Strabismus Research Update

ARVO Asia
Brisbane 2017

Lionel Kowal  Melbourne

With help from recent Fellows
Drs Sheth, Kini, Mitchell

No financial conflicts
Today: Some of the advances in recent years that have changed or are changing the understanding & treatment protocols and options in sensory and motor strabismus
AMBLYOPIA
PEDIG & MOTAS

- Rx often based on quantifying the sensory asymmetry & treating it with asymmetric treatments
- Glasses alone sometimes effective
- Less treatment is often as effective as more
- **Atropine** [used for 100 yrs] & **opaque occlusion** [used for 300 yrs] : equivalent effect for many pts
- **There are NO other treatments still used in medicine that are as old as these**
21st Century Amblyopia Rx
High Tech Asymmetric Rx

Binocular treatment of amblyopia using videogames (BRAVO): study protocol for a randomised controlled trial.
Guo CX, ….Kowal L….Trials. 2016 Oct 18;17(1):504

Using asymmetric high tech inputs
• Blurred video game to good eye
• Clear video game to amblyopic eye

Results expected in next few weeks
21st Century Amblyopia Rx 2

- **Effect of a Binocular iPad Game vs Part-time Patching in Children Aged 5 to 12 Years With Amblyopia: A Randomized Clinical Trial.**
  - Holmes JM, ......PEDIG
  - JAMA Ophthalmol. 2016 Dec 1;134(12):1391-1400

- **Binocular Treatment of Amblyopia in Children: Teething Problems on the Path to Clinical Practice.**
  - Dahlmann-Noor A. JAMA Ophthalmol. 2016 Nov 3
The role of Interactive Binocular Treatment system in amblyopia therapy.


Rajavi Z1, et alii

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- Invited Commentary
- New Treatments for Amblyopia—To Patch or Play?
- John Sloper, Moorfields, London
For more than 100 years, ophthalmologists have been wagging their fingers at reluctant, amblyopic children, urging them to wear an eye patch that they do not want to wear, while frightened parents have coaxed, begged, bribed, threatened, and cajoled their stubborn little ones to comply. In the current issue of the Journal of AAPOS and in a recent issue of the journal Eye, Birch and colleagues have conducted the first pediatric studies of a binocular therapy for amblyopia that allows parents to offer their children not an eye patch but an iPad. In both studies, children who played games on a handheld tablet computer equipped with red-green glasses had a statistically significant improvement in visual acuity in the amblyopic eye. Vision was gained with as little as 4 hours of treatment in some cases, with a few patients improving to 20/20. Are we ready, then, to abandon the burden of forced monocular occlusion in favor of encouraging our young patients to play binocular video games on their tablet computers? Unfortunately, no, we are not—not just yet, anyway.
• Some published data. Many more papers presented @ meetings
• I expect this will a popular 1\textsuperscript{st} treatment for amblyopia
• Commercial competition ++ expected.
Radiology of strabismus: Orbital pulleys

• Orbital pulleys have been recognised >100 yrs ['poules' in 19C French literature].
• Clinical relevance has been appreciated for ~20 years and practical application of the knowledge is growing fast

Today:
• Childhood pulley disorders
• Acquired pulley disorders – elderly and high myopes

Pulley surgeries:
• The medial rectus pulley
• The lateral rectus
Radiology of strabismus
Recent findings: orbital pulleys

Incomitant Strabismus Update

The Role of Extraocular Muscle Pulleys in Incomitant Non-Paralytic Strabismus

Robert A. Clark

Abnormal location of the pulleys could explain many cases of incomitant strabismus, conventionally [& without scientific justification] attributed to ‘oblique muscle dysfunction’.

“A” pattern ET
LR displaced sup to MR and
SR displaced nasal to IR
As if orbital contents INtorted

“V” pattern ET
Inf displacement of LR (R>L)
As if orbital contents EXtorted

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J AAPOS

A 12-Year, Prospective Study of Extraocular Muscle Imaging in Complex Strabismus

Joseph L. Demer, MD, PhD, Robert A. Clark, MD, Reika Kono, MD, PhD, Weldon Wright, MD, Federico Velez, MD, and Arthur L. Rosenbaum, MD
Childhood pulley disorders

12 yo
previous LR & IO
weakening for V - XT

Recurrent / Residual
V- XT [UG 30, DG 10]
Minimal IO OA
No fundus torsion
Coronal MRI T1: inf positioning of LR (L>R), and nasal shift of IR
Infraplaced Lateral rectus seen before the medial rectus
At upper edge of MR, LR no longer seen

Childhood pulley disorders: unknown % of childhood strabismus esp incommotant strabismus
Acquired pulley disorders
Common(?est) cause of small angle ET +/- vertical in the healthy elderly

Sagging Eye Syndrome

Connective Tissue Involution as a Cause of Horizontal and Vertical Strabismus in Older Patients

Zia Chaudhuri, MS, FRCS(Glasg); Joseph L. Demer, MD, PhD

Figure 2. Fast spin-echo T2-weighted sequence quasi-coronal plane magnetic resonance imaging. Left, Younger control participant showing lateral rectus (LR)-superior rectus (SR) band. Note the normal morphology of LR muscle with respect to a horizontal reference line drawn through the globe center. Middle, Elderly control participant demonstrated marked elongation of LR-SR band associated with LR muscle sag. Right, Rupture of LR-SR band in sagging eye syndrome (SES) with resultant LR sag. IR indicates inferior rectus; MR, medial rectus; and SO, superior oblique.
Acquired LET: Sagging LLR

Downward displacement of the LLR changes its vector and causes an abduction deficit.

Surgical correction of an inferiorly displaced lateral rectus with equatorial myopexy

Tiana Y. Clark, Robert A. Clark, MD
Family Eye Medical Group, Long Beach, California
Acquired pulley disorders: Extreme Esotropia of High Myopia [aka Heavy Eye]

Preoperative

Postoperative (52 days after surgery)

Dr Yokoyama’s case
Extreme myopia:

LR displaced down, SR nasal

Preoperative: LR displaced down, SR nasal

Postoperative: LR displaced up, SR nasal

From Tsuranu Yokoyama
Pulley surgeries:
The medial rectus pulley suture:
a safer alternative to posterior scleral fixation

Medial rectus muscle pulley posterior fixation sutures in accommodative and partially accommodative esotropia with convergence excess

Logan Mitchell, MBChB, FRANZCO, a and Lionel Kowel, MBBS, FRANZCO a,b,c

BACKGROUND

The use of medial rectus pulley posterior fixation sutures to treat esotropia with convergence excess has limited support in the literature. We describe our results using this technique to treat patients with large near-distance disparities.

J AAPOS 2012
Each half of each EOM* has a unique non-overlapping nerve supply

Compartmentalization of extraocular muscle function

* Compartmentalization not [yet?] demonstrated for superior rectus
abducens motor neuron pools. Humans, monkeys, and other mammals demonstrate separate, nonoverlapping intramuscular nerve arborizations in the superior vs inferior compartments of the medial rectus (MR) and lateral rectus (LR) EOMs that could apply force at the superior vs inferior portions of scleral insertions, and in the medial vs lateral compartments of the superior oblique that act at the equatorial vs posterior scleral insertions that might preferentially implement incycloduction vs inftraduction.
Clinical implications of EOM compartmentalization

- 1. Sup compartment LR atrophy:
  Esotropia – of – obscure - cause, not- quite LR palsy
  Not rare

- 2. Sup compartment MR atrophy
  Progressive exotropia – of- obscure- cause
  Rare – no published cases yet

- 3. Medial / Lateral Sup Obl compartment atrophy
  Probably explains why some have vertical diplopia vs torsional diplopia vs both V & T

4. Probably Many more incomitant clinical scenarios waiting to be appreciated
1. Esotropia & compartment hemi-atrophy

• Superior compartment atrophy of the LR produces a clinical condition that resembles LR paresis

• Clinical picture: more LR function than complete palsy. Treatment implications uncertain
Lateral Rectus Superior Compartment Palsy

ROBERT A. CLARK AND JOSEPH L. DEMER


Total RLR atrophy = palsy

Sup compartment RLR palsy

Clinical picture: more LR function than complete palsy. Treatment implications uncertain
Adult ET of uncertain cause

Delayed diagnosis of superior compartment LR paresis

9 years after first presentation and 3 years after the last MRI and the 3rd horizontal rectus surgery, the diagnosis is clearer.

2012 MRI re-read for purpose of a talk. RLR has a triangular appearance

Clark & Demer Lateral Rectus Sup compartment palsy. Amer J Ophth 2014; 157: Fig 2 on page 481
2. Exotropia and compartment hemi-atrophy

- Diplopia onset 66 yo
- 68yo: increased prism to 10Δ
- 69yo: ...to 24Δ
- 70yo: D:50Δ, N: 60Δ

- MR -2mm OU
Bilateral asymmetric atrophy of the superior half of medical rectus compared to inferior.

Asymmetry can be expected to produce a small vertical.

Effective lowering of the MR vector might cause an ‘A’ pattern.

Surgery and Course.
MR plicate/resect OU. LR recess x1. Adjustables.
10 w followup: single vision, small phorias
Medial compartment of SO [=SOm] controls torsion
Lateral ....[=SOl] controls vertical movement
B,C,D: 20% develop floppy tendons requiring tendon tightening surgery
Flap tear of the inferior rectus
Common / ? Commonest cause of vertical diplopia after orbital trauma

Normal [contralateral] IR

Superficial layer of inferior rectus ‘shaved’ off

Affected IR
Outer ½ has been shaved off
From Irene Ludwig

Example where ‘divot’ of muscle has been ‘sliced’ off

Large divot ~50%

Normal contralateral IR
Flap Tear Hypothesis

From Irene Ludwig

• Blunt trauma causes outward traction on orbital septae.
• Orbital connective tissue, which attaches onto EOM pulls away portion of muscle & weakens it.
• Flap can acts as a tether, further restricting & complicating motility.
MRI images of the flap tear

048 Magnetic resonance imaging (MRI) of inferior rectus (IR) flap tears. Tina G. Damarjian, Joseph L. Demer
Introduction: Thus far, the only evidence for existence of flap tears in EOMs has been observations during surgical repair. This study employed MRI to investigate anatomy of flap tears of the IR.
Methods: Five adults (ages 25-68 years) who sustained trauma to the IR were studied prospectively using a 1.5 Tesla MRI scanner with surface coils and fixation targets. Sagittal and coronal T2 sequences were performed in upward, downward, and central gaze for each eye, permitting comparison with age-matched controls.
Results: Patients exhibited infraduction limitation greatest in abduction, and concomitant ipsilateral hypertropia greatest in infradversion. All but one had associated orbital fractures. Three torn IRs exhibited a longitudinal fissure separating the orbital (OL) and global layers (GL), with avulsion of the GL from the sclera in one case, and avulsion of the OL from its pulley in two cases. Two involved IRs exhibited a longitudinal fissure separating medial portion of the GL that was attached to the sclera, from the avulsed lateral portion. All cases manifested extensive inferior orbital scarring. Surgical repair was possible in 3 cases.
Discussion: Blunt trauma may cause longitudinal tears in the IR having variable orientations: longitudinal separation of GL from

Validates Irene Ludwig’s observations first published in 2001!
Intramuscular injection of 3% Bupivacaine BP with Botox BT to treat strabismus

- The unwanted changes caused by accidental injection of local anaesthetic agents like Bupivacaine into EOM during ocular anaesthetic procedures can be exploited to treat the ‘weak’ muscle in strabismus eg the MR in consecutive XT [combined with Botox BT to the LR].

Pharmacologic injection treatment of comitant strabismus

Iara Debert, MD, PhD, Joel M. Miller, PhD, Kenneth K. Danh, BS, and Alan B. Scott, MD

PURPOSE

To report the magnitude and stability of corrections in comitant horizontal strabismus achieved by injecting bupivacaine (BPX, optionally with epinephrine) and botulinum A toxin (BTXA) into extraocular muscles of alert adult subjects with electromyographic (EMG) guidance.

J AAPOS 2016
<table>
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<tr>
<th>Time post injection</th>
<th>Evolution of RLR changes (post BT)</th>
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BT yet to show effect, BP showing full anesthetic effect. Full effect of BT. Anesthetic effect of BP worn off, secondary changes to RMR begin.

BT effect worn off fully.

ET 4△ Near, 6△ Distance

XT 12△ Near, Ortho Distance

ET 1△ Near(Variable), Ortho Distance
Why does this transposition often work so well?

Superior Rectus Transposition and Medial Rectus Recession for Duane Syndrome and Sixth Nerve Palsy

Reshma A. Mehendale, MD; Linda R. Dagi, MD; Carolyn Wu, MD; Danielle Ledoux, MD; Suzanne Johnston, MD; David G. Hunter, MD, PhD

Innovator: Dr Earl Crouch
Validated by this paper from Harvard
Technique of SR transposition & LMR Rc

WHY DOES SUCH AN ASYMMETRIC OPERATION HAVE SO FEW UNEXPECTED CYCLOVERTICAL COMPLICATIONS?
Useful in traumatic 6ths

Augmented superior rectus transposition with medial rectus recession in patients with abducens nerve palsy

Preeti Patil-Chhablani, DNB, Krishnapriya Kothamasu, VR, DO, Ramesh Kekunnaya, FRCS, Virender Sachdeva, MS, and Vivek Warkad, MS

PURPOSE
To evaluate the surgical outcome of augmented superior rectus transposition (SRT) and medial rectus recession (MRe) in patients with abducens nerve palsy.

J AAPOS Dec 2016

WHY DOES SUCH AN ASYMMETRIC OPERATION HAVE SO FEW UNEXPECTED CYCLOVERTICAL COMPLICATIONS? ........SO MUCH MORE TO KNOW
THANK YOU

készöm  děkuji
mahalo       고맙습니다
thank you     merci   谢谢 danke
so little time   so much to learn

どうもあ