

AVC 2017

A primer on palsies

aka

*The science and art of
handball :
when have to do it fast,
when you can do it slow*



LIONEL KOWAL

MELBOURNE

MAGPIE

A PRIMER ON PALSIES

- This is a Very difficult area
- A condition of the same appearance can be innocent in one pt & life threatening in another
- It's not just the palsy, it's what goes with it
- Always be worried by your ignorance and your lack of clinical experience in this difficult area of motor n-ophthalmology: your patients will benefit

THREE SEMINAL SLIDES ABOUT WHEN TO HANDBALL QUICKLY

- 1. SUSPECTED SIXTH N PALSY IN INFANCY
- 2. CAN'T DO A CONFIDENT EXAM ON A CHILD WITH ET
- 3. 3rd N PALSY full or partial



Ptosis, dilated pupil, XT, hypo

FIRST OF THREE SEMINAL SLIDES

SIXTH N PALSY IN CHILDHOOD esp <12 mo old

- Abduction deficit is the key diagnostic finding
- Rarely it's a 6th
- PLEASE PANIC in case it is a 6thbut all longstanding large angle ET of *any* cause will have aBduction deficit

*Don't be embarrassed to refer:
EVERYONE finds this difficult*

FIRST OF THREE SEMINAL SLIDES

SIXTH N PALSY IN CHILDHOOD esp <12 mo old

- Abduction deficit is the key.
- Sometimes post viral.
- RARELY a tumour.
- Can be 2ary to raised ICP
- Sometimes is Duane's.
- Sometimes is 'regular' large angle ET with amblyopia
- They can all look **exactly** the same.

*Don't be embarrassed to refer:
EVERYONE finds this difficult*

SECOND OF THREE SEMINAL SLIDES CAN'T DO A CONFIDENT EXAM ON A CHILD WITH ET

- See again tomorrow or the next day
- If still can't do a confident exam, arrange child to be seen by someone else in 1-2 days

Don't miss:

- afferent pupil defect
- raised ICP/ swollen discs
- unilateral intraocular tumor
- optic atrophy [as clue for intracranial pathology]

*Don't be embarrassed to refer:
EVERYONE finds this difficult*

HOW OFTEN IS 'TYPICAL' ACQUIRED CHILDHOOD ET WITH NO CNS CLUES DUE TO BRAIN TUMOUR?

- 1. MAYO / MOHNEY: 0
- 2. RVEEH CONSULTANT: 0
- 3. RCH CONSULTANT: 0
- 4. WESTMEAD CHILDRENS VMO : n=7!
- 5. **MANY** ANECDOTAL CASES IN LITERATURE

- *This makes us all nervous & anxious - will the next pt be the one?*

THIRD OF THREE SEMINAL SLIDES

3rd N PALSY full or partial

- Ptosis, big [blown] pupil
- XT, hypo
- Some [adults] with 3rds: due to aneurysm & will die or have terrible neurological outcomes if not fixed in a few days.

• *Painful 3rd n palsy is an emergency*

THREE SEMINAL SLIDES ABOUT WHAT EYE DRS OFTEN GET WRONG WITH PALSIES

- 1. VERTICALS
 - 2. OCCULT THYROID EYE DISEASE
 - 3. STRABISMUS OF AGING
 - 4. TERMINOLOGY: PARESIS vs PALSY
 - 5. STRABISMUS OF MYOPIA
-
- 1-3: CAN LOOK LIKE PALSY

I. VERTICALS

Sudden onset vertical diplopia

- Is it a 4th n palsy? FNP
- Is it a skew deviation? SKD
- Is it important to know?
- SKD: accompanying vestibular symptoms / signs
- FNP: preferred head tilt fixes vertical diplopia
- SKD: preferred head tilt doesn't fix diplopia
- SKD: diplopia goes away when pt lies down;
FNP: no change when pt lies down

Is it a Sup obl palsy?

**Clinically diagnosed SOP =
'true' SOP only ~ 1/2 the time**

Demer & Herzau:

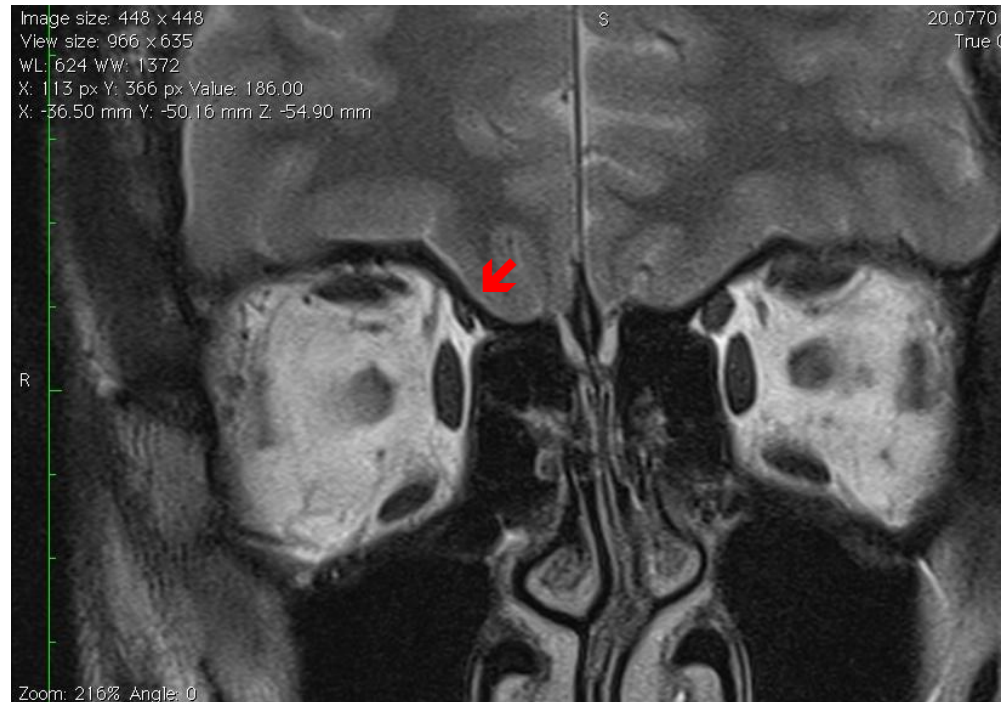
- **Radiological SOP found in ~ 50% of those with Clinical SOP**

Demer JL et al MRI of the functional anatomy of the sup obl muscle.

IOVS 1995 & 1994 AAPOS / ISA joint meeting proceedings

Siepmann K, Herzau V

Klin Monatsbl Augenheilkd. 2005
May



2. THYROID EYE DISEASE

- There are many cases of TED where the ONLY presentation is diplopia:
- Can look like 4th or 6th
- Need MRI for diagnosis

A few will have:

- +ve TRAB
- remote history of Graves'
- recent onset Graves'
- progressive eye disease

3/I. STRABISMUS OF AGING

- Very difficult : must segregate serious pathology from ‘interesting’ aging changes
- Some are due to cranial arteritis
- Can rarely cause just diplopia
- You have to ‘fish’ for: Headache, scalp tenderness, malaise: urgent ESR [ED, GP or ophthalmologist]

THREE SEMINAL SLIDES ABOUT WHAT EYE DRS OFTEN GET WRONG

3/2. STRABISMUS OF AGING: PSEUDO 6thS

- Symmetric sagging of horizontal rectus pulleys: reason for reduced upgaze in the elderly
- ***Asymmetric sagging of horizontal rectus pulleys: common[est?] reason for small angle esotropia, distance > near, in the elderly***
- Can look like a 6th n paresis: sudden onset of abduction defect

Acquired pulley disorders

Common(?est) cause of small angle ET +/- vertical in the healthy elderly

CLINICAL SCIENCES

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

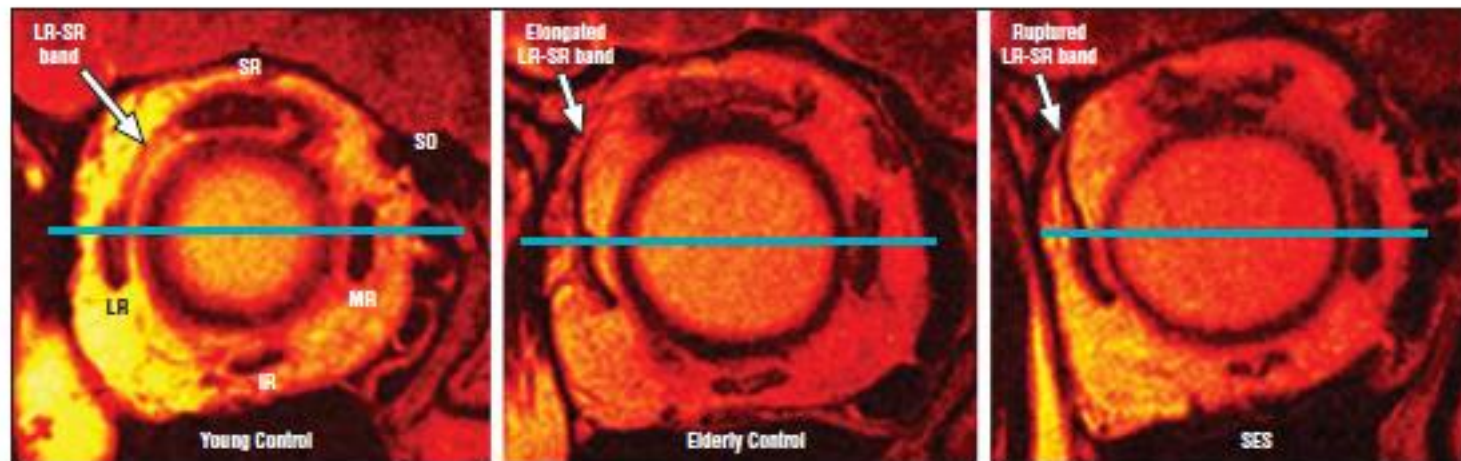
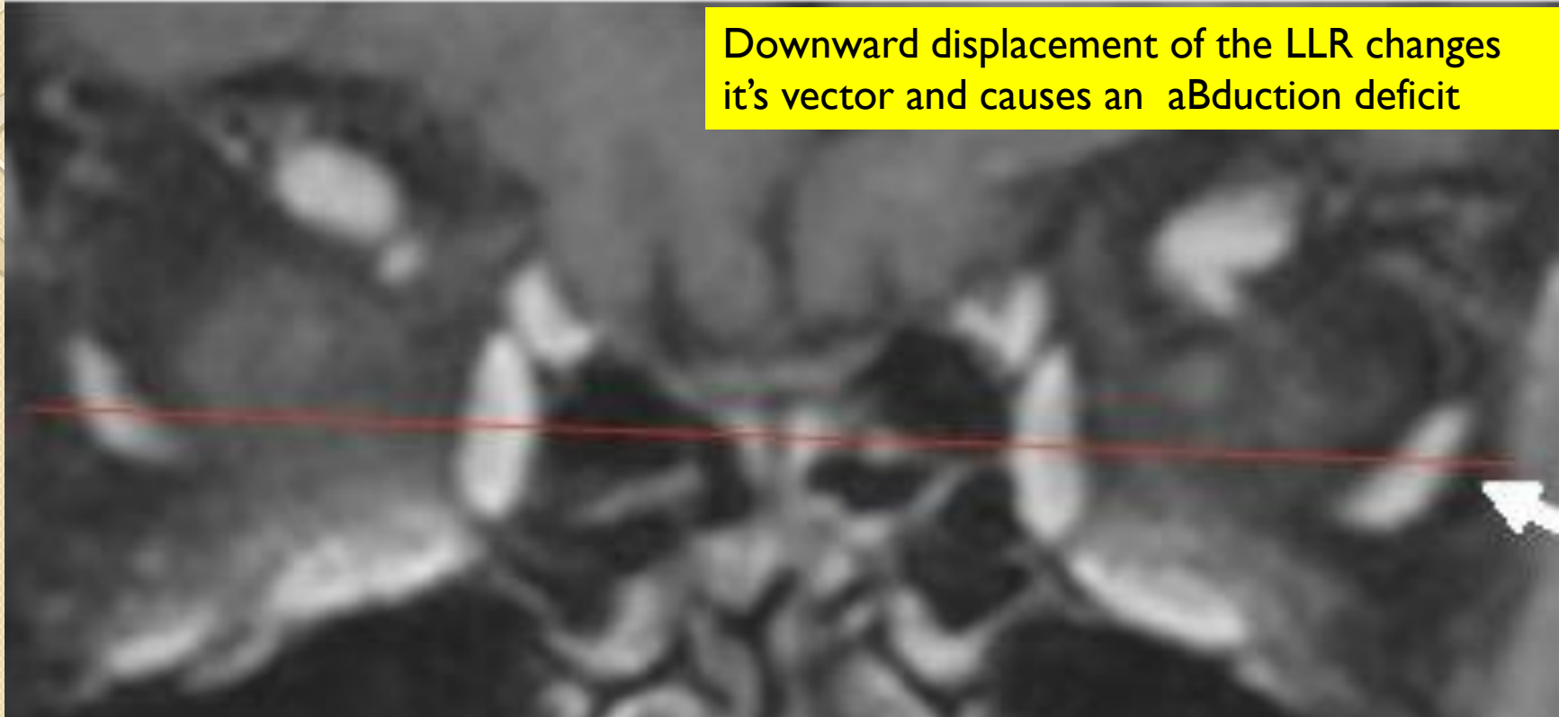


Figure 2. Fast spin-echo T2-weighted sequence quasi-coronal plane magnetic resonance imaging. Left, Younger control participant showing lateral rectus (LR)–superior rectus (SR) band. Note the normal morphology of LR muscle with respect to a horizontal reference line drawn through the globe center. Middle, 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 its vector and causes an abduction deficit



[< Previous Article](#)

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

[Next Article >](#)

JAAPOS

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



A 'Real' 6th: may not follow classical 'rules'

Diplopia 10+ yrs

R 6th. No definite cause

R gaze 45 Δ ET,

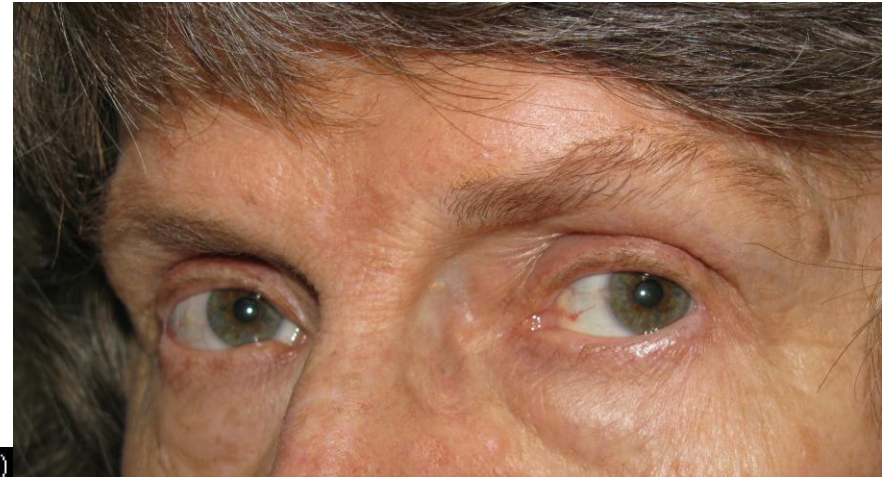
Primary 30 Δ [R fixation 45 Δ].

L gaze 10 Δ

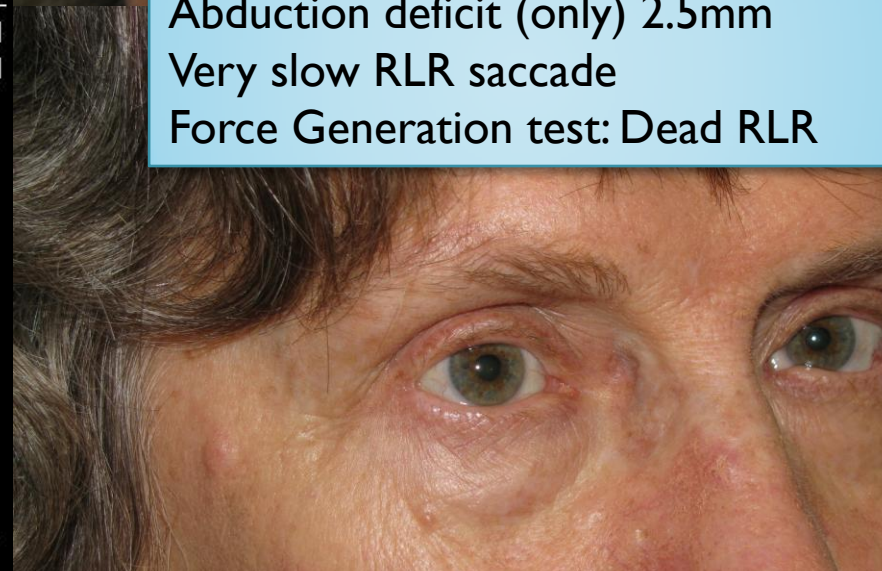
512 x 512 R K4213471 (68 y , 68 y)
45 x 635 T1 SPIR Ax Thins Post Gad SENSE
W: 3510 00001
497 px Value: 0.00 901
m Y: 62.67 mm Z: -9.58 mm



Shoe lace RLR



Abduction deficit (only) 2.5mm
Very slow RLR saccade
Force Generation test: Dead RLR



The 21st century talk on palsies

- Traditional teaching on sup obl palsy is wrong
- Traditional teaching on rectus and oblique innervation is wrong

Is it a Sup obl palsy?

**Clinically diagnosed SOP =
'true' SOP only ~ 1/2 the time**

Demer & Herzau:

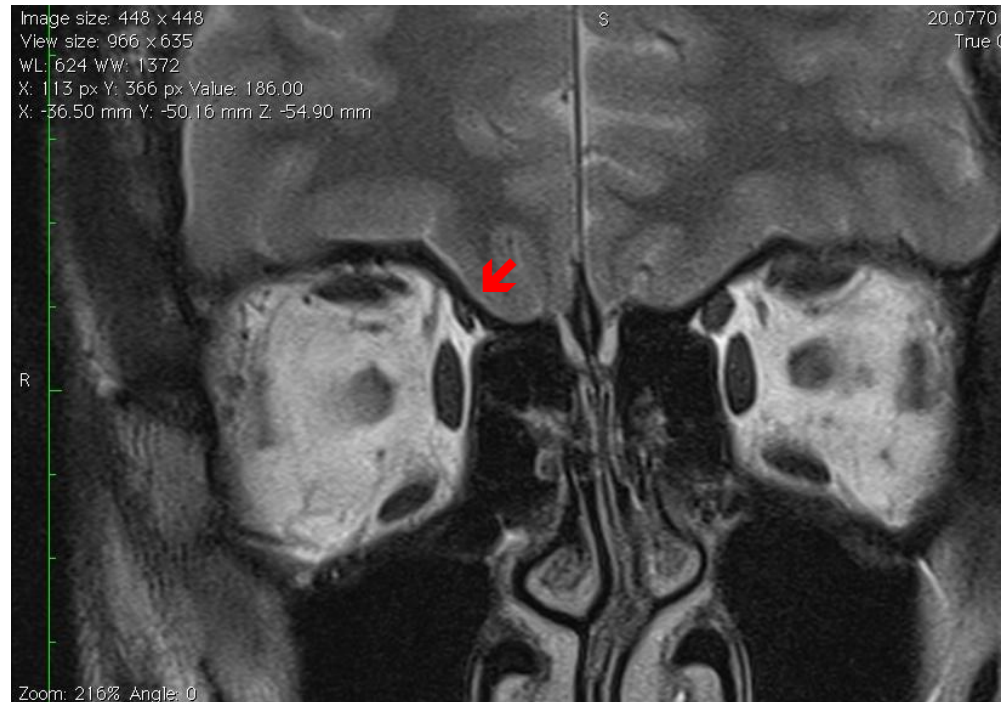
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May



It looks like a FNP but it isn't: what else could it be

- Simulating lesions

- Pulley disorders (Velez et al, 2000)
- Graves orbitopathy (Chen et al, 2008)
- Anatomical causes (Siepmann and Herzau, 2005)
- Tumour – an extremely rare cause
- Posteroplaced trochlea (Bagolini, 1982)
- Anomalous anatomy (Fink, 1962)
- Resolved 4NP leaving no radiological sequelae

Rumour: real FNP not caused by pathology

- Wrong because diagnostic criteria can be 50% off
- Wrong because no-one looks too hard, and keeps on looking

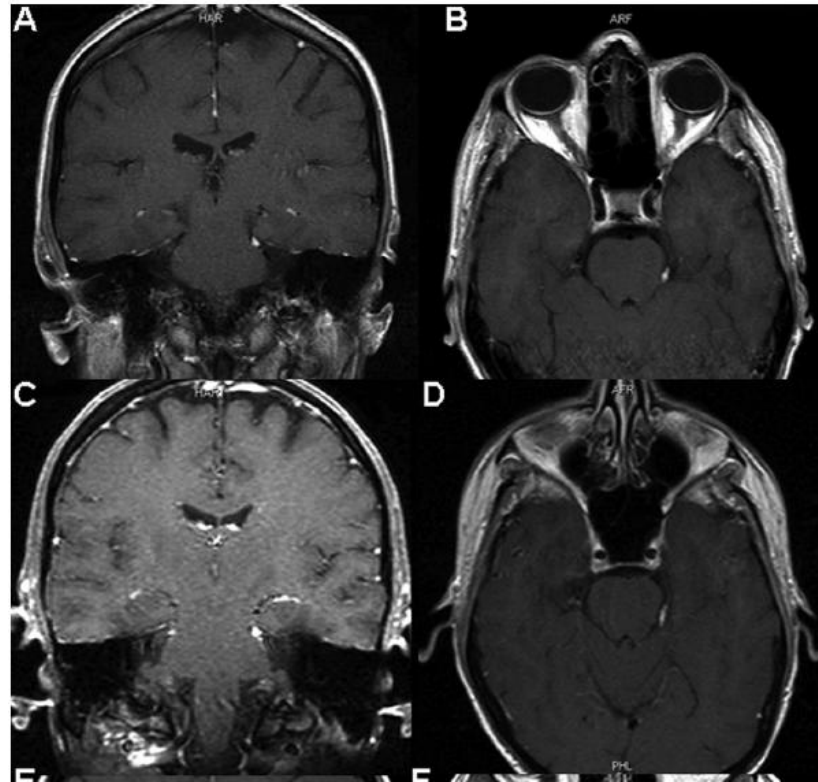
Ophthalmology 2009

Clinical Course and Prognosis of Trochlear Nerve Schwannomas

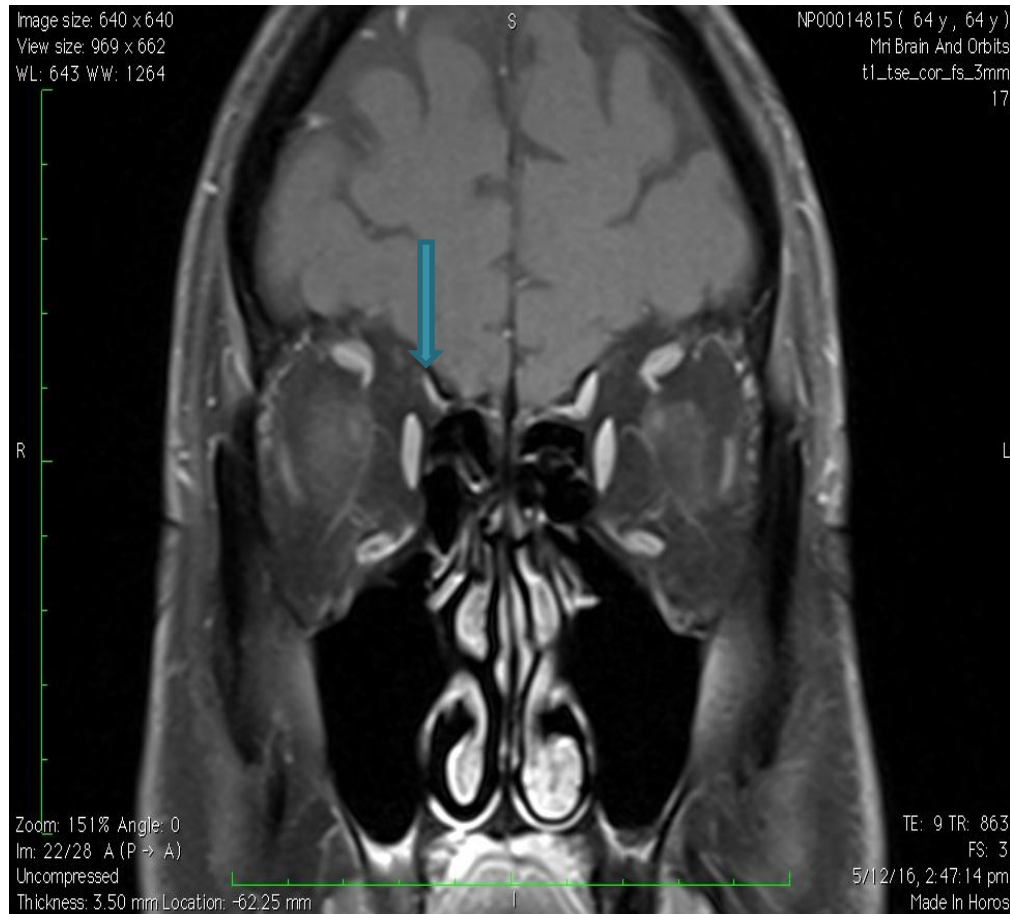
Valerie I. Elmalem, MD,¹ Brian R. Younge, MD,⁴ Valérie Biousse, MD,^{1,2} Jacqueline A. Leavitt, MD,⁴ Mark L. Moster, MD,⁵ Judith Warner, MD,⁶ Mark J. Kupersmith, MD,^{7,8,9} Klara Landau, MD,¹⁰ Michael C. Brodsky, MD,⁴ Larry P. Frohman, MD,¹¹ Eugene F. May, MD,¹² Robert L. Tomsak, MD, PhD,¹³ Nancy J. Newman, MD^{1,2,3}

Little tumours Easy to miss

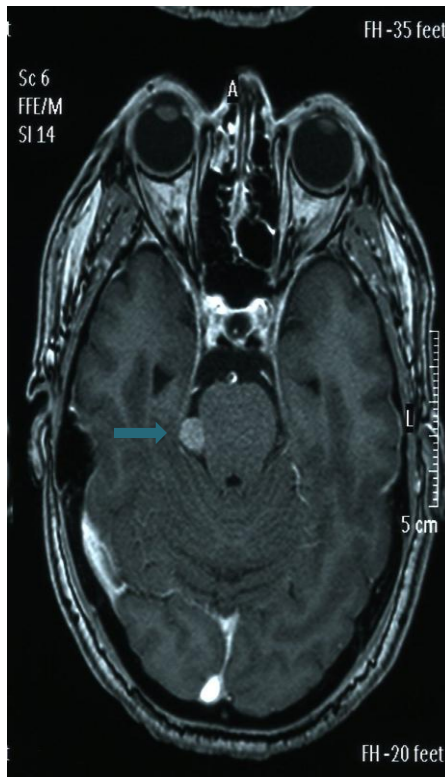
Elmalem et al · Clinical Course and Prognosis of Trochlear Nerve Schwannomas



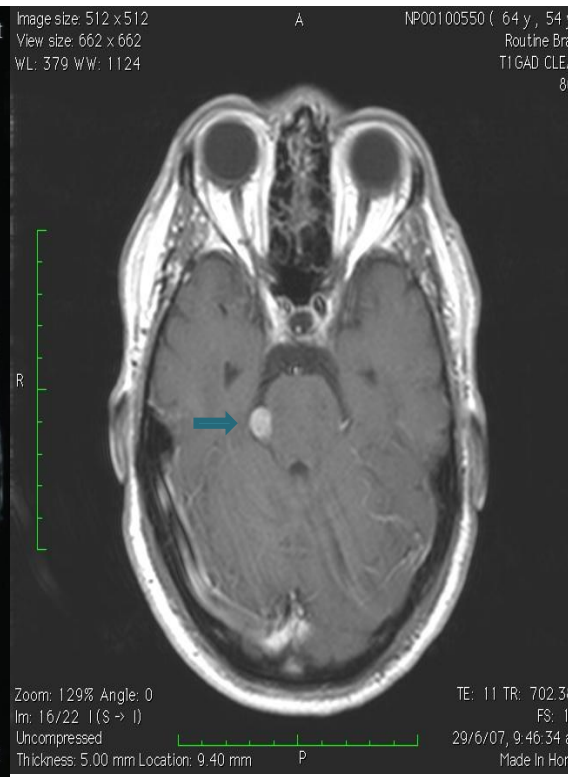
MRI showing R superior oblique atrophy not previously described (orbits not included on previous imaging)



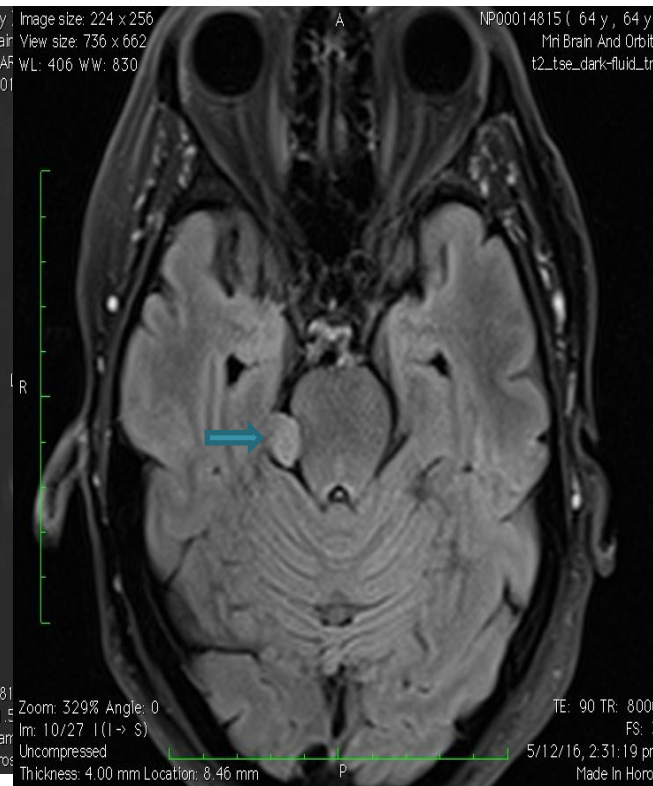
Minimal growth overtime (2mm over eleven years)



2006



2012



2016

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

Eye (2015) 29, 157–162
© 2015 Macmillan Publishers Limited |
www.nature.com/eye

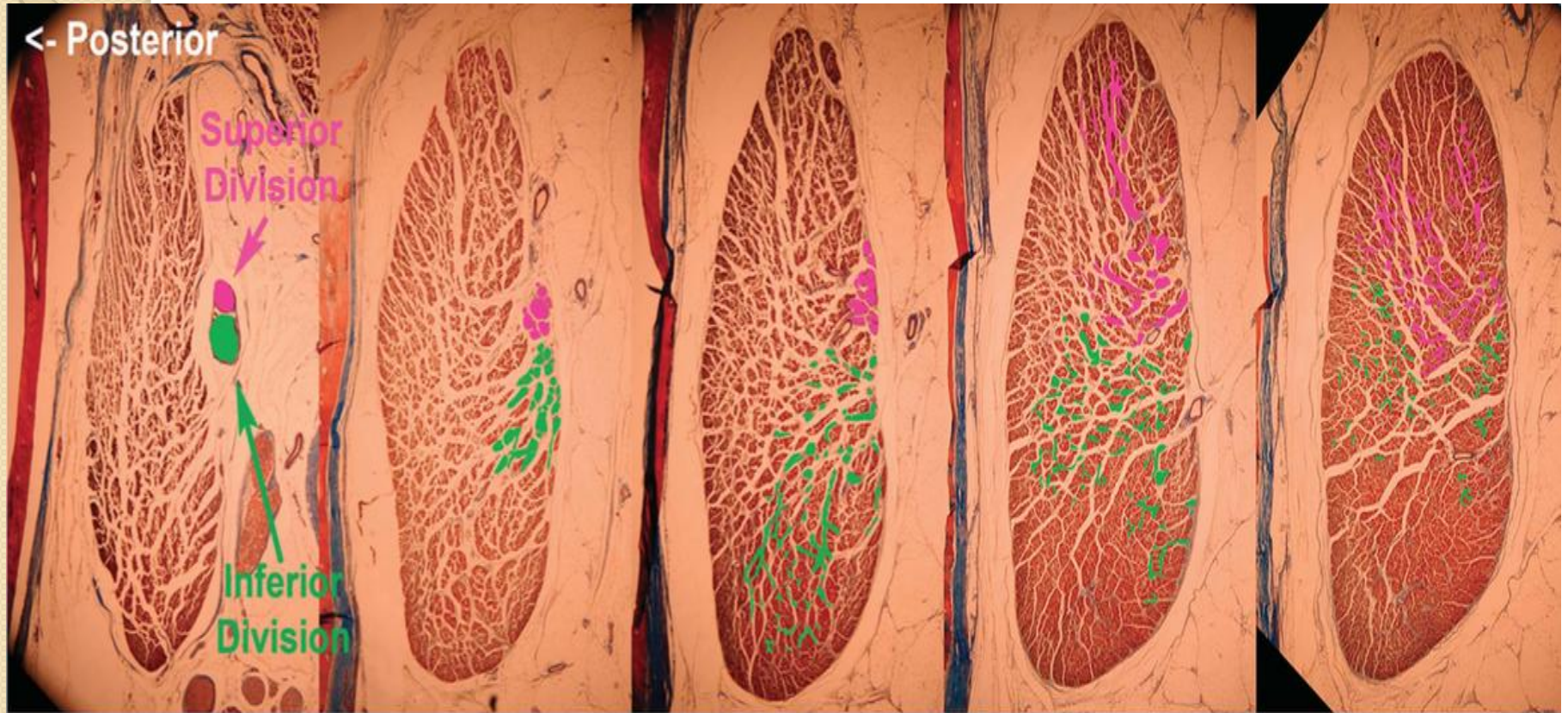
Compartmental-
ization of
extraocular muscle
function

JL Demer



* Compartmentalization not [yet?] demonstrated for superior rectus

“Humans, monkeys, and other mammals demonstrate separate, non-overlapping intramuscular nerve arborizations in the superior vs inferior compartments of the medial rectus (MR) and lateral rectus (LR)”



Demer, J. L. (2014). Compartmentalization of extraocular muscle function. *Eye (London, England)*, 29(August), 1–6.

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

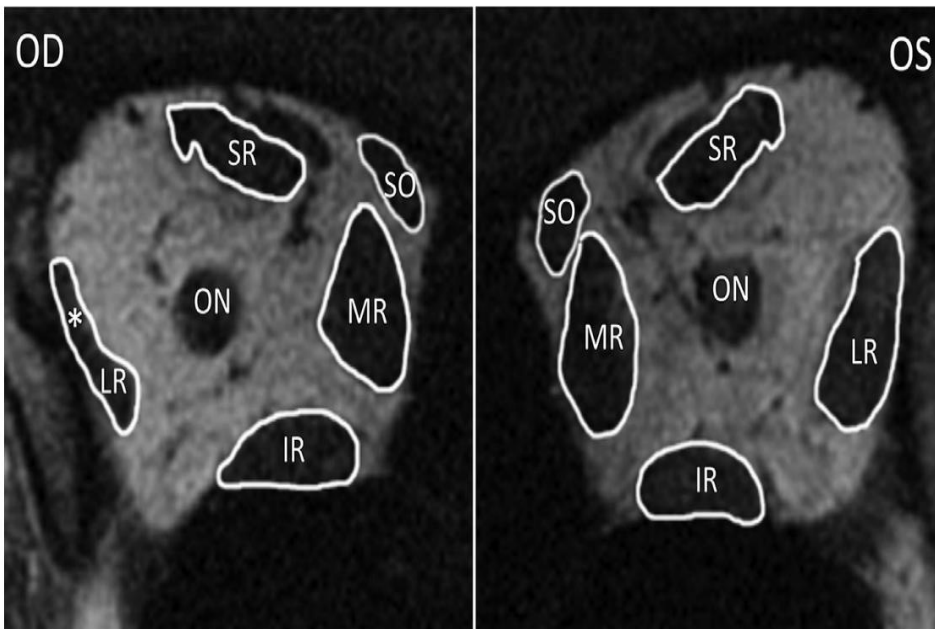
I. Esotropia & LR compartment hemi-atrophy

- 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 – watch this space**

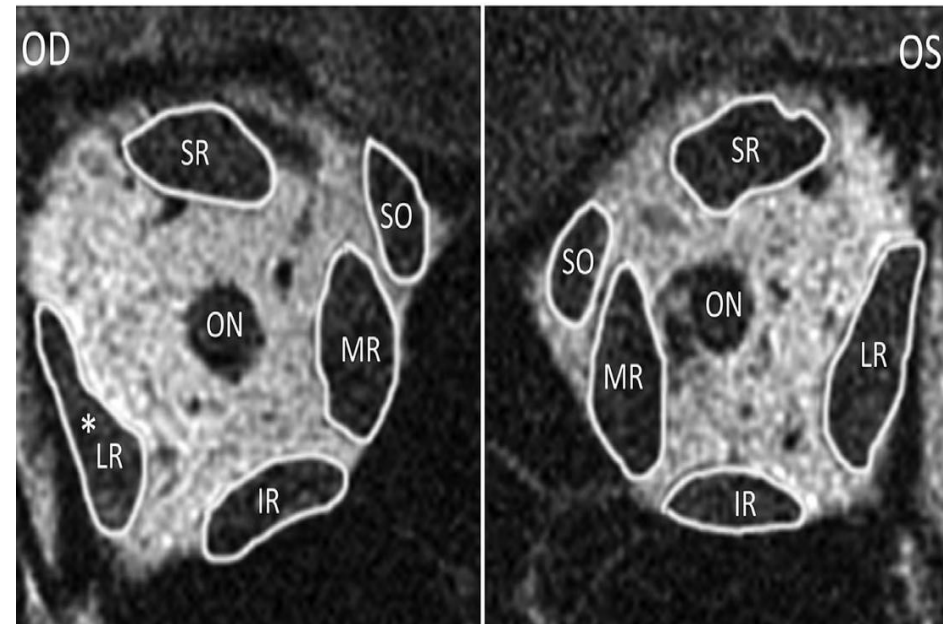
Lateral Rectus Superior Compartment Palsy

ROBERT A. CLARK AND JOSEPH L. DEMER

American Journal of Ophthalmology, 157(2) (2014).



Total RLR atrophy = palsy

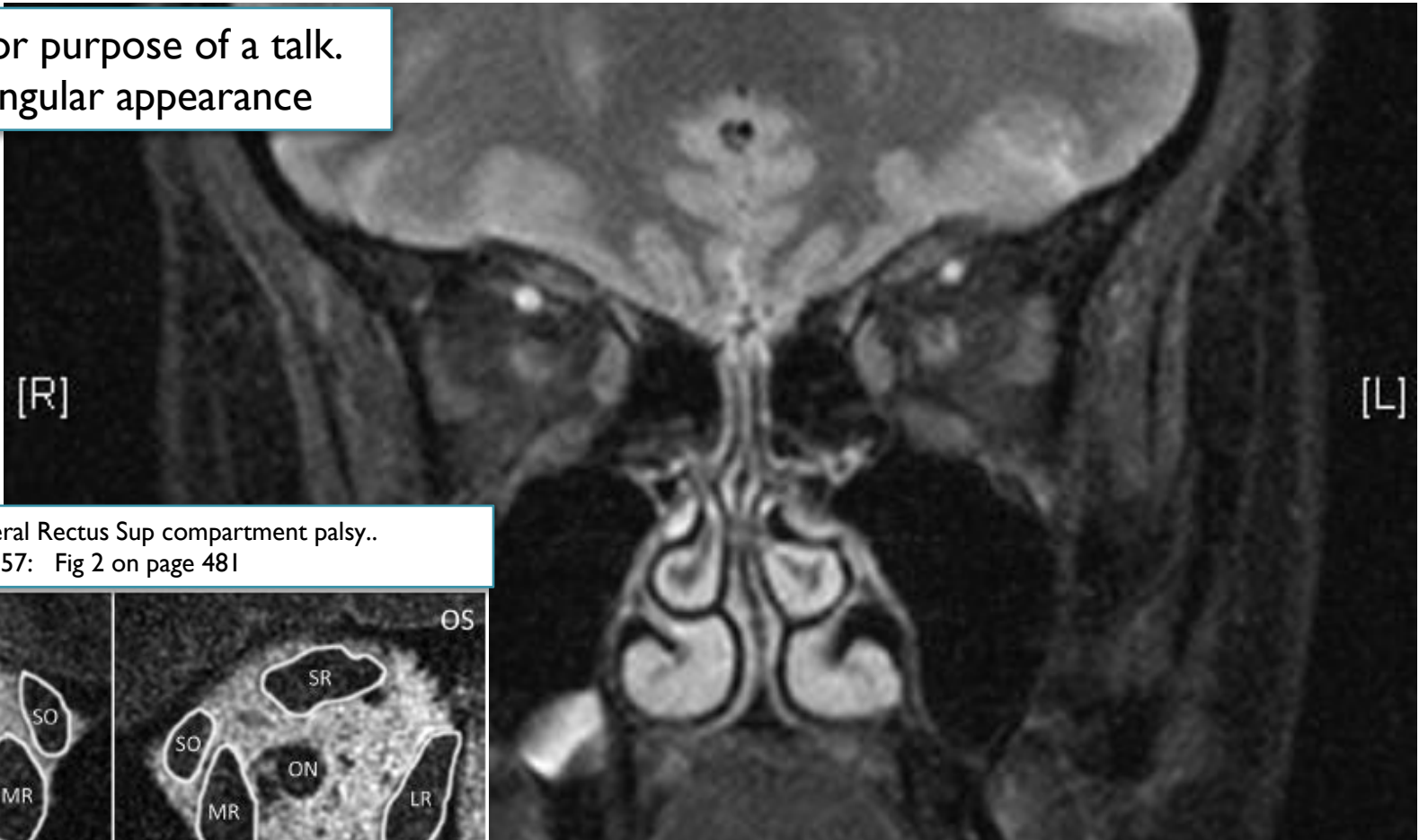


Sup compartment RLR palsy

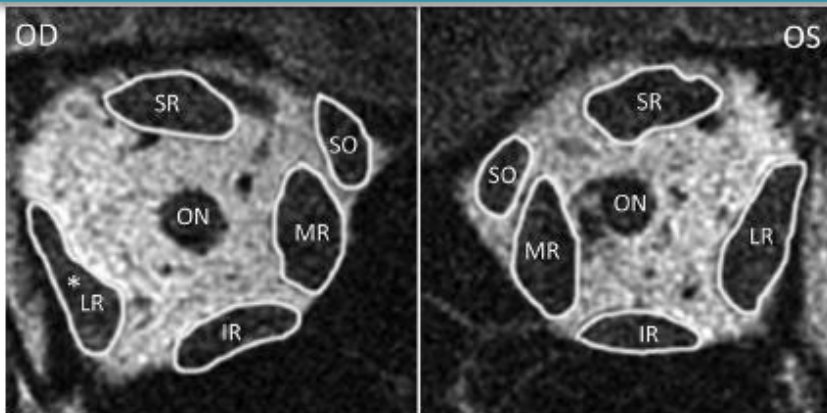
Clinical picture: more LR function than complete palsy. Treatment implications uncl

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

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



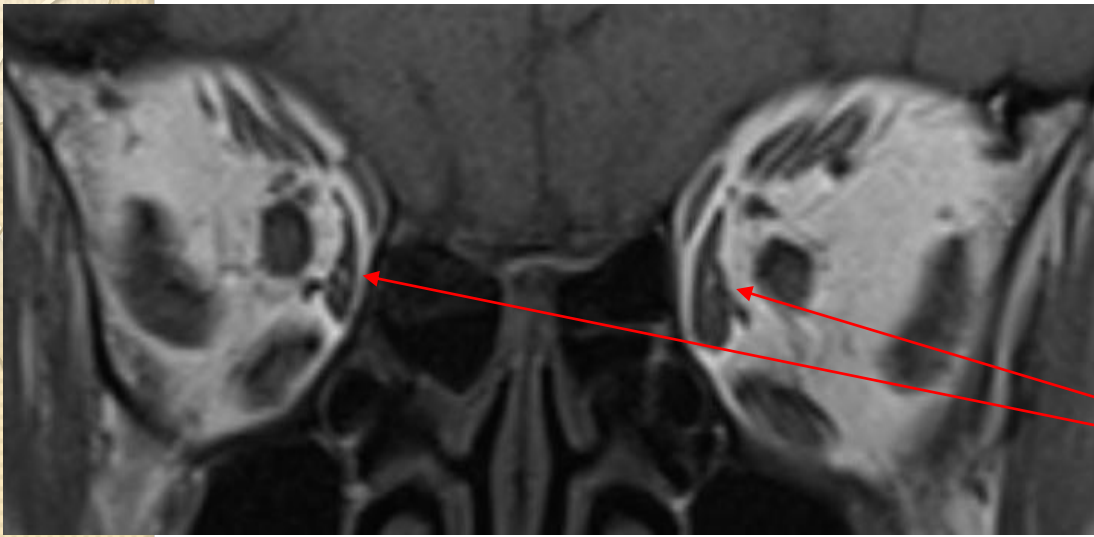
Clark & Demer Lateral Rectus Sup compartment palsy..
Amer J Ophth 2014; 157: Fig 2 on page 481



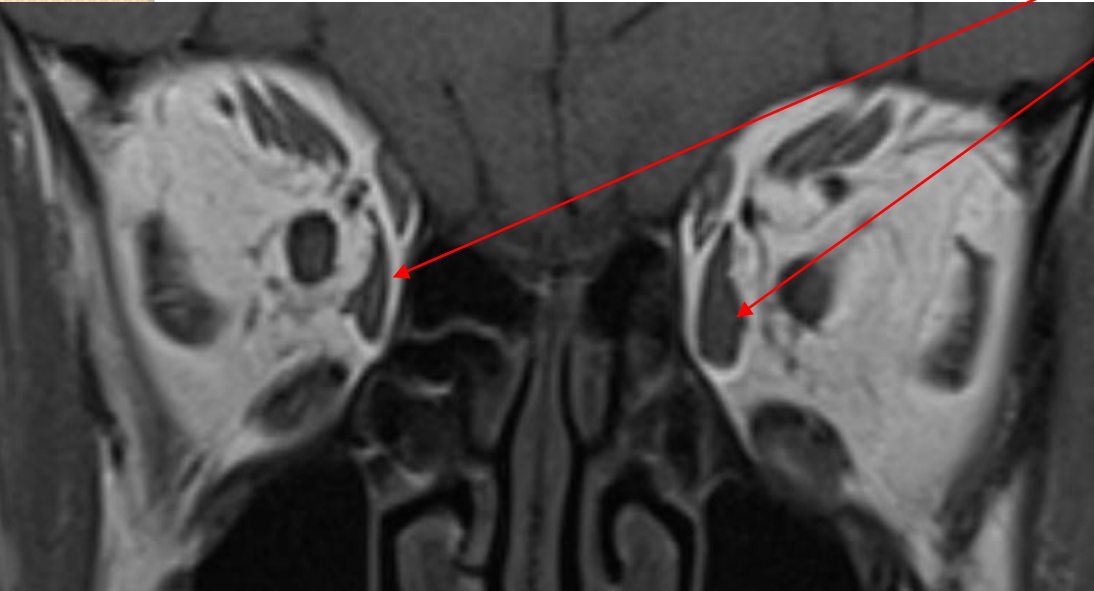
2. Exotropia & MR compartment hemi-atrophy

- 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 medial rectus compared to inferior



Asymmetry can be expected to produce a small vertical.

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

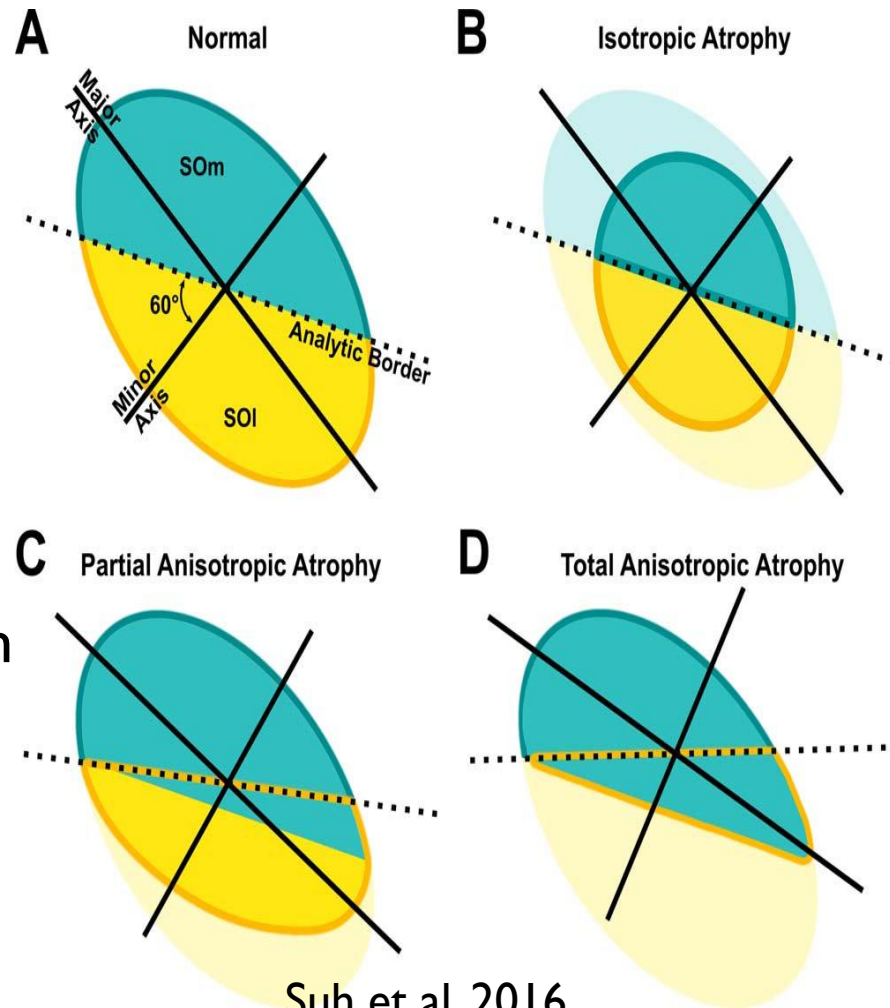
Surgery and Course.
MR plicate/resect OU. LR recess x1. Adjustables.
10 w followup: single vision, small phorias

- **3. SO: medial & lateral compartment innervation**

- Lateral compartment inserts behind the equator and is responsible for vertical globe movement

- Medial compartment inserts in front of the equator and is responsible for incyclotorsion

Muscle Compartments in Superior Oblique Palsy



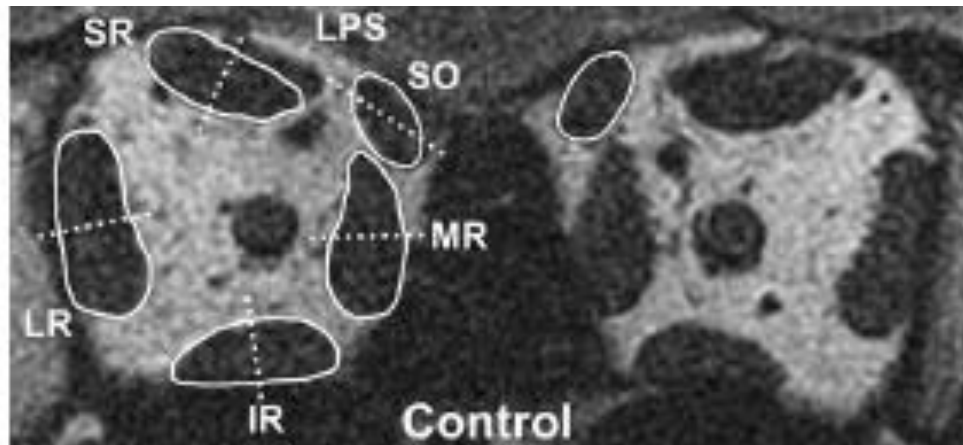
Suh et al, 2016

Medial compartment of SO controls *torsion*

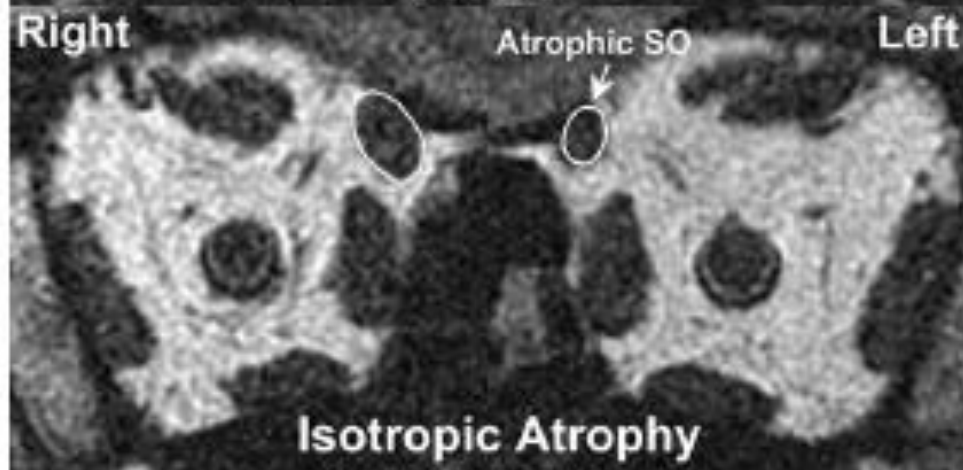
Lateral ...controls *vertical* movement

B,C: 20% develop *floppy tendons* requiring tendon tightening surgery

= A



= B




= C



SLOW HANDBALL OK : PRISM

- You prescribe a prism & 'fix' the pt but you don't have a 'proper' diagnosis
- 'Symptomatic' or 'broken down' phoria is hardly ever a 'proper' diagnosis
- 'Esodeviation of uncertain cause' [etc] is the honest diagnosis
- You must refer for a proper diagnosis – **fixing symptoms with prism is only a part of the treatment**
- *All these pts who you 'fix' can sometimes have a sinister cause / association*
- *There is a lot of selection bias in this slide*

- 
- Thank you for your attention
 - Now go read some more...you'll love it!