Classification and Various Presentations of Duanes Retraction Syndrome

DR SUMA GANESH
DEPUTY MEDICAL DIRECTOR
HEAD OF PAEDIATRIC OPHTHALMOLOGY AND STRABISMUS
DR. SHROFF’S CHARITY EYE HOSPITAL,
NEW DELHI, INDIA
Duane’s Retraction Syndrome ---- Classification

Duanes retraction syndrome is a spectrum of neurological, mechanical, innervational and genetic abnormalities.

Various Systems of classification were proposed to understand the mechanism and presentation of DRS
Duane’s Retraction Syndrome ---- Huber Classification

- Huber classification based on EMG. But it does not cover all types of cases. Lot of overlaps.

- Ahluwalia et al modified the classification depending on the alignment in primary gaze.

Huber Classification
Duanes Type 1

- Limitation of abduction
- Esotropia
- Normal or slightly defective adduction
- Narrowing of the palpebral fissure and retraction of globe on adduction

Duane TYPE 1 (70 to 80%)

- Widening of the fissure on attempted abduction
- Head turn to the involved side
Type 1 - Large Esotropia with Large face turn

- Esotropia 30 prism diopters
- Abduction deficit better than – 4
- Minimal adduction deficit and retraction

30 Δ BO
Type 1 - Small esotropia with small face turn with minimal globe retraction

PBCT for Distance

PBCT for Near : 6 ET
Huber Classification

DRS TYPE 2

- Limitation or absence of adduction
- Exotropia
- Narrowing of the fissure on attempted adduction

DRS Type 2 (7%)

- Normal or reduced abduction
- Face turn to the normal side
- EMR lateral rectus showed peaked impulses on abduction and paradoxical impulse on adduction
Type 2 : XL Exo DRS with Downshoot

Type 2: Small exotropia XT with upshoot and marked globe retraction
Hubers classification Type 3 DRS

Duanes Type 3 (15%)

- Limited abduction and adduction
- Retraction of the globe and narrowing of the fissure on adduction
- Esotropia, exotropia or orthotropia equally
- Frequent upshoot and downshoot with attempted adduction
- Usually no face turn
- EMG shows simultaneous innervation of lateral rectus and medial rectus in primary gaze, adduction and abduction
Type 3 DRS – Orthotropia with upshoot and downshoot
Type 3 - Esotropia with upshoot and downshoot
Minimal face turn
Globe retraction

4 BO
30 ET
-4 abduction deficit
30 BO
Type 4

- Synergistic divergence or Divergence Splits
- Large exotropia
- Adduction deficit
- Simultaneous abduction on attempted adduction
- EMG data demonstrated co-contraction and excessive LR firing on adduction.
Type 5: Vertical retraction syndrome: Combined severe horizontal and vertical

- Horizontal DS plus a globe retraction on vertical positions of gaze with or without limitation of vertical eye movements.
- Vertical recti contracted on abduction
- Limitation on depression

Acknowledgement: Dr Tanmayi Dhamankar Bose for the video
Bilateral ESO DRS without fusion

Note the narrowed fissure in each eye during adduction and the limited abduction demonstrated by each eye.
Polling Question:

Most common Type of Refractive error seen in DRS

a. Myopia
b. Myopic astigmatism
c. Hyperopia
d. Hyperopic astigmatism
Refractive error

- Anisometropic and ametropic Amblyopia most common
- Hypermetropia most common
- Check for accommodative component too, it should be corrected (cycloplegic refraction)

Important to correct these to restore binocularity
Globe retraction: Co-contracture of extraocular muscles

Grading of globe retractions
0: no narrowing
1: <25%
2: 25-<50%
3: 50-<75%
4: >75%
GRADING OF Globe Retraction

No Narrowing
Grade 0

10 mm
9 mm

12 mm
12 mm

Globe retraction 10%
Grade 1

Globe retraction 30.8%
Grade 2

13 mm
9 mm

Globe retraction 73%
Grade 3

3 mm
11 mm
Grading of upshoot

Grade 0
Line bisects pupil of involved eye

Grade 1
Line lies b/w pupillary centre n pupillary margin
A line drawn from pupillary centre of normal eye

Grade 2
Line lies b/w pupillary margin n limbus

Grade 3
Line lies at the limbus or over the sclera

Grade 4
Cornea disappears below the lid
Anomalous vertical movements in DRS

### Upshoots and Downshoots

<table>
<thead>
<tr>
<th>Mechanical</th>
<th>Innervational</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abrupt up and down movement</td>
<td>Gradual elevation and depression</td>
</tr>
<tr>
<td>No vertical deviation in primary position</td>
<td>May have vertical deviation in primary position</td>
</tr>
<tr>
<td>Leash effect</td>
<td>Aberrant innervation of muscles</td>
</tr>
</tbody>
</table>

Mohan et al have found the mechanical type to be more common than the innervational type (26% vs 12%). More common in type 3.

Upshoots and downshoots more common in unilateral DRS types I and III.
Polling Question:

Question: Most common Pattern seen in DRS

a. Y pattern
b. X pattern
c. V pattern
d. A pattern
Differentials:

Infantile esotropia

Doll's head maneuver

Duane retraction syndrome (OS)

Sixth nerve palsy (OD)

Mobius syndrome
Case 1: Eso Duanes

- Esotropia with head turn
- Abduction deficit – 4
- Forced Duction test negative
- No upshoot or downshoot
- Minimal globe retraction

25 PD of Esotropia
Review of Literature

Comparison of augmented superior rectus transposition with medial rectus recession for surgical management of esotropic Duane retraction syndrome

Shalja Tibrewal, MS, a Virender Sachdeva, MS, DNB, b Mohammed Hasnat Ali, MBA, c and Ramesh Kekunnaya, MD, FRCS d

Superior Rectus Transposition vs Medial Rectus Recession for Treatment of Esotropic Duane Syndrome

Shiqiang Yang, MD; Sarah Mackinnon, MSc, OC(C); Linda R. Dagi, MD; David G. Hunter, MD, PhD

Surgical outcome of superior rectus transposition in esotropic Duane syndrome and abducens nerve palsy

Rohit Agarwal, MD, Medha Sharma, MD, Rohit Saxena, MD, and Pradeep Sharma, MD

Superior or inferior rectus transposition in esotropic Duane syndrome: a longitudinal analysis.

Sener EC, Yilmaz PT, Fathioglu OU

Superior rectus transposition combined with medial rectus recession for Duane syndrome and sixth nerve palsy

Reshma A. Mehendale, MD, Linda R. Dagi, MD, Carolyn Wu, MD, Danielle Ledoux, MD, Suzanne Johnston, MD, and David G. Hunter, MD, PhD
Department of Ophthalmology, Children's Hospital Boston & Harvard Medical School, Boston, Massachusetts

Surgery: MR Recess with SRT

Postoperative picture
Case 2 : History

- 6 yrs old girl
- Resident of Delhi
- **C/C : Abnormal head posture and squinting of eyes since the age of 1 year**
- H/O wearing glasses since 2 years
- No H/O ocular trauma/ head trauma
- Birth History : Full term, NVD, No complications
- Family History : Not Significant
- Past Medical history : Not significant
<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual Acuity (BCVA)</td>
<td>6/9 (-4.50x 180°)</td>
<td>6/9p (-4.00 x 180°)</td>
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<tr>
<td>Near Vision</td>
<td>N6</td>
<td>N6</td>
</tr>
<tr>
<td>Lids and Adenexa</td>
<td>Normal</td>
<td>Normal</td>
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<tr>
<td>Conjunctiva</td>
<td>Normal</td>
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<tr>
<td>Cornea</td>
<td>Clear</td>
<td>Clear</td>
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<tr>
<td>AC</td>
<td>VH Gr 4</td>
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<tr>
<td>Lens</td>
<td>Clear</td>
<td>Clear</td>
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<tr>
<td>Fundus</td>
<td>Normal</td>
<td>Extorsion</td>
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</tbody>
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DRS with Vertical deviations
---Exotropia with hypertropia and innervational Upshoot

Face turn to right side 15°
Minimal globe retraction
Left eye Upshoot on adduction

Right Head tilt: XT18 PD

XT 45 PD + LHT 10 PD
XT 45 PD + LHT 10 PD
XT 45 PD + LHT 10 PD

XT 30 PD + LHT 6 PD
XT 18 PD + LHT 6 PD
XT 12 PD + LHT 3 PD

Left Head tilt: XT18 PD+ LHT 3 PD

Left eye Upshoot on adduction

PBCT Near : XT 18PD
Sensory Examination

- **Worth 4 Dot Test**: Distance – OS Suppression  
  Near – Alternate suppression

- **Stereopsis**: 80 sec of arc
Questions?

- Is there associated upshoot or is it inferior oblique overaction?

- Do I touch the inferior oblique muscle?

Awadein A. Inferior oblique myectomy for upshoots mimicking inferior oblique overaction in Duane retraction syndrome. J AAPOS. 2013;
Surgery ---

Surgery done: Left eye IO Recession 3:2 + LR
Recession 16 mm
Post op 2 months
Surgery done: Left eye IO Rec 3:2 + LR Rec 16 mm

Orthophoria

10 PD XT

8 PD ET

3 PD XT

PBCT Near: 8 PD XT

W4DT: BSV, Stereopsis: 80 sec of arc
Surgery: Lateral rectus recession with Y split
Postoperative pics
CASE 3:

Globe retraction with narrowing of palpebral aperture with upshoot 35 BI

25 BI for distance 25 BI for near

4 BI
Thank You

Dr Suma Ganesh
Questions:
drsumaganesh@yahoo.com