A case of A-pattern esotropia with dissociated vertical deviation with bilateral congenital ptosis

Archana Gupta, DNB; Suma Ganesh, MS, DNB; Saman Adil, MS

Dissociated vertical deviation (DVD) is an enigmatic strabismus entity for which various theories of causation have been proposed. Commonly seen in association with infantile esotropia, it responds unpredictably to surgical intervention hence fuelling theories for a supranuclear center defect albeit at the midbrain level. Cases of DVD associated with A-pattern esotropia and superior oblique overaction have been reported, although rarely. We report a case of bilateral congenital ptosis associated with this complex. Not reported earlier, we hope this case may help in better understanding of the obscure etiology of this symptom complex.

Key words: A-pattern, dissociated vertical deviation, pathophysiology, ptosis, superior oblique overaction

Indian J Ophthalmol 2008;56:521-3

An eight-year-old boy presented with alternating esotropia noticed since three or four months of age and drooping of both eyelids since birth. On examination, he had an uncorrected visual acuity of 20/20p in both eyes. Full cycloplegic refraction under cyclopentolate 1% eye drops of +4.00 diopters (D) in both eyes was prescribed. However, as this caused a marked blurring of vision for the eight-year-old child, interfering with his studies, a post-cycloplegic test with fogging was done and he was then prescribed +1.50 D (20/20) in both eyes. On external examination, he had bilateral severe ptosis [Figure 1 left] with a palpebral fissure height of 4 mm, the marginal reflex distance-1 (MRD 1) was -1, the levator function was 8 mm and Bell's phenomenon was good.

On orthoptic evaluation with glasses, he had a chin up head position and on cover test, there was alternating esotropia with DVD in both eyes. On alternate prism cover test for distance, there was an esotropia of 25 prism diopters (pd) in the primary position increasing to 35 pd in up gaze and no horizontal deviation in down gaze. The right eye had a DVD of 20 pd in the primary gaze, 25 pd in dextroversion and absent in levoversion. The left eye had a DVD of 18 pd in the primary gaze, 25 pd in levoversion and absent in dextroversion. For near, he had an esotropia of 35 pd and a DVD of 20 pd and 18 pd in the right and left eye respectively. Both eyes had a superior oblique overaction of +4 and an inferior oblique underaction of -2 [Figure 2].

There was no change in the deviation on head tilt to either side. Sensory evaluation showed: alternate suppression on Worth four-dot test and no stereopsis as measured by the Randot E test. The rest of the anterior and posterior segment evaluation was unremarkable.

An impression of A-pattern esotropia with incomitant DVD with bilateral superior oblique overaction with ptosis was made and a two-stage surgery was planned; squint surgery in the first stage followed by ptosis surgery later.

Forced duction test under anesthesia did not demonstrate any restrictions. He underwent bilateral medial rectus recession of 4 mm, superior rectus recession of 8 mm and superior oblique posterior 7/8th tenectomy. We used a conjunctival limbal approach and standard surgical technique.

Postoperatively, on orthoptic evaluation, the esotropia for distance had decreased to 5 pd in the primary position, 10 pd in up gaze and there was an esotropia of 8 pd in down gaze. The A-pattern had decreased from 35 to 18 pd [Figure 3]. The DVD had decreased in the right and left eyes to 7 and 10 pd respectively; superior oblique overaction was still present. On sensory evaluation there was no fusion or stereopsis. There was no change in the ptosis after strabismus surgery.

In both eyes levator resection of 28 mm (supermaximal) was done three weeks after the squint surgery. Postoperatively [Figure 1 right], the palpebral fissure increased to 12 mm and 11 mm in the right and left eye respectively; the MRD1 was 3 mm both eyes and minimal lagophthalmos was present.
Infantile esotropia may be associated with DVD in 40-90% of cases. A or V patterns are generally associated with oblique muscle dysfunction. Our case was most likely infantile esotropia untreated and undiagnosed in early childhood with associated ptosis. The fact that he was alternating well and had similar DVD both eyes helped him to maintain good vision.

The DVD associated with superior oblique overaction is generally incomitant, increasing in abduction and decreasing in adduction as in our patient. In such cases, overaction of the superior obliques causes the incomitance.

This strabismus complex is in itself relatively rare. Helveston first described this symptom complex in a case report in 1967. This case gained significance as it was believed to disprove the theory of underaction of superior obliques in the causation of DVD. Rosenbaum reported seven such cases in 1991; he maintained that this entity was still possible in the presence of superior oblique underaction based on the fact that the eye elevates and undergoes excyclotorsion in DVD. In a commentary on this article, Helveston however used the occurrence of this group of symptoms to further his supranuclear theory for the causation of DVD.

The unique feature in this case was the bilateral congenital ptosis. A thorough literature search did not yield any reported case of ptosis with this constellation of signs. Out of two large case series of ptosis associated with strabismus, only one reports a case of esotropia with unilateral DVD with bilateral congenital ptosis. However, there is no comment on the pattern and the levator function in this case was poor at 2 mm as compared to 8 mm in our case.

Infantile esotropia, according to one school of thought is believed to be an essentially cortical defect; it appears to be a plausible explanation considering its association with birth asphyxia and neonatal insults. Dissociated vertical deviation in its present state is less understood; it's pathogenesis still remains elusive although various theories have been proposed; one widely accepted theory is the supranuclear theory proposed by Biedlowsky and expanded by Helveston.

A possible explanation for the ptosis in this case can be a common supranuclear etiology for the entire symptom complex; further investigation is required. If this complex does originate in the higher centers, why this association has not been previously reported cannot be explained. One explanation can be that because of lack of documented association, practitioners may dismiss the ptosis as only a coincidental myogenic ptosis in most cases.

Although by a single case report we cannot comment on the etiology of the entire symptom complex we would like to highlight this constellation of signs. Such case reports may spark a debate and help in better understanding the obscure etiology of this symptom complex.

### References
1. Santiago AP, Rosenbaum AL. Dissociated vertical deviation. In: Rosenbaum AL, Santiago AP, editor. Clinical strabismus management. Principles and surgical techniques. Philadelphia: W B Saunders Company; 1999. p. 237-47.
2. Von Noorden G K. Esodeviations. In: Von Noorden G K, editors. Burian Von Noorden's Binocular vision and ocular motility: Theory and Management of strabismus. 3rd ed. St Louis: CV Mosby; 1985. p. 320-3.

---

**Figure 1:** External photograph of the child showing severe bilateral ptosis. The picture on the right shows the postoperative appearance after both the surgeries.

**Figure 2:** Extraocular movements preoperatively illustrating the superior oblique overaction.

**Figure 3:** Extraocular movements postoperatively illustrate the improved ocular alignment.
3. Helveston EM. Discussion on: Incomitant dissociated vertical deviation and superior oblique overaction. Ophthalmology 1991;98:917-8.

4. Mehta A. Chief complaint, history and physical examination. In: Rosenbaum AL, Santiago AP, editors. Clinical strabismus Management. Principles and surgical techniques. Philadelphia: W B Saunders Company; 1999. p. 17.

5. Helveston EM. Surgical management of strabismus. In: Helveston EM, editor. Surgical management of strabismus. An Atlas of Strabismus surgery. 4th ed. St Louis: Mosby Inc; p. 222-3, 404-14.

6. Shin GS, Elliot RL, Rosenbaum AL. Posterior superior oblique tenectomy at the scleral insertion for collapse of A pattern Strabismus. J AAPOS 1996;33:211-8.

7. McCall LC, Rosenbaum AL. Incomitant dissociated vertical deviation and superior oblique overaction. Ophthalmology 1991;98:911-7.

8. Helvston EM. A-exotropia, alternating sursumduction and superior oblique overaction. Am J Ophthalmol 1969;67:377-80.

9. Anderson RL, Baumgartner SA. Strabismus in ptosis. Arch Ophthalmol 1980;98:1062-7.

10. Harrad RA, Graham CM, Collin JR. Amblyopia and strabismus in congenital ptosis. Eye 1998;2:625-7.