DUANE’S SYNDROME: CLINICAL FEATURES AND SURGICAL MANAGEMENT

Ananda (Andy) Kalevar, MD
Michael Flanders, MD
McGill University - MUHC, Ophthalmology

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Definition

- Congenital eye-movement disorder
- Failure of CN VI to develop normally
- Limitation of abduction/adduction
- Narrowing of palpebral fissure and retraction of the globe on adduction
Background - History

- 1879-1905: Hueck, Stilling, Turk, Duane
  - Abduction/adduction deficit; Head turn
  - Globe retraction/fissure narrowing on adduction
  - Upshoot/downshoot on adduction

- 1974: Huber-Types I, II, III (EMG); miswiring

- 1980: Hotchkiss - absence of VIth nerve and nucleus in a bilateral case (autopsy)

- 2002: CCDD – neurodevelopmental disease of brainstem and cranial nerves
Duane type I

ADD - 1/2
ET in FPP
ABD -3 1/2

Head turn (L)
Duane Type II
Upshoot with “V” pattern

Head turn (R)

UPSHOOT
ADD -3
XT in FPP
ABD -2
Duane type III
Upshoot, Downshoot, X pattern

- ABD -3.5, ET 50
- Ortho
- XT 20
- ADD -3, XT 40
- Normal head posture
- XT 15
- Upshoot
- Downshoot
Background - Epidemiology

- Duane syndrome: 1-5% of strabismus
- Type I: 75-80%, II: 5-10%, III: 10-20%
- Unilateral 80% (left eye 68%)
- Females: Males 3:2
“Y” spit + Recess Lateral Rectus OS
Background - Surgery

- **Indications:**
  - significant primary position misalignment
  - significant abnormal head posture
  - unsightly fissure narrowing
  - unsightly upshoots or downshoots

- **Strategies:**
  - ET → MR Recess, XT → LR Recess
  - Globe retraction → LR/MR Recess
  - Up/Down shoots → LR Surgery
  - Transposition surgery
Purpose

- Report clinical findings and surgical results in 75 patients with Duane syndrome
- Classify with emphasis on forced primary position alignment
- Explore relationship of up/down shoots with A, V and X patterns
- Examine alignment in adduction in Type I Duane
Study design

- Retrospective chart reviews including clinical series and interventional subset

- Patients from private practice of Dr Michael Flanders seen during the period 1986-2011
Selection & Methodology

- Names of 93 patients with Duane syndrome were extracted from Dr Flanders’ strabismus database.
- 75 patients remained after exclusion criteria applied.
Selection & Methodology

- Ophthalmologic & orthoptic exam data collected as follows:
  - Age, sex, laterality
  - Pre & post-op head position, fissure narrowing/globe retraction
  - Pre & post-op ocular alignment (forced primary position, up/down gaze, lateral gaze
  - Pre & post-op motility abnormalities (abduction, adduction, Up/Down shoots)

- Category of Duane assigned based on type of forced primary position alignment: ET=Type I, XT=Type II; Ortho=Type III
Selection & Methodology

- 18 patients underwent strabismus surgery
- MR Recess ??, LR Recess ?? etc
- Criteria for levels surgical success
  - Excellent: Forced primary position (FPP) alignment equal to or <10 PD; Head position (HP) significantly improved
  - Fair: FPP >10 PD +/- some improvement in HP
  - Poor: no improvement
Results, observational (n=75)

**Types**

- Type I: 56%
- Type II: 28%
- Type III: 16%

**Tropia, PD**

- Type I (eso): 6
- Type II (exo): 15
- Type III (exo): 0

**Head Turn**

- Type I (toward affected eye): 85%
- Type II (away from affected): 80%
- Type III (no head turn): 74%
- Male:Female
- Unilat vs Bilat
- Right vs Left eye
Results, observational

**Horizontal Motility Deficits**

- **Abduction deficit**
  - Type I: 3.8
  - Type II: 1.5
  - Type III: 3.0

- **Adduction deficit**
  - Type I: 0.2
  - Type II: 2.4
  - Type III: 1.0
Results, observational

- 96% had fissure-narrowing/globe retraction
- 67% had an upshoot and/or downshoot
- 63% had an “A”, “V”, or “X” pattern
Emanuel Maris – Duane type I – pre-op
Emanuel Maris – Duane type I – post-op

(L) Medial rectus recess 6 mm
Duane type I – Bilateral
Results, surgical (n = 18)

Types

- Type I: 14
- Type II: 4
- Type III: 0
Results, surgical (n=18)

Forced Primary Position

Type I: 27
Type II: 15

Pre-op (PD): 27
Post-op (PD): 8

50

0
Results, surgical (n=18)

Head Position Improvement

- Type I: 71%
- Type II: 100%
Duane type I - Preop
Duane type I - Post-op
Duane Type II
Upshoot with “V” pattern

Head turn (R)

UPSHOOT
ADD -3
XT in FPP
ABD -2
Results, surgical (n=18)

Surgical outcomes

- Excellent: 81% Both types, 75% Type I, 100% Type II
- Partial: 6% Both types, 8% Type I, 0% Type II
- Failure: 13% Both types, 17% Type I, 0% Type II
Discussion

- Classification modified for surgery
- “Shoots” correlation with the A, V, X patterns
- Contralateral gaze
- Overall surgical success
- Bilateral cases, comment???
- Abd > Add in type III, why?
Conclusion

- Majority are unilateral, female, OS affected, consistent relationship between type, FFP and in turn head turn and motility defects.
- “A”, ”V” and “X” syndromes correlate with the type of up/downshoot present.
- Surgery can result in significant improvement of abnormal head turn and reduction of primary position alignment in types I, II
References

Results, observational (n=75)

Types

- Type I: 56%
- Type II: 16%
- Type III: 28%

Laterality

- Unilateral: 91%
- Bilateral: 9%

Sex

- Female: 57%
- Male: 43%

Laterality

- OS: 66%
- OD: 34%
Results, observational

**Tropia, PD**

<table>
<thead>
<tr>
<th>Type</th>
<th>I (eso)</th>
<th>II (exo)</th>
<th>III</th>
</tr>
</thead>
<tbody>
<tr>
<td>Count</td>
<td>6</td>
<td>15</td>
<td>0</td>
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**Head Turn**

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