

Duane retraction syndrome, Egyptian study

Abstract

Objective: The evaluation of patients of Duane Retraction Syndrome regarding the clinical and anatomical aspects, putting them in consideration for the surgical plan of management.

Subjects and methods: Prospective study of 198 patients having Duane retraction syndrome. Surgery was aimed to eliminate the up shoot or down shoot, face turn with a horizontal deviation, and the globe retraction.

Results: Type I DRS was more common, slight male preponderance, Ninety three cases are operated, by recessing MR and/or LR muscles, Up shoot, globe retraction, ocular deviation and head posture are greatly ameliorated.

Conclusion: The surgical management in a case of Duane syndrome should be tailored according to the severity of the problem and specific clinical appearance of the patient and modified according to the anatomical findings during surgery.

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Introduction

Duane retraction syndrome a congenital ocular motility disorder most commonly characterized by the inability of the eye to abduct, variable limitation of adduction, and globe retraction with narrowing of the palpebral fissure on adduction.¹⁻³ Huber's classification system was based upon clinical findings: Type I is characterized by marked limitation of abduction with normal or minimally defective adduction, type II is characterized by marked limitation of adduction with normal or minimally defective abduction, and type III is characterized by marked limitation of both abduction and adduction.⁴

Subjects & methods

Prospective study, started at 1998 until 2010, in Alexandria, Egypt, for patients with a diagnosis of Duane syndrome who had no prior ocular surgery. Data collected data included type of Duane syndrome, gender, family history for strabismus, abnormal head position, versions, strabismus measurements, presence of amblyopia, presence of up shoot or down shoot and associated ocular and/or non ocular abnormalities. The cause of the up shoots and down shoots was thought being mechanical, innervational, or both on the basis of clinical characteristics described by Kraft,⁵ and by EMG in cooperative patients (Figure 1A-1K, Figure 2A-2D).



Figure 1 DRS type I.

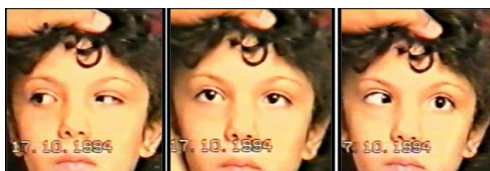


Figure 1A Left DRS-I with upshoot.

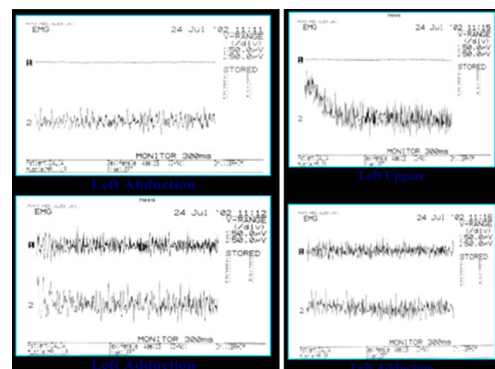


Figure 1B Left DRS-I.

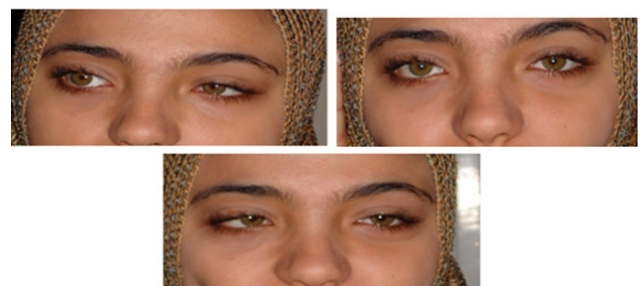


Figure 1C EMG of Left DRS-I.



Figure 1D Left DRS-I.



Figure 1E Left DRS-I.

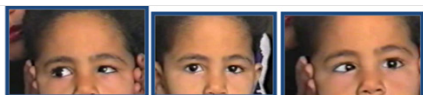


Figure 1F Left DRS-I with upshoot.



Figure 1G Left DRS-I preoperative.



Figure 1H Left DRS-I with upshoot preoperative.



Figure 1I DRS-I with upshoot preoperative.



Figure 1J DRS-I with upshoot preoperative.



Figure 1K DRS-I with upshoot and downshoot.



Figure 2 DRS type III.



Figure 2A DRS-III with upshoot preoperative.



Figure 2B DRS-III with exotropia and upshoot preoperative.

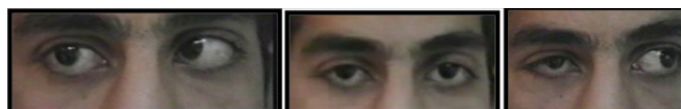
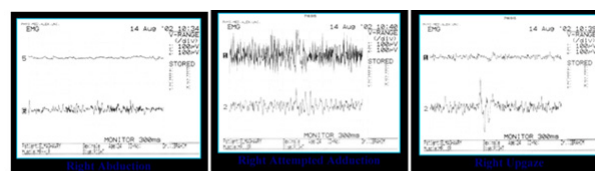
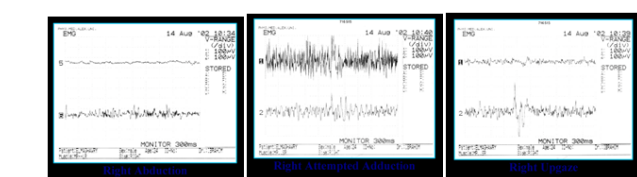


Figure 2C Right DRS-III.





EMG

**Figure 2D** Left DRS-III with upshoot preoperative.

Strabismus surgery was performed on patients to eliminate the upshoot or downshoot, face turn with a horizontal deviation, and the globe retraction. One surgeon (DS) performed all of the surgeries. Different surgical techniques depending on preoperative condition, EMG, anatomical findings and forced duction test. Surgeries performed included: Recession of MR muscle (one or two), Recession of LR muscle, Recession of MR & LR of the same eye, Y splitting of lateral rectus muscle.⁶

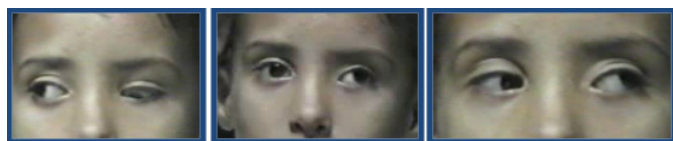
Results

The study included 198 patients, there was a slight male preponderance in contrast to a female preponderance reported elsewhere⁷⁻¹⁰ there were 94 (48%) female patients and 104 (52%) male patients. In our group of patients there was a male preponderance among type I and type II Duane while there was a female preponderance among type III. Like other reports left eye was more commonly affected^{3,8,11,12} and 114 patients (58%) had involvement of left eye; 56 patients (28%) had involvement of the right eye; and 28 patients (14%) had involvement of both eyes. The ages of the patients ranged from 6 months to 44 years (mean 10.4 years). There was no significant difference between the different types of Duane with regards the age of presentation.

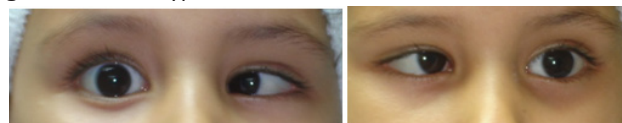
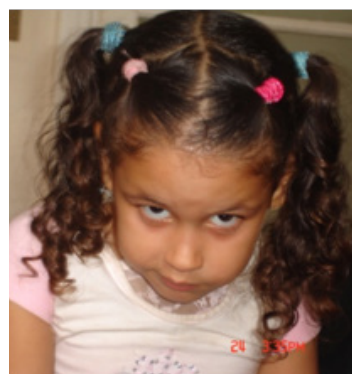
All of the patients were categorized into types I, II, and III Duane syndrome, based on Huber's classification: Type I: 145 patients (73%) (Figure 1A–1K), Type II: Ten patients (5%) (Figure 3) Type III: 43 patients (22%). Similar to other studies type I DRS occurred more frequently. Patients with bilateral Duane syndrome (Figures 4A & 4B) constituted (14%) of our series, most of which were type I (68%) and none were type II. Patients having bilateral Duane syndrome had, a lower incidence of abnormal head posture, and one patient with bilateral DRS I with V pattern esotropia had an abnormal head posture in the form of chin down position (Figure 5) and associated congenital anomalies.



Postoperative.

**Figure 3** DRS type II.
DRS-II with left downshoot preoperative.

Postoperative.

**Figure 4A** Bilateral type III DRS.**Figure 4B** Bilateral type I DRS.**Figure 5** Bilateral DRS I with esotropia V pattern and abnormal head posture.

Preoperative.



Postoperative.

Amblyopia was present in 15% of patients in comparison to the study by Chua et al 16.9% had amblyopia.¹³ There was no difference in amblyopia among various types of Duane syndrome. The incidence of both anisometropia and amblyopia was somewhat higher than for the normal population.¹¹ Ninety three patients underwent strabismus surgery: type I: 63 patients, type II: 5 patients, type III 25 patients. Among the patients with type I Duane, 32 patients who had a horizontal deviation with a mild degree of globe retraction and esotropia in primary position, were operated by MR recession, a single medial rectus muscle recession was performed in 15 patients with esotropia of small angle and biomedical surgery was performed in 17 patients for larger angle of esotropia where recession of one muscle was not sufficient. Thirty one patients of DRS I had MR and LR recession in the same eye, for severe retraction and upshoot. Recession of the LR was performed in five patients with DRS II presenting with exotropia, and globe retraction, in two of them Y-splitting is added to stabilize the globe as well.

Recession of the two horizontal recti was performed in 59 patients with severe retraction or with upshoot; 31 patients type I, three patients type II and 25 patients type III. In these patients simultaneous medial and lateral rectus muscle recession was performed to reduce the upshoot, the recession was asymmetrical.^{14,15} The LR muscle was recessed 50% more than the MR. In the beginning of the study, we had a complication; 2 patients of DRS III who were orthotropic prior to surgery and became exotropic after simultaneous MR and LR recession.

EMG was performed on 12 patients and all these patients have revealed that simultaneous activation of the MR and LR muscles is associated with co-contraction and globe retraction, similar to previous reports.^{16,17} In our series there were 3 patients who had family members with Duane but no genetic studying was performed.

Discussion

In our series, more than 50 % of cases of DRS did not need any interference, and this was due to the absence of deviation of the eyes in primary gaze position, no abnormal head posture, no upshoot, mild or negligible globe retraction on adduction, good binocular functions. These cases are left alone and only followed up for any changes that might appear. Esotropia in DRS was managed by recession of one MR or two MR muscles for larger angles where LR resection is absolutely contraindicated preventing more globe retraction. Severe globe retraction and upshoot of the globe was managed by simultaneous recession of the two horizontal recti of the same eye, this was based upon the EMG study that proved simultaneous co-contraction of these muscles and not with the vertical recti muscles. The amount of relative recession was controversial, but after getting two cases of post operative exotropia in the beginning of the study, and after putting into consideration the anatomical changes, the lateral rectus is recessed 50 % more than the medial rectus, putting in consideration the pre-operative deviation as well in order to preserve the same preoperative position for orthotropic patients but eliminating or greatly reducing the globe retraction and the upshoot, modifying these numbers in front of a pre existing esotropia or exotropia and face turn.

Conclusion

The surgical management in a case of Duane syndrome should be tailored according to the severity of the problem and specific clinical

appearance and anatomical findings in each patient. The treatment options and realistic expectations should be discussed with the patient and the family in detail as cases of DRS syndrome could be greatly ameliorated but not reaching complete cure.

Acknowledgments

None.

Conflicts of interest

The author declares there is no conflict of interest.

References

1. Duane A. Congenital deficiency of abduction, associated with impairment of adduction, retraction movements, contraction of the palpebral fissure and oblique movements of the eye. 1905. *Arch Ophthalmol*. 1996;114(10):1255–1256.
2. Miller NR, Kiel SM, Green WR, et al. Unilateral Duane's retraction syndrome (Type 1). *Arch Ophthalmol*. 1982;100(9):1468–1472.
3. Kowal VO, McKeown CA. Duane's syndrome. *Int Ophthalmol Clin*. 1992;32(1):51–62.
4. Huber A. Electrophysiology of the retraction syndrome. *Br J Ophthalmol*. 1974;58(3):293–300.
5. Kraft SP. A Surgical approach for Duane syndrome. *J Pediatr Ophthalmol Strabismus*. 1998;25(3):119–130.
6. Rao VB, Helveston EM, Sahare P. Treatment of upshoot and downshoot in Duane syndrome by recession and y-splitting of the lateral rectus muscle. *J AAPOS*. 2003;7(6):389–395.
7. Shauly Y, Weissman A, Meyer E. Ocular and systemic characteristics of Duane Syndrome. *J Pediatr Ophthalmol Strabismus*. 1993;30(3):64–68.
8. Mehel E, Quere MA, Lavenant F, et al. Epidemiological and clinical aspects of Stilling-Turk-Duane syndrome. *J Fr Ophtalmol*. 1996;19(8–9):533–542.
9. Rutstein RP. Duane's retraction syndrome. *J Am Optom Assoc*. 1992;63(6):419–429.
10. Alexandrakis G, Saunders RA. Duane retraction syndrome. *Ophthalmol Clin North Am*. 2001;14(3):407–417.
11. Sloper JS, Collins AD. Effects of Duane's retraction syndrome on sensory visual development. *Strabismus*. 1999;7(1):25–36.
12. DeRespini PA, Caputo AR, Wagner RS, et al. Duane's retraction syndrome. *Surv Ophthalmol*. 1993;38(3):257–288.
13. Chua B, Johnson K, Donaldson C, et al. Management of Duane retraction syndrome. *J Pediatr Ophthalmol Strabismus*. 2005;42(1):13–17.
14. Von Noorden GK. Recession of both horizontal recti muscles in Duane's retraction syndrome with elevation and depression of the adducted eye. *Am J Ophthalmol*. 1992;114(3):311–333.
15. Sprunger DT. Recession of both horizontal rectus muscles in Duane syndrome with globe retraction in primary position. *J AAPOS*. 1997;1(1):31–33.
16. Hunderson T, Zeavin B. Observations on the retraction syndrome of Duane. *Arch Ophthalmol*. 1956;55:576.
17. Huber A. *Duane's Retraction Syndrome: Consideration of Pathophysiology and Etiology*. In: Grune et al, editors. Florida, USA, 1984. p. 345–361.