Original Article

Refractive Profile in Duane Retraction Syndrome

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Abstract

Aim: To study the refractive error profile of patients with Duane retraction syndrome (DRS). **Methods:** We conducted a retrospective analysis of patients diagnosed with DRS between January 2015 and December 2018 at a tertiary eye center. All case files of patients diagnosed with DRS were retrieved and analyzed. Following parameters were collected from the records: demographic data, laterality of involvement, type of DRS, type of refractive error, presence of anisometropia, and presence of amblyopia, if any. Type of refractive error was correlated with type of DRS. **Results:** Seventy-seven eyes of 74 patients were included with age range of 2 to 65 years. There were 42 female patients and 32 male patients. Exotropic DRS with unilateral presentation was the commonest pattern. Left eye was predominantly involved in unilateral DRS. Hyperopia (58.1%) was the most common refractive error in both esotropic and exotropic DRS. Anisometropia was present in 16% cases, with anisoastigmatism being the most common refractive error in them. Amblyopia was reported in only three cases with anisometropia being the cause. Abnormal head posture was reported in most of the patients (81%). Upshoots and downshoots were common in exotropic DRS. **Conclusion:** Hyperopia was the most common refractive error in both esotropic and exotropic DRS. Amblyopia was reported in a few cases and was due to anisometropia. This underscores the importance of proper evaluation of refractive error in patients with DRS.

Keywords: Duane syndrome, exotropic DRS, hyperopia

INTRODUCTION

Duane retraction syndrome (DRS) is a congenital anomaly of the sixth cranial nerve nuclei along with aberrant innervation from the third cranial nerve. It is a rare condition with a prevalence of approximately 0.1% of the general population[1,2] and less than 5% of all strabismic cases.[1-4]

Binocular vision is preserved in DRS despite the restriction of eye movements owing to occurrence of compensatory head posture in most of the cases. Hyperopia and anisometropia are refractive errors commonly reported in esotropic DRS (eso DRS). The presence of amblyopia is variable ranging from 3% to 40%.[1,5,6] Anisometropia, not strabismus, is commonly the cause of amblyopia in such cases.

Though the occurrence of hyperopia in eso DRS and importance of its correction is documented, the exact type of refractive error in various types of DRS has not been widely studied or reported. Hence, the aim of this study was to analyze the refractive error profile of patients with DRS and also to correlate it with type of DRS.

MATERIALS AND METHODS

Retrospective review of case records of patients diagnosed with DRS from January 2015 to September 2018 presenting to outpatient department of our pediatric ophthalmology and strabismus unit was done. The records that were complete in terms of documentation of oculomotor evaluation and cycloplegic refraction were included, whereas those with incomplete documentation were excluded

Parameters including demographic data such as age and sex, laterality (unilateral/bilateral), type of DRS (exotropic/ esotropic/orthotropic), cycloplegic refraction values, presence of anisometropia, and amblyopia were collected from the records.

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The DRS was classified into eso DRS, exotropic (exo DRS), and orthotropic (ortho DRS) on the basis of deviation in primary gaze. Deviation was measured with alternate prism cover test (APCT) or modified Krimsky test depending upon the visual acuity; APCT was used if visual acuity was better than 6/18 in both eyes.

Documentation of cycloplegic refraction readings was noted. Cycloplegic refraction was performed with Auropent (Aurolab, Madurai, India; cyclopentolate1%) or Homide (Indoco Remedies, Mumbai, India; homoatropine 2%) and Tropicacyl plus (Sunways, Mumbai, India; phenylephrine 5% + tropicamide 0.8%).

Refractive error was categorized into myopia, hyperopia, myopic astigmatism, hyperopic astigmatism, and emmetropia on the basis of cycloplegic refraction. Spherical equivalent was considered for the classification of refractive error. We classified as emmetropia when refraction was Plano +/− 0.5 diopters sphere (DS); myopic astigmatism if refraction showed cylinder of >- 0.5 with no spherical component; hyperopic astigmatism if refraction showed cylinder of more than +0.5 with no spherical component; myopia when spherical error (SE) was more than − 0.5DS which was further categorized into mild, moderate, and high if SE was less than −3.00DS, −3.00DS to less than −6.00 DS, and greater than or equal to −6.00DS, respectively. Finally, hyperopia if SE more than +0.5DS and was further classified as mild, moderate, and high depending on SE less than +2.50, +2.50 to less than 5.0, and greater than

Anisometropia was said to be present if there was a difference of 1 DS in spherical equivalent between two eyes.[7] The frequency of type of refractive error in DRS patients and its correlation with type of DRS was noted. Other associated clinical features such as up/downshoots, globe retraction, and presence of abnormal head posture recorded also was noted.

RESULTS

Seventy-seven eyes of 74 patients with DRS were included with age range of 2 to 65 years. Majority of patients were in the age group of 0 to 9 years . Females were predominantly involved in all types of DRS with male to female ratio of 1:1.3 [Figure 1, Table 1].

Considering laterality of occurrence, 71 cases were unilateral (95%), whereas 3 (5%) cases were bilateral. Left eye (OS) was more commonly affected (53/71) when compared with right eye (18/71) [Table 2].

Exotropic DRS was the most common type reported (46/74 patients) followed by eso DRS (19/74) patients and ortho DRS (9/74) [Table 3]. Further vertical strabismus was noted in primary gaze in eight cases.

Hyperopia (58.1%) was the commonest refractive error followed by myopic astigmatism (27.27%) and myopia (7%) in all types of DRS.

Hyperopia ranged from +0.50DS to +4.0DS and myopia ranged from −0.50DS to − 6.0 DS.
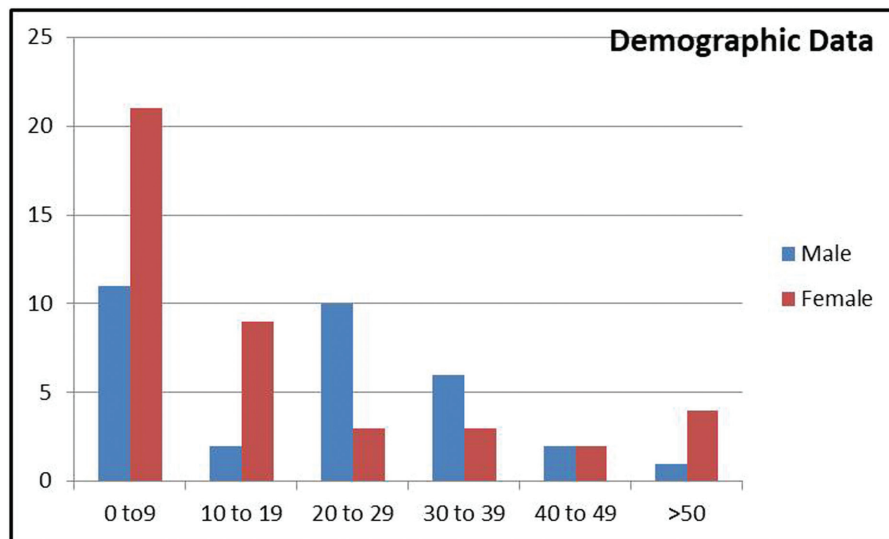

Figure 1: Age and sex distribution of the Duane retraction syndrome cases.

or equal to +5.00 DS, respectively.

Table 1: Sex distribution according to type of Duane retraction syndrome

Sex	Exo DRS	Eso DRS	Ortho DRS	Total
Male	22 (47.8%)	5 (26.3%)	5 (55.5%)	32 (43.24%)
Female	24 (52.1%)	14 (73.6%)	4 (44.4%)	42 (56.76%)

Data are presented as *n* (%). DRS, Duane retraction syndrome; Eso DRS, esotropic DRS; Exo DRS, exotropic DRS; Ortho DRS, orthotropic DRS.

Table 2: Laterality of involvement in relation to type of Duane retraction syndrome

	RE*	LE*	Total
Exo DRS	14 (30.4%)	31 (67.3%)	45
Eso DRS	1 (5.2%)	17 (89.4%)	18
Ortho DRS	3 (33.3%)	5 (55.5%)	8
Total	18 (24.3%)	53 (71.6%)	71 (95.9%)

Data are presented as *n* (%). DRS, Duane retraction syndrome; Eso DRS, esotropic DRS; Exo DRS, exotropic DRS; ortho DRS, orthotropic DRS. *Bilateral cases are not included in this table.

Table 3: Type of refractive error and correlation in various types of Duane retraction syndrome

Type of refractive error	Eso DRS	Exo DRS	Ortho DRS	Total	Percentage
Hyperopia	14	26	5	45	58.44%
Myopia	2	3	1	6	7%
Myopic astigmatism	4	16	1	21	27.27%
Plano	0	1	2	3	3.8%
NA	0	1	1	2	
Total	20	47	10	77*	

DRS, Duane retraction syndrome; Eso DRS, esotropic DRS; Exo DRS, exotropic DRS; ortho DRS, orthotropic DRS; NA, not available. *Refers to number of eyes analyzed.

In subcategories of refractive error on the basis of spherical equivalent, mild hyperopia was most common. Hyperopia was the most common refractive error both in eso DRS and exo DRS [Table 3].

Anisometropia was present in nine cases, anisoastigmatism being the most common (seven cases) followed by anisohyperopia (four cases) and anisomyopia (three cases). Amblyopia was noted in three cases, of which two were anisohyperopia and one with anisoastigmatism. No case of strabismic amblyopia was noted in our study.

Abnormal head posture was seen in 60 cases in our study, face turn (92%) being the most common followed by head tilt (7%) and chin down (1%). Signs of aberrant innervation in the form of upshoots and globe retraction were reported in 28 and 60 patients, respectively. Upshoot (23 out of 28) downshoots (5 cases) accounting to total of 37.8% and being more common in exo DRS (50%) was noted.

DISCUSSION

The present study explored the refractive error profile and other clinical characteristics of patients diagnosed with DRS.

Kekunnaya et al.[8] noted female preponderance in 60% cases especially in unilateral type I DRS. Kirkham[9] and O'Malley et al.[10] have found DRS types II and III to be more prevalent in females compared to males. Our study showed female predominance (57/74 cases) in all types of DRS (eso, exo, and ortho DRS) similar to other studies of individual types of DRS.

Several studies have noted unilateral presentation being common than bilateral occurrence and with left eye being predominantly involved.[11-13] Similar findings with respect to laterality and predominant eye involved were noted in our study population.

Kekunnaya et al.[8] reported esotropia as most common primary position deviation, followed by orthotropia. According to Isenberg and Urist,[14] in patients with unilateral type I DRS, esotropia occurred more frequently than exotropia; in type II, exotropia was common whereas esotropia, exotropia, and orthotropia occurred equally in type III cases. We classified DRS according to type of primary gaze deviation and not as types I, II, and III unlike previous studies. Our study showed exotropia as the most common primary gaze deviation in contrast to other studies attributable to difference in classification used. Exotropia was probably more common in our study because exotropia is more common in India unlike earlier studies from western countries where the commonest type of strabismus is esotropia.

Kekunnaya et al.[12] reported an occurrence of upshoot or downshoot in 43% of their cases and occurring commonly in unilateral DRS types I and III. Mohan et al.[13] found the prevalence to be significantly greater in DRS types II and III. Upshoots and downshoots amounting to a total of 37.8% were more common in exo DRS in our study comparable to its occurrence in DRS types II and III in the report by Mohan et al.[13]

Kekunnaya et al.[15] have shown that 30% to 80% of patients with eso DRS have hypermetropia or hypermetropic astigmatism greater than +1.50DS, some even more than +4.00DS. Hyperopia and associated accommodative component in eso DRS have also been reported. The importance of hyperopic correction to correct the accommodative component in eso DRS before surgery has been stressed. Hyperopia (43%) was the most common

refractive error followed by myopic astigmatism (22%) and myopia (5%) both in exo DRS and eso DRS in our study. This finding underscores the importance of evaluation and appropriate management of refractive error in both eso DRS and exo DRS.

Tredici and Von Noorden[16] found a 17% prevalence of anisometropia and 3% prevalence of amblyopia in these patients. Kirkham[9] found anisometropia >1D in about 40% of cases. O'Malley et al.[10] reported strabismic amblyopia in 11% and anisometropic amblyopia in 3% of their patients. In present study, anisometropia (1.0DS) was noted in 12% cases, anisoastigmatism being most common (9.4%). Amblyopia was reported only in three cases, out of which two had anisohyperopia and one had anisoastigmatism. The difference from other studies may be due to smaller sample size and retrospective nature of our study.

There are few limitations in our series in terms of sample size and retrospective nature of the study. In addition, the impact of refractive correction on the abnormal posture or primary gaze deviation in these patients was not studied.

CONCLUSION

Refractive errors were reported in most of the patients of DRS. Hyperopia being the commonest type of refractive error, both in eso DRS and exo DRS. Upshoots and downshoots were common in exo DRS. Exo DRS and unilateral occurrence was more common in our series. Amblyopia was reported in very few cases. Proper evaluation of refractive error in patients with DRS is important.

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Conflicts of interest

There are no conflicts of interest.

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