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Original Article

SURGICAL OUTCOMES OF DUANE RETRACTION SYNDROME IN SOHAG UNIVERSITY HOSPITAL

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Abstract

Introduction: *Duane retraction syndrome is a congenital complex strabismus. It was first described in 1887. It is characterized mainly by 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.* ***Aim of the work***: *to study the surgical outcomes of Duane retraction sundrome (DRS) in Sohag University Hospital.* ***Patients and Methods***: *A prospective non-randomized interventional clinical study done in Sohag University Hospital.* ***Results***: *35 patients had DRS out of 960 strabismus patients, only twenty patients were indicated for surgery. Marked improvement of primary ocular deviation, globe retraction and overshooting was noticed.*

Keywords: *Duane, Sohag, Surgical outcomes*.

1. Introduction

Duane retraction syndrome (DRS) is a congenital syndrome which is characterized by limited eye movement either inwards, outwards or both. The eyeball retracts and the palpebral fissure narrows as the affected eye shifts inward towards the nose. When a patient tries to look inside, their eye may shoot upward or downward (upshoot or downshoot) [1]. DRS is a congenital ocular motility condition that represents sporadically in about one to five percent of all strabismus cases [2]. To achieve fusion and correct the binocular dysfunction that may be brought on by vertical deviations, abnormal head posture is frequently maintained by the patient. Usually, the head is tilted in the direction of the weak muscle. In this manner, binocularity can be maintained while diplopia can be avoided [3]. While 10% of instances are bilateral, the majority are sporadic and unilateral. Left eye is affected more commonly than right eye, and the majority of those affected are females [4]. In addition to a higher risk of inflammation, it has been cleared that increased oestrogen levels in females during embryogenesis increase the risk of venous thromboembolic events. In addition, right to left shunts frequently result in embolic events that impact the left carotid artery, leading to dysregulated apoptosis and left

eye misinnervation [5]. Etiopathogenesis: Congenital cranial dysinnervation diseases (CCDD), which include DRS, are developmental malformations of the cranial nerves that are characterized by aberrant movements of the eye, eyelids, and/or face [6]. The third nerve's branches are diverted to the lateral rectus. The fundamental pathophysiology of Duane retraction syndrome is an incorrect or paradoxical innervational impulse to the horizontal recti. From paradoxical to subnormal innervation, the LR may display a spectrum of aberrant innervational patterns. Because growing nerve fibres are diverted to the sixth nerve, even the medial rectus (MR) may have abnormal innervations. Constant esotropia and upshoots, respectively, might lead to secondary muscle alterations in the form of contracture and subsequent fibrosis of the MR and superior rectus [7]. Classification of DRS: The commonest and widely used classification of DRS is Huber's classification [8]; which depends on electromyography to detect limitation of movement. Type I has limited abduction, type II is characterized by limited adduction, while in type III both abduction and adduction are limted. Bilateral DRS is less common and represents about 10-24% of all cases of DRS [9]. The treatment of DRS is considered a challenge, and surgeons across the world have different favoured methods. Generally speaking, the surgical strategy and goal of DRS strabismus surgery usually depends on primary position deviation, degree of abnormal head posture, severity of globe retraction and overshoots, degree of limitation of ductions, forced duction testing (FDT) and extent of field of binocular single vision [10]. Purpose: to study the surgical outcomes of indicated cases of DRS diagnosed in Sohag University Hospital.

2. Patients and Methods

2.1. *Patients*

A prospective non-randomized interventional 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 about the planned procedure, prognosis, the nature and the aim of the study. Approval of the ethical committee of Sohag faculty of medicine is fulfilled. Preoperatice evaluation: History taking from all subjects including age, gender, family history, systemic diseases, previous medications or ophthalmic surgery. All patients are subjected to medical assessment to detect or exclude any systemic disorders. Detailed ophthalmological examination would be done including: uncorrected visual acuity, best corrected visual acuity, amblyopia detection, cycloplegic refraction, ocular motility examination in all directions, anterior and posterior segment examination. Postoperative examination and follow up: For detection of improvement in ocular alignment, globe retraction, over shooting, abnormal head posture, and follow up for 6 months. Patients are divided into three groups: group A: patients with DRS type I, group B: patients with DRS type II and group C: patients with DRS type III. Surgery is done to patients who had one or more of the following indications of surgery:

1) Strabismus in primary position.

2) Abnormal head posture.

3) Retraction of the globe on adduction (grade 2 or more).

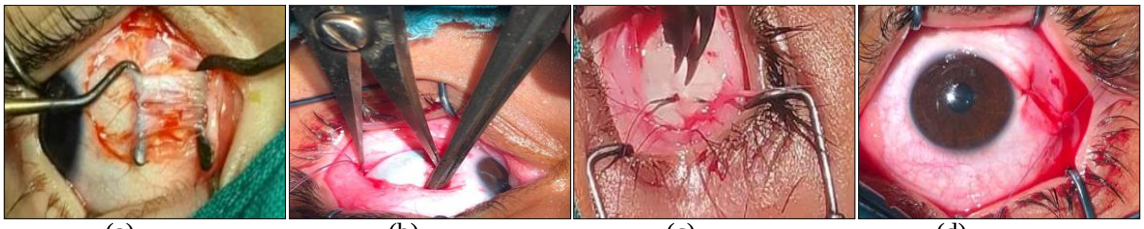
4) Upshoot or downshoot on attempted adduction (grade 2 or more).

Informed consent was taken from all patients before surgery, with special emphasis on the predictability of limited movement of the globe even after surgery. Consents included the agreement of photographing patients before and after surgery. All surgeries were done under general anesthesia.

Types of operations planned in this study:
***)** LR recession in one or both eyes.
***)** MR recession in one or both eyes.
***)** Simultaneous MR and LR recession.
***)** SR transposition.

2.2. *Surgical principles*

Povidone Iodine is used to sterilize the eye. Conjunctival incision: limbal incison is used, with relaxing incisions, then incising the tenon capsule. Catching the muscle by muscle hook. Meticulous muscle exposure and dissection by removing the intermuscular septum and check ligament. Using the double armed absorbable sutures (vicryl 6/0) to hold the muscle by one partial thickness and another full thickness suture to secure the muscle. Cutting the insertion of the muscle by sharp scissor, with avoidance of cutting the sutures. Then, measure the intended distance of recession and fixing the muscle to the new insertion site by partial thickness scleral sutures. Suturing the conjunctiva is done by inverted absorbable suture to avoid ocular irritation. Splitting of lateral rectus muscle if needed; in cases of over shooting is done after exposure of the muscle all the way to its entrance through Tenon's capsule, which is dissected as far back as possible. Then, LR is bisected into two halves. The upper half is hung on a suture, removed from its insertion, and sutured in sclera in the new site after recession, in the supero-temporal quadrant. The same maneuver is repeated with the lower half to be sutured in the inferotemporal quadrant. Superior rectus transposition in cases of eso-DRS with marked limitation of abduction, either alone or combined with MR recession. FDT is done before surgery to detect where the restriction to abduction starts. A limbal conjunctival incision is made between the superior and lateral rectus muscles and the SR is isolated. Then, SR is secured using double armed 6-0 vicryl sutured. Its attachments to the levator palpebrae superioris and the superior oblique (SO) tendon are observed by elevating the SR after dis-insertion and these are carefully dissected. This dis-inserted SR is then attached adjacent to the superior border of the LR along the spiral of Tillaux. Figures (2-11) show five patients before surgery with their full diagnosis and after surgery, with surgical decision



(a) (b) (c) (d)

Figure 1: Surgical steps; (a) Muscle hooking, (b) Measuring intended distance of recession by caliber in recession, (c) Partial thickness scleral suture in recession, (d) Conjunctival suturing by absorbable sutures

Figure 2: Patient No. 1: Female 16 years old pre-operative photos: Rt. DRS type I, Rt. Esotropia 20 PD

Figure 3: Patient No. 1: postoperative photos after Rt. MR recession 6 mm

Figure 4: Patient No. 2: male 10 years old preoperative photos: Lt. DRS type III, exotro-pia shooting in adduction, grade 1 globe retraction15 prism diopter in 1ry position, grade 1 over

Figure 5: Patient No. 2: postoperative photos after Lt. simultaneous MR recession 4 mm, LR recession 5 mm Residual exotropia 5 prism diopter in 1ry position

Figure 6: Patient No. 3: female 2 years old 30 prism diopter preoperative photos: Lt. DRS type II, exotropia

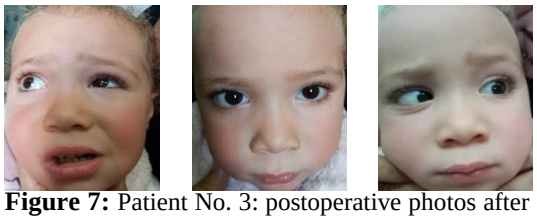


Figure 7: Patient No. 3: postoperative photos after bilateral LR recession 6 mm orthophoria in 1ry position

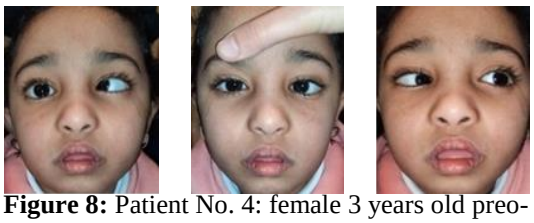


Figure 8: Patient No. 4: female 3 years old preoperative photos: Rt. DRS type I, Rt. Esotropia 25 prism diopter

Figure 9: Patient No. 4: postoperative photos after Rt. MR recession 4 mm, SR transposition to LR. Orthophoria in 1ry position

Figure 10: Patient No. 5: male 9 years preoperative photos: Lt. DRS type I, grade 4 old over shooting in adduction, grade 3 globe retraction in a mentally retarded child

Figure 11: Patient No. 5: postoperative photos after mm, with Y splitting, orthophoria in 1ry position Lt. simultaneous MR recession 3 mm, LR recession 6

3. Results

This study was conducted in Sohag University, 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). Twenty patients (57.1%) were indicated for surgery, while fifteen patients (42.9%) were not. Intervention was done to thirteen patients, and seven patients refused surgery due to unrealistic high expectation of postoperative

full ocular motility. 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 2: Postoperative results for cases with strabismus in primary position

Ocular deviation in 1ry position	Postoperative result
Exotropia in 6 cases	Orthophoria in 5 cases Residual exotropia (10 PD) in 1 case
Exotropia, with hypertropia in 1 case	Orthophoria
Esotropia in 4 cases	Orthophoria

Table 2: Postoperative results of Esotropic DRS

	Minimum	Maximum	Range	Mean	SD	P value
Preoperative Esotropia	20 PD	35 PD	15 PD	25 PD	7.07	<0.05
Postoperative result (of ET-duane)	Orthophoria 0 PD	5 PD	5 PD	1.25 PD	2.5	
Preoperative Exotropia (of XT-duane)	15 PD	35 PD	20 PD	22.86 PD	9.94	<0.05
Postoperative result	Orthophoria 0 PD	10 PD	10 PD	2.14 PD	3.93	

Table 3: Pre- and postoperative results of globe retraction, overshooting

	Preoperative	Postoperative	P value
Globe Retraction	2.0± 1.78	0.71±0.61	<0.05
Over Shooting	1.36± 1.78	0.23±0.44	<0.05

Table 4: Pre- and postoperative results of abnormal head posture

Abnormal head posture	Postoperative result
Face turn to the opposite side in four cases of exotropic DRS	Total improvement
Face turn to the same side in two cases of esotropic DRS	Total improvement

4. Discussion

Regarding management of esotropic DRS in this study, ipsilateral MR recession 4.5± 1.29mm was done. It was associated with LR recession in a case of severe globe retraction. Bilateral MR recession was done to larger angle deviation (35 PD). While SR transposition was added in one case, with marked abduction limitation. So, MR recession was the gold standard for treatment of esotropic Duane. Kekunnaya et al described that unilateral MR recession of the affected eye can correct up to 20 prism diopters (PD) of esotropia. They suggested keeping MR recession of the DRS eye to less than 6 mm. This is because the possibility of an iatrogenic adduction limitation is increased by a larger MR recession, by inducing an exotropia in the contralateral gaze, this impairs the field of binocular single visio [11]. In instances of esotropic DRS with moderate to severe co-contraction, simultaneous MR recession with LR recession has been documented. This lessens abnormal innervation of the LR during adduction [12]. In their study, all patients exhibited improved globe retraction after simultaneous MR recession 6± 0.5 mm, LR recession 8±1 mm. So, there is agreement with other studies that MR recession is the mainstay of treating eso-DRS. When globe retraction is marked, simultaneous MR, LR recession is done; and when the angle of deviation is large, bilateral MR recession is conducted. In managing exotropic DRS in this study, unilateral LR recession 7 mm was done in small angle of deviation; 15 PD. Bilateral LR recession was addressed in larger angles up to 35 PD. While, simultaneous MR recession and LR recession was used in a case with severe globe retraction. And Y-splitting was done in all cases with overshooting, with satisfactory results. Gaur et al emphasized that over shooting must be noticed first, as this will change the decision for surgery, with the need of Y

splitting or not. Such patients can be treated with LR recession with Y-split, a supramaximal LR recession or a periosteal fixation of LR. An in cases with normal LR activity during abduction,. When there is typical LR activity during abduction, and the exotropia and overshoots are the problems, LR recession with Y-split is the preferrable option [13]. A study done in 2019 by Mohamed Fared doing asymmetrical bilateral lateral rectus recession combined with augmented partial vertical rectus transposition (VRT) in the management of exotropic Duane retraction syndrome (XT-DRS) [14]. He demonstrated that exodeviation at far and near fixation were corrected by means of 26.4 PD and 24.8 PD respectively. Head turn was improved by a mean of 17.3°. Limited abduction and abnormal vertical movements were corrected by means of 1.6 and 1.5 units, respectively. So, studies of Gaur et al, Mohamed Fared agreed with this study that managing exo-DRS is done mainly by unilateral or bilateral LR recession, according to angle of deviation and Y-splitting is mandatory in over shooting. However, Gaur et al preferred combining LR recession with periosteal fixation in cases with large angle exotropia. Mohamed Fared added partial VRT to LR recession. Regarding cases of ortho-Duane, these patients are orthophoric in the primary position. These patients however present with retraction, palpebral aperture narrowing, and upshoots and/or downshoots on adduction, which may be cosmetically very disfiguring. In this study, all cases of ortho-Duane associated with globe retraction were managed by simultaneous MR, LR recession. Globe retraction grade improved from 2.0±1.78 to 0.71±0.61, which is statistically significant. Managing of over shooting by LR splitting improved its grade from 1.36±1.78 to 0.23±0.44. This agrees with Gaur et al results who described treating such patients by equal recession of both LR, MR. However, they recommended adjustable sutures on MR, and Y-split on LR to control the risk of undesirable under or over correction [13].

5. Conclusion

Duane retraction syndrome (DRS) is an eye movement disorder present at birth (congenital) characterized by horizontal eye movement limitation, with globe retraction. There are specific indications to be considered in managing DRS. Treatment should be tailored to each case according to the findings and degree of each of them. Preoperative consultation is a must about the non-correctable signs. There is marked postoperative improvement in the correctable signs, and it is advised to proceed in surgery in indicated cases.

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