Monocular elevation deficiency: when the involved eye takes up fixation

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Introduction

Monocular elevation deficiency (MED) is a form of restrictive strabismus, characterized by the unilateral limitation of elevation and associated ptosis.1 The inability of the eye to elevate persists across the horizontal plane, from adduction to abduction. It is usually a sporadic condition.2 This condition was previously described by Dunlap3 as Double Elevator Palsy and the limitation of elevation was attributed to the paralysis of both the superior rectus and the inferior oblique muscle. MED can be congenital or acquired. Congenital MED is often seen in identical twins and it is caused by either of the following4: (a) paralysis of the superior rectus muscle without any involvement of the inferior oblique muscle. (b) Primary inferior rectus restriction. Metz found inferior rectus restriction without elevator weakness in 73% of MED patients.5 (c) Lesion in the supranuclear pathway of upgaze located in the pretectum. (d) Inferior rectus restriction secondary to long-standing superior rectus palsy.

Acquired MED is seen in (a) cerebrovascular diseases, (b) tumours of the mid-brain, (c) syphilis, (d) sarcoidosis.1-4 Congenital MED presents with (i) the inability to elevate the eye above midline in abduction, adduction or from primary position of gaze.4 (ii) Hypotropia of the affected eye on the affected side. However, one-third of the cases present as orthophoria in primary gaze.2 Rarely the affected eye may fixate. (iii) Ptosis of the affected eye or sometimes pseudoptosis may be seen. (iv) Chin-up position.

Important investigations include forced duction test, active force generation test, saccadic velocity measurements, and magnetic resonance imaging of brain.5 Some other conditions may mimic MED such as Brown syndrome, thyroid eye disease, congenital fibrosis of inferior rectus muscle, orbital floor fracture and myasthenia gravis.4,5

The indications for surgical interventions are (a) large vertical deviation in primary gaze, (b) vertical deviation causing suppression and amblyopia, especially in children and (c) abnormal head posture.4 Now we are going to describe a rare case of MED where the patient is fixating with the affected eye.

Case Report

An 11-year-old male child presented with diminution of vision and upward deviation of right eye for 3 years. He had undergone a surgery for bronchial fistula at 7 years of age. He was a full-term baby with normal birth weight. There was no history of delayed developmental milestones. There was neither any history of ocular disorder in the family nor any history of consanguinity.

Figure 1: (a) Chin-up position. (b) Left eye taking up fixation.
Examination revealed visual acuity in the right eye to be 6/36 and in the left eye to be 6/6. He had a chin-up (~30°) position as shown in Figure 1(a). At primary position of gaze, the right eye was hypertropic and the left eye was hypotropic. Left eye was the fixating eye (Figure 1(b)). Ocular motility was full in the right eye, but there was marked limitation of elevation from primary position, in abduction and adduction in the left eye. There was globe retraction and unnatural convergence on attempted elevation. Mild to moderate ptosis was seen in the left eye which remained constant in all gaze (Figure 2). Fusional status was assessed by Worth 4-Dot test for distance as well as for near, and it revealed right eye suppression. Stereo acuity was about 140 s of arc. The alternate prism cover test showed 35RHT, 20ET at primary gaze, 40RHT, 30ET at upgaze, 20RHT at downgaze, 30RHT, 18ET at right gaze and 30RHT, 20ET at left gaze for distance and 12RHT, 15XT for near (Figure 3). Rotatory nystagmus was seen in both of the eyes. Pupillary reaction was normal. Slit lamp findings, intraocular pressure measured by Goldmann Applanation Tonometry and fundus findings of both of the eyes were within normal limit. MRI brain and orbit revealed cord-like thick inferior rectus muscle of left eye suggestive of fibrosis. FDT, performed preoperatively, was positive for left inferior rectus and right superior rectus under anaesthesia. Left inferior rectus recession (5 mm) and right eye superior rectus (3 mm) recession were performed under general anaesthesia. Postoperatively there was improvement in

Figure 2: Preoperative photos showing left eye hypotropia in primary gaze, limitation of elevation in upgaze from primary position, in adduction as well as in abduction.

Figure 3: Strabismus evaluation by alternate prism cover test.
elevation in the left eye as shown in Figure (4). He could fixate with both eyes. He was advised occlusion of left eye.

Discussion
The child shows fixation preference for the involved eye, i.e. the left eye. When the hypotropic left eye fixates, there is an overshoot in the right eye. This is based on the Hering’s law, where an excess innervation is required by the left superior rectus muscle to fixate leading to concomitant excess supply to the elevators of the right eye, thus causing hypertropia.5, 6 The hypertropia in the right eye has been long standing which is justified by the chin-up position adapted by the child to facilitate fusion.5 Eventually amblyopia has developed in the right eye leading to low vision. Ptosis is due to levator palpebrae superioris weakness. The persistence of ptosis when left eye is fixating rules out pseudoptosis which is seen in hypotropia due to the fascial attachments between the levator palpebrae superioris and superior rectus and disappears as the affected eye takes up fixation.4 The fact that it is long standing has been reinforced by the MRI findings suggestive of inferior rectus fibrosis of the left eye. Preoperative FDT indicates restriction of left inferior rectus, which may be primary or secondary.5 Surgical intervention is indicated as the child has already developed suppression and amblyopia. To align the hypertropic right eye in primary position weakening of right eye superior rectus is needed in addition to left eye inferior rectus recession.5, 7

Occlusion therapy should be continued and the child should be followed up regularly.

Conclusion
A thorough ocular examination should be performed, including ocular motility, test for fusion and stereo acuity, cover, uncover and alternate prism cover test to reach a diagnosis. FDT holds importance in diagnosing the cause and management is based on the results of FDT. Early detection is important to prevent suppression and amblyopia of the normal eye.

References

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