

Case Report

Successful management of Duane Retraction Syndrome type III with significant upshoot and concurrent superior rectus contracture

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Abstract

Background: Duane Retraction Syndrome (DRS) Type III is an uncommon condition, which remains surgically challenging.

Objective: To report a rare case of DRS Type III with superior rectus (SR) contracture and its successful surgical management.

Method: Case report

Results: A 31- year old gentleman presented to our clinic with abnormal head posture and double vision. On examination, he had right face turn. Left exotropia and hypertropia were noted. Adduction of the left eye revealed severe upshoot of the left eye and narrowing of the palpebral aperture. Limitation of left eye adduction, abduction and depression was noted. The patient was diagnosed with DRS type III with SR contracture which is very rare. He later underwent left eye Y-split lateral rectus recession with superior rectus recession for his condition. Post-operatively, there was resolution of head posture and diplopia.

Conclusion: The authors has reported a rare occurrence of DRS Type III with SR contracture and their surgical technique adopted for the patient. This surgical procedure improved patient's ocular deviation, cosmetic appearance and functional ability.

Keywords: DRS Type III, Duane's retraction syndrome, Lateral rectus Y split, Strabismus, Superior rectus contracture

Background

Duane Retraction Syndrome (DRS) is an innervational ocular motility disorder characterized by deficient horizontal movements of the affected eye with associated enophthalmos with globe retraction on attempted adduction. The overall prevalence of DRS among patients with strabismus has been reported to be around 1-4%.¹ DRS Type III is an uncommon strabismus representing 15% of all DRS.² The occurrence of superior rectus contracture with DRS III is very rare.³ We report successful surgical management of a case of DRS Type III with significant upshoot and superior rectus contracture.

Case report

A 31-year old gentleman complained of double vision which progressively worsened over the past 10 years. He subsequently developed abnormal head posture, which lead to severe neck pain and the inability to drive. The patient also claimed he had a left eye squint since birth where he was unable to move his left eye outwards. There were no other ocular complaints and he denied any history of trauma. The patient was otherwise well and had no medical illness.

Visual acuity was 6/6 in both eyes on presentation. A right face turn of 45 degrees was noted. Upon correction of abnormal head posture, cover test revealed a 40-prism diopter (PD) left exotropia for near and 25PD for distance. The patient also had left hypertropia of 25PD which improved on left gaze and left head tilt. Versions revealed limitation of adduction and abduction of the left eye with severe upshoot and narrowing of palpebral fissure on attempted adduction (Figure 1). The patient had a stereoacuity of 55" with abnormal head posture. The rest of the anterior and posterior segment assessment was found to be unremarkable. He was diagnosed with left eye DRS Type III with significant upshoot and possible superior rectus

contracture. The patient was scheduled for left eye squint surgery under general anesthesia.



Figure 1. Photograph of patient showing a right face turn in primary position with the 9 preoperative cardinal positions

Intra-operatively, forced duction test of the left eye revealed a tight superior rectus and tight lateral rectus. The patient underwent lateral rectus recession 9mm from insertion with Y-split using the hang-back technique with 6/0 vicryl sutures. The bifurcation of the 2 arms of the Y split was 20mm apart. In addition, he also had left superior rectus recession of 6mm from insertion (direct scleral fixation).

During immediate postoperative period, the right face turn and upshoot improved significantly. Six weeks post-operatively, there was resolution of abnormal head posture and reduction of his left exotropia to 16PD for near and 12 PD for distance and hypertropia reduced to 14PD for near and 9 PD for distance (Figure 2). The patient also reported that his neck pain had resolved and he was able to drive again.



Figure 2. Post-operative photograph showing an improvement of the abnormal head posture and also the 9 cardinal positions

Discussion

Various methods have been suggested for the treatment of upshoot in DRS such as horizontal recti posterior fixation sutures, ipsilateral recession of both horizontal recti and vertical recti recession.⁴⁻⁶ We chose to Y-split the lateral rectus as this would preserve lateral rectus function while the bifurcated arms would prevent slippage of the eye on adduction.⁷ Hang-back technique was chosen due to the large amount of lateral rectus recession.

Clinically our patient did not show classical superior rectus contracture as he did not have an increase in hypertropia of >5PD on version and head tilt to the left side,⁸ however, we confirmed superior rectus contracture from forced duction test intraoperatively. The clinical signs of superior rectus contracture may have been masked by abnormal innervation of DRS. We proceeded to recess the superior rectus to relieve this restriction to correct the patients' hypertropia. As the patient had stereopsis with abnormal head posture pre-operatively, we aimed to correct his exotropia and hypertropia to within 10PD to prevent overcorrection and allow for fusion as in our experience, these patients usually have good fusional reserve.

Superior rectus contracture in association with DRS III has only been reported once in literature.⁹ To the best of our knowledge, this will be the second reported case of successful management DRS III with superior rectus contracture. Superior rectus contracture in our patient may have existed concurrently with DRS III or could be secondary to longstanding ocular upshoot. Further anatomical studies will be necessary to confirm this theory. We hope that the findings from our case will help contribute to literature to allow for better understanding and management of this very rare condition.

Conclusion

DRS III with significant upshoot and superior rectus contracture is a very rare condition which can be successfully treated with Y-splitting and recession of the lateral rectus combined with simultaneous recession of the superior rectus. This procedure improves ocular deviation, cosmetic appearance and most importantly the functional ability of the patient.

References

1. Ahluwalia BK, Gupta NC, Goel SR, Khurana AK. Study of Duane's retraction syndrome. *Acta Ophthalmologica*. 1988;66(6):728-30.
2. DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane's retraction syndrome. NEW YORK: Elsevier Inc; 1993. p. 257-88.
3. Rosenbaum AL, Santiago AP. Clinical strabismus management: principles and surgical techniques. Philadelphia: Saunders; 1999.
4. Rao VB, Helveston EM, Sahare P. Treatment of upshoot and downshoot in Duane syndrome by recession and y-splitting of the lateral rectus muscle. *Journal of AAPOS*. 2003;7(6):389-95.

5. Rogers GL, Bremer DL. Surgical treatment of the upshoot and downshoot in Duane's retraction syndrome. *Ophthalmology*. 1984;91(11):1380-3.
6. Von Noorden GK. Recession of both horizontal recti muscles in Duane's retraction syndrome with elevation and depression of the adducted eye. *American Journal of Ophthalmology*. 1992;114(3):311-3.
7. Kekunnaya R, Kraft S, Rao VB, Velez FG, Sachdeva V, Hunter DG. Surgical management of strabismus in Duane retraction syndrome. *Journal of AAPOS*: the official publication of the American Association for Pediatric Ophthalmology and Strabismus / American Association for Pediatric Ophthalmology and Strabismus. 2015;19(1):63-9.
8. Molinari A, Ugrin MC. Frequency of the superior rectus muscle overaction/contracture syndrome in unilateral fourth nerve palsy. *Journal of AAPOS*. 2009;13(6):571-4.
9. Arora P, Ganesh S, Shanker V. Duane's retraction syndrome with severe upshoot and ipsilateral superior rectus contracture: A rare presentation. *Journal of Clinical Ophthalmology and Research*. 2014;2(2):108