Available online at www.sciencedirect.com

ScienceDirect

Journal of Current Ophthalmology 29 (2017) 248–257

http://www.journals.elsevier.com/journal-of-current-ophthalmology

Review

Surgical treatment of Duane retraction syndrome

Mohammad Reza Akbari [a,*], Vahideh Manouchehri [b], Arash Mirmohammadsadeghi [a]

[a] Eye Research Center, Farabi Eye Hospital, Tehran University of Medical Sciences, Tehran, Iran
[b] Department of Ophthalmology, Nikookari Eye Hospital, Tabriz University of Medical Sciences, Tabriz, Iran

Received 6 May 2017; revised 2 August 2017; accepted 10 August 2017
Available online 11 September 2017

Abstract

Purpose: Surgical treatment in Duane retraction syndrome (DRS) can be very challenging even for the strabismus specialists because of a wide spectrum of diversity in clinical manifestations. The purpose of this article is to review these different surgical treatments.
Methods: A comprehensive search was performed using PubMed database with the different keywords of "Duane retraction syndrome" and "surgery". Articles were selected from original English papers published since 2000. The full text of the selected articles was reviewed, and some articles were added based upon the references of the initial articles. We also provided selected case examples about some of these procedures.
Results: 125 articles were found in the initial search of which 37 articles were mostly related to the topic of this review. The number finally increased to 59 articles after considering the relative references of the initial articles. Different surgical methods performed on horizontal and vertical rectus muscles (recession, resection, transposition, Y splitting, periosteal fixation and posterior fixation suture) are reviewed. Careful selection of the surgical technique is important to achieve optimal results.
Conclusion: With accurate diagnosis of patients with DRS and proper surgical management, several adverse situations associated with this syndrome (amblyopia, abnormal head posture, upshoot, downshoot, and muscle underaction) can be prevented.

Keywords: Duane retraction syndrome; Surgery

Introduction

Duane retraction syndrome (DRS) is one of the congenital cranial dysinnervation disorders characterized by limitation of horizontal eye movements, globe retraction, and vertical movements in adduction.[1,2] The incidence of DRS is 1%–4% of strabismus patients.[3] This syndrome is caused by congenital hypoplasia or absence of sixth nerve and nuclei with aberrant innervations by branches from the oculomotor nerve resulting in co-contraction between medial and lateral rectus muscles on adduction.[4–7] Using electrophysiology tests, Huber classified DRS into three types, but because of variability in misinnervation and resultant clinical features, some clinicians have simplified Duane syndrome patients to either esotropic, exotropic, or orthotropic DRS. This simple classification is easier to use for surgical planning.[8,9]

Various surgical techniques have been described that strabismus surgeons should be aware of the potential benefits and limitations of these procedures. In this article, we review different surgical treatments for DRS with selected case examples for some of them.

Methods

A PubMed search was performed in November 2016 using each of the following key words: "Duane retraction syndrome" and "surgery". Articles were selected from original English papers published since 2000. All article types including original articles, reviews, case reports, and book chapters about

Institution at which the study was conducted: Farabi Eye Hospital.
Authors obtained consents from the patients for publishing the photos.

* Corresponding author. Farabi Eye Hospital, Eye Research Center, South Kargar, Qazvin SQ, Tehran, Iran.
E-mail address: mrakbari83@hotmail.com (M.R. Akbari).

Peer review under responsibility of the Iranian Society of Ophthalmology.

http://dx.doi.org/10.1016/j.joco.2017.08.008

surgery in DRS were identified. The full text of the selected articles was reviewed, and some articles were added based upon the references of the initial articles. We also provided selected case examples about some of these procedures.

Results

After reviewing English literature using PubMed database, 125 articles were found in the initial search of which 37 articles were mostly related to the topic of this review. The number finally increased to 59 articles after considering the relative references of the initial articles. The results were divided into five main sections of esotropic DRS, exotropic DRS, globe retraction and overshoots, bilateral DRS, and simultaneous abduction. Different surgical methods are categorized and summarized in Table 1.

Surgical approaches

Esotropic Duane retraction syndrome

If strabismus is present, esotropia occurs more frequently than exotropia in patients with DRS. Esotropia is usually less than 30 prism diopters (PD) in primary position.[10] It presents when the limitation in abduction is greater than the limitation in adduction and when the tonus of the lateral rectus muscle in primary position is less than that of the medial rectus muscle. An approach to the surgical treatment of esotropic DRS depends on the analysis of important anomalies observed in this group of patients such as primary position alignment, abnormal head posture, limitation of ductions, and severity of retraction and overshoots.[11] Patients with DRS can have high hyperopia and resulting accommodative esotropia, which is important to correct prior to surgery. Sometimes, spectacles alone can correct abnormal head posture in DRS.[12]

Case 1 (Fig. 1). The patient was a 4-year-old child with left eye esotropic DRS who presented severe left face turn, left eye esotropia, and limitation of abduction. His cycloplegic refraction was +7.00 in both eyes. After prescription of

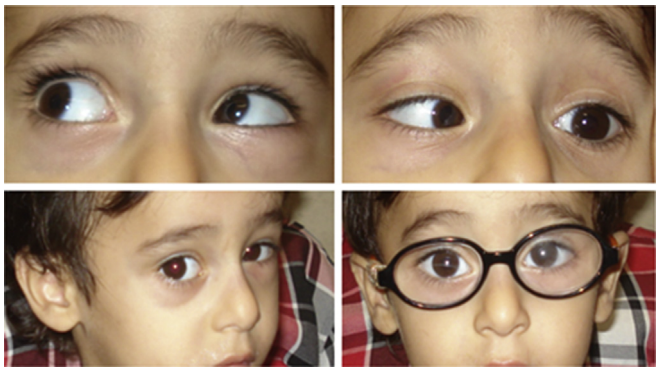

Fig. 1. *Case 1*. Photograph (upper row) shows motility in a patient with left eye esotropic Duane retraction syndrome (DRS) and hyperopic refractive error. Photograph (lower row) shows head posture before and after prescription of glasses in the same patient.

Table 1
Summary of different surgical approaches for Duane retraction syndrome (DRS).

Type	Intervention
Esotropic DRS	**Horizontal rectus surgery:** - Ipsilateral MR recession - Symmetric or asymmetric bilateral MR recession - Ipsilateral MR recession and contralateral MR posterior fixation suture - Ipsilateral MR recession and LR resection **Transposition surgery:** - Full tendon or half tendon VRT with or without fixation suture - SRT with and without MR recession
Exotropic DRS	- Ipsilateral LR recession - Bilateral lateral rectus recession - Periosteal fixation of the LR - Full tendon or half tendon nasal VRT with or without fixation suture
Overshoots & Globe retraction	**Mechanical type:** - Large LR recession - Periosteal fixation of the LR - Recession of the ipsilateral LR and MR - Posterior fixation suture of the ipsilateral LR alone or both the LR & MR - Splitting of the LR into a Y configuration **Innervational type:** - Recession of the appropriate vertical rectus muscle
Bilateral DRS	- Horizontal muscle recession - VRT
Simultaneous abduction	- Large LR recession - Periosteal fixation of the LR - MR resection

DRS: Duane retraction syndrome, MR: Medial rectus, LR: Lateral rectus, VRT: Vertical rectus transposition, SRT: Superior rectus transposition.

glasses, the eyes were orthotropic, and abnormal head posture was improved.

Usually, surgery is performed around age 3–8 years, as these patients have excellent fusion with abnormal head posture.[13] Evidence exists that facial musculoskeletal asymmetry may develop in the context of abnormal face turns.[14,15] Thus, surgery in younger age, as soon as one year old, could prevent the complications resulting from prolonged abnormal head posture.

Case 2 (Fig. 2). Parents of a 1-year-old girl had noted abnormal head posture and complete limitation of abduction in the left eye. She had left eye esotropic DRS with severe left face turn. She underwent early surgery, and left medial rectus was recessed 4 mm. After three years, she was orthotropic with no abnormal head posture. Her abduction was significantly improved.

Surgeries for management of primary position deviation and abnormal head position in esotropic DRS can be classified in three categories: (1) surgery on horizontal rectus muscles, including unilateral or bilateral medial rectus muscle recession, recession of the medical rectus muscle of the affected eye combined with Faden operation of the contralateral medial rectus muscle, unilateral medial rectus recession and lateral rectus resection; (2) surgery on vertical rectus muscles, including vertical rectus transposition (VRT) that can be full tendon or partial tendon and superior rectus transposition (SRT) only; and (3) combination of horizontal and vertical rectus surgery.

Horizontal rectus surgery

Medial rectus muscle recession was proposed by Duane in 1905 and has been used extensively with success for eliminating an abnormal head position and improving primary position deviation.[1] Medial rectus muscle recession of the eye with DRS should be part of the surgical plan if positive forced duction to abduction is present. Based on its simplicity, reliable correction of abnormal head posture and low rate of complications, single medial rectus muscle recession of the affected eye remains a reasonable and effective surgical treatment.[16] Unilateral recession of the medial rectus muscle is recommended for deviations less than 20 PD.[11,17] The amount of recession depends on the preoperative measurements in primary position, the degree of restriction on forced duction testing, and the limitations noted on versions and ductions.[17] Medial rectus recession of the DRS eye should not be more than 5–6 mm because of inducing adduction limitation.[18] When adduction is limited to a significant degree, the exotropia in contralateral gaze can cause disabling diplopia. Large medial rectus recession may improve abduction mildly but often leads to even more severe limitation of adduction, which compromises the field of binocular single vision and increases the risk of consecutive exotropia, synergistic divergence and difficulty in convergence. Moreover, once the single medial rectus is recessed, there may be a gradual recurrence of the deviation and head posture due to lack of abduction tone.[19]

Bilateral medial rectus recession has been recommended in unilateral DRS with more than 20 PD esotropia.[11–17] In some patients with severe globe retraction in which surgical plan is ipsilateral medial and lateral rectus recessions, medial rectus of unaffected eye can be recessed for correction of residual deviation. Also, in patients who have limited adduction and very slow adduction velocities in the affected eye, recession of medial rectus of contralateral eye can be done for decreasing the amount of ipsilateral medial rectus recession.[11] Jampolsky proposed asymmetric recession of the medial rectus of both eyes with more on the unaffected eye. This large medial rectus recession in unaffected eye results in more innervation of the normal eye's medial rectus and less innervation of normal eye's lateral rectus according to Sherrington's law and less innervation of the affected eye's medial rectus, according to Hering's law. This “fixation duress” prevents contracture of medial rectus in affected eye.[17] The approach remains controversial. Several authors stated that symmetric or asymmetric bilateral medial rectus recession has resulted in improvement of deviation, abnormal head position, and globe retraction in patients with unilateral esotropic DRS.[20,21]

Nevertheless, Greenberg and Pollard reported four patients with small-angle esotropic DRS that medial rectus recession of their normal eyes decreased the positive effects of recessing the Duane's medial rectus muscle on abnormal head posture and increased the risk of consecutive exotropia. They concluded that adding a simultaneous normal eye medial rectus muscle recession produces a bilateral gaze weakness in

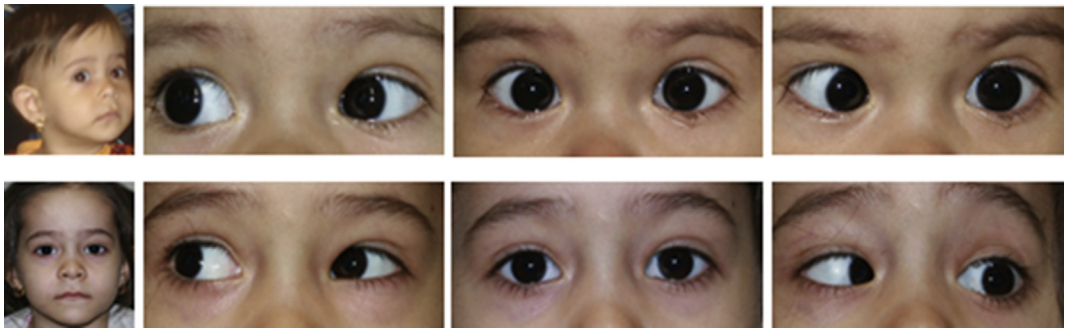

Fig. 2. *Case 2*. Preoperative (upper row) and postoperative (lower row) head posture and motility in a patient with left esotropic Duane retraction syndrome (DRS). She was orthotropic without abnormal head posture after 4 mm medial rectus recession that was performed when she was one year old.

side of affected eye, thus easing any tendency for unaffected side-gaze preference and increasing any residual face turn.[22]

Medial rectus muscle recession of the affected eye and placement of contralateral medial rectus posterior fixation suture has been advocated on the basis of matching duction limitations between two eyes that allow a wider field of diplopia-free field and improve abduction in the affected eye.[23,24] Experience with this method is inadequate and has seen various results. Nevertheless, Hering's law may not apply in DRS because of abnormally innervated lateral rectus in this syndrome.[16]

Traditionally, resection of the lateral rectus muscle of the affected eye in patients with esotropic DRS and limited adduction is not suggested owing to the risk of severely limiting adduction and worsening globe retraction on adduction. Kraft compared unilateral recession and resection with bilateral medial rectus recession for esotropic DRS and showed that small lateral rectus resection can be a safe and effective component of surgery. It has a low risk of worsening retraction or limiting adduction when performed in appropriate cases, including: 1) patients with esotropia of at least 25 PD; 2) mild retraction on adduction; 3) clinically normal adduction; 4) significantly limited abduction; 5) no or mild up/down shoots. Surgery involved medial rectus recession up to 5.0 mm and lateral rectus resection of maximum 3.5 mm. This approach led to clinically significant improvement in abduction.[25]

There are limited experiences about botulinum toxin injection in DRS. In most patients, the results of injection have been relative and short-term. The most important role of injection is that it can be used to simulate the effect of surgery. However, the permanent effects of botulinum after repeated injections have been reported.[26–28]

The patients with amblyopia in the fellow eye prefer to fixate with the DRS eye despite an imbalance of muscle forces, and with the mechanism of fixation duress, large deviations are created in non-DRS eye. Surgery should be performed first in the fixing DRS eye for balancing the forces in the primary position that cause improvement of face turns.[17]

Case 3 (Fig. 3). A 16-year-old male was referred for the left eye esodeviation and right face turn. His best corrected visual acuity was 10/10 in the right eye with cycloplegic refraction of +1.00 and 5/10 in the left eye with cycloplegic refraction of +4.00. In examination, he had limitation in right eye's abduction and adduction and globe retraction in adduction. This means the patient was using right DRS eye as fixator eye. With prism on the left eye, the amount of esotropia was 40 PD (secondary deviation). Primary deviation was 25 PD. First, surgery was performed on the right DRS eye, and the medial rectus was recessed 5 mm. After surgery, his face turn was improved, but he had 20 PD residual esotropia in the left eye. In the second surgery, medial rectus of the left eye was recessed 6.5 mm. One year after second surgery, he was orthotropic with no face turn.

Transposition surgery

In 1974, for the first time, Gobin used the vertical rectus muscle transposition with a recession of the medial rectus of the affected eye for managing esotropic DRS.[29] The procedure was further popularized for use by Molarte and Rosenbaum, but without medial rectus recession in primary surgery to avoid the possible complication of anterior segment ischemia.[19] However, nearly fifty percent of their patients required a secondary procedure involving recession of the ipsilateral medial rectus to achieve acceptable eye alignment and head position. Foster reported that the addition of lateral fixation sutures to full VRT improved the resulting tonic abducting force for patients with lateral rectus (LR) palsy or type 1 DRS even in the presence of medial rectus (MR) restriction.[30] The major advantage of augmented VRT surgery was a significant reduction in the need to weaken the ipsilateral MR muscle, as is required in most other transposition procedures. Foster warned that ipsilateral medial rectus recession should be avoided in the primary transposition procedure and even as a secondary procedure because of the risk of decreased adduction and the possibility of late overcorrection. Botulinum toxin injection of the ipsilateral medial rectus muscle or a contralateral medial rectus muscle recession done secondarily was suggested for notable residual deviation even after this augmented transposition procedure.[30] The other advantages of augmented VRT compared with ipsilateral medial rectus recession include increased abduction ability and diplopia-free visual field and decreased risk of creating limitation to adduction.[31–33] On the other hand, it has been noted that VRT may lead to worsening of upshoots and

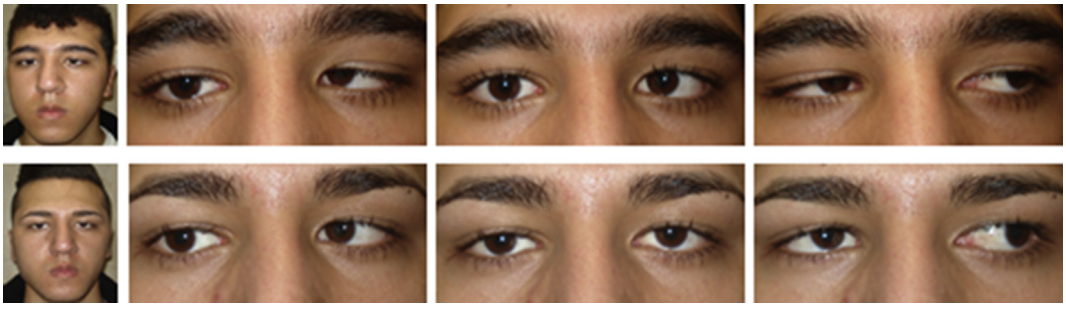

Fig. 3. *Case 3*. Preoperative (upper row) and postoperative (lower row) alignment in a patient with right fixator Duane eye and resulting secondary esodeviation in left eye. After asymmetric medial rectus recession in two stage surgery, he was orthotropic in primary position.

downshoots, breaking fusional ability because of induced vertical deviations and increasing retraction on adduction.[16]

The main risk factor for residual esotropia after VRT is restriction of the medial rectus muscle. If forced duction test (FDT) demonstrates significant restriction to abduction, the medial rectus muscle should be recessed. Otherwise, findings such as migration of the muscle insertion toward its original insertion, muscle recession or slippage might be seen in exploration that needs revision. Risk factor for overcorrection and resulting exotropia in patients treated with VRT is large exotropia or less esotropia in adduction preoperatively. Overcorrection is treated according FDT to adduction. If the FDT is negative, management includes ipsilateral medial rectus advancement. When FDT is positive, repositioning of the transposed muscles or maximal ipsilateral lateral rectus recession should be done.[33]

Currently, the newest technique for transposition surgery includes only SRT temporally that was proposed by Johnston and colleagues in 2006.[34] SRT allows for the option of simultaneous recession of the medial rectus in patients with tight medial rectus who need transposition surgery. This technique may induce some intorsion; however, the risk for symptomatic torsional diplopia seems to be very low. With SRT, a portion of the muscle force vector directed vertically is displaced laterally. But this loss of vertical force does not cause significant vertical deviation because it is also advanced slightly, which compensates the reduced vertical force.[35–37] If esotropia is less than 15 PD, SRT alone may be adequate. For larger deviation SRT should be combined with the medial rectus recession. The results are better when adjustable medial rectus recession is performed along with SRT. This method seems to be more effective than medial rectus recession alone in improving abduction.[36]

Case 4 (Fig. 4). A 6-year-old girl presented with left DRS, esotropia of 20 PD in primary position, left face turn, and complete limitation of abduction. At surgery, she had moderate positive forced duction testing for abduction of the left eye. The left medial rectus was recessed 4 mm, and the superior rectus was transposed temporally with augmentation suture. Four months after surgery, she was orthotropic in primary position with no abnormal head posture. Abduction was significantly improved.

Exotropic Duane retraction syndrome

Exotropic DRS is a relatively rare form of DRS that happens when the power of the lateral rectus, through innervation or lateral rectus contracture, is greater than the corresponding medial rectus force. The abduction can be normal or restricted.[38] In motility examination, the exotropic eye position could be mistaken for true lateral rectus function. Commonly, there is severe lateral rectus co-contracture with tight lateral rectus that causing severe globe retraction and overshoots. The patients usually have a compensatory head turn away from the affected eye to obtain fusion.[17]

Indications for surgery are the same as esotropic DRS. Here, a selection of the type of surgery is straightforward, and the lateral rectus weakening should be done, anyway. The classical treatment is an ipsilateral lateral rectus recession for deviations less than 20 diopters and bilateral lateral rectus recession for deviations more than 20 diopters with the surgical dosages more than standard surgical tables.[11–17] Recession of tight and shortened lateral rectus can correct the deviation and head turn and simultaneously diminishes globe retraction and overshoots in the cost of increased abduction limitation.[17] Profound weakening technique such as periosteal fixation of the lateral rectus muscle may be needed if the globe retraction is severe.[39] This procedure can be done in combination with VRT toward the lateral rectus muscle.[40] The other approach for the management of these patients with severe retraction consists of simultaneous recession of the lateral and medial rectus muscle. In orthotropic DRS, the lateral rectus muscle recession should be about 1–2 mm more than the medial rectus muscle recession.[41] Therefore, in patients with exotropia in primary position, the larger amount of lateral rectus should be recessed if recession of both medial and lateral rectus is planned. In patients with mild globe retraction and residual exotropia, after lateral rectus recession, VRT toward the medial rectus muscle could be performed.

Case 5 (Fig. 5). The patient was a 22-year-old male with left eye exotropic DRS. He had severe right face turn, 77 PD exotropia of the left eye and limitation of adduction with mild down shoot in adduction. Intraoperatively, forced duction test for the lateral rectus was positive. The left lateral rectus was recessed 10 mm, and two third of the superior rectus and the

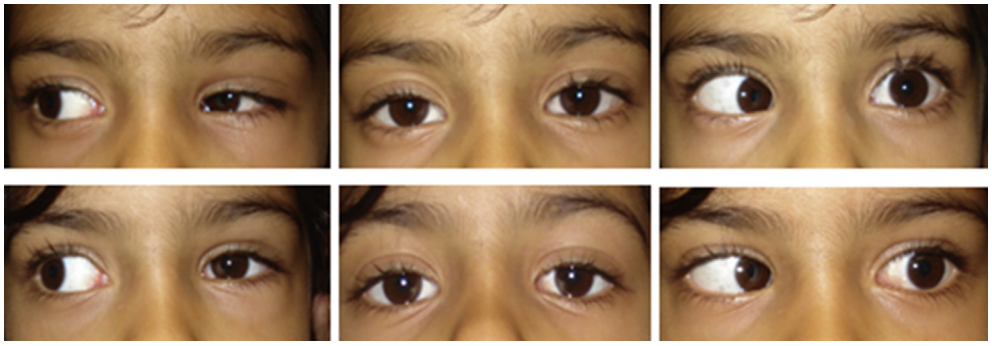

Fig. 4. *Case 4*. Preoperative (upper row) and postoperative (lower row) alignment and motility in a patient with left esotropic Duane retraction syndrome (DRS). She was orthotropic after 4 mm medial rectus recession and superior rectus transposition (SRT).

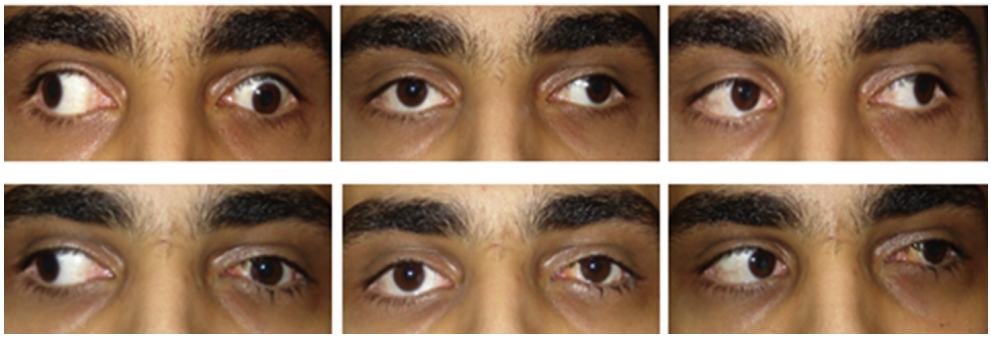

Fig. 5. *Case 5*. Preoperative (upper row) and postoperative (lower row) alignment in a patient with left exotropic Duane retraction syndrome (DRS). He had good alignment in primary gaze following left lateral rectus recession and transposition two third of the superior rectus and the inferior rectus muscles medially.

inferior rectus muscles were transposed medially with augmentation suture after 4 mm resection of both muscles. Three months after surgery he had 8 PD exotropia.

Anomalous orbital structures that attach to the globe and restrict motility may be found in some cases of DRS. These structures could limit the motility and cause severe globe retraction even after recession of the lateral rectus. The forced duction testing were not freed until these bands were carefully dissected out and released.[42,43]

Globe retraction and overshoots

Abnormal lateral rectus innervation in DRS leading to co-contraction with the medial rectus on attempted adduction. This co-contracture along with tight lateral rectus create globe retraction in DRS.[10]

About 25%–39% of patients with DRS have over elevation or over depression in adduction.[44] The mechanism of these vertical movements can be mechanical, innervational, or both.[11]

In the mechanical type, overshoot occurs abruptly as the eye moves into adduction. Sometimes, milder form of overshoot occurs when the eye moves above or below the horizontal plane in adduction. The mechanical type of upshoot and downshoot is due to tight lateral rectus attached to the crest of the globe, which causes the muscle to slip off the globe when the eye is adducting and termed the "bridle effect" or "knife-edge" overshoots.[10,11,17] This overshoots may look cosmetically very disfiguring even in patients with orthotropic DRS. Any procedure that would remove the tight lateral rectus from the crest of the globe or stabilize it on the globe would eliminate the mechanical overshoots, including: large lateral rectus recession, lateral rectus periosteal fixation, recession of the ipsilateral lateral and medial rectus muscles, posterior fixation suture of the ipsilateral lateral rectus muscle alone or both the lateral and medial rectus muscles and splitting of the lateral rectus tendon into a Y configuration. All of these procedures except posterior fixation suture can improve globe retraction, too. Both retraction and overshoots improve when lateral rectus recession and Y splitting done simultaneously.[11,35,38,45–50]

Case 6 (Fig. 6). A 25-year-old woman presented with left orthotropic DRS without any abnormal head posture and with severe globe retraction in adduction. To treat her globe retraction, the medial rectus muscle was recessed 5 mm and the lateral rectus was recessed 7 mm. Postoperatively, she was orthotropic in primary position, and her globe retraction was significantly improved.

Case 7 (Fig. 7). A 19-year-old woman presented with a left exotropic DRS, severe upshoot and mild downshoot in adduction. She had an XT of 10 PD at far and 18 PD at near and a right face turn. To correct her XT, face turn and upshoot, the left lateral rectus was recessed 7 mm, and Y-splitting was accomplished.

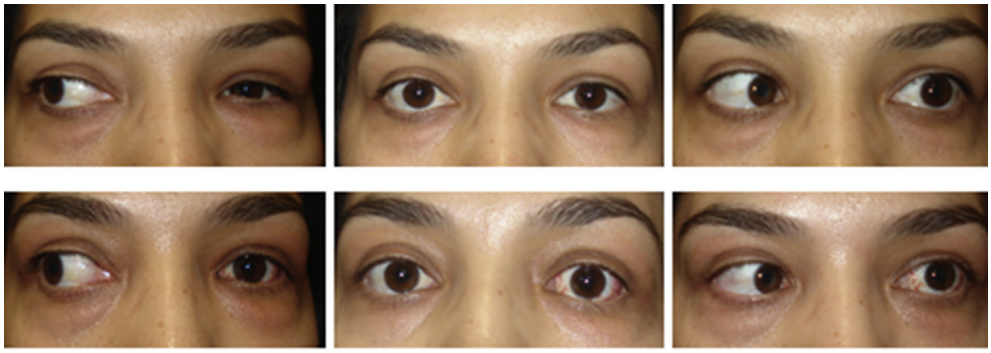

Fig. 6. *Case 6*. Preoperative (upper row) and postoperative (lower row) alignment and motility in a patient with severe globe retraction and orthotropic Duane retraction syndrome (DRS) showing good alignment in primary gaze and reduction in the globe retraction following medial and lateral rectus recession of the left eye.

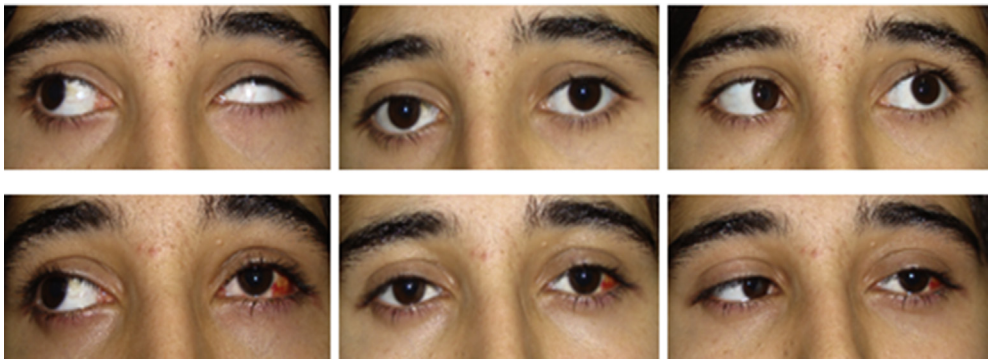

Fig. 7. *Case 7*. Preoperative (upper row) and postoperative (lower row) alignment and motility in a patient with severe upshoot in adduction. Upshoot decreased significantly after recession and Y-splitting of the lateral rectus.

After surgery, she had no abnormal head posture and upshoot, and downshoot were improved considerably.

In the innervational type, there is a gradually increasing overshoots in the horizontal position as it moves into adduction. This is possibly due to co-innervation of the vertical or oblique muscles with the lateral rectus muscle. Vertical deviation in the primary position is not commonly seen in the mechanical type and if it exists, is usually small angle. In contrast, there is typically a large vertical deviation in the primary position in patients with innervational overshoots.[11] The mechanical upshoot and downshoot can occur simultaneously in same patient, but in the innervational type only one of the upshoot or downshoot can occur. The innervational type can be improved with recession of the appropriate vertical rectus muscle.[51]

Bilateral Duane retraction syndrome

The incidence of bilateral DRS is reported to range from 10% to 24%. Recent reports support the different characteristics in terms of sex distribution, horizontal strabismus and abnormal head posture prevalence, amblyopia, positive strabismus family history of strabismus, and associated congenital abnormalities in patients with bilateral DRS. Esotropia is the most common deviation, and some patients may exhibit an alphabet pattern.[52,53] Bilateral DRS can be associated with fusion. Such cases usually have little or no deviation in primary position, limited eye movements, and relatively immobile globes. Surgery can be unpredictable in these patients, and prisms are better treatment if needed. Head position is determined by the resting position of the dominant eye. The aim of surgery in these patients is to correct the abnormal head posture by balancing medial and lateral rectus muscle function in the fixating eye. Patients without fusion have noticeable deviation and abnormal head posture. The disease can be very asymmetric with different types of DRS in two eyes.[17]

Case 8 (Fig. 8). Parents of a 4-year-old girl had noted bilateral limitation of eye movements from childhood. In first examination, visual acuity of both eyes were 8/10, and cycloplegic refraction were +1.00. There was 8 PD esotropia in primary position, globe retraction of both eyes in adduction

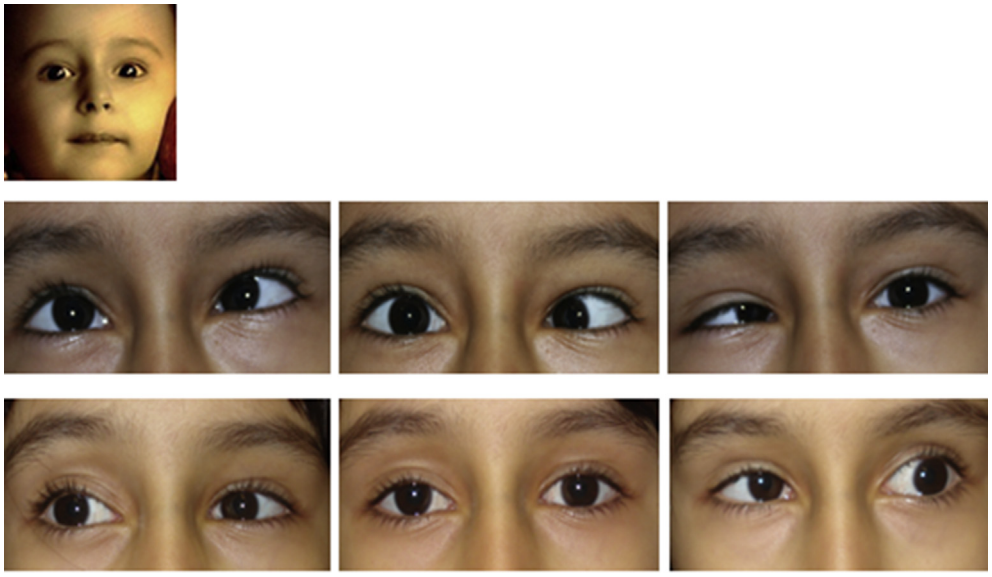

Fig. 8. *Case 8*. Photograph (upper row) shows near orthotropia position of eyes in bilateral Duane retraction syndrome (DRS) when patient was 4 years old. Preoperative (middle row) and postoperative (lower row) alignment of same patient after two years with decompensated bilateral esotropic DRS who underwent bilateral medial rectus recession. She was orthotropic in primary position and abduction significantly improved.

and complete bilateral limitation of abduction without any abnormal head posture. She had 80 s of arc stereopsis by titmus stereo test. Surgery was not planned for her because of small deviation and good fusion. During two years follow-up, esotropia increased gradually, and fusion was disrupted. When she became 6 years old, she had 30 PD esotropia. Bilateral medial rectus recession of 5.5 mm was performed. Six months after surgery, she was orthotropic in primary position. So, bilateral DRS with fusion may decompensate with increasing deviation.

Limited literature exists on the bilateral DRS treatment because of lower incidence and variability in presentation. Horizontal muscle recession is proven to improve abnormal head posture and correct strabismus in primary position. Bilateral medial rectus muscle recession can be done in patients with bilateral esotropic DRS with good results.[54] Transposition of the vertical rectus muscles to the lateral rectus muscle, with or without recession of the medial rectus muscle in patients with bilateral esotropic DRS have been reported.[55] Unilateral or bilateral lateral rectus recession in bilateral DRS associated with exotropia have good outcomes.[56]

Case 9 (Fig. 9). A 13-year-old boy presented with limitation of adduction and abduction in both eyes since childhood compatible with bilateral DRS. Visual acuity was 10/10 in both eyes. He had left face turn when he fixed with the right eye, and he had right face turn when the left eye was the fixator. For deviation measurement, we had to use two prisms on both eyes simultaneously because he had severe globe retraction in primary position and could not have foveal fixation with each of eyes during measurement. With this method he had 25 + 25 PD exodeviation. Intraoperatively, forced duction testing for both lateral rectus muscles was severely positive. He underwent bilateral lateral rectus recession of 8.5 mm. Two months after surgery, he had no face turn and was orthotropic in primary position.

Simultaneous abduction

"Simultaneous abduction", "synergistic divergence", the "splits" and "type 4 DRS" are synonyms for the condition that characterized by outward rotation of Duane eye as the normal fixing eye goes to abduction. Patients usually have exotropia, head turn opposite the involved eye, and absent or decreased adduction of involved eye. Weakness of the medial rectus is responsible for this type of DRS. It can occur congenitally due to misrouting of third nerve to the lateral rectus, causing the medial rectus to be deprived of its usual amount of innervation. Simultaneous abduction has been reported in patients who have undergone large medial rectus recession for esotropic DRS.[17] The surgical procedure of choice for this condition is weakening of the ipsilateral lateral rectus muscle along with medial rectus resection.[57–59]

Fig. 9. *Case 9*. Preoperative (upper row) and postoperative (lower row) alignment and motility of a patient with bilateral exotropic Duane retraction syndrome (DRS) showing good alignment in primary gaze and reduction of globe retraction following bilateral lateral rectus recession.

Fig. 10. *Case 10*. Preoperative (upper row) and postoperative (lower row) alignment and head posture of a patient with the left eye synergist divergence type Duane retraction syndrome (DRS). After anchoring of lateral rectus to the lateral orbital wall and medial rectus resection, she had good alignment in primary position and elimination of simultaneous abduction. In left gaze, she had no abduction.

Medial rectus resection can be performed because there is no globe retraction.

Case 10 (Fig. 10). A 4-year-old girl had a severe right face turn, left eye's exotropia and simultaneous abduction of the left eye in right gaze. Intraoperatively, forced duction testing for lateral rectus muscle was positive. The left lateral rectus muscle was disinserted and sutured to the lateral orbital wall periosteum, and the left medial rectus was resected 6 mm. Postoperatively, she had no abnormal head posture, and eyes were orthotropic in primary position with no simultaneous abduction of the left eye.

Discussion

Currently older surgical techniques including horizontal rectus recession are effectively used for different manifestations of DRS. VRT surgeries were popularized because they could improve ductions. In the recent years, novel techniques such as SRT and orbital fixation of the lateral rectus have been introduced with good acceptance. With accurate diagnosis of patients with DRS and proper surgical management, we can overcome several adverse situations with this syndrome, i.e. amblyopia, abnormal head posture, upshoot, downshoot, and muscle underaction.

References

1. Duane A. Congenital deficiency of abduction, associated with impairment of adduction, retraction movement, contraction of the palpebral fissure and oblique movement of the eye. 1905. *Arch Ophthalmol*. 1996;114(10):1255–1256. discussion 1257.
2. Oystreck DT, Engle EC, Bosley TM. Recent progress in understanding congenital cranial dysinnervation disorders. *J Neuro-Ophthalmol*. 2011;31(1):69–77.
3. De Respinis PA, Caputo AR, Wagner RS, Guo S. Duane's retraction syndrome. *Surv Ophthalmol*. 1993;38(3):257–288.
4. Matteucci P. I difetti congeniti di abduzione con particolare riguardo alla patogenesi. *Rassegna Ital Ottalmol*. 1946;15:345–380.
5. Hotchkiss MG, Miller NR, Clark AW, Green WR. Bilateral Duane's retraction syndrome. A clinical-pathologic case report. *Arch Ophthalmol*. 1980;98(5):870–874.
6. Miller NR, Kiel SM, Green WR, Clark AW. Unilateral Duane's retraction syndrome (type 1). *Arch Ophthalmol*. 1982;100(9):1468–1472.
7. Breinin GM. Electromyography; a tool in ocular and neurologic diagnosis. II. Muscle palsies. *AMA Arch Ophthalmol*. 1957;57(2):165–175.
8. Huber A. Electrophysiology of the retraction syndromes. *Br J Ophthalmol*. 1974;58(3):293–300.
9. Ahluwalia BK, Gupta NC, Goel SR, Khurana AK. Study of Duane's retraction syndrome. *Acta Ophthalmol (Copenh)*. 1988;66(6):728–730.
10. Von Noorden GK. *Binocular Vision and Ocular Motility*. 4th ed. St Louis, MO: Mosby; 1990:399.
11. Kraft SP. A surgical approach for Duane syndrome. *J Pediatr Ophthalmol Strabismus*. 1988;25(3):119–130.
12. Kekunnaya R, Velez FG, Pineles SL. Outcomes in patients with esotropicduane retraction syndrome and a partially accommodative component. *Indian J Ophthalmol*. 2013;61(12):701–704.
13. Wright KW. Pediatric Ophthalmology and Strabismus. In: Wright KW, Spiegel PH, eds. 2nd ed. New York: Springer-Verlag Inc; 2003:267.
14. Greenberg MF, Pollard ZF. Ocular plagiocephaly: ocular torticollis with skull and facial asymmetry. *Ophthalmology*. 2000;107(1):173–179.
15. Akbari MR, Khorraminejad M, Askarizadeh F, Pour FF, Ranjbar Pazooki M, Moeinitabar MR. Facial asymmetry in ocular torticollis. *J Curr Ophthalmol*. 2015;27(1-2):4–11.
16. Barbe ME, Scott WE, Kutschke PJ. A simplified approach to the treatment of Duane's syndrome. *Br J Ophthalmol*. 2004;88(1):131–138.
17. Jampolsky A. Duane syndrome. In: Rosenbaum AL, Santiago AP, eds. *Clinical Strabismus Management*. Philadelphia: WB Saunders; 1999:325–346.
18. Shiratori A, Kameyama C, Sibasaki K. Adduction deficiency following a large medial rectus recession in Duane's retraction syndrome type 1. *J Pediatr Ophthalmol Strabismus*. 1999;36(2):98–100.
19. Molarte AB, Rosenbaum AL. Vertical rectus muscle transposition surgery for Duane's syndrome. *J Pediatr Ophthalmol Strabismus*. 1990;27(4):171–177.
20. Farvardin M, Rad AH, Ashrafzadeh A. Results of bilateral medial rectus muscle recession in unilateral esotropic Duane syndrome. *J AAPOS*. 2009;13(4):339–342.
21. Dotan G, Klein A, Ela-Dalman N, Shulman S, Stolovitch C. The efficacy of asymmetric bilateral medial rectus muscle recession surgery in unilateral, esotropic, type 1 Duane syndrome. *J AAPOS*. 2012;16(6):543–547.
22. Greenberg MF, Pollard ZF. Poor results after recession of both medial rectus muscles in unilateral small angle Duane's syndrome type 1. *J AAPOS*. 2003;7(2):142–145.
23. Saunders RA, Wilson ME, Bluestein EC, Sinatra RB. Surgery on the normal eye in Duane retraction syndrome. *J Pediatr Ophthalmol Strabismus*. 1994;31(3):162–169.
24. Puerto-Hernandez B, Lopez-Caballero C, Rodriguez-Sanchez JM, González-Manrique M, Contreras I. Surgical treatment of Duane's syndrome type 1 by recession of the affected eye and faden operation of the contralateral medial rectus. *Arch Soc Esp Oftalmol*. 2008;83(2):113–116.
25. Kraft SP. Resecting the lateral rectus in unilateral Duane syndrome with esotropia and limited abduction. *Binocul Vis Strabismus Q*. 2010;25(3):149–157.
26. Dawson ELM, Maino A, Lee JP. Diagnostic use of botulinum toxin in patients with Duane syndrome. *Strabismus*. 2010;18(1):21–23.
27. Maya JF, Liano RG, Catalan MRG, Rayward O. Botulinum toxin treatment in patients up to 3 years of age who have esotropic Duane retraction syndrome. *Strabismus*. 2013;21(1):4–7.
28. Ameri A, Farzbod F, Bazvand F, et al. Botulinum toxin injection in the patients with Duane syndrome type. *J Curr Ophthalmol*. 2016;29(1):50–53.
29. Gobin MH. Surgical management of Duane's syndrome. *Br J Ophthalmol*. 1974;58(3):301–306.
30. Foster RS. Vertical muscle transposition augmented with lateral fixation. *J AAPOS*. 1997;1(1):20–30.
31. Yazdian Z, Rajabi MT, Yazdian MA, Rajabi MB, Akbari MR. Vertical rectus muscle transposition for correcting abduction deficiency in Duane's syndrome type 1 and sixth nerve palsy. *J Pediatr Ophthalmol Strabismus*. 2010;47(2):96–100.
32. Velez FG, Foster RS, Rosenbaum AL. Vertical rectus muscle augmented transposition in Duane syndrome. *J AAPOS*. 2001;5(2):105–113.
33. Rosenbaum AL. Costenbader Lecture. The efficacy of rectus muscle transposition surgery in esotropic Duane syndrome and VI nerve palsy. *J AAPOS*. 2004;8(5):409–419.
34. Johnston SC, Crouch Jr ER, Crouch ER. An innovative approach to transposition surgery is effective in treatment of Duane's syndrome with esotropia [ARVO abstract 2475-B403]. *Invest Ophthalmol Vis Sci*. 2006;47. eAbstract 2475.
35. Mehendale RA, Dagi LR, Wu C, Ledoux D, Johnston S, Hunter DG. Superior rectus transposition and medial rectus recession for Duane syndrome and sixth nerve palsy. *Arch Ophthalmol*. 2012;130:195–201.
36. Yang S, MacKinnon S, Dagi LR, Hunter DG. Superior rectus transposition vs medial rectus recession for treatment of esotropic Duane syndrome. *JAMA Ophthalmol*. 2014;132(6):669–675.
37. Velez FG, Oltra E, Isenberg SJ, Pineles SL. Assessment of torsion after superior rectus transposition with or without medial rectus recession for Duane syndrome and abducens nerve palsy. *J AAPOS*. 2014;18(5):457–460.
38. Velez FG, Velez G, Hendler K, Pineles SL. Isolated Y-splitting and recession of the lateral rectus muscle in patients with Exo-Duane syndrome. *Strabismus*. 2012;20(3):109–114.

39. Velez FG, Thacker N, Britt MT, Alcorn D, Foster RS, Rosenbaum AL. Rectus muscle orbital wall fixation: a reversible profound weakening procedure. *J AAPOS*. 2004;8(5):473–480.
40. Sharma P, Tomer R, Menon V, Saxena R, Sharma A. Evaluation of periosteal fixation of lateral rectus and partial VRT for cases of exotropic Duane retraction syndrome. *Indian J Ophthalmol*. 2014;62(2):204–208.
41. Sprunger DT. Recession of both horizontal rectus muscles in Duane syndrome with globe retraction in primary position. *J AAPOS*. 1997;1(1):31–33.
42. Lueder GT. Anomalous orbital structures resulting in unusual strabismus. *Surv Ophthalmol*. 2002;47(1):27–35.
43. Pineles SL, Velez FG. Accessory fibrotic lateral rectus muscles in exotropic Duane syndrome with severe retraction and upshot. *J AAPOS*. 2015;19(6):549–550.
44. Isenberg S, Urist MJ. Clinical observation in 101 consecutive patients with Duane retraction syndrome. *Am J Ophthalmol*. 1977;84(3):419–425.
45. Von Noorden GK. Recession of both horizontal recti muscles in Duane's retraction syndrome with elevation and depression of the adducted eye. *Am J Ophthalmol*. 1992;114(3):311–313.
46. Jampolsky A, Eisenbaum AM, Parks MM. *A Study of Various Surgical Approaches to the Leash Effect in Duane's Syndrome*. Presented at the American Academy of Ophthalmology, Chicago, Nov 2–7. 1980.
47. Von Noorden GK, Murray E. Up- and downshoot in Duane's retraction syndrome. *J Pediatr Ophthalmol Strabismus*. 1986;23(5):212–215.
48. Scott AB. Upshoots and downshoots. In: Souza-Diaz C, ed. *Smith-Kettlewell Symposium on Basic Sciences in Strabismus, Annex to Congresode1 ConsejoLatinoamericano de Estrabismo (C.L.A.D.E.), p 60*. Sao Paolo: Oficinas das Edicoes Loyola; 1978.
49. Rao VB, Helveston EM, Sahare P. Treatment of upshoot and downshoot in Duane syndrome by recession and Y-splitting of the lateral rectus muscle. *J AAPOS*. 2003;7(6):389–395.
50. Sukhija J, Kaur S, Singh U. Isolated lateral rectus recession with Y splitting versus anchoring of the lateral rectus muscle in patients with exotropic Duane syndrome. *J AAPOS*. 2014;18(2):147–150.
51. Mohan K, Saroha V. Vertical rectus recession for the innervational upshoot and downshoot in Duane's retraction syndrome. *J Pediatr Ophthalmol Strabismus*. 2002;39(2):94–99.
52. Khan AO, Oystreck D. Clinical characteristics of bilateral Duane syndrome. *J AAPOS*. 2006;10(3):198–201.
53. Mohan K, Sharma A, Pandav SS. Differences in epidemiological and clinical characteristics between various types of Duane retraction syndrome in 331 patients. *J AAPOS*. 2008;12(6):576–580.
54. Sachdeva V, Kekunnaya R, Gupta A, Bhoompally VR. Surgical management of bilateral esotropic Duane syndrome. *J AAPOS*. 2012;16(5):445–448.
55. Britt MT, Velez FG, Velez G, Rosenbaum AL. Vertical rectus muscle transposition for bilateral Duane syndrome. *J AAPOS*. 2005;9(5):416–421.
56. Theodorou N, Burke J. Surgical and functional outcomes in bilateral exotropic Duane's retraction syndrome. *Br J Ophthalmol*. 2013;97(9):1134–1137.
57. Hamed L, Lingua R, Fanous M, Saunders TG, Lusby FW. Synergystic divergence: saccadic velocity analysis and surgical results. *J Pediatr Ophthalmol Strabismus*. 1992;29(1):30–37.
58. Khawam E, Terro A, Hamadeh I. Surgical correction of synergistic divergence strabismus. A report of three cases. *Binocul Vis Strabismus Q*. 2007;22(4):227–234.
59. Schliesser JA, Sprunger DT, Helveston EM. Type 4 Duane syndrome. *J AAPOS*. 2016;20(4):301–304.