How and When Imaging Improves Strabismus Management.

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RVEEH Melbourne
WSPOS Symposium
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The radiology of strabismus is 25+ years old. Where are we heading?

- It has increased our understanding of the pathophysiology of many strabismus conditions
- We have introduced new treatment paradigms
- Some ‘abnormal’ radiological findings may have no clinical significance
- Alphabet patterns: confusing data
- We need to educate & enthuse ophthalmologists and radiologists

Today's talk

- Sup oblique palsy **
- Inf Obl overaction *
- ABduction deficits ***
- Alphabet patterns *
- Strabismus in high myopia ***

*Number of * indicate how useful radiology is*

When it changed how I defined & evaluated apparent sup obl palsy

UPDATE
on
STRABISMUS
and
PEDIATRIC
OPHTHALMOLOGY

Proceedings of the
Joint ISA and AAPO&S Meeting
Vancouver, Canada
June 19 to 23, 1994

Edited by
Gunnar Lennerstrand

CHAPTER 7

TRUE VERSUS MASQUERADING SUPERIOR OBLIQUE PALSIES: MUSCLE MECHANISMS REVEALED BY MAGNETIC RESONANCE IMAGING

Joseph L. Demer, Joel M. Miller, Edward Y. Koo, Arthur L. Rosenbaum, and J. Bronwyn Bateman

I. Introduction

The superior oblique (SO) muscle cannot be clinically visualized and has mechanical actions involving complex cooperation with ipsilateral and contralateral agonists and antagonists. Thus, SO palsy is currently diagnosed indirectly based on clinical observations of ocular versions, as well as on measurements of binocular misalignment. The present study was conducted to validate diagnostic criteria for SO palsy against magnetic resonance imaging (MRI), an independent measure of SO contractile function.

II. Methods

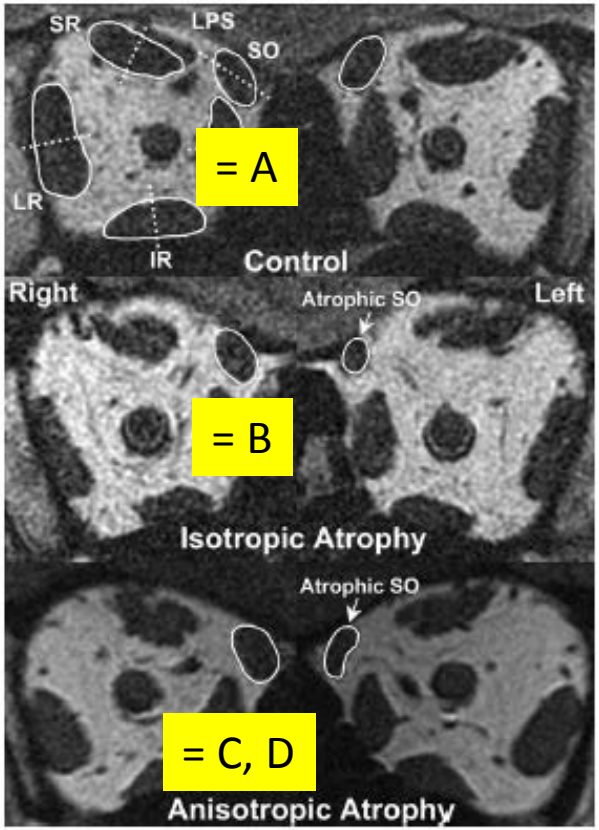
High-resolution, surface coil MRI was performed on 16 normal orbits of 12 volunteers, and on 34 orbits of 17 patients with hypertropia in whom the diagnosis of SO palsy (2 bilateral, 15 unilateral) had been made by a pediatric ophthalmologist. Diagnostic criteria included: hypertropia increasing in contralateral gaze and with ipsilateral head tilt; ipsilateral SO underaction on version testing; and cyclohetropia

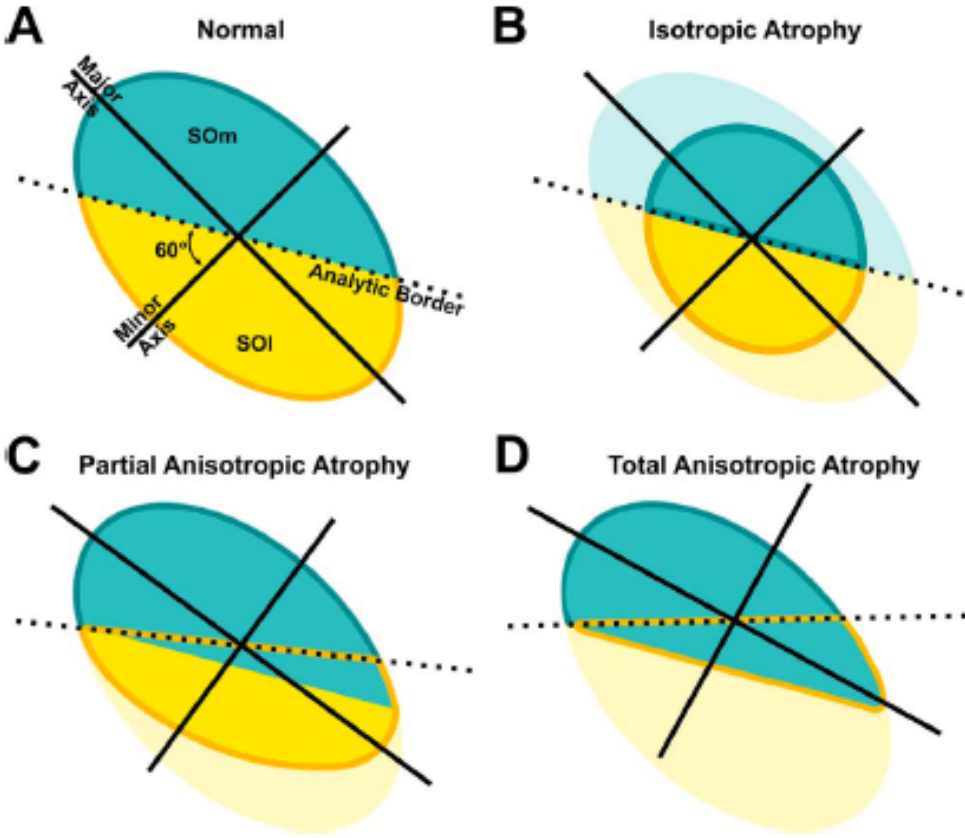
On Downgaze, RSO is much thicker than on Upgaze

Different types of Superior Oblique Palsy/Paresis

UCLA 2016

IOVS | October 2016 | Vol. 57 | No. 13 | 5536





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

Real /apparent / pseudo SOP

SOP = Superior Oblique Palsy

- ’SOP’ often used as a synonym for ‘a condition that resembles SOP’
- When ‘SOP’ is diagnosed by strabismus Drs, it is wrong ~50% of the time [Demer, Simonsz, Herzau]
- The most accessible technique to reliably diagnose SOP is to demonstrate atrophy on coronal scans

Q: Is it important to segregate 'real' from 'pseudo' SOP?

- ***..is it important to know the most accurate diagnosis …. most of the time we are going to do Inf Obl weakening irrespective of the precise cause?***
- Some pseudo- SOP causes need different Sx eg LR sup myopexy
- **True SOP** has other implications:

- some are rarely due to tumour
- natural history is probably to get worse

Imaging in Congenital Superior Oblique Palsy: Is it important?

Lionel Kowal, Melbourne, Australia Logan Mitchell, Dunedin, New Zealand

INTRODUCTION: There are many diagnostic criteria for superior oblique palsy (SOP). Demer and others [1,2] have suggested that radiological atrophy is a necessary condition for a definitive diagnosis of SOP [Fig 1].

Fig 1. RSO atrophy

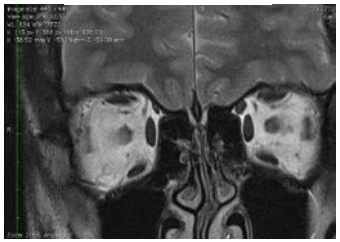


Fig 2. Sato's results

TABLE 2. Relationship Between Tendon Looseness and Muscle Atrophy

	Atrophy of SO Muscle		
	Normal	Small	Total
Tendon looseness			
Normal	8	6	14
Loose	0	9	9
Total	8	15	

SO = superior oblique.

Many believe that if the Superior Oblique (SO) in cases of SOP has a clearly floppy tendon then superior oblique tightening (SOT) is the preferred procedure. Our experience is that floppy SO cannot be predicted from pre-operative characteristics [Fig 3, Table 1]
SOT is more difficult and has greater morbidity than the inferior oblique surgeries that are more commonly performed for SOP. It would be good to predict which patients are likely to require SOT so that those who are not expert in performing SOT can avoid these cases, and so ap
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SO
this was seen in only four patients.

20% of pts with atrophic Sup Obl on scans have unequivocally floppy tendons @ surgery, probably requiring Sup Obl tightening.
0% floppy tendons if Sup Obl normal on MRI

Fig 3. Floppy R SO, not suspected on rotations

Table 1: 3 of the 4 cases of floppy superior oblique

005 M		RH	RSO plicate
2			24
8		14	20
8			20
015 D		LH	LSO plicate
13			0
20		6	0
18			0
022 W		LH	LSO+, ATLIO
45			16
45		45	3
45			20

- 3 of the SOT operations were combined with ipsilateral IO weakening surgery. One patient required subsequent SO scar excision and ipsilateral IO weakening after SOT alone produced an inadequate result.
- 14 patients underwent primary ipsilateral inferior oblique weakening, none of whom required further cyclovertical strabismus surgery.

further cyclovertical

measurements, with
Δ (range 0-45) to 3.6
34).
s with radiologically

s of SOP are probably

loppy SO tendon on
ato results of 9/15

c.f. 0/20 in our series
(p=0.02)

CONCLUSION: Imaging of the SO tendon is recommended as part of the evaluation of patients with (probable) SOP to segregate those with atrophy. 20% of the group with radiological atrophy are likely to have floppy tendons and possibly require SOT, a more difficult and higher morbidity procedure than the more commonly performed inferior oblique surgeries for SOP.

References:

1. Demer et al. *MRI of the functional anatomy of the superior oblique muscle.* IOVS 1995 & 1994 AAPOS / ISA joint meeting proceedings.
2. Siepmann, K et al. *Is congenital superior oblique strabismus a paretic disorder?* Klin Monatsbl Augenheilkd. 2005; 222(5): 413-8.
3. Sato M. *Magnetic resonance imaging and tendon anomaly associated with congenital superior oblique palsy.* Am J Ophthalmol. 1999

The authors have no financial interest

Inferior Oblique Overaction

IO OA may be due to Sup Obl Palsy, but there are many other causes & the best treatment may not always be Inf Obl weakening

Table. Conditions Causing Overelevation in Adduction Not Related to the Inferior Oblique in the Adducting Eye

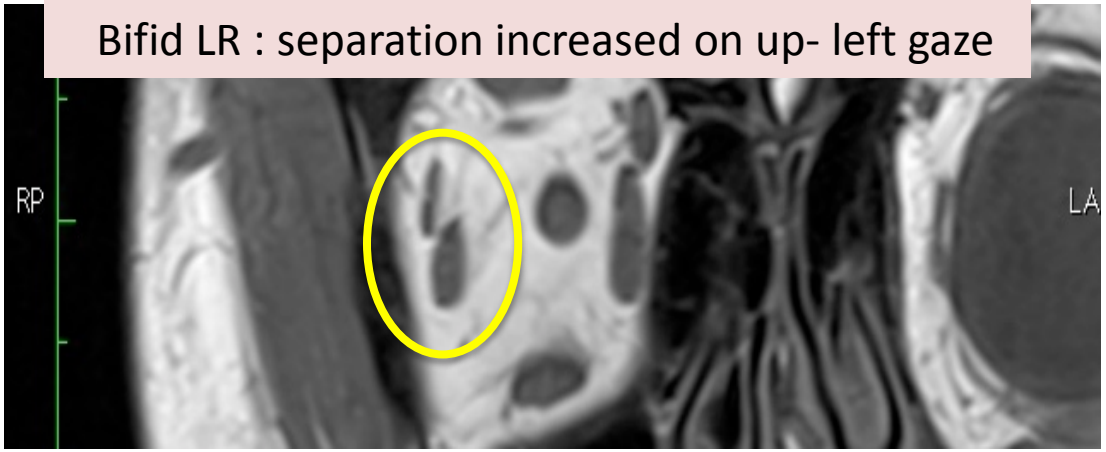
Clinical Condition	Comment
Duane syndrome[1,2]	Elevation in adduction caused by cocontraction of lateral rectus
Dissociated vertical divergence[3]	Dissociation by the nose can bring out a manifest dissociated vertical divergence in adduction
Craniofacial syndromes[4-6]	Excyclorotation of all the rectus muscles can cause elevation in adduction
Antielevation syndrome after IO anterior transposition[7]	The antielevating effect of the IO anterior transposition restricts elevation in abduction, causing fixation duress to the contralateral eye and resulting in overelevation
Mechanical restriction of the inferior rectus[3]	Restriction of elevation in abduction causes fixation duress to the contralateral eye, resulting in overelevation
Pseudo IO overaction with Y or V pattern[8]	Cocontraction of the lateral rectus on attempted upgaze mimics IO overaction
Pulley heterotopia[9,10]	Abnormal position of one or more of the rectus muscle pulleys can cause overelevation in adduction

& Graves'

Kushner BJ Arch Ophthal 2006

#1: A IO OA 2ary to bifid LR

Case 1



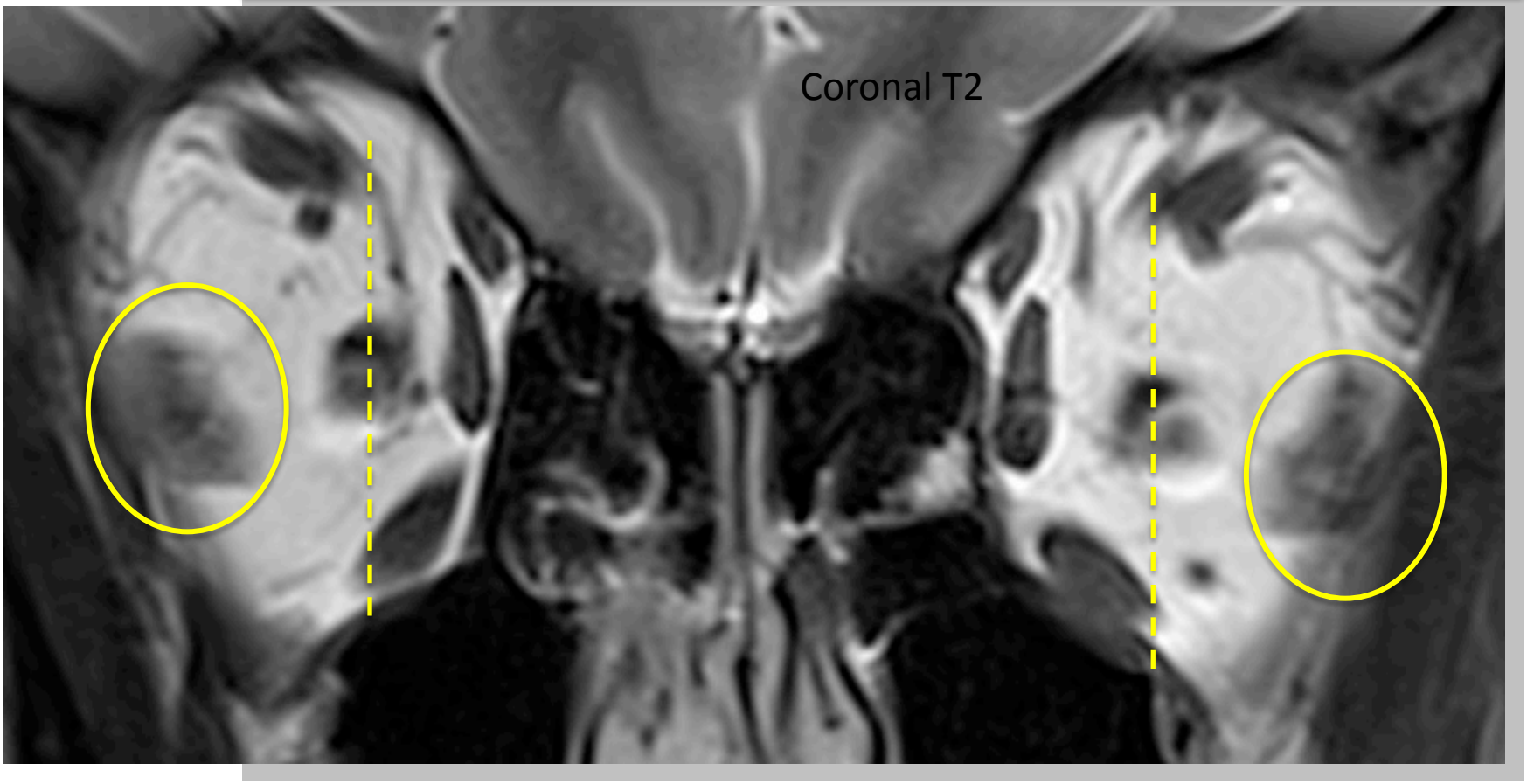
Case 2

Acq Inferior Oblique Overaction

21 yo PhD student:
Recent vertical diplopia with prolonged reading

Acquired Inferior Oblique Overaction

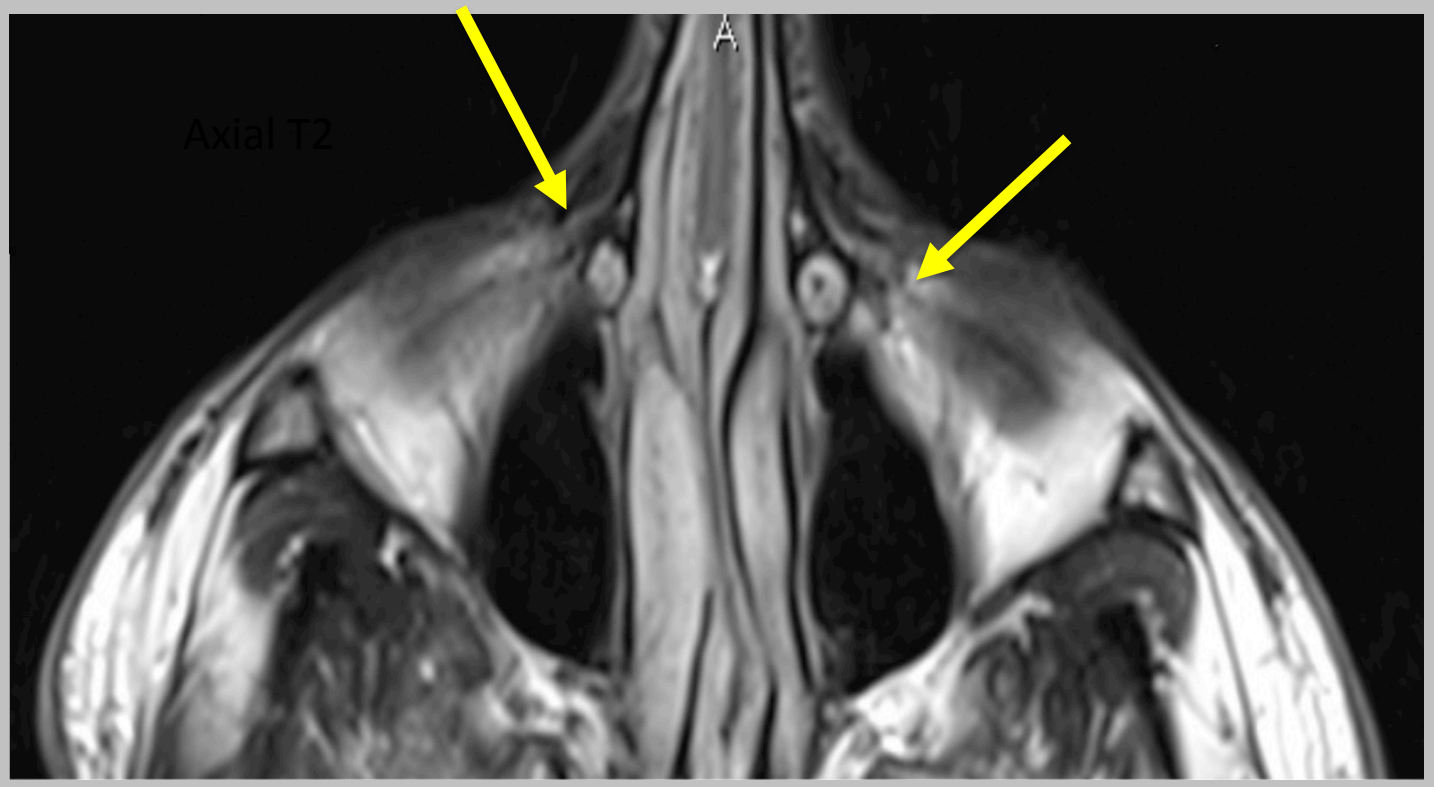
1. Extorsion of both orbital contents.
2. SR more lateral than IR which is normally only slightly nasal
3. LR also appear slightly sagging / heterotopic



Acquired Inferior Oblique Overaction

Case 2

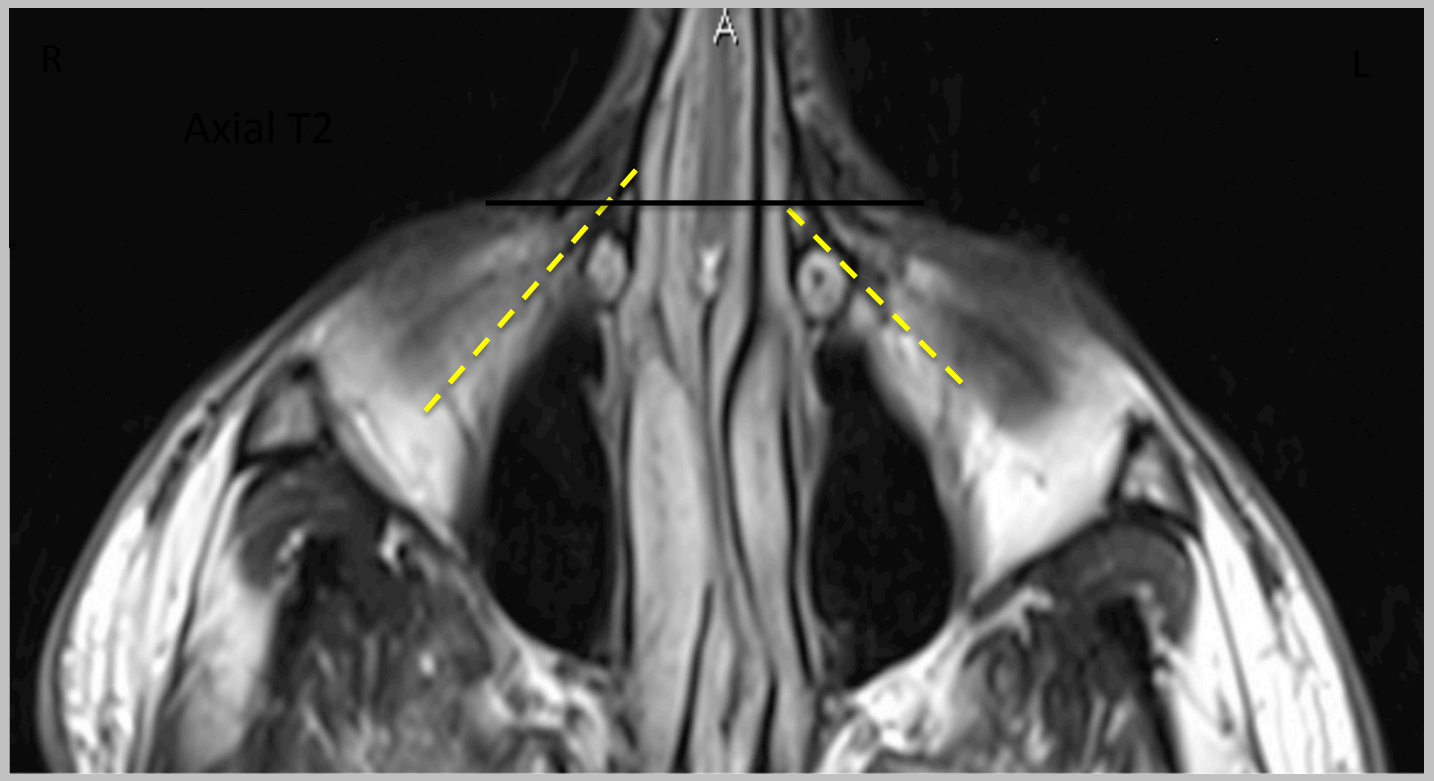
Origins of inferior obliques not symmetric



Acquired Inferior Oblique Overaction

Case 2

Origins of inferior obliques not symmetric



Acquired Inferior Oblique Overaction
Fink 1954 !

Significant asymmetry occurs in 20% of cadaveric cases

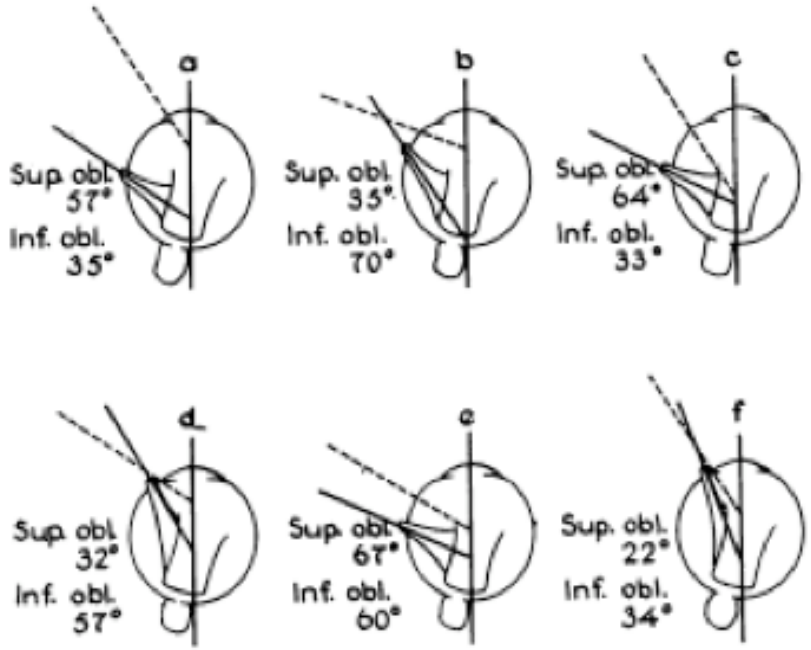


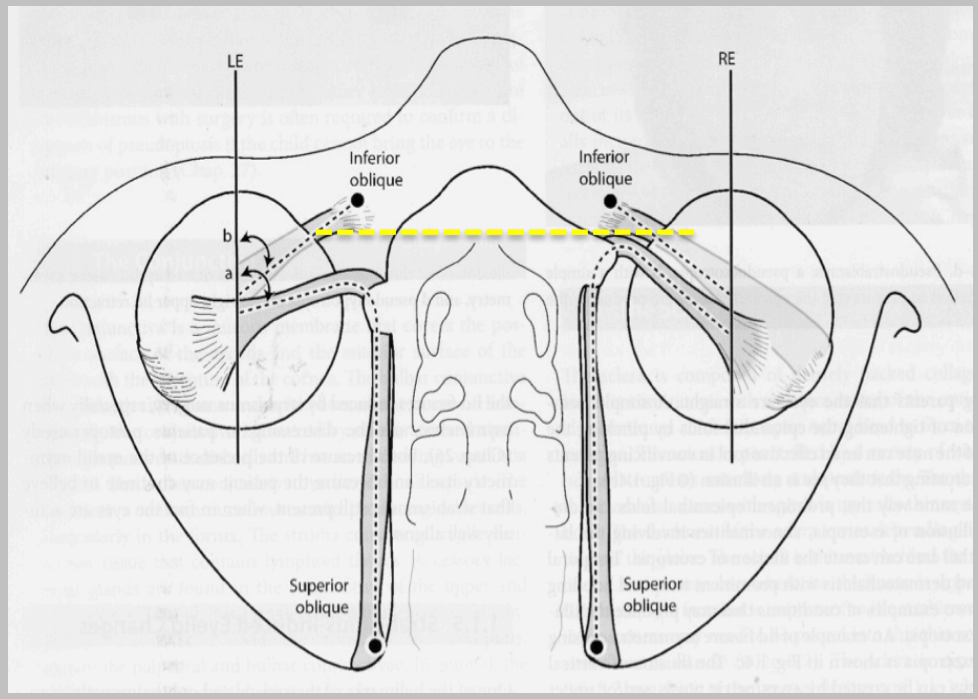
FIGURE 5. DRAWINGS OF SPECIMENS IN WHICH THERE ARE PRONOUNCED VARIATIONS OF THE OBLIQUE MUSCLE PLANES OF ACTION Continuous line indicates the superior oblique plane of action. Broken line indicates the inferior oblique plane of action.

Discussion

Fink, W. “The role of developmental abnormalities in vertical muscle defects”. Trans Am Ophth Soc 1954

Apparent oblique dysfunction [inc cases that look like SOP] is often due to orbital abnormalities

"Plagiocephaly causing superior oblique deficiency and ocular torticollis. A new clinical entity." Bagolini et al. Arch Ophth 1982



Key:

'Possible SOP' needs imaging if you need an accurate diagnosis or if you plan to operate

ABduction deficits

- 6th n paresis
- 6th n palsy
- Sup compartment LR palsy
- Acquired LR sag
- Occult Graves'
- Post surgical [over-recessed, stretched scar, lost muscle]
- In moderate & high myopes

…can all look the same, but have different causes, natural history and surgical treatments, and need imaging

1. Is it a paresis or is it a palsy?

Paresis: horizontal surgery

Palsy: will need transposition

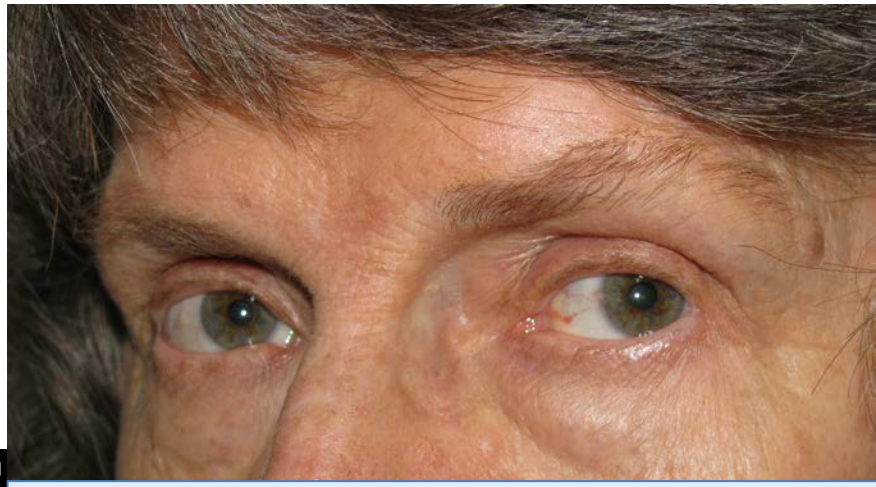
<u>Diplopia 10+ yrs</u>

R 6th. No definite cause

R gaze 45Δ ET,

Primary 30Δ [R fixation 45Δ] .

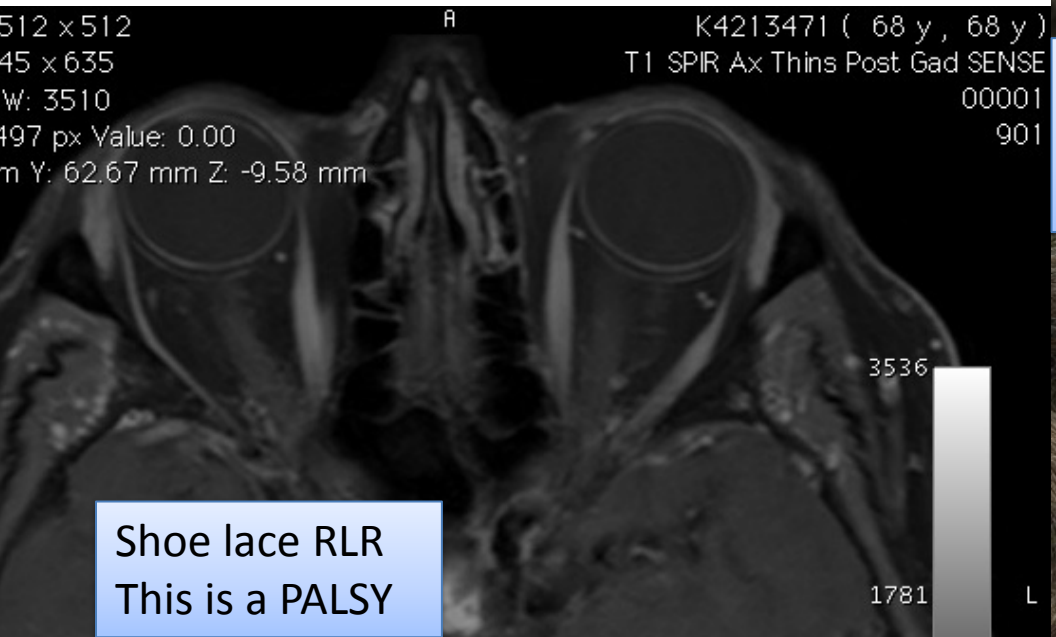
L gaze 10Δ



R Abduction deficit (only) 2.5mm

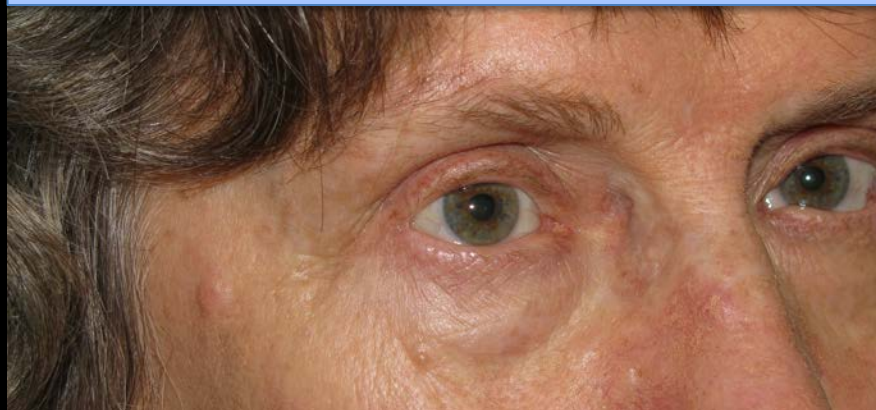
Very slow RLR saccade

Force Generation test: Dead RLR



Shoe lace RLR

This is a PALSY

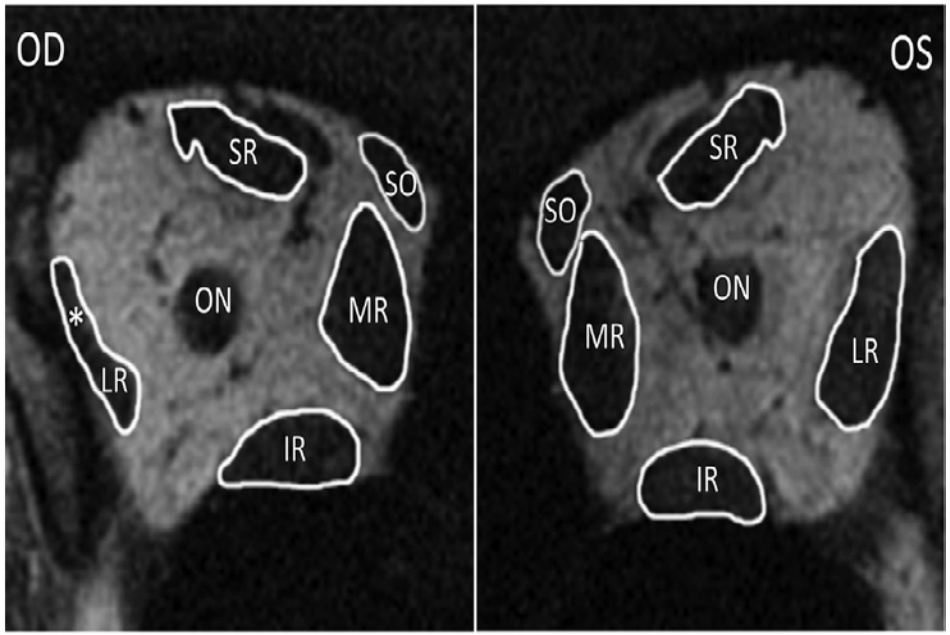


2

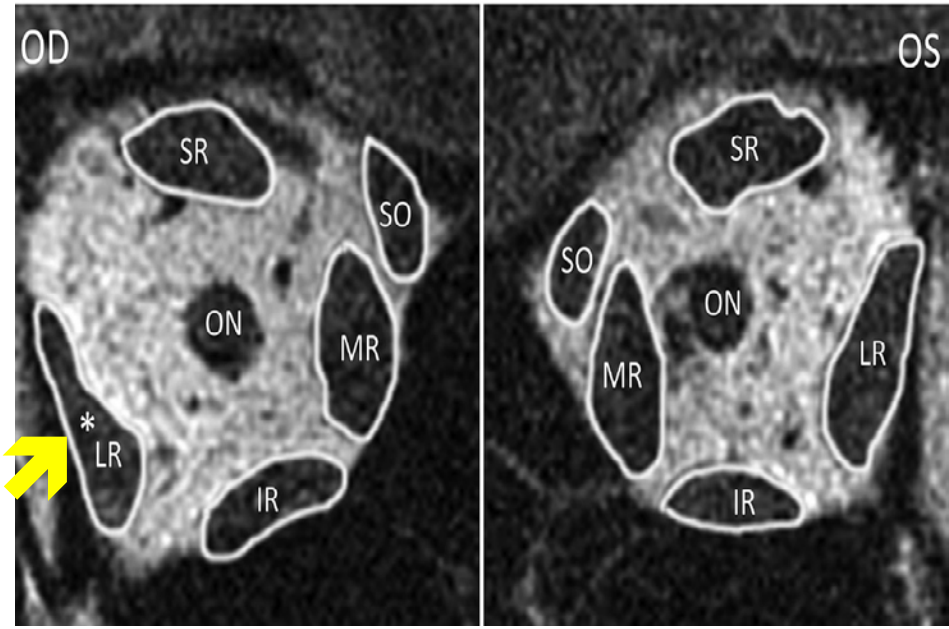
Lateral Rectus Superior Compartment Palsy

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)



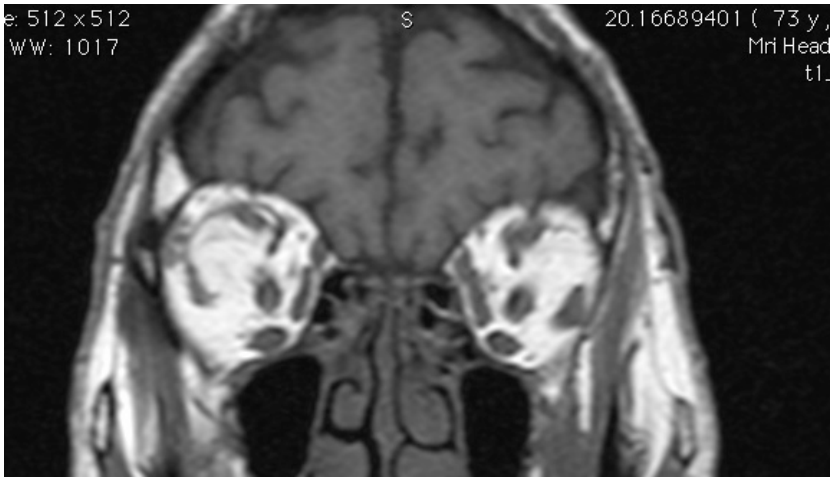
Total RLR atrophy = palsy

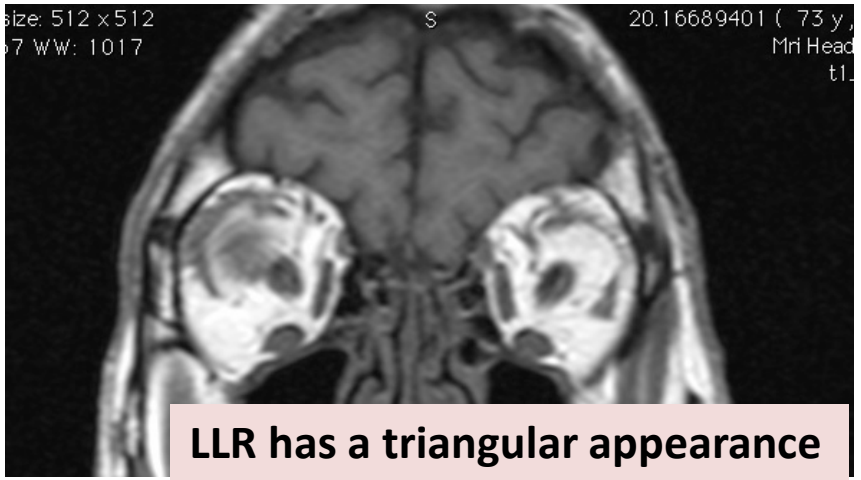


Sup compartment RLR palsy

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

2. Sup compartment LLR paresis 63019





LLR has a triangular appearance

71 yrs old

Diplopia onset >1 yr ago.

ET 16Δ, ET’ 4Δ

LLR UA

Spectrum of Lateral Rectus Heterotopy

- Abnormally positioned or directed lateral recti (demonstrated radiologically or intra-operatively) can cause or contribute to strabismus, including alphabet pattern strabismus.[1]

Congenital LR Heterotopy	Acquired LR Heterotopy

• **Isolated LR Heterotopy:**
  • Normal orbits
  • No other extraocular muscle heterotopy
  • Not syndromic/No Craniosynostosis
	• **Heavy Eye syndrome:**
	  • High Myopia

• **LR heterotopy with rotated orbits**
  • +/- Heterotopy of other Extraocular muscles
  • Not Syndromic/No Craniosynostosis
	• **Sagging Eye Syndrome**
	  • **Disruption of LR-SR Band**
	[Heavy Eye and Sagging Eye can both be seen in myopia]

• **LR Heterotopy as a feature of Syndromic features/Craniosynostosis/Plagiocephaly** (*or from it's corrective surgery eg: Frontal Orbital Advancement*)	• Iatrogenic with augmented inferior or superior transposition of lateral rectus during strabismus surgery

[1] Demer JL, Clark RA, Kono R, Wright W, Velez F, Rosenbaum AL. A 12-year, prospective study of extraocular muscle imaging in complex strabismus. J AAPOS 2002; 6(6): 337–347.

[2] Tan KP, Sargent MA, Poskitt KJ, Lyons CJ. Ocular overelevation in adduction in craniosynostosis: is it the result of excyclorotation of the extraocular muscles? J AAPOS 2005; 9(6): 550–557.

[3] Chaudhuri Z, Demer JL. Sagging eye syndrome: Connective tissue involution as a cause of horizontal and vertical strabismus in older patients. JAMA Ophthalmology 2013;131(5): 619–625.

[4] Tan RJ, Demer, JL. Heavy eye syndrome versus sagging eye syndrome in high myopia. J AAPOS *2015; 19*(6): 500–506

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

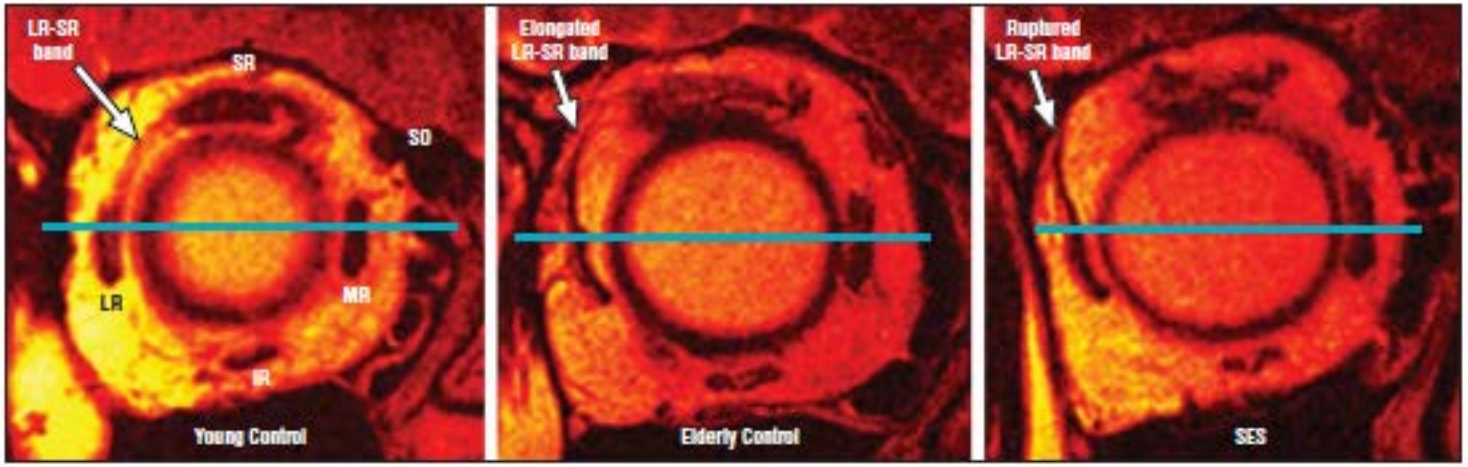


Figure 2. Fast spin-echo T2-weighted sequence quasi-coronal plane magnetic resonance imaging. Left, Younger control participant showing lateral rectus (LR)–superior rectus (SR) band. Note the normal morphology of LR muscle with respect to a horizontal reference line drawn through the globe center. Middle,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

Case #3. 82 y o
Intermittent Horizontal diplopia,

…..mainly on left gaze, started 8 y ago

- Horizontal Deviation:

0

0 6ET 12ET

6ET

- Prescribed glasses:

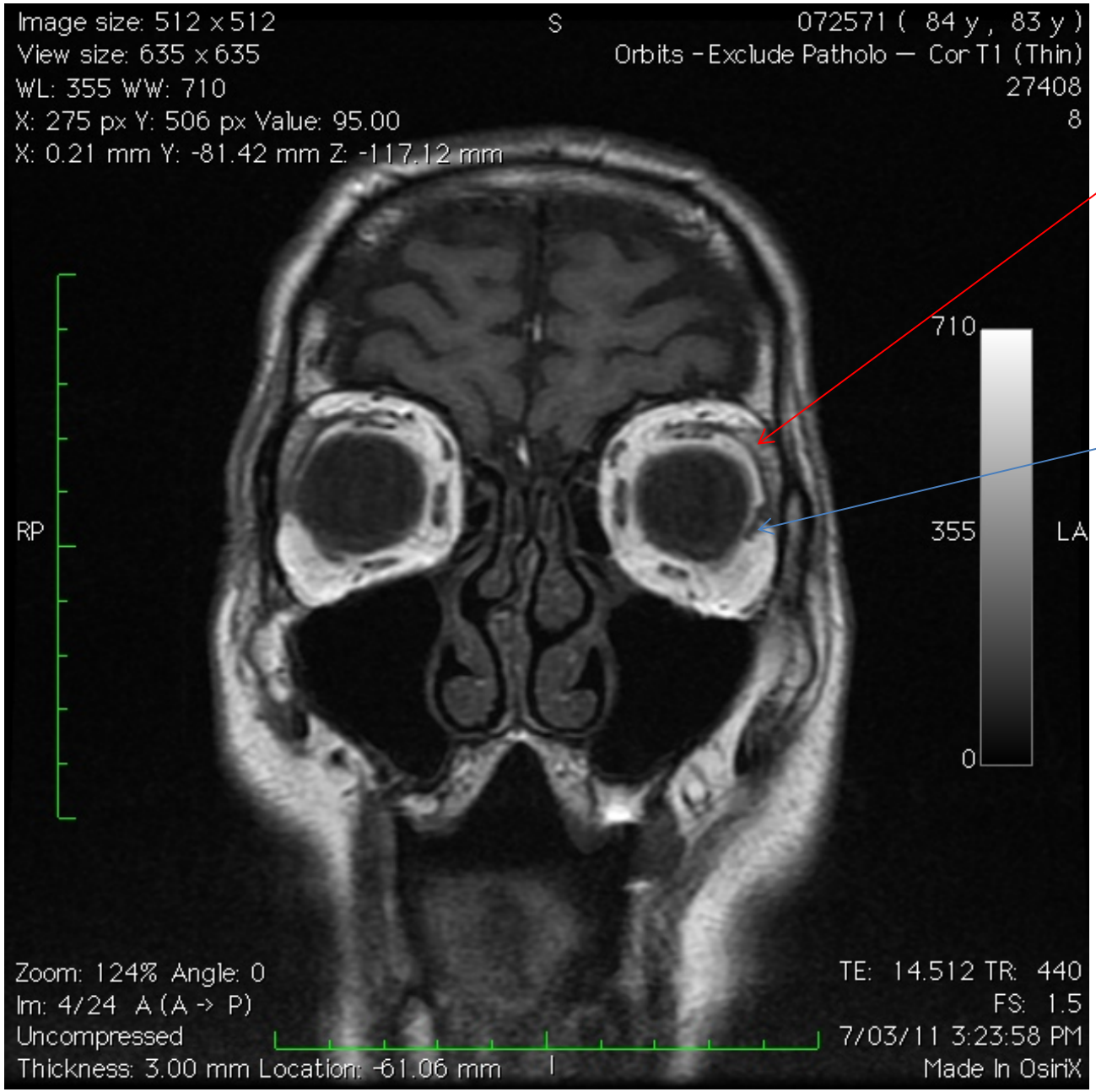
8Δ BO, 2Δ BU LE (single vision).

BE elevate incompletely.

R gaze – L elevation reduced

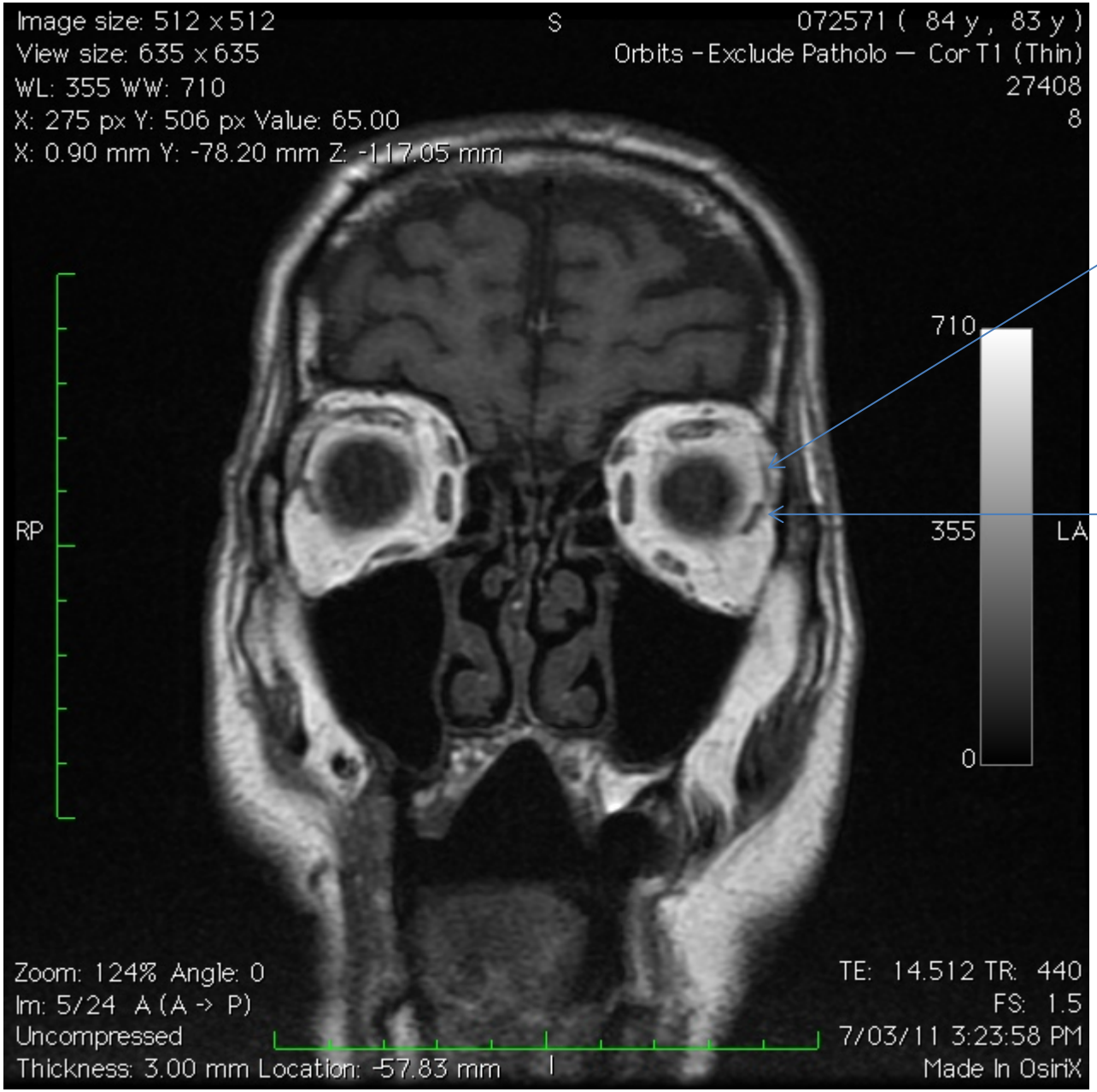
L gaze – L depression reduced.

Tiny aBduction deficit
No LT gaze paresis



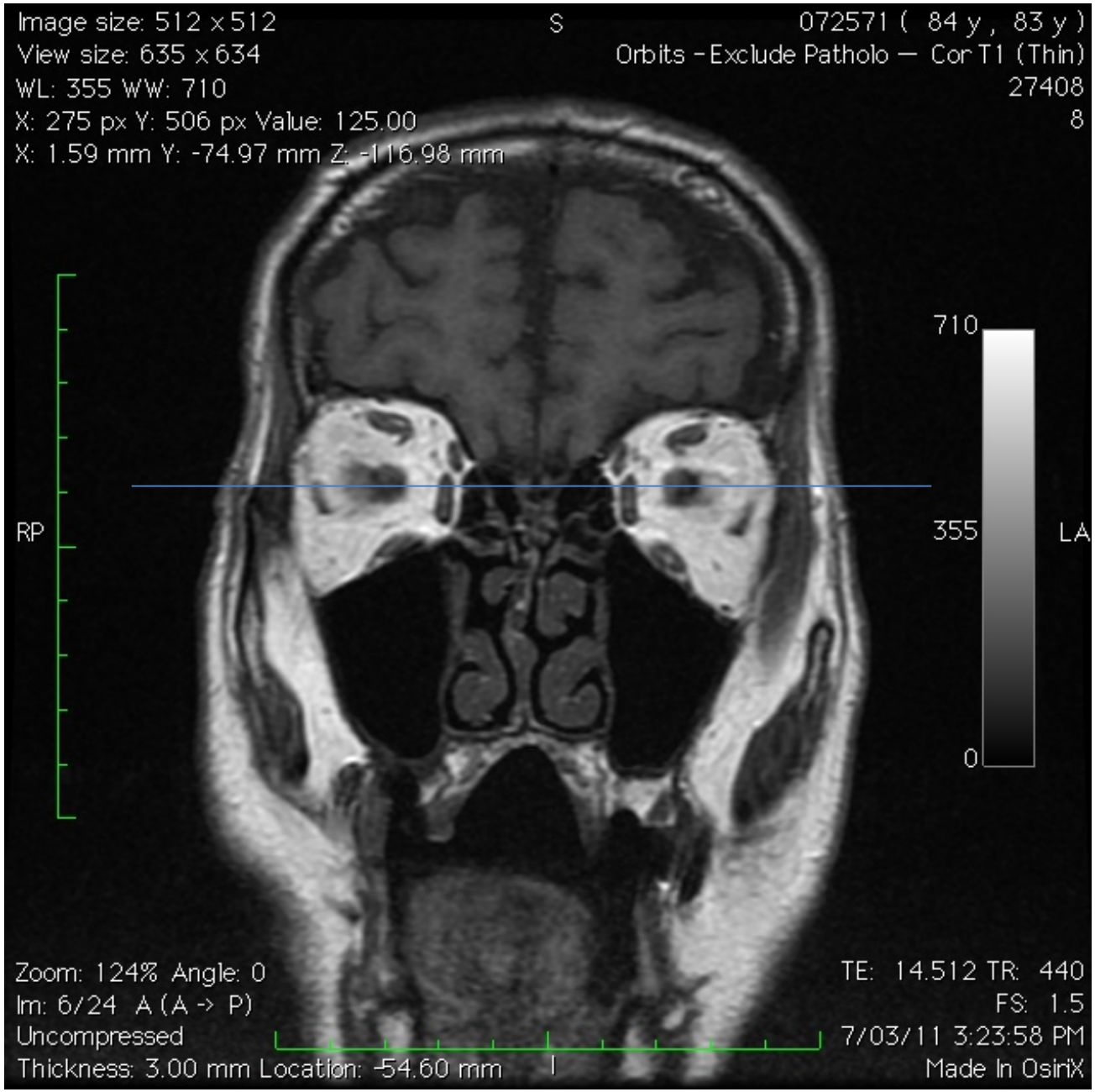
Inter-muscular septum (LLR-LSR Band)

Lower & tilted position of LLR



Atrophy of Inter-muscular septum (LLR-LSR Band)

Lower & tilted position of LLR



Lower & tilted position of LLR

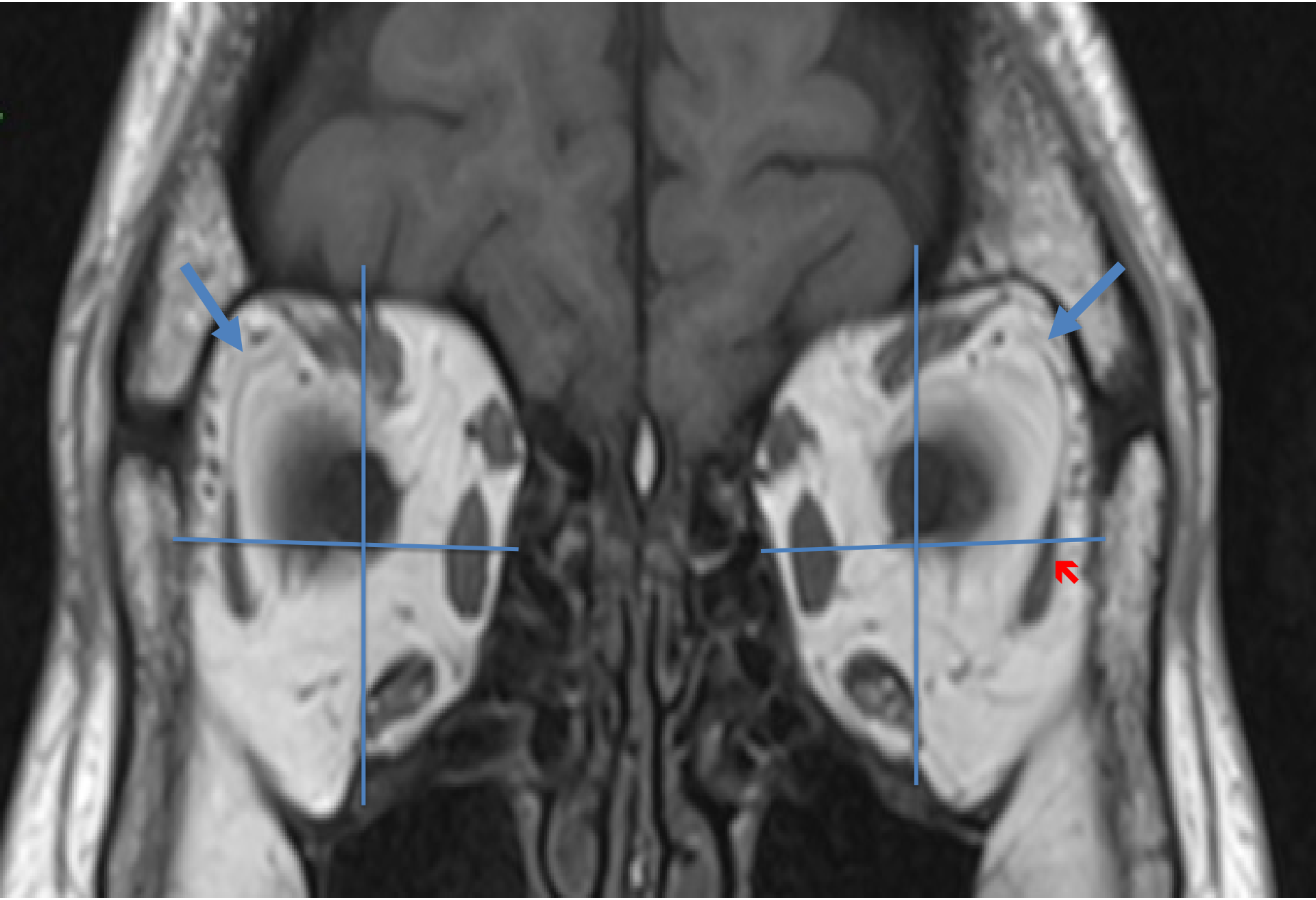
LR-SR band

- By MRI, LR-SR band thinning, discontinuity, or displacement, was seen in:
  - 100% of strabismic elderly orbits,
  - 67% of strabismic myopic orbits,
  - and 12.5% of control (ortho) elderly.

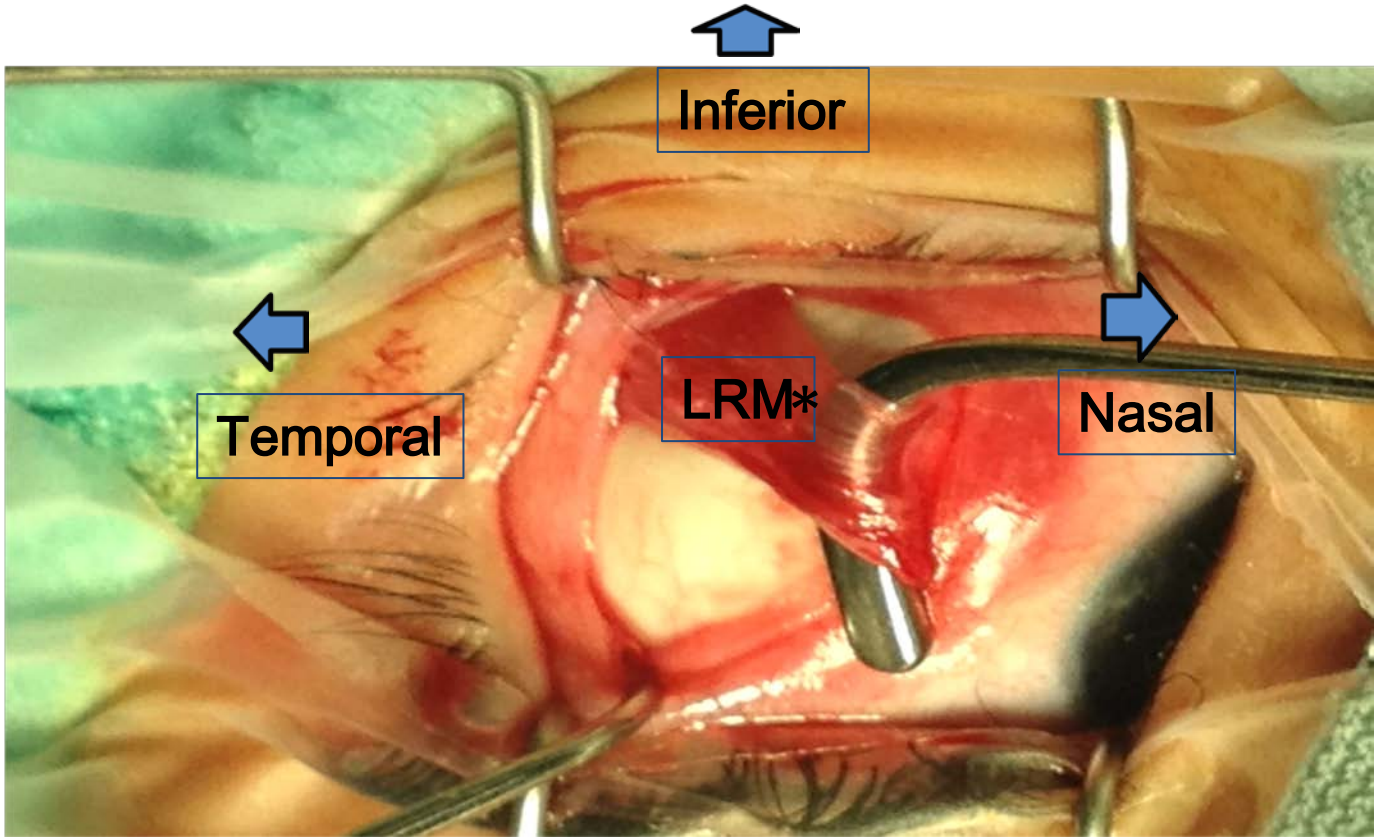
"Heavy Eye" Syndrome in the Absence of High Myopia: A Connective Tissue Degeneration in Elderly Strabismic Patients. J AAPOS. 2009 February; 13(1): 36–44.

Case #4. 47 yo, myope, sudden horizontal diplopia

- Intermittent for 6 weeks, then 6 weeks of constant horizontal diplopia.
- Glasses from age 10
- R -9 [29mm], 6/6
- L -6 [28.5mm], 6/12.
- Left esotropia 40Δ for distance, 35Δ for near.



Inferior path of sagging LLR (surgeon's view)



Demer, Clark, Miller: Advances in Strabismology

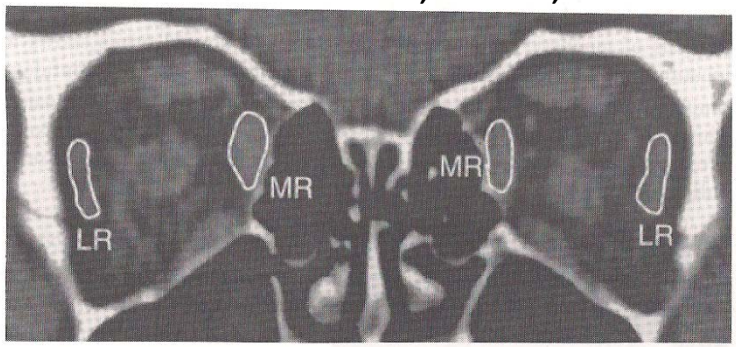


Fig. 2. 211 µm resolution, 1 mm thick coronal CT scan of the orbits of a 5 year old girl with large "V" esotropia and marked overelevation and underdepression of the right eye in adduction. Note inferior displacement of right LR more than left LR.

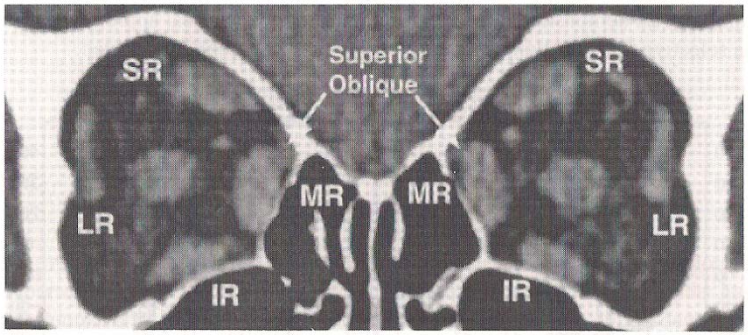


Fig. 3. CT scan of 6 year old girl with "A" pattern esotropia 60Δ greater in elevation than depression. Note LR displaced superiorly and SR displaced nasally in both eyes.

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

"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]

1

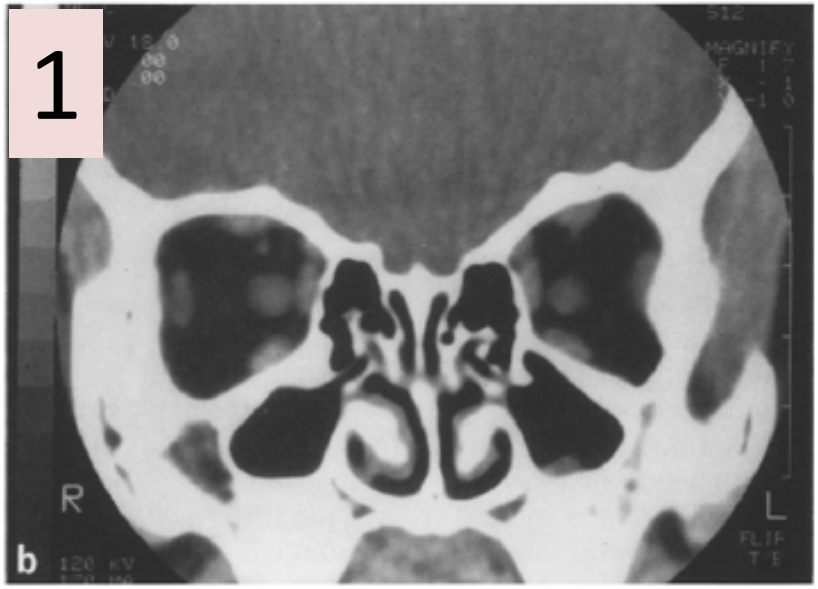


Fig. 1 a, b. Preoperative computed tomography (CT) scan in 3-year-old patient with V-pattern strabismus. **a** Scout film demonstrates plane of section perpendicular to the hard palate; **b** coronal section just posterior to the globe. The location, relationship, and dimensions of the four rectus muscles can easily be determined

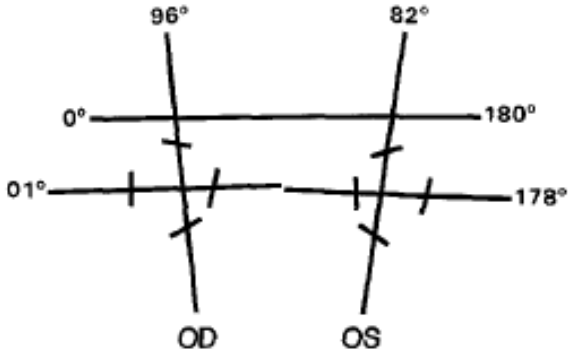


Fig. 2. Position tracing (using cylinder axis notation) of the rectus muscles (*bars*) shown in Fig. 1. Lines have been constructed between the midpoints of the medial rectus and lateral rectus muscles (*horizontal axis*) and the superior rectus and inferior rectus muscles (*vertical axis*). A *horizontal reference line* (0°–180°) is drawn along the floor of the anterior cranial fossa

2

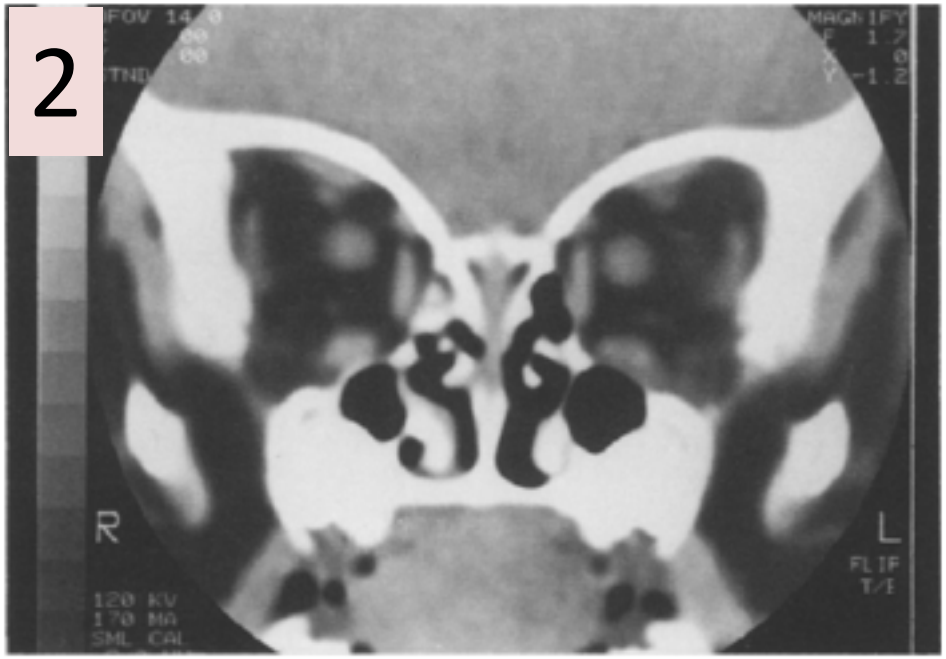


Fig. 3. CT scan in 6-month-old infant without strabismus; rectus muscles appear obliquely positioned, but have a normal relationship for the patient's age

Saunders showed:

1. **Normal arrangement of orbital pulleys in some pts with V patterns**
2. **Very abnormal pulley arrangement in some pts with NO strabismus**

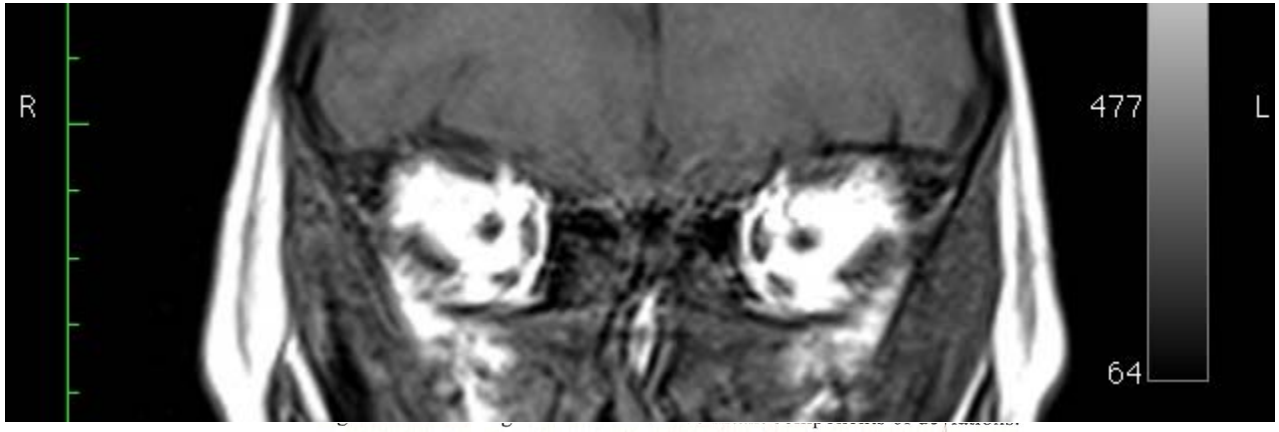
Childhood pulley disorders

12 yo
previous LR & IO weakening for V - XT

Recurrent / Residual
V- XT [UG 30, DG 10]
Minimal IO OA
No fundus torsion

Pulleys & Alphabet patterns: A work in progress

Coronal MRI T1: inf positioning of LR (L>R), and nasal shift of IR



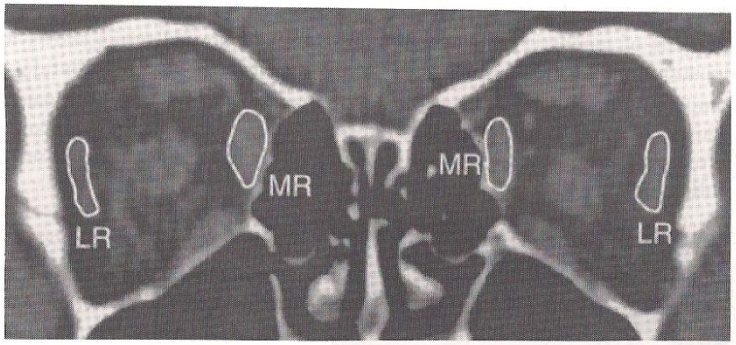
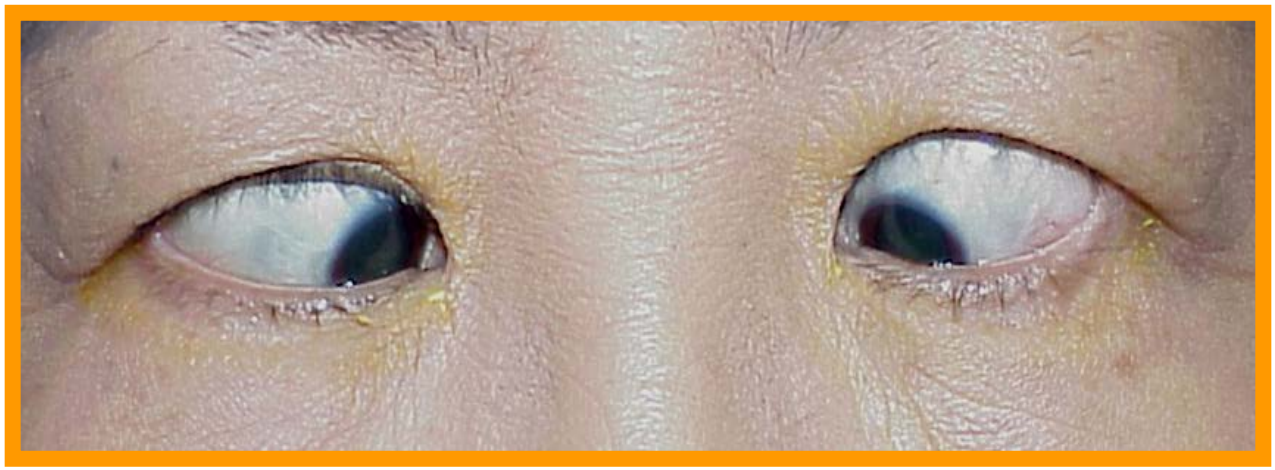


Fig. 2. 211 µm resolution, 1 mm thick coronal CT scan of the orbits of a 5 year old girl with large "V" esotropia and marked overelevation and underdepression of the right eye in adduction. Note inferior displacement of right LR more than left LR.

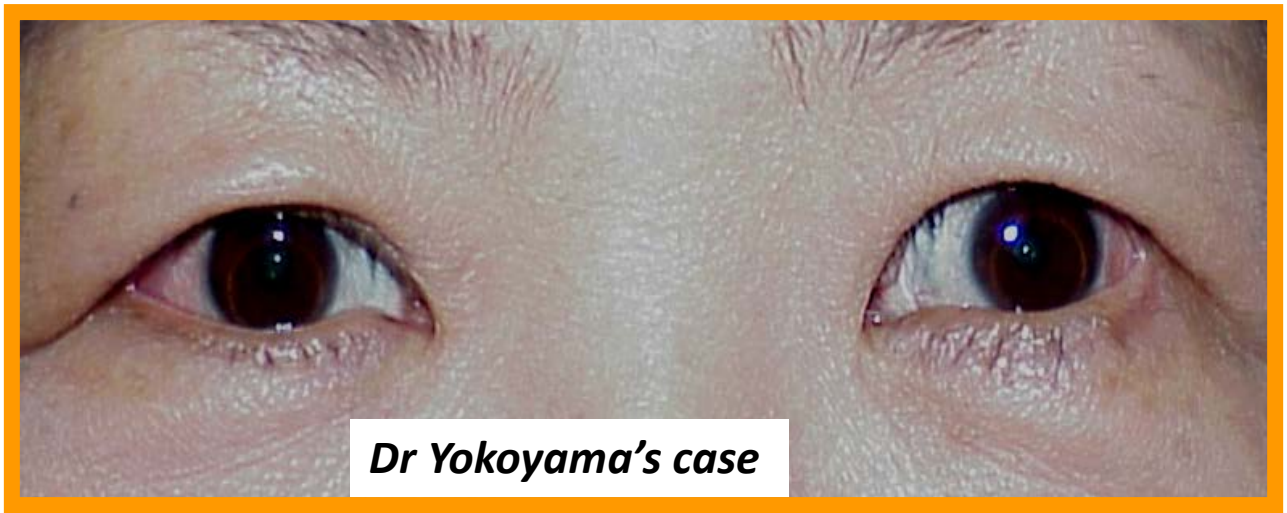
Extreme Esotropia of High Myopia

[aka Heavy Eye]

Preoperative



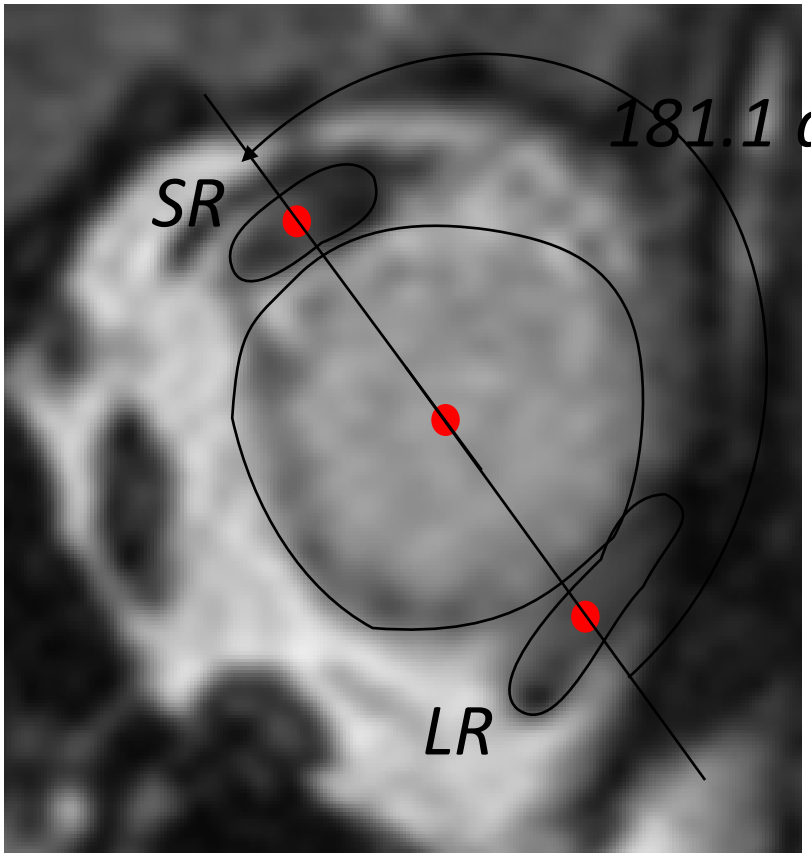
Postoperative (52 days after surgery)



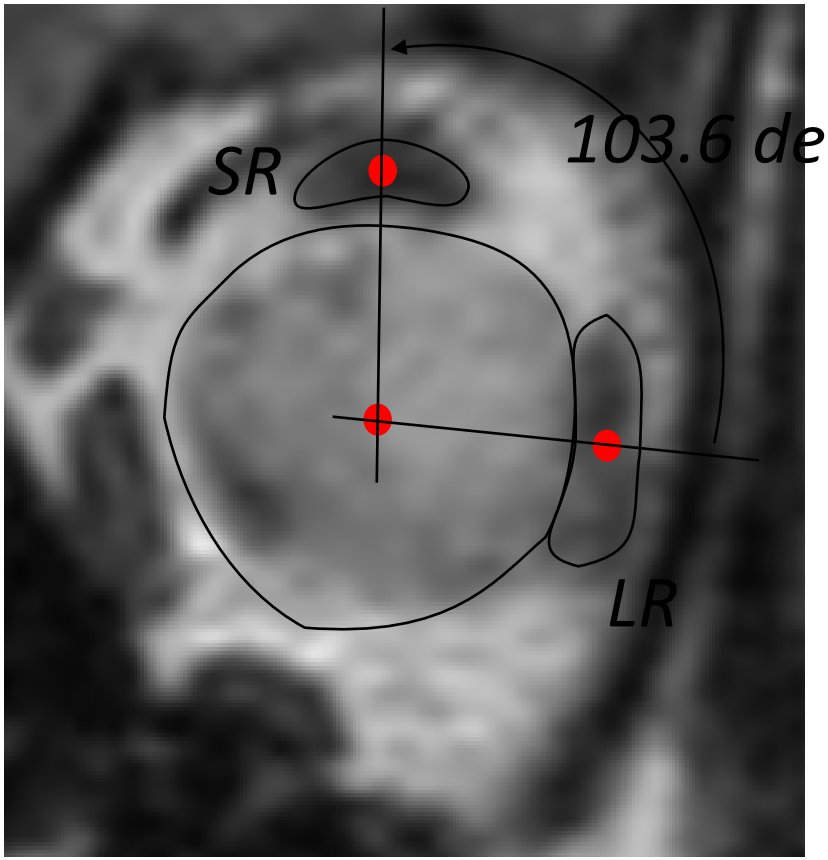
Dr Yokoyama's case

Extreme myopia:

LR displaced down, SR nasal



Preoperative



Postoperative

From Tsuranu Yokoyama

Strabismus of high myopia

Heavy eye syndrome versus sagging eye syndrome in high myopia

Roland Joseph D. Tan, MD,[a] and Joseph L. Demer, MD, PhD[a,b,c,d]

BACKGROUND Heavy eye syndrome (HES) presents with esotropia and limited abduction due to superotemporal globe shift relative to the extraocular muscles. Sagging eye syndrome (SES) was originally described in nonmyopic patients exhibiting distance esotropia and cyclovertical strabismus with limited supraduction due to lateral rectus muscle inferodisplacement caused by degeneration of the lateral rectus–superior rectus (LR-SR) band. We hypothesized that SES might also cause strabismus in high myopia.

METHODS Eleven strabismic subjects with high myopia underwent ophthalmological examination and orbital magnetic resonance imaging (MRI) to assess quantitative orbital anatomy.

RESULTS Of 11 subjects, 5 had HES; 6, SES. Mean axial length in subjects with HES was 32 ± 5 mm; in subjects with SES, 32 ± 6 mm. Average distance esotropia in subjects with HES was $61^{\Delta} \pm 39^{\Delta}$; hypotropia was $26^{\Delta} \pm 21^{\Delta}$. Average distance esotropia in subjects with SES was $23^{\Delta} \pm 57^{\Delta}$; hypertropia was $2^{\Delta} \pm 2^{\Delta}$. All 5 subjects with HES had superotemporal globe prolapse; the LR-SR band was thinned in 6 orbits and ruptured in 2. The mean angle between the lateral rectus and superior rectus muscles in HES was 121° ± 7°. In SES the LR-SR band was thinned in 7 orbits and ruptured in 5, with superotemporal soft tissue prolapse. The mean angle between the lateral rectus and superior rectus muscles was 104° ± 11°, significantly less than in HES ($P < 0.001$).

CONCLUSIONS SES occurs in highly myopic patients who also exhibit less relative globe dislocation than in HES. Unlike HES, SES exhibits superotemporal soft tissue prolapse that may limit superotemporal globe shift. The distinction is important because surgery for HES uniquely requires creation of a surgical connection between the superior rectus and lateral rectus muscles, whereas SES may be treated with conventional surgery. SES can cause strabismus in high axial myopia. Orbital MRI is useful in differentiating SES from HES. (J AAPOS 2015;19:500-506)

Strabismus of high myopia

E-Poster Viewer

POSTER LIST | SEARCH

84. "KNOBBY EYE SYNDROME:" IRREGULAR POSTERIOR STAPHYLOMA AS A FACTOR COMPLICATING STRABISMUS IN HIGH MYOPIA.

by Joseph L. Demer, MD, PhD, Stein Eye Institute and Department of Neurology, UCLA.

SteinEye

Introduction

Traditional concepts of strabismus assume a spherical globe. This study employed magnetic resonance imaging (MRI) to demonstrate complex effects of irregular posterior staphylomata in axial high myopes with strabismus.

Methods

High resolution (2 mm planes, 312 micron resolution), surface coil axial MRI was obtained in central gaze in 18 highly myopic subjects with strabismus. Most were esotropic. Scans were repeated in dextro- and levoversion in most subjects. Images were analyzed for axial length, scleral shape, and deflection of muscle paths.

Results

Knobby Eyes Are Older

Six of 18 myopes had uniformly shaped

Example of Uniform Globes

36 year old esotropic woman with axial length 28.8 mm OS, 27.4 mm OS. Muscle paths were uniform during horizontal duction in 6 patients like this.

Knobby Posterior Staphyloma

36 year old woman with > 100Δ ET and axial length 33.7 mm OD, 34.4 mm OS. Lateral rectus was stretched by staphyloma in adduction.

Asymmetrical Equatorial Staphyloma Deflects Lateral Rectus Path

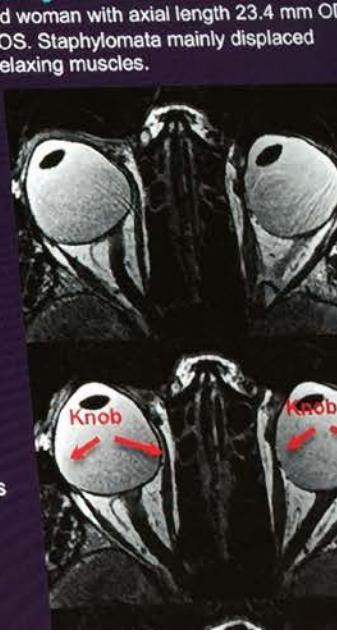
57 year old woman with axial length 25.8 mm OD, 27.0 mm OS. Lateral rectus was stretched over equatorial staphyloma.

Discussion

Irregular equatorial or posterior staphylomata are common in strabismic axial high myopes. Staphylomata act like "cams" affixed to the normally spherical globe; thus staphylomata have no mechanical effect until they eccentrically rotate against muscles. After rotational contact, staphylomata would add tension to the muscle that increases non-linearly with further duction. This

Symmetrical Equatorial Staphyloma Deflect Relaxing Horizontal Rectus Paths

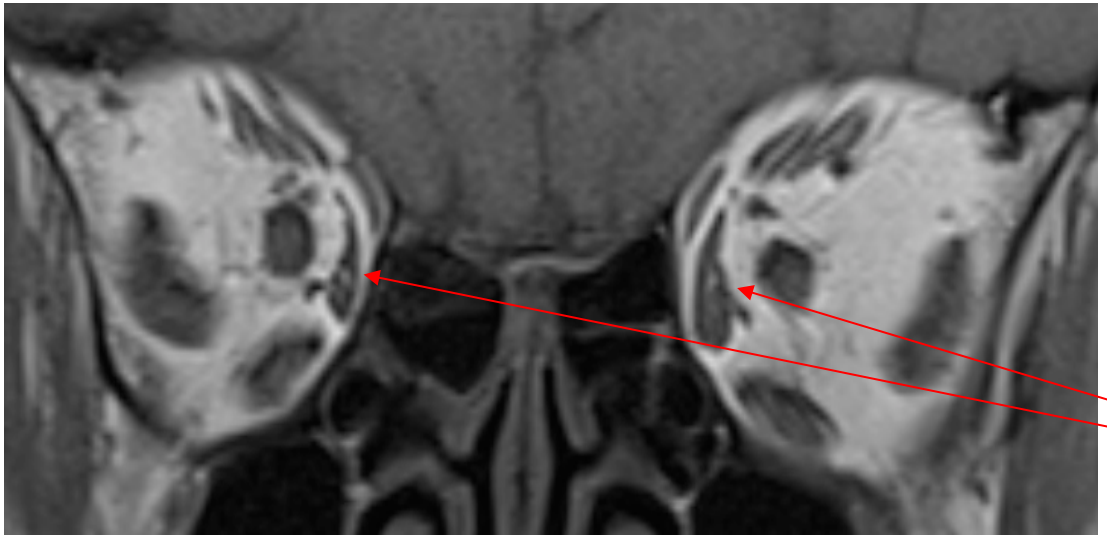
57 year old woman with axial length 23.4 mm O 27.8 mm OS. Staphylomata mainly displaced paths of relaxing muscles.

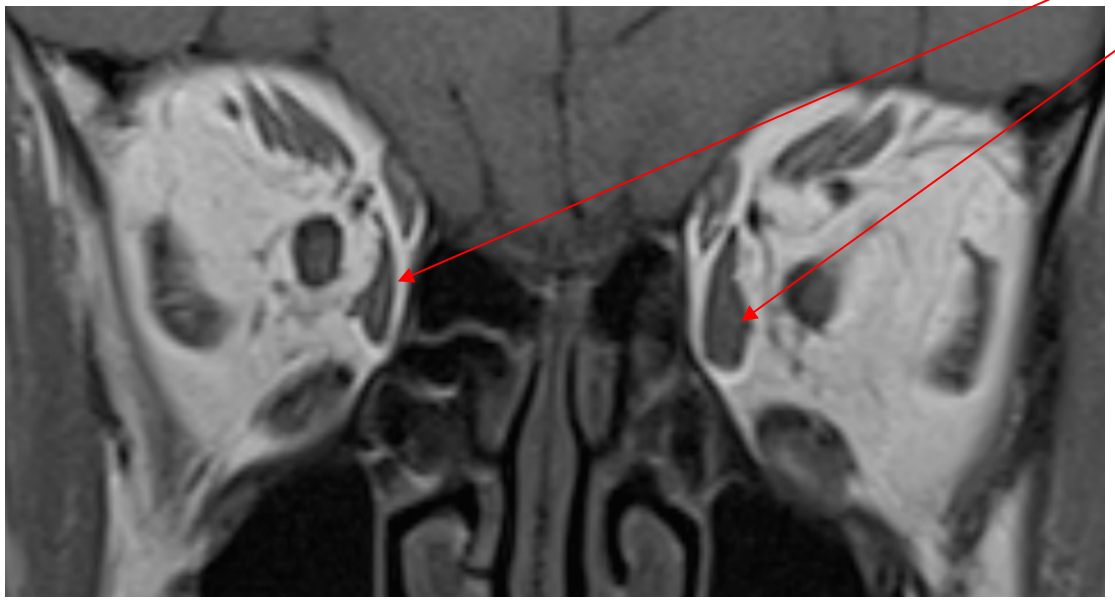


Exotropia & MR compartment hemi-atrophy

- 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 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

The radiology of strabismus is 25+ years old. Where are we heading?

- It has increased our understanding of the pathophysiology of many strabismus conditions
- We have introduced new treatment paradigms
- Abnormal radiological findings may have no clinical significance
- Alphabet patterns: confusing data
- We need to educate & enthuse ophthalmologists and radiologists