

AVC 2017

***Surgical treatment of childhood strabismus***

**LIONEL KOWAL MELBOURNE**



This is Amy getting ready to go to sleep for surgery. Flat James on the other hand

# TODAY'S TALK

- If / when / why surgery is a good next step for your child
- Esotropia ET
- Exotropia XT

## Complex strabismus

- Cyclovertical problems CV
- Developmentally Delayed Kids DDK

# Why surgery?

The child has symptoms or signs that surgery can be expected to improve & after a discussion about:

- Benefits
- Risks
- Hassle / Costs
- Alternative treatments

....I proceed, with the parents' blessings

# Parents' expectations have to = mine I

- Realignment fixes part - a large necessary part, but only a part - of the problem
- Often, the only reliable outcome is improved appearance

# Parents' expectations have to = mine 2

- ET: improved alignment: improved field
- Perfect alignment necessary for 3D
- Glasses may still be needed
- Amblyopia Rx may still be needed and may be more effective if the eyes are straight[er]

## KEY TO SUCCESSFUL MNGMT OF PARENTS' EXPECTATIONS: **EDUCATION**

- ***'HIGHLY RECOMMENDED [FREE!] E-BOOK FOR YOU TO READ'***
- **Eye Muscle Problems in Children and Adults: A Guide to Understanding**
- **Burton J. Kushner, MD**  
The John W. and Helen Doolittle Professor of Ophthalmology  
University of Wisconsin Department of Ophthalmology and Visual Sciences, Madison  
*...LINK ON MY WEBSITE*

## Burton J. Kushner, MD

- If knowledge is power, one of its powers is to enable us to make wise and informed decisions that influence our future.
- Hopefully after reading this book you will feel more empowered to make considered choices regarding the treatment of your child, yourself, or your loved one.

# Overview of Surgery expectations

## Two main groups

- Low risk for 2<sup>nd</sup> surgery, good prognosis for some sensorimotor fusion.
- Higher risk [for 2<sup>nd</sup> surgery] & poorer prognosis for some sensorimotor fusion.





Low risk for 2<sup>nd</sup> surgery

Good prognosis for some sensorimotor fusion.

- Acquired ET.
- Stable measurements.
- Pre-op sensory assessment suggests postop fusion [PAT 1987 study].
- Many cases of intermittent XT
- Healthy child



Higher risk for 2<sup>nd</sup> surgery

Poorer prognosis for some sensorimotor fusion

- Infantile onset strab [IOS].
- [Apparent] Cyclo Vertical Problems CVP
- Delay till 1<sup>st</sup> straight.
- Developmentally Delayed Kids DDK.
- ASD/ADHD
- Many cases of intermittent XT
- Huge misalignments
- Recurrent strabismus

# These parents need LOTS of time

- Parents with expectations that will never be met: expect / demand that one surgery will give a perfect cure - perfect alignment, appearance, 3D
- Child has had unconventional ineffective treatment for some years : need total recalibration of parental 'religion'.
- Albinism: +ve angle Kappa common: when aligned, look XT - beware

# Mentioning disaster outcomes:

## Tailor to parent

- Most: Surgery is 99+% safe. Do you want more detail?

Some parents:

- Anesthetic disaster 1/100,000
- Blind [usually infection] 1/10,000 – I have never seen it in Melbourne
- Pedestrian/ passenger in a car: death rate 1/20,000 pa
- New discussion: developmental problems after general anesthesia in young children - several references on my website

# DEVELOPING AN ESOTROPIA...

THE UNCORRECTED HYPEROPE or  
'Convergence Excess' aka High AC/A

Prolonged inappropriate  
accommodation → tendency to  
inappropriate convergence and  
**increased tone in medial recti**  
[vergence tonus]

# Developing an esotropia...2

- Increased tone will lead to changes in Tension / Length ratio and eventually to structural changes inc changes in sarcomere density in MR that eventually exceeds motor fusional reserve and → **esotropia!**
- Then the MR starts to permanently shorten

# Esotropia

- Core defect becomes a mechanically abnormal MR.
- Making the MR mechanically normal is a necessary part of the fix
- Accurate measurements, accurate surgery: early alignment success in 90%

# BENEFITS OF REALIGNMENT OF ET

- Normal appearance
- Better peripheral field
- Chance for sensory fusion
- Better chance to treat resistant amblyopia



# TIMING OF ESOTROPIA SURGERY

- Von Noorden
- Ing
- Europe
- Wright
- Costenbader
- Parks
- Tychsen & the Texans

...so many eminent contributors for decades – nearly all point to early or very early alignment as important

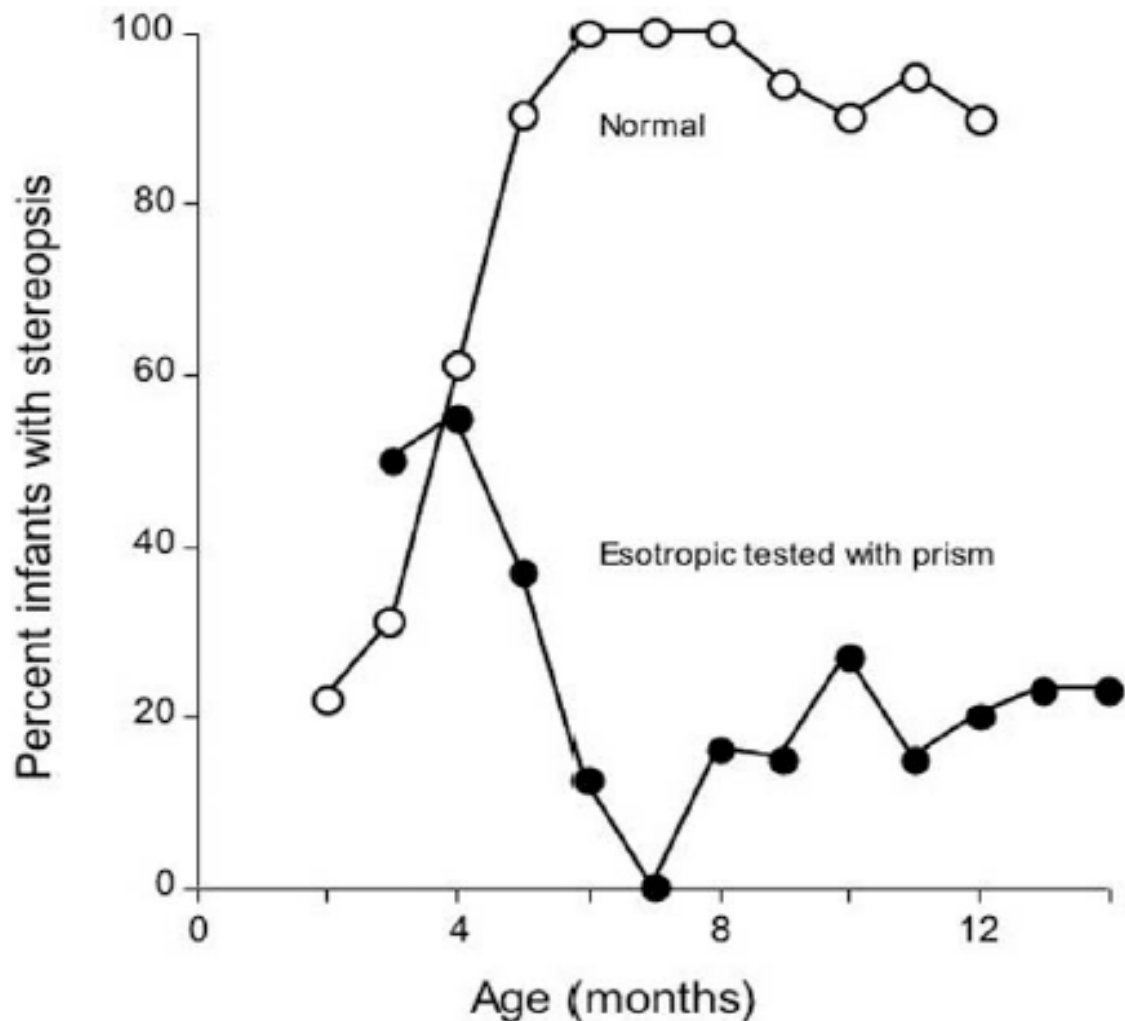
Benefits : Appropriate surgery  
makes for better neurology

*Marshall Parks Lecture*

Can Ophthalmologists Repair the Brain in  
Infantile Esotropia? Early Surgery,  
Stereopsis, Monofixation Syndrome, and the  
Legacy of Marshall Parks

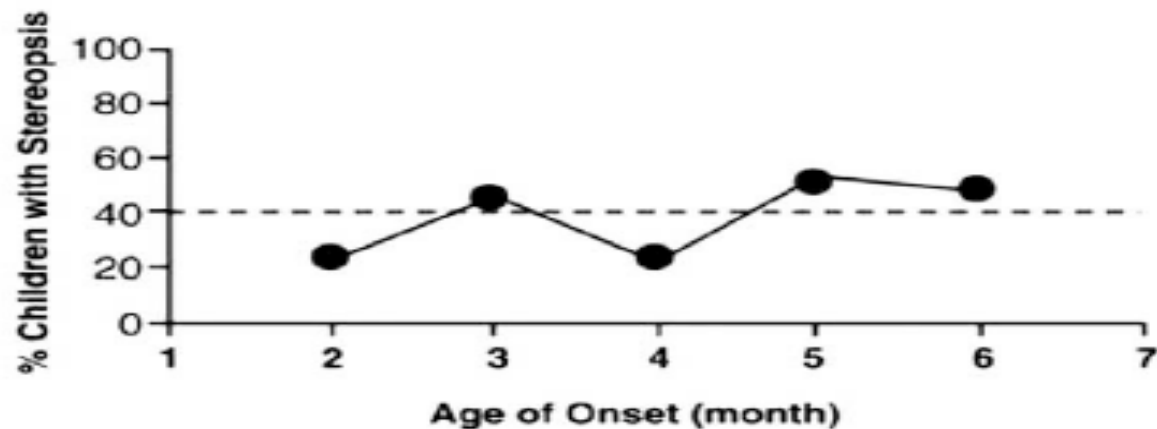
Lawrence Tychsen, MD

JAAPOS 2005

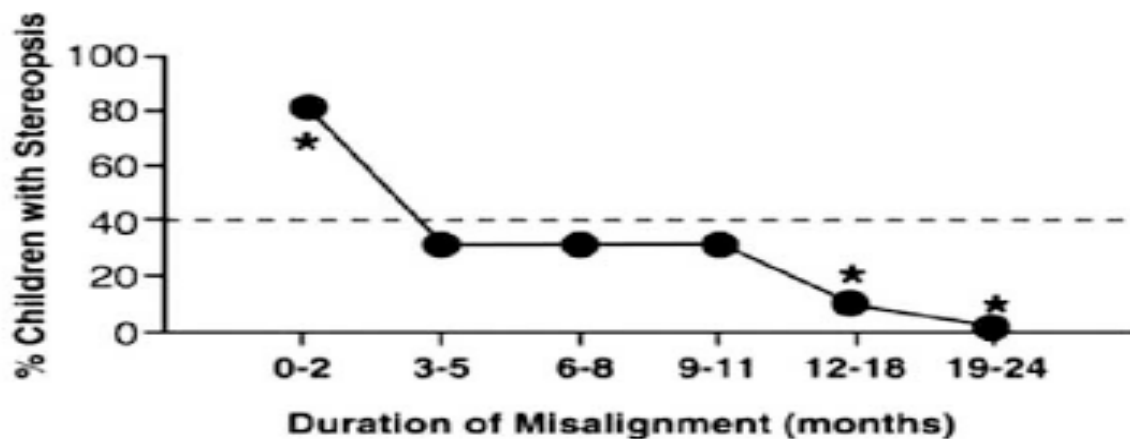


**FIG 4.** Prevalence of stereopsis as a function of postnatal age in a population of normal ( $n > 50$ ) versus esotropic infants ( $n = 85$ ). Tested using dichoptic viewing (polarized goggles and images) by the preferential looking method. Esotropic infants were aligned using prisms and tested before any surgery. Data replotted from Birch and Stager<sup>6</sup> and Stager and Birch.<sup>10</sup>

### A Stereopsis vs Age-of-Onset



### B Stereopsis vs Duration



**FIG 5.** Prevalence of stereopsis after surgical realignment in children with infantile esotropia as (A) a function of age-of-onset of esotropia and (B) as a function of duration of esotropia before realignment. Testing performed at age 5 years. Surgical realignment achieved generally by age 1 year for the population as a whole. Dashed line at 40% indicates average prevalence for all the infants.

# Surgery timing

ET: Infantile onset, Acquired

- Improved % of good alignment and sensorimotor fusion if realigned within some months in all cases of ET inc IOS

# Exotropia

- Infantile: common association with developmental issues
- Older: intermittent XT
- Sensory XT
  
- Indications for surgery less well defined than for ET

# EXOTROPIA XT

- Core problem is usually **subtle anomaly in orbital anatomy**, not a tight LR
- A common 2<sup>o</sup> problem:  
hemiretinal suppression that 'allows' XT without diplopia

Fixing the LR length & tension tries to compensate for the XT and improve the alignment & mechanics, but:

- 1. does not return the mechanics of this abnormal orbit to normal - this 'allows' some recurrence of XT
- 2. may not alter the suppression pattern even when straight - this 'allows' recurrent XT

# ESOTROPIA & EXOTROPIA

## ET & XT

### ET:

- core problem is [or becomes] a tight medial rectus, driven by normal or Xs accom convergence

### XT:

- core problem is usually subtle anomaly in orbital anatomy [*not a tight lateral rectus*] &/or
- ‘soft’ neurological issues &/or
- sensory adaptation to the XT
- **ET / XT ARE NOT MIRROR IMAGE CONDITIONS**



# Exotropia

For this next child:

- when and for how long do I use over-minus?
- when should I refer for surgery?
  
- EBM = Evidence Based Medicine
- ..or is it Eminence Based Medicine

LK: Refer if:

- A cosmetic issue [ XT or MEC]
- Spectacle intolerance

## BASICS OF TREATMENT : XT SURGERY

>50% early ET [5-10 $\Delta$  desirable]  
<10% persistent ET  $\Rightarrow$  risk of  
amblyopia / troublesome  
diplopia depending on age

*Some sense in deferring surgery  
till out of the amblyogenic age,  
hence minus lenses*

# Exotropia: surgery results

- 2<sup>nd</sup> surgery
- MI 2%
- YI 10%
- YIO 30% [Baker]
  
- Regain sensory fusion: many
- Lose sensory fusion: some

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## ***Surgical treatment of childhood strabismus***

- Real and pseudo- oblique dysfunctions are common accompaniments of childhood ET [& XT].
- Add an extra layer/s of complexity to cause, natural history, treatment planning and expectations

# APPARENT ABNORMAL OBLIQUE ANATOMY / FUNCTION



# CYCLO VERTICAL PROBLEMS

- REAL AND PSEUDO – ALL LOOK THE SAME
- There are subtle ways to tell the differences:
- Fundus torsion
- Commutative vs non-commutative versions
- Imaging

**SOME:  
TRULY ABNORMAL OBLIQUE ANATOMY / FUNCTION**

The 4 oblique muscles need to be ***built, grow and work in perfect 3D symmetry.***

At BEST they are very finely tuned with little room for error, hence vertical fusional range only  $\pm 2-3 \Delta$ .

Any imperfection will interfere with motor fusion, and predispose to tropia – if hyperopic, ET

Extreme example: Cong sup obl palsy

# ABNORMAL OBLIQUE ANATOMY / FUNCTION – NON PARETIC WALTER FINK 1954! ANATOMY TEXTBOOKS OFTEN WRONG

20% of cadavers:  $> 30^\circ$  difference b/w course of SO & IO

Trans Am Ophthalmol Soc. 1954; 52: 305–350.

PMCID: F

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## The Role of Developmental Anomalies in Vertical Muscle Defects

Walter H. Fink

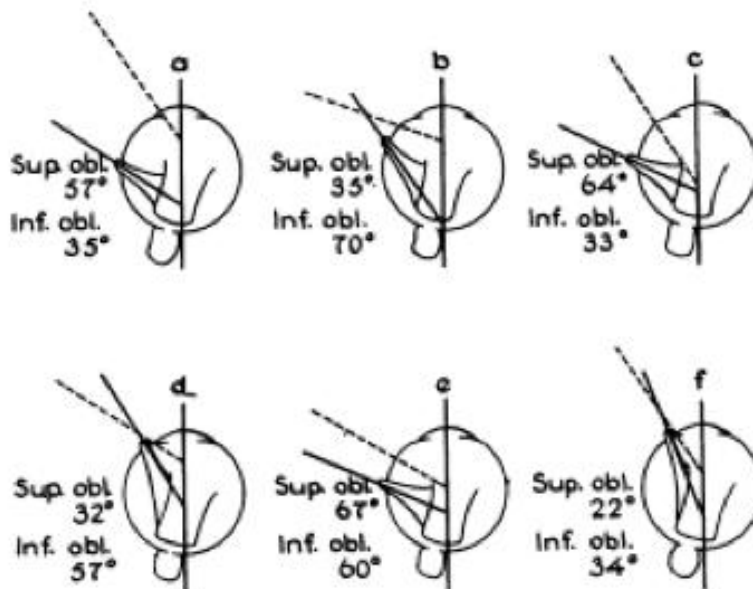


FIGURE 5. DRAWINGS OF SPECIMENS IN WHICH THERE ARE PRO-  
NOUNCED VARIATIONS OF THE OBLIQUE MUS-  
CLE PLANES OF ACTION. Continuous line indicates  
the superior oblique plane of action. Broken line in-  
dicates the inferior oblique plane of action.



# SUBTLE ABNORMALITIES IN ORBITAL ANATOMY THAT CAUSE ABNORMAL OBLIQUE FUNCTION

Unicoronal synostosis [ premature fusion of a coronal suture] : ~  
**slightly misshapen forehead.**

**Apparent IO OA ~50%**

Manifest strabismus in primary >**50%**

BAGOLINI 1982:  
Isolated posteroplaced  
trochlea is an  
underrecognised cause of  
idiopathic oblique  
dysfunction

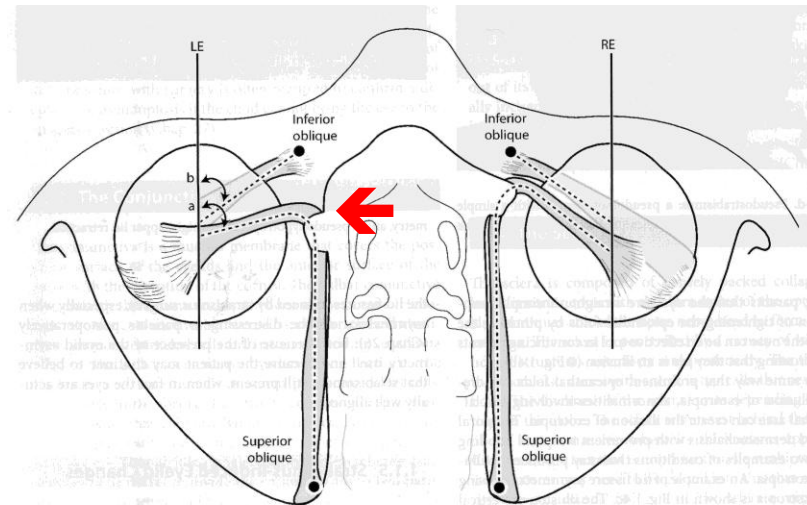
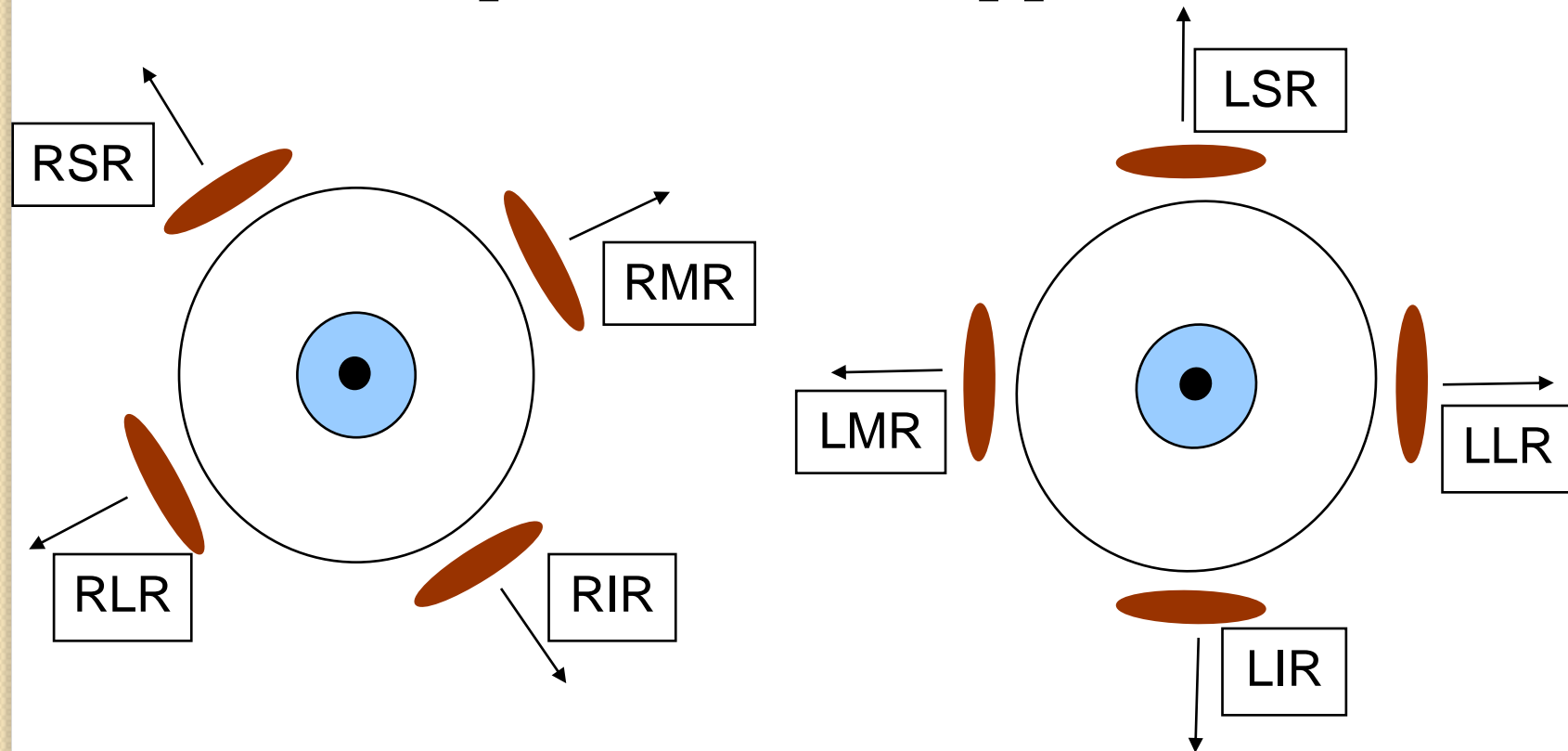


Fig. 1.3. Failure of the trochlea to advance anterior to the equator in a patient with unilateral coronal synostosis may result in reduction of d pressing action on the globe with contraction of the superior oblique muscle

## EXTORTED ORBIT

- Extorted right orbit and globe will cause a V-pattern and apparent IO-OA



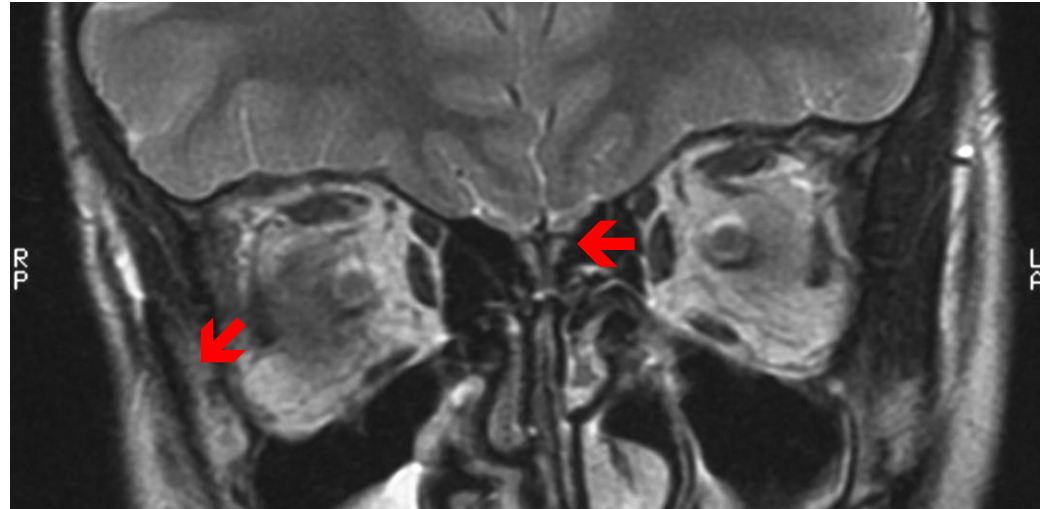
## ORBITAL PULLEY HETEROTOPY

RLR lower than RMR

R gaze: RLR will pull  
RE to R & **down**

LMR will adduct on the  
horizon: LE will then  
be higher than RE:  
**Resembles LIOOA**

Will be no fundus  
torsion: LIO surgery  
not expected to be  
effective





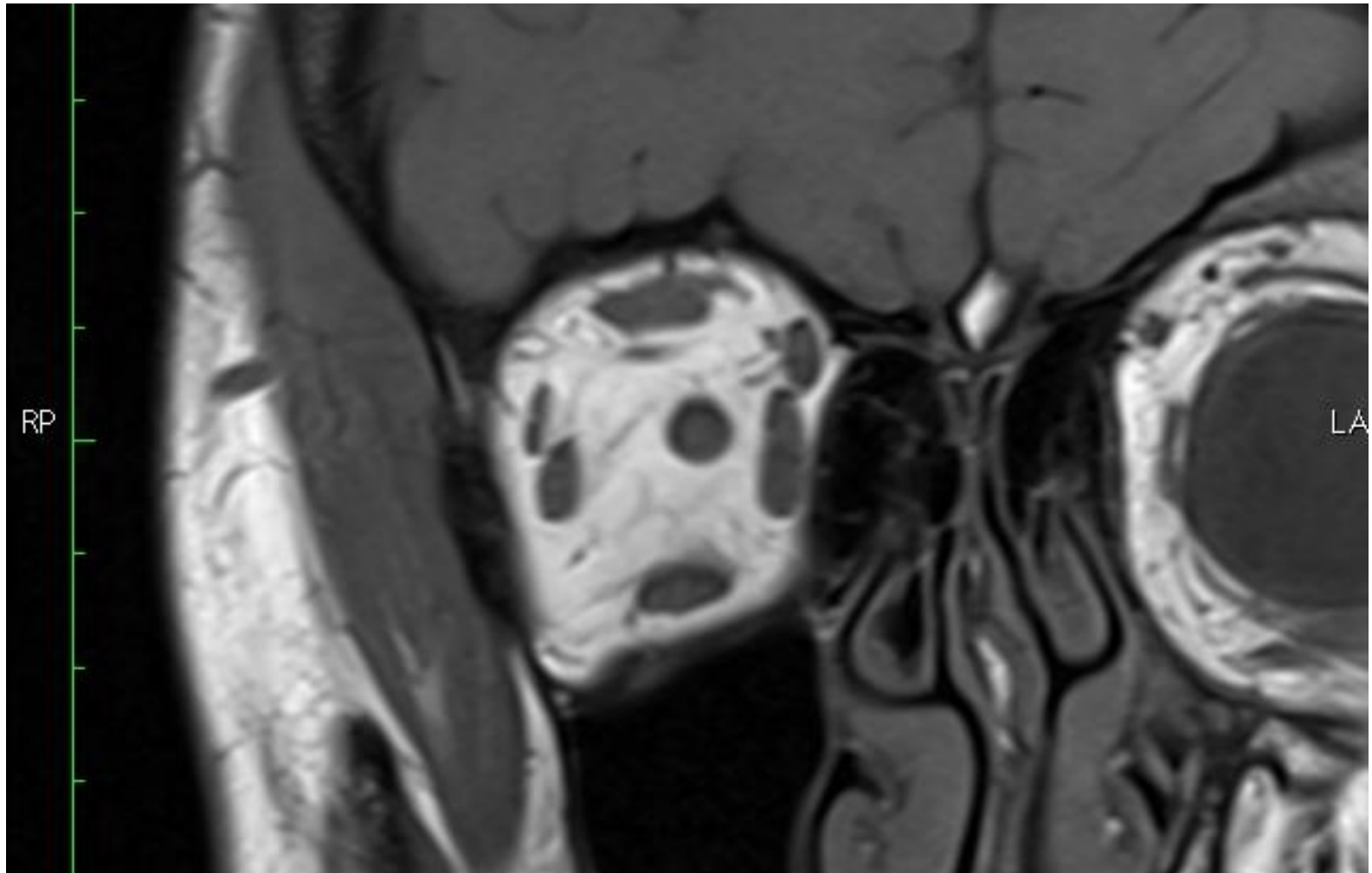
RIGHT HYPERTROPIA



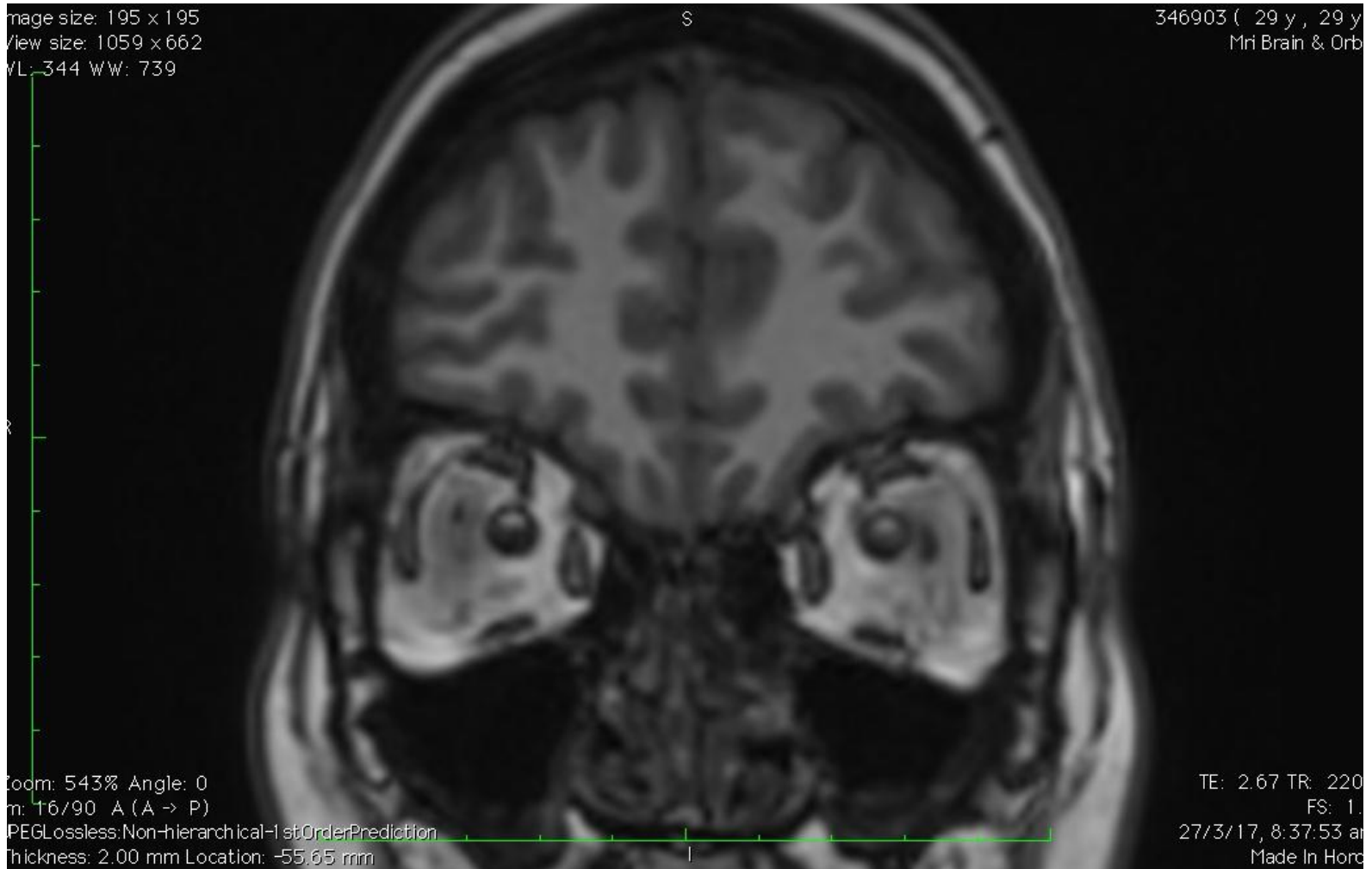
OVERELEVATION IN ADDUCTION

**IDIOPATHIC RIGHT INFERIOR OBLIQUE OVERACTION?**

# 'RIO OA' DUE TO BIFID RLR



# A – XT : superiorly displaced LR's



# Real and pseudo- oblique dysfunctions

- ....are common accompaniments of childhood ET [& XT].
- Add an extra layer of complexity to cause, natural history, treatment planning and expectations
- ***Some definitely get better with time***

## Preparation for the hospital experience

### My website:

- 1. Ella's Eye Surgery Experience
- 2. Amy's adventure.
- 3. Gabriel's Eye Surgery Adventures
- 4. Briannah's Book
- 5. Kara's adventure
- 6. Noah's adventure



# Hospital experience

View Kara's visit to the Eye and Ear:



# Special situations

- Incomitant
- Duane's Syndrome
- Trisomy 21
- Other developmentally delayed kids  
DDK
- Getting out of glasses

# Special situations

## Incomitant:

- Distance/ near
- Left / right
- Up / down
- All need an explanation, an understanding, and tailored surgery

# Special situations

## Duane's Syndrome

- Complex++
- Many false +ve & -ve diagnoses in Y I
- Surgery still evolving: different transpositions helpful in many pts

# Special situations

## T21/ Downs

- Expectations of surgery close to 'normal' kids

# Special situations

## Other developmentally delayed kids DDK

- Typically need 2-3 surgeries to achieve good stable alignment

# Special situations

## Getting out of glasses

- ET with full +, &  $<+2.50$  DSOU : realistic to aim for uncorrected ET. Often successful
- Kids who still need full+ @ age 8 for normal alignment & are +3 DSOU or less: 50+% will grow out of glasses in next 10y.
- Measure BIFR for quantitative guidelines



*That's all Folks!*

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wallpapers