Review Article - Narrative Review

Taiwan J Ophthalmol 2023;13: 489-499

Access this article online

Quick Response Code:

Website: http://journals.lww.com/TJOP

DOI: 10.4103/tjo.TJO-D-23-00078

# Duane syndrome: An overview on the current management

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**Abstract:**

Duane syndrome is one of the most common restrictive congenital strabismus characterized by variable horizontal duction deficits with globe retraction and shoots on attempted adduction and narrowing of the palpebral aperture. It is now listed as a congenital cranial dysinnervation disorder. The disease is usually unilateral with female preponderance. Basic etiopathogenesis involves dysinnervation of the lateral rectus (LR) due to aplastic/hypoplastic abducens nucleus with a secondary aberrant supply to the LR by the medial rectus (MR) subnucleus of the oculomotor nerve. Diagnosis of the disease is usually clinical. Due to the variable presentation of the disease, surgical management is a challenge and has to be individualized to achieve alignment in the primary gaze, reduction in globe retraction, upshoots and downshoots, and correction of any abnormal head posture. Differential recessions of the lateral and MR muscles are done to correct esotropia or exotropia in the primary gaze. For globe retraction and shoots, Y-split or periosteal fixation of the LR muscles is done depending on the severity.

**Keywords:**

Duane syndrome, globe retraction, restrictive strabismus, strabismus surgery, Y-splitting

## Introduction

Duane syndrome (DS), also known as Stilling-Turk-DS, is a complex form of strabismus characterized by limited horizontal ductions, narrowing palpebral aperture, and possible upshoots/downshoots on attempted abduction/adduction.[1,2] The syndrome with all its features was first described in detail by Duane in 1905 through a series of 54 cases, with possible etiopathogenesis proposed to be mis-innervation and co-contraction.[3] According to the new Classification of Eye Movement Abnormalities and Strabismus classification, DS has been renamed as co-contraction retraction syndrome. With advances in imaging and a better understanding of the disease pathophysiology, it is now considered a form of congenital cranial dysinnervation disorder, a term introduced by Gutowski and Ellard in 2003. It describes a number of congenital restrictive nonprogressive strabismus conditions, caused by impaired innervation of the extraocular muscles as a consequence to dysgenesis of the cranial nerve nuclei at the level of the brainstem and not due to primary fibrosis of the extraocular muscles.[4-6]

## Epidemiology

DS is one of the most common restrictive strabismus encountered in clinical practices among all special strabismus with documented prevalence among the strabismic population to be 1%–4%. The disease has a female preponderance (60%). Unilateral presentation is common (80% of cases) with the left eye more commonly affected. Bilateral DS is seen in 10% of cases. Patients with bilateral disease may be asymmetrically affected. The majority of cases of DS are sporadic in occurrence. The prevalence of familial cases has been

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Submission: 10-06-2023
Accepted: 26-08-2023
Published: 20-12-2023



**How to cite this article:** Prasad P, Saxena A, Saxena R. Duane syndrome: An overview on the current management. Taiwan J Ophthalmol 2023;13:489-99.

reported to vary from 10% to 23%.[1-3,7] Although the study of the autosomal dominant form of DS has identified CHN1 as a DS gene, autosomal recessive forms of inheritance have also been documented. Mutations in the CHN1 gene alter the development of abducens, and to some extent, the oculomotor axons, and are believed to be the cause of dysinnervation associated with the disease.[8,9] Ocular[10-12] and syndromic[13-15] associations of the disease have been summarized in Tables 1, 2 and Figure 1a and b.

## Etiopathogenesis

The disease is thought to result from an insult to the developing embryo during 4–8 weeks of gestation leading to aplasia or hypoplasia of the sixth nerve nucleus, which is the primary dysinnervation.[6] This is accompanied by an aberrant supply to the lateral rectus (LR) by the medial rectus (MR) subnucleus of the third nerve, which is the secondary dysinnervation. This leads to co-contraction of both the MR and LR during adduction and, therefore, retraction of the globe on attempted adduction, which is the first classical feature of this complex strabismus.[16-18] A case of Duane may exhibit a spectrum of abnormal innervational patterns ranging from moderate-to-severe subnormal innervation of the LR to varying paradoxical contraction on adduction. As a corollary, less than normal fibers will innervate the MR muscle, and hence, there will be essentially normal, subnormal, or markedly reduced MR innervations and strength. Over time, the noninnervated fibers of the MR become fibrotic, and as they contract, they lead to a tight MR. Secondary fibrosis of the medial and superior/inferior recti is a possible cause of persistent esotropia and upshoot/downshoot, respectively, due to slippage of the globe over the tight muscle on attempted adduction, which forms the second subset of the characteristic feature of the disease.[19,20] Duane's syndrome has also been associated with fetal alcohol syndrome, in cases of vascular hypofunction during embryogenesis and in patients with thalidomide syndrome.[21]

**Table 1: Ocular associations of Duane retraction syndrome**

| Anterior segment | Posterior segment |
|---|---|
| Ptosis | Coloboma |
| Nystagmus | Optic nerve hypoplasia |
| Epibulbar dermoid | Morning glory disc |
| Microcornea | Myelinated nerve |
| Keratoconus | Staphyloma |
| Coloboma | Situs inversus disc |
| Cataract | Persistent fetal vasculature |

**Table 2: Syndromic associations of Duane retraction syndrome**

| Syndromes | Associated features in addition to DS |
|---|---|
| Okihiro syndrome | Radial ray defects |
| Wildervanck syndrome | Klippel–Feil anomaly and deafness |
| Moebius | Congenital paresis of facial and abducens cranial nerve |
| Holt–Oram syndrome | Abnormalities of the upper limbs and heart |
| Morning glory syndrome | Abnormalities of the optic disc |
| Goldenhar syndrome | Malformation of the jaw, cheek, and ear, usually on one side of the face |

DS=Duane syndrome

## Clinical Features

Diagnosis of DS is mainly clinical. The clinical features of DS are variable depending on the amount of normal as well as anomalous innervations received by the LR muscle. A child with DS may present with anomalous head posture, affected eye appearing smaller due to globe retraction, and pseudoptosis in adduction. Ocular upshoot or downshoot may be noted in adduction. Refractive error and amblyopia can be associated, and hence, cycloplegic refraction is mandatory in all patients with DS [Figure 2a and b].

Limitation in abduction is the most notable and one of the most consistent features in almost all cases of DS. The primary deviation is usually less compared to the extent of abduction limitation. Anomalous head posture is in the form of a face turn and is a motor adaptation so as to maintain binocular single vision. These patients have good stereopsis and usually do not complain of

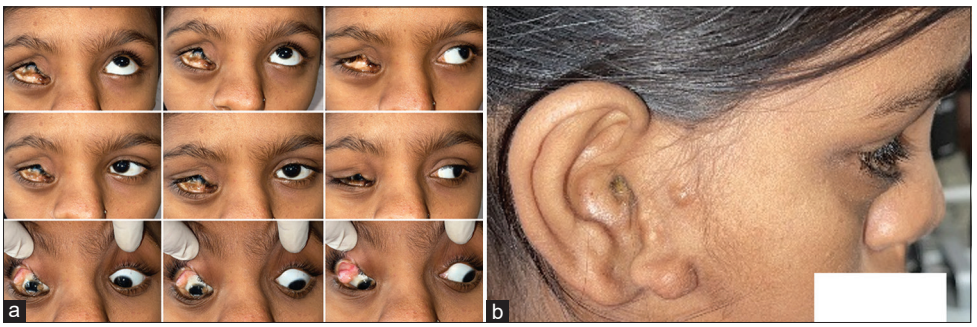


**Figure 1:** (a) Right eye exotropia–Duane syndrome with associated Goldenhar syndrome. (b) Note lid coloboma, limbal and forniceal lipodermoid, and preauricular skin tags

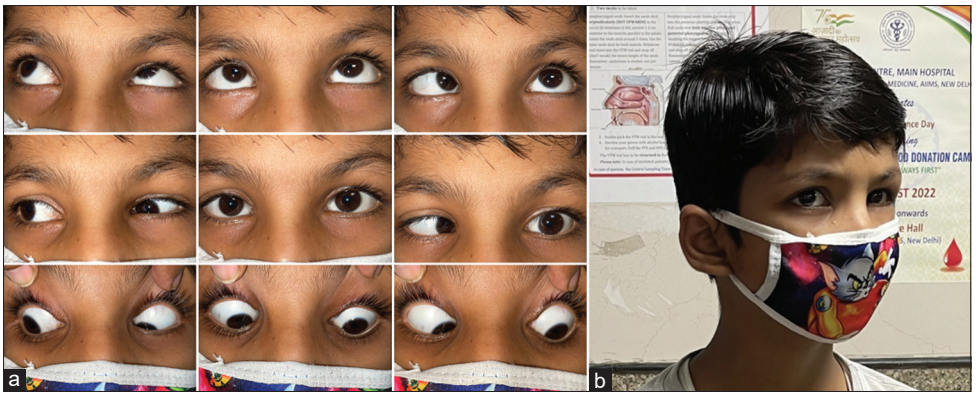

**Figure 2:** (a) Left eye esotropic Duane syndrome with shoots and globe retraction. (b) Note the left face turn

diplopia. These features help to differentiate it from the 6th nerve palsy.[22,23] Secondary muscle changes in the form of fibrosis can occur due to long-standing eye position. Usually, the shoots in adduction are due to the bridle effect of the co-contracting LR; however, rarely, there may be associated oblique muscle overaction.[19,20] In case of the presence of shoots, the eye will show upshoot when the eye is adducted being above the midline and downshoot if it is below the midline. The oblique muscle overaction is unlikely to be the cause of this ocular movement as is more gradual compared to the sudden vertical deviation seen in this form of strabismus due to shoots. Sometimes, an X-pattern can also be seen due to the shoots. Fundus torsion in adduction may hint toward a possible oblique overaction giving a clinical A- or V-pattern.[24]

## Classification of Duane Retraction Syndrome

Several classification systems have been proposed for DS –

- Papst classification[25,26] – Abnormal co-contraction of
  1. MR and LR
  2. SR and LR
  3. IR and LR
  4. LR and other muscles.
- Lyle and Bridgeman classification[27]
  - A. Abduction is more deficient than adduction. Adduction causes globe retraction and pseudoptosis
  - B. Abduction is deficient, but adduction is not
  - C. Adduction is more deficient than abduction. Adduction causes globe retraction and pseudoptosis.
- Malbran classification[28]
  - I. Abduction palsy
  - II. Adduction palsy
  - III. Elevation and depression limitation. Horizontal duction normal.
- Huber's classification[29] – Based on the above with electromyography (EMG) findings,
  - I. (70%–80%) Marked limitation or complete absence of abduction with widening of the palpebral fissure on attempted abduction; normal or only slightly defective adduction; narrowing of the palpebral fissure; and retraction on adduction. This is associated with a moderate amount of esotropia (~30 prism diopters). EMG recordings show paradoxical innervations of the LR with maximum impulses on adduction and defective impulses in attempted abduction, the normal electrical behavior of the MR [Figure 3]
  - II. (7%) Limitation or absence of adduction with exotropia of the affected eye; normal or slightly limited abduction; narrowing of the palpebral fissure; and retraction of the globe on attempted adduction. EMG recordings show peak impulses in the LR on abduction and a second paradoxical impulse on attempted adduction, the normal electrical behavior of the MR [Figure 4]
  - III. (15%) Combination of limitation or absence of both abduction and adduction; retraction of the globe; and narrowing of the palpebral fissure on attempted adduction. EMG recordings show simultaneous innervation of the MR and LR muscles in the primary gaze, adduction, and abduction. Alphabet patterns have also been described with documented synergistic innervation of the vertical recti on EMG [Figure 5].

There are certain other forms of strabismus that similarly violate Sherrington's law of reciprocal innervation. Synergistic divergence/simultaneous abduction: synergistic divergence is a rare type of Duane syndrome with abduction on attempted adduction.[30] This involves agenesis of the abducens nucleus, and the uninnervated LR muscle paradoxically receives almost

all the innervation from the MR nerve. The DS eye turns outward as the normal fixing eye moves in the opposite gaze (abduction). Acquired DS may follow trauma with medial orbital wall fracture (inverse Duane), myositis or metastasis, or any other infiltration. Duane *et al*. have described this as pseudo-DS.[31-38]

## Grading for Globe Retraction

With the DS eye in maximum adduction, a scale is used at the center of the palpebral fissure to measure the height and compare it with the fellow eye in abduction [Table 3 and Figure 6].[39]

## Grading for Overshoots

With the involved eye in adduction, a straight line parallel to the intermedial canthal line is drawn from the pupillary center of the fellow eye [Table 4].[39]

## Surgical Management

Surgery in DS is sought for the correction of head posture, significant deviation in the primary gaze, severe upshoots/downshoots, and globe retraction/narrowing of the palpebral fissure. Surgical options vary according to the amount of the normal LR function, abnormal LR function (co-contraction/retraction), presence of shoots, deviation in the primary gaze (ortho/eso/exo), compensatory head posture, and force duction test of the

**Table 3: Grading of globe retraction for Duane syndrome**

| Grades | Palpebral fissure change |
|---|---|
| Grade 0 | No narrowing |
| Grade 1 | <25% |
| Grade 2 | 25%–<50% |
| Grade 3 | 50%–<75% |
| Grade 4 | ≥75% |

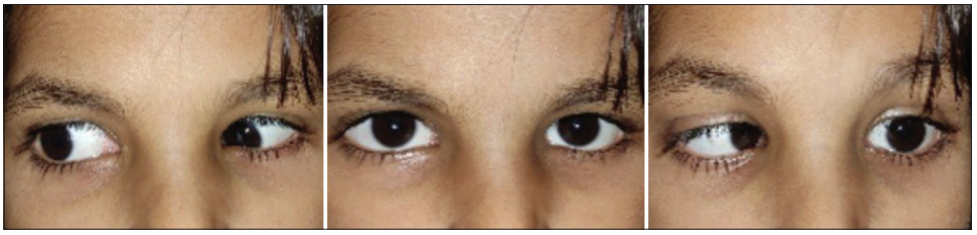

**Figure 3:** Left esotropic Duane syndrome

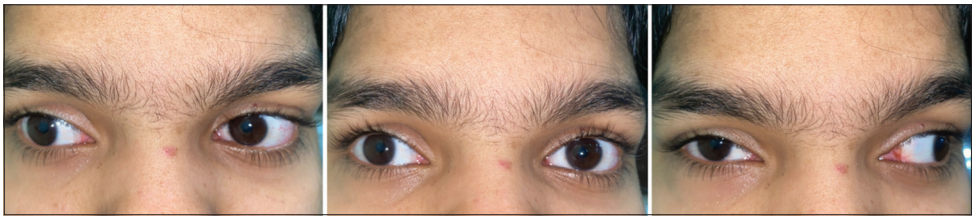

**Figure 4:** Right exotropia–Duane syndrome

**Figure 5:** Left orthotropic Duane syndrome

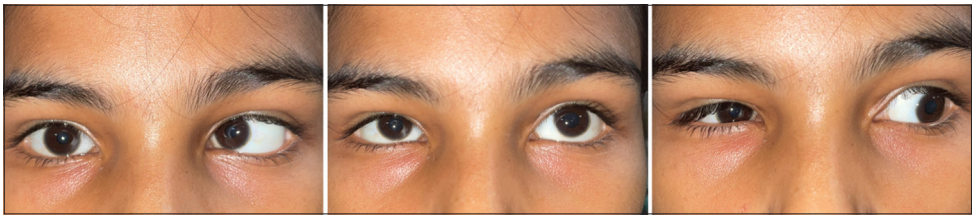

**Figure 6:** Right esotropic Duane syndrome with Grade 3 globe retraction

recti. It should be understood that even after successful surgery, the normal ductions and versions cannot be achieved; upshoots, downshoots, and enophthalmos can be greatly reduced, but not completely eliminated; fusing patient will continue to find areas of diplopia after treatment. This is because the primary anomaly cannot be practically corrected. The aim of the surgery is just to provide binocular single vision in the primary gaze with cosmetically acceptable reduced shoots/enophthalmos.

## Management of esotropic Duane Syndrome

### *Unilateral or bilateral medial rectus recession*

Recessing the MR is an effective surgery for esotropic DS (Eso-DS) as the LR almost always has some tone due to either some residual normal innervation from the VI nerve nucleus or aberrant abnormal innervation from the MR subnucleus of the III nerve. Long-standing Eso-Duane subjects are bound to have a tight MR; therefore, weakening it alone should suffice to hold the eye in the primary position.[40-44] However, it must be kept in mind that the MR is already deficient in innervation due to its fibers being misdirected to the LR. Hence, weakening MR further with recession will almost always cause an adduction deficit which will be proportional to the amount of recession. Further, in cases of severe co-contraction, recessing MR alone/a very large MR recession (MRc) can make the anomalously contracting LR win the tug-of-war on attempted adduction and can result in simultaneous abduction/splits/synergistic divergence. Therefore, in such a condition, both the MR and LR need to be recessed. In the presence of large esotropia in the primary gaze, asymmetric recession of bilateral MR is done.[44] LR resection is not a preferred option, especially when anomalous innervation is present, because it will worsen globe retraction.[45] MRc of the other eye can help improve the esotropia in the primary gaze, but does not improve the abduction deficit, as the LR is not innervated by the VI nerve. Ipsilateral MR should not be excessively recessed as that will exacerbate the adduction deficit as mentioned above. The least amount of MRc to free the forced duction test (FDT) in Eso-DS is recommended [Figures 7a, b and 8a, b].

**Table 4: Grading of upshoot/downshoot for Duane syndrome**

| Grades | Shooting eye position relative to intermedial canthal line |
|---|---|
| Grade 0 | Line bisects the pupil of the involved eye |
| Grade 1 | Line lies between the pupillary center and the pupillary margin |
| Grade 2 | Line lies between the pupillary margin and the limbus |
| Grade 3 | Line lies at the limbus or over the sclera |
| Grade 4 | The cornea disappears below the lid (pumpkin seed sign) |

### *Transposition of the vertical recti*

In case of no severe LR innervational anomaly (retraction/shoots), transposition of SR and IR to the LR (vertical rectus transposition [VRT]) or only SR to the superior border of the LR (superior rectus transposition [SRT]) is a viable option, to create at least some alternate abducting force. The VRT/SRT has been shown to improve abduction besides the correction of primary deviation.[46-48] Single VRT to LR has not been shown to cause significant vertical deviation, and when coupled with a small amount of MRc can correct moderate-to-large esotropia with an increase in abduction. However, these transposition procedures will paradoxically worsen retraction/shoots, where LR dysinnervational abduction is itself the culprit. Hence, in the presence of co-contraction or shoots, the LR also has to be recessed [Figure 9a and b].

### *Summarizing preferred surgical options for various scenarios of esotropic Duane Syndrome*

1. Unilateral, small esotropia in one position without co-contraction: ipsilateral MRc/VRT/SRT
2. Unilateral, small esotropia in one position with co-contraction: ipsilateral MRc + LR recession
3. Unilateral, large esotropia in one position without co-contraction: B/l MRc (asymmetric)/MRc + SRT/partial tendon VRT (p-VRT)
4. Unilateral, large esotropia in one position with


**Figure 7:** Preoperative (a) and postoperative (b) horizontal gaze photographs of left esotropic Duane syndrome. Surgery done – Left medial rectus recession of 5 mm

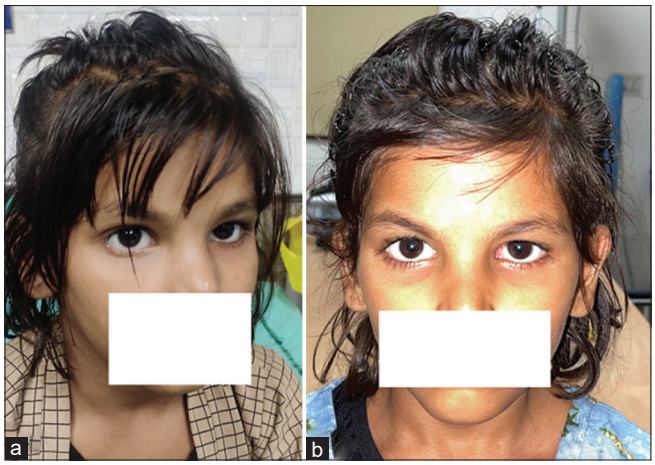


**Figure 8:** Preoperative (a) left face turn of the same subject corrected after surgery (b)

co-contraction/shoots: B/l MRc (asymmetric) + ipsilateral LR recession + Y-split

5. Bilateral Eso-DS: B/L MRc ± SRT/p-VRT [Figure 10].

## Management of exotropic Duane Syndrome

Exotropic DS has a head posture away from the affected side. The surgical goal is to correct the deviation, improve the head posture, reduce the anomalous movement, improve ocular rotations, and enlarge the binocular field of vision [Figure 11a and b].

Surgical options include large recession of the LR, vertical rectus muscle transposition, and orbital wall fixation of the LR muscle.[17,49-51] These procedures primarily correct the exodeviation in the primary position and, thus, the head posture. Since the aberrant innervational force is

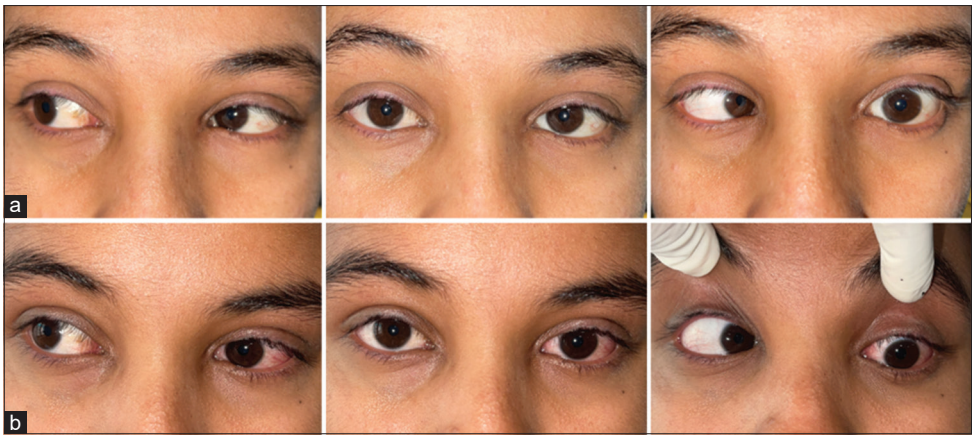


**Figure 9:** Preoperative (a) and postoperative (b) photographs of left esotropic Duane syndrome. Surgery done – Left medial rectus recession 4 mm and superior rectus transposition. Note the alignment in the primary gaze with improvement in left abduction

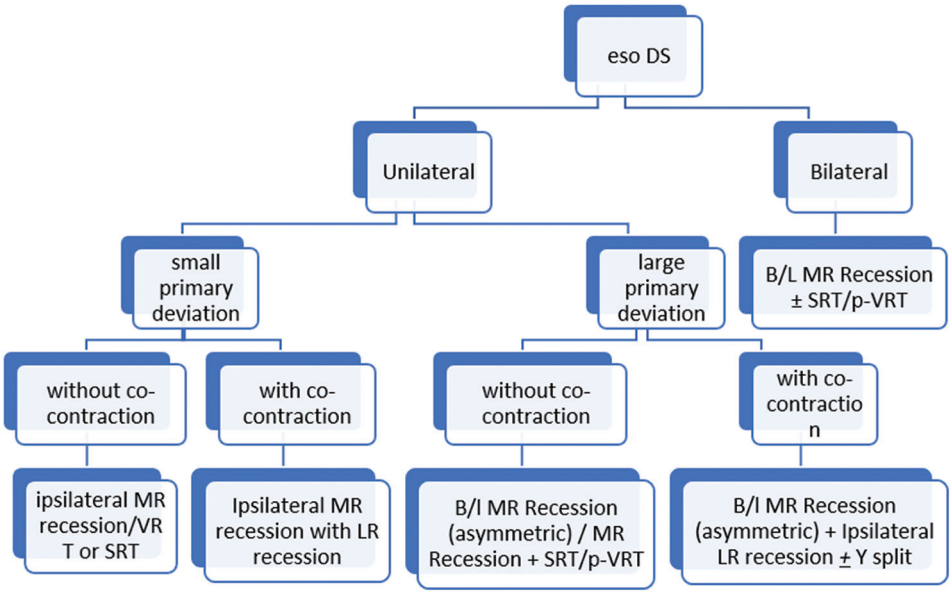


**Figure 10:** Management of esotropic Duane syndrome. DS: Duane syndrome, MR: Medial rectus, VRT: Vertical rectus transposition, SRT: Superior rectus transposition, LR: Lateral rectus, pVRT: Partial tendon vertical rectus transposition, Eso-DS: Esotropic DS

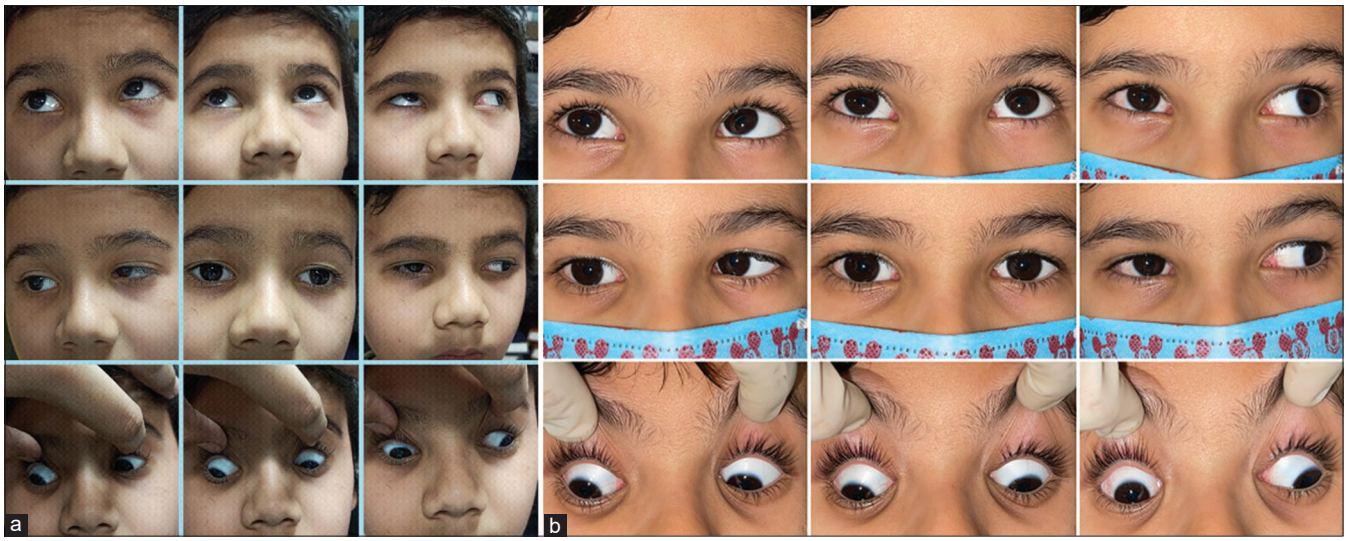


**Figure 11:** (a) Preoperative photographs of right exotropia–Duane syndrome with upshoots. (b) Postoperative photographs of the same subject. Surgery done – Right lateral rectus recession 9 mm with Y-split. Note alignment in the primary gaze with a marked reduction in shoots

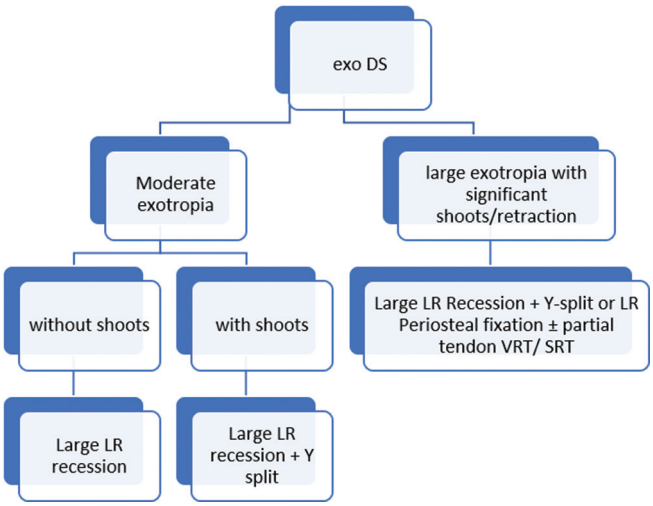


**Figure 12:** Management of exotropia–Duane syndrome. LR: Lateral rectus, VRT: Vertical rectus transposition, SRT: Superior rectus transposition, DS: Duane syndrome, Exo DS: Exotropia DS

not being tackled and will remain residual even after the surgery, there is often persistence or recurrence of the effects of anomalous LR. A complete inactivation of LR muscle force by orbital wall fixation, however, can eliminate all residual aberrant functions such as globe retraction, slippage, upshoots, and downshoots, and also addresses exotropia at the same time.

### *Indications of periosteal fixation*

Complete functional inactivation of the LR, although seldom required, is beneficial in those with severe LR contraction on attempted adduction.[52-54]

### *Advantages and limitations*

Periosteal fixation is a profound weakening procedure and results in a permanent disinsertion of the muscle from the globe and prevents any residual function. Suturing the rectus muscle to the orbital wall also reduces the risk of globe perforation compared to technically challenging maximal recessions. Another advantage is the possibility to reverse the procedure or convert it into a different weakening procedure. Potential complications of this procedure may include injury to the lacrimal gland, eyelid position changes, periocular inflammation, and bleeding. It may be technically difficult to expose the adjacent periosteum. There is a complete loss of movement in the gaze of muscle action. Some authors have noted a slight improvement in abduction over time, which could probably happen due to force transmitted via posterior connections of the LR through Tenon tissues. Consecutive esotropia is occasionally noted in patients with small preoperative exotropia.

### *Summarizing preferred surgical options for various scenarios of exotropia–Duane Retraction Syndrome*

1. Moderate exotropia: large LR recession
2. Moderate exotropia with shoots: large LR recession + Y-split
3. Large exotropia with significant upshoot/retraction: large LR Recession + Y-split or LR Periosteal fixation ± p-VRT/SRT [Figure 12].

## Management of orthotropic Duane Syndrome

No intervention is required if the eyes are aligned in the primary gaze without any significant co-contraction [Figure 13a and b].

Associated overshoots can be tackled by the recession of both MR (adjustable) and LR (with Y-split). Some amount of recession in the LR is necessary with the Y-split as the Y-splitting procedure tightens the muscle to some extent. Associated co-contraction retraction can


**Figure 13:** (a) Bilateral orthotropic Duane syndrome. (b) Horizontal gaze photographs of the same subject. No intervention was done in view of no face turn/shoots/retraction

be treated by asymmetric MR and LR recession in the affected eye. The same is a viable option for small eso/exo with severe retraction [Figure 14].

## Management of synergistic divergence

Synergistic divergence is characterized by abrupt abduction on attempted adduction; hence, both the eyes abduct on gaze away from Duane's eye. Treatment can be very difficult; however, LR transposition to the MR may be beneficial.[55]

## Management of upshoots/downshoots

Upshoots/downshoots are clinical manifestations of globe slippage over an inflexible anomalously innervated LR, and thus, its recession alone can be helpful in mild cases. LR recession with bifurcation or Y-split is helpful for severe upshoots and/or downshoots. Recession amounts have to be larger as splitting, and suturing the muscle itself tightens it requiring an additional recession amount. Another option for managing shoots is the LR posterior fixation suture (described by Scott). However, it is less effective than a Y-split, and the associated retraction is not corrected unless the LR muscle is recessed simultaneously [Figure 15a and b].

### *Bifurcation of the lateral rectus (Y-split) with or without recession*

The splitting and spreading of the muscle into two halves stabilize the globe and prevent it from slipping up or down in the contralateral gaze and, at the same time, reduce the retraction in the primary position. The muscle is split into two parts from its insertion as far posterior as possible, and each half is reinserted superior and inferior to the original insertion with around 20 mm of distance between them. The Y-split may or may not be combined with recession; although if coexisting exotropia is present or retraction is severe, it may be advisable to recess each half by 5–10 mm.[56-58]

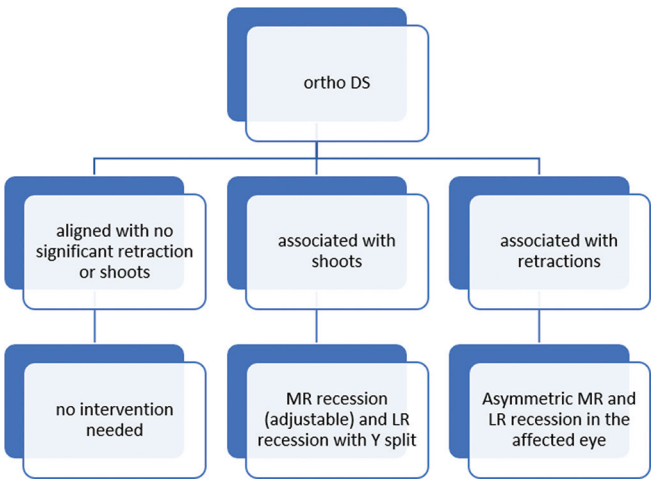


**Figure 14:** Management of orthotropic Duane syndrome. DS: Duane syndrome, Ortho DS: Orthotropic DS, MR: Medial rectus, LR: Lateral rectus

## Management of bilateral Duane Syndrome

In patients with bilateral DS with fusion (binocularity), the eyes may be aligned in a primary position with a straight head with or without retraction. In these cases, all four horizontal recti need to be recessed depending on the deviation in the primary position and the presence of shoot/retraction. In cases where there is no binocularity, the patient either has marked eso or exo deviation in the primary position. These cases are treated as unilateral DS, and balancing of MR and LR forces is done depending on the deviation governed by the dominant eye.[59]

It is important to remember that the results of surgery for DS can be disappointing.

It is wise not to opt for surgery if clear indications do not exist. One should avoid surgery in cases with binocular vision in the primary gaze or with a slight head posture. Furthermore, preoperatively, the patient should be made aware that complete and normal ocular eye movements in all gazes will not be possible with any surgery.

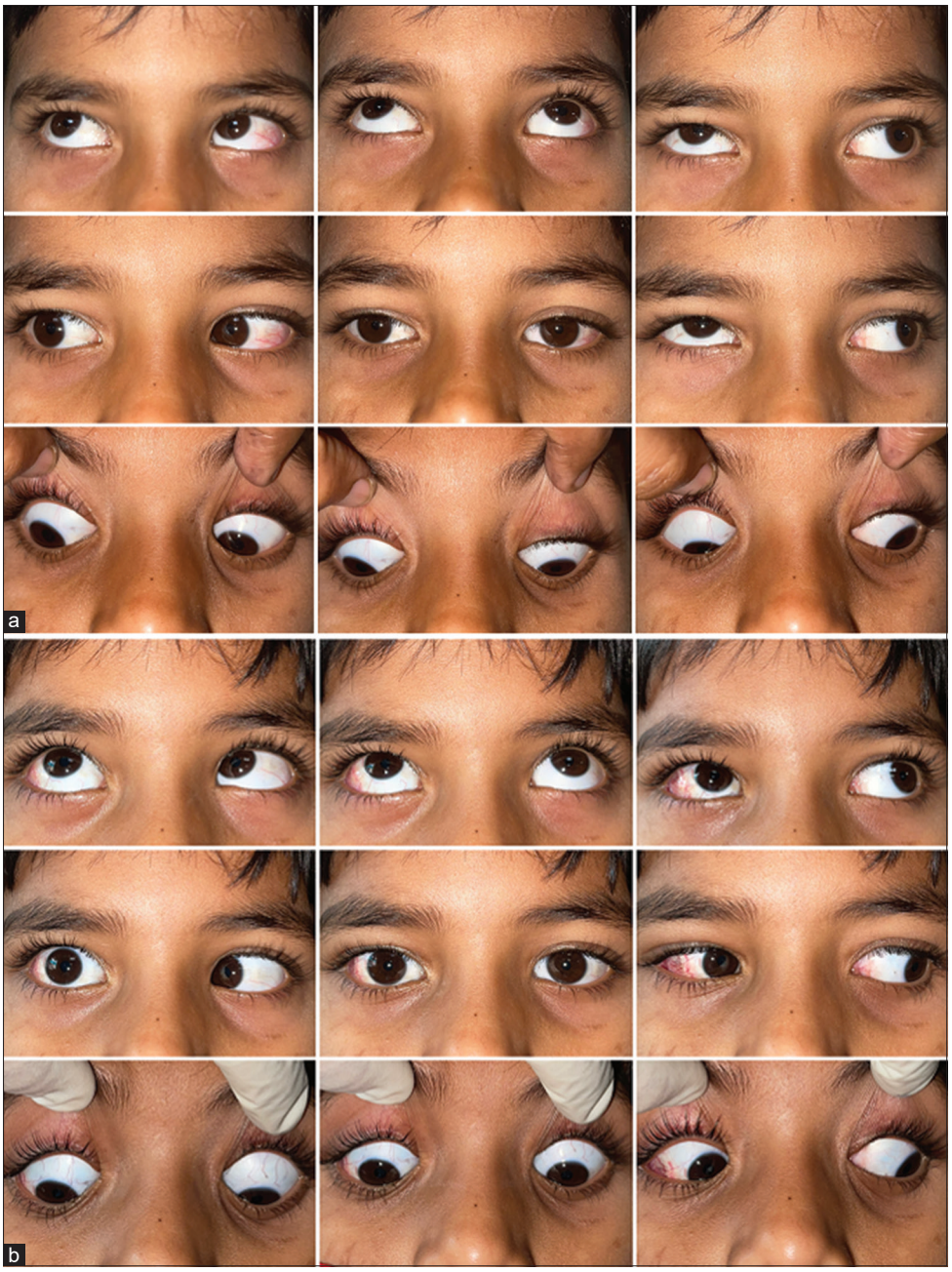


**Figure 15:** (a) Preoperative photographs of right exotropia–Duane syndrome with Grade 3 upshoots and downshoots. (b) Postoperative photographs of the same subject. Surgery done – Right lateral rectus recession 12 mm with Y-split. Note alignment in the primary gaze and disappearance of shoots

## Conclusion

Duane co-contraction-retraction syndrome is a nonprogressive congenital cranial dysinnervation syndrome. With our better understanding of the disease pathogenesis and presentations, and the advent of new surgical options, it is now possible to provide cosmetically acceptable outcomes, decent binocularity, and some amount of abduction. It is important to understand that the primary pathology can never be fully treated, and thus, the cases with a minimal amount of deviation/head posture, globe retraction, and shoots need only observation.

### Declaration of patient consent

The authors certify that they have obtained all appropriate consent forms from the legal guardians of the patients. In the form, the guardians have given the consents for the images and other clinical information of the patients to be reported in the journal. The guardians understand that the names and initials of the patients will not be published and due efforts will

be made to conceal the identity, but anonymity cannot be guaranteed.

## Data availability statement

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.


## Financial support and sponsorship

Nil.


## Conflicts of interest

The authors declare that there are no conflicts of interests in this article.

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