Case report

Bilateral lateral rectus recession in exotropic Duane syndrome with downshoot

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Abstract

Objective: To report that maximum weakening of lateral rectus muscles can improve significant exotropia in primary position, abnormal head posture, retraction, narrowing of palpebral fissure and downshoot in exotropic duannes retraction syndrome. **Case:** A 12-year-old boy with exotropic Duane syndrome presented with downshoot and globe retraction in attempted adduction. Squint surgery was undertaken to correct the alignment and treat the secondary aberrant movements doing maximum weakening of the lateral rectus muscles by hang- back method. At 6 months follow up visit, there was improvement in abnormal head posture, reduction of ocular deviation with downshoot and stereoacuity. **Conclusion:** Supramaximal recession of lateral rectus muscles can correct exotropia with down shoot in a patient with Duane retraction syndrome.

Keywords: Exotropic Duane Syndrome with Downshoot, Supramaximal recession

Introduction

Duane retraction syndrome (DRS) is a congenital cranial dysinnervation disorder (CCDD), first described by Jakob Stilling (1887) and subsequently by Alexender Duane in 1905 (Duane A, 1905). Exotropic Duane syndrome is a relatively rare form of Duane retraction syndrome often associated with upshoot and downshoot, as well as globe retraction on adduction. Upshoots and downshoots are likely to occur due to slippage of tight lateral rectus over the crest of the globe on attempted adduction (Kraft SP, 1988). Various surgical approaches have been described- including recession of lateral and medial rectus muscles (Von Noorden GK, 1992), Posterior fixation suture of horizontal recti (Von Noorden GK, 1986), lowering of insertion of lateral rectus muscle, vertical rectus recession (Mohan K, 2002) and Y- splitting of lateral rectus muscle at the insertion (Rao VB, 2003) for the treatment of upshoot and downsoot in DRS.

We report a case of exotropic Duane syndrome with downshoot successfully managed by bilateral supramaximal recession of lateral rectus muscles.

Clinical case

A 12-year-old boy presented to the strabismus clinic of Fateh- Bal Eye Hospital, Nepalgunj, Nepal, with chief symptom of outward deviation of right eye since childhood. Past medical and family history were unremarkable with no history of trauma or neurological disease.

On ocular examination, best corrected visual acuity was 6/6 in both eyes. He preferred his left eye to fixate and adopted face turn to left side (Figure 1a). The patient had a left head turn of 25°. Ocular motility test was notable for limitation of adduction (-2) in right eye, with narrowing of palpebral fissure and downshoot of the eye in attempted adduction (Figure

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2a). Prism alternate cover test in straight-ahead revealed an exotropia (XT) of 60Δ in primary position, 50Δ XT in right and 55Δ XT in left gaze. The upgaze and downgaze showed 50Δ XT and at near 55 Δ XT. Sensory evaluation showed right eye suppression for both distance and near with Worth 4 dot test and gross stereopsis of 4800 arcs second noted with Titmus fly test. The boy was diagnosed as Exotropic Duane retraction syndrome with downshoot in right eye.

Squint surgery was performed under general anesthesia. Tight lateral rectus muscle was noted in the right eye. The patient underwent both eyes maximum recession of lateral rectus muscle. In the right eye 19 mm and in left eye 10 mm recession of lateral rectus by hang-back method.

Six months after the surgery orthoptic evaluation showed exotropia of 14Δ at distance and 10Δ at near with improved downshoot. There was no head turn (Figure 1b) and narrowing of palpebral fissure was also no longer in evidence (Figure 2b). Improvement in stereopsis was noted with 400 arcs second. The patient is on regular follow up till date.

Discussion

The main goals in the treatment of Duane retraction syndrome is to eliminate abnormal head posture, to correct strabismus without causing overcorrection or additional limitation of movement, and to decrease globe retraction, upshoot and downshoot (Pressman SH, 1986 and Kraft SP, 1988). Various surgical procedures are recommended for the correction of strabismus and co-contraction in Duane syndrome. As described by Venkeshwar B. Rao et. al. recession of lateral rectus muscle with Y-split is effective in treatment of upsdoot and downshoot with globe retraction in Duane syndrome (Venkateshwar B Rao, 2003). The splitting of lateral rectus muscle leads to less slippage of muscle from the 'crest' of the globe thereby decreasing up-shoot or down-shoot and recession leads to reduction of torque (Haslwanter T et al, 2004) which improves rotation of the globe. But Y- splitting requires greater tissue dissection and large recessions, there is a potential risk of inadvertent tearing of tight lateral rectus muscle (Jaspreet Sukhija et al, 2014). Weakening of either superior or inferior oblique muscle to treat upshoot or downshoot has been disappointing and indicates that these abnormal movements are not related to oblique dysfunction (Parks MM, 1982) but are caused by co-contraction of medial and lateral rectus muscle and taut lateral rectus muscle (Jampolsky A, 1978 and Magoon E, 1982). The outcome of maximally weakening the lateral rectus muscle by disinserting it from the globe and fixating it on the adjacent periosteal wall in the treatment of Duane retraction syndrome with up and downshoots was excellent but exposing the adjacent periosteum and passing the needle though it was technically difficult (Velenz FG, 2004). In the present case, the maximum weakening of the lateral rectus muscles and suturing it back on to the globe eliminated abnormal head turn, decreased exotropia and globe retraction with downshoot in the affected eye. It also showed improvement in stereopsis after surgery. This procedure removes the lateral rectus muscle from the crest of the globe, thereby diminishing slippage and improving downshoot.

Conclusion

Supramaximal recession of lateral rectus muscles is an effective procedure in the management of exotropia with downshoot associated with globe retraction in patient with Duane retraction syndrome.

Figure 1a: Preoperative photograph showing Head turn to the left side.

Figure 1b: Postoperative photograph showing resolution of abnormal head posture.

Figure 2a: Preoperative photograph showing exotropia in primary position with downshoot in adduction of the right eye.

Figure 2b: Post-operative photograph showing decreased exotropia in primary position and improvement in the downshoot in adduction.

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