

# Treatment modalities in Duane's Retraction Syndrome

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## Abstract

• **AIM:** To study the different treatment modalities needed in cases of Duane's Retraction Syndrome (DRS).

• **METHODS:** This prospective study undergone in more than four years, in Alexandria, included 238 patients of DRS, including type I, 162 patients (68%), type II 12 patients (5%) and type III 64 patients (27%). Surgery was indicated in 98 (41%) of them, to eliminate abnormal head posture, deviation of the eye in primary position, severe retraction of the globe or cosmetically unacceptable upshoot with attempted adduction.

• **RESULTS:** Type I was the most common and type II was the least. Females were predominant in this study, 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%). Amblyopia was found in 27 patients (11.3%) and treated in 13 patients under 10 years of age, by patching the normal eye. Ninety-eight patients (41%) were operated, the results were most satisfactory and a nomogram is followed in the surgical plan.

• **CONCLUSION:** The surgical management is needed in less than half of the cases and should be planned for every case individually according to the clinical findings, planned nomogram and modified intraoperatively according to the anatomical findings during surgery.

• **KEYWORDS:** Duane's Retraction Syndrome; globe retraction; globe upshoot; medial rectus; lateral rectus; forced duction test

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## INTRODUCTION

Duane's Retraction Syndrome (DRS) is a congenital complex strabismus characterized by variable limitation of abduction, variable limitation of adduction, and globe retraction with narrowing of the palpebral fissure on attempted adduction<sup>[1-3]</sup>. It is characterized by co-contraction of horizontal recti on attempted adduction causing globe retraction along with variable amounts of upshoot or downshoot. The prevalence is about 1/1000 in general population, females are more affected, constituting 60% of cases, and is the most common type of aberrant ocular innervation. About 80% of cases are unilateral, showing a predominance of the left eye<sup>[4]</sup>. Huber has classified this syndrome depending upon clinical findings: type I characterized by defective abduction, with normal or minimally defective adduction; type II characterized mainly by defective adduction with normal or minimal defective abduction and type III characterized by marked limitation of both abduction and adduction<sup>[5]</sup>. It may have limited abduction or adduction or both and present as esotropic, exotropic, or orthotropic Duane. So, It can be also classified on the basis of the primary position deviation into esotropic-DRS (eso-DRS), exotropic-DRS (exo-DRS), and orthotropic-DRS (ortho-DRS). This classification is easier to use for surgical planning<sup>[6-7]</sup>. The indications for surgery are abnormal head posture, deviations in the primary position, severe retraction and narrowing of the palpebral fissure and upshoot or downshoot during adduction and sometimes also to improve abduction<sup>[8]</sup>. Before planning the surgery in DRS it is important to perform the forced duction test to detect the tightness of medial rectus muscle (MR) and lateral rectus muscle (LR). The main surgical procedures, in eso-DRS, a single MR recession is done in the affected eye in case of deviations less than 20 prism diopter<sup>[9]</sup>, other treatment options in larger angle include asymmetric bilateral MR recessions<sup>[10]</sup>. In cases with upshoot or downshoot, recession of the ipsilateral LR in addition to the recession of MR. Y-splitting of the recessed LR has been shown to improve the upshoot and downshoot<sup>[11]</sup>. Also, some authors perform vertical rectus transposition to improve the abduction movements<sup>[12]</sup>. In exo-DRS, many procedures have been performed by different authors including LR recession, with or without Ysplitting, or LR periosteal fixation<sup>[13]</sup>. The aim of this study was to plan the different modalities of treatment of DRS so as to improve as much as possible both the functional and the cosmetic appearance.

## SUBJECTS AND METHODS

**Ethical Approval** This prospective study started January 2015 till the end of February 2019, in Alexandria, Egypt. Patients diagnosed as DRS were included in this study. An informed written consent was obtained from patients, or guardians of those aged less than 18y, according to Ethical Approval Committee in Helwan University, prior to inclusion in this study. The total number of cases included in this study was 238 cases of DRS.

Data collected included age, sex, family history of strabismus or aberrant ocular innervation. Examination included general appearance, any abnormal head posture, any systemic congenital anomalies, parallel eyes or presence of strabismus in primary position, ocular motility examination in different directions of gaze. Cases were classified according to Huber's classification, depending upon limitation of abduction, limitation of adduction, or limitation of both abduction and adduction into type I, II and III respectively.

The degree of globe retraction and narrowing of the palpebral fissure with adduction was graded into mild, moderate or severe (mild insignificant, moderate: detected and complained of by patients, severe: cosmetically unacceptable). Presence or absence of upshoot or downshoot with adduction was also noted.

Sensory or visual functions were evaluated in all cases, by detecting errors of refraction, anisometropia, amblyopia which is at least two lines difference between the two eyes and managed in young patients as usual in strabismus cases by appropriate glasses and patching of the dominant eye.

The cases were managed regarding sensory functions first. Suppression or amblyopia were treated in patients in the critical period of age; whereas motor functions and cosmetic appearance were targeted thereafter. Surgery was planned in presence of abnormal head posture, esotropia or exotropia in primary position, moderate and severe globe retraction or cosmetically unacceptable upshoot or downshoot with adduction.

The surgical procedure of cases was planned before management according to clinical findings, forced duction test before surgery, and modified according to anatomical findings intraoperatively.

In cases of esotropia in primary position and/or face turn, the plan of surgery was to recess the MR, after forced duction test (FDT), for the tightness of the muscle and the anatomical findings during surgery might affect the surgical outcome. LR is not touched, and resection is contraindicated. In large angle esotropia, the contralateral MR is recessed as well.

In cases of exotropia in primary position, and /or face turn the plan of surgery was to recess the LR, after FDT, for the tight muscle when recessed will greatly change the position of the

**Table 1 Sex distribution of Duane's Retraction Syndrome**

Parameters	Females	Males	Total
Number	125	113	238
Percentage	52.5%	47.5%	100%

globe, and also the surgical technique is modified according to findings during surgery, the MR is not touched.

In cases of severe globe retraction and cases of cosmetically unaccepted upshoot or downshoot, the management was to recess the two horizontal recti in the same eye, and according to primary position of the globe, whether no deviation, esotropia or exotropia in primary position, a nomogram is followed.

Recessing the MR by (X) mm and the LR 1.5(X) mm, that is to recess the LR 50% more than the MR. This is to preserve the same preoperative ocular alignment while curing the retraction and upshoot, in ortho-DRS.

If additional esotropia is present with the severe globe retraction and upshoot, the nomogram is modified so as to recess the MR more, in order to correct the esotropia as well and the nomogram will be: MR recession (X) mm + 1 mm/5 prism diopters esotropia, LR recession 1.5(X) mm. This modification is to cure the esotropia in addition to globe retraction and upshoot.

If additional exotropia is present with the severe globe retraction and upshoot, the nomogram is modified so as to recess the LR more, in order to correct the exotropia as well and the nomogram will be: MR recession (X) mm and LR recession 1.5(X) mm + 1 mm/3 prism diopters exotropia, this modification is to cure the exotropia in addition to globe retraction and upshoot.

## RESULTS

The total number of cases included in this study was 238 cases. Their age ranged from 12mo to 41y, mean 12.4y. Females constituted the majority and were 125 cases (52.5%) while males were 113 cases (47.5%; Table 1).

According to Huber's classification, type I constituted 162 patients (68%; Figures 1-4), type II 12 patients (5%; Figure 5), and type III 64 patients (27%; Figure 6, Table 2).

As regards laterality, left eye was affected in 110 cases (46.2%), right eye in 91 cases (38.2%) while bilateral cases were 37 (15.6%; Table 2).

The bilateral cases of DRS, were 37 patients, most of them were type I, 20 cases (54%), type III, 16 cases (43.3%), and type II, only one case (2.7%).

In this study, although DRS is known as one of the known congenital motor disorders, the sensory functions were studied and managed first.

Regarding the sensory functions, and visual functions including anisometropia, suppression and amblyopia, they

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Parameters	Type I	Type II	Type III	Total	<i>n</i> (%)
No.	162 (68)	12 (5)	64 (27)	238 (100)	
Unilateral	142	11	48	201 (84.4)	
Left eye	75	6	29	110	
Right eye	67	5	19	91	
Bilateral	20	1	16	37 (15.6)	
Amblyopia	16	2	9	27 (11.3)	
Operated	61	6	31	98 (41)	

Parameters	No.	Percentage
Type of surgery in DRS I		
Recession of one MR	29	
Recession of two MR	8	
Recession of MR & LR	24	
Total	61	62.24%
Residual esotropia	3	
Consecutive exotropia	1	
Type of surgery in DRS II		
Recession of one LR	3	
Recession, Y-split LR	2	
Recession of two LR	1	
Total	6	6.12%
Residual exotropia	2	
Consecutive esotropia	0	
Type of surgery in DRS III		
Recession MR & LR	31	31.64%
Postoperative esotropia	1	
Postoperative exotropia	1	
Total operated patients	98	100%

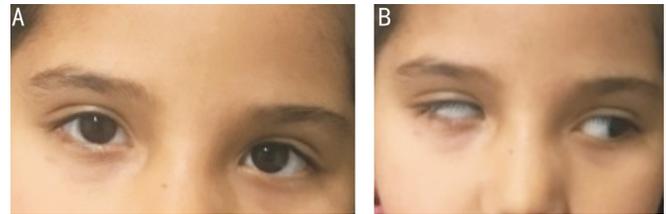
were first considered and targeted, as other types of strabismus. Amblyopia was recorded in 27 cases only (11.3%) and was managed by part time patching of the dominant eye in 13 patients under 10y of age.

Then, the cosmetic appearance, that motivated the patients to be included in this study, was targeted. Patients were operated in presence of deviation of the eye in primary position, abnormal head posture, moderate and severe globe retraction and narrowing of the palpebral fissure or the presence of a cosmetically unacceptable upshoot or downshoot of the globe with attempted adduction. The patients who needed operation were 98 patients, 61 patients type I, six patients type II and 31 patients type III (Tables 3, 4). The type of surgery used was, recession of one MR in 29 patients, with esotropic type I, recession of two medial recti muscles in eight patients with large angle esotropia type I (Figures 7, 8).

Patients with disfiguring upshoot of the globe or severe globe retraction with attempted adduction, where operated by recession of MR and LR of the same eye to relieve the tension



**Figure 1 Left DRS type I** A: Looking right, left narrowing of palpebral fissure; B: Looking straight, eyes parallel; C: Looking left, defective abduction of left eye.



**Figure 2 Right DRS type I** A: Looking straight, eyes are parallel; B: Looking left, right eye showing narrowing of palpebral fissure, globe upshoot.



**Figure 3 Right DRS type I with upshoot, severe globe retraction** A: Right defective abduction when looking right; B: Looking straight forwards; C: Looking left, right eye severe narrowing of palpebral fissure, globe upshoot.



**Figure 4 Right DRS type I with upshoot and downshoot** A: Looking straight forwards eyes are parallel; B: Looking left, above midline, right globe upshoot; C: Looking left, below midline, right globe downshoot.

on the globe (Figure 9), and was performed in 24 patients of type I, and 31 patients of type III (Tables 3, 4).

Recession of one LR in three patients with type II exo-DRS, recession of one LR with Y-splitting in two patients, type II with mild upshoot (Figure 5), recession of two lateral recti muscles in one patient with bilateral type II DRS, with disfiguring large angle exotropia (Figure 10, Tables 3, 4).

The plan of surgery was tailored for every patient according to clinical findings, amount of ocular deviation in primary position, ocular motility and degree of limitation of movements, forced duction test, degree of globe retraction and globe upshoot in attempted adduction, anatomical findings during surgery (Figure 11), the recession procedure is modified to be performed by hang-back technique in cases of tight muscle causing indentation of sclera, and thin sclera.

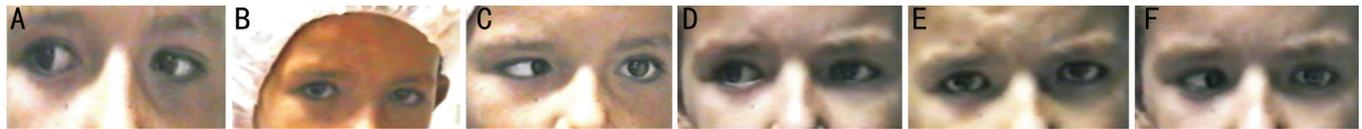
The nomogram that was used in surgery in cases of severe globe retraction and upshoot of the globe where the two horizontal recti are recessed in the same eye, to cure or greatly



**Figure 5 Left DRS type II, exotropic, with upshoot, and globe retraction with attempted adduction** A-C: Preoperative; D, E: Postoperative, after LR recession with Y-splitting.



**Figure 6 Left DRS type III, defective abduction and adduction with severe globe retraction and upshoot** A: Looking right, left eye shows severe narrowing of palpebral fissure with upshoot; B: Looking straight forwards, eyes parallel, ortho-DRS; C: Looking left, defective abduction.



**Figure 7 Left DRS type I, preoperative and postoperative after left MR recession** A: Preoperative, looking right; B: Preoperative looking straight, left esotropia, left face turn; C: Preoperative, looking left, defective abduction of left eye; D: Postoperative looking right; E: Postoperative, looking straight forward, eyes parallel, no face turn; F: Postoperative, looking left.



**Figure 8 Left DRS type I, esotropic, preoperative, and postoperative residual esotropia** A: Left DRS I preoperative; B-E: Postoperative, residual esotropia.



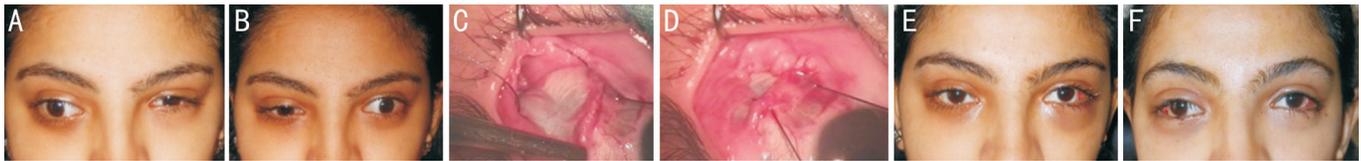
**Figure 9 Left DRS type III preoperative and postoperative, after ipsilateral left MR & LR recession** A: Left DRS III preoperative looking right, severe palpebral fissure narrowing, severe upshoot; B: Preoperative looking straight forward; C: Preoperative looking left, defective abduction; D: Postoperative looking right, after ipsilateral MR & LR recession, great amelioration of globe retraction and no upshoot of the globe; E: Postoperative looking straight; F: Post operative looking left, still defective abduction.

**Table 4 Descriptive analysis of the operated cases according to DRS type**

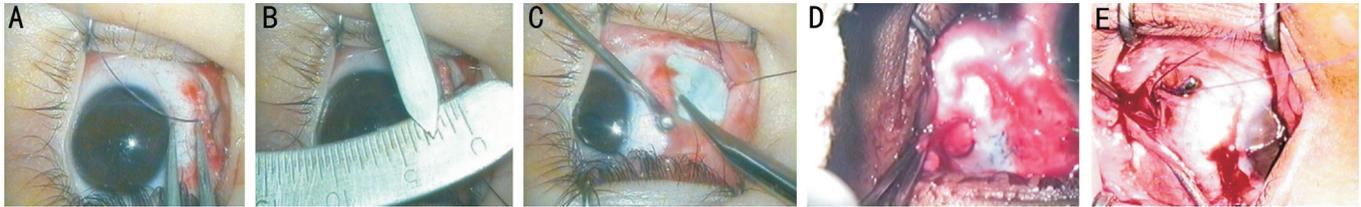
Parameters	Group DRS I (n=61)			Group DRS II (n=6)	Group DRS III (n=31)
	One MR (n=29)	Two MR (n=8)	MR & LR (n=24)		
Preoperative					
Median (min-max)	20 (15-30)	43.5 (40-48)	20 (15-25)	20 (15-80)	5 (0-15)
Mean±SD	21.5±4.5	43.8±3.2	20.9±2.9	29.2±25.2	7.03±5.6
Postoperative					
Median (min-max)	2 (-5-25)	2.5 (2-7)	2 (0-5)	2 (0-25)	1 (-5-6)
Mean±SD	2.6±4.7	3.5±2	2.4±1.5	5.7±9.7	0.9±1.6

improve the narrowing of palpebral fissure and upshoot and at the same time preserve the same preoperative ocular alignment, the nomogram used was the following: 1) Recessing the MR by (X) mm and the LR 1.5(X) mm; 2) Recessing the MR by (X) mm+1 mm/5 prism diopters esotropia, LR recession

1.5(X) mm, This is to cure the esotropia in addition to globe retraction and upshoot. 3) Recessing the MR by (X) mm and LR recession 1.5(X) mm+1 mm/3 prism diopters exotropia, this is to cure the exotropia in addition to globe retraction and upshoot.



**Figure 10 Bilateral DRS type II, exotropia, downshoot, preoperative, operative, and postoperative after bilateral LR hang back recession with Y-splitting** A: Bilateral DRS type II, looking right, defective left adduction; B: Looking left, defective right adduction; C: Intraoperative, scleral indentation due to tight LR muscles; D: Recession with hang-back of LR; E: Postoperative, much improvement; F: Residual exotropia.



**Figure 11 Abnormal anatomical findings during surgery** A: Anterior insertion of MR; B: Anterior insertion of MR; C: Thin sclera; D: Scleral pigmentation; E: Abnormal direction of LR.



**Figure 12 Left DRS type I syndrome, esotropia, difficult surgery** A: DRS type I looking right; B: Looking straight forwards, left esotropia (eso-DRS); C: Looking left, defective abduction; D: Intraoperative hypertrophic MR muscle; E: Intraoperative abnormal anterior insertion of MR.

This nomogram is also modified according to the anatomical findings during surgery. The postoperative results were satisfactory for most patients, residual deviation resulted in six patients, and consecutive deviation in two patients, where they are knowing before surgery that the aim is to greatly improve the appearance and not to completely cure DRS<sup>[14]</sup>.

All operated patients were satisfied with their postoperative results except two patients who were only fairly satisfied, one of them, was the one having bilateral type II DRS, presenting with very large angle of exotropia the angle of preoperative exotropia was 80 prisms and at the same time severe narrowing of the palpebral fissure and globe retraction with attempted adduction, in this patient, the two lateral recti muscles were targeted with large recession with hang-back technique, and the exotropia was greatly ameliorated to be 25 prisms postoperatively, but the two medial recti could not be recessed to ameliorate the narrowing of the palpebral fissure for this would worsen the exotropia which was the main concern of this patient and also could not be resected to diminish the exotropia, for carrying the risk of aggravating the enophthalmos (Figure 10). The other difficult patient, eso-DRS type I, with 25 prisms where the preoperative plan was left MR recession, but intraoperatively it was technically very difficult and risky for the MR was very tight and sutures application had a great risk of globe perforation, and the deviation remained the same (Figure 12A-12C).

The findings during surgery were taken into consideration in modifying the plan of surgery, and included very tight muscle and scleral indentation underneath (Figure 10D, 10E), thin sclera, scleral pigmentation, abnormal direction of LR muscle, very tight muscle (Figure 11) and abnormal anterior insertion of MR muscle (Figure 12D, 12E).

#### DISCUSSION

In this series of cases of DRS, only 41% of cases of DRS needed to be operated and about 60% did not need any intervention. Cases where no indication for surgery, where binocular functions were good, and absence of deviation in primary position, no abnormal head posture, and mild globe retraction with no upshoot of the globe, these cases were 140 patients, and constituted 59% and were followed up every six months during the study period, with no changes.

Eso-DRS was managed by recession of one MR in 29 patients, recession of the two medial recti muscles in eight patients with large angle esotropia, considering the forced duction test that showed MR tightness, in many cases and where resection of the LR is contraindicated to avoid aggravation of globe retraction. This was in contrary to other studies as Kraft<sup>[15]</sup> series who advised resection of LR to be added to recession of MR.

In exo-DRS, LR recession was performed in three patients, LR recession with Y-split in two cases, and bilateral LR hang-back recession in a case of bilateral type II<sup>[11]</sup>. MR is not included in surgery in exotropia without upshoot.

In ortho-DRS, in presence of severe globe Retraction and upshoot, recession of the two ipsilateral horizontal recti was performed, using the nomogram, with satisfactory results, in 24 patients in type I, and in 31 patients with Duane III who suffered from severe globe retraction and upshoot with attempted adduction.

We did not follow the studies that performed vertical recti transposition for upshoot or downshoot of Agarwal *et al*<sup>[12]</sup>, and this procedure was not performed in this study, depending on EMG study in cases of Duane Retraction Syndrome, showing co-contraction of MR and LR, and demonstrating the cause of upshoot as a leash effect of tight horizontal recti muscles, or co-contraction of the two ipsilateral horizontal recti muscles, rather than activity of vertical recti and so planning and targeting the ipsilateral MR and LR recession accordingly<sup>[14]</sup>.

The nomogram used for ipsilateral horizontal recti recession, to preserve the same preoperative globe position, in ortho-DRS, but targeting the narrowing of the palpebral fissure and upshoot with attempted adduction, was recessing the MR (X) mm, recessing the LR 1.5(X) mm, to preserve the same preoperative alignment in ortho-DRS, modifying this nomogram according to deviation in primary position and to anatomical changes found during surgery.

Not all cases of DRS needed to be treated. Those who needed management, sensory functions were managed first, then the plan of surgery was tailored for each patient, depending upon the preoperative clinical picture, planned nomogram, modified by FDT and by intraoperative findings during surgery. In cases of esotropia or exotropia in primary position, recession of MR or LR respectively, mainly by hang-back technique for safety of the thin sclera under the tight muscles. In cases of severe globe retraction and upshoot of the globe, recession of the ipsilateral medial and lateral recti, following the nomogram to preserve the preoperative alignment in ortho-DRS, and to put in consideration preoperative deviation to be added to the nomogram.

#### ACKNOWLEDGEMENTS

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