Case Report

Augmented superior rectus transposition surgery for vertical strabismus in moebius syndrome

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Abstract

Introduction: Moebius syndrome is a rare disease characterized by unilateral or bilateral congenital nonprogressive facial nerve palsy along with limitation of ocular abductions. Vertical Rectus Transpositions with posterior fixation suture is known to correct abduction deficiencies in case of Moebius syndrome. Traditionally both superior and inferior rectus transposition are done to prevent any post operative vertical imbalance. The purpose of reporting this case is to evaluate superior rectus transposition augmented with posterior fixation suture along with bilateral recession of medial rectus as a useful and safe alternative for treating large esotropia and abduction limitation with a significant vertical deviation in patients of Moebius syndrome. **Case:** We report a rare case of a seven year old male child with large esotropia and with limited ocular abductions along with a significant vertical deviation which is not common in classic Moebius syndrome. We performed a superior rectus transposition in the eye with vertical deviation along with bimedial recession and our post operative results indicated a significant correction in the horizontal as well as vertical deviation along with an improvement in head posture. Conclusion: We advocate a superior rectus transposition surgery in cases of moebius syndrom whenever there is a significant vertical deviation.

Keywords: moebius syndrome, transposition, superior rectus

Introduction

Moebius syndrome is a rare congenital, non-progressive neurological disorder characterised by complete or partial bilateral facial paralysis with bilateral or unilateral abducens paralysis. It is named after Paul Julius Moebius, a neurologist, who first described the syndrome in 1888. Hypoplasia of tongue, swallowing and speech difficulties owing to multiple cranial nerve palsies and anomalies of musculoskeletal system are be the associated features of this syndrome. It is usually a consequence of a sporadic ischemic event resulting in degeneration of nuclei of sixth and seventh cranial nerves during early foetal development,There are few documented familial caseshave also been identified.. It is unlikely to be a single entity, as variety of pathological disturbances may produce a phenotype recognisable as Moebius syndrome.

Clinically patients present with esotropia and limitation of abduction with incomplete closure of eyes.

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Case History

A seven year old male child visited our hospital with complaints of incomplete closure, watering, and inward deviation of eyes. History revealed a full-term uncomplicated pregnancy with no history of maternal alcohol or drug ingestion. Systemic evaluation did not reveal any abnormality except for a bifid tongue. **Figure 1A.**

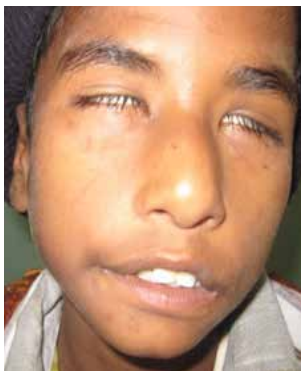

Figure 1A

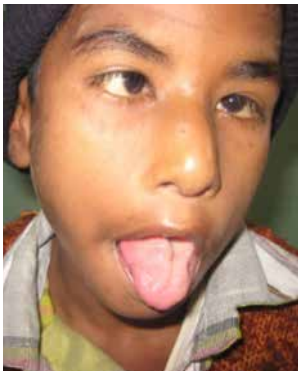

Figure 1B

Oculofacial examination showed expressionless face, loss of nasolabial folds, entropion of upper lids in both the eyes with misdirected lashes and lagophthalmos as shown in **Figure 1B**

His best corrected visual acuity was 6/24 with +1.5DS in the right eye and 6/36 with +2DS/-1.75 DC 180?in the left eye with a binocular visual acuity of 6/12 for which he adopted an abnormal head posture with a 25 degree right face turn and a 10 degree left head tilt measured by a Goniometer. **Figure 2**

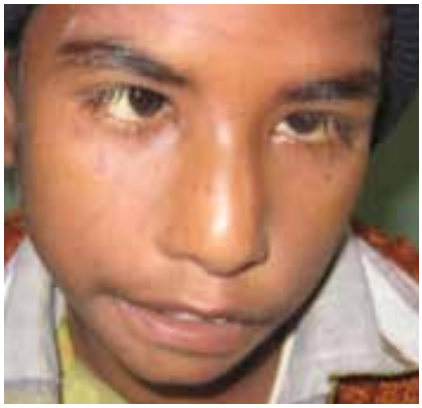

Figure 2: anomalous head posture

Ocular motility examination revealed bilateral abduction limitation of -5 (not reaching up to midline) and adduction limitation of (- 2) in both eyes which made the patient fixate with the deviated eye resulting in an anomalous head posture **Figure 2**

Krimsky test revealed a large esotropia of 60 PD with a left hypertropia of 12PD **Figure 3.**

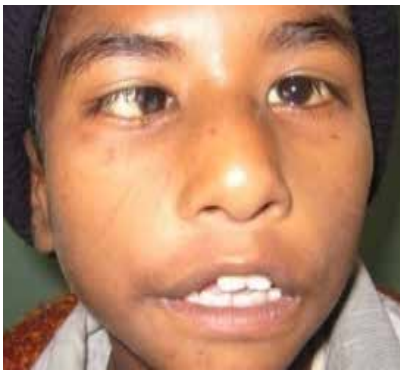

Figure 3: large Esotropia

Sensory evaluation and stereo testing was done using a Worth Four Dot test and Titmus fly test both of which revealed left eye suppression and absent stereopsis. Since the child complained of watering of eyes he was operated for entropion of the upper lids in both eyes about a month before he was taken up for strabismus surgery. Intraoperative forced duction test under general anaesthesia revealed tight (forced abduction possible just upto midline) medial rectus muscles in both eyes. Bilateral large (7mm) hemi hangback recession of medial rectus was performed along with augmented superior rectus transposition in the left eye.

The child was followed up post operatively on day 1 **(Fig 4 a)**, 1 week, 4 weeks **(Fig 4 b)** and after 3months.

Figure 4a: primary position postoperative day 1

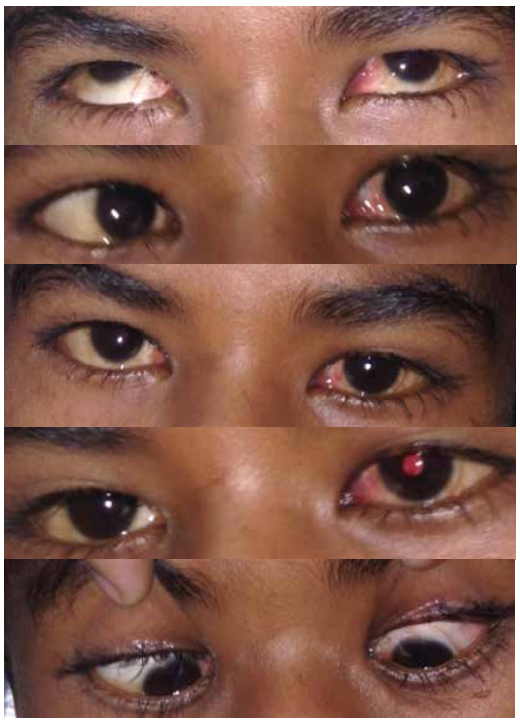

Figure 4b: Cardinal gazes postoperative at 4 weeks

Figure 4

On his final follow up at 3 months, his best corrected visual acuity was 6/9 in the right eye and 6/12 in the left eye and binocular vision was 6/9. He had a mild head tilt with glasses **(FIG 5)**

Figure 5: Orthotropia with a small head tilt

Without spectacles he had a residual esotropia of 16 PD and right hypertropia of 7 PD in the primary gaze but with spectacles he was orthotropic horizontally with a small right hypertropia. His ocular motility showed mild improvement of abduction (improving to -3) in the left eye. **Figure 6**

Figure 6: postoperative results without spectacles

Discussion

Vertical deviations are not very common in classic Moebius syndrome. Amaya et al reported a comprehensive series of 18 patients of Moebius syndrome of whom 9 had a horizontal gaze pattern and 3 had vertical limitations. There is a paucity of studies designed to investigate whether different phenotypical expressions of the disease exists worldwide. Carta et al1 has described three patterns of presentation of Moebius syndrome: the type C pattern exhibits exotropia and vertical deviation. The type A and Type B with esotropia don't exhibit a vertical deviation.

Even though many reports have described ocular and systemic features of Moebius syndrome, only a few articles have reported the results of strabismus surgery in these children. The types of surgery mentioned are: bilateral medial rectus recession, medial rectus recession with lateral rectus resection, and transposition of vertical rectus muscles Our case had a large esotropia with abduction deficits and a significant vertical deviation which was corrected using large bilateral medial rectus recession (7mm) along with unilateral augmented superior rectus transposition to improve abduction and to treat vertical deviation. Sun et al in 2011 described the surgical outcomes of augmented vertical rectus

transposition in 3 patients of Moebius syndrome who presented with moderate to large angle esotropia. According to them, with the absence of medial rectus contracture, the augmented vertical rectus transposition (VRT)procedure alone appears to be more efficacious in correcting moderate- to large-angle esotropia as compared to medial rectus recession alone. However, when significant medial rectus muscle contracture is present, medial rectus muscle weakening by either chemodenervation or surgical recession is likely to be required. Since our case had a significant medial rectus contracture we decided to combine medial rectus recession with augmented superior rectus transposition using technique similar to one described by Sun etal; the only difference being we did one vertical (superior) rectus transposition only in one eye.

Till date there is no published literature on advocating superior rectus transposition for correcting abduction deficit in such cases. This procedure may also be safer in terms of preventing anterior segment ischaemia.

The limitations of our case report are that we were not been able to do a preoperative MRI to look for specific areas of damage, and to confirm Moebius syndrome. Also binocular diplopia free fields, useful for the functional evaluation of patients with paralytic or restrictive strabismus, could not be performed. Cognitive evaluation was not investigated specifically in the present study but was based on clinical observation

We advocate large bilateral medial rectus recession in patients presenting with large esotropia due to medial rectus contracture along with augmented superior rectus ransposition for significantly enhancing outcome in patients of Moebius syndrome.

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