Management of duane retraction syndrome
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Background
Duane retraction syndrome (DRS) is a congenital ocular motility disorder most commonly characterized by the inability of the eye to abduct, sometimes limitation of adduction, and globe retraction with palpebral fissure narrowing on adduction.

Aim
To study the clinical features, management, and outcome of DRS.

Patients and methods
A prospective, nonrandomized study was carried out on patients attending the Outpatient Clinic of Ophthalmology in Menoufia University Hospital during the period of study from June 2017 to July 2019. This study was conducted on 51 patients with DRS. Ocular and systemic features were studied. Surgery was done when indicated to eliminate upshoot or downshoot, face turn with a horizontal deviation, and globe retraction. Preoperative and postoperative ocular deviation, angle of head turn, and severity of limitation in abduction and adduction were detected and analyzed.

Results
Type I DRS was more common (70.6%), with female preponderance (52.9%) and a predilection for the left eye (70.6%). Twenty-one (41.2%) cases were operated, by recessing MR and/or lateral rectus (LR) muscles and/or Y-splitting and acceptable improvement was detected. Mean esotropic DRS improved from 18±4.02 to 3±2.8 (P<0.001) and mean exotropic DRS improved from 14±2 to 5±1.4 (P<0.01). Upshooting/downshooting and narrowing of the palpebral fissure showed significant improvement (P<0.001 and <0.03, respectively).

Conclusion
Proper diagnosis of patients with DRS and proper plan for surgical management can reduce several complications with satisfactory postoperative outcomes.

Keywords: Duane retraction syndrome, restrictive strabismus, strabismus surgery
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Introduction
Duane retraction syndrome (DRS) is mainly restrictive strabismus and is now considered as a congenital cranial dysinnervation disorder. It is characterized by simultaneous contraction of the horizontal recti on adduction resulting in globe retraction. It may be orthotropic, esotropic, or exotropic [1].

DRS was classified by Huber into three types. Type I in which there is defective abduction with normal or minimally defective adduction; type II is characterized by a marked limitation of adduction with normal or minimally defective abduction, and type III is characterized by marked limitation of both abduction and adduction. This classification is based on prominent deficiency duction. Type I is the most common variety (78%), followed by type III (15%) and type II (7%) [2].

Patients and methods
A prospective, nonrandomized study was held on cases seeking medical consultation in the strabismus clinic in Menoufia Hospital attending during the period of study from July 2017 to July 2019. The study protocol was approved by the Ethics Committee of Menoufia Faculty of Medicine. Participant’s names were secret. All patients in this study received a detailed explanation about the aim, objectives, and methodology of the study before enrollment. The principal investigator was responsible for obtaining the participants’ approval and written informed consent.

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This study was applied on 51 patients diagnosed with DRS who had no prior ocular surgery or orbital trauma. Demographic data gathered included the type of Duane syndrome, laterality, sex, family history for strabismus, abnormal head position, primary position, presence of amblyopia, and presence of globe shift on adduction.

Spectacles were prescribed for patients with refractive errors. Patching of the nonamblyopic eye was advised. Minimum follow-up period of patients who did not undergo surgery was at least 6 months.

**Surgical management**

Twenty-one patients underwent strabismus surgery. Indication for surgery in DRS is directed to the correction of primary position deviation and anomalous face turn. Surgery can also be done to eliminate cosmetically unacceptable globe retraction and upshoots or downshoots.

Surgical techniques used were the following:

Bilateral MR recession in esotropic DRS when the deviation was more than 20 prism diopter (PD); unilateral MR and lateral rectus (LR) recession with Y-splitting in esotropic DRS when the deviation was less than 20 PD and associated with up/downshooting; lateral rectus recession in exotropic DRS; and Y-splitting of the LR in cases with orthotropia and upshooting/downshooting (Fig. 1).

The amount of recession done was according to strabismus surgery tables, but when bimedial rectus recession was indicated it was done asymmetrical with more recession on the healthy side.

**Results**

Our study included 51 patients in which there were 24 (47.1%) men and 27 (52.9%) women. Family history of DRS was positive in 5.9% of cases while strabismus other than DRS was positive in 11.8% of the patients. A family history of nystagmus was found in three (5.9%) patients while a family history of ptosis was found in three (5.9%) patients.

All patients were carefully evaluated for refractive errors and 33 patients were given glasses. Out of the 51 patients, 18 (35.3%) patients were emmetropic, 12 (23.6%) were hypermetropic, three (5.9%) patients were myopic, nine (17.6%) patients with simple myopic astigmatism, three (5.9%) patients with compound myopic astigmatism, three (5.9%) patients with compound hyperopic astigmatism, and three (5.9%) patients were with mixed astigmatism.

Amblyopia was found in 21 (41.2%) cases, six (11.8%) cases have amblyopia in the right eye, three (5.9%) cases have amblyopia in the left eye, and 12 (23.5%) cases were bilateral. Regarding the cause of amblyopia, strabismic amblyopia was present in three (14.3%), anisometropic amblyopia was present in 15 (71.4%), and mixed amblyopia was present in three (14.3%).

Our 51 patients were divided into different types according to Huber classification: type I DRS (Fig. 2) was present in 36 (70.6%) patients, type II (Fig. 3) was present in six (11.8%) patients, and type III (Fig. 4) was present in nine (17.6%) patients. Out of the 51 patients, 48 (94.1%) had unilateral DRS: 12 (23.5%) had right eye DRS, 36 (70.6%) had left eye while three (5.9%) cases had bilateral DRS (Fig. 5). In unilateral cases, a predilection for the left eye (70.6%) was seen in our study (Table 1).

Of the 51 patients studied we found that the number of orthophoric DRS patients were 15 (29.64%), while
patients with orthotropia associated with upshooting or downshooting were three (5.9%). Esotropic DRS patients were 15 (29.4%), while those with esotropia associated with upshooting or downshooting were nine (17.6%). Exotropic DRS patients were six (11.8%), while those with exotropia associated with upshooting or downshooting were three (5.9%) patients (Table 2).

The deviation in primary position in each type was the following:

1) Of 36 patients of DRS type I, 12 (33.3%) cases were orthotropic, 15 (41.7%) cases were esotropic, and nine (25%) cases were esotropic with upshooting/downshooting.
Six patients were of DRS type II, three (50%) cases were orthotropic with upshooting/downshooting while other three (50%) cases were exotropic.

Of the nine patients of DRS type III, three (33.3%) cases were orthotropic, three (33.3%) cases were exotropic, and other three (33.3%) cases were exotropic with upshooting/downshooting.

Twenty-one patients underwent surgeries, of which 15 patients were of type I (Fig. 6), three patients were of type II (Fig. 7), and three patients were of type III. Surgical techniques used were the following:

1. Nine cases of esotropia in the primary position underwent bilateral MR recession.
2. Six cases with esotropia and upshooting/downshooting had unilateral MR and LR recession with Y-splitting.
3. Three cases with exotropia had lateral rectus recession.
4. Three cases with orthotropia and upshooting/downshooting had Y-splitting of the LR (Table 3).

Table 1 Features of Duane syndrome in the study population

| Variables                | n (%) |
|--------------------------|-------|
| Type                     |       |
| Type I                   | 36 (70.6) |
| Type II                  | 6 (11.8)  |
| Type III                 | 9 (17.6)  |
| Laterality               |       |
| Bilateral (OU)           | 3 (5.9)  |
| Unilateral               | 48 (94.1) |
| OD                       | 12 (23.5) |
| OS                       | 36 (70.6) |

Table 2 Ocular alignment status in the study population

| Status                          | n (%) |
|---------------------------------|-------|
| Orthophoria                     | 15 (29.4) |
| Orthophoria with upshooting/downshooting | 3 (5.9)  |
| Esotropia                        | 15 (29.4) |
| Esotropia with upshooting/downshooting | 9 (17.6) |
| Exotropia                        | 6 (11.8)  |
| Exotropia with upshooting/downshooting | 3 (5.9)  |

Preoperative and postoperative case of DRS type I of the left eye after bimedial rectus recession. DRS, Duane retraction syndrome.
Among patients with horizontal deviation there was significant improvement. Mean esotropic DRS improved from 18±4.02 to 3±2.8 ($P<0.001$) and mean exotropic DRS improved from 14±2 to 5±1.4 ($P<0.01$). Narrowing of the palpebral fissure showed significant improvement ($P<0.03$). Patients underwent lateral rectus Y-splitting showed elimination of upshot/downshoot postoperatively ($P<0.001$) (Table 4).

**Discussion**

O’Malley et al. [3] reported dominance of the left eye and female patients in all types of unilateral Duane syndrome. We had a similar observation in unilateral types I Duane syndrome but not in type II Duane syndrome in which the number of male patient equals the number of female patients and also not in type III in which all the patients were men. Few studies showed male preponderance like an Egyptian study in 2015 which included 198 patients; there was slight male preponderance (52%) [4].

In our study, type I was found in 70.58%, type II in 11.76%, and type III in 17.65% eyes. Mohan and Saroha [5] and Kekunnaya et al. [6] also noticed type I to be the most common in unilateral cases and type II to be least common in 331 and 441 patients studied, respectively. The mean age at presentation was higher in DRS type III compared with those with type I DRS and type II DRS. This finding was also reported in a previous study [7].

Shauly et al. [8] noticed that cases with unilateral DRS type I have esotropia in forced primary position, while those with type II have exotropia in forced primary position, and those with type III have equal esotropia and exotropia. In our study, unilateral type I Duane syndrome had esotropia in 66.7% of cases of type I while the remaining cases (33.3%) were orthotropic. Type II had exotropia in 50% while the remaining cases (50%) were orthotropic. Type III had exotropia in 66.7% and other cases (33.3%) were orthotropic and no cases of esotropia.

The incidence of amblyopia in DRS in this study falls within the range reported in the literature. Mehel et al. [9] reported that 50% of patients with unilateral DRS have amblyopia and ~25% of those with bilateral involvement have amblyopia. Amblyopia in DRS has been said to be the result of anisometropia more often than strabismus [10]. In our study, the incidence of amblyopia was 41% and it was bilateral in more than half of the cases of amblyopia. The main cause of amblyopia in our study was due to error of refraction (71.4%), while strabismus was responsible for 14.3% of cases and mixed cause was found in 14.3% of cases of amblyopia.
Table 3 Preoperative and postoperative details

| Type of duane | N | Preoperative | Operation | Postoperative |
|---------------|---|--------------|-----------|--------------|
|               | PP | ABD | ADD | AHP | Upshoot/downshoot | PP | ABD | ADD | AHP | Upshoot/downshoot |
| Type I        | 1  | ET  | −4.00 | 0  | L++ | No  | Bi-MR Recession | ET 4 | Δ | −3.00 | −1.00 | N  | No  |
|               | 2  | ET  | −4.00 | 0  | L++ | No  | ET 2 | Δ | −3.00 | −1.00 | L+ | No  |
|               | 3  | ET  | −4.00 | 0  | R++ | No  | ET 6 | Δ | −3.00 | −1.00 | R+ | No  |
|               | 4  | ET  | −4.00 | 0  | L++ | No  | XT 6 | Δ | −2.00 | −2.00 | N  | No  |
|               | 5  | ET  | −3.50 | 0  | L++ | No  | XT 6 | Δ | −3.00 | −1.00 | N  | No  |
|               | 6  | ET  | −4.00 | 0  | R++ | No  | XT 4 | Δ | −3.00 | −2.00 | N  | No  |
|               | 7  | ET  | −4.00 | 0  | L++ | No  | ET 5 | Δ | −3.00 | −1.00 | L+ | No  |
|               | 8  | ET  | −3.00 | 0  | L++ | No  | ET 4 | Δ | −3.00 | −1.00 | N  | No  |
|               | 9  | ET  | −4.00 | 0  | R++ | No  | ET 6 | Δ | −3.00 | −1.00 | R+ | No  |
|               | 10 | ET  | −4.00 | 0  | L+  | +3.00 | unilateral MR Recession +LR recession +Y-splitting | Ortho | 0  | −2.00 | −2.00 | N  | +1.00 |
|               | 11 | ET  | −3.00 | 0  | R+  | +3.00 | Ortho | 0  | −2.00 | −1.00 | N  | +1.00 |
|               | 12 | ET  | −4.00 | 0  | L+  | +3.00 | Ortho | 0  | −3.00 | −2.00 | N  | +1.00 |
|               | 13 | ET  | −3.00 | 0  | R+  | +4.00 | Ortho | 0  | −1.00 | −1.00 | N  | +1.00 |
|               | 14 | ET  | −3.00 | 0  | L+  | +4.00 | Ortho | 0  | −1.00 | −1.00 | N  | +2.00 |
|               | 15 | ET  | −3.00 | 0  | L+  | +4.00 | Ortho | 0  | −1.00 | −1.00 | N  | +2.00 |
| Type II       | 16 | Ortho | 0  | −2.00 | N  | +4.00 | Y-splitting of the LR | Ortho | 0  | −1.00 | −1.00 | N  | +2.00 |
|               | 17 | Ortho | 0  | −2.00 | N  | +4.00 | Ortho | 0  | −1.00 | −1.00 | N  | +2.00 |
|               | 18 | Ortho | 0  | −2.00 | N  | +4.00 | Ortho | 0  | −1.00 | −1.00 | N  | +1.00 |
| Type III      | 19 | XT  | −2.00 | −2.00 | N  | No  | Unilateral LR recession | Ortho | 0  | −1.00 | −1.00 | N  | No  |
|               | 20 | XT  | −2.00 | −2.00 | N  | No  | XT6 | Δ | −1.00 | −1.00 | N  | No  |
|               | 21 | XT  | −3.00 | −2.00 | N  | No  | XT 4 | Δ | −2.00 | −1.00 | N  | No  |

+, mild; ++, moderate; ABD, limited abduction; ADD, limited adduction; AHP, abnormal head posture; L, left face turn; LR, lateral rectus; PP, primary position; R, right face turn.

Table 4 Preoperative and postoperative deviation

| Variables                      | Preoperative | Postoperative | P value |
|--------------------------------|--------------|---------------|---------|
| Angle of deviation (Δ)         |              |               |         |
| ET                             | 18±4.02      | 3±2.8         | 0.001*  |
| XT                             | 14±2         | 5±1.4         | 0.01*  |
| Upshooting/downshooting        | 1.7±1.8      | 0.6±0.8       | 0.001*  |
| Narrowing of the palpebral fissure |            |               |         |
| G0                             | 0            | 0             | 0.03*  |
| G1                             | 10           | 0             |         |
| G2                             | 11           | 3             |         |
| G3                             | 0            | 18            |         |
| G4                             | 0            | 0             |         |

*a*Paired t test. *b*One-way repeated measures analysis of variance. *P* value is significant.
In a review of DRS patients in previous studies regarding errors of refraction, it was found that hyperopia was the major error of refraction while myopia and emmetropia were relatively equal [11]. This may be due to the fact that most of patients in these studies were children under the age of 4 years and it was reasonable to find that hyperopia is the major error in these patients. In our study, astigmatic error was found in nearly 35% and emmetropia was found in 35% of patients while hyperopia was found in 23% of patients, which differs slightly from previous studies and that may be due to the higher mean age of children in our study or due to the different geographical area.

The average incidence of anisometropia in major studies performed was 23%, although Tredici and van Noorden [12] found it to be only 3%. In our study, the incidence of anisometropia was nearly 6%. In our 21 surgical patients (15 of type I, three of type II, and three of type III), the majority had an acceptable surgical outcome with significant improvements in AHP and forced primary position alignment. It was found that compromising the rotation of the medial rectus balances to some extent the limitation of the lateral rectus [13]. In our 15 patients with esotropia (all of them were unilateral type I DRS) nine underwent bilateral medial rectus recession while the other six patients underwent unilateral MR recession and LR recession with Y-splitting to treat associated upshoot or downshoot. Surgical outcomes in our 15 esotropic patients were comparable to those in a study by Mehendale et al. [14] where improvements in esotropia and head posture have occurred. Three exotropic DRS patients underwent surgery with small angle and all of them were of type III and received unilateral lateral rectus recession showing improvement in the angle of deviation postoperatively.

Upshoot and downshoot in DRS patients are believed to be due to co-contracture of the medial and LR muscles and a taut LR muscle [15]. Splitting of an LR muscle lessens lateral slippage of the muscle [16]. In our study, there was nine patients with upshoot or downshooting; six of them were of type I with associated esotropia while the remaining three patients were of type II and orthotropic. These patients underwent Y-splitting of the lateral rectus and we found significant improvement in most of the features in all the patients after performing the operation.

Conclusion and recommendation
Unilateral types I, II, and III Duane syndrome differ in the mean age at presentation, primary position horizontal deviation, upshoot and downshoot, and associated ocular and systemic abnormalities. Anisometropia and amblyopia should be searched for and treated aggressively in DRS patients. The surgical management of a case of Duane syndrome should be tailored according to the severity of the problem and specific clinical criteria of the patient. Proper diagnosis of patients with DRS and surgical management help in avoiding several adverse situations.

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Conflicts of interest
There are no conflicts of interest.

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