Case Report

Surgical Options in Managing Convergent Strabismus Fixus Related to High Myopia

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ABSTRACT

Background: *Convergent strabismus fixus is a condition in which one or both eyes are anchored in extreme adduction. This condition is caused by superotemporal herniation of the enlarged globe from the muscle cone through the space between the superior and lateral rectus muscle. It is commonly related to high myopia. Recently, transposition type procedure which unite the superior and lateral rectus muscles were proposed as the treatment of choice in this condition. However, there is no clear consensus regarding the best surgical procedure in convergent strabismus fixus. This case series reported three rare cases of convergent strabismus fixus related to high myopia and highlight the option of surgical procedure that can be done in these cases.*

Case Illustration: *Three patients presented with convergent strabismus fixus of both eyes with history of bilateral high myopia since childhood. All patients showed Krimsky tests of >95$^{\Delta}$ET preoperatively, with medial displacement of superior rectus and inferior displacement of lateral rectus of both eyes in all patients on orbital imaging. The bilateral Yokoma procedure both resulted in an orthophoric postoperative result with marked improvement in abduction and elevation. The hemi-Jensen also showed a significant improvement of over than 60$^{\Delta}$ on the operated eye, resorting it to a central position during primary gaze.*

Conclusion: *Convergent strabismus fixus is a rare condition and frequently associated with high myopia. Both techniques of Yokoyama procedure or hemi-Jensen procedure combined with medial rectus recession showed similar good results with improvement in position and ocular motility of the eye*

Keywords: *convergent strabismus fixus, high myopia, Yokoyoma procedure, hemi-Jensen procedure*

Strabismus fixus is a rare ocular motor abnormality in which one or both eyes are anchored in extreme adduction or abduction. The eyes may be so firmly fixed in their deviated horizontal position that it is impossible to displace the eyes horizontally, either with volitional movement or with forced traction. When the affected eye is so tightly fixed in an esotropic and hypotropic position, the condition is called as convergent strabismus fixus. This condition could develop from other various causes such as intraocular tumor, amyloidosis and abducens nerve palsy. However, convergent strabismus fixus is more frequently associated with high myopia.[1-9]

Many different theories on the underlying cause of this restrictive disease can be found in the literature but the exact mechanism is not well understood. However, the most recent explanation was provided by Aoki[3] and Yokoyama[10] who noted the superotemporal herniation of the enlarged globe from the muscle cone through the space between the superior and lateral rectus muscles. This dislocation happened because of the axial elongation due to severe myopia. On Magnetic Resonance Imaging (MRI), the superior rectus (SR) muscle was seen to be displaced nasally and the lateral rectus (LR) muscle was displaced inferiorly.[3,4,10,11,12]

Convergent strabismus fixus can only be treated by surgery, but there is no clear consensus on what is the best surgical option in these patients. Generally, there have been two main procedures performed to help correct the restrictive strabismus associated with high myopia. These are resection-recession procedures which mainly alter muscle forces or transposition type procedures which affect the direction of muscle actions. Based on the pathogenesis of a superotemporal herniation of the enlarged globe from the muscle cone through the space between the SR and LR muscle, the transposition type procedures which unite the SR and LR mucles were aimed to restore the dislocated globe back into the muscle cone. Yokoyama et al[17] described a surgical procedure involving loop myopexy of the SR and LR muscles without muscle splitting to restore the dislocated globe back to the muscle cone. While Larsen et al[13] treated convergent strabismus fixus patients with a hemi-Jensen's procedure which unite the lateral half of the SR muscle and the superior half of the LR muscle with muscle splitting.[1,10,13-16]

This case series reports three patients with convergent strabismus fixus related to high myopia. Two cases were treated with Yokoyama procedure combined with MR muscle recession and one case treated with hemi-Jensen's procedure. This case series highlightings the options of surgical procedures that can be done in a rare cases of convergent strabismus fixus.

CASE ILLUSTRATION

Case 1

A 48 years old female with history of high myopia in both eyes since childhood presented with progressive esotropia for several years. From the ophthalmological examination, her visual acuity was hand movement in both eyes uncorrected with pinhole. The streak retinoscopy showed a refraction status of S-25.00 C-4.50x130° (right eye, RE) and S-27.50 C-2.00x90° (left eye, LE). The Krimsky test revealed an esotropia of more than 95$^{\Delta}$ET and both eyes were fixed in a position of esotropia and hypotropia. Ocular motility showed limitation in elevation and abduction on both eyes, confirmed by the forced duction test (FDT). The funduscopy of both eyes revealed myopic fundus with staphyloma posticum. Axial length from the biometry examination were 35.38 mm for the RE and 34.33 mm for the LE. The orbital MRI revealed medial displacement of SR muscle and inferior displacement of LR muscle of both eyes.

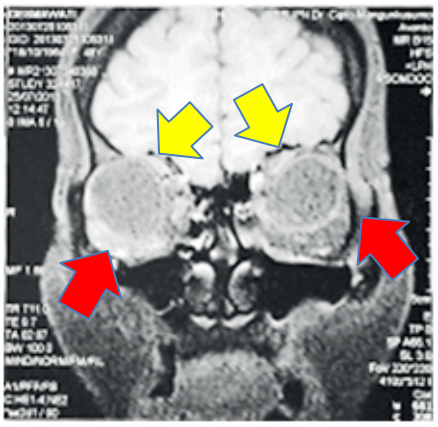

Fig 1. Orbital MRI revealed medial displacement of SR (yellow/upper row arrows) and inferior displacement of LR (red/lower row arrows) of both eyes

The patient was diagnosed as convergent strabismus fixus of both eyes and high myopia of both eyes. She was planned to undergo medial rectus recession and Yokoyama procedure for both eyes. The surgery consisted of a 7 mm medial rectus (MR) recession and transposition of the superior rectus (SR) and lateral rectus (LR), with a loop myopexy at 12 mm from the limbus and anchoring of the suture to the sclera. The surgery was performed under general anesthesia on the left eye and 3 weeks later the surgery was performed on the right eye as well. Four months postoperatively, the patient had no complaints and was orthophoric. Ocular movement was improved but with residual restriction, especially in abduction and elevation.

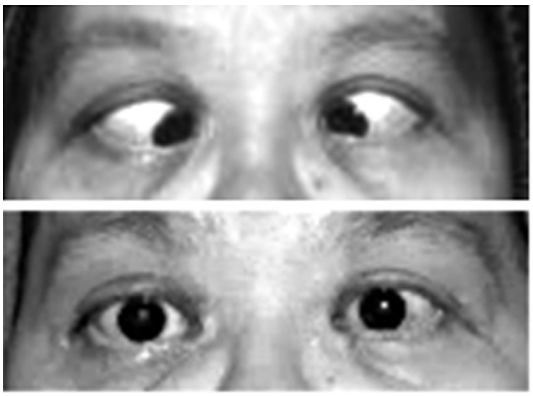

Fig 2. Improvement of eye position (upper image: pre-operative; and lower image: 4-months post-operative)

Case 2

A 31 years old male with history of high myopia in both eyes since 8 years ago presented with progressive esotropia for 5 years. He also noticed the decreasing ability to abduct and elevate both eyes. He neither had a history of trauma nor prior ocular surgery. From the ophthalmological examination, visual acuity of the RE was 0.5/60 uncorrected with pinhole and visual acuity of the LE was 1/60, corrected with S-23.00 into 6/20f$_2$. The Krimsky test revealed more than 95$^{\Delta}$ET. Both eyes were fixed in a position of esotropic and hypotropic condition. Ocular motility of both eyes were restricted in elevation and abduction, confirmed by FDT. Anterior segment of both eyes were within normal limits. The funduscopy of RE showed positive fundus reflex but the details were hard to be evaluated since the pupil was fixed in an inward position, while the LE showed myopic chorioretinal degeneration. The RE axial length was 32.62 mm and 31.46 mm for the LE. The orbital computed tomography scan (CT scan) revealed medial displacement of SR muscle and inferior displacement of LR muscle of both eyes.

The patient was diagnosed as convergent strabismus fixus and high myopia of both eyes. He was planned to undergo medial rectus recession and Yokoyama procedure for both eyes. The surgery consisted of an 8 mm MR recession and transposition of the SR and LR with loop myopexy at 12 mm from the limbus, and anchoring the suture to the sclera. The surgery was performed under general anesthesia on the left eye and 2 weeks later the surgery was performed on the right eye as well. Three months postoperatively, the patient had no complaint and was orthophoric. Krimsky test showed 8$^{\Delta}$ ET. As the first patient, ocular movement was improved but restriction was found especially in abduction and elevation.

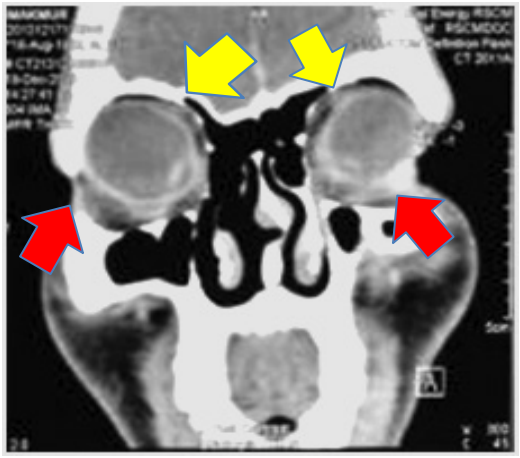

Fig 3. Orbital CT-scan revealed medial displacement of SR (yellow/upper row arrows) and inferior displacement of LR (red/lower row arrows) of both eyes.

Fig 4(a). Pre-operative pictures of case 2 showing Hirschberg >45^{o}ET and limited movement of both eyes especially in abduction and elevation, compared to **Fig 4(b)** Three months post-operatively, showing residual Krimsky about 8$^{\Delta}$ET and improvement of ocular movement

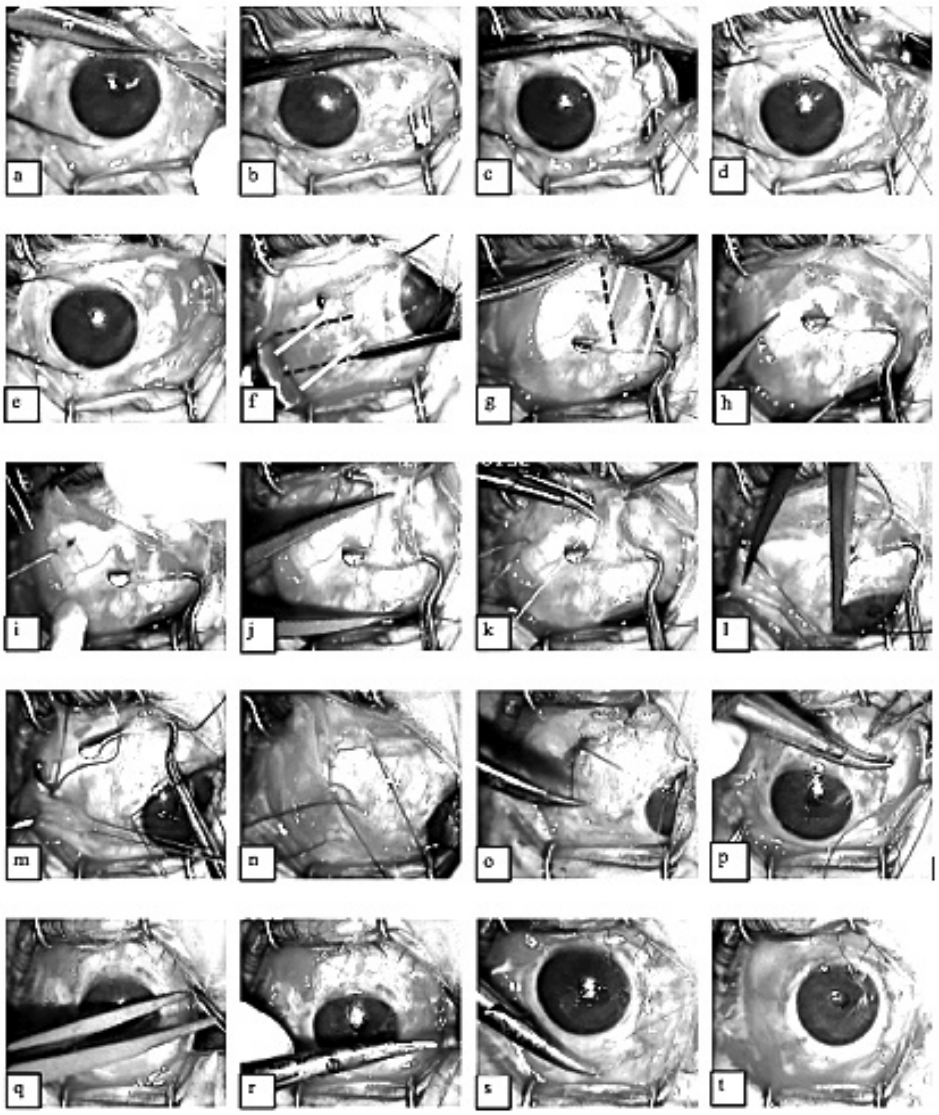

Fig 5. Medial rectus recession and Yokoyama procedure of the RE in case 2. **Fig 5a,b**. A limbal based peritomy was made at the medial quadrant and MR muscle was identified. **Fig 5c,d,e**. Djoko Sarwono's suture was made at the MR muscle and the muscle was cut near its insertion. **Fig 5f**. Identification of the LR muscle and the muscle looked displaced inferiorly (yellow line). **Fig 5g.** Identification of the SR muscle and the muscle looked displaced nasally (yellow line). **Fig 7h-o.** A mersilene 4.0 suture was used to unite the lateral part of SR and superior part of the LR at 12 mm from the limbus (without splitting the muscle). The suture was anchored to the sclera. **Fig 7p,q,r.** A 8 mm recess procedure was made at the MR muscle. **Fig 7s**. The conjunctiva was sutured. **Fig 7t.** The operation was done.

Case 3

A 46-years old female with history of high myopia in both eyes since 15 years ago presented with progressive esotropia for 5 years. She also noticed the decreasing ability to abduct and elevate both eyes. Previously, she complained that her left eye was turning to inward since 12 years before admission. Two years later, she went to an ophthalmologist and underwent a squint surgery on her left eye. After the operation, the position of the LE was straight. Past medical history were unremarkable with no history of trauma, head injury, diabetes mellitus nor malignancies. She had already undergone cataract extraction surgery for her right eye. From the ophthalmological examination, her visual acuity was hand movement in both eyes, the Krimsky test revealed more than 95$^{\Delta}$ET. With slight hypotropia, and both eyes were restricted

in elevation and abduction. Anterior segment of both eyes were within normal limits. There was intraocular lens in the RE with posterior capsul opacification (PCO) and grade 4 haziness of the lens of the LE. Funduscopy of the RE showed myopic chorioretinal degeneration, while funduscopy of the LE was hard to be evaluated due to haziness of the lens. The forced duction test revealed restriction in abduction and elevation of both eyes. Axial length from the biometry examination were 27.91 mm for the RE and 29.01 mm for the LE. The orbital CT-Scan revealed medial displacement of right superior rectus of both eyes.

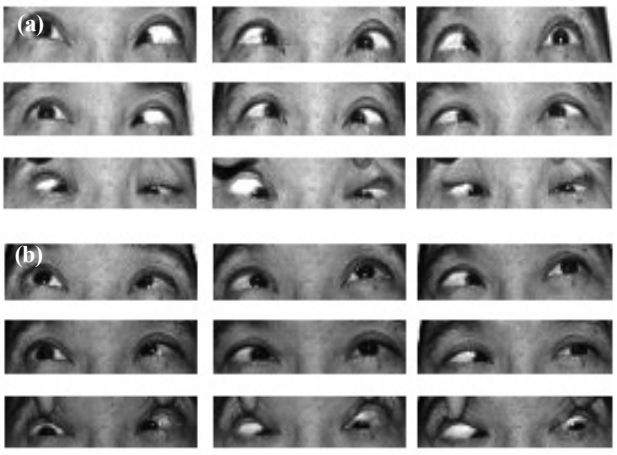

Fig 6(a). Pre-operative pictures of case 3 showing Hirschberg >45°ET and limited movement of both eyes especially in abduction and elevation, compared to **Fig 6(b).** Three weeks post-operative picture showing a residual Krimsky of about 35ΔET but fixating central LE with improved ocular movement towards abduction and elevation.

The patient was diagnosed with convergent strabismus fixus of both eyes, pseudophakia of the RE with PCO and mature cataract of the LE. She was planned to undergo hemi-Jensen's procedure for both eyes, and the first surgery was performed on her LE consisting of transpositions of the SR and LR muscle. Unfortunately, the patient refused to do the surgery on her RE, accounting for a residual esotropia of 35ΔET (by Krimsky) three weeks post-operatively. The LE regained its central position in primary gaze, with much improved abduction and elevation compared to the unoperated RE.

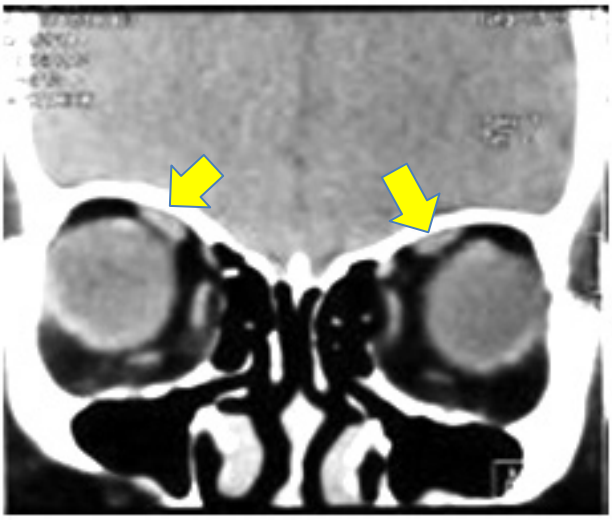

Fig 7. Orbital CT-scan revealed medial displacement of SR (yellow/upper row arrows) of both eye

DISCUSSION

Strabismus fixus is a rare ocular motor abnormality in which one or both eyes are anchored in extreme adduction or abduction. When the affected eye is so tightly fixed in an esotropic and hypotropic position, the condition called as convergent strabismus fixus. Diagnosis of convergent strabismus fixus could be made clinically because of the typical clinical presentation which is fixed esotropic and hypotropic position of the involved eye with the limitation of movement especially in abduction and elevation. This diagnosis also supported by positive force duction test. The forced duction test will reveal loss of elasticity and contracture of the medial rectus.[1-4,7] Three cases that presented in this case series had characterisic features of convergent strabismus fixus such as both eyes are fixed in esotropic and hypotropic condition with limitation in abduction and elevation. All cases in this case series also had positive result of force duction test that showed restriction in abduction and elevation. There were also history of high myopia of both eyes in all cases. There were no histories of tauma, head injury, diabetes mellitus nor malignancies in all cases.

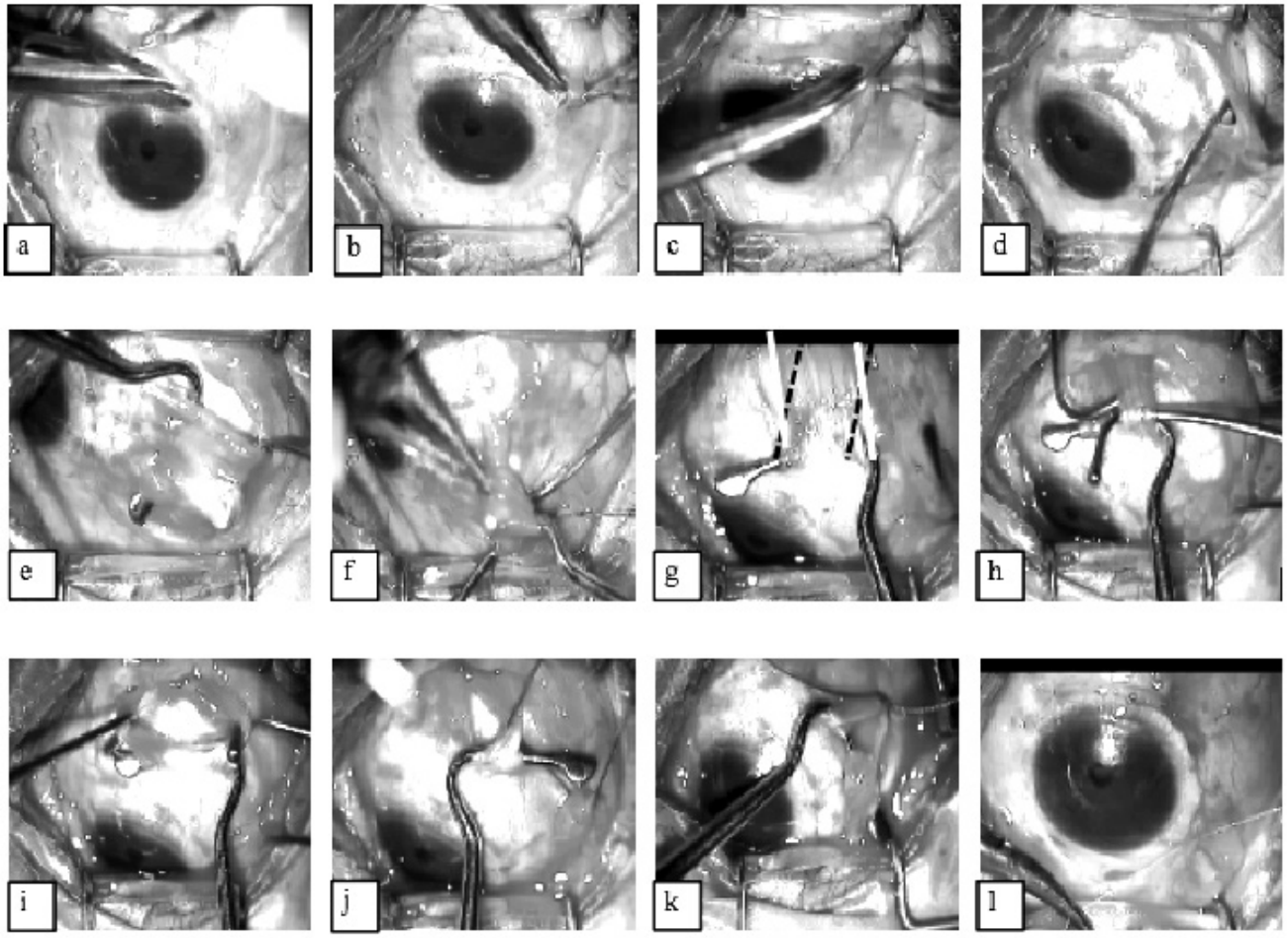

Fig 8. Hemi-Jensen procedure of the LE in case 3. **Fig 8a,b,c.** A limbal based peritomy was made at the superior quadrant. **Fig 8d,e.** Identification of the LR muscle. **Fig 8f.** A muscle hook penetrate through the middle part of the LR and splitted the muscle from the insertion to the equator. **Fig 8g.** Identification of the SR muscle and the muscle looked displaced nasally (yellow line). **Fig 8h,i.** The SR muscle was splitted with a muscle hook from the insertion to the equator. **Fig 8j,k.** The lateral half of the SR muscle was united to the superior half of the LR muscle with mersilene 4.0 suture. **Fig 8l.** The conjunctiva was sutured

Many different theories regarding the etiology of acquired esotropia associated with high myopia have been reported but the exact mechanism is not well understood. Bagolini et al[7] assumed that in this condition, there was myopathic paralysis of the LR by the pressure from the lateral orbital wall. In a study using MRI by Demer and Von Noorden, globe elongation was the cause of restricted ocular motility. Rotation of the globe was limited due to contact between the posterior aspects of the elongated globe and the bones of the orbital apex.[8]

Miller first demonstrated MRI evidence that the rectus muscles have paths that are constrained anteriorly in the orbit by connective tissues called as "pulleys." Miller proposed that pulleys were responsible for maintaining the stability of the deep paths of the rectus muscles, preventing them from side-slipping over the globe in secondary and tertiary gazes. The superior rectus (SR) and lateral rectus (LR) pulleys are normally joined by a ligament, the "LR-SR band," that suspends the LR pulley and prevents the globe from dislocating superotemporally. A connective tissue abnormality expressed in both the sclera and LR-SR band may be the common factor allowing both development of axial high myopia and LR-SR band degeneration permitting superotemporal globe dislocation. There is likely to exist a genetic association between the axial high myopia and LR-SR band degeneration.[17,18]

Recently, several studies have paid special attention to the shift of the extraocular muscle (EOM) path. These studies reported that shifting of the EOM path was caused by a posterior prolapse of an elongated eyeball beyond the muscle cone in the orbit, which stretches and shifts the EOMs.[3,10,11,16,17,19,20] Krzizok et al[11] reported marked inferior displacement of the LR path on MRI in patients with esotropia and hypotropia. This displacement reduces abducting torque of the LR and creates depressing torques. Yokoyama et al[10] reported that both the LR and SR path were significantly displaced. The LR muscle was displaced inferiorly and the SR was displaced nasally. Aoki et al[3] also reported that LR shifts inferiorly and SR shifts nasally in acquired esotropia with high myopia. Both Yokoyama and Aoki suppose that both shifts are caused by the elongated posterior portion of the eyeball due to high myopia. They found superatemporal eyeball prolapse from the muscle cone.[3,10] Aoki et al[3] hypothesized that inferior shift of the LR causes reduced abduction and increased infraduction. Nasal shift of SR causes reduced supraduction and increased adduction. Consequently, these patients show esodeviation and hypodeviation due to increased adduction, reduced abduction, increased infraduction and reduced supraduction.[1,3]

In this case series, the displacement of the extraocular muscle path were confirmed with MRI or CT scan and observation during the surgery. The first and second case had medial displacement of SR and inferior displacement of LR of both eyes, while the third case had medial displacement of right superior rectus of both eyes. These displacement of extraocular muscle path allowing superotemporal herniation of the enlarged globe from the muscle cone and made a a condition of fixed esotropia and hypotropia with restriction of elevation and abduction in all cases.

Surgical management of convergent strabismus fixus can be difficult. There is no clear consensus on what is the best surgical option in these patients. The correction of this misalignment has frequently been disappointing. A good surgical result is considered repositioning the eyes in the primary position, but it had to be acknowledged that a full rotations were cannot be restored. A number of different surgical procedures have been reported. Conventional manouvers are resection-recession procedure that mainly affect muscle force. Modified procedures includes transposition techniques that alter muscle paths.[1,20,21]

Hayashi et al[15] found that the traditional recession and resection surgery may be effective only in early stages for acquired esotropia associated with high myopia patient. If the imaging findings show the abnormal LR path, the common procedure of MR recession and LR resection may aggravate hypodeviation. The resection of the dislocated LR muscle enhances abduction and infraduction, esotropia reduces, but hypotropia increases. Because of this, imaging studies is needed to confirm whether there is a displacement of extraocular muscle path or not because this findings will determine is a resection-recession procedure still has position as the therapy or not.[20,21]

As mention earlier, recent studies have paid special attention to the shift in extraocular muscle path in convergent strabismus fixus. This led to the consideration of transposition procedures. Yokoyama et al[10] performed loop myopexy of the LR and SR muscles to treat patients who had progressive esotropia with high myopia. Later, Larsen et al[13] treated patients with highly myopic acquired esotropia using hemi-Jensen procedure of the SR and LR muscles without MR muscle recession. Their results were effective and practical. Such surgeries may re-establish the physiological muscle plane.[10,13]

Loop myopexy of the LR and SR without muscle splitting was first reported by Yokoyama et al[10] and then Wong and Basmak also used this technique in cases of myopic strabismus fixus.[10,22,23] The surgery changed the SR and LR's muscle position and redirected their motility's plane, thus there were considerable limitations in abduction and elevation postoperatively. If the loop was anchored to the sclera, the risk of scleral perforation existed, noting that in high myopic patient the sclera was also thinner than normal condition. This procedure can be combined with MR recession or can be done independently. Several studies said that an additional recession of MR especially recommend in cases where forced duction confirmed tight muscle.[21] In this case series, the first and second patient was

treated with Yokoyama procedure combined with recession of MR of both eyes. The Yokoyama suture were all anchored to the sclera in these two cases and there were no scleral perforation. The Yokoyama procedures were combined with MR recession because in these cases, the forced duction revealed restriction in abduction in all eyes. Post operative, these two cases achieve good results. The first case showed only residual 6$^{\Delta}$ET with improvement in abduction and elevation, while the second case showed residual 8$^{\Delta}$ET, also with improvement in abduction and elevation. Anterior segment of both cases were within normal limits with no sign of anterior segment ischemia.

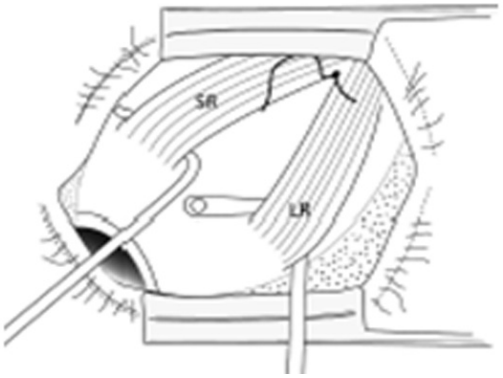

Fig 9. The SR and LR joined with small bites from both muscle bellies without splitting the muscle[21]

Other alternatives in transposition procedure was the hemi-Jensen's procedure with or without MR recession in convergent strabismus fixus cases. The hemi-Jensen's procedure, compared with loop myopexy, preserves the unsecured halves of the SR and LR muscles, which may contribute to the circulation of the anterior segment. Thus, we consider the hemi-Jensen's procedure to have a lower risk of ocular ischemic syndrome than loop myopexy. In addition, the benefit of this procedure was also the elimination of the risk of scleral perforation as no suture was placed on the sclera.[2,13,16] The third case in this case series was treated with hemi-Jensen's procedure without MR recession. The MR recession was not performed due to the consideration of the patient already had a history of squint surgery to correct esotropia 10 years ago, presumably a resection-recession procedure, so the operator only done the hemi-Jensen's procedure in this patient. The hemi-Jensen's procedure was only carried out on the LE but has already shown marked improvement of the overall deviation from more than 95$^{\Delta}$ET to 35$^{\Delta}$ET (over 60$^{\Delta}$). Ocular movements were also significantly improved. Inspite of that, the second surgery for the right eye is still needed to complete the alignment of both eyes and improve function in terms of visual field and motility.

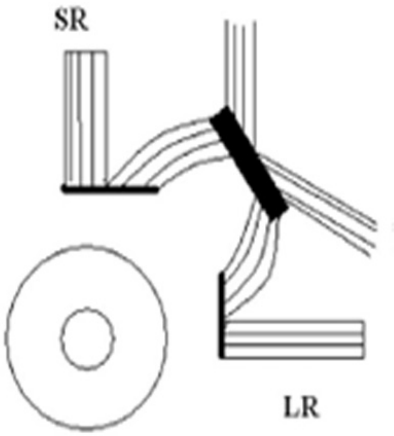

Fig 10. Hemi-Jensen's procedures[2]

CONCLUSION

Convergent strabismus fixus is a rare condition and frequently associated with high myopia. There has been no clear consensus regarding the best surgical treatment for this condition. This case series presents two different techniques of transposition surgery performed on three patients, which is combination of medial rectus recession and Yokoyama procedure tha was performed on three patients, and a hemi-Jensen's procedure without MR recession on one patient. Both techniques gave good results with improvements in terms of position and ocular motility range of the eyes.

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