Monitoring and Management of Progressive Exotropia and Significant Anomalous Head Turn in Duane Retraction Syndrome Type 3

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Abstract:
This case addresses appropriate monitoring and management of a teenage boy with progressive large angle exotropia in primary gaze and compensatory head turn secondary to Duane Retraction Syndrome Type 3.

Case History:
17 yo Hispanic male followed yearly since 10 months of age for Duane Retraction Syndrome Type 3 of the right eye presented for potential strabismus repair of a progressive exotropia in primary gaze with compensatory head turn. His ocular history is significant for a small left head turn of about 5 degrees that was noted as early as 10 months of age. He compensated well and was orthophoric in primary gaze until 10 years of age. At age 10, he developed a small right XT (5-10Δ) in primary position, as well as a larger left head turn (approximately 10 degrees). At this time, 6Δ BI was prescribed in spectacles, resulting in a minimally phoric alignment, with improved head alignment. Patient’s ocular history is also significant for low myopia OU.

Pertinent findings:
At age 17yo, examination showed corrected VA of 20/20 OD and OS with habitual correction: -1.75-0.75x148, 6ΔBI OD; and -2.75sph OS. Extraocular muscle testing and ductions revealed signs of Duane’s Type 3: limited abduction and adduction OD, globe retraction and narrowing of the palpebral fissure OD on adduction, widening of the palpebral fissure in abduction, and downshoot of OD on attempted adduction. Cover test at distance with prism spectacles revealed ‘A-pattern’ exotropia. At distance, the XT was larger in left gaze (35 RXT) than in primary gaze (25 RXT). Significant left head turn was present, with orthophoria in right gaze. A small right hypotropia in up and down gaze was also present at distance.

Diagnosis and Discussion:
Duane Retraction Syndrome is a congenital cranial dysinnervation disorder (CCDD) caused by an absent or dysplastic abducens nerve, with aberrant innervation of the extraocular muscles by the oculomotor nerve. It is characterized by limitation of horizontal ductions, as well as globe retraction and narrowing of the palpebral fissure with adduction. There are three types of Duane Retraction Syndrome, classified by the direction of limitation in horizontal duction: inability of the eye to move out (Type 1), in towards the nose (Type 2), or both (Type 3). Excess co-contraction with slippage of the lateral rectus muscle on adduction can lead to an up shoot or downshoot of the eye on attempted adduction. This condition is more common in females, and occurs more often in the left eye than the right. The vast majority of cases are unilateral. Amblyopia is more
common in bilateral Duane’s cases, while unilateral amblyopia has a frequency similar to the general population. Traditional therapy for amblyopia (patching, spectacles) is indicated in these patients\(^3\). Isolated Type 3 is the 2\(^{nd}\) most common of the three types. It can present with strabismus in primary gaze with a compensatory head turn, for which prism can be prescribed in spectacles, unless head turn is >15 degrees, where surgery is indicated. Some studies suggest strabismus surgery for exotropic Duane’s through ipsilateral lateral rectus recession to improve head alignment\(^2\), while one other study suggests contralateral lateral rectus recession\(^4\). Strabismus surgery will not however remediate the limitation in horizontal ductions.

**Treatment and Management:**
At this visit, the patient had a significant left head turn (>15 degrees) with large angle right exotropia in primary gaze. A successful prism adaptation test in-office with 12\(^{\Delta}\)BI OD over his current 6 BI prism (Total 18 BI), lead to the recommendation for strabismus surgery (right lateral rectus recession). Follow-up 6 days after uneventful strabismus surgery revealed extinction of the head turn. Cover test without correction was orthophoria at distance and near in primary gaze. Downshoot on adduction was eliminated.

**Conclusion:**
Thorough investigation of the ocular motilities through ductions testing and observation for worsening of an anomalous head turn, with cover test in all cardinal positions of gaze, is necessary for annual follow-up of Duane’s Type 3 patients. Treatment includes observation, prisms or strabismus repair. Although less common, anomalous innervation into the lateral rectus muscle by the oculomotor nerve can lead to exotropia in primary position. In this case recession of the lateral rectus muscle can improve primary position head position and anomalous downshoot on adduction despite continued lateral rectus dysfunction.

Keywords: Duane Retraction Syndrome Type 3, Exotropia, anomalous head turn

**Bibliography:**