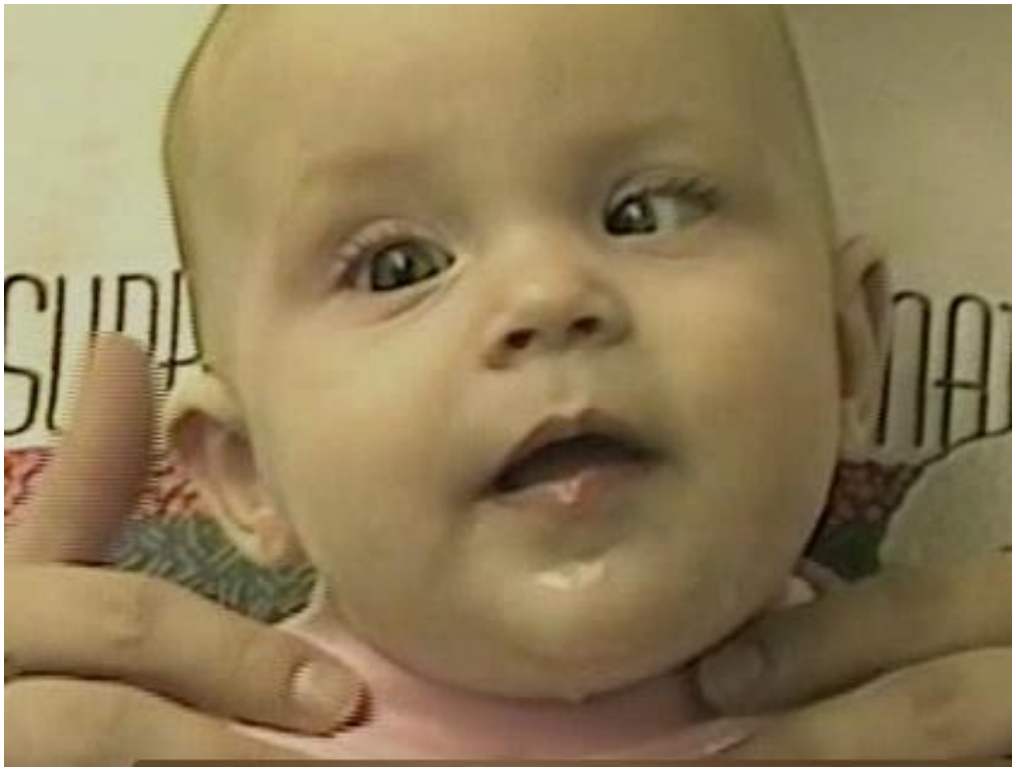
CHILDHOOD ESOTROPIA 2020

LIONEL KOWAL



Childhood Esotropia ET

..is all about:

1. Abnormal medial rectus : stiffer / tighter ± eventually shorter

2. Abnormal convergence, often driven by under- corrected hyperopia

Childhood Esotropia ET

‘*Abnormal convergence, often driven by under corrected hyperopia*‘

is the usual cause of

‘*Abnormal medial rectus : stiffer /tighter ± shorter*‘

There is another (less) common cause: the aDduction null of the type of nystagmus seen with Infantile Onset Esotropia IOE - *more later*

Some terminology

- Phoria
- Tropia
- Motor fusion

Esodeviation
Phoria - Tropia

Eso**tropia** ET

- Obvious misalignment
- Demonstrated by cover test
- Caused by a tighter &/or shorter MR

Eso**phoria** E

- Pre-tropia
- Esodeviation not obvious – 'hidden' from view by motor fusion [=orthotropia]
- *Only becomes apparent with a search using a dissociating examination technique: alternate cover test, Maddox rod, prism challenge, Hess test, prolonged patching…*

Motor fusion

- The ability to keep the eyes straight despite attempts to disrupt the alignment esp with a prism
- Here, using BI Δ to measure the BIFR for near

Motor fusion

- Using BI Δ to measure the BIFR for near measures the pre-disposition to ET
- Normal >20 Δ
- Safe > 5-6 Δ
- ‘Almost ET’ 1-2 Δ

Childhood Esotropia ET 1

There are 2 main groups:

- Infantile Onset ET = IOE

= ET caused (?largely) by the effects of persisting immature supranuclear eye muscle control

Childhood Esotropia ET 2

There are 2 main groups:

- Infantile Onset ET = IOE = ET caused by persisting immature supranuclear eye muscle control

- Acquired ET in a child who does not have Infantile Onset Strabismus,

…or who previously had successful alignment surgery for IOE

& a tiny scary 3rd group: ACUTE ESOTROPIA …… *More later*

ESOTROPIA ET
Final common pathway

- Common factor in ALL ET is increased tension in the medial rectus (abnormal T-L ratio) and eventually a TIGHT & shorter MEDIAL RECTUS.
- Shorter = tropia
- Tighter = phoria or tropia

ET: TIGHT MEDIAL RECTUS
Group 1: Acquired ET

How do we end up with a tight MR:

- **1A: Increased demands on horizontal motor fusion**, &/or
- **1B: Reduced quality of horizontal motor fusion**

…will eventually change the T-L ratio in the MR & cause a tight MR

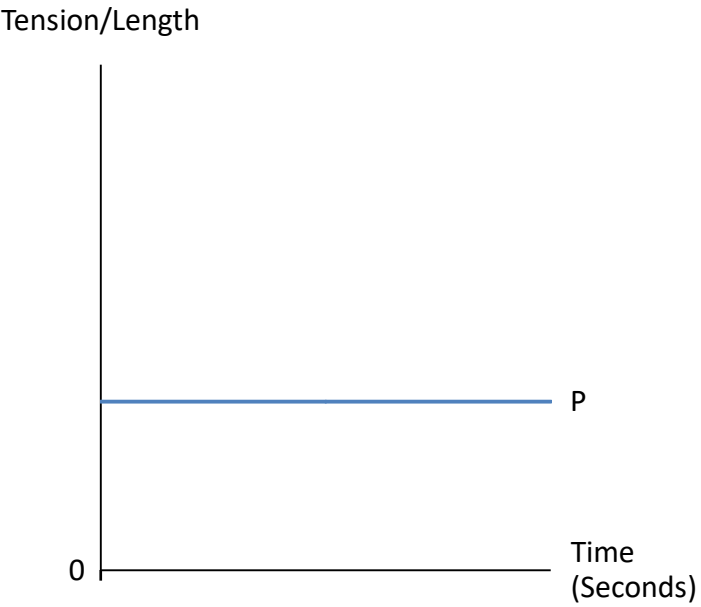
ET: TIGHT MEDIAL RECTUS
Group 2: IOE Infantile Onset Esotropia

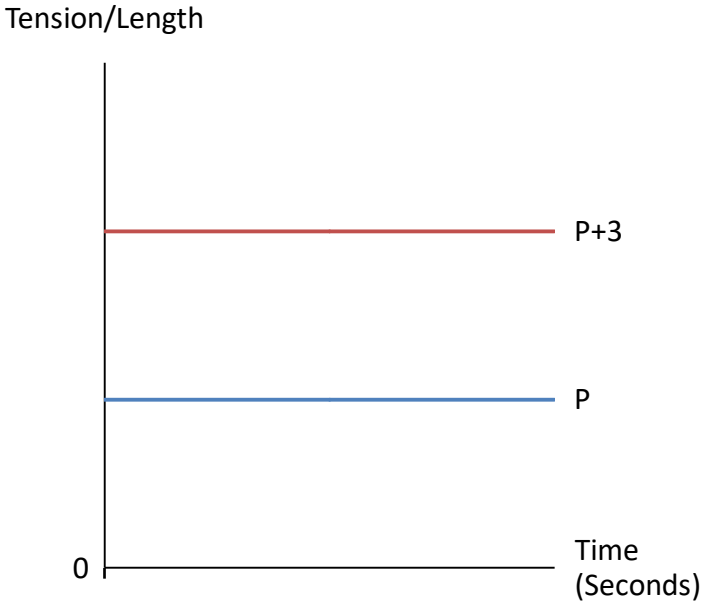
- These pts have reduced potential for excellent sensory-motor fusion.
- The IOS-associated nystagmus [FMN - see later] has an **aDduction null** & this probably drives the ET
- ± Neo-natal & infantile hyperopia [25% normal Caucasian babies ≥+4DS before age 3mo]

This lecture

- Why do we get ET
- Important associated findings
- When to treat
- How to treat
- Expectations of treatment

How to get a tight MR: **Need persistent changes to the T-L curve**

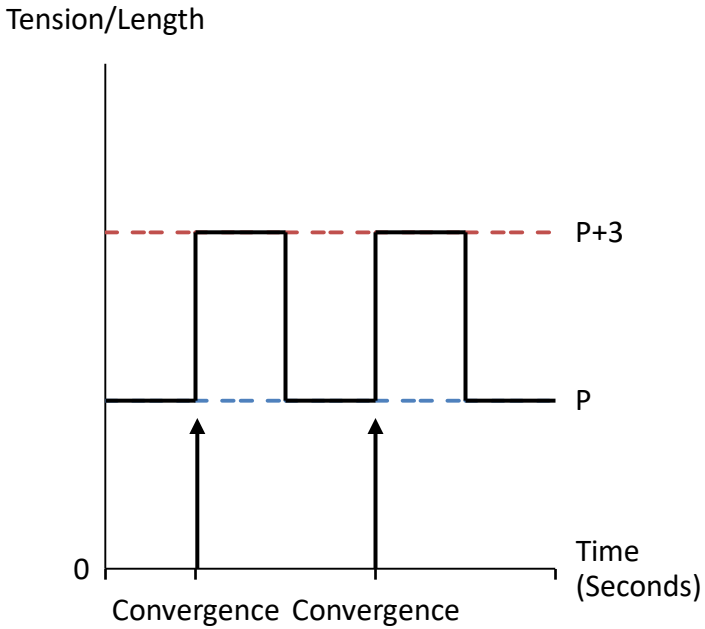




Distance viewing
Plano refraction
Normal T-L ratio

Distance viewing +3 refraction
Increased (accommodative) convergence
More T per L
Abnormal T-L ratio

Repeated near viewing for seconds or minutes



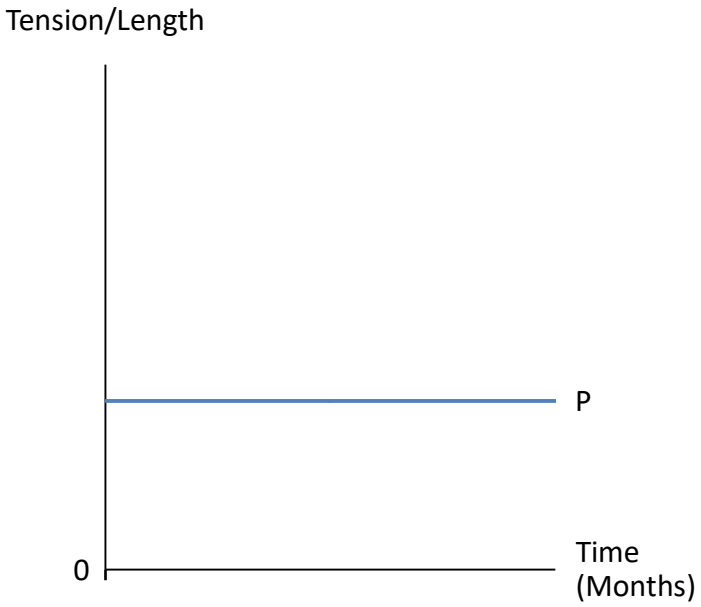
Plano refraction Normal T-L ratio for distance

Appropriately elevated T-L ratio when converging

This does NOT cause permanent long term changes in the T-L ratio

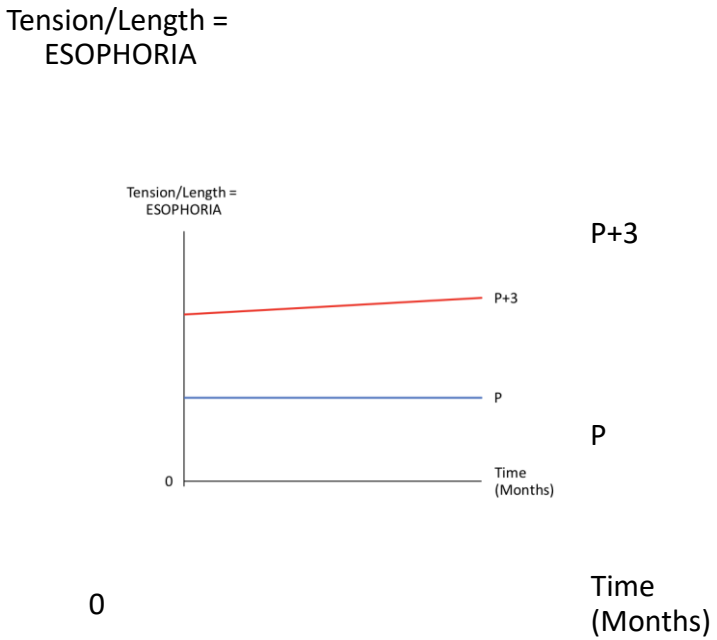
Increased T-L ratio when converging can be minimised with extra + for near

How to get a tight MR



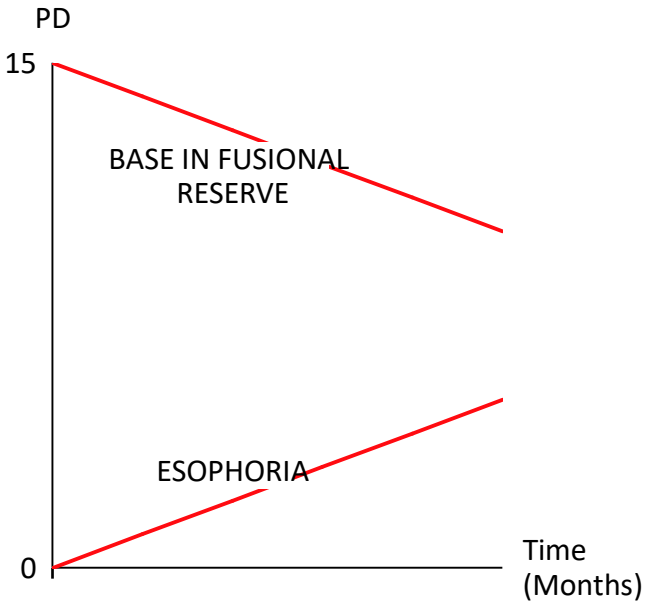
Distance viewing
Plano refraction
Normal T-L ratio

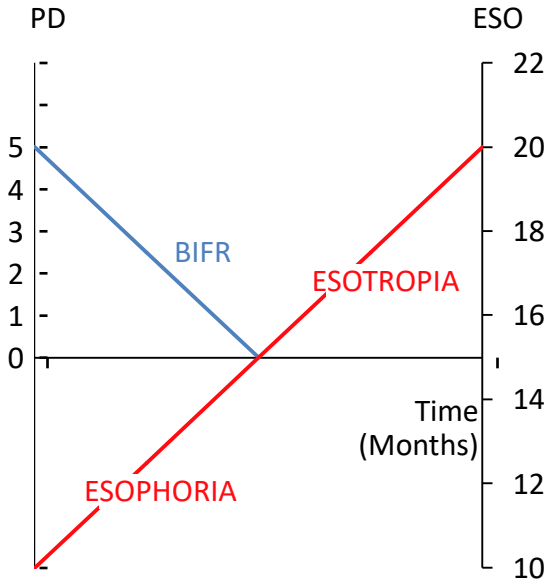
Uncorrected +3 refraction for <u>months</u>



Distance viewing
Progressively abnormal T-L ratio = increased stiffness of MR = esodeviation develops

Progressive mechanical changes to the MR muscle in an uncorrected +3 hyperope

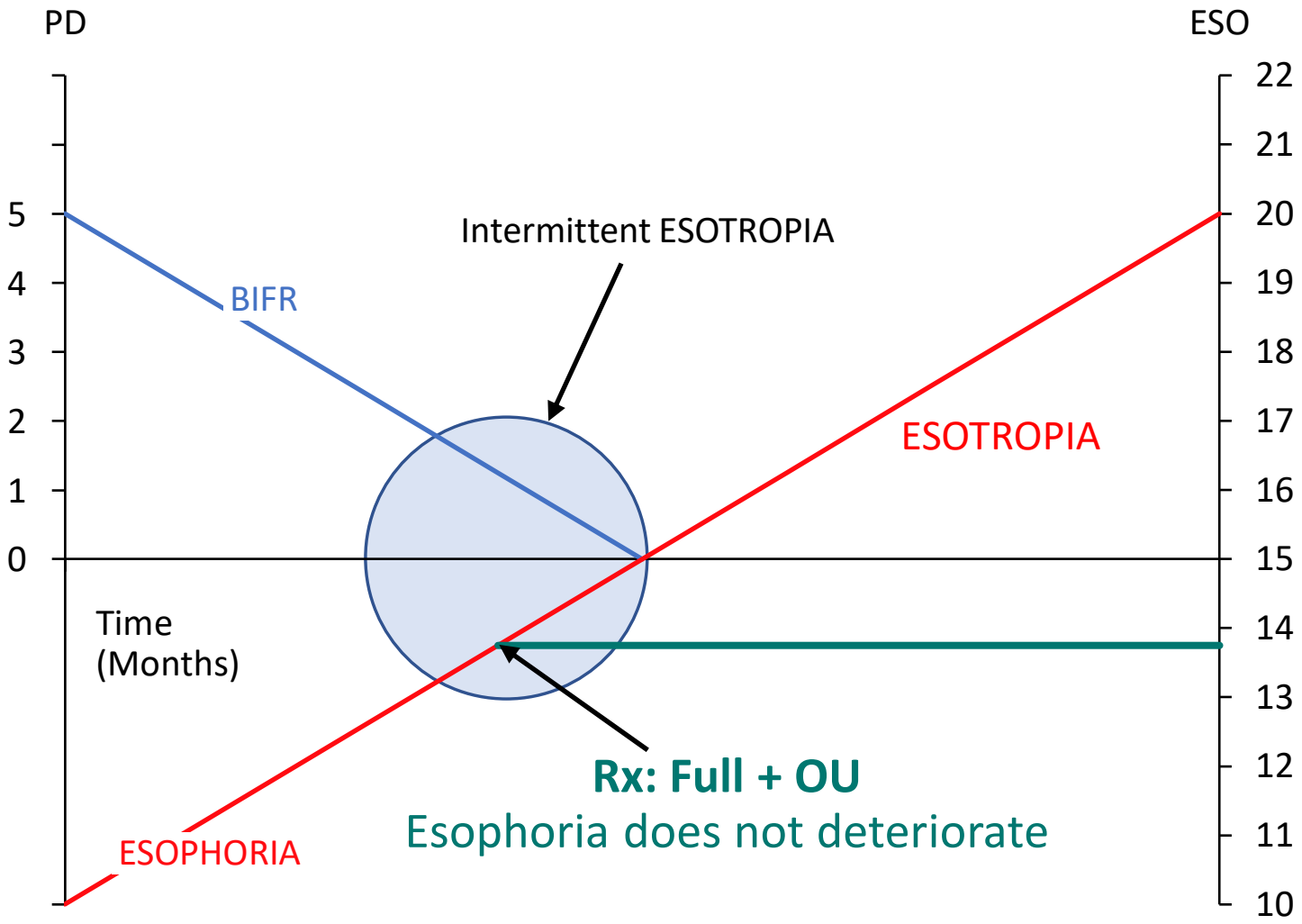




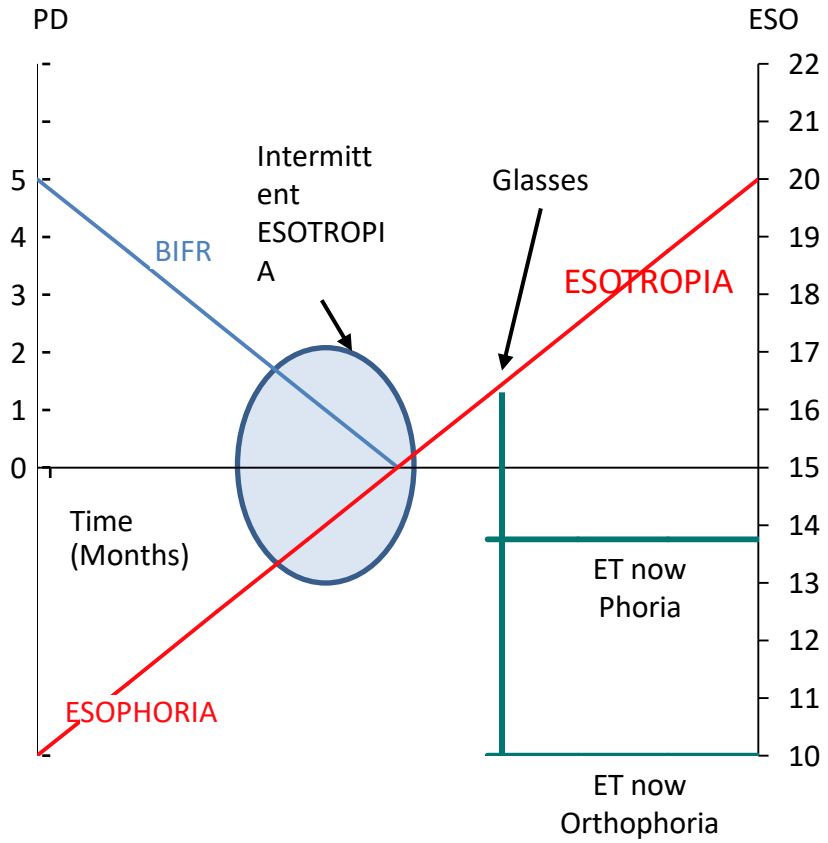
As MR gets stiffer, the esophoria gets worse & the BIFR gets less

As esophoria gets worse, the BIFR gets less.
When BIFR approaches zero, the esodeviation changes from esoPHORIA to esoTROPIA

+ prevents deterioration



Giving + ‘Cures’ ET = Refractive ET



Giving the + converts Esotropia into an asymptomatic Esophoria (= Orthotropia) or (even better) Orthophoria: **Spectacle ‘Cure’**

‘Rescue’ ET with ≥ +6 DS

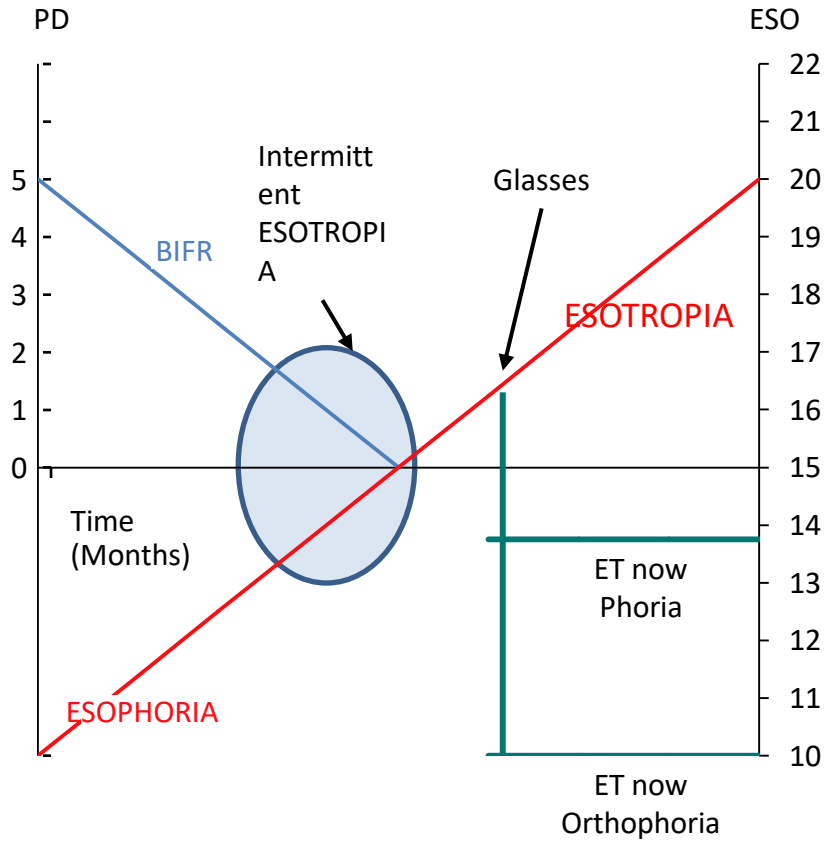
Cohort of ET with high +

Kowal, Poon, Chen

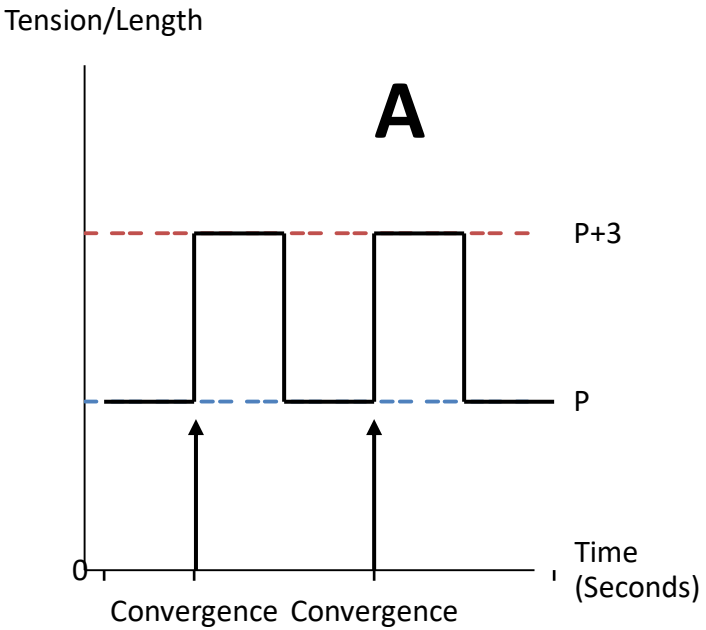
ET, ≥ +6 DS n=86

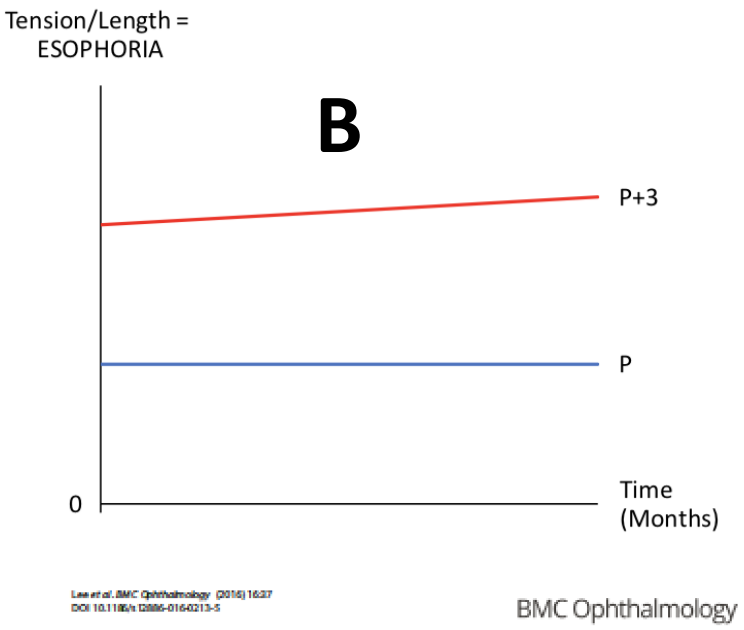
Initial result orthotropia:
Subsequent need for
ET surgery in only 2.5%

Mulvihill 2.4%



Can using a Smartphone for 6h a day change the T-L ratio? Maybe….





A

Intermittent near viewing

Normal T-L ratio for distance & increased for near

B

If converges constantly, will be like uncorrected +3 & develop progressive eso .

Lee et al. BMC Ophthalmology (2016) 16:27
DOI 10.1186/s12886-016-0213-5

BMC Ophthalmology

RESEARCH ARTICLE Open Access

Acute acquired comitant esotropia related to excessive Smartphone use

Hyo Seok Lee, Sang Woo Park and Hwan Heo*

Abstract

Background: To describe the clinical characteristics and outcomes of acute acquired comitant esotropia (AACE) related to excessive smartphone use in adolescents.

Methods: The medical records of 12 patients with AACE and a history of excessive smartphone use were retrospectively reviewed, and the duration of smartphone use, angle of deviation, refractive error, stereopsis, and treatment options were analyzed.

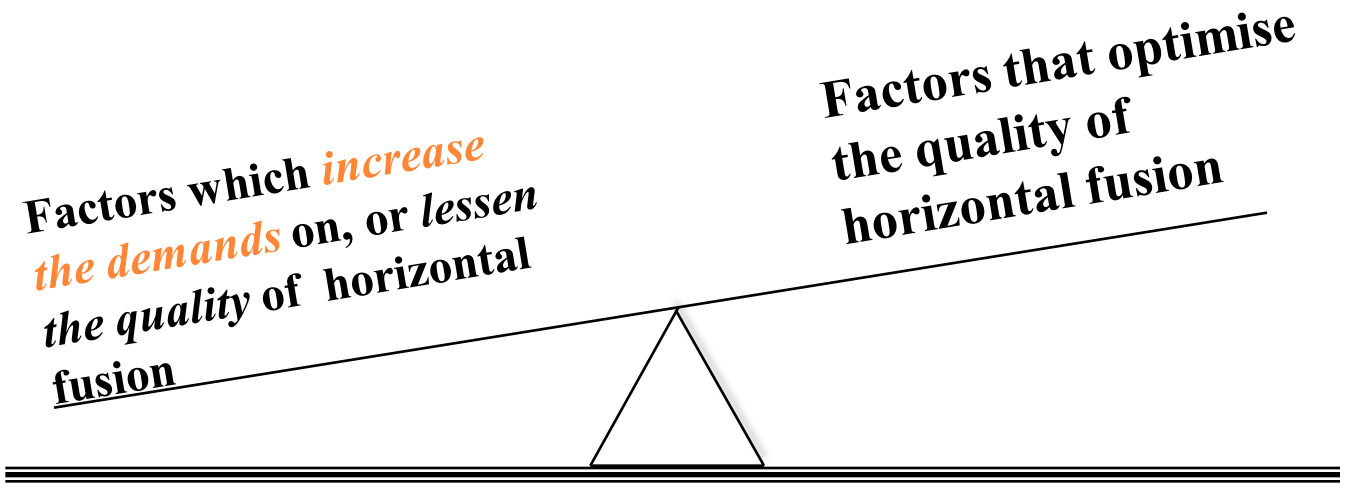
Results: All patients showed convergent and comitant esotropia ranging from 15 to 45 prism diopters (PD; average: 27.75 ± 11.47 PD) at far fixation. The angle of deviation was nearly equivalent for far and near fixation. Every patient used a smartphone for more than 4 h a day over a period of several months (minimum 4 months). Myopic refractive errors were detected in eight patients (average: −3.84 ± 1.68 diopters (D)), and the remaining four patients showed mild hyperopic refractive error (average: +0.84 ± 0.53 D). Reductions in esodeviation were noted in all patients after refraining from smartphone use, and bilateral medial rectus recession was performed in three patients with considerable remnant esodeviation. Postoperative exams showed orthophoria with good stereoacuity in these patients.

Conclusion: Excessive smartphone use might influence AACE development in adolescents. Refraining from smartphone use can decrease the degree of esodeviation in these patients, and remnant deviation can be successfully managed with surgical correction.

Keyword: Esotropia, Medial rectus recession, Smartphone, Video display terminal

1. Acquired ET
Pts who had/have good sensori-motor fusion potential
Not IOS

ET develops due to an imbalance between two groups of factors



If this side is heavier, there will be strabismus

If this side is heavier, there will be no strabismus

FACTORS THAT INCREASE THE DEMANDS ON FUSION

1. Hyperopia

INGRAM:

- ≥ + 3.50 DS in one axis @ age 12 mo:

50% risk of strabismus / amblyopia

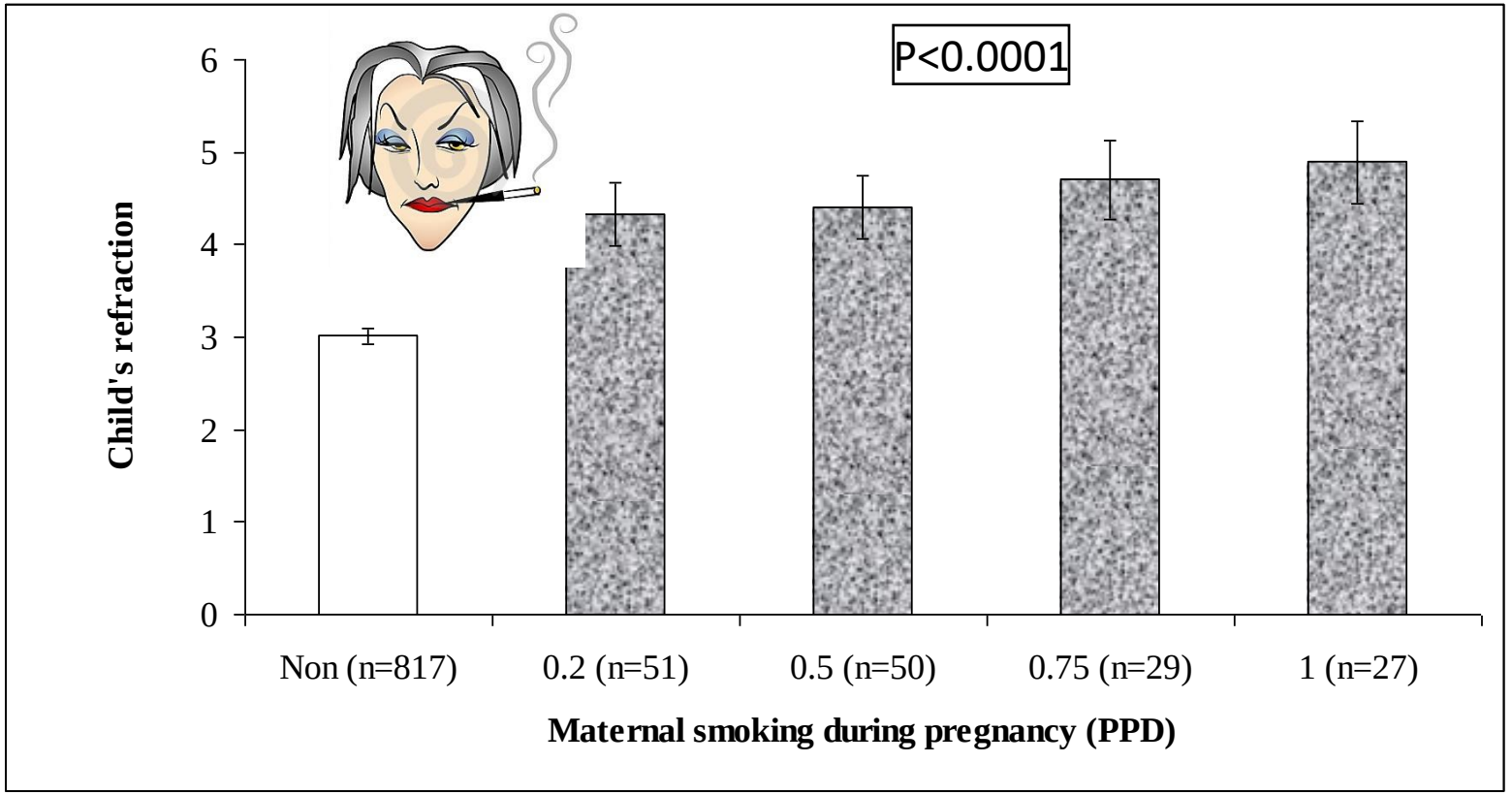
2. High AC / A

HYPEROPIA

- Hyperopia is present in some infants
- <3 mo: 25%!* ≥ +4 DS OU
- 12 mo: 5%
- Ethnicity affects prevalence
- Higher in certain subgroups esp. family history of hyperopia or accommodative ET.

* inc a cohort in China!

MATERNAL SMOKING DURING PREGNANCY INCREASES RISK OF CHILDHOOD HYPEROPIA [ISRAEL, 2012]



Morad et al, presented WSPOS Milan 2012

All ET: Need to find & give full +

Pre-subjective refraction age:

- Full cyclo refraction

Subjective refraction:

- Most + that doesn't cause distance blur

= **Manifest +**

- Giving an extra +0.50 DS: being a low myope is no disability as a child & a small price to pay if it fixes the alignment, or prevents deterioration

FACTORS THAT INCREASE THE DEMAND ON FUSION

ABNORMAL ACCOM - CONV RELATIONSHIP

- **High AC/A ratio**, abn CA/C ratio, proximal convergence, proximal fusion,.. all have precise definitions, but common usage is not precise.

- **USA: 'High AC/A'** = near eso > distance eso by ≥10Δ

- LK preference : **Convergence Excess** as synonym for all of these terms [after GvN].

BIFOCALS IN THE TREATMENT OF CONVERGENCE EXCESS

- Some uncertainty about using bifocals
- Selection bias: Average optometrist pt cf average pt attending strabismus clinic @ Boston Childrens*
- Typical Optometry pt with conv Xs ET: bifocals
- BCH pt: bifocals do not improve outcome
- LK preference : **Bifocals are used if they can be shown to improve sensory &/or motor status, and are dispensed, worn and used correctly**

**Whitman et al*

Bifocals fail to improve stereopsis outcomes in high AC/A accommodative esotropia *Ophthalmology April 2016*

ABNORMAL AC/A
Presbyopia

Second age peak for accomm ET

Seen in pts with impaired motor fusion eg previously straightened strabismus

Prsebyopia complicating pre-existing strabismus
Oystreck & Lyons
Can J Ophthalmol 2003

ABNORMAL AC/A
Drugs

- Drugs that interfere with accommodation e.g. Ditropan, some antidepressants / other psychotropics

Parents don't think of mentioning an enuresis tablet to the eye Dr

Oxybutynin-associated esotropia.
Wong EY, **Harding** A, **Kowal** L.
J AAPOS. 2007 Dec;11(6):624-5.
PMID: 18086433
Similar articles

ASD/ ADHD …. &/or their treatments

- **Labile convergence and accommodation** in many
- ASD + CP: increased ophthalmic morbidity
- May not accept / respond ‘normally’ to sensible glasses
- Surgery less reliable

J AAPOS. 2019 Dec;23(6):337.e1-337.e6. doi: 10.1016/j.jaapos.2019.09.008. Epub 2019 Oct 30.

Ophthalmologic disorders and risk factors in children with autism spectrum disorder.

Chang MY[1], Gandhi N[2], O'Hara M[2].

Screenshot ormation

Abstract

PURPOSE: To report the results of our review of all children with autism spectrum disorder (ASD) who underwent complete pediatric ophthalmologic examination at our institution over a 10-year period.

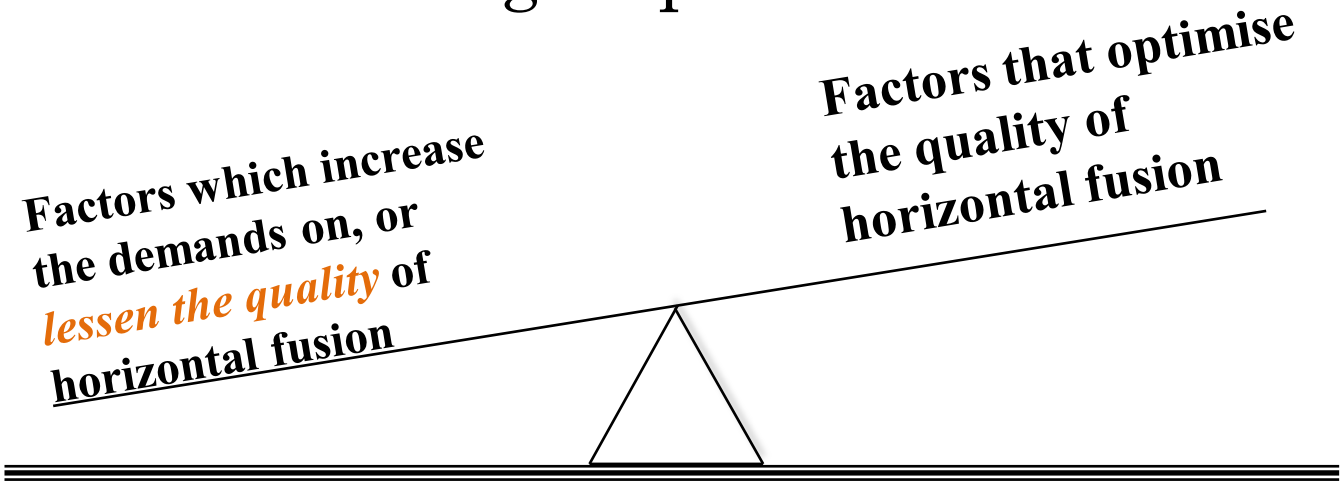
METHODS: The medical records of all children (0-17 years of age) with a diagnosis of ASD seen at University of California, Davis, over a 10-year period were reviewed retrospectively. Demographic data, birth history, genetic testing results, neuropsychiatric comorbidities, and ophthalmologic findings were extracted from the record. Multiple logistic regression was used to identify risk factors for ophthalmologic disorders.

RESULTS: A total of 2,555 children with ASD were seen at the university over the study period, of whom 380 (15%) were evaluated in the ophthalmology clinic. Eye examination revealed an ophthalmic diagnosis in 71% of children, of which the most common were significant refractive error (42%), strabismus (32%), and amblyopia (19%). Optic neuropathy occurred in 14 children (4%). Cerebral palsy was a significant risk factor for refractive error (OR = 3.22; P = 0.016), strabismus (OR = 3.59; P = 0.012), amblyopia (OR = 3.49; P = 0.0097), and optic neuropathy (OR = 14.0; P = 0.0009).

CONCLUSIONS: Ophthalmic disorders were found in 71% of children with ASD evaluated at our university-based ophthalmology clinic. The rates of significant refractive error, strabismus, amblyopia, and optic neuropathy exceeded those of the general pediatric population. ASD and cerebral palsy may have additive risk for these disorders.

FACTORS THAT DECREASE THE QUALITY OF FUSION

Strabismus develops due to an imbalance between two groups of factors



If this side is heavier, there will be strabismus

If this side is heavier, there will be no strabismus

LOOONG LIST OF FACTORS THAT DECREASE THE QUALITY OF FUSION

Mechanical: Abnormal orbital anatomy

- Abnormal oblique muscle anatomy / function
- Abnormal orbital pulleys
- Abnormal orbit - torted or shallow

Neurological

- **Abnormal innervation inc IOS**
- **Abnormal cortical factors inc IOS**
- **Amblyopia**
- **Organic visual loss**
- **Head injury**

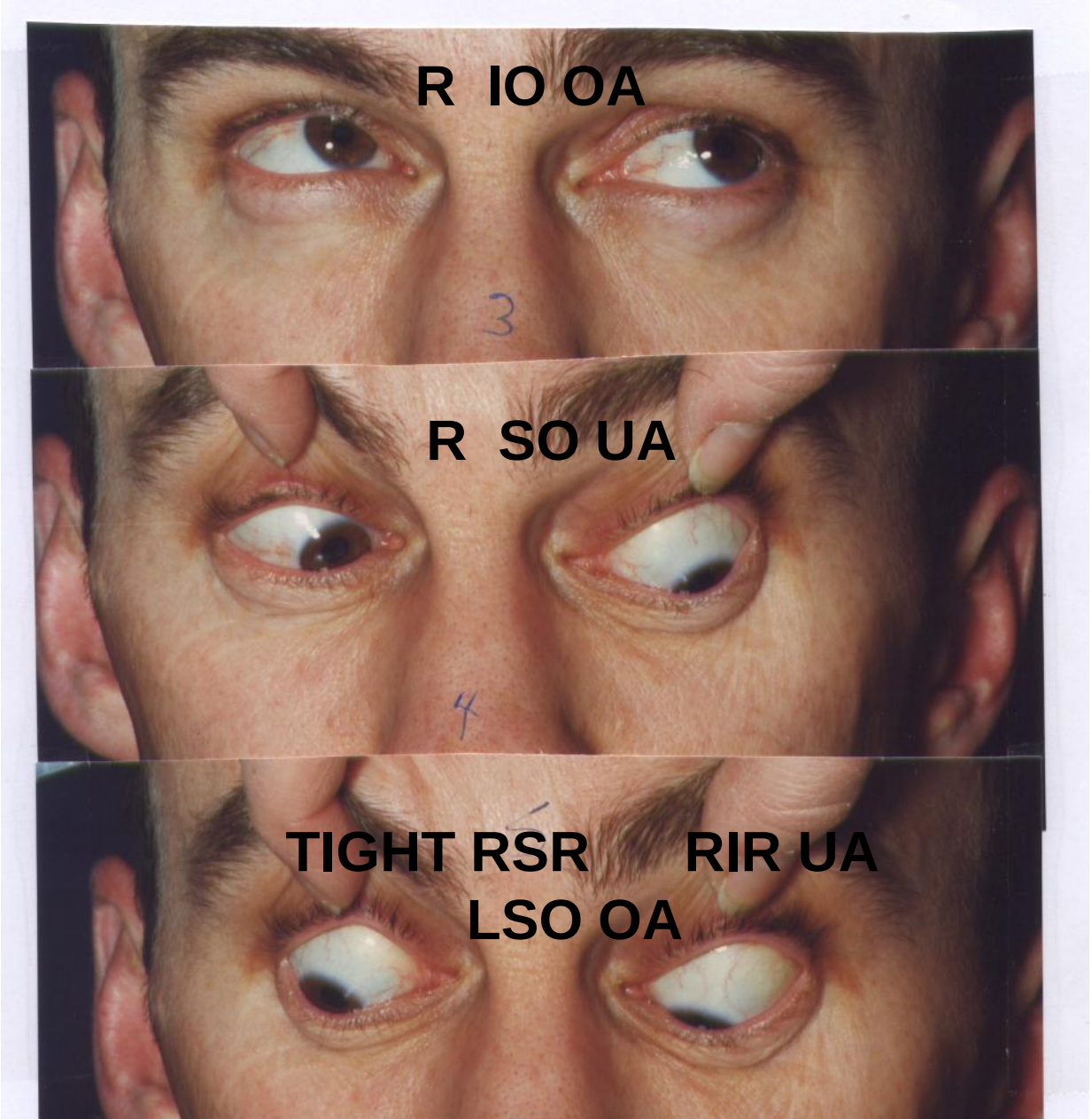
MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION 1
ABNORMAL OBLIQUE ANATOMY / FUNCTION

These 4 complex 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 ± 2-3 Δ.

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

ABNORMAL OBLIQUE ANATOMY / FUNCTION



MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION - SUBTLE ABNORMALITIES IN ORBITAL ANATOMY 2

ABNORMAL OBLIQUE ANATOMY / FUNCTION – NON PARETIC

FINK: 20% of cadavers: > 30° difference b/w course of SO & IO

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

The Role of Developmental Anomalies in Vertical Muscle Defects

Walter H. Fink

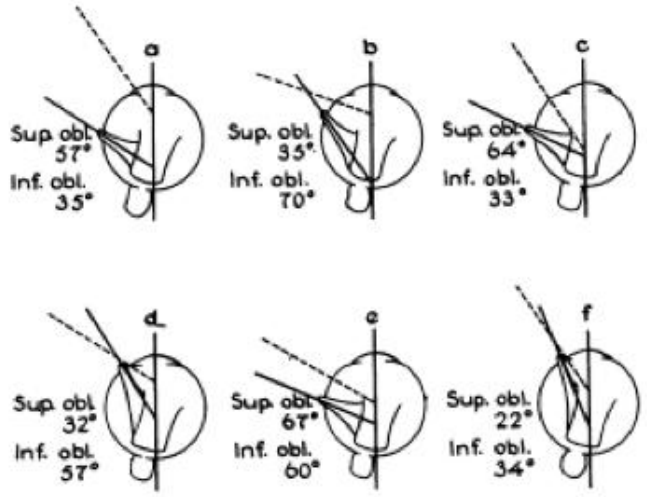
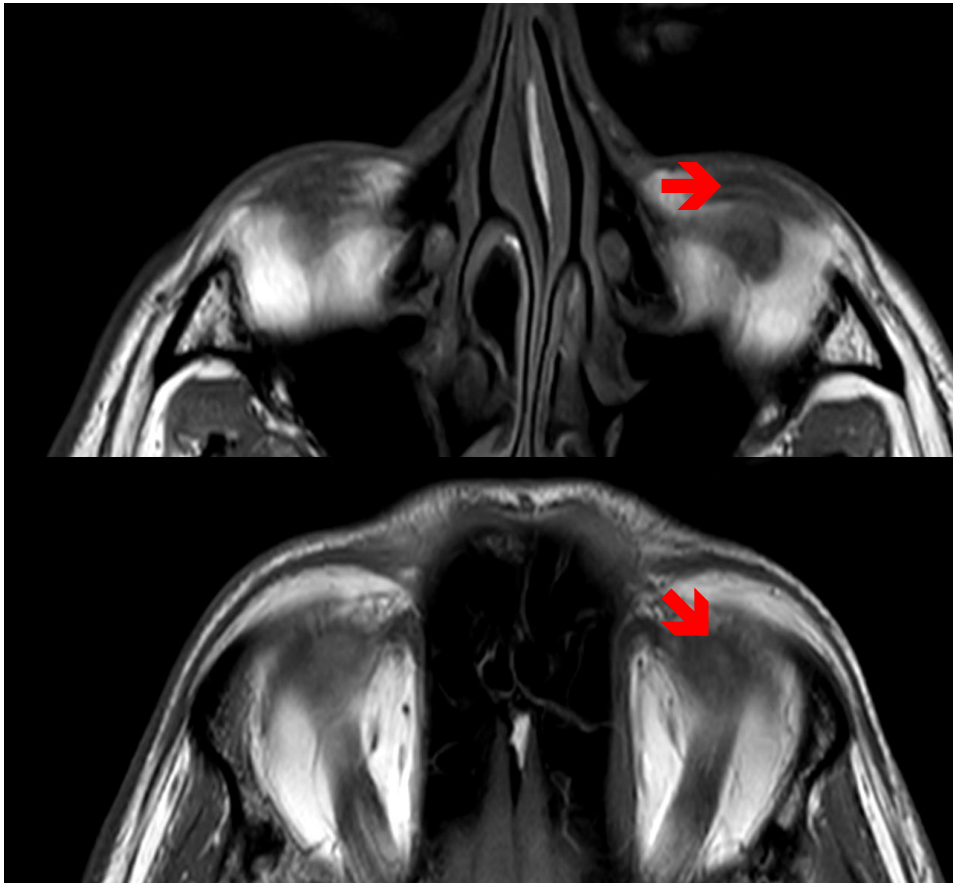


FIGURE 5. DRAWINGS OF SPECIMENS IN WHICH THERE ARE PRONOUNCED VARIATIONS OF THE OBLIQUE MUSCLE PLANES OF ACTION Continuous line indicates the superior oblique plane of action. Broken line indicates the inferior oblique plane of action.

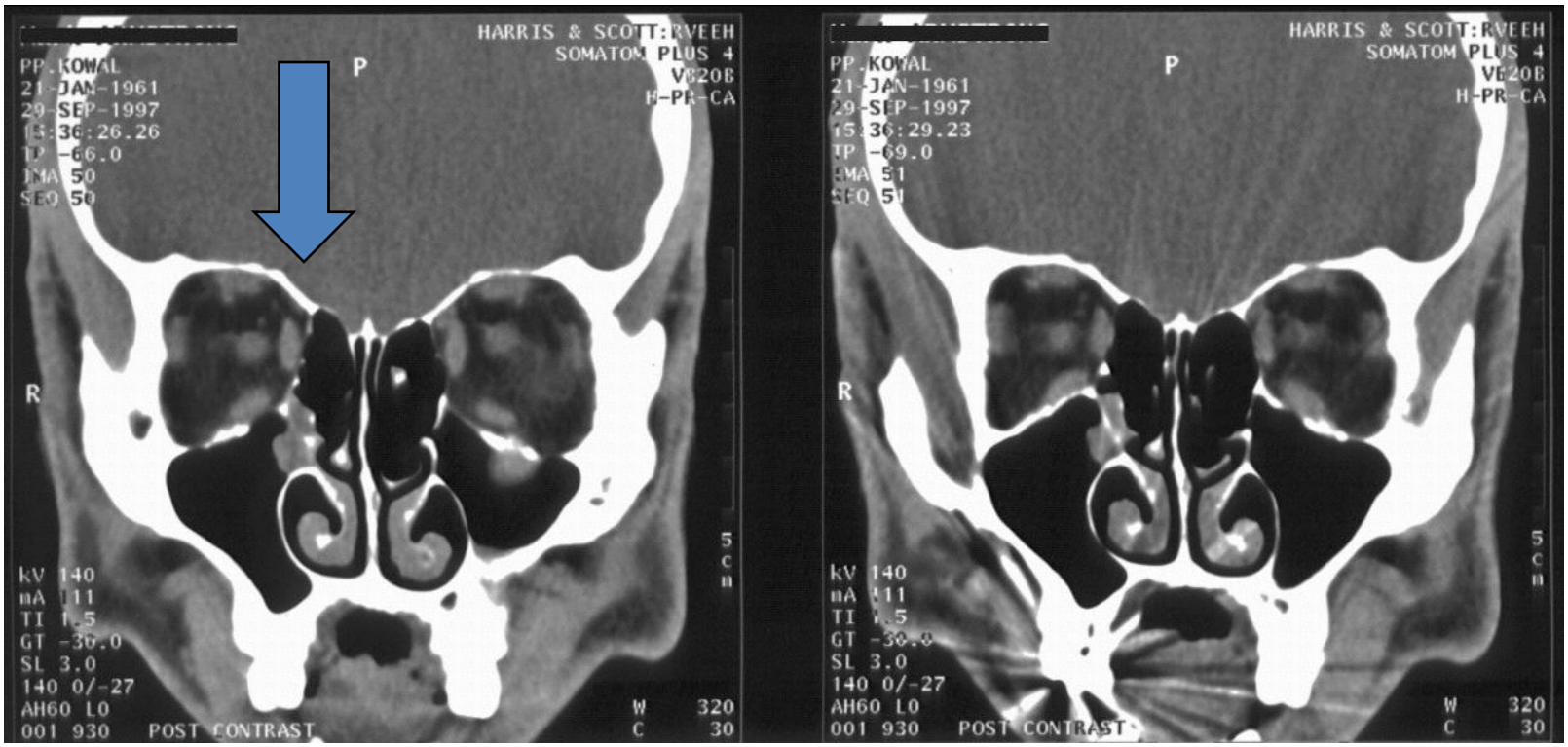
INFERIOR / SUPERIOR OBLIQUE ANATOMY IN VIVO

- IF ANATOMY IS VERY ASYMMETRIC IN VIVO, EXPECT OBLIQUE DYSFUNCTION



MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION: PARETIC

SUPERIOR OBLIQUE ATROPHY



LSO OK RSO ?absent

MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION - NON- PARETIC SUBTLE ABNORMALITIES IN ORBITAL ANATOMY

ABNORMAL OBLIQUE ANATOMY / FUNCTION

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

Apparent IO OA ~50%

Manifest strabismus in primary >50%

ET with vertical: 61% of all strabismus

BAGOLINI:
isolated posteroplaced trochlea is a cause of idiopathic oblique dysfunction

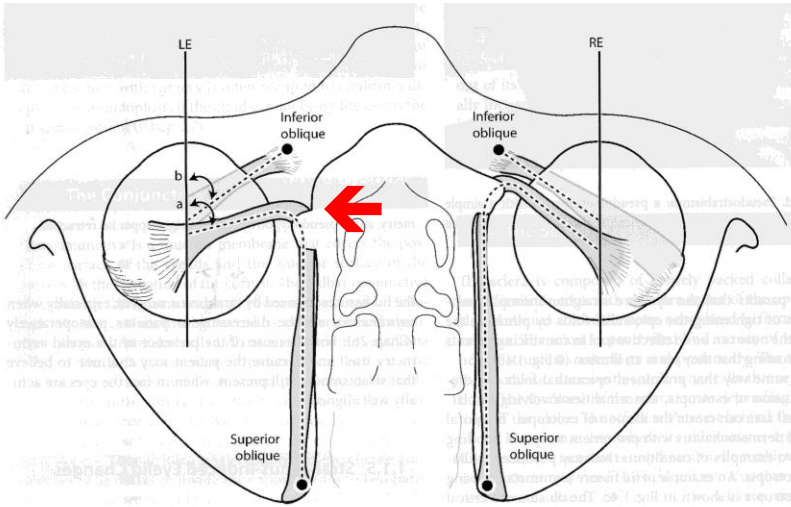
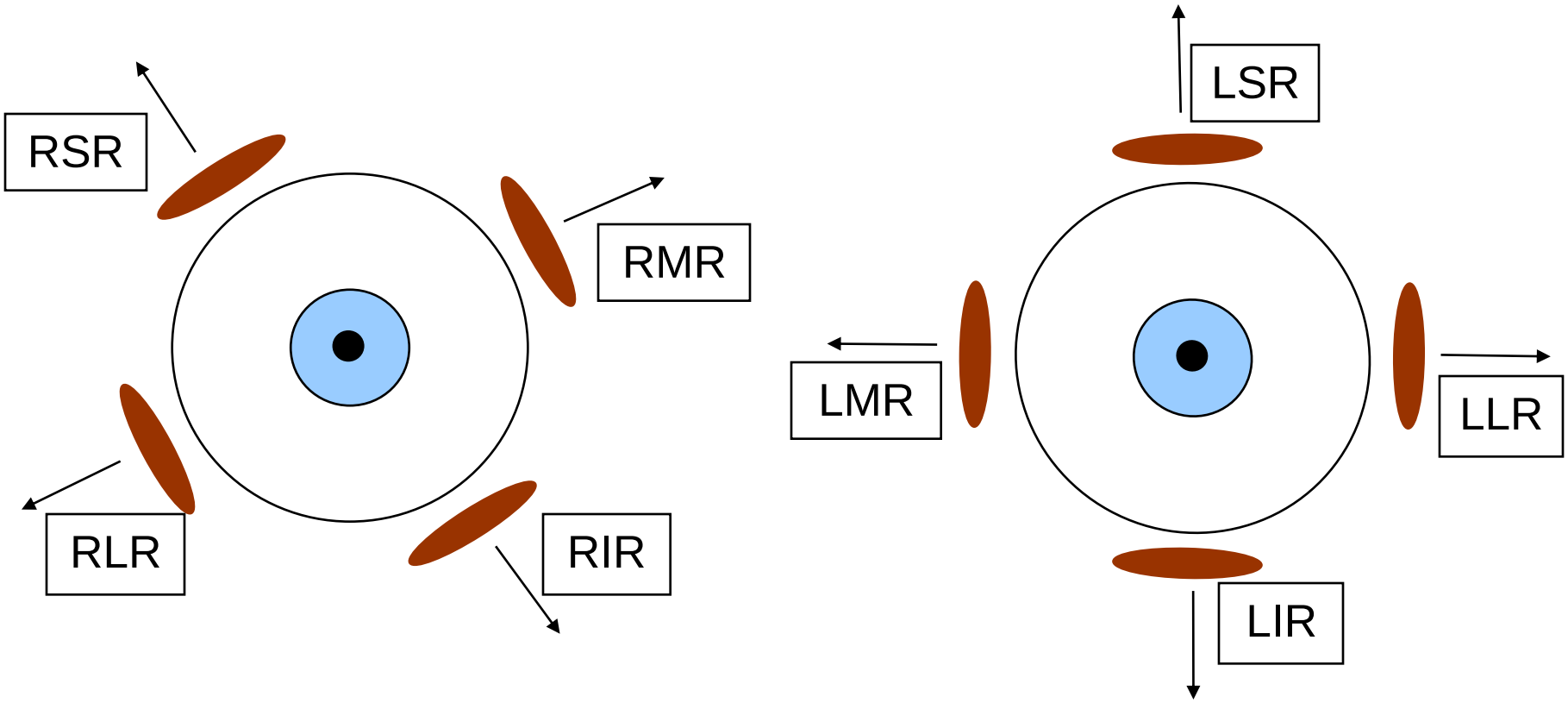


Fig. 1.3. Failure of the trochlea to advance anterior to the equator in a patient with unilateral coronal synostosis may result in reduction of d
pressing action on the globe with contraction of the superior oblique muscle

MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION – SUBTLE ABNORMALITIES IN ORBITAL ANATOMY

EXTORTED ORBIT

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



MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION - SUBTLE ABNORMALITIES IN ORBITAL ANATOMY

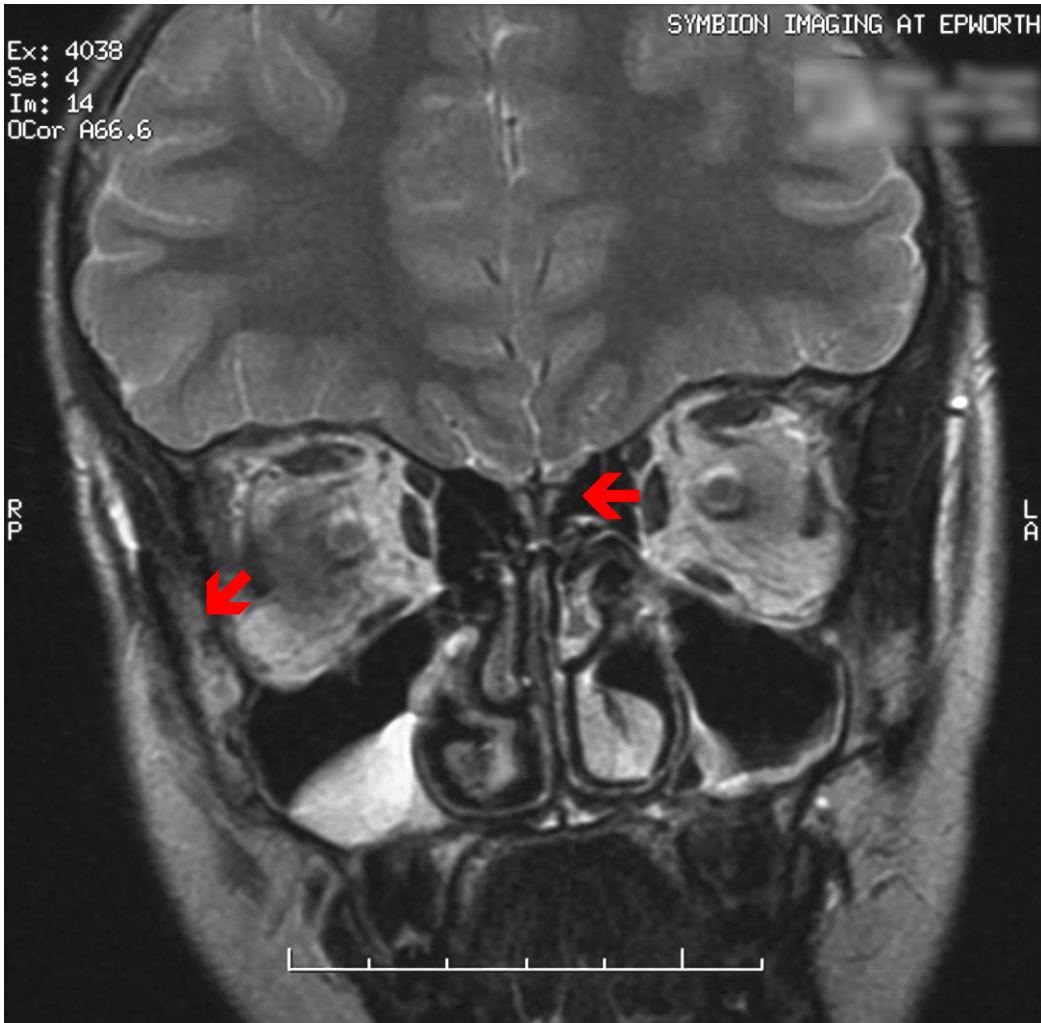
ORBITAL PULLEY HETEROTOPY

RLR lower than RMR

<u>R gaze</u>:

RLR will pull RE to R & down

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



Abnormal orbit anatomy is probably the commonest cause of alphabet patterns in ET

Journal of AAPOS
Volume 6 Number 6 December 2002

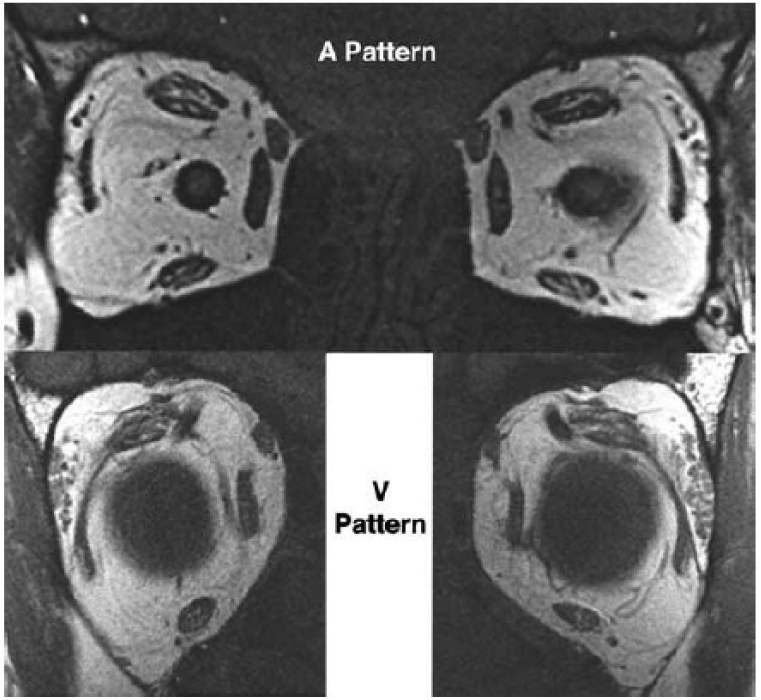


FIG 9. Typical coronal MRI from 2 representative patients showing heterotopic rectus pulleys associated with A-pattern (*top*) and V-pattern (*bottom*) strabismus.

FACTORS THAT DECREASE QUALITY OF FUSION

Mechanical

- Abnormal oblique anatomy / function
- Abnormal orbital pulleys
- Extreme myopia
- Abnormal orbit - torted or shallow

Neurological /sensory:

- Abnormal cortical factors esp motor aspects of IOS, PVL
- Amblyopia
- Organic visual loss
- Head injury
- Abnormal innervation

2. Manifestations of Infantile Onset Strabismus IOS [nearly always Esotropia]

- Persisting immature Supranuclear innervation of medial rectus muscles
- Tends to cause INFANTILE ONSET ET = CONGENITAL ET = ESSENTIAL INFANTILE ET = ….

IOS: Persisting immature Supranuclear innervation of medial rectus muscles

with apologies for the terminology

2 main persisting types of immature supranuclear eye muscle control:

1. **FMN** – Fusion Maldevelopment Nystagmus [aka LMLN]

2. **MNTSPA** – Monocular Naso Temporal Smooth Pursuit Asymmetry aka **SPA**

IOS: Persisting immature Supranuclear innervation of medial rectus muscles

1. FMN – Fusion Maldevelopment Nystagmus [aka LMLN]

– **N to fixing eye** – unique type of N

- **N increases to lateral gaze** (also seen in GPN) – acuity worse on lateral gaze & prefers to fix in aDduction:

<u>ADduction null :</u>

- **L fixation: face turn to L because of aDduction null,**
- **R fixation: ……R……**

….causes progressive tightness / shortening of MR & probably drives the ET in IOE

IOS: Persisting immature Supranuclear innervation of medial rectus muscles

1. **FMN** – Fusion Maldevelopment Nystagmus

– N to fixing eye

- N increases to lateral gaze
- aDduction null probably drives the ET in IOE

2. **MNTSPA** – Monocular Naso Temporal Smooth Pursuit Asymmetry aka **SPA**

An IOS Key feature
.. present in all neonates.

- **Smooth pursuit asymmetry SPA**
- SPA normally matures [asymmetry becomes symmetric & disappears] by age 4-6 mo, and if it doesn't, then IOS develops in ?many ?most [but not all]
- SPA is sometimes found in asymptomatic 1st degree relatives of children with IOS

IOS

Complicated supranuclear abnormalities
ET is one part of this

1. Usually: Large angle ET.
Think: *Bilateral Monocular Esotropia* caused by preference for fixation in aDduction (=null)

2. FMN Fusional Maldevelopment Nystagmus:
N beating to fixing eye
Without cover = Manifest Latent N

3. SPA Smooth pursuit asymmetry
Larry Tychsen Thank you for video on nextslide

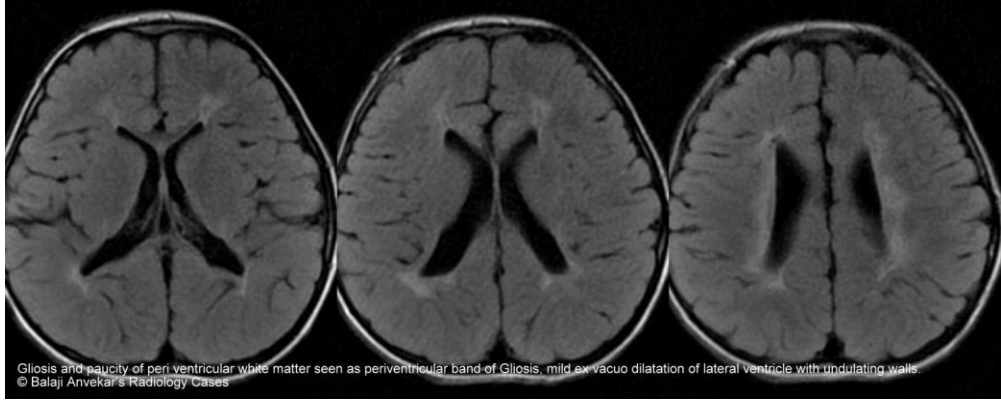
1. Large angle ET. 2. FMN Fusional Maldevelopment N : N beating to fixing eye. With cover L, N to R, then…
3. Without cover [= Manifest Latent N] N to R, then… 4. cover R, see N to L, then… 5. SPA Smooth pursuit asymmetry R then L, jerky going temporal, smooth(er) going nasal

IOS : Key Features

- The FMN & SPA are core abnormalities
- Reduced potential for sensory fusion, even when perfectly straightened
- Reduced potential for motor fusion, even when perfectly straightened, **hence predisposition to recurrent ET even with low +**

- **WHY do we get FMNS & SPA?**

IOS Causes & Associations: **PVL**



- **PVL PeriVentricular Leukomalacia**
- Common in premature babies
- The periventricular area is a watershed area for circulation @ ≤32 w
- PVL increases the risk of IOS by a factor of ≥ 30!
- Many possible manifestations of PVL other than IOS: cerebral palsy, poor reading, ….

IOS Causes & Associations: Genetic associations

- Williams syndrome [rare] ≈ 100% have IOS
- Down's syndrome many have IOS
- Many other chromosomal disorders associated with impaired devpt have high % of IOS

IOS Different subtypes

1. Ciancia's syndrome:

IOE phenotype with main features: face turns, head tilts

2. IO XT. Sometimes behave like mirror image of IOE

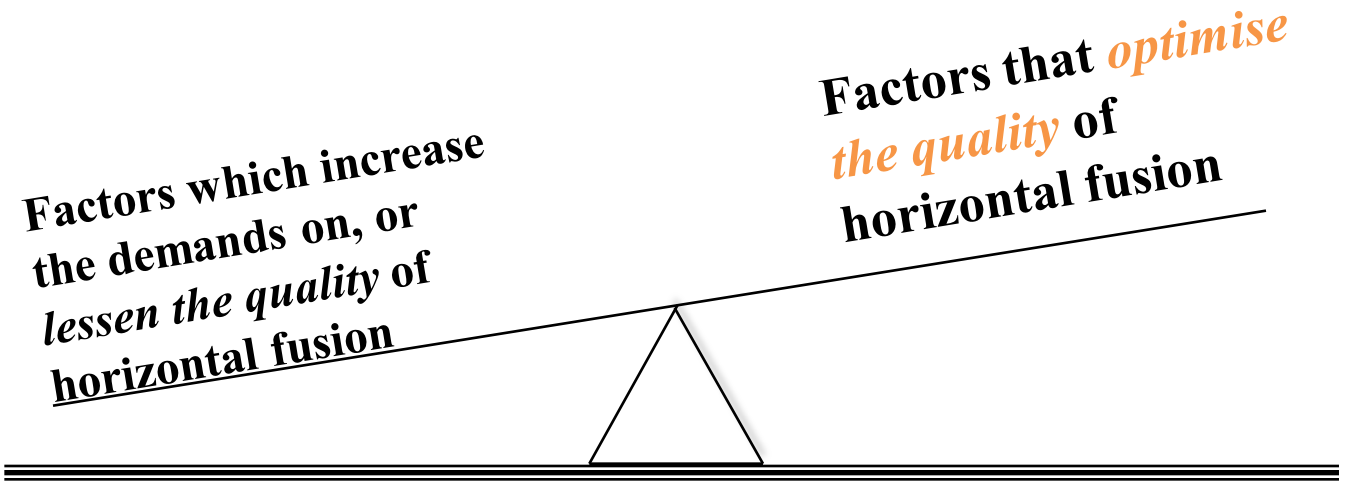
IOS Different subtypes
Dissociated deviations

3 Horizontal [usually eXo = **DXD**]…*manifestations of horizontal component of FMNS :* looks like XT, but RF often ≠ LF

4. Vertical = **DVD** *manifestations of the T component of FMNS :* looks like any hyper, but RF often ≠ LF & there is no Hypo of other eye

Optimising quality of motor fusion

ET develops due to an imbalance between two groups of factors



If this side is heavier, there will be strabismus

If this side is heavier, there will be no strabismus

Improve quality of motor fusion

1. Optimise refraction
2. Improve amblyopia
3. Straighten eyes
4. Improve the orbital factors that have impaired motor fusion…..if you can do this reliably and safely

Benefits of alignment

- 1. normal appearance – important for self esteem and normal social relationships
- 2. ± best chance for stereo
- 3. ± better peripheral field
- 4. ± easier amblyopia treatments

SEMINAL PAPER

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

Lawrence Tychsen, MD

Can ophthalmologists repair defects of visual cortex circuitry in infants who have esotropia? The answer to this question encompasses both sensory and motor behaviors because the clinical hallmarks of the disorder are stereoblindness and absence of motor fusion, which manifests as convergently deviated eyes. Functional recovery of sensory and motor fusion in infantile esotropia was a consuming interest, if not career-defining passion, of Marshall Parks. The purpose of this work is to pay tribute to Parks' legacy by showing how human and animal studies, conducted largely during the last 25 years, support both his clinical insights and treatment philosophy. (J AAPOS 2005;9:510-521)

A discussion of Dr. Parks' (Figure 1) contributions to fusion recovery can begin with the men whose ideas helped refine his thinking. Marshall Parks' clinical education—at the end of World War II—was shaped by a debate between 2 competing 20th-century schools of treatment philosophy, derived from the eminent British strabismologists, Claude Worth and Bernard Chavasse (Figure 2). Worth postulated, in 1903, that esotropic infants suffered "an irreparable defect of the fusion faculty."[1] Their brain was congenitally incapable of achieving substantial binocular vision. Early surgical treatment was therefore unfounded because it was futile. Chavasse, on the other hand—attracted by the Pavlovian physiology of the 1920s and 1930s—believed that the brain machinery for fusion was present in esotropic infants, but the development of "conditioned reflexes" for binocular fusion were impeded by factors such as weakness of the motor limb.[2] He postulated (in his text published in 1939) that if the eyes could be realigned during what he believed to be a period of reflex learning, binocularity of some degree could be restored.

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COSTENBADER-PARKS AND THE ROOTS OF EARLY REPAIR

The publications of Chavasse had impressed Frank Costenbader, an American ophthalmologist practicing in Washington, DC. He had, uniquely for that time (the 1940s) established the only ophthalmology practice devoted to pediatrics in the United States.[3] Costenbader summarized his clinical observations of strabismic infants, gathered over a period of 2 decades, in a landmark article published in the 1961 *Transactions of the American Ophthalmologic Society*.[4] The article defined infantile esotropia and reported that 1 in 5 children could develop gross stereopsis if surgically aligned by age 1 year.

Marshall Parks was Costenbader's first fellow (circa 1947) and first practice partner (1949) (Figure 3). As such, he was tutored in the Chavasse-Costenbader school, which favored early surgery. In the early 1960s, he succeeded Costenbader as director–mentor to what would become in time a long succession of distinguished fellows-in-training (see Appendix 1). By the time the article "Early Surgery for Congenital Esotropia" was published (in 1966, written with the 7th Parks fellow, Malcolm Ing[5]), Dr. Parks was imprinting his own early surgery rationale on the conscience of each Washington trainee.

INFANT PSYCHOPHYSICS IN DALLAS, TEXAS

The 19th Parks fellow was David Stager, Sr., who had established himself in Dallas, and was carrying out the Parks' mandate to contribute to strabismus research in a private practice. In the mid-1980s, Dr. Stager teamed up with a gifted infant psychophysicist, Eileen Birch, to study the development and maldevelopment of stereopsis.[6] The study was possible because of the invention a few years

When to straighten the eyes? Tychsen

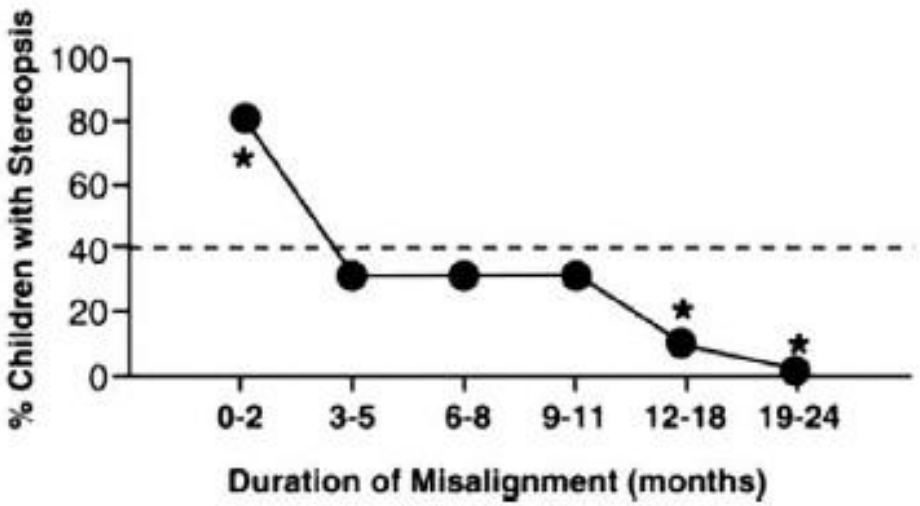


FIG 5. Prevalence of stereopsis after surgical realignment in children with infantile esotropia as (A) a function of age-of-onset of esotropia and (B) as a function of duration of esotropia before realignment. Testing performed at age 5 years. Surgical realignment achieved generally by age 1 year for the population as a whole. Dashed line at 40% indicates average prevalence for all the infants. Data from more than 100 consecutive infants, replotted from Birch et al.[25]

Tychsen: BENEFITS of earlier alignment Improve quality of fusion

- Better sensory fusion
- Better motor fusion
- Less need for 2nd surgery
- …these outcomes are optimised if re-alignment occurs within a few months of constant misalignment

Alignment – principles in ET

- Check that child is wearing full +
- Check again that child is wearing full +
- Has amblyopia Rx been optimised?
- A little + can have a big effect in

- Convergence Xs ET

- A child with poor motor fusion reserve eg after previous Infantile Onset ET surgery

Alignment : When is the child ready? Hyperopia & the child who won't wear glasses

- *<u>Check refraction</u>: what to do with low + eg +2.50 DS?*

If the child won't wear glasses, I use pilocarpine 1% OU to check if the + is strabismically important *no data,* or

'Force' glasses wear with g.Atropine 0.5% daily for a few days

Alignment – principles in ET

- Most careful measurements for D&N.
- D: also UG, DG
- Check versions, ductions
- Large ET: expect tight MR, and LR UA
- Look for real / apparent oblique dysfunction*

** 'oblique dysfunction', 'IO OA', 'SO UA' etc = strabismus Dr's shorthand for 'abnormal vertical version when the eye is aDducted' and can but may not have anything to do with **real** oblique dysfunction*

When do I book an operation?

- 1. I have been able to examine the child confidently and accurately twice: *'measure twice, cut once'*

- 2. I have a surgical plan that is 90+% reliable for early orthotropia

THINKING OF 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 1

- Realignment often only fixes part - a large necessary part, but only a part - of the problem

- Often, the only reliable outcome is improved appearance : abnormal appearance has to be an issue before proceeding with surgery

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]

These parents need LOTS of time

- Parental expectations will never be met: one surgery perfect cure - perfect alignment, appearance, 3D, no need for glasses any more

- Child has had unconventional ineffective treatment for some years : need recalibration of parental beliefs ± management of guilt for ?depriving my child of effective treatment: a great result will fix guilt issues

- Albinism: +ve angle Kappa common: when aligned, will look XT

Surgery doses / techniques

- Do whatever works…
- Most of the world: BMR Bimedial recession, Parks' mm doses for ET ≤50 Δ

<u>Many variations:</u>

- Hangback* vs. fixed recession
- Convergence Xs : near angle*, distance mm + 1mm, BMR + pulley suture*, BMR + scleral Faden, …
- Huge ET: BMR + another muscle*[s], BMR + Botox
- Augment the mm dose for large globes*
- Rc-Rs or Rc-Plicate* for unilateral amblyopia

* = LK preference

Alignment: How

- Some cities: bimedial Botox [no randomised series]

Cochrane review: unenthusiastic

- Some uncertainty and no EBM regarding:

- Simultaneous oblique/ vertical surgery
- Huge angles

Alignment – principles in ET
LK guidelines for A-V patterns

- V
- If have IO OA, SO UA, V, fundus extorsion: add ATIO OU to the horizontal surgery
- If do not have full hand, offsets of horizontals, and do bilateral symmetric surgery

- A
- If have SO OA, IO UA, A, fundus intorsion: superior oblique weakening OU.
- Higher morbidity than other surgeries – leave for experts
- If do not have full hand, offsets of horizontals, and do bilateral symmetric surgery

Problems with surgery
Need for 2nd surgery

- Month 1: 2% - something has gone wrong – wrong measurements, stitches break, ….
- Year 1: 10% - something goes wrong with scar maturation, wrong choice of operation, ….
- By year 20: 30% - commonest problem becomes consecutive eXotropia.

The amount of muscle re-positioning required to give optimal early alignment and best early sensory and motor visual development sometimes does not ‘grow’ with growth of the eye and orbit and ends up being out of perfect ‘engineering’ balance in 20%

Best possible outcome after surgery in IOS

- Straight with reduced sensori - motor fusion – this means :

- Increased risk of subsequent strabismus
- Fragile hold on orthotropia
- Alignment easily disrupted eg with uncorrected + or presbyopia

Acute esotropia – sudden onset moderate-large ET 1

Acute esotropia – sudden onset moderate-large ET…or stuttering onset over a day or so

Why worry? **Tumour** in :

- brainstem [causing direct damage to the oculomotor nerves] or
- posterior fossa [causing damage indirectly through raised intracranial pressure]

<u>Commonest identifiable cause of acute esotropia:</u>

- Uncorrected +

Acute esotropia – sudden onset 2
The literature

- MANY case reports of acute comitant ET with **no apparent neurological overlay** caused by tumour

- **Mayo clinic:** large series [nearly 300] with good follow up **– NO tumours** in this population

Acute esotropia – sudden onset 3
The conversation: true story

- 3 pediatric ophthalmologists chatting at a conference…Σ 75 years of experience
- Thousands of cases of ET
- Hundreds of **acute ET with no neurological hints** – no pale disc, sudden aBduction deficit, swollen disc(s).
- #1 [me]: NO tumours
- #2 [Melbourne]: NO tumours
- #3 [Sydney]: SEVEN tumours in this group

??Weird unrecognized selection bias…

For the general ophthalmologist

- ‘I’m only going to do the easy cases’

<u>Easy case:</u>

- Deteriorated accomodative ET
- Range 20 - 40Δ
- With prism compensation in the office, has evidence of sensory fusion
- Same angle on UG, PP, DG, Near, & LF = RF

For the general ophthalmologist
The Easy Case

- Give full +
- Check that you have given full +
- BMR using Parks' or Wright's tables
- Good luck

INFANTILE ONSET ESOTROPIA

IT'S UNCOMMON,
IT'S DIFFICULT
EXAMINE WITH **GREAT** CARE

Slide courtesy of Larry Tychsen

6th NERVE PALSY - looks the same

R +5, L +6 : looks the same as IOE

Brainstem tumour - looks the same as IOE

DUANE'S - looks the same as IOE

Benefits of early alignment in acquired ET

- Wilson ME et al
- BINOCULARITY IN ACCOMMODATIVE ESOTROPIA *JPOS JULY-AUG 1993*

- No pt with stereo had constant ET > 4 mo

When to straighten the eyes? Tychsen

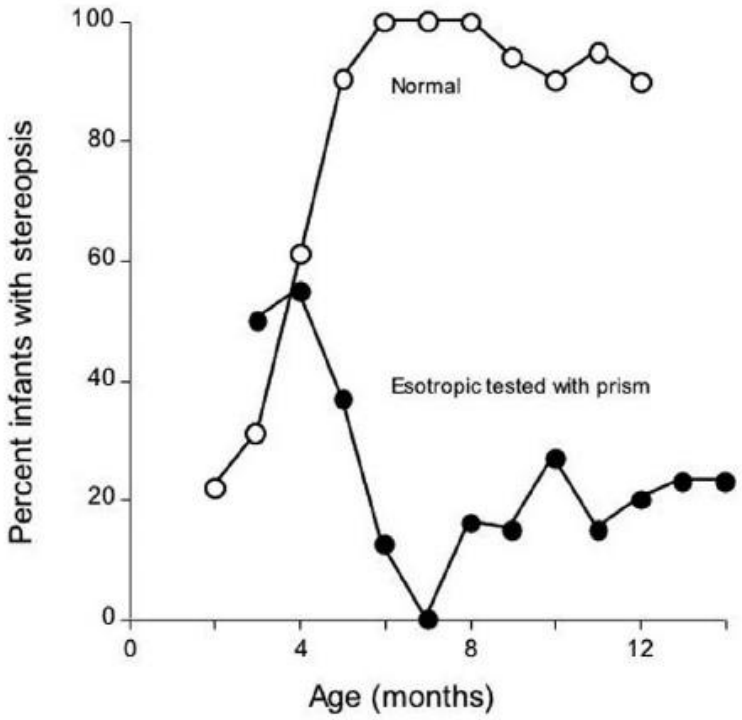
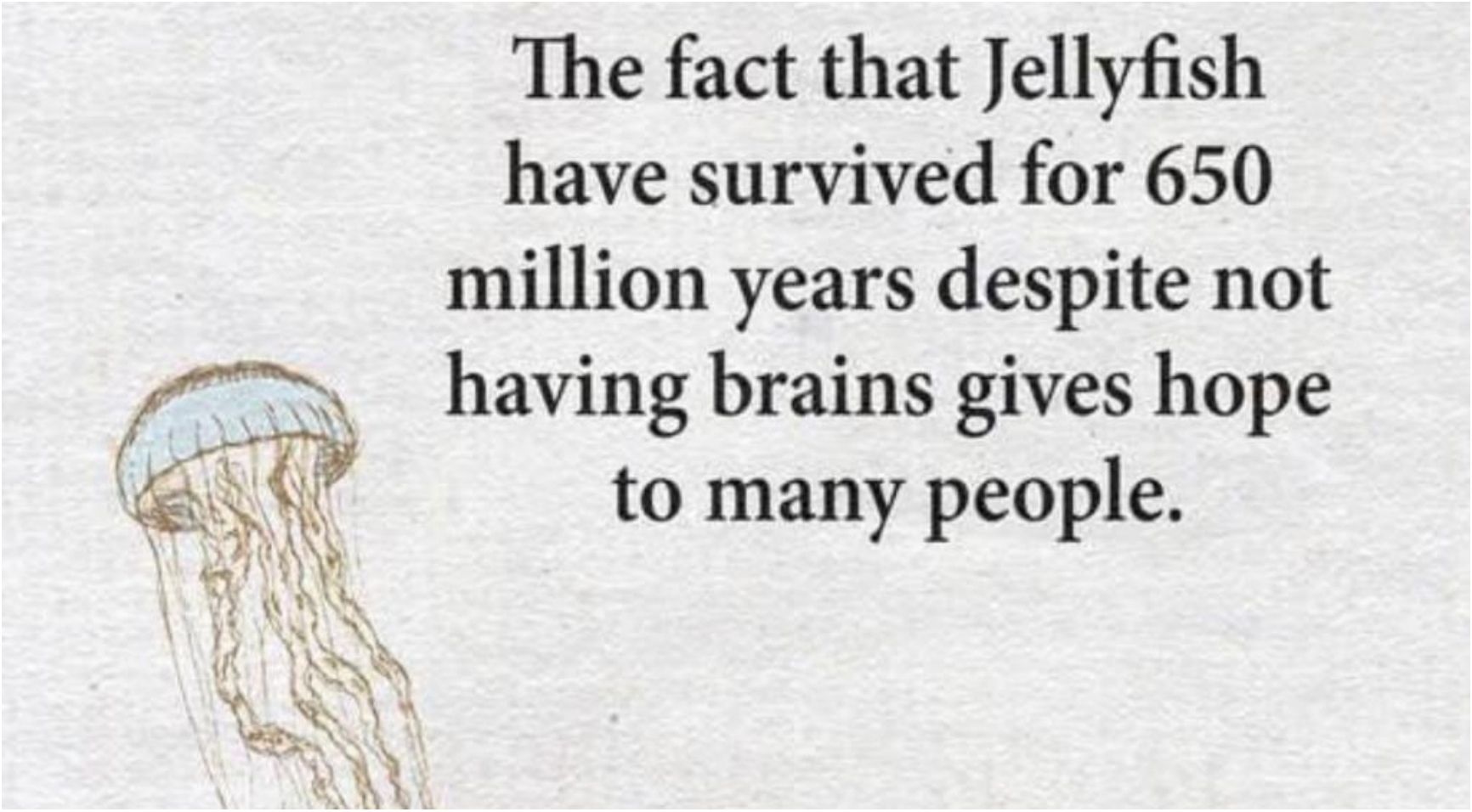


FIG 4. Prevalence of stereopsis as a function of postnatal age in a population of normal ($n > 50$) versus esotropic infants ($n = 85$). Tested using dichoptic viewing (polarized goggles and images) by the preferential looking method. Esotropic infants were aligned using prisms and tested before any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


The fact that Jellyfish
have survived for 650
million years despite not
having brains gives hope
to many people.