MANAGEMENT OF STRABISMUS & AMBLYOPIA 2018



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ORANGE IS FOR

REALLY

IMPORTANT

INFORMATION

OVERVIEW

- Introduction
- Why to treat & when to treat
- Why strabismus happens
- How to treat
- Amblyopia: Causes and treatment

WHY IS IT SO DIFFICULT?

Every clinical decision depends on accurate evaluation of:

- o 1. Alignment
- o 2. Acuity
- o 3. Refraction
-all difficult to do reliably in children.
- Lectures / textbooks necessary, one-one tuition
 - & supervised examination is essential

REQUIREMENTS OF A PERFECT VISUAL SYSTEM. WE NEED ALL OF:

- 1. Straight eyes
- 2. Good & equal vision
- 3. Low [or no] & symmetric refractive error
- 4. Normal EOM anatomy / innervation / physiology
- 5. Normal occipital lobe anatomy & physiology (required for normal motor fusion, normal sensory fusion)
- 6. Normal visual pathways
- 7. Normal early visual development

STRABISMUS: END RESULT OF ANY IMPERFECTION IN THIS COMPLEX JIGSAW PUZZLE

Abnormalities in one / more of...

- Sensory development
- Refraction
- Orbital anatomy
- EOM anatomy / physiology
- Relevant brain anatomy, function and development

Visual system takes up ~ ½ the brain!

Accommodation / convergence..can cause or be caused by strabismus

TIME DEPENDENT RESULTS

- Delay in starting & completing effective treatment can have negative life- long outcomes
- You WILL in your career see children and adults with visual loss that is /was reversible only with timely & effective treatment
- o IF YOU HAVEN' T FIXED IT IN 3 MONTHS, REFER

KEY TO SUCCESSFUL MANAGEMENT OF EXPECTATIONS: EDUCATION

'HIGHLY RECOMMENDED [FREE!] E-BOOK FOR PATIENTS & PARENTS TO READ'

- •Eye Muscle Problems in Children and Adults: A Guide to Understanding
- OBurton J. Kushner, MD

University of Wisconsin Department of Ophthalmology and Visual Sciences, Madison

LINK ON MY WEBSITE: SHOULD BE ON YOURS

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.
- ...after reading this book you will feel more empowered to make considered choices regarding the treatment of your child, yourself, or your loved one.

2 STEP MANAGEMENT OF STRABISMUS

..in either order, or simultaneously

- Straighten the eye(s) or otherwise compensate for misalignment inc FULL +/Prisms / Surgery / Botox (infrequent option)
- 2. Improve /equalize acuity

SLOW DECLINE IN NUMBERS OF SURGERIES MEDICARE STATS

12 months	To July 14	To July 15	To July 16	To July 17
Total strab surgeries	2482	2440	2304	2261 9% decline in 3y
Under 15 yo	1521	1479	1423	1369 10% decline in 3y
Botox for strabismus	147	141	135	209 40+% increase in 2017

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WHY STRAIGHTEN THE EYES?

Age < 12 mo:

• Best chance for some sensorimotor fusion

Commonest good result:

- Straight most / all of the time
- Reduced sensorimotor fusion
- Normal appearance

WHY STRAIGHTEN THE EYES?

Age 3-7:

- Best chance to develop / to regain sensorimotor fusion
- Normal appearance, self esteem & psychological and social devpt [important from prep year]
- Better motor skills
- Better reading speeds

STEREO VISION ENHANCES THE LEARNING OF A CATCHING SKILL.

MONTAGNE G ET AL EXP BRAIN RES. 2007 JUN;179(4):723-6.

- Poor catchers with good stereo (N = 8; Stereo+) and weak stereo (N = 6; Stereo-) participated in an intensive training program over 2w, during which they caught >1,400 tennis balls.
- Stereo +: improved 18% to 59%
- Stereo : 10 to 31% not significant similar to control group (N = 9) that did not practice at all.

WHY STRAIGHTEN THE EYES?

Age >10:

- Best chance to regain some sensory fusion, usually subnormal
- Normal appearance / self esteem / social interactions
- Better field [if ET; worse if XT]

Opinions of dating agents about strabismic subjects' ability to find a partner

S M Mojon-Azzi,1 W Potnik,2 D S Mojon3

2008;92;765-769 Br. J. Ophthalmol.

ABSTRACT

Aims: To determine the influence of strabismus on the ability to find a partner.

Methods: We interviewed Swiss dating agents retrieved from two Swiss online telephone directories using a validated questionnaire to determine whether strabismus has any impact on the ability to find a partner. During the interviews, subjects with internet access could view downloadable, digitally altered photographs of a strabismic man and women, as well as images of other computer-generated facial anomalies.

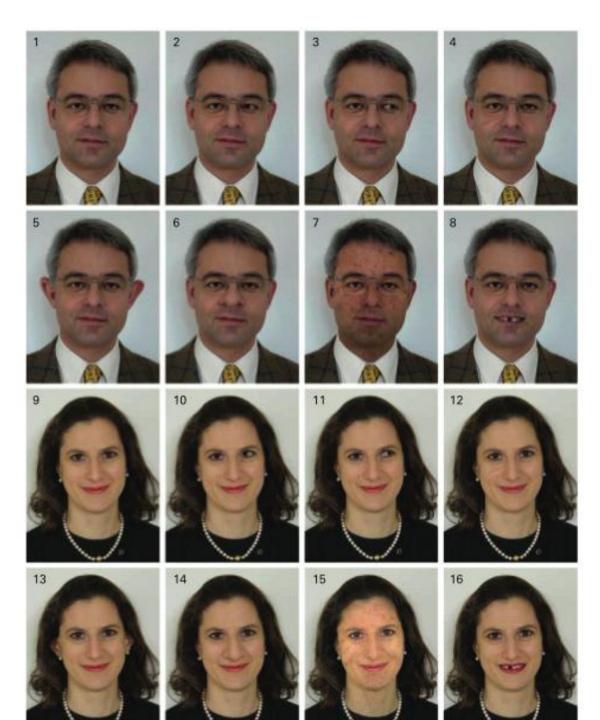
Results: Of the 40 dating agents, 92.5% judged that strabismic subjects have more difficulty finding a partner (p<0.001). Such difficulty was not associated with either gender or age but was perceived as being greater in exotropic than in esotropic persons (p<0.001). Among the seven facial disfigurements, strabismus was believed to have the third largest negative impact on finding a partner, after strong acne and a visible missing tooth. Dating agents also believed that potential partners perceive persons with strabismus as significantly less attractive (p<0.001), erotic (p<0.001), likeable

(p<0.001), interesting (p<0.001), successful (p<0.001), intelligent (p = 0.001) and sporty (p = 0.01).

Conclusions: Visible strabismus negatively influences the ability to find a partner. Because strabismus surgery in adults restores a normal functioning condition and reduces not only physical but also psychosocial difficulties, it cannot be considered a cosmetic procedure. distress, particularly during social interactions that expose the disfigurement to others' gaze and can result in displays of ignorance and negative comments.

The psychosocial problems experienced by strabismic individuals are similar to those of persons with other craniofacial anomalies. Jackson et al6 measured anxiety and depression, social anxiety and QoL 6 weeks before and 3 months after strabismus surgery. The researchers found not only that strabismic individuals experience greater social anxiety and use more social avoidance strategies but that these subject's scores reduce to normal levels following surgery. This finding of strabismus negative impact was confirmed by Satterfield et al,7 who found evidence of problems related to strabismus during school, work, play or sports in subjects over age 14. Nonetheless, the authors identified no difference in the amount of psychosocial impairment between esotropic and exotropic subjects. In a similar study, Menon et al8 showed that patients aged 15-25 who had had a constant squint since childhood had difficulties with self-image and interpersonal relationships, faced ridicule at school and work, and generally avoided activities that brought attention to their defect. Burke et al9 showed that strabismus surgery reduced the psychosocial difficulties reported before surgery and improved the quality of nevchosocial functioning. Resuchamn et allo also

Figure 1 Photographs of a man and woman with and without seven computer-generated facial anomalies. Subject consent has been obtained for publication of this figure.



WHEN TO STRAIGHTEN THE EYES? TYCHSEN (I/IV)

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

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)

WHEN TO STRAIGHTEN THE EYES? TYCHSEN (II/IV)

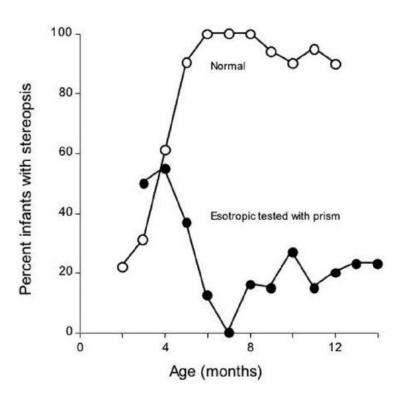


FIG 4. Prevalence of stereopsis as a function of postnatal age in a population of normal (n > 50) versus esotropic infants (n = 85). Tested using dichoptic viewing (polarized goggles and images) by the preferential looking method. Esotropic infants were aligned using prisms and tested before any surgery.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

WHEN TO STRAIGHTEN THE EYES? TYCHSEN (III/IV)

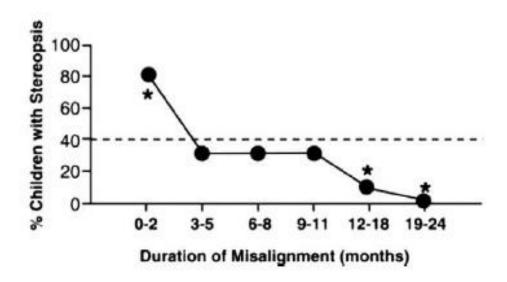


FIG 5. Prevalence of stereopsis after surgical realignment in children with infantile esotropia as (A) a function of age-of-onset of esotropia and (B) as a function of duration of esotropia before realignment. Testing performed at age 5 years. Surgical realignment achieved generally by age 1 year for 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

WHEN TO STRAIGHTEN THE EYES?

Kids:

orealign within a few ?4 mo of constant misalignment to regain best sensorimotor fusion ...usually not achieved

Adults:

o...≤ 12mo of constant misalignment to frequently regain measurable sensorimotor fusion...usually not achieved

Many exceptions: many great results can also be seen after prolonged delays to alignment

oKushner: 40% of adult 'cosmetic' realignments: measurable improvement in sensory fusion

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CLUES TO THE CAUSES OF STRABISMUS

1. GENETIC

Frequent strabismus:

- William's syndrome 75% have congenital ET Chrom 7
- ⇒ genetic factor
- Many families with frequent strabismus
 & no defined genetic explanation

CLUES TO THE CAUSES OF STRABISMUS 2: NEUROLOGICAL

Frequent strabismus:

- Neonatal brain injury IVH / HC: most have Infantile Onset Strabismus [IOS]
- 1. Developmental delay of any sort: genetic / acquired 25%
- 2. ASD / ADD/ ADHD population Increased frequency

THE CAUSE OF INFANTILE STRABISMUS LIES UPSTAIRS IN THE CEREBRAL CORTEX, NOT DOWNSTAIRS IN THE BRAINSTEM

TYCHSEN, L EDITORIAL ARCHIVES OPHTHAL AUG '12

• Infantile-onset strabismus IOS.. a combo of abnormal ocular motor behaviors: eye misalignment, subnormal binocular fusion, a type of nystagmus, dissociated vertical & horizontal deviations.

THE CAUSE OF INFANTILE STRABISMUS LIES UPSTAIRS IN THE CEREBRAL CORTEX, NOT DOWNSTAIRS IN THE BRAINSTEM

TYCHSEN, L EDITORIAL ARCHIVES OPHTHAL AUG '12

• Children at greatest risk are those who suffer cerebral lesions around the time of birth, esp PVL = Peri Ventricular Leuko Malacia, damage to the posterior-most fibers of the optic radiations, the binocular inputs to striate cortex).

o PVL: >30 fold greater risk of IOS

CLUES TO THE CAUSES OF STRABISMUS 3 GENETIC & ORBITAL

Comitant Horizontal Strabismus: an Asian perspective. Chia A, et al. BJO. 2007 May 2; Singapore.

2ce as many Singaporean children present with XT than ET

Caucasians ET >> XT.

Within the XT and ET groups, the distribution, characteristics and treatment responses of various strabismus subtypes are similar to Caucasians

4. NON- SYNDROMIC / NON-NEUROLOGICAL CAUSES OF STRABISMUS

Strabismus develops due to an imbalance between two groups of factors

Factors which increase
the demands on, or lessen
the quality of horizontal
fusion

Factors that optimise the quality of horizontal fusion

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

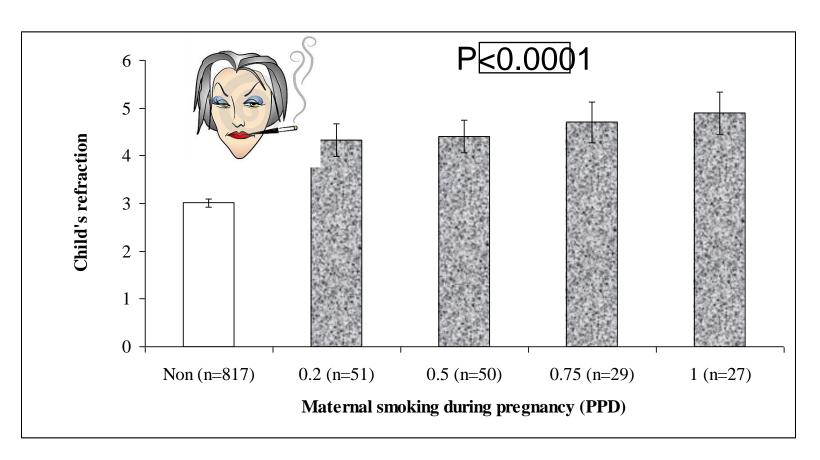
- Hyperopia
- Abnormal accomm convergence relationship [high AC / A & other /similar factors]

HYPEROPIA

- Hyperopia is present in a small proportion of children age 6-12 mo
- ethnicity affects prevalence
- higher in certain subgroups esp. family history of hyperopia or accommodative ET.

 \circ 20% of hyperopic infants \Rightarrow esotropia

MATERNAL SMOKING DURING PREGNANCY [ISRAEL; 2012]



INGRAM UK

 $\circ \ge +3.50$ DS in one axis @ age 12 mo:

o 50% risk of strabismus / amblyopia

FACTORS THAT INCREASE THE DEMAND ON FUSION 2 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.
- o USA: 'high AC/A' = near eso > distance eso by $\geq 10\Delta$
- All these subtypes have same 'final common pathway'.
- o LK preference: convergence excess as synonym for all of these terms [after GvN].

ABNORMAL ACCOM - CONV RELATIONSHIP

• Presbyopia

Another age where accomm ET can be seen in pts with fragile motor fusion

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

ABNORMAL ACCOM - CONV RELATIONSHIP

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

Parents don't think of mentioning an enuresis [bed wetting] tablet to the eye Dr

UNDERSTUDIED SUBGROUPS

ASD/ ADHD/....&/or their treatments

- Labile convergence and accommodation
- Will not accept / respond 'normally' to sensible glasses
- Surgery less reliable

HEAD INJURY

 Labile / inappropriate accommodation [under ≈ presbyopia, over = pseudomyopia] & convergence [under ≈ XT or CI, over ≈ convergence Xs ET].

FACTORS THAT DECREASE THE QUALITY OF FUSION

Strabismus develops due to an imbalance between two groups of factors

Factors which increase
the demands on, or lessen
the quality of horizontal
fusion

Factors that optimise the quality of horizontal fusion

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

Neurological

- Abnormal innervation
- Abnormal cortical factors
- 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 \pm 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 1 ABNORMAL OBLIQUE ANATOMY / FUNCTION

1. Atrophic superior oblique

It never developed or Damaged by falling off change table / bike ...

MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION 1

SUPERIOR OBLIQUE ATROPHY



LSO OK RSO ?absent

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

ABNORMAL OBLIQUE ANATOMY / FUNCTION – NON PARETIC

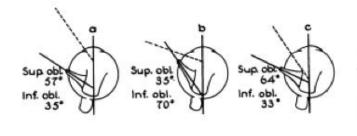
FINK: 20% of cadavers: > 30° difference b/w course of SO & IO

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

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The Role of Developmental Anomalies in Vertical Muscle Defects

Walter H. Fink



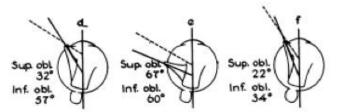


FIGURE 5. DRAWINGS
OF SPECIMENS IN
WHICH THERE ARE PRONOUNCED VARIATIONS
OF THE OBLIQUE MUSCLE PLANES OF ACTION
Continuous line indicates
the superior oblique plane
of action, Broken line indicates the inferior oblique
plane of action.

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

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

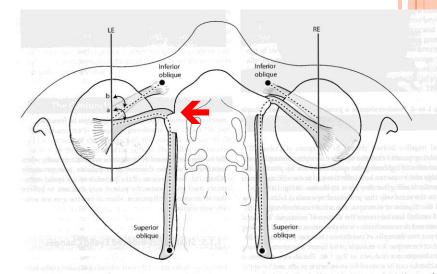


Fig. 1.3. Failure of the trochlea to advance anterior to the equator in a patient with unilateral coronal synostosis may result in reduction of pressing action on the globe with contraction of the superior oblique muscle

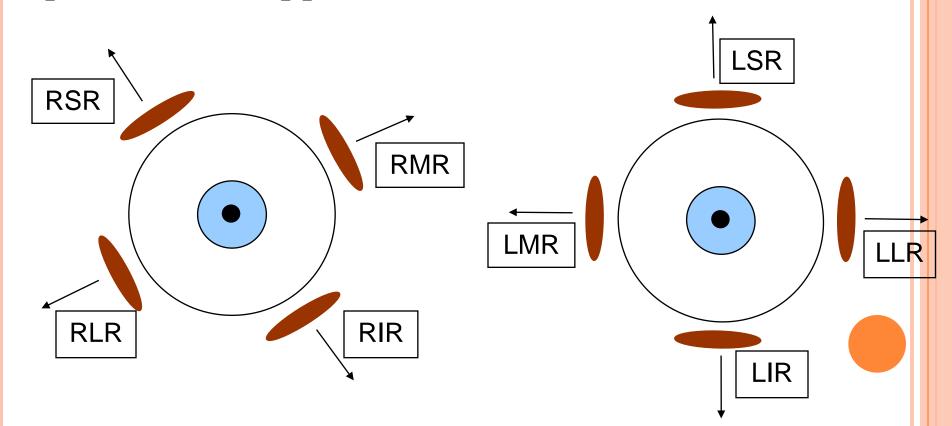
MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION SUBTLE ABNORMALITIES IN ORBITAL ANATOMY 3

- Orbital pulley heterotopyChanges muscle actions
- Intorted / extorted orbit
 More prone to alphabet patterns

...some overlap

MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION – SUBTLE ABNORMALITIES IN ORBITAL ANATOMY 3 EXTORTED ORBIT

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



MECHANICAL FACTORS THAT DECREASE THE QUALITY OF FUSION -

SUBTLE ABNORMALITIES IN ORBITAL ANATOMY 3

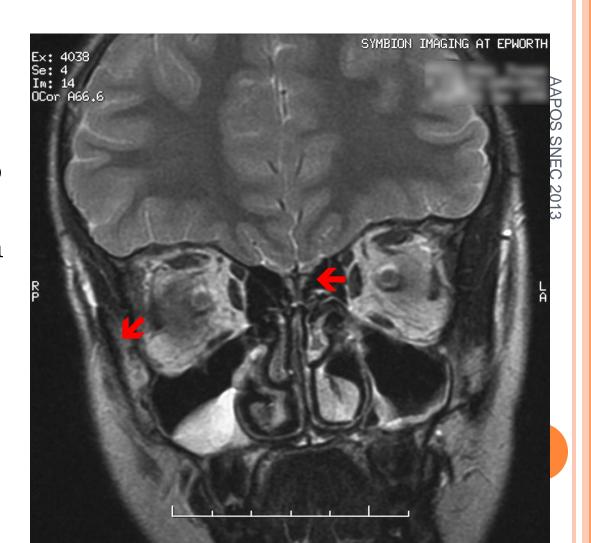
ORBITAL PULLEY HETEROTOPY

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



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
- Amblyopia
- Organic visual loss
- Head injury
- Abnormal innervation

CORTICAL FACTORS WHICH DECREASE THE QUALITY OF FUSION 1

Poor Sensorimotor Fusion

- ↓ motor fusion
 oculomotor 'shock absorber' / 'glue' that tries to keep eyes straight despite pressure to misalign them
- \$\forall \text{ sensory fusion} \text{ stereopsis}
- Abnormal binocular columns

Cortical Factors 2: Structural abnormalities in the brain

Maternal drug use

Usually multiple drugs

Periventricular LeukoMalacia

•PVL

Cortical Factors 2: PVL Peri Ventricular Leukomalacia

Circulation problems @ 32 w gestation.

Causes one/ more of:

Cong ET PVL: 30+ times greater risk of IOS

Congenital nystagmus [both types]

Optic n hypoplasia

Reading problems

Reduced acuity for cortical reasons [CVI]

&

NON-MECHANICAL FACTORS WHICH DECREASE THE QUALITY OF FUSION 3

- Amblyopia
 - oanisometropic amblyopia, amblyopia from congenital cataract, strabismic amblyopia
- Decreased vision from organic causes
 - •Retinal disease any visual pathway disease
- Head injury

IMPAIRED SENSORIMOTOR FUSION: ET happens more readily [with lower or no +]

 Chromosomal defect / devptl delay Amblyopia
 Orbital anomaly
 PVL etc

Factors which increase the demands on fusion

Factors which improve the quality of fusion

THIS
SIDE
NOW
LIGHT
ER

If this side is heavier, there will be strabismus If this side is heavier, there will be no strabismus

TYPES OF STRABISMUS

- 1. Derived from refractive disorders : ESOTROPIA
- 2. ... from abnormal early visual development
- o 3. Orbital causes
- 4. Neurological

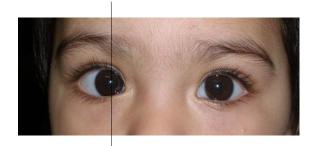
PSEUDO-ET: BEWARE OF DISMISSING AN? ET (NOT PRESENT DURING YOUR TESTING) AS A PSEUDO-ET

- Demonstrate to parents how to interpret light reflexes
- Offer email follow up of any suspicious photos
- 10% will end up with strabismus, ~ 3 TIMES THE BACKGROUND RATE

R PSEUDO ET

Do a thorough search for strabismogenic & amblyogenic factors

MUST include cycloplegic retinoscopy for latent hyperopia





PSEUDO-ET

Determine if 6^ BI will ⇒ ET [poor fusional divergence = 'almost ET']

•MUST check for oblique dysfunction - predisposes to ET in a hyperope

•Every 'ET by history, normal by exam' could have the rare cyclic ET: one day ET, one day straight

PSEUDO STRABISMUS: IS IT?

- o 51 children
- \circ Av age, 1.5 \pm 0.8 y range, 3-36 mo
- Refractive accommodative ET developed in 16% of the children @ mean age of 2.8 ± 1 y.

ET developed in

- 54% of children with pseudoesotropia who were > + 1.5 D c.f.
- **o** 3% of those \leq + 1.50 D (P=0.0001).



This is not Chandigarh, but isn't it a beautiful photo?

PSEUDO STRABISMUS: IS IT?

• Family history of strabismus (P= 0.193) and age @ presentation with pseudoesotropia (P = 0.571) were not predisposing factors.



This is not Chandigarh, but isn't it a beautiful photo?

- Development of refractive accommodative esotropia in children initially diagnosed with pseudoesotropia
- Mohan & Sharma, J AAPOS 2012;16:266-268 Chandigarh

DEVELOPING AN ESOTROPIA...1

THE UNCORRECTED HYPEROPE

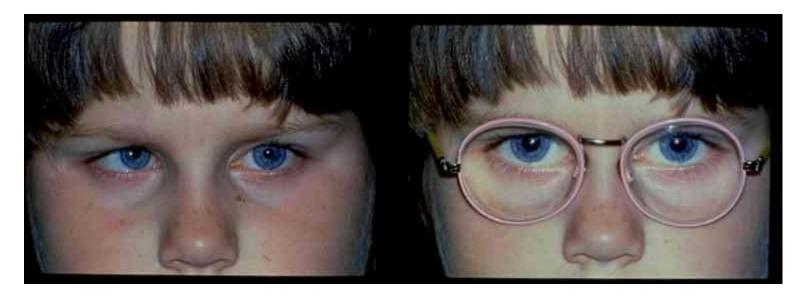
Prolonged accommodation → tendency to prolonged inappropriate convergence and increased tone in medial recti [vergence tonus]

Developing an esotropia...2

THE UNCORRECTED HYPEROPE

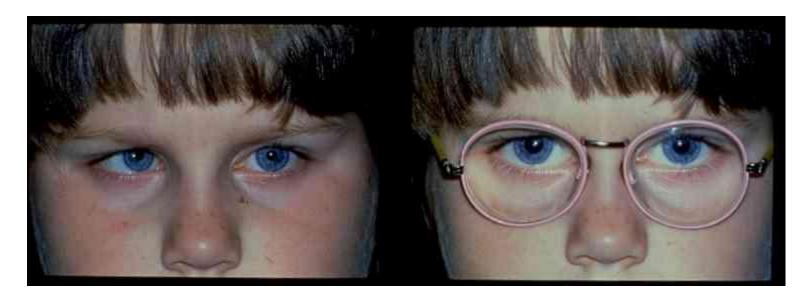
- o Increased tone will lead to changes in Tension / Length ratio and eventually to structural changes in muscle changes in sarcomere density that eventually exceed motor fusional reserve and → esotropia!
- Then muscle starts to permanently shorten
- o SEMINAL SLIDE

'OPTOMETRIC' ESOTROPIA say, +4 DSOU



- Abnormal [& appropriate!] degree of accommodation is required to see clearly
- Abnormal amount of accommodative convergence is generated
- Glasses required to make the child normal
- If you wait too long before you fully compensate with +, you will get structural changes in the MR and glasses alone will be insufficient to straighten the eyes

'OPTOMETRIC' ESOTROPIA



- Exactly the same can happen with low + and abnormal accommodative - convergence relationship = convergence excess.
- If you wait too long before you fully compensate with +, you will get structural changes in the MR and glasses alone will be insufficient to straighten the eyes

ACCOMMODATIVE ESOTROPIA

- Usually 2-5 yrs old
 Second small peak in middle age
- Usually moderate +
- Sometimes low / normal + with convergence Xs
- Background of normal visual devpt in first 6mo of life - normal sensorimotor fusion can be regained

ESOTROPIA ET

ET: core problem is [or becomes] a tight medial rectus

Fixing the abnormal medial rectus length/ tension should return the alignment & mechanics to normal. Often need surgery to do this.

Then you need to keep it normal. Often need strongest-possible hyperopic glasses to do this.

TYPES OF STRABISMUS

- 1. Derived from refractive disorders ESOTROPIA
- 2. Derived from abnormal early visual development
- o 3. Orbital causes
- 4. Neurological

CONGENITAL ESOTROPIA = IOS INFANTILE ONSET STRABISMUS, USU ET





ASSOCIATIONS OF CONGENITAL ET

o Down's 30%

• Bad neonatal course

IVH / HC >>50%

oPVL ?%

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PRINCIPLES OF TREATMENT OF ANY ET

- 1. Give full + [cyclo if young, manifest if older].
- + for amblyopic eye is to optimise vision in the amblyopic eye
- + for fixing eye is optimise alignment of amblyopic eye
- 2. Rx any amblyopia
- 3. Consider realignment for any residual ET after best amblyopia result and + has been rechecked

- + IN ET
- o Always give full +
- o Then check that you have given full +

Over 8-10 yo: a new Q

- Does this child still need full + to stay this good?
- If BIFR > 6, consider cutting by 0.5 DS every 4-6 months

BENEFITS OF REALIGNMENT OF ET

Normal appearance

• Better peripheral field

Chance for sensory fusion

• Better chance to treat resistant amblyopia

THINKING OF SURGERY....

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

- Obenefits
- oRisks
- oHassle / Costs
- Alternative treatments
-I proceed, with the parents' blessings

Parents' expectations have to = mine 1

• 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

o 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

• Child has had unconventional ineffective treatment for some years : need total recalibration of 'religion'

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

MENTIONING DISASTER OUTCOMES: TAILOR TO PARENT

• Most: surgery is 99+% safe – do you want to talk about the rare problems?

Some:

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

Preparation for the hospital experience

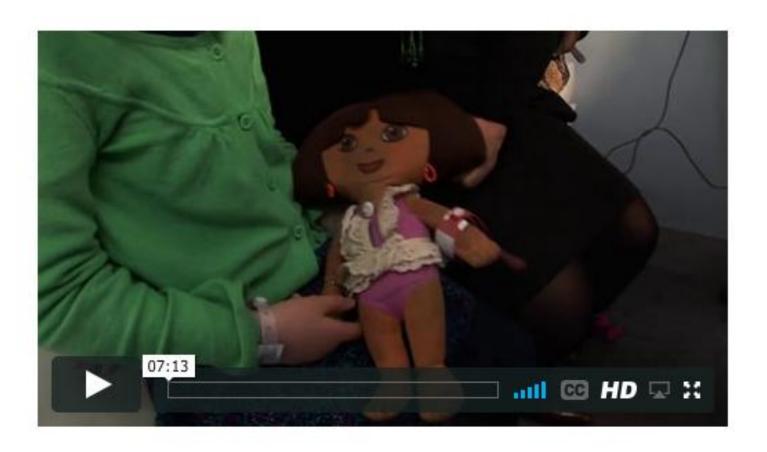
My website:

- o1. Ella's Eye Surgery Experience
- •2. Amy's adventure.
- **3.** Gabriel's Eye Surgery Adventures *
- o4. Briannah's Book
- o5. Kara's adventure *
- o6. Noah's adventure

^{*} not my patient: all others are

HOSPITAL EXPERIENCE

View Kara's visit to the Eye and Ear:



SURGERY FOR REALIGNMENT OF ET

BIMEDIAL RECESSION or

RECESS / RESECT ONE EYE

Conv Xs: BMR

Amblyopia: R-R

 $<35\Delta$ same results

Other:

Botox

Prism



SURGERY

AIM: perfect early alignment

• Expectation: 80- 90%

• IF operating for ET /XT, improve the 'other' factors that have compromised fusion esp. anomalous oblique anatomy /function

SURGERY FOR ET MEDIUM TERM EXPECTATIONS:

Depends on:

- Sensorimotor fusion
- o 1st 12 mo: 10% reoperation − issues with healing, bell curve for surgical doses
- Subsequent: 1% per year consec XT the operation that has repositioned the muscles doesn't 'grow with the patient'

REALIGNMENT OF ET: 2

MEDIAL RECTUS BOTOX

- \circ 50+% success for 10 -20 Δ ET
- 15% temporary ptosis
- 1% permanent acquired vertical

Small number of Drs get GREAT results

• LK 20 p.a. [= 20% of country]

CONGENITAL ET / IOS

Poor motor fusion: insufficient 'capture range' to 'collect' a near- perfect mechanical realignment.

Alignment has to be mechanically perfect.

- Expectation of alignment: 80-90%
- The repositioned muscles may not grow in perfect mechanical balance with growth in the eye & orbit; recurrent tropia more common
- No cortical 'glue' = no motor fusion to help maintain the mechanical alignment in some

ACQUIRED ET:

• Expectation of alignment: 80-90%

Alignment has to be CLOSE. Presence of motor fusion: sufficient 'capture range' to 'collect' a near- perfect mechanical realignment. If a large tropia is improved to a small phoria: success*.

The repositioned muscles may not grow in perfect mechanical balance with growth in the eye & orbit, and motor fusion will often look after that, and keep the deviation as a phoria.

^{*}if there was no motor fusion, this would be tropia=failure

TYPES OF STRABISMUS

- 1. Derived from refractive disorders : ESOTROPIA
- 2. Derived from abnormal early visual development
- 3. Orbital causes: EXOTROPIA
- 4. Neurological

SEMINAL SLIDE 1 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
- 1. subtle anomaly in orbital anatomy [not a tight lateral rectus] &/or
- 2. sensory adaptation to the XT &/or
- 3. 'soft' neurological issues

ET / XT ARE NOT MIRROR IMAGE CONDITIONS

EXOTROPIA XT SEMINAL SLIDE 2

- Core problem is usually subtle anomaly in orbital anatomy, not a tight LR
- A common 2° 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' recurrent XT
- 2. may not alter the suppression pattern even when straight this 'allows' recurrent XT

EXOTROPIA XT SEMINAL SLIDE 3

Common associations can be:

- 1. Neurological problems, both 'soft' & obvious
- o 'Infantile' XT: frequent neurological / developmental issues
- 2. Unilateral visual loss [often amblyopia]
- Poorer prognosis for maintaining good alignment after surgery

EXOTROPIA - BASICS

• Abnormal mechanical balance of orbital tissues & other factors vs. motor fusion & other factors

Divergent orbit & orbital tissues
Unilateral poor VA
Abnormal CNS

Motor fusion

Convergence factors esp. uncorrected +

If this side is heavier, there will be exotropia If this side is heavier, there will be no exotropia

TYPES OF XT: INTERMITTENT XT, D > N

- o Usu 2-7 yo *
- Little / no amblyopia Because often straight
- Motor fusion is typically better for N, so XT worse for D
- Hemiretinal suppression that 'allows' XT without diplopia

*but can deteriorate to 'clinically significant' @ any later age

INTERMITTENT XT: MAYO CLINIC STUDY

• Very high incidence of late myopia

 Higher incidence of adult psychiatric disease

BASICS OF TREATMENT OF XT

Check manifest / cyclo refraction

 High +: give full + to improve peripheral fusion - Paradoxical effect

Treat any amblyopia

BASICS OF TREATMENT OF XT LOOSE GUIDELINES

o < 4y: patching</pre>

• 4-8: minus lenses

• > 6: surgery

BASICS OF TREATMENT: MINUS LENS TREATMENT...TO PROMOTE ACCOMM CONVERGENCE

LK: as much minus as will not interfere with near threshold

Typically -1.5 over the cyclo to start

WHY?: only good alternative is surgery ⇒ >10% have persistent ET ⇒ risk of amblyopia / troublesome diplopia depending on age

BASICS OF TREATMENT: MINUS LENS TREATMENT...TO PROMOTE ACCOMM CONVERGENCE

Usually NOT a long term solution

? risk of promoting / exacerbating any myopic tendency. Wisconsin study: little / no risk

Useful temporising measure to age 7-8

WHO GETS XT SURGERY?

Better outcome if:

- o not quite constant XT
- Medium angle rather than large angle
- Pre-op stereo

BASICS OF TREATMENT: XT SURGERY

- >50% early ET [5-10 Δ desirable]
- <10% persistent ET ⇒ risk of amblyopia / troublesome diplopia depending on age

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

BASICS OF TREATMENT: XT SURGERY OUTCOMES

12 mo results:

10% have needed 2nd surgery 80% excellent

10 yr results:

30% have needed 2nd surgery

OTHER TYPES OF EXODEVIATION

 SENSORY – surgery when it looks bad.
 Sometimes needs multiple surgeries in a lifetime

- o CONVERGENCE INSUFFICIENCY very difficult issues with selection bias
- Mild/ moderate / severe
- CITT trial: did not control for ADHD
- LK: never see pts for whom pencil pushups are useful

TYPES OF STRABISMUS

- 1. Derives from refractive disorders : ESOTROPIA
- 2. Derives from abnormal early visual development
- o 3. Orbital causes
- •4. Neurological: RED FLAGS

RED FLAGS IN STRABISMUS

- ET greater for distance than near
- ET or XT greater to lateral gaze
- Strabismus that varies a lot from morning to evening
- \circ Any vertical $> 5^{\land}$
- A recently symptomatic vertical of any size
- Recent onset nystagmus / oscillopsia
- Recent / variable ptosis

SEMINAL NOTES ON THERAPEUTIC PRISMS

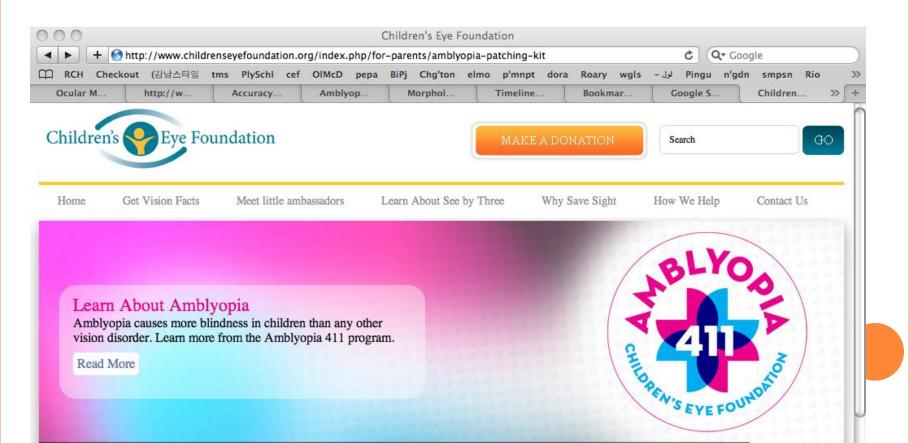
- o Do not use prisms unless you have a diagnosis or are about to get one
- 'Esodeviation' is not an acceptable diagnosis: could be due to thyroid eye disease, presbyopia, 6th nerve palsy, underplussed,

OVERVIEW

- Introduction
- Why to treat & when to treat
- Why strabismus happens
- How to treat
- Amblyopia: Causes and treatment

THIS WILL BE DIFFICULT FOR YOU AND PARENTS

CHILDREN'S EYE FOUNDATION .ORG



AMBLYOPIA

- Normal ocular morphology
- Reversible to some degree
- Often ?usually very asymmetric bilateral condition

Small list of associated / causative factors:

- 1. Anisometropia, astigmatism
- 2. Strabismus
- 3. Any vision- reducing pathology, on wch amblyopia is superimposed

WHY TREAT AMBLYOPIA?

Better spare tyre

More accurate presurgical strabismus measurements

Better sensory fusion : ↑ stereo ⇒ better function

AMBLYOPIA STUDIES: ACRONYMS

PEDIG [USA]:

- Large numbers of clinics / patients
- Simulates community treatment

MOTAS [UK]:

- Few clinics
- High tech electronic patch

WHEN TO TREAT AMBLYOPIA? SUCCESS RATES @ DIFFERENT AGES

3-7 y	75- 85%
7-17 y	25 - 50%
Adult	≤10%

AMBLYOPIA TREATMENTS *WELL STUDIED

Monocular occlusion

o ** Opaque patch popularised by Erasmus Darwin Charles Darwin's grandfather

Asymmetric binocular input

- ** Glasses / CLs
- **Atropine near penalisation. Late 19th century.
- o* Bangerter filters
- o * Optical penalisation
- * Binocular Video games[LK: investigator]

PEDIG: GLASSES ALONE

6/12 to 6/75

o27% cured

•Another $50\% \ge 2$ lines better

•Took up to 7 mo

MOTAS GLASSES ALONE

- o 65 newly diagnosed children
- VA improved from 0.67 [6/24-] to 0.43 [6/15-] (p=0.001)
- 'REFRACTIVE ADAPTATION'

*is this why the CAM stimulator 'worked'?

Br J Ophthalmol 2004;88:1552-1556.

PEDIG 6/12 - 6/24 OCCLUSION & ATROPINE

 \circ 2h/d = 6h/d

 \circ Weekend A = daily A

• 10%: change in strabismus - better or worse

PEDIG 6/30 -6/120

 \circ 6h/d = full time or FT-1h

o 6/15 usual endpoint

SEMINAL SLIDE MOTAS ...SEVERAL STUDIES

1 line gain:

o needs ~ 120h occlusion

2 line gain:

• 4y: needs 170h

o 6y: needs 236h

DOSE-RESPONSE OF OPAQUE PATCH @ DIFFERENT AGES

< 4 years old:

• low doses (<3 h/d) are effective, slight (p=0.54) additional gains for doses >3h/d

4-6 years old:

- significant differences between <3h/d & 3-6h/d
- no difference between 3-6h/d & 6-12h/d
- > 6 years old:
 - <3h/d has little effect; need >3h/d

CONCLUSIONS OF AMBLYOPIA RECURRENCE STUDY

- o ¼ of successfully treated amblyopic children experience a recurrence over 1 year of f/u
- Recurrence risk similar for stopping patching and stopping atropine
- Most recurrences occur < 3 mo early follow-up is critical, but long term follow-up is also important
- o If ≥ 6h of patching stopped recurrence risk is lower if patching is reduced to 2h/d before cessation "weaning" is beneficial

WHEN IT DOESN'T WORK FOR YOUR PATIENT: IS IT THE PARENTS?

 Parents avoid parading an obviously defective child & will not patch in public

• Parents do not want to inflict discomfort on their child

RECRUITING PARENTS TO TREAT THEIR CHILD / YOUR PATIENT



Types of parents

- Type A no excuses:
- on Thursday we only did 5h 20m, so we made up for it on Friday with 6h 40m
- Type B:
- We're careful to do it all the time.. but we forget sometimes when we're busy....
- Type C great excuses:
- s/he hates it.... we haven't managed for the last week.... s/he was sick... we were on vacation... we let the nanny look after it.... s/he only does it @ school...

AWAN M, PROUDLOCK FA, GOTTLOB I THE EFFECT AND COMPLIANCE OF STRABISMIC AMBLYOPIA MONITORED WITH THE ODM.

INVEST OPHTHALMOL VIS SCI 44[SUPPL]: S199, 2003]164,483).

• Parent diaries overestimate actual patching time by a factor of 2-3 even when they know it is monitored by an electronic Occlusion Dose Monitor and will be checked!

STRABISMIC AMBLYOPIA

• Alignment can result in better response to amblyopia therapy...or no need for amblyopia therapy in 20%?

TIMING OF AMBLYOPIA THERAPY RELATIVE TO STRABISMUS SURGERY

Guyton et al, Ophthalmology, Dec 1993

- 47 children < 8 y with both amblyopia and esotropia.
- o 26: amblyopia fully treated before surgery
- 21 : surgery before completing amblyopia therapy.
- o 5/21 did not require amblyopia therapy after surgery even though they were amblyopic before operation.

HELPING THE PARENTS: THERAPEUTIC ENVIRONMENT

 Some parents need help to maintain enthusiasm for a task which everyone finds difficult

• Keep the therapeutic environment alive / active e.g. ring daily

NEW/ UPCOMING TREATMENTS

- Handheld device based games BRAVO study, etc: no better than patch
- Video goggles based treatment: no good data
- Electronic shutter glasses AmblyZ: no better than patch
- Pharmacotherapy Levodopa, Citicholine: largely abandoned even by enthusiasts
- Perceptual learning vision therapy -NeuroVision/ RevitalVision: no good data
- Transcranial Random Noise Stimulation (tRNS): no good data

REMEMBER THE BASIC 2 STEP MANAGEMENT OF STRABISMUS

- 1. Improve /equalize acuity
- 2. Straighten the eyes
 - Optically
 - Botox
 - Surgically

