ORIGINAL ARTICLE

# DUANE'S SYNDROME: SURGICAL OUTCOME AND NON OPHTHALMOLOGIC ASSOCIATIONS

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**Background:** Duane retraction syndrome (DRS) is the most common of the ocular congenital cranial dysinnervation disorders .This study evaluates the types of Duane syndrome and its management in patients presenting to the paediatric and strabismus unit of a tertiary care eye hospital. **Methods:** This case series study involved 41 patients diagnosed with Duane syndrome between January 2007 and December 2009. History of presenting complaints, past treatment and family history were recorded. Ocular examination and orthoptic assessment was carried out **Results:** Forty one patients were included in this case series study. It involved 10 right eyes, 27 left eyes and both eyes of 4 patients. There were 26 females and 15 males. Type-1 Duane syndrome was present in 28 (68.3%), type 2 in 8 (19.5%), Type-3 in 4 (9.8%) and type-4 with synergistic divergence was present in 1 (2.4%) patient. Comorbidity was present in 6(14.6%) patients. Surgery was carried out in 26 (63.4%) patients either for abnormal head posturing or significant upshoots or down shoots. Upshoots noted in 21 eyes, were completely or partially resolved in 15 cases. Among 4 patients with down shoots on adduction, complete resolution was seen in 1. The pre and post-operative measurements of horizontal deviation showed statistically significant difference in Duane type-1 and 2, where as in Duane type-3 it was not significant. One patient with type-4 Duane did not undergo surgery. **Conclusions:** Recession of the horizontal recti is more effective in treating the upshoot or down shoot associated with DRS as compared to recession and y-split of the horizontal muscle.

**Keywords:** Duane retraction syndrome, upshoots and downshoots, congenital cranial dysinnervation disorder

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## INTRODUCTION

Duane retraction syndrome (DRS) is the most common of the ocular congenital cranial dysinnervation disorders.[1] It is named after an ophthalmologist Alexander Duane (1858–1926). Originally its aetiology was thought to be myogenic but MRI findings support a neurogenic origin.[2] This has led to renaming it as co-contractive retraction syndrome.[3] Presenting features and results of surgical treatment have been reported in different studies. We present the associated non ocular features and results of surgical treatment in patients with different types of DRS, seen at our tertiary care eye hospital.

## MATERIAL AND METHODS

The case series study involved 41 patients diagnosed with Duane retraction syndrome (DRS) between January 2007 and December 2009. History of presenting complaints, past treatment and family history were recorded. Ocular examination including cycloplegic refraction in children and auto-refraction in adults, best corrected visual acuity, fundus examination and orthoptic assessment was carried out. Types of DRS were based on the prominent deficiency of duction, as classified by Huber.[4] Presence of any associated non ocular feature was recorded. Patients having previous squint surgery and other causes of restrictive strabismus were excluded from the study. The study was approved by the ethical review board of our institution. SPSS-10 for windows was used for statistical analysis.

## RESULTS

A total of 41 patients were included in the study, out of which 26 (63.4%) were females and 15 (36.6%) were males. Mean age at presentation was 10.87±7.07 (range 1–30) years. The right eye was involved in 10 (24.4%), left eye in 27 (65.9%) and both eyes were involved in 4 (9.8%) of the patients. Type 1 Duane syndrome was present in 28 (68.3%), type 2 in 8 (19.5%), type 3 in 4 (9.8%) and type 4 with synergistic divergence was present in 1 (2.4%) patient. Table-1 describes the associated vertical tropia with each type of Duane noted in our study.

Face turn was noted in 31 (75.6%), head tilt in 2 (4.9%) whereas no compensatory head posture was present in 8 (19.5%) patients. Upshoot on adduction was present in 21 (51.2%) and downshoot on adduction was observed in 4 (9.8%) and no vertical abnormality in extra-ocular movements was noted in 16 (39%) cases. Surgery was carried out in 26 (63.4%) patients either for abnormal head posturing or significant upshoots or downshoots. Horizontal muscle recessions were performed in the range of 5–16 mm mean (6.75±5.35 mm). Among these, Y-split of the lateral rectus muscle along with recession was done in 5 cases.

Exotropia (XT) in the range of 6–50 (mean 33.45±12.85) prism diopters (PD) was found in

22(53.7%) of patients. Fourteen patients with upshoot and 3 patients with down shoot had exotropia. Post-operative mean exotropia was 12.21±8.38 PD ($p$=0.000). Esotropia (ET) was present in 15 (36.6%) patients in the range of 2–50 (mean 21.07±17.31) PD. Upshoot was present in 6 and downshoot in 1patient having ET. Mean post-operative esotropia was 6.25±4.78 PD ($p$=0.011). Four (9.8%) patients were orthophoric. One of these orthophoric patients had mild upshoot on adduction. Upshoots were completely (n=9) or partially (n=6) resolved in 15 out of 21 cases. In 3 cases it was not resolved and 3 patients did not have surgery for mild upshoot. Among 4 patients with down shoots on adduction, complete resolution was seen in1 patient. It was not resolved in 2 patients, while one patient did not undergo any surgery.

Out of 5 patients who were treated with y-split and recession of horizontal recti, 60% of patients (n=3) had partial or complete resolution of the upshoot while in 40% (n=2) the upshoot was not resolved. With horizontal muscle recession without y-split, the upshoot and down shoot was completely or partially resolved in 13 out of 16 patients (81.25%) and it was not resolved in 3 (18.75%) patients.

Comorbidity was present in 6(14.6%) of patients. Torticollis and congenital heart disease, cleft palate, impaired hearing and cataract, were present in 1 patient each. Facial asymmetry was present in 2 patients. Family history was positive in 1 patient. There was no significant correlation between type of duane and presence of compensatory head posture ($p$=0.167), presence of upshoot or down shoot ($p$=0.191), type of Duane and sex ($p$=0.697) or presence of non-ophthalmological features and unilateral or bilateral disease ($p$=0.656) using the Chi square test.

Out of the 10 patients with DRS affecting the right eye, 2 had systemic associations. Similarly, 3 among the 27 patients with DRS in left eye and 1 out of 4 bilateral DRS cases had associated non ocular features. The pre and post-operative measurements of horizontal deviation showed significant difference in Duane type-1 and 2 ($p$=0.000 in each), where as in Duane type-3 it was not significant ($p$= 0.184) One patient with type-4 Duane did not undergo surgery.

**Table-1: Primary position horizontal deviation associated with each type of Duane syndrome**

| DRS type | Horizontal deviation | N | % |
|---|---|---|---|
| **1** | XT | 11 | 39.3 |
| | ET | 14 | 50 |
| | Orthophoria | 3 | 10.7 |
| | Total | 28 | 100 |
| **2** | XT | 8 | 100 |
| **3** | XT | 2 | 50 |
| | ET | 1 | 25 |
| | Orthophoria | 1 | 25 |
| | Total | 4 | 100 |
| **4** | XT | 1 | 100 |

DRS= Duane retraction syndrome, XT=exotropia, ET=esotropia

**Table-2: Results of surgical treatment in patients with different types of Duane syndrome**

| Duane type | | Mean(PD) | n | SD | $p$-value |
|---|---|---|---|---|---|
| **1** | Pre-operative | 22.4583 | 24 | 17.99270 | .000 |
| | Post-operative | 8.2500 | 24 | 6.68711 | |
| **2** | Pre-operative | 41.2500 | 8 | 6.40870 | .000 |
| | Post-operative | 12.1250 | 8 | 10.93406 | |
| **3** | Pre-operative | 13.0000 | 3 | 12.52996 | |
| | Post-operative | 4.0000 | 3 | 6.92820 | .184 |
| **4** | No surgery done | | 1 | | |

PD=prism diopters, n=number of patients

## DISCUSSION

Most cases of Duane syndrome represent simplex cases being a single occurrence in the family.[5] In the same way, only 1 case in our study had a positive family history. A large series of DRS patients, reports equal frequency of primary position esotropia and exotropia in type-1 Duane, exotropia in type 2 and more XT than ET associated with Type 3 DRS.[4] Our study has similar results with 11 (39.3%) patients with XT, 14 (50%) patients with ET and 3 (10.7%) patients with orthophoria in type-1 Duane. The findings in Type 2 and 3 DRS are also comparable to the above study (Table-1).[4] Our study is comparable to the same retrospective study, in frequency of unilateral and bilateral cases of DRS also. Mohan and colleagues have observed 12% bilateral involvement in 40 out of 331 patients.[4] Our study included 9.8% (n=4) cases with bilateral DRS. One of these four patients had associated non ocular feature of torticollis and congenital heart disease.

In a study involving 124 patients, success was achieved after recession of horizontal recti in all types of DRS.[6] A retrospective study of 17 patients described success of horizontal recession in 70.58% of cases.[7] In the same study results of surgery were unsuccessful in 4 cases of Duane type-1 and 1 patient of Duane type 3.[7] Our study showed significant difference between the pre and post-operative measurements of horizontal deviation in type-1 and 2 Duane syndrome, while in type 3 Duane syndrome the results were insignificant (Table-2).

Up to 70% of patients with Duane syndrome do not have any associated congenital anomaly.[8] Isolated Duane syndrome was seen in 85.4% of patients (n=35) in our study without any co morbidity. Some of the CCDDs are known to have non-ocular associations involving neurologic, neuro-anatomic, cerebrovascular, cardiovascular and skeletal abnormalities.[9] Congenital abnormalities associated with DRS include those involving the ear, kidney, heart upper limbs and skeleton. Most of the associated findings in our study have been reported before. Klippel-Feil anomaly has been described as part of Wildervanck syndrome in female patients with Duane syndrome and deafness.[10] The patient in our study with

Klippel-Feil anomaly alone did not have deafness and was a male patient. Deafness however, was a separate feature in another patient of duane syndrome.

Analysis of 67 cases of Duane syndrome shows that upshoot and downshoot phenomenon is more common in eyes with exotropia in a study.[11] This observation is similar to our results in which 63.6% of eyes (n=14) with exotropia had associated upshoot and 13.6% (n=3) of eyes had downshoot, compared to upshoot in 40% (n=6) and downshoot in 6.7% (n=1) in eyes with ET. Orthophoria in 59% of patients with DRS was the commonest presentation in a study.[12] In contrast, our study population had orthophoria in 9.8% (n=4) as the least common presentation.

Upshoot and down shoot of the affected globe on attempted adduction is seen more commonly in type-3 Duane as compared to type-1 and 2.[13] Another study describes upshoots and downshoots more common in unilateral disease and types-1 and 3 compared with type-2.[14] We did not find any statistically significant association between the type of Duane and presence of any upshoot or downshoot.

Our pre-operative and post-operative horizontal deviation was statistically significant in cases of both preoperative esotropia and exotropia. However, better results, with the mean horizontal tropia within 10 PD, resulted after surgery in cases with preoperative esotropia. This may be attributed to lesser mean preoperative esotropia (21.07±17.31) compared to mean preoperative exotropia (33.45PD±12.86). Fewer patients had y-splitting of the recessed muscle compared to conventional recession of the horizontal recti.

We experienced better resolution of the upshoots and down shoots with the later procedure. The limitations in our study is that due to the large difference in number of patients (5 vs 13) treated with either procedure the statistical results cannot be confirmed with the present sample size. Our study also shows that upshoots partially or completely resolved in 15 (83.33%) patients out of 18 surgically treated patients, as compared to downshoots which were resolved post operatively in 1(33.33%) out of three cases. Here again the number of patients treated in both categories are fairly different. Small number of patients in the less common type of DRS has affected the comparison of results in retrospective series of large sample size as well.[4]

## CONCLUSION

In our study the surgical outcome was better in patients with preoperative esotropia in DRS patients. Standard recessions were more effective than combining y-splitting and recession of the horizontal muscles to treat the associated upshoots and downshoots. The upshoots are more amenable to treatment as compared to downshoots in DRS patients.

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