

NYSTAGMUS DECODED PART 2

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Thank you for having me

VERTICAL HEAD POSTURE MANAGEMENT

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No \$,£,¥,€ & conflict disclosures



Multiple Nulls in Infantile Nystagmus IN

First few slides:

- Version null
- Vergence null
- Version surgery
- Vergence surgery

...

Multiple Nulls in Infantile Nystagmus IN

Version nulls: The 3 T's*

- Any one patient with Infantile Nystagmus can have several nulls:
- 3 T's: Turn / **Tip** / Tilt * are **Version Nulls** that drive Abnormal Head Posture
- The patient will tend to use the null that :
 - Gives better vision**
 - Is *orthopedically* more comfortable***
 - Often: a *compromise* between vision & comfort

*** Many pts do not posture most of the time because they cannot ...**and if they could, they would** [Hertle]

** Spectacle frames and correctly centred lenses can influence this

* Thank you Annette Spielmann

➤ Nulls in Infantile Nystagmus IN

Vergence nulls: Convergence null for near and distance

- You can see / measure a Turn / **Tip** / Tilt across the room
- You have to get close to examine a child to find a Con**Vergence Null**
- If you find a Convergence Null for Near CNN, you must now look for a **Convergence Null for Distance CND**
- Add 7Δ BO OU +/- -1 DSOU to look for a CND in the office
- If present, wear these glasses in real life for 1-2 weeks to see **if patient prefers this vergence null to a version 3T null.**
- Is so, Vergence surgery (BMR = Artificial Divergence Surgery) is now an option

How common is CND?

- n=88 consecutive Eye Movement Recordings
- 29/88 = **1/3 have CND**
- *True % CND > EMR- demonstrated % CND*

<u>Type of nystagmus</u>	<u>CND*</u>	<u>No CND *</u>
IN 53 [60%]	23 [43%]	30
PAN ** 18 [20%]	6 [33%]	12
IN & FMN 4	1	
FMN 8	0	
Uncertain 5	0	

** PAN is an underestimate - some children labelled IN will later turn out to have PAN

* CND is an underestimate – some children are too young to assess

Why bother looking for a CND in someone with a Tip from IN?

Vergence surgery vs Version surgery in IN

- ..because Artificial Di**Vergence Surgery** in IN is often more effective than **Version surgery** [eg Kestenbaum surgery to shift the null] to obtain & maintain a large broader primary position null

Joshua age 5

- Moderate hyperope
- Wearing CLs R +5.5, L +4.5 Cyclo R +6.5, L +6
- Acuity LH BEO 20/50, either eye 20/70 OU
- Intermittent R eye closure
- sc ET, cc EX/EX'=0
- Findings of OCA: TIDs, ?small discs, some macular definition with indirect

'Always' looks like....

Reading 20/80 LH line on M&S, refreshing every 4 sec

Wearing his CLs



With protractor, face turn to L 25+ °, **tip up 20°**

7Δ BO OU & -1 DS OU

creates distance conv null & *straightens both turn & tip*



Next steps....

Optical Treatment

- Trial of glasses:
7Δ BO OU & -1 DS OU over his full + CLs to wear all the time
- If effective, continue glasses or consider VERGENCE SURGERY
- If ineffective, VERSION surgery (IR Rc OU + SO tenotomy OU + RLR/LMR Rc)

Vergence Surgery

- If spectacle trial in real life shows that CND is frequently used: BMR 3mm +/- Tenotomy- Resuture LR OU.
- If spectacle trial shows that he still has some tip up, both Vergence surgery (BMR 3mm) and Version surgery (IR Rc OU + SO tenotomy OU)

Tip UP

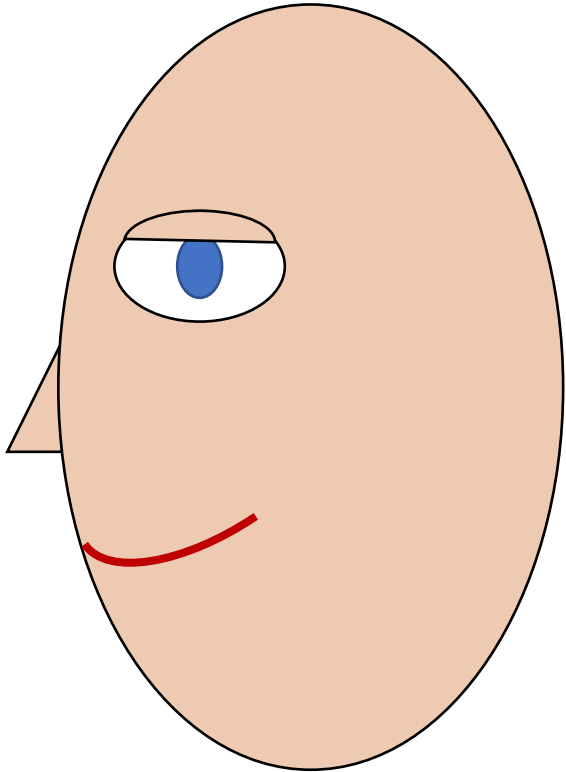
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- Tip up: depressors recruited, esp the IR & SO.
- This causes a reduction in the nystagmus = null zone on downgaze

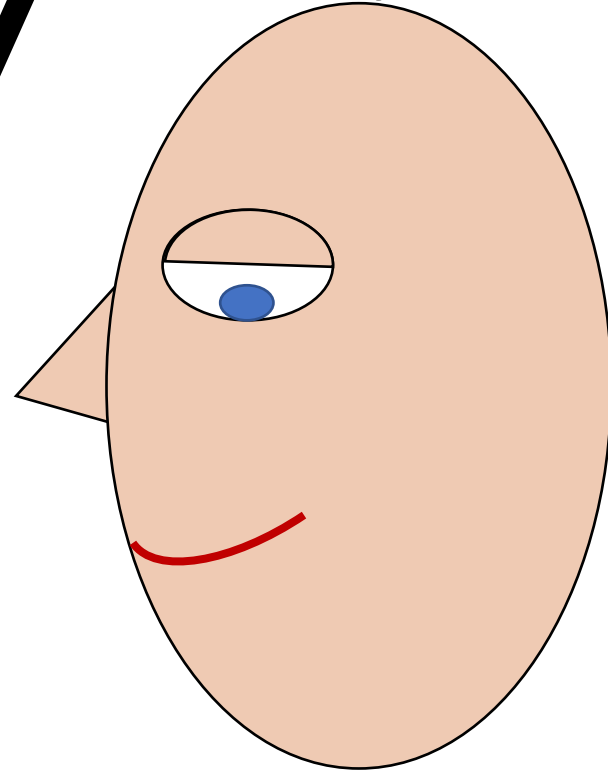
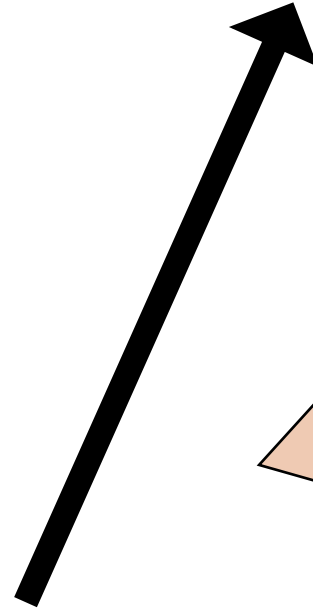
How?

- Possibility 1: **Excess innervation by IR acts as a brake on the IN**
- Possibility 2: Some other innervational magic stops the nystagmus in this position

6/30 Obvious IN



Tip up
Acuity 6/12



IR recruited
Excess innervation acts
as a brake on the IN

Tip UP

2

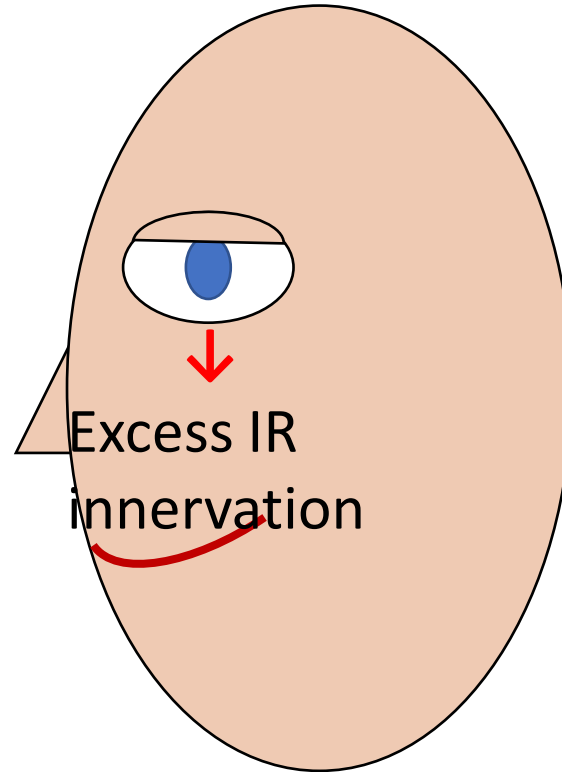
- We normally recess an IR for an eye that has hypotropia.
- IR recess for an eye that is not hypotropic will produce fixation duress in primary position.

Fixation duress?

- This results in excess innervation to the IR to maintain primary position, similar to the excess innervation that acts as a brake on IN.
- This may be how IR recess re-creates the effects of downgaze null in primary position.

After IR recess, Fixation Duress in Primary Position causes excess innervation in IR

This acts as a brake on the IN & the downgaze null is now shifted to Primary Position



We will see later how IR Rc alone is inadequate about ½ the time in producing the desired effect

Tip UP

3

- So we achieve the tip-up effect on IR innervation by recessing IR and no tip-up is now needed to brake the nystagmus = fixation duress

How can we augment this effect if needed?

1. Larger IR recess
2. SR resect or plicate
3. SO weakening

....all of these cause more fixation duress

A look at the literature Hertle 2010

Clinical & Experimental Ophthalmology

Surgical Procedure For Treatment Of Vertical Head Posturing Associated With Infantile Nystagmus Syndrome (INS): Results in 24 Patients

Journal:	<i>Clinical and Experimental Ophthalmology</i>
Manuscript ID:	CEO-10-01-0071
Manuscript Category:	Original Article - Clinical Science
Date Submitted by the Author:	29-Jan-2010
Complete List of Authors:	Hertle, Richard; UPMC Eye Center, Ophthalmology Yang, Dongsheng; UPMC Eye Center Adams, Kenneth; UPMC Eye Center Caterino, Roxanne; UPMC Eye Center
Keywords:	Nystagmus, Surgery, Strabismus, Strabismus surgery

Hertle series n=24
13/24 have albinism

13 tip down

- SR Rc 5mm OU
- IO myectomy
- + one horizontal muscle per eye

11 tip up

- IR Rc 5mm
- SO tenectomy 5mm nasal to SR
- + one horizontal muscle surgery per eye

Hertle series

Follow the surgical recipe with great care

- IO myectomy
 - I clamp the muscle where it penetrates through posterior Tenon's with a hemostat, stretch the muscle between the hemostat and the LR then cut it off the globe near the LR, then cut muscle anterior to the hemostat, then cauterize the stump, release stump into orbit.....big myectomy

Postop

- 1. All pts had mild vertical gaze paresis [in intended direction]
- 2. Average acuity improved from 0.6 [6/24] to 0.4 [6/15]
- 3. 20/24 pts had improved acuity
- 4. 5/24 had 0.3 Logmar or greater improvement in acuity

Hertle - acquired torsion

- Large IR Rc : likely to have a net incyclo effect
- SO tenectomy: ...net excyclo...
- Net expected torsion: 0
- *If the operation is performed symmetrically and the patient fuses before, the patient will fuse after, if the patient does not fuse before there may be a long term secondary bilateral OAIO, which may or may not need another procedure.*
- ...& vice versa for tip down head posture

A look at the literature 2

Major Articles

Vertical Rectus Muscle Surgery for Nystagmus Patients With Vertical Abnormal Head Posture

shot

Michael B. Yang, MD,^a Carlos R. Pou-Vendrell, MD,^b Steven M. Archer, MD,^c E. Jean Martonyi, CO,^c and Monte A. Del Monte, MD^c

Purpose: To characterize the results of vertical rectus muscle recession, or recession and resection surgery for vertical abnormal head posture (VAHP) due to null-point nystagmus. **Method:** This is a retrospective, noncomparative, consecutive case series of patients who underwent vertical rectus muscle recession alone, recession then resection, or combined recession/resection surgery for VAHP due to null-point nystagmus. The primary outcome measure was the amount of residual VAHP. **Results:** Twenty patients with VAHP due to null-point nystagmus were studied. The mean follow-up interval after surgery was 49 months (median: 44 months; range 9 to 124 months). Preoperative VAHP ranged from 10° to 45°, and the total amount of bilateral vertical rectus muscle recession, or recession and resection surgery, ranged from 8.5 to 20.0 mm per eye. After recession alone, 5/11 had residual VAHP ≤ 5°. The remaining six underwent subsequent resection or plication for residual VAHP > 10° that resulted in residual VAHP ≤ 5° in each case. After combined recession/resection, 6/9 had residual VAHP ≤ 5° and the remaining three had VAHP < 10°. **Conclusion:** Large amounts of vertical rectus muscle surgery are indicated to successfully correct the VAHP associated with nystagmus with a vertical null point. Combined recession/resection of the appropriate vertical rectus muscles, instead of recession alone, appears to be a more effective procedure. (J AAPOS 2004;8:299-309)

Michigan n=20

- 4/20 Acquired neurological nystagmus, not IN
- **11/20: Recession alone as 1st procedure**
- **5/11: 2nd resect surgery follows**
- 9/20: Recess/ resect surgery as 1st procedure

300 *Yang et al*

TABLE 1. Diagnoses of nystagmus patients with null point and VAHP

Diagnosis	No. of Patients
A. Congenital Motor Nystagmus	8
B. Sensory Nystagmus	8
Aniridia	1
Ocular albinism	3
Optic atrophy	3
Hydrocephalus	1
Inherited	1
Idiopathic	1
Visual deprivation	1
Congenital cataracts	1
C. Acquired Nystagmus	4
Cerebellar degeneration	1
Brainstem tumor/WEBINO	1
Myelomeningocele/hydrocephalus (without optic atrophy)	1
Traumatic brain injury/MVA	1
	Total 20

VAHP: vertical abnormal head posture; MVA: motor vehicle accident; WEBINO: wall-eyed bilateral internuclear ophthalmoplegia.

Michigan – surgical results

- Recess- alone [total dose $\leq 10\text{mm}$] n=11:
- 6/11 tip no better or worse- needed 2nd surgery
- Better results with combined Rc-Rs

Recommend

- 10-15 deg: 12mm [6mm each on IR & SR]
- 20-25 deg: 16mm
- 30+ deg: 20mm

306 *Yang et al*

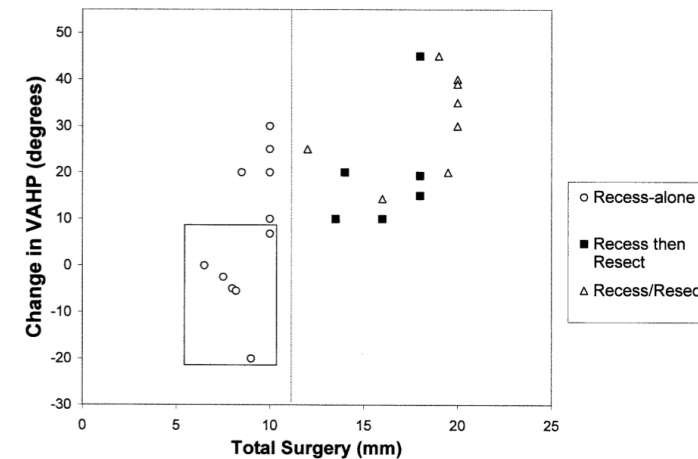


FIG 3. Change in VAHP was plotted against the total amount of surgery (recession and resection in mm). All recession-alone patients had 10 mm or less of surgery (○). All patients had residual VAHP $< 10^\circ$ except for the six recession-alone patients in the demarcation square, who had residual VAHP $> 10^\circ$ and required operation. The results after reoperation are designated as recession then resection (■) and show a similar trend as the patients who had combined recession/resection (△) in one operation.

Michigan - acquired torsion

- Large IR Rc : likely to have a net incyclo effect
- Large SR Rs: likely to have a net incyclo effect
- ...& vice versa for tip down head posture
- 0/16 [presumed] infantile nystagmus had acquired torsion

A look at the literature 3

> [J Pediatr Ophthalmol Strabismus](#). 2018 Jul 1;55(4):234-239.
doi: 10.3928/01913913-20180327-01. Epub 2018 May 29.

The Role of Superior Oblique Posterior Tenectomy Along With Inferior Rectus Recessions for the Treatment of Chin-up Head Positioning in Patients With Nystagmus

[Anna G Escuder](#), [Milan P Ranka](#), [Kathy Lee](#), [Julie N Nam](#), [Mark A Steele](#)

PMID: 29809265 DOI: [10.3928/01913913-20180327-01](#)

IR recess doses combined with
SO post 7/8 tenectomies:
< 30 deg chin up 6mm
30 deg chin up 7mm
45 deg chin up 7.5mm

Abstract

Purpose: To evaluate the clinical outcomes of bilateral superior oblique posterior 7/8th tenectomy with inferior rectus recession on improving chin-up head positioning in patients with horizontal nystagmus.

Methods: Medical records were reviewed from 2007 to 2017 for patients with nystagmus and chin-up positioning of 15° or more who underwent combined bilateral superior oblique posterior 7/8th tenectomy with an inferior rectus recession of at least 5 mm.

Results: Thirteen patients (9 males and 4 females) were included, with an average age of 7.3 years (range: 1.8 to 15 years). Chin-up positioning ranged from 15° to 45° degrees (average: 30°). Three patients had prior horizontal muscle surgeries, 1 for esotropia and 2 for horizontal null zones causing anomalous face turns. Ten patients underwent other concomitant eye muscle surgery: 3 had esotropia, 1 had exotropia, and 2 had biplanar nystagmus null point requiring a horizontal Anderson procedure. Four patients underwent simultaneous bilateral medial rectus tenotomy and reattachment. All patients had improved chin-up positioning. Eight patients had complete resolution, whereas 5 had minimal residual chin-up positioning. Three patients developed an eccentric horizontal gaze null point with compensatory anomalous face turn with onset 2 weeks, 2 years, and 3 years postoperatively. Average follow-up was 42.7 months. No postoperative pattern deviations, cyclodeviations, or inferior oblique overaction were seen. No surgical complications were noted.

Conclusions: Bilateral superior oblique posterior 7/8th tenectomy in conjunction with bilateral inferior rectus recession is a safe and effective procedure for improving chin-up head positioning in patients with horizontal nystagmus with a down gaze null point. [[J Pediatr Ophthalmol Strabismus](#). 2018;55(4):234-239.].

NY acquired torsion

- IR Recess : net intorter
- SO intorsion effect: not lessened because ant 1/8 still intact
- No torsional problems in this series
- Shetty: reported 1 torsional complication to AAPOS server 2015

Subsequent surgeries

Hertle, Steele

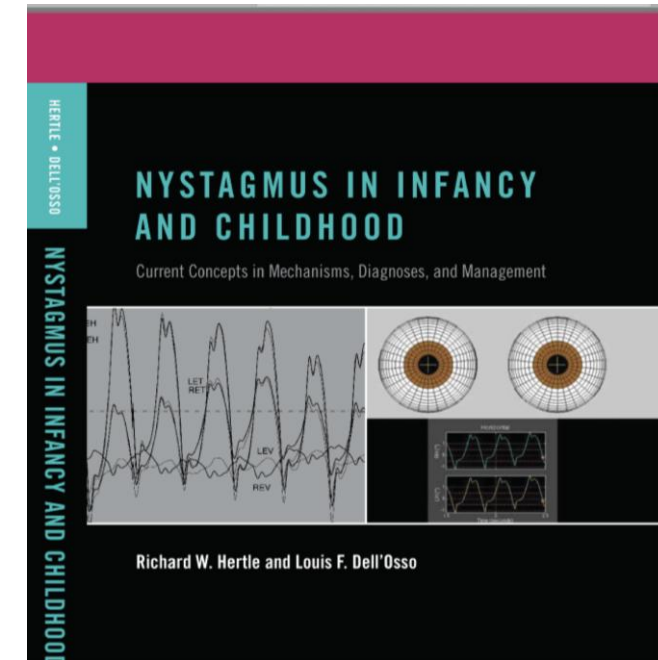
- Room for horizontal surgery with less worry about ASI

Michigan

- Worry about ASI if pt needs horizontal surgery after vertical Rc-Rs

My best recommendations to you for tip up AHP

- 1. Look hard for conv null for distance. If present and proved useful in real life, BMR is the best surgery
 - 2. Tip up: Large IR Rc effective $\sim 1/2$ the time
 - 3. IR Rc + bilateral SO weakening effective nearly all the time
 - 4. Follow published surgical recipes carefully.
 - 5. Get Rich Hertle & Lou Dell'Osso's book.
- OMLAB.org. Link is on 1st page. Good price – free.



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- Annette Spielmann, Nancy, France
- Richard Hertle, Everywhere, USA

köszönöm !תודה *děkuji*

mahalo 고맙습니다

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thank you

merci 谢谢 *danke*

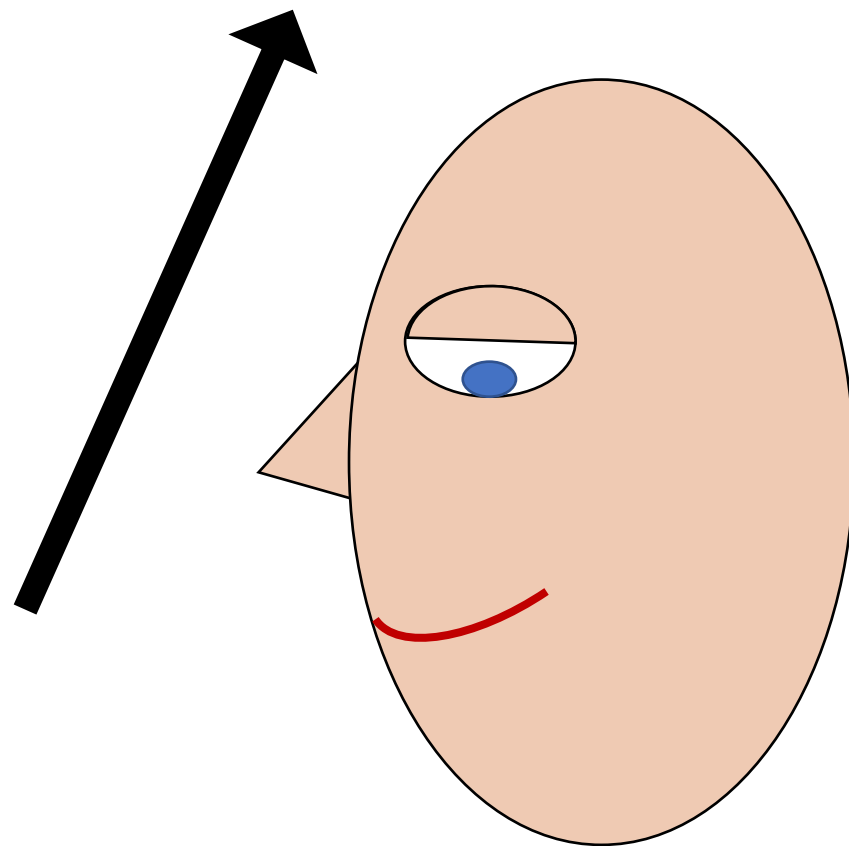
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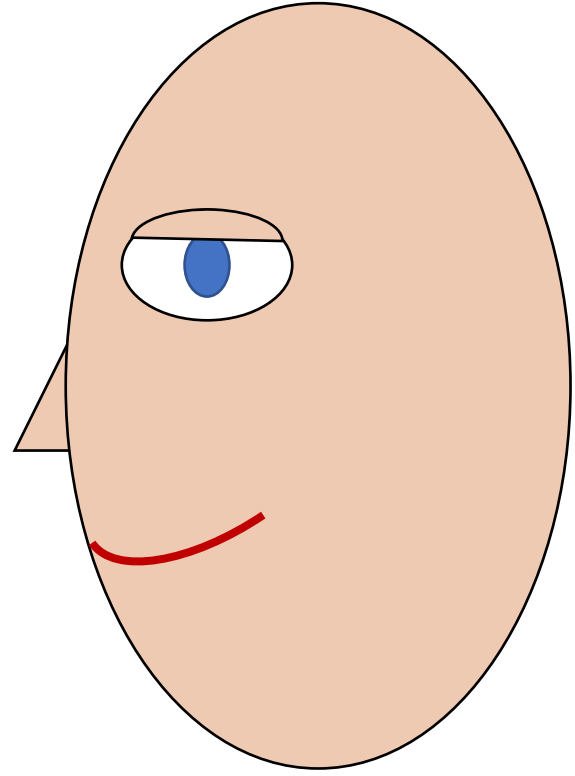
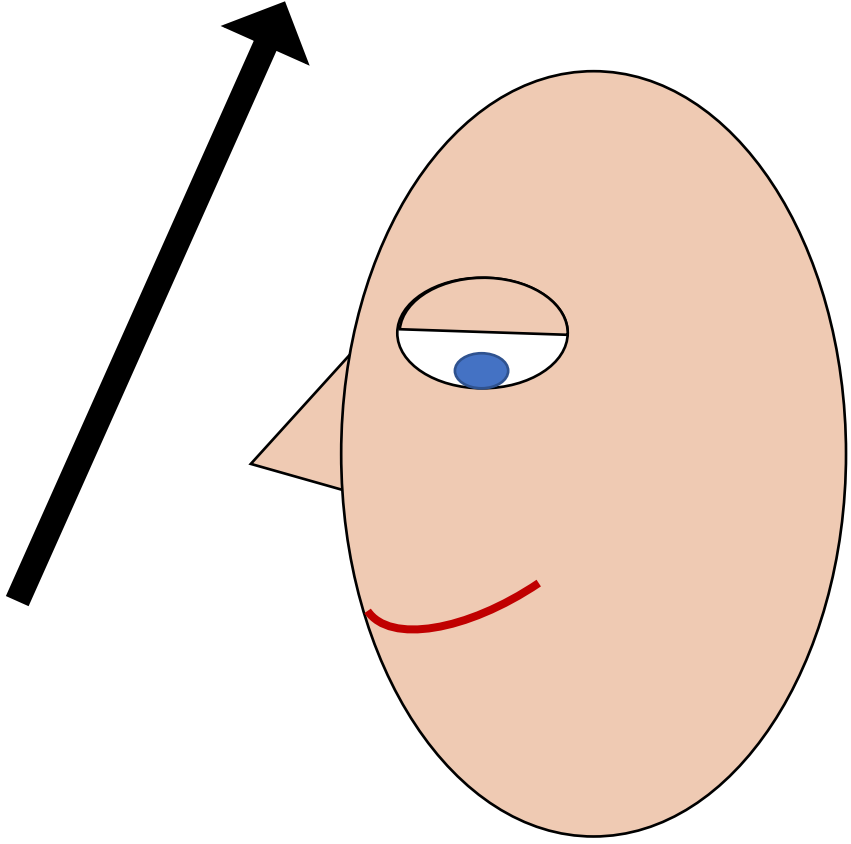
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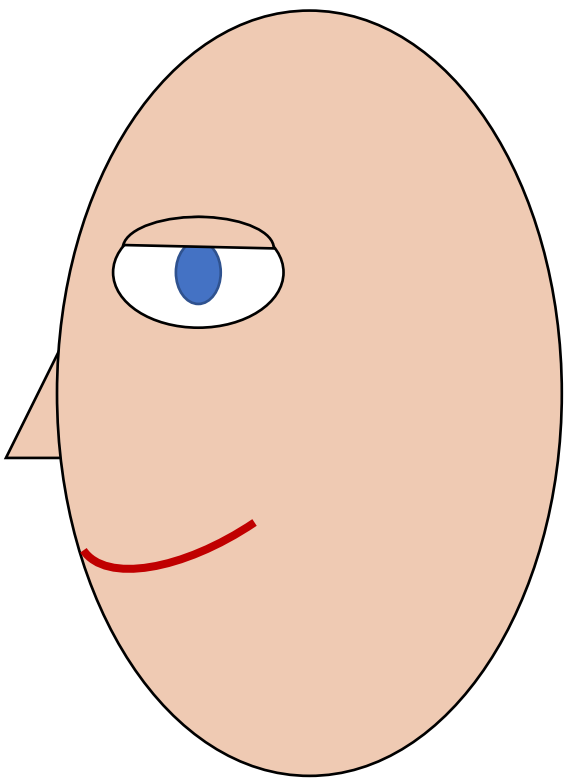
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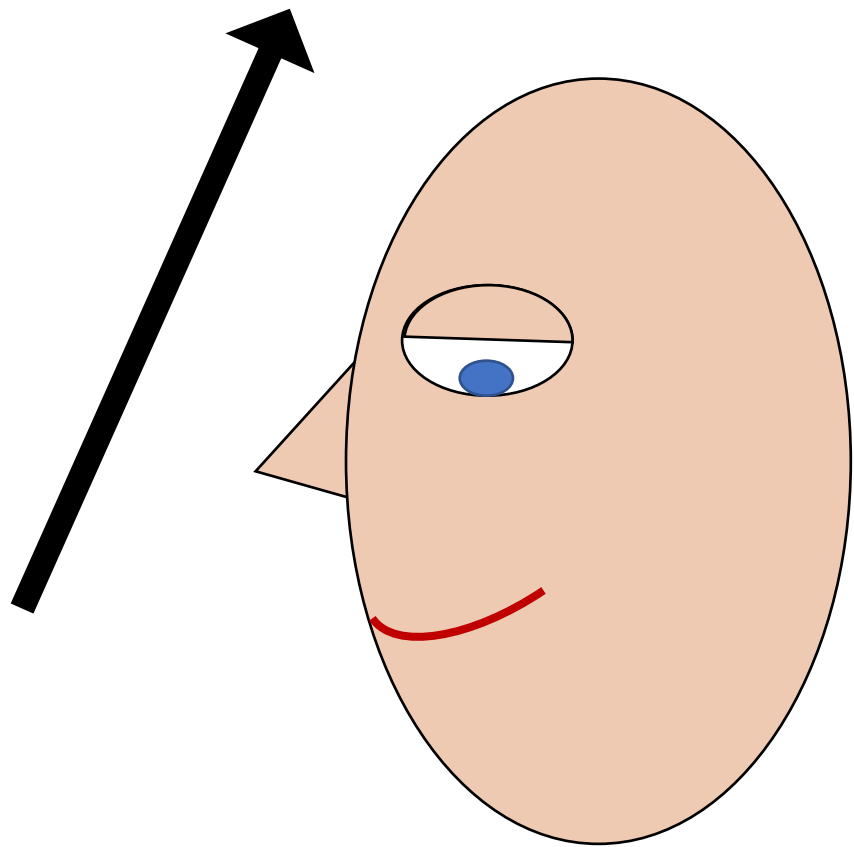




Problems with each muscle surgery and how to minimize/ avoid these problems

- 1. IR Rc: torsion, lid position
- 2. SO tenotomy: torsion
- 3. IO surgery: torsion
- 4. SR Rs/plicate: torsion, lid position





Albinism & Vertical Nulls

Albinism

- More likely to have vertical nulls
- More likely to have PAN
- More likely to have CND

Vertical nulls. More common in:

- Albinism
 - Optic n hypoplasia
 - Retinal dystrophy
- ...anterior visual pathway disease