ISA Symposium: CHALLENGING DUANE CASES FROM AROUND THE WORLD

HANDOUT

Introduction

Duane Retraction Syndrome (DRS) is a distinctive form of restrictive strabismus marked by co-contraction of the medial and lateral rectus muscles, resulting from congenital anomalous innervation of the extraocular muscles.

It is among the most common forms of special strabismus and its prevalence in strabismic populations has been reported to range from 1% to 4% across different studies.

This syndrome is characterized by variable limitations or absence of abduction, varying degrees of adduction restriction, and globe retraction with narrowing of the palpebral fissure on adduction. These features are often accompanied by abrupt elevation or depression of the eye in adduction, commonly referred to as upshoots and downshoots.

The clinical manifestations in DRS can be quite variable and will depend upon the wide array of innervational abnormalities of the lateral rectus that can occur in these patients.

In this workshop, ISA members from around the world will discuss how to evaluate patients with Duane syndrome to obtain the most meaningful clinical information for selecting the optimal management strategy. The session will also explore the diverse clinical manifestations associated with this condition, along with the range of available treatment options.

CLINICAL INVESTIGATION OF DUANE RETRACTION SYNDROME

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The clinical presentation of DRS varies based on the pattern of anomalous innervation. The clinical investigation is directed at deducing that pattern.

Strabismus and Anomalous Head Posture

- PACT and SPCT should be done in forced primary position with the uninvolved (or less involved) eye fixating (prism over the Duane eye).
- Measurement of the secondary deviation in primary position is helpful, as large secondary angles suggest a more anomalous innervation pattern.
- Always measure to reversal of the shift to avoid under-estimation of the deviation.
- *Bilateral DRS is an important exception*!
    - With limited rotations OU, neither eye may be in the fixating position.

    - Measure with a prism over each eye in primary, with the larger prism over the more restricted eye, if applicable.

Globe Retraction and Lid Fissure Narrowing

- Retraction graded as a % difference in lid fissure height with the involved eye in maximum adduction and/or primary position
- Significant globe retraction (grade 3 to 4) in *primary* position suggests:
    - Greater % of MR fibers to LR
    - Presence of fibrous bands
    - Involvement of other EOMs

Shoots and Associated Vertical Deviations

- "Shoots" on adduction can be graded on a 1 to 4 scale
    - Horizontal line from pupillary center of uninvolved or fellow eye to judge vertical position of limbus of Duane eye
- Increasing vertical deviation as the eye moves into adduction or abduction may be innervational or mechanical
    - Abrupt change in vertical position on adduction is more likely mechanical
    - Gradual increase in vertical deviation as eye moves into adduction implicates oblique muscles
    - Gradual increase in vertical deviation as eye moves into abduction suggests dysinnervation involves the vertical recti
        - More common in Type I
        - Larger vertical with more severe limitation of abduction
        - More likely HYPOtropia in abduction

Refractive Error

- Hyperopia and astigmatism are common. The more anomalous the innervation pattern, the more likely there will be significant refractive error and anisometropia
    - DRS eyes have shorter axial length than uninvolved eye
    - DRS eyes have higher cylinder power than uninvolved eye
    - DRS eyes more likely to have oblique or ATR astigmatism

Conclusion

- Clinical findings correlate with innervation pattern; the more anomalous the innervation, the greater the deviation from clinical norms. More anomalous innervation patterns are also more likely to have associated fibrous bands.
- Indicators of highly anomalous innervation pattern include:

- Exotropia, regardless of classification
- Bilateral DRS, regardless of classification
- Larger secondary horizontal deviations in primary position
- Severe “shoots” or associated vertical deviations, particularly in side gazes
- Severe globe retraction (Grade 3 or 4)
- Anisometropia with significant hyperopic astigmatism in DRS eye

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GENETIC CONSIDERATIONS IN DUANE RETRACTION SYNDROME (DRS) Outline

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1) DRS is common (1/1000)

2) Most DRS is not syndromic or from gene mutation

3) Concept of pseudo-syndromic DRS

4) DRS that is truly syndromic or from gene mutation

ESOTROPIC DUANE

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Esotropic Duane retraction syndrome (DRS) is more common than the exotropic variant and typically presents with an esodeviation in the primary position. In most cases, the angle of deviation in primary position is modest, usually less than 30 prism dioptres. A characteristic clinical feature is that the limitation of abduction is greater than the limitation of adduction, reflecting the underlying co-contraction and anomalous innervation of the lateral rectus muscle.

Clinical assessment should be systematic and guide management planning. The surgical (or conservative) approach depends on several key factors: alignment in primary position, the presence and degree of abnormal head posture, the extent of limitation of ductions, and the severity of globe retraction and any upshoots or downshoots on adduction. Patients with good primary position alignment and minimal head posture may be managed conservatively, whereas significant compensatory head turn or cosmetically concerning retraction and overshoots may warrant surgical intervention tailored to the individual motility pattern.

It is also essential to ensure that hypermetropia is fully corrected, particularly if there is any suspicion of an accommodative component contributing to the esotropia. A cycloplegic refraction should be performed, and appropriate spectacle correction prescribed before considering surgical options, as some patients may demonstrate improvement in primary position alignment with optical management alone.

Management of esotropic Duane Retraction Syndrome (DRS) involves several surgical strategies based on the severity of deviation and the patient's motility pattern:

Unilateral and Bilateral Medial Rectus (MR) Recession

- **Unilateral Recession**: Typically used for primary position deviations under 20 prism diopters (PD). Recession is generally limited to 5–6 mm to avoid compromising adduction.
- **Bilateral Recession**: Considered for deviations exceeding 20 PD. Using both eyes helps reduce the amount of recession needed on the affected eye.
- **Asymmetric Approach**: Jampolsky suggests asymmetric MR recession, as unilateral surgery often fails to fully correct compensatory head turns.

Lateral Rectus (LR) Resection

- **Indications**: Effective for patients with esotropia of at least 25 PD, significant abduction limitation, but clinically normal adduction and minimal retraction.
- **Procedure**: Combines an MR recession of $\leq$ 5.0 mm with an LR resection of maximum 3.5 mm.
- **Outcome**: Frequently results in a clinically significant improvement in abduction.

Vertical Rectus Transposition (VRT)

- **Technique**: Involves transposing one or both vertical recti, sometimes augmented with a posterior fixation suture (PFS).
- **Safety**: Surgeons should use Forced Duction Testing (FDT) on the MR to assess the risk of Anterior Segment Ischemia (ASI). Ipsilateral MR recession should generally be avoided during the primary procedure.
- **Disadvantages**: May induce vertical deviations, increased globe retraction on abduction, and more pronounced up-shoots or down-shoots.

Surgery on the Normal Eye

- **Rationale**: Saunders et al. propose that large recessions or PFS on the unaffected eye can create comparable duction limitations, widening the diplopia-free field.
- **Innervation**: While Hering's law suggests equal innervation to yoke muscles, it does not apply normally in DRS due to anomalous innervation.

EXOTROPIC DUANE

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Exotropic Duane syndrome is characterized by an exotropia in primary gaze, a limitation of adduction that is usually greater than the limitation of abduction, abduction being almost always normal—though not invariably—and a head turn toward the side opposite the affected eye.

Generally, although not always, it corresponds to Type II in Huber's classification. It is the least frequently seen type of Duane Syndrome.

A characteristic clinical feature of exotropic Duane syndrome is the marked globe retraction observed during attempted adduction. This phenomenon is driven by the strong innervation of the lateral rectus, supplied by a sixth nerve that is often nearly normal, combined with the anomalous branch of the third nerve. As a result, the lateral rectus receives both normal tone from the sixth nerve and aberrant innervation from the third, producing a more powerful co-contraction and consequently greater enophthalmos.

In some cases of exotropic Duane syndrome, co-innervation between the medial and superior rectus muscles can be observed, producing an elevation during attempted adduction that is distinct from a typical mechanical upshoot.

Anomalous orbital structures—such as fibrous bands located beneath or adjacent to the extraocular muscles, particularly the lateral rectus—are identified more frequently in this subtype than in other forms of Duane syndrome.

Treatment of these patients always involves weakening of the ipsilateral rectus muscles. Conventional surgical tables based on the primary-position deviation are not applicable here. Larger recessions are usually needed, in some cases associated with a Y split, especially in cases with significant retraction or up and down-shoot.

Recession of the lateral rectus in the fellow eye can be considered when the primary-position deviation exceeds 25 prism diopters. However, it should never be performed when the patient exhibits marked globe retraction on attempted adduction, as this indicates strong anomalous innervation. In such cases, recessing the normal eye's lateral rectus increases innervation to its yoked medial rectus by Hering's law, and due to the anomalous pathway, the affected eye's lateral rectus receives additional innervation, worsening the exotropia.

The medial rectus of the affected eye should never be recessed for the same reason. Weakening the medial rectus requires increased innervation to maintain primary-position alignment, and this additional drive is transmitted to the lateral rectus through the anomalous branch, further increasing the exotropia.

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Duanes syndrome associated with Vertical Deviations

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Duane syndrome is one of the most common restrictive congenital strabismus characterized by variable horizontal duction deficits with globe retraction and upshoots or downshoots on attempted adduction and narrowing of the palpebral aperture. It is now listed as a congenital cranial dysinnervation disorder.

Anomalous Vertical movements :

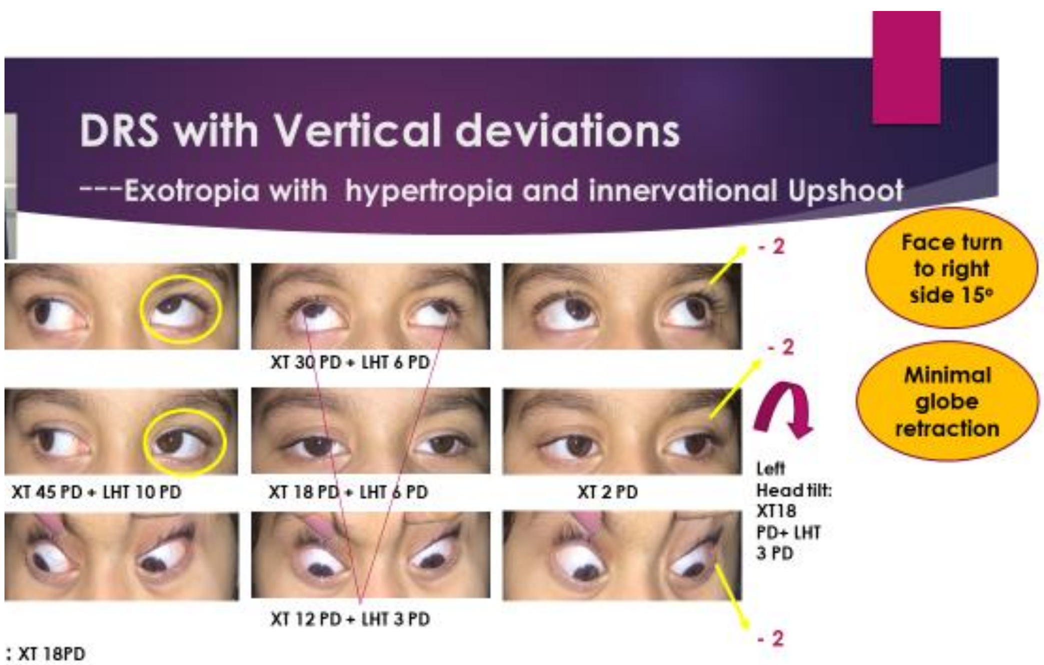
Mechanical	Innervational
Abrupt up and down movement	Gradual elevation and depression
No vertical deviation in primary position	May have vertical deviation in primary position
Leash effect	Aberrant innervation of muscles

Kekunnaya et al (1) have reported an incidence of upshoot or downshoot in 43% of their cases. They found these movements to be more common in unilateral DRS types I and III, unlike Mohan et al (2)who found the incidence to be significantly greater in type III. In this study, Mohan et al have found the mechanical type to be more common than the innervational type (26% vs 12%). They also found a statistically significant association between exotropic DRS and mechanical upshoots and primary position vertical tropia with innervational upshoots.[2]

Along with horizontal strabismus, vertical strabismus is also seen in type III DRS. The vertical deviations may be due to fibrosis of the vertical muscles, innervational anomalies, or tethering effect of the horizontal muscles.

In the study by Isenberg and Urist, (3) they found that there is drooping of upper lid along with elevation of lower lid in about 52% of cases of DRS. In 18%, only upper lid changes were noted, while 21% had only lower lid elevation. A small proportion of cases (7%) did not show any lid changes with motility in DRS. (Video shall be shown during presentation).

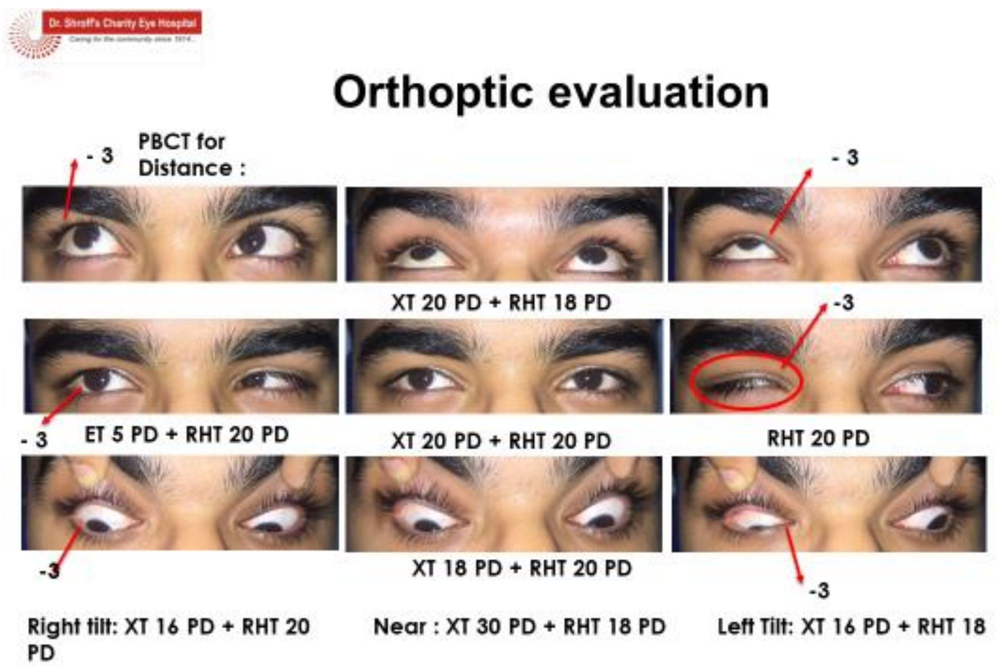
Case 1 for discussion ; EXO DRS with Hypertropia with V pattern with Innervational upshoot



Is there associated upshoot or is it inferior oblique overaction ?

Surgery done : **Left eye IO Rec 3:2 + LR Rec16 mm**

Case 2:

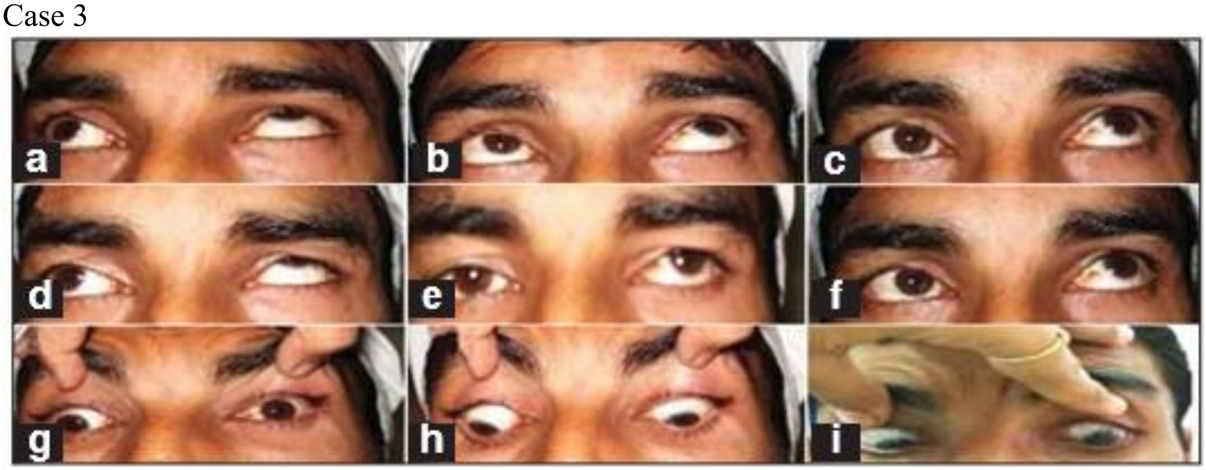


Surgery done

Superior rectus recession combined with lateral rectus recession and Y-split surgery for exotropia with large comitant innervational hypertropia in a case of type III Duane retraction syndrome

Case 3

Case 3



Surgery done :
Superior rectus recession along with Lateral rectus recession and Y split surgery

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   Virender Sachdeva,Tishya Vepakomma,Anjali Chandrasekharan,Ramesh Kekunnaya .Published online: 28 Aug 2025 STRABISMUS

ANOMALOUS ORBITAL STRUCTURES IN DUANE SYNDROME

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- Anomalous orbital structures in Duane syndrome could be muscular structures or fibrous bands.
- The presence of accessory muscles or fibrous bands results in unusual/ atypical patterns of strabismus.
- Atypical Duane syndrome cases should undergo orbital imaging, repeated forced duction testing after disinsertion of each muscle and

careful exploration to verify presence or absence of anomalous structures.

- Management includes excision/disinsertion of accessory muscles to normalize ocular movements together with weakening of other muscles that contribute to restriction of motility.
- Two cases of atypical Duane syndrome with anomalous orbital structures will be presented. The clinical picture and management of each case will be discussed.

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