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

חננו מאתך דעה בינה והשכל

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 1

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

- <u>Effect of a Binocular iPad Game vs Part-time Patching in</u> <u>Children Aged 5 to 12 Years With **Amblyopia**: A <u>Randomized Clinical Trial.</u>
 </u>
- 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.

• <u>Dahlmann-Noor A¹</u>. <u>JAMA Ophthalmol</u>. 2016 Nov 3

21st Century Amblyopia Rx 3

The role of Interactive Binocular Treatment system in amblyopia therapy.

<u>J Curr Ophthalmol. 2016 Aug 9;28(4):217-222.</u> <u>Rajavi Z¹, et alii</u>

- Invited Commentary
- New Treatments for Amblyopia—To Patch or Play?
- John Sloper, Moorfields, London
- JAMA Ophthalmol. Published online November 10, 2016. doi:10.1001/jamaophthalmol.2016.4296

Editorial

Treatment of amblyopia: the "eye pad," or the iPad?

David G. Hunter, MD, PhD

or 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 AAPOS⁴ and in a recent issue of the journal Eye,² 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.

iter strabismus surgery thanks to binocular summa-

Editorial J AAPOS Feb 2015



- Some published data. Many more papers presented @ meetings
- I expect this will a popular 1st 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

<u>Today:</u>

- Childhood pulley disorders
- Acquired pulley disorders elderly and high myopes
 <u>Pulley surgeries:</u>
- 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

Middle East African Journal of Ophthalmology, July-Sept, 2015, pp 279-285

Demer, Clark, Miller:



Fig. 2. $211 \,\mu\text{m}$ resolution, 1 mm thick coronal CT scan of the orbits of a 5 year old girl with large "V" esotropia and marked overelevation and underdepression of the right eye in adduction. Note inferior displacement of right LR more than left LR.

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

Advances in Strabismology



Fig. 3. CT scan of 6 year old girl with "A" pattern esotropia 60Δ greater in elevation than depression. Note LR displaced superiorly and SR displaced nasally in both eyes.

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

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

> J AAPOS Major Articles A 12-Year, Prospective Study of Extraocular Muscle Imaging in Complex Strabismus

Joseph L. Demer, MD, PhD,^{a,b} Robert A. Clark, MD,^a Reika Kono, MD, PhD,^c Weldon Wright, MD,^{a,d} Federico Velez, MD,^a and Arthur L. Rosenbaum, MD^a

Childhood pulley disorders

<u>12 yo</u> 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





Fig. 2. 211 μ m resolution, 1 mm thick coronal CT scan of the orbits of a 5 year old girl with large "V" esotropia and marked overelevation and underdepression of the right eye in adduction. Note inferior displacement of right LR more than left LR.





Childhood pulley disorders: unknown % of childhood strabismus esp incomitant 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 L ET : Sagging LLR

Downward displacement of the LLR changes it's vector and causes an aBduction deficit



October 2016 Volume 20, Issue 5, Pages 446.e1-446.e3

Next Article >

Surgical correction of an inferiorly displaced lateral rectus with equatorial myopexy

Tiana Y. Clark, Robert A. Clark, MD Market Family Eye Medical Group, Long Beach, California

< Previous Article



Acquired pulley disorders: Extreme Esotropia of High Myopia[aka Heavy Eye]

Preoperative



Postoperative (52 days after surgery)



Extreme myopia:

LR displaced down, SR nasal



Preoperative

Postoperative

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 Kowal, 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

Eye (2015) 29, 157–162 © 2015 Macmillan Publishers Limited All rights reserved 0950-2

www.nature.com/eye

Compartmentalization of extraocular muscle function JL Demer



* 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 infraduction.

Clinical implications of EOM compartmentalization

• <u>1. Sup compartment LR atrophy:</u>

Esotropia – of – obscure - cause, not- quite LR palsy Not rare

• <u>2. Sup compartment MR atrophy</u>

Progressive exotropia – of- obscure- cause

Rare – no published cases yet

• <u>3. Medial / Lateral Sup Obl compartment atrophy</u>

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

Clark, R. A., & Demer, J. L. (2014). Lateral rectus superior compartment palsy. American Journal of Ophthalmology, 157(2)



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

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



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

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



Surgery and Course. MR plicate/resect OU. LR recess x1. Adjustables. 10 w followup: single vision, small phorias Asymmetry can be expected to produce a small vertical.

Effective lowering of the MR vector might cause an 'A' pattern

Different types of Superior Oblique Palsy/Paresis

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Medial compartment of SO [=SOm] controls *torsion* Lateral[=SOI] controls *vertical* movement B,C,D: 20% develop *floppy tendons* requiring tendon tightening surgery

<u>Flap tear of the inferior rectus</u> 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



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.





ns have a c ophthalc dramati-

nts with cle on.

in thyroid nt vertical tal rectus

e in 67 pahypotropia cle recesssion plus ided: age, sition, dist for each d 2. and 24.5^{Δ} 05). Mean 2.5^{Δ} and 9^{Δ} \pm 4.5^{Δ} postoper- 2 , respecto increased actin-myosin lattice spacing, so that density decreases.

This Chan LR, t mm³ Conc cont 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 incomitant ipsilateral hypertropia greatest in infraversion. 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

NEX

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,^{a,b,c,d} Joel M. Miller, PhD,^{a,b} Kenneth K. Danh, BS,^{a,b} and Alan B. Scott, MD^{a,b}

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.



Time post injection	Evolution of RLR changes (post BT)	Evolution of RMR changes (post BP)	Evolution of alignment
PRE	INJECTION		138
4 days			00
2 weeks		00	00
3 weeks	00	00	
4 weeks	66,	120	00
5 weeks	60	60	
6 weeks	661	60	66
7 weeks	66.	66	66
9 weeks	66,	66	00
14 weeks	1010	0	
17 weeks		60.	60
6.5 months	60	62.	66

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 \triangle Near, 6 \triangle Distance

XT 12∆ Near, Ortho Distance

ET 1∆ Near(Variable), Ortho Distance

Why does this transposition often work so well?

CLINICAL SCIENCES

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



Useful in traumatic 6ths

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



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

PURPOSE To evaluate the surgical outcome of augmented superior rectus transposition (SRT) and medial rectus recession (MRc) 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

