Sohag University Sohag Medical Journal Faculty of Medicine

Prevalence of Duane Retraction Syndrome in Sohag University Hospital

Hatem Ammar, Al-Ahmady Hamad, Adham Salah, Mohamed Ezz Eldawla

*** Department of Ophthalmology, Faculty of Medicine, Sohag University**

Abstract

Introduction: Duane retraction syndrome is a congenital complex strabismus. It was first described in 1887. It presents mainly with limitation of ocular movement, with globe retraction. It may be associated with over shooting of the eye. DRS is one of congenital cranial dysinnervation disorders (CCDD), which are congenital, nonprogressive, sporadic, or familial developmental anomalies of the cranial nerves characterized by abnormal eye, eyelid, and/or facial movements.

It has been hypothesized that there is a maldevelopment which may occur due to any insult to the development of the sixth nerve nucleus and/or nerve at 4–8 weeks of pregnancy has been demonstrated to play a causal role in the emergence of this disease process.

Aim of the work: to study the prevalence of Duane retraction sundrome (DRS) among cases of strabismus in Sohag university hospital Patients and Methods: A prospective non-randomized interventional clinical study done in Sohag university hospital. All patients presented with Duane syndrome in strabismus clinic in Sohag university hospital were included.

Results: Thirty five patients had DRS out of 960 strabismus patients, type I was the commonest. DRS is more common in females.

Keywords: prevalence, duane, sohag

DOI: 10.21608/SMJ.2023.204760.1381

Introduction

Strabismus is one of the often occuring dysmorphic ocular disease types in the outpatient ophthalmology clinic. It can occur in 0.5% to 5% of people. In addition to having detrimental impacts on the patient's self-confidence and interpersonal connections, strabismus has a number of effects of its own, includeing the dissociation of binocular vision and stereopsis. With increased information, strabismologists now aim to restore "stereopsis," which is the ultimate goal, in addition to ocular alignment. (1)

Duane retraction syndrome (DRS) is a congenital complex strabismus. Stilling initially outlined this condition in 1887. Then, American ophthalmologist Alexander Duane reported a group of 54 patients who had this problem . (1)

The prevalence of Duane retraction syndrome in the general population of strabismus patients is 1–4% .(2) It has been determined that over 60% of those affected are female . (3) Regarding laterality, it has been discovered that 80% of cases are unilateral and that at the left eye is typically affected . (4)

DRS is one of congenital cranial dysinnervation disorders (CCDD), which are congenital non-progressive sporadic or familial developmental malformations of the cranial nerves that are characterrised by aberrant movements of the eye, eyelids, and/or face.[5]

It has been hypothesized that any insult to the sixth nerve nucleus or nerve during its development between weeks 4 and 8 of pregnancy may result in maldevelopment, which would then lead to the onset of this disease process. [6]

DRS is often diagnosed clinically. According to the EMG pattern and related ocular movement impairments, the conventional Huber's classification.[7] suggests three kinds of DRS. Currently, Huber's classification is among the most popular.[8] and proposes three types of DRS according to presenting electromyographic (EMG) pattern as follows:

A pronounced limited abduction and a normal or mild adduction limitation are characteristics of Huber type I DRS. Adduction limitation and normal or mild abduction limitation are characteristics of Huber type II DRS, while Huber type III DRS is characterized by both abduction and adduction limitations. [9]

Purpose: to study the prevalence of DRS among cases of strabismus in Sohag University Hospital, study age, sex and laterality distribution.

Patients and Methods

A prospective non-randomized clinical study. All patients presented with Duane syndrome in strabismus clinic in Sohag university hospital were included. Surgeries done in the department of ophthalmology, Sohag university hospitals.

A written informed consent is taken from all patients after understanding the nature and the aim of the study. Approval of the ethical committee of Sohag faculty of medicine is fulfilled.

Inclusion criteria:

All cases with Duane syndrome coming to strabismus clinic in Sohag university hospital are included.

Exclusion criteria:

Previous strabismus surgery. Associated eye diseases; other congenital anomalies.

Patients are divided into three groups

Group A: patients with DRS type I

Group B: patients with DRS type II

Group C: patients with DRS type III

Informed consent was taken from all patients diagnosed as DRS. Consents included the agreement of photographing patients included in the study.

Examples of patients with different groups of DRS

.

Figure (1): Patient No.1: Female 21 years old Preoperative photos: Lt. DRS type I, grade 4 over shooting in adduction,grade 3 globe retraction

Figure (2): Patient No. 2: Female 15 years old Preoperative photos: Lt. DRS type II, exotropia 15 prism diopter, Rt. Face turn, grade 2 globe retraction

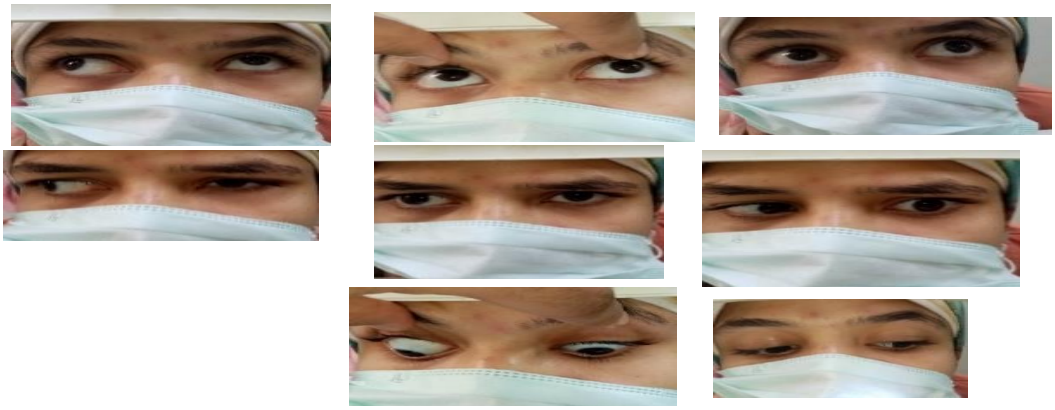

Figure (3): Patient No. 3: Female 16 years old Preoperative photos: Lt. DRS type III, grade 1 over shooting in adduction, grade 2 globe retraction

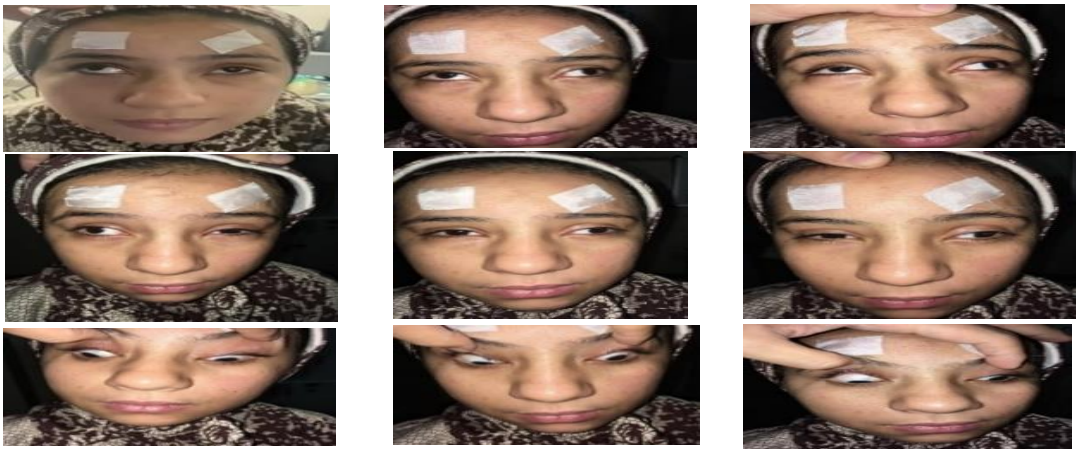

Figure (4): Patient No. 4: Female 20 years old Preoperative photos: Bilateral DRS type III, Lt. exotropia 35 prism diopter in 1ry position, V pattern exotropia, grade 3 globe retraction

Results:

This study was conducted in Sohag ophthalmology department over the duration of about two years in the period from June 2020 to July 2022

Strabismus clinic received 960 patients in this period. Only thirty-five patients had DRS (3.64% of all strabismus cases). Patients were divided into three groups:

Group A: 17 patients (48.6%) with DRS type I

Group B: 7 patients (20%) with DRS type II

Group C: 11 patients (31.4%) with DRS type III

Only 5 patients (14.3%) had bilateral DRS

Table (1): Distribution of DRS

Sex	Male	Female	All cases
Number, percentage of patients	12 (34.3%)	23 (65.7%)	35 (100%)
Range of age	5-25 years	2-26 years	2-26 years
Clinical types, corresponding age:			
Ortho-Duane	15.3±6.32	13.6±7.82	14±7.45
Exotropic Duane	14.9±5.8	12.92±8.2	13.68±8.67
Esotropic Duane	11.2±4.6	8.83±5.9	9.20±5.40
All types	13.8±5.57	11.78±7.3	12.69±7.65

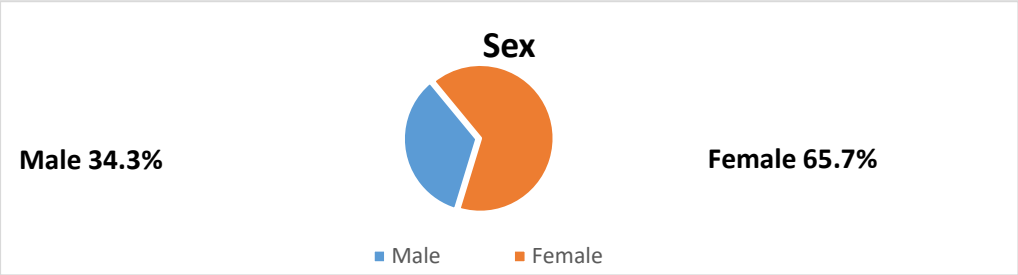

Figure (5): Sex distribution

Table (2): Distribution according to laterality

Rt.	8	22.8%
Lt.	22	62.9%
Bilateral	5	14.3%

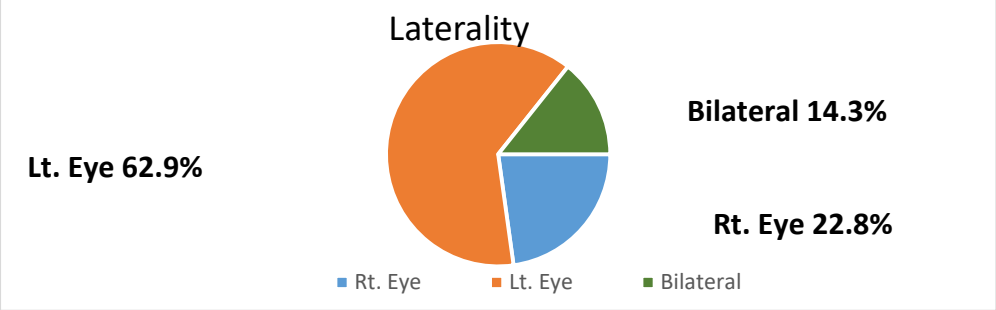

Figure (6): Distribution according to laterality

Table (3): Strabismus in primary position

Strabsimus in 1ry position	Number of patients	Percentage
Orthophoric DRS	9	25.7%
Esotropic DRS	8	22.9%
Exotropic DRS	18	51.4%

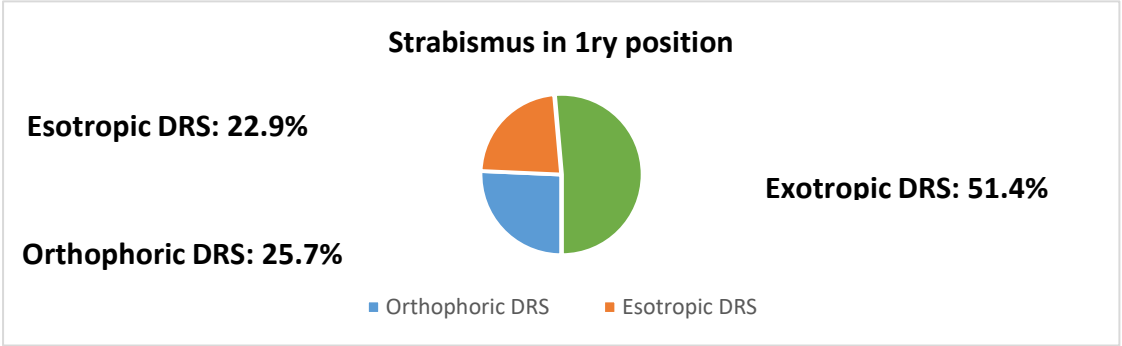

Figure (7): Strabismus in primary position

Discussion

Prevalence of DRS in this study was 35 patients out of 960 strabismus cases (3.64%). The mean age at diagnosis was 12.69±7.65 years in all cases. Patients with exotropic Duane were 13.68±8.67 years, while patients with esotropic Duane were 9.20±5.40 years. Cases with ortho-Duane were 14±7.45 years. Sex distribution was much higher in females by 65.7%, while male percentage was 34.3%.

According to laterality in this study, DRS in Lt. Eye had the highest percentage by 62.9%, then Rt. eye and both eyes in 22.8%, and 14.3% of patients respectively. Concerning prevalence according to its clinical type, it was proved that type I is the commonest type by 48.6%, while types II, III were much less by 20 %, 31.4% respectively. Concerning strabismus in primary position, most cases were exotropic Duane by 51.4% of all patients, then ortho-Duane and esotropic Duane by 25.7%, 22.9% respectively. As regard to association with systemic diseases, only one case had mental retardation (2.9%). No patients had amblyopia, and three patients only had refractive errors, one was myope, and two were hypermetropes.

According to Murillo-Corea et al,[10] prevalence of DRS is about 1/1000 in general population, with 60% female predominance. DRS accounts for 4% of all strabismus cases, most commonly is due to congenital aberrant ocular innervation

A study conducted in 2020 by Yun Jeong Lee et al[11] showed that DRS was more in males with a percentage of 56.9% compared to females by 43.1%. The prevalence of ortho-Duane were 53.85% of cases. Esotropic Duane comprised 33.85%, while exotropic Duane were only 12.3% of all DRS cases. In regard to laterality, Lt. eye was the most affected by 70.8% of cases, Rt. Eye was included in 24.6% of cases, while the disease was bilateral in 4.6% of cases. The mean age at diagnosis was 4.4±4.6 years (range: 4-19 years) to all cases. The youngest group were esotropic Duane 2.8±2.4 years (range: 0-8 years), exotropic Duane were the oldest: 10.4±5.6 years (range: 2-18 years), while ortho-Duane were 4.3±4.5 years (range: 0-19 years). Concerning systemic association, only 10.8% of all cases showed systemic disease, of which only 1.5% had neurological delay.[11]

In Egypt, another study made in 2020, including 238 DRS cases. [12] Gaballah K. et al identified type I to be the most common and type II was the least. Type I, 162 patients (68%), type II 12 patients (5%) and type III 64 patients (27%). Surgery was indicated in 98 (41%) of them.[12] In their study, females were predominant, constituting 125 patients (52.5%), and males 113 patients (47.5%). Left eye was more affected, in 110 patients (46.2%), right eye in 91 patients (38.2%) and bilateral in 37 patients (15.6%).

Regarding sex distribution, Gaballah K. et al.[12] and O'Malley ER et al. had cl-ose results to this study, but against the result of Yun Jeong Lee et al.[11] who described male predominance. Concer-ning Laterality, this study proves the Lt. Eye predominance that was also evidenced by other studies.

In regard to prevalence according to its clinical type, results of this study are close to results of Gaballah et al in 2020 in which type I was the commonest, then type III,[12] while type II was the least. Concerning association with systemic diseases, this study had only one case with mental retardation that comprises 2.9% of all cases; a result more or less near to Yun Jeong Lee et al. results.[11]

In this study, no patients had amblyopia, and three patients only had ref-

ractive errors, one was myope, and two were hypermetropes.

Association with amblyopia was found in only one case (8.3%) in a study conducted in Turkey by Aygit, E. et al,[13] so this agrees with this study proving the very low incidence of amblyopia. However, in Gaballah K. et al study, they noted more percentage of amblyopia; in 27 patients (11.3%) and treated in 13 patients under 10 years of age, by patching the normal eye.[12]

References

1. Sharma P., N. Gaur, S. Phuljhele, et al. What's new for us in strabismus? Indian journal of ophthalmology2017.90-184(3)65;

2. DeRespinis PA Caputo AR, Wagner RS, Guo S. . Duane's retraction syndrome. . Surv Ophthalmol 1993:38:257–88.

3. O'Malley ER Helveston EM, Ellis FD. Duane's retraction syndrome-plus. J Pediatr Ophthalmol Strabismus 1982:19:161–5.

4. Pfaffenbach DD Cross HE, Kearns TP. Congenital anomalies in Duane's retraction syndrome. Arch Ophthalmol 1972:;88:635–9.

5. Singh A Pandey PK, Agrawal A, et al. . Congenital cranial dysinnervation disorders. Int Ophthalmol 2017,81-37:1369.

6. Hoyt WF Nachtigäller H. . Anomalies of ocular motor nerves. Neuroanatomic correlates of paradoxical innervation in Duane's syndrome and related congenital ocular motor disorders. Am J Ophthalmol 1965:60:443–8.

7. A. Huber. Electrophysiology of the retraction syndromes. . Br J Ophthalmol 1974:58:293–300

8. Yuksel D Orban de Xivry JJ, Lefevre P. . Review of the major findings about Duane retraction syndrome (DRS) leading to an updated form of classification. . Vision Res 2010:50:2334–47.

9. Mohan K Sharma A, Pandav Differences in epidemiological and clinical characteristics between various types of Duane retraction syndrome in 331 patients. J AAPOS 2008:SS12:576–8.

10. Murillo-Correa C. E., V. Kon-Jara, E. C. Engle, et al. Clinical features associated with an I126M alpha2-chimaerin mutation in a family with autosomal-dominant Duane retraction syndrome. Journal of AAPOS : the official publication of the American Association for Pediatric Ophthalmology and Strabismus. 2009;13(3):245-8.

11. Lee Y. J., H. J. Lee, S. J. Kim. Clinical Features of Duane Retraction Syndrome: A New Classification. Korean J Ophthalmol. 2020;34(2):158-65.

12. Gaballah K. A., D. Shawky. Treatment modalities in Duane's Retraction Syndrome. Int J Ophthalmol. 2020;13(2):278-83.

13. Aygit E. D., M. Kocamaz, A. Inal, et al. Management of Duane retraction syndrome with prismatic glasses. Clinical ophthalmology (Auckland, NZ). 2017;11:697-700.