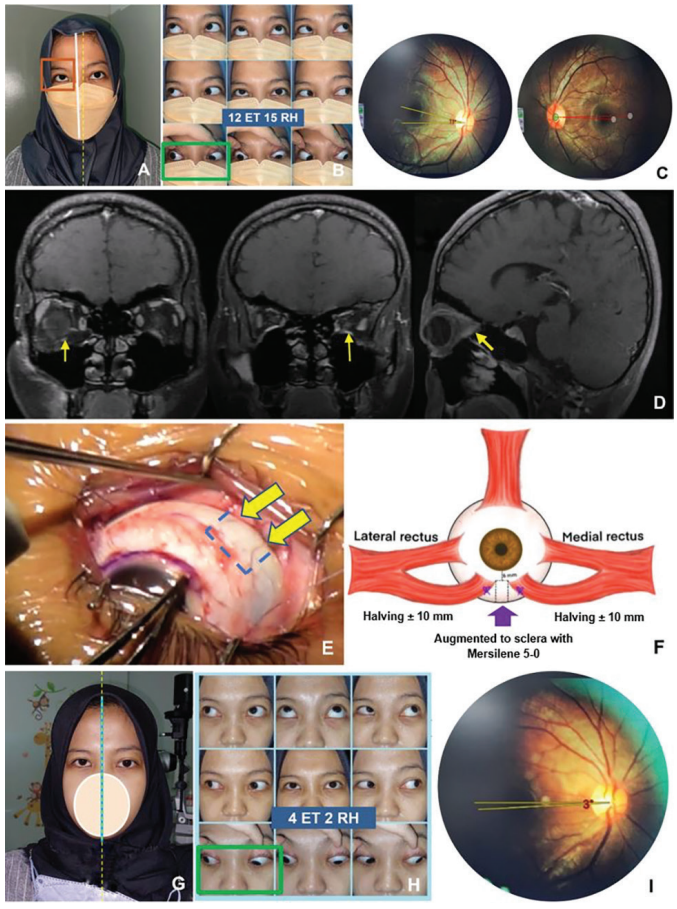

Figure 1. A) Right head tilt and face turn. B) Ocular movement limitation of -4 in depression, more prominent during inferotemporal gaze (green box). C) Posterior pole imaging: incyclotorsion of 11 degrees in right eye (RE), normal in left eye. D) Coronal and sagittal magnetic resonance imaging: a thin muscle sheath was found on the anterior segment of the RE. It became thicker, forming a muscle band on the posterior segment in lieu of the inferior rectus (IR) (yellow arrow). E) Intraoperative findings: two ciliary arteries (two yellow arrows) where the IR should be (blue dash line). F) Augmented modified Hummelsheim procedure: special approaches included transposing the horizontal muscle with a gap between the muscle halves to simulate the presence of an IR. Additionally, Mersilene (polyester) suture augmentation to the sclera (purple knots) was implemented due to the absence of the IR belly (purple arrow). G) At postoperative 6 months, no abnormal head posture was observed. H) Deviation and ocular movement (green box) were improved. I) Incyclotorsion was reduced to 3 degrees (RE)

ET: Esotropia, RH: Right hypertropia

and maximize tonic force. Couser et al.[6] performed an augmented Hummelsheim procedure for total abducens nerve palsy and reported consistently good outcomes. Additionally, the modified Hummelsheim procedure resulted in a lower incidence of anterior segment ischemia, a complication linked to inadequate blood supply to the anterior ciliary arteries caused by muscle manipulation.[4,5,6] Other modification techniques include the modified Nishida, which involves splitting the temporal halves of the muscle without tenotomy. Although this technique requires further evaluation of its efficacy, it is reported to be more beneficial for incomitant horizontal strabismus.[7,8]

In conclusion, a thorough examination and intraoperative exploration are essential for establishing a definitive diagnosis in rare cases of IR hypoplasia. The augmented modified Hummelsheim procedure has demonstrated promising outcomes in improving ocular alignment in the primary position and correcting AHP, consistent with the main goal of the procedure.

Ethics

Informed Consent: Written informed consent for publication obtained.

Declarations

Authorship Contributions

Surgical and Medical Practices: A.P.B., Concept: F.P., A.P.B., Design: F.P., A.P.B., Data Collection or Processing: F.P., A.P.B., Analysis or Interpretation: F.P., A.P.B., Literature Search: F.P., Writing: F.P., A.P.B.

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References

1. Matsuo T, Watanabe T, Furuse T, Hasebe S, Ohtsuki H. Case report and literature review of inferior rectus muscle aplasia in 16 Japanese patients. Strabismus. 2009;17:66-74.
2. Astle WF, Hill VE, Ells AL, Chi NT, Martinovic E. Congenital absence of the inferior rectus muscle--diagnosis and management. J AAPOS. 2003;7:339-344.
3. Solanes F, Demer JL. Clinical and imaging features of congenital and acquired isolated inferior rectus muscle hypofunction. J AAPOS. 2021;25:11.
4. Sheth J, Shinde A, Kekunnaya R. Congenital anomalies of inferior rectus and its surgical outcomes. Strabismus. 2022;30:150-158.
5. Fogel-Tempelhof O, Bachar Zipori A, Stolovitch C, Spierer O. Outcomes of half-width vertical rectus transposition augmented with posterior fixation sutures for sixth cranial nerve palsy. Int J Ophthalmol. 2021;14:1921-1927.
6. Couser NL, Lenhart PD, Hutchinson AK. Augmented Hummelsheim procedure to treat complete abducens nerve palsy. J AAPOS. 2012;16:331-335.
7. Ramkumar AV, Agarkar S, Mailankody S. Modified Nishida's procedure in management of unilateral isolated inferior rectus aplasia – a case report. Indian J Ophthalmol Case Rep. 2021;1:363-364.
8. Nishikawa N, Ito H, Kawaguchi Y, Sato M, Yoshida A. Resection and anterior transposition of the inferior oblique muscle for treatment of inferior rectus muscle hypoplasia with esotropia. Am J Ophthalmol Case Rep. 22017;7:70-73.