Duane's Syndrome: Clinical Features and Surgical Management

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Definition

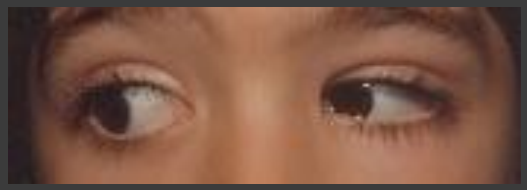
- Congenital eye-movement disorder
- Failure of CN VI to develop normally
- Limitation of abduction/adduction
- Narrowing of palpebral fissure and retraction of the globe on adduction

Background - History

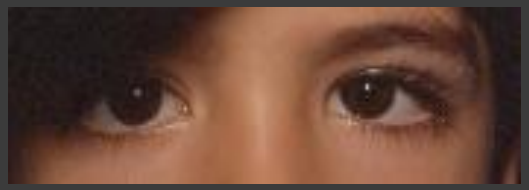
- 1879-1905: Hueck, Stilling, Turk, Duane
  - Abduction/adduction deficit; Head turn
  - Globe retraction/fissure narrowing on adduction
  - Upshoot/downshoot on adduction
- 1974: Huber-Types I, II, III (EMG); miswiring
- 1980: Hotchkiss - absence of VIth nerve and nucleus in a bilateral case (autopsy)
- 2002: CCDD – neurodevelopmental disease of brainstem and cranial nerves

Duane type I

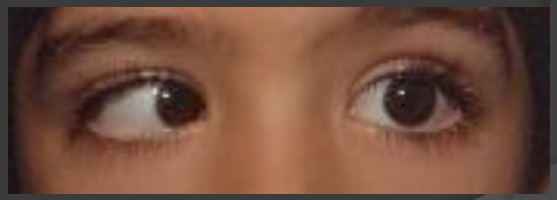
Head turn (L)



ADD - 1/2

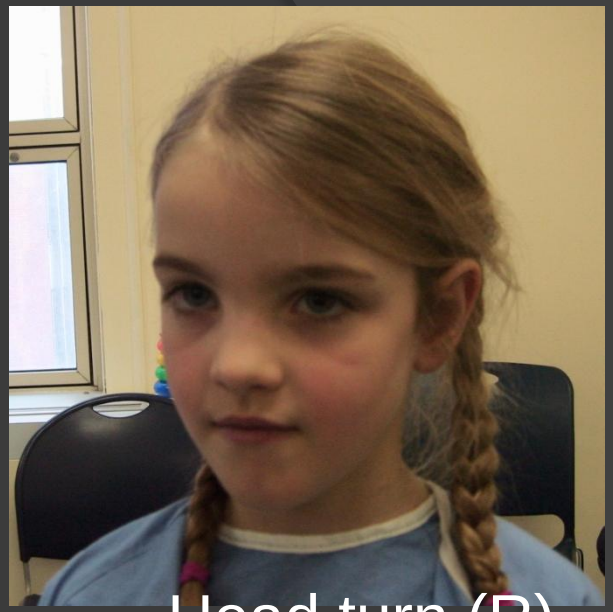


ET in FPP



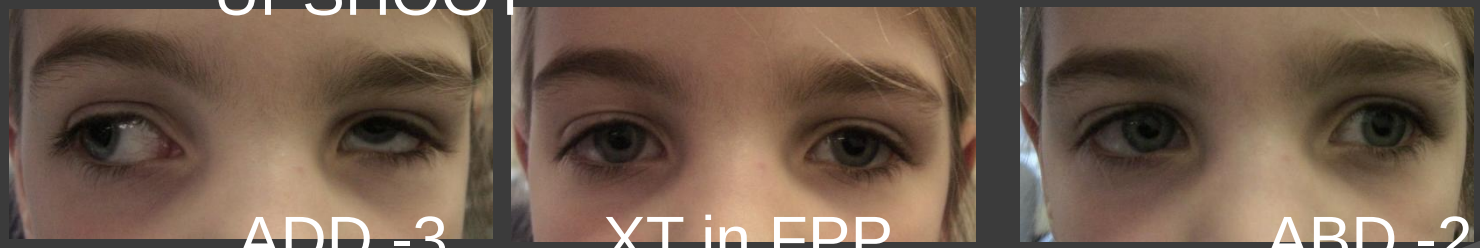
ABD -3 1/2

Duane Type II
Upshoot with “V” pattern



Head turn (R)

UPSHOOT

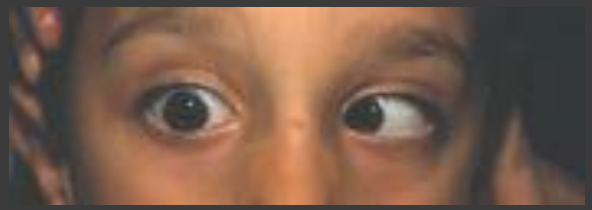


ADD -3 XT in FPP ABD -2

Duane type III
Upshoot, Downshoot, X pattern

XT 15

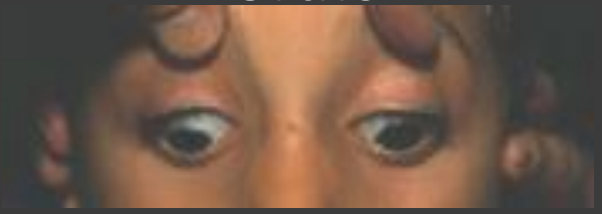
Upshoot



ABD -3.5, ET 50

Ortho

ADD -3, XT 40



XT 20

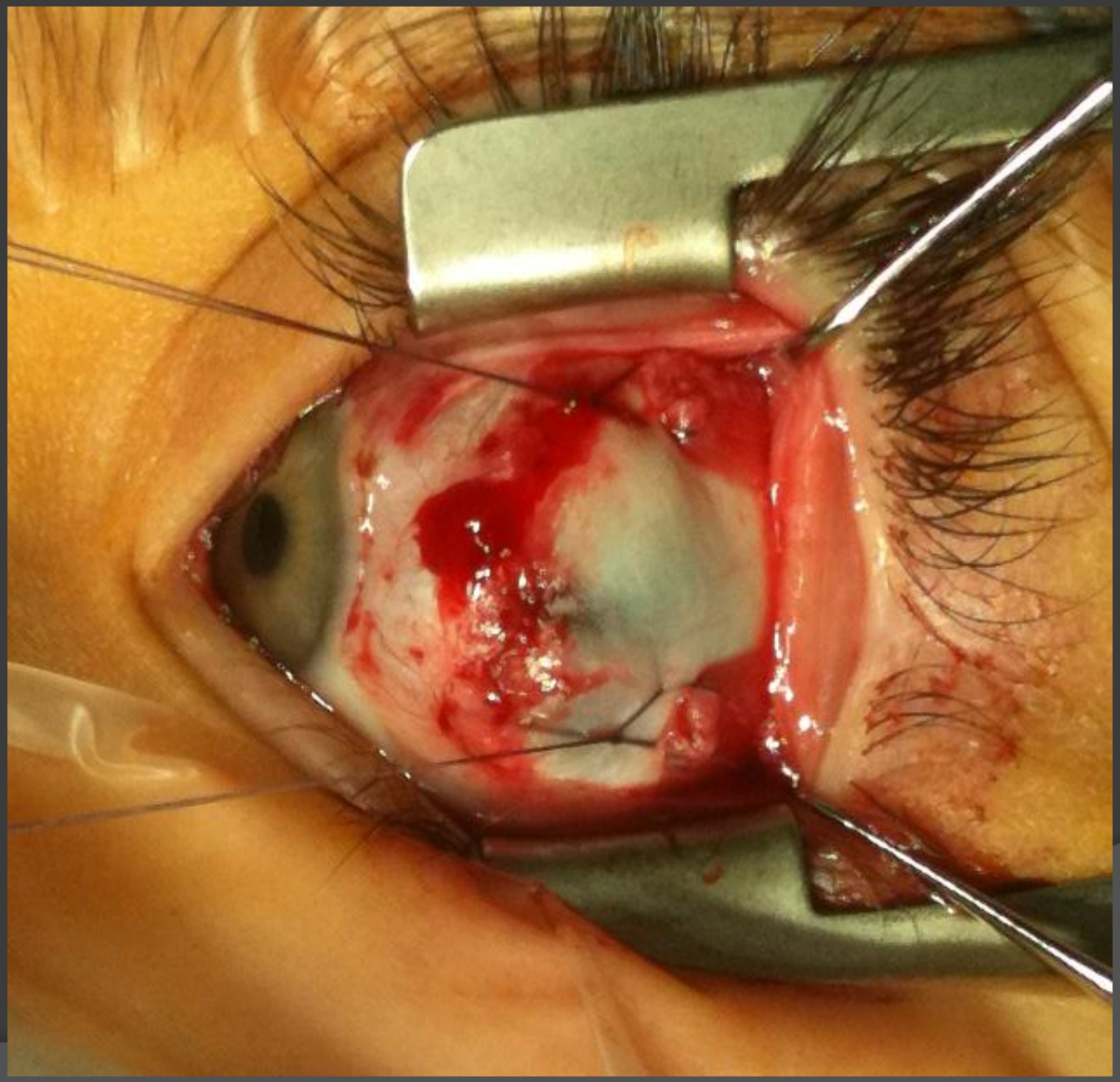
Downshoot

Normal head posture

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

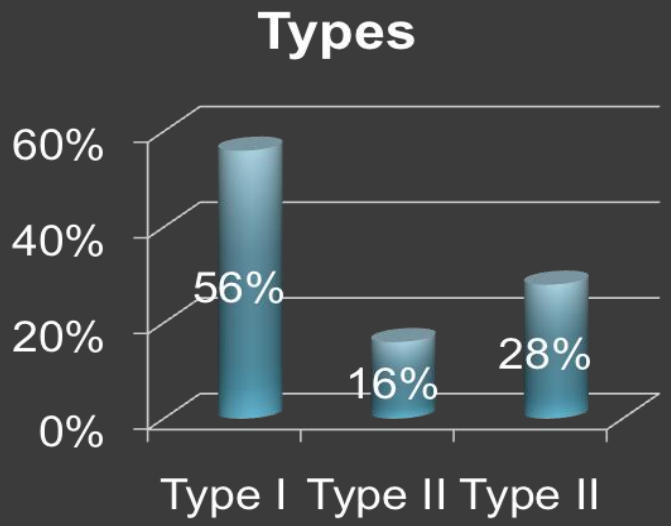
Selection & Methodology

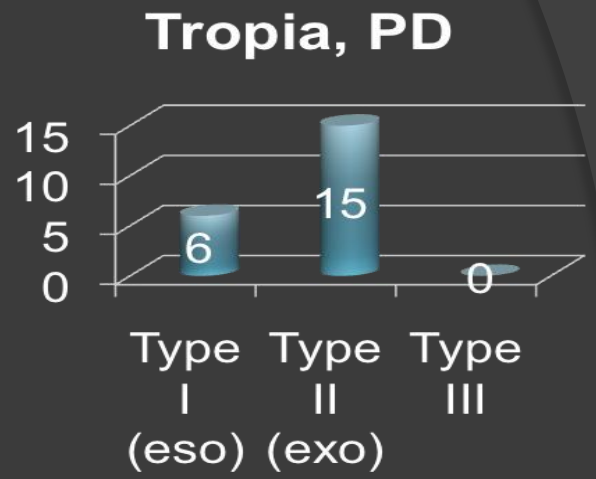
- Ophthalmologic & orthoptic exam data collected as follows:
  - Age, sex, laterality
  - Pre & post-op head position, fissure narrowing/globe retraction
  - Pre & post-op ocular alignment (forced primary position, up/down gaze, lateral gaze
  - Pre & post-op motility abnormalities (abduction, adduction, Up/Down shoots)
- Category of Duane assigned based on type of forced primary position alignment: ET=Type I, XT=Type II; Ortho=Type III

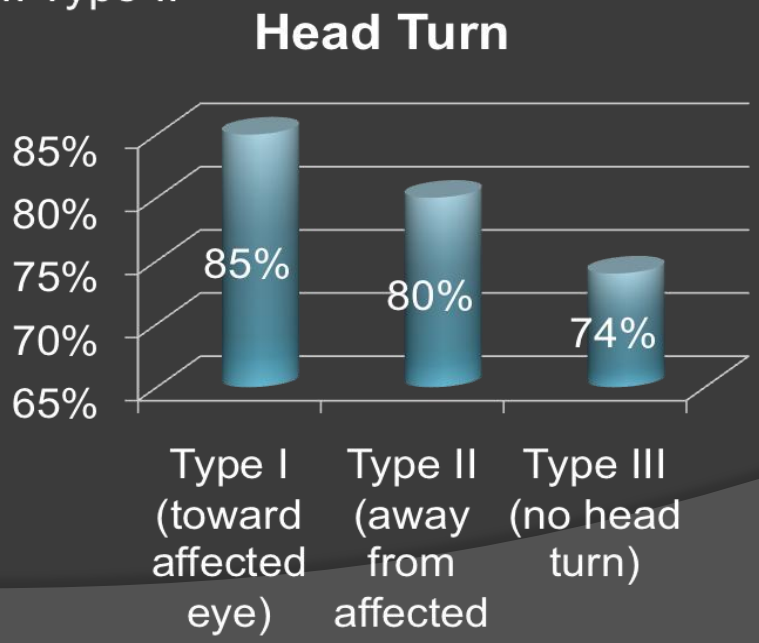
Selection & Methodology

- 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)

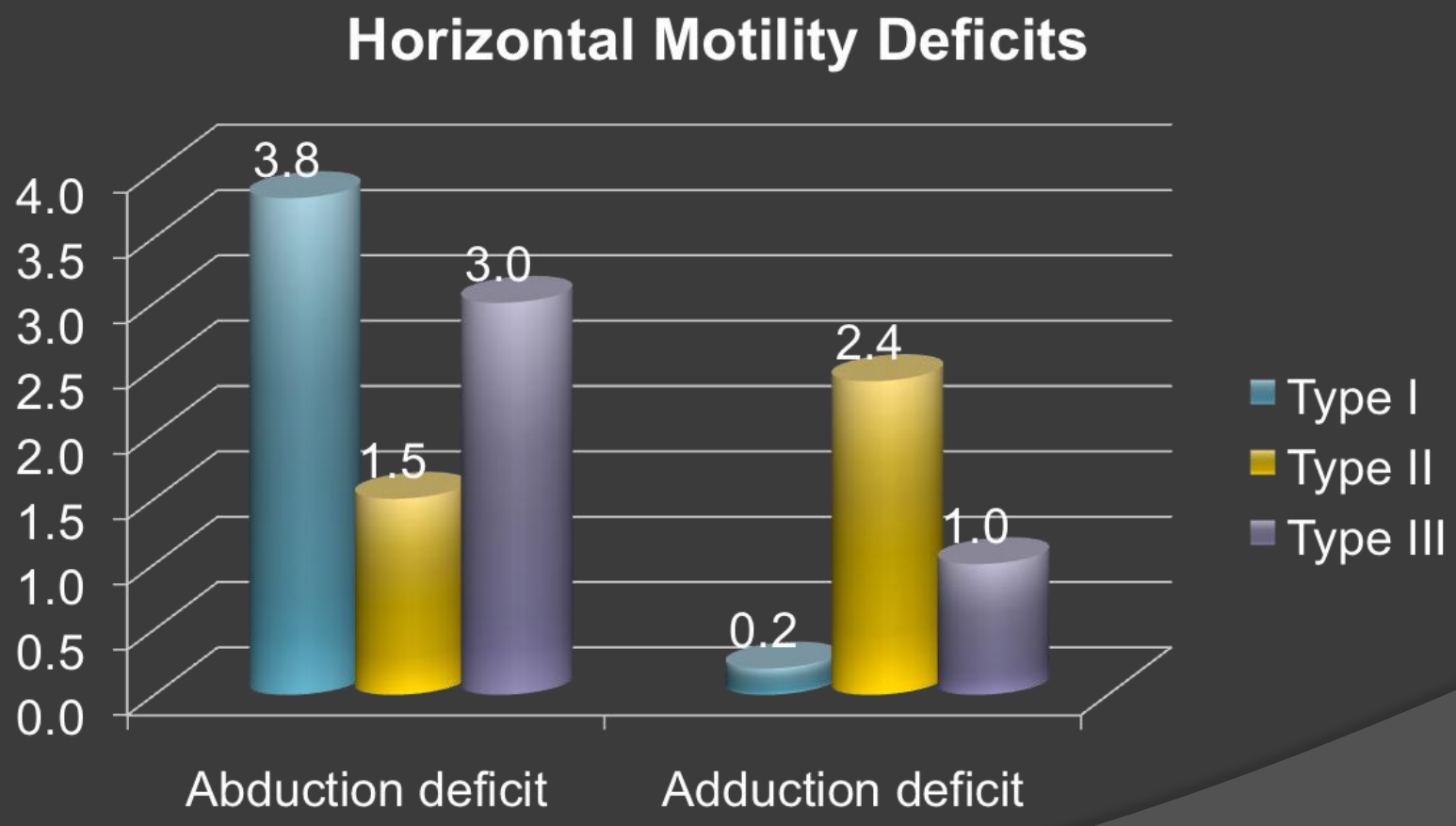






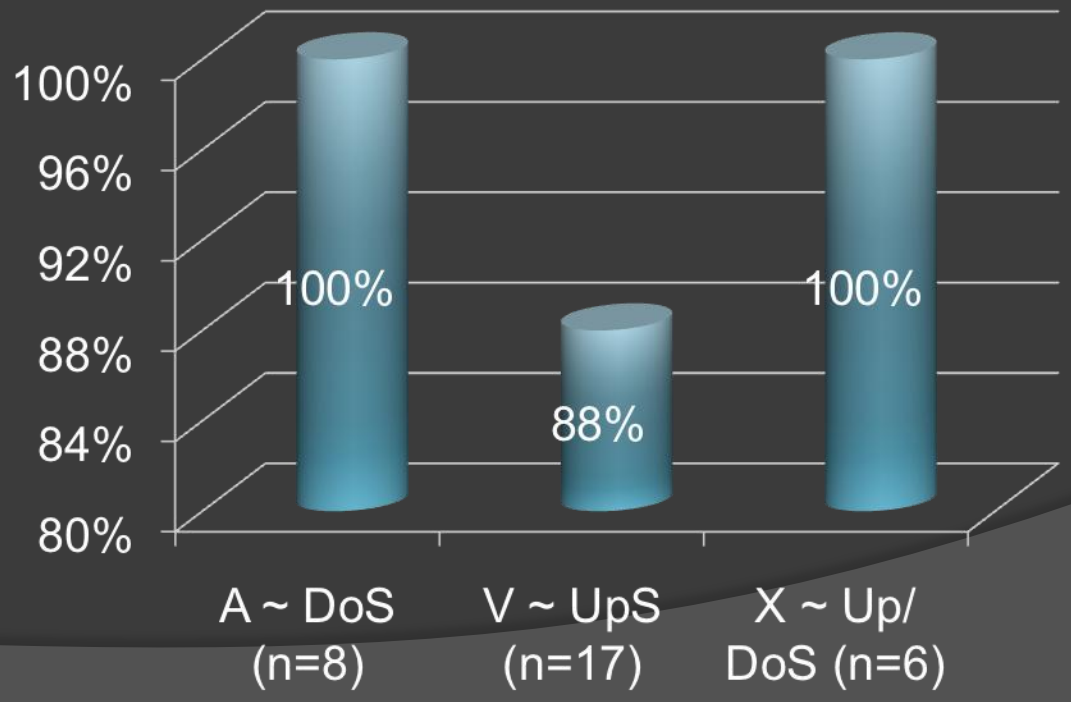
- Male:Female
- Unilat vs Bilat
- Right vs Left eye

Results, observational

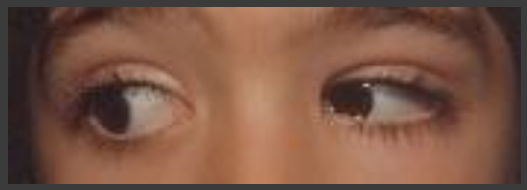


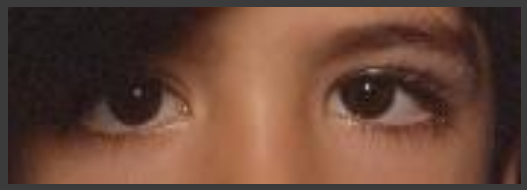
Results, observational

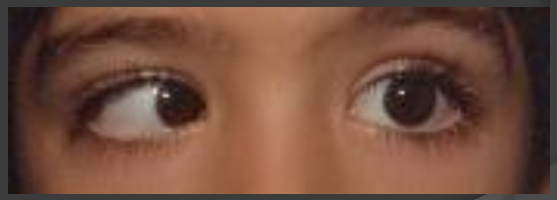
- 96% had fissure-narrowing/globe retraction
- 67% had an upshoot and/or downshoot
- 63% had an “A”, “V”, or “X” pattern



Emanuel Maris – Duane type I – pre-op

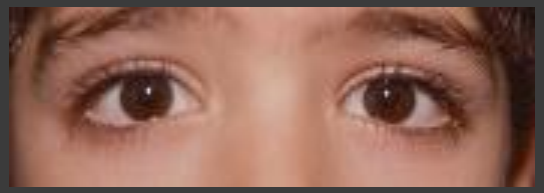






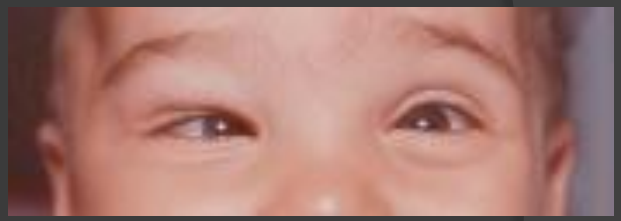
Emanuel Maris – Duane type I – post-op

(L) Medial rectus recess 6 mm

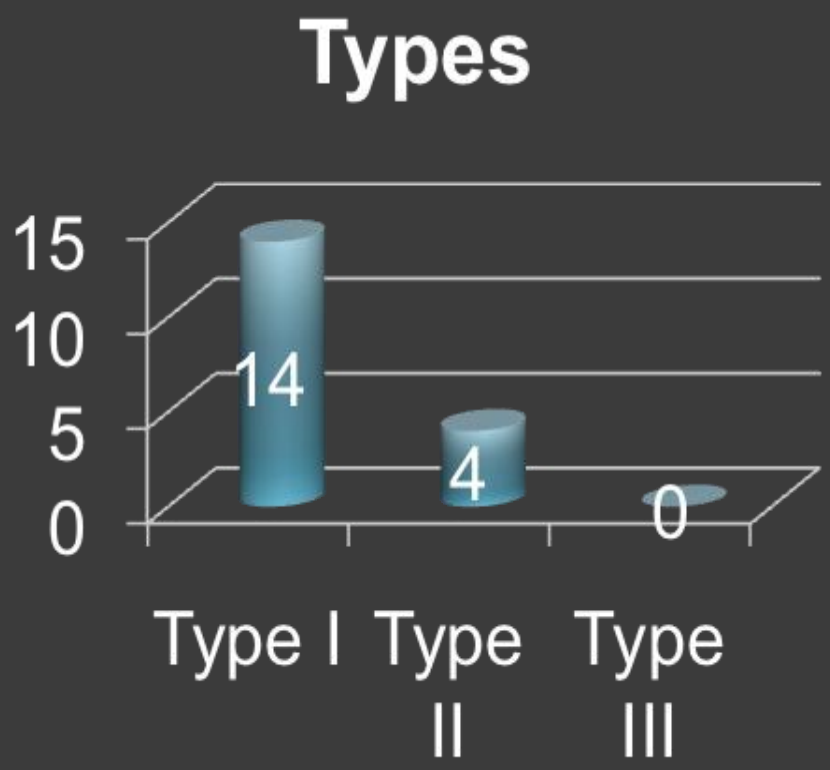


Duane type I – Bilateral





Results, surgical (n = 18)



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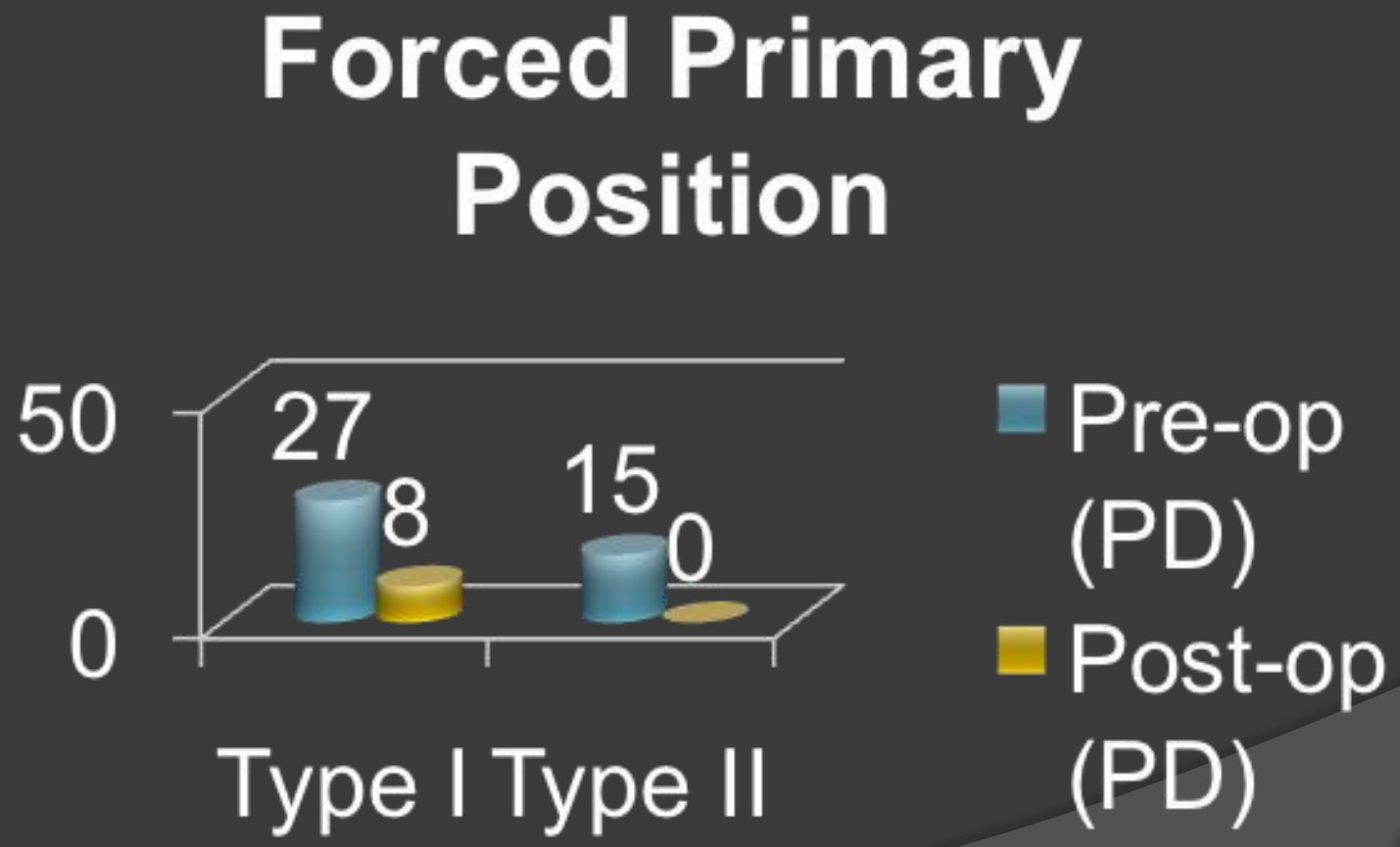

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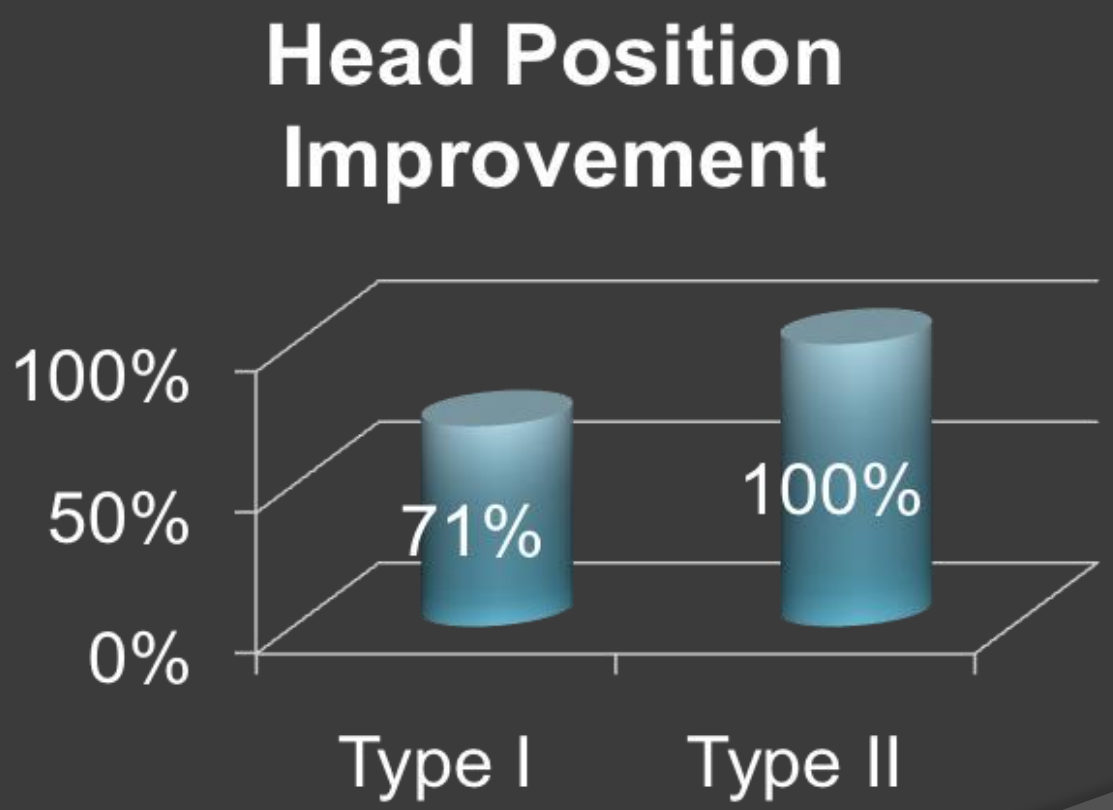
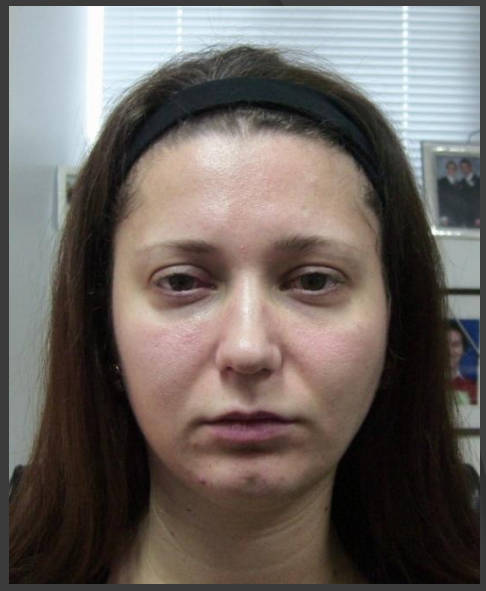
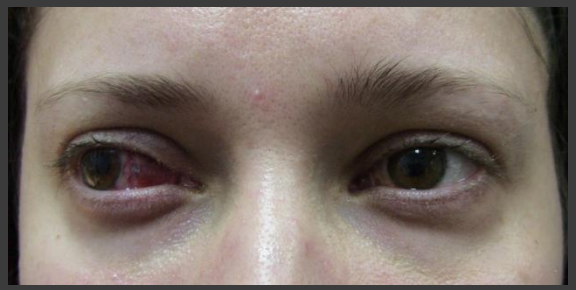


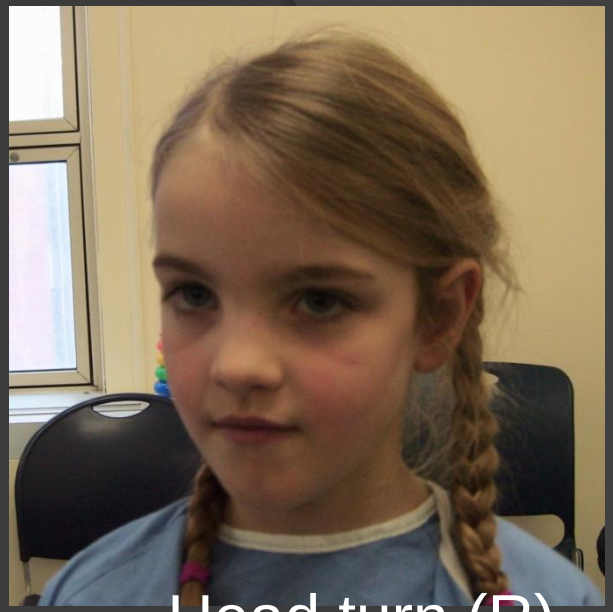
Duane type I -Preop

Duane type I - Post-op



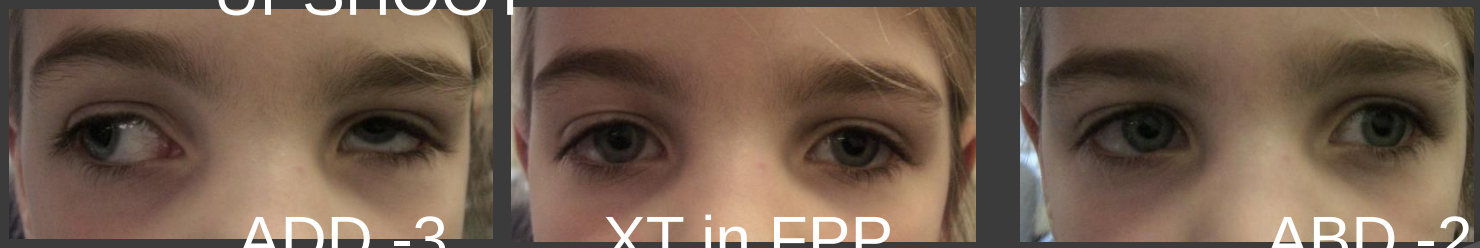


Duane Type II
Upshoot with “V” pattern



Head turn (R)

UPSHOOT



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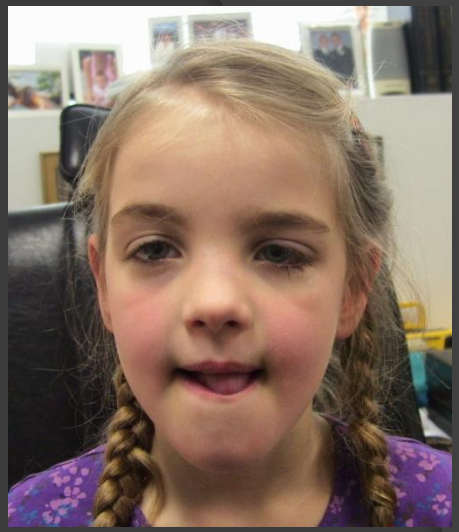

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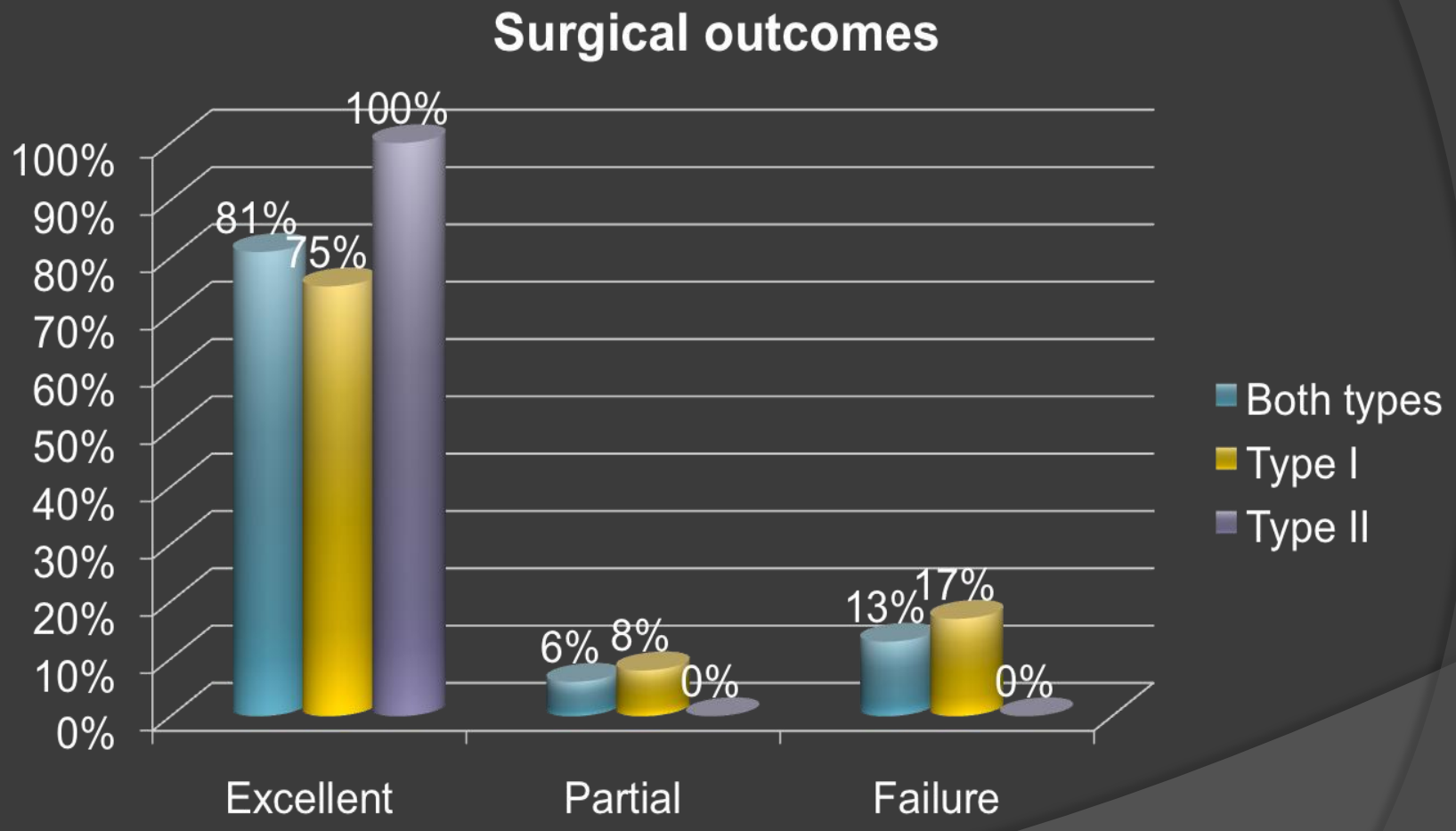

Discussion

- 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

- Kraft S.P.: A Surgical Approach for Duane Syndrome. *Journal of Pediatric Ophthalmology & Strabismus* 1998; 25:119-130
- Chung M. Stout JT. Borchert MS:Ophthalmology. 107(3):500-3, 2000 Mar.

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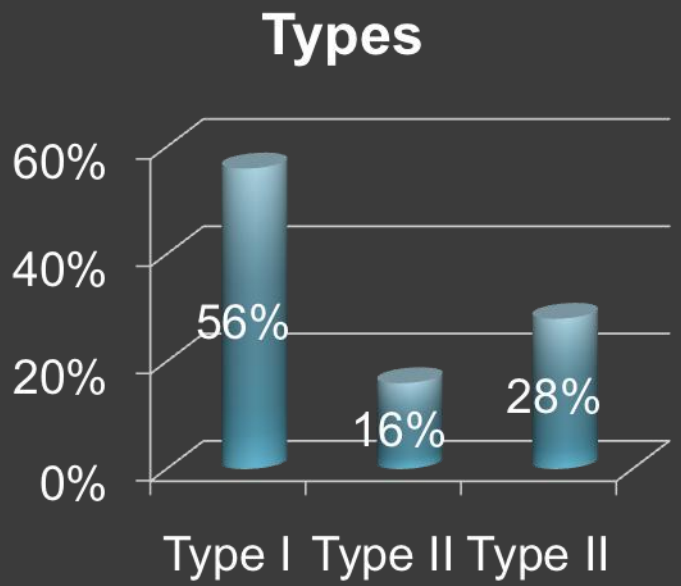

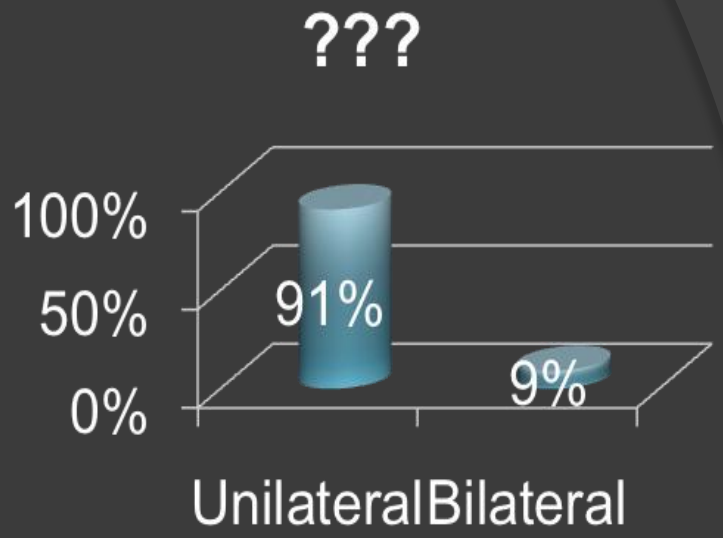

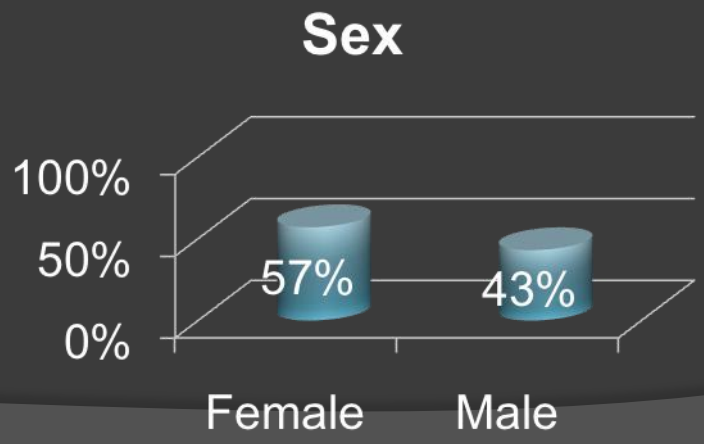

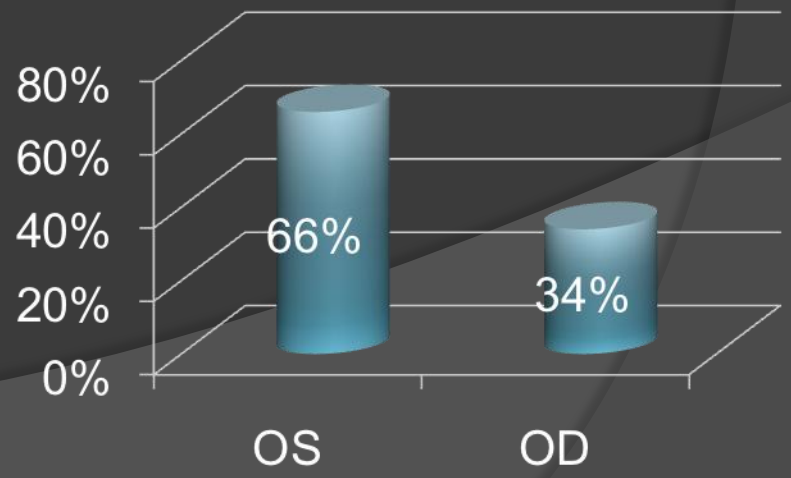

Results, observational