Childhood nystagmus:

in children and adults

What to do when you don't know what to do = DKWTD

Lionel Kowal

Melbourne, Australia

Case #1 **14 year old girl, rhythmic gymnast**

- **Vision**: Right eye: 20/20; Left Eye: 20/20

- Straight for D & N

- **Acuity with both eyes open:**
  - Primary Position: 20/40
  - **Left Gaze: 20/20, so Face Turn FT to R**
  - Right gaze: 20/80

- **Cyclo:** +0.50 D OU

- **Titmus:** 40 “

#1: **14 year old girl, rhythmic gymnast**

- Nystagmus with null in left gaze
- ***Right face turn*** 35° & small head tilt L
- No face turn with 45Δ BR OU
- **No face turn with 7Δ BO OU & -1 DSOU**
- Family history of Nystagmus: older brother, many cousins & other relatives

Mom: Problem Description

…has to throw an apparatus, make a number of spins, pivots or tumbles and then catch the apparatus before it hits the ground.

….as she approaches the international levels the elements and apparatus use is becoming more complex, faster and involving more maneuvers with apparatus (ball, hoop, ribbon, clubs and rope) that involve spin or rotation..

….spinning and/or tumbling sets in the process the rapid and erratic eye movement and the finish of the element is spoilt through lack of balance or vision

….is required to spot straight ahead whilst she pivots…. difficult because her position of null movement is out to the left of the straight ahead position

Patient Description

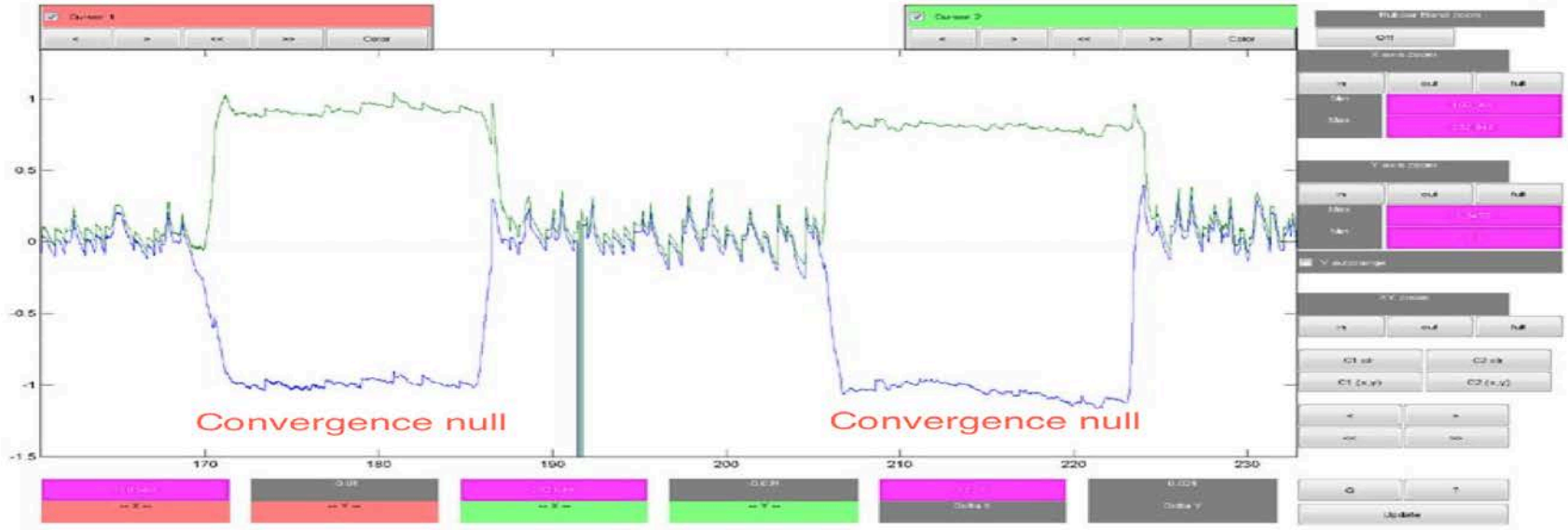
“I catch the apparatus by feel and practice, but not by using my vision”

Dr’s Problem

Nystagmus interfering with highly skilled performance in a highly talented girl

Eye Movement Recordings: Left Gaze Null

Eye Movement Recordings: Convergence Null



Treatment Plan

- 7Δ BO OU & -1.50 DS OU glasses to assess the effect of a convergence null for distance (CND)

Result all +ve:

- Head Position improved
- Vision clearer in primary position
- Less eye movement and rapid eye adjustment after spin and pivot
- But “objects appeared smaller and farther away”: ?effect of over-minus glasses

Surgical plan

1) *Artificial Divergence Surgery*[1] to duplicate the effect of the Δ glasses and exploit convergence null for distance.

2) Lateral Rectus tenotomy and re-attach muscle to sclera to dampen nystagmus and ?further improve foveation time[2]

[1] **Clinical rationale for manifest congenital nystagmus surgery.**
Spielmann A.
J AAPOS. 2000 Apr;4(2):67-74.

[2] **Horizontal rectus muscle tenotomy in children with infantile nystagmus syndrome: a pilot study.**
J AAPOS. 2004 Dec;8(6):539-48.
Hertle RW, Dell'Osso LF, FitzGibbon EJ, Yang D, Mellow SD

Post – op Results

- “An amazing and immediate improvement. She came straight over to me after the first evening training session and said this is excellent…..She can see!” Coach

- “She pivots and spins and we notice that her eyes are essentially still with much less wobble and eye movement compared to previous observations”
  Mom

Retrospectoscope …

- Typical ‘old’ CMN
- Once I had determined that BOΔ fixed her problem, there was no need to record her nystagmus.
- Rx options then were

1. Continue with Δ glasses
2. BMR
3. BMR +/- T-R LR OU

DKWTD
Look for Convergence Null for Distance = CND

PRE-REQUISITES

- Horizontal jerk N
- Eccentric null
- No strabismus
- Some sensory fusion
- Convergence null for near

LOOK FOR

- **Convergence null for Distance.**
- **Does 7Δ BO & -1 DS OU** over present correction give the pt the same or greater benefit as the eccentric null in real life?
- Give & wear **Δ** glasses: do not rely on in-office findings alone – some false +ves

N = Nystagmus

Convergence Null for Distance = CND

- Many pts with IN & PAN
- have CND
- do not have CND
- Look for CND : if there is conv null for near, orthotropia, some sensory fusion
- If Δ work in ‘real life’, the effect can be reliably reproduced by Artificial Divergence Surgery [BMR 3mm]
- Don’t need recordings

Convergence Null for Distance = CND

- Near-zero morbidity from BMR in these pts
- Spielmann FR 🏠: 10% re-op rate for consec XT.
- LK: 0% □
- Δ glasses usu need some minus [CA/C]. Same effect from BMR seems to never? needs minus

- +ve Kappa in OCA causes pseudo-XT: parents may think your BMR has done this
- *Apparent CND: also with small angle ET with FMN*
- *FMN = LMLN : the N associated with Infantile Onset Strabismus*

Convergence Null for Distance CND: Can mean IN or PAN
When it IS important to know whether it is IN or PAN

- Surgery other than BMR for CND, eg if the CND improves but doesn't cure the Abn Head Posture, and A-K surgery modified for CND is contemplated

- Acuity with Δ <20/40 in adolescent approaching driving test:

1. Baclofen may help with acuity in PAN
2. Gabapentin, memantine may help with acuity in IN

How often will you find CND?

- N=88 consecutive EMR's
- 29/88 = 1/3 have CND
- *True % CND > EMR-proven-N % CND*

Type of nystagmus	CND*	No CND *
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 with IN will later turn out to have PAN**

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

Usefulness of CND 1

- 1/3 or more pts have this
- Some have IN [80%], some PAN [20%]
- They should have a trial of **Δ** glasses in real life
- If CND is confirmed in real life then BMR is likely to also produce the same null, with:
- 1. no abn head posture
- 2. widened null zone
- 3. better acuity if sensory anatomy allows

Usefulness of CND 2

- If your pt has CND and this normalises Abn Head Posture and acuity is 20/40 or better:

- No recordings required

- Rx: Δ glasses or BMR

Case #2. 17 yr old
OCA, reduced acuity, poor N waveform

See how her 20/100 acuity improves to 20/40+

- Bothered by:
- 1. Appearance of wobbly eyes in primary position
- 2. Reduced acuity. Wants to do nursing [later: she didn't]
- 3. Intermittent oscillopsia

- 1,2,3: all fixed with Rx.

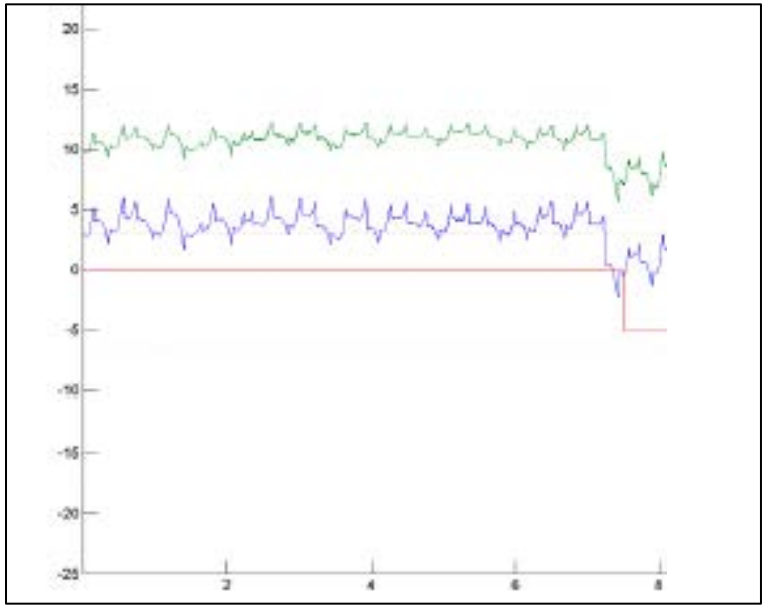
- PMH: Bipolar. Lexapro, Na Valproate

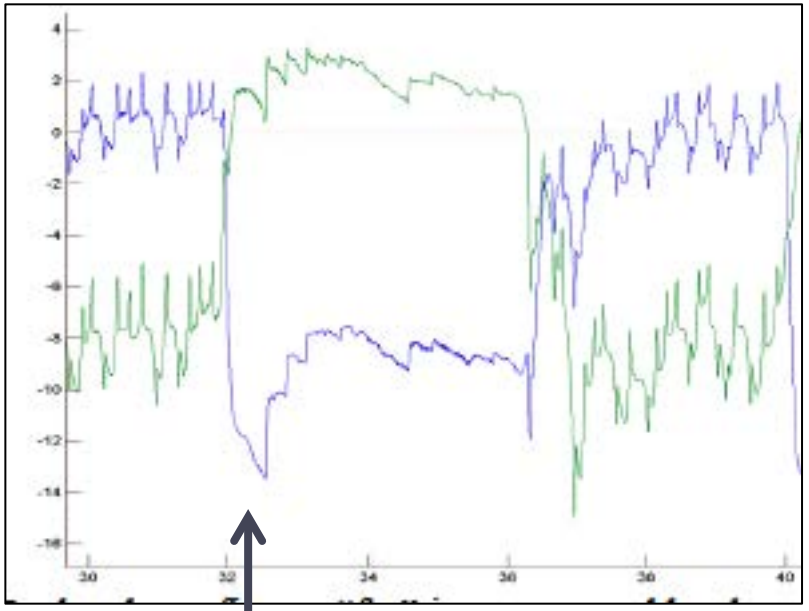
Findings

- **R 20/400** (-2-1.5x180) , **L 20/100** (-2.75)
- **BEO 20/100**
- OCA. TIDs. Normal disc size / morphology.
- Indirect: normal macular morphology
- Convergence null for near.
- L gaze: N to L. R gaze: N to R.
- Titmus ‘Fly’
- **DKWTD:**

CND: Add -1.5 DS & 7Δ BOOU: **BEO 20/80+**

Recordings: IN, Convergence null





Attempted Convergence

Foveation times mostly bad

Near fixation marked effect

Treatment plan

- 1. **CLs**: not successful
- 2. Gabapentin 2.7g/d improves acuity to **20/60+** ☺
- 3. Extra -1 & 7Δ BO OU : now **20/50+** ☺

- **As acuity improves, notices a new null for 1st time: 20° tip down**

Surgery

- BMR 3.5mm*
- LR tenotomy – resuture
- IO myectomy

Hertle ‘recipe’: distal myectomy 5mm from insertion

** BMR 3mm augmented for large globes*

Outcome

- **20/60- OU**
- **Both eyes open 20/50+**
- Intermittent small (<10°) tip up
- Fuses BSG* for distance, Fly for near
- Add Gabapentin: 20/40+ and passes driver's licence vision test 

*Bagolini Striated Glasses

Summary

- CND improves acuity*, both with Δ & Artificial Divergence Surgery
- Effect of Gabapentin is additive
- **Poor foveation times on EMR**: N may be contributing to poor acuity.
- CND improves the foveation time: this may improve acuity *

** to the extent that sensory anatomy allows*

Summary : Acuity – improving treatments 1*

- Surgery in IN

ANY surgery (BMR for CND, Anderson +/ – Kestenbaum, Tenotomy-Resuture) improves acuity in 25-50% of pts

- Surgery in PAN

BMR for CND, Tenotomy-Resuture, large recessions of all horizontal recti improves acuity in ?% of pts

** to the extent that sensory anatomy allows*

Summary : Acuity – improving treatments 2*

- Surgery in FMNS

Straightening the eyes converts N to Latent N

- Drugs in IN

Gabapentin, memantine

- Drugs in PAN

Baclofen

** to the extent that sensory anatomy allows*

Seminal paper 1987 : CN surgery often improves acuity

Australian and New Zealand Journal of Ophthalmology 1987; 15: 25-34

SURGICAL MANAGEMENT OF CONGENITAL NYSTAGMUS

J. NORTON TAYLOR FRACO, FRACS, FRCS
Ophthalmologist in Charge, Ocular Motility Clinic, The Royal Victorian Eye and Ear Hospital, Melbourne

K. JESSE DAppSc (Linc), DOBA
Research Orthoptist, Ocular Motility Clinic, The Royal Victorian Eye and Ear Hospital, Melbourne

TABLE 4
Nystagmus with Horizontal Abnormal Head Posture

	Improvement — Average of surgical results for all cases		
	Anderson	Kestenbaum	Anderson-Kestenbaum
AHP	70%	75%	75%
BV with AHP	1¼ lines	1¼ lines	1½ lines
BV in PP	2 lines	1½ lines	2¼ lines

AHP = abnormal head posture. BV = binocular vision. PP = primary position.

No recordings. Didn't know about PAN. Probably some FMN cases.

Cases : 3 siblings with reduced acuity and nystagmus N #3,4,5

- #3 F [oldest] 20/60
- Previous ET surgery, residual ET. SPA. N.

- #4 M 20/80
- XT. SPA. Face turn to L. N.

- #5 M [youngest] 20/100
- Oscillopsia. ET. N.

Cases #3,4,5

- What is unusual about this family?
  - Childhood Nystagmus - did NOT have typical IN office findings, hence EMR
  - Reduced acuity

Typical findings of IN

- *Jerk N*
- *R Gaze: N to R*
- *L Gaze: N to L*
- *Null zones – eccentric [can be >1] and convergence*

Cases #3,4,5

- EMR

  “All 3 young people have:
  - Strabismus
  - quite similar jerk nystagmus …. waveform of FMNS = Infantile-Onset-Strabismus – associated waveform

Cases #3,4,5

	F	M	M
BCVA OU	20/60 Low hyperopia + astigm	20/80 Low hyperopia + astigm	20/100 Low hyperopia + astigm
Allignment	ET	XT	ET
Colour vision	CUCVT OK. HRR no correct responses	CUCVT OK. HRR only #4 is correct	CUCVT & HRR tests OK.
Imaging and electrophysiology	Abnormal OCT, photopic ERG, pERG & FAF Partially preserved cone function on ERG 30Hz flicker good	Abnormal OCT, photopic ERG, pERG & FAF	Abnormal OCT, photopic ERG & FAF pERG at lower end of normal spectrum 30Hz flicker attenuated

Cases #3,4,5

- What is unusual about this family?
  - Achromatopsia – can ‘cause’ IN, FMNS or both
  - Incomplete achromatopsia, with variable expressivity between family members, or X-linked variant given the female sibling has residual cone function?
  - Achromatopsia - mutations in CNGB3 in 50% of patients with achromatopsia (AR)

Case #3,4,5

- Rx options
  - To convert FMN to Latent N by perfect alignment
  - Likely to improve acuity [*to the extent that sensory anatomy allows]*

Zubcov AA et al
Treatment of manifest latent nystagmus
Am J Ophthalmol 1990

Strabismus with IN/ PAN

- CND excluded [can't have strabismus]
- Overall, 38% have strabismus
- FMN: [nearly] all have strabismus

Type of nystagmus	Total [no CND]	Strabismus
IN	30	10 [30%]
PAN	12	6 [50%]

Case #6: Late diagnosis of PAN 44898

LM, dob June 2000, first seen age 5.

- 20/160, BEO, Either Eye. Near Lea 20/200 OU
- Face turn to L 20 deg
- M also describes tip down & tilt R: not seen by me
- RG: N to R. LG: N to L.
- Conv null for near & D
- TID's
- +3 DSOU. Later myopic astigmatism

#6: Late diagnosis of PAN 2$_{44898}$

- RG: 30 deg 20/200
- RG 20 deg 20/100
- PP 20/300
- LG < 20/400
- 7Δ BOOU: no face turn, but VA 20/120 [not as good as eccentric null]

#6: Late diagnosis of PAN 3$_{44898}$

- EMR: no PAN after 280 sec.
- IN with null 10 deg into R gaze

Surgery 7/07 for eccentric null, modified for vegence null

- RMR+7.5, RLR -8.5 [less for vergence damping], LMR-6, LLR+9.5
- Improved VA 20/80
- 7/07: Small FT to R
- 9/07: small FT to L
- M describes i/mitt tip down, L FT > R FT.

#6: Late diagnosis of PAN 4_{44898}

2017

- Myopic astig OU [-3-3, -2.5-2.5] 20/80
- No better with 7Δ BO OU
- Needs 20/40 to drive
- Repeat EMR: PAN
- Baclofen can improve foveation time in PAN & improve acuity to the extent that sensory anatomy allows
- Starts 5mgm Baclofen hs, now 40 mgm/d [3 doses]
- Building up…20/40-….not quite good enough [yet?]
- ?try CL's as well

What went ‘wrong’? How did I miss PAN?

2 main groups of PAN:

- 1. change in direction of N on recording because of change in position of gaze null
- 2. change in direction of N on recording without change in position of gaze null – will always miss this in the office
- 1A: classical IN recording with good video evidence of change in position of gaze null

1. Classical PAN: Change in direction of N on recording because of change in position of gaze null

- If eccentric null in phase A is 20 deg to L of primary, will have FT to R.
- **In primary, N to R**
- RG: to R. LG to L .
- If eccentric null shifts in phase B to 20 deg to R of primary, will have FT to L.
- **In primary, N to L**
- RG: to R. LG to L .

1. What went ‘wrong’? How did I miss PAN? Change in direction of N on recording because of change in position of gaze null

More likely to miss phase B in the office if:

- Very short eg A is 8 minutes, B is 45 sec
- B is at primary position
- B is close to A

1A: classical IN recording with good video evidence of change in position of gaze null

- The EMR didn't go for long enough or the pt didn't concentrate for long enough to show PAN on EMR
- Need some voluntary effort to maintain visual interest on a target for 10 minutes to determine whether s/he has PAN. Some kids won't do this.
- Some cases of apparent IN later turn out to be PAN

What went ‘wrong’? How did I miss PAN?

2. change in direction of recording **without change in position of gaze null**

- Sensory defect PAN [#17: BOA].

no sensory benefit in changing gaze position

Can only diagnose PAN here with EMR

What went 'wrong'? How did I miss PAN?

2. change in direction of recording without detectable change in position of gaze null

- PAN with strabismus
- sometimes a motor disadvantage to changing gaze position, or
- changes fixation to change gaze null – can look like FMNS, or
- fixation switch can be spontaneous, unrelated to N, and change the gaze position and the N.

Can only diagnose PAN with EMR

"..those patients who have Infantile Aperiodic PAN with strabismus were more likely to have a static head posture, even with a periodic rhythm detected on eye movement recordings."

- Hertle et alii Infantile APeriodic Alternating Nystagmus JPOS. 2009 Mar-Apr;46(2):93-103.

PAN with strab can have EMR-only PAN #16

- Age 4: OCA. EMR IN
- Age 16: EMR for PhD study on effect of stress in IN: PAN.
- Has small angle ET
- No clinical evidence of PAN
- This type of PAN prob not diagnosable on office exam

- Also #41. Large ET. No suggestion PAN on OE

Acknowledgements

Robert Reinecke

- ..whose nystagmus lab was the 1st I'd ever seen.
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- He makes me look like an expert, and helps my patients receive the best care.