CONGENITAL NYSTAGMUS WHEN TO RECORD HOW TO TREAT   2009

LIONEL KOWAL

Royal Victorian Eye and Ear Hospital

Center for Eye Research Australia

Melbourne, Australia

TYPES OF CONGENITAL NYSTAGMUS cN

- **c**N: any type of early onset N
- <u>Congenital N = CN</u>

aka Congenital Motor N, Congenital Sensory N, Idiopathic Infantile N *IIN*

Common subtype: Periodic Alternating N   PAN

- <u>LMLN</u>

Latent Manifest Latent Nystagmus aka FMNS = Fixation Maldevelopment N Syndrome.

Can be MLN or 'pure' LN.

Other: spasmusnutans, vestibular, …

Clinical Features

CN

1. L beat on L gaze
2. R beat on R gaze
3. Eccentric null
4. Convergence null
5. Latent component
6. Usu. Horizontal, can be torsional or vertical or all 3
7. Oscillopsia uncommon
8. Strabismus in some

LMLN

1. Fast beat to fixing eye
2. Adduction null
3. N on lateral gaze
4. Latent component
5. Usu horizontal, can be torsional or both
6. Oscillopsia uncommon
7. Strabismus in all

…all well known

Why does CN occur?   1

- Poor symmetric acuity @ a critical time of visual development

Sometimes there is continuing evidence of the initiating cause – eg bilateral optic n hypoplasia, symmetric terrible refraction, cone dystrophy, cataract OU, macular hypoplasia of albinism, …and sometimes there is no evidence of the initiating cause – macular hemorrhages, terrible refraction that got better, …

Why does CN occur?   2

- Genetic reasons – the N waveform itself can be inherited

- Abnormal CNS : Peri Ventricular Leukomalacia PVL

Many hypotheses / ?Multiple causes

- ? abnormal circuit between fixation and ocular stabilisation systems

- Abnormal proprioceptors in enthesis* [? cause, ?effect]

* where tendon inserts into sclera

Why does LMLN occur?

- Asymmetry in motor or sensory development @ a critical time

  Congenital strabismus, amblyopia, monocular cataract, PVL, unilateral optic n hypoplasia,…

Whatever caused it, cNwill then degrade acuity further.

In CN, the amount of further degradation is:

1. reliably estimated by mathematically dissecting the waveform for NAFX factor [esp. in Cleveland?]

2. estimated by assessing foveation time [the duration when N speed <5° / sec as it changes direction]

Acuity is *not* related to the *amplitude* or *frequency* of N

Other effects of CN:

- 2. Abnormal head posture

e.g. sees 6/12 in 15° RG, 6/24 in 30º RG, 6/18 in PP, 6/30 in 15º LG, 6/48 in 30º LG… will have FT to L ~ 15°

- 3. Reduced visual field

e.g. sees 6/12 in 15° RG, 6/24 in 30º RG, 6/18 in PP, 6/30 in 15º LG, 6/48 in 30º LG… will have FT to L ~ 15°, and effectively restricted field of best acuity

- 4. Strabismus

Principles of treatment of CN

- 1. Improve the waveform

- 2. Improve 2ary effects such as face turn

1. Improve the waveform

- Drugs – gabapentin, memantine

Fairly new. Often effective.

Medium – long term results?

- Contact lenses

SCL & HCL have an effect > optical effect

- BO Δ to induce convergence

- Surgery

- Are the effects of these sometimes additive?

- Prism glasses

- Surgery

Effects of treatment of CN

- Improved waveform may result in improved acuity limited by any associated pathology

- Expanded null zone = improved field of same or better acuity

- Improved face turn – improved appearance and improved field

Effects / Treatment of LMLN

- Reduced acuity : make perfectly straight and convert MLN to LN
- Face turn from adduction null: MR surgery
- Head tilt from intorsion null: torsional surgery

- Do we need Eye movement recordings EMR ?

Clinical Features

CN

1. L beat in L gaze / R beat in R gaze (either side of null zone)
2. Eccentric null
3. Convergence null (better reading VA)
4. Latent component (can be pseudo-latent)
5. Usu. horizontal, can be torsionalor vertical or all

LMLN

1. Nystagmus on lateral gaze (moving away from adduction null)
2. Fast beat to fixing eye
3. Adduction null – can cause face turn or head tilt
4. Latent component
5. Can be horizontal or torsional or both

CN c.f. LMLN

- CONVERGENCE NULL :
- Both can have this – different mechanisms
- Adduction null of LMLN can look like conv null of CN

- ECCENTRIC NULL :
- Both can have this – different mechanisms:
- LMLN: null in aDuction or Intorsion; if RE dominant, will have FT to R or head tilt to R
- CN: RE = LE

CN c.f. LMLN

- STRABISMUS :

  LMLN 100%

  CN ?30%

- VERTICAL / TORSIONAL COMPONENT:

- LMLN: T more than V

- CN: V more than T

CN c.f. LMLN : L beat on L gaze, R beat on R gaze [Alexander's Law]

- CN: N $\rightarrow$ L on L of null zone, N $\rightarrow$ R on R of null zone

- <u>LMLN :</u>

- LE is fixing & is in LG: BE have N $\rightarrow$ L

- RE is fixing & is in RG: BE have N $\rightarrow$ R

- LATENT COMPONENT:
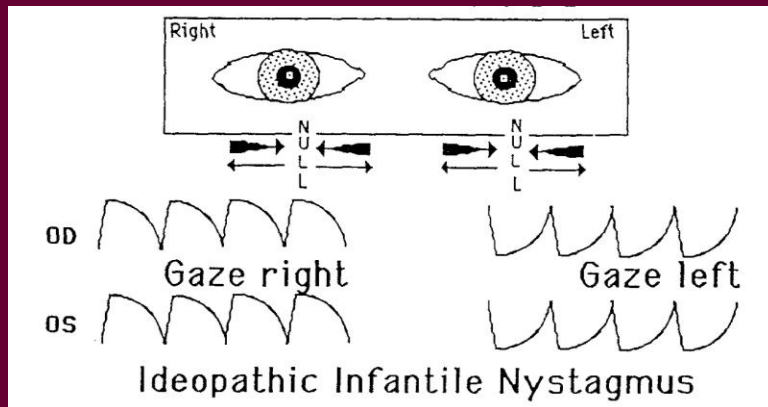
Both can have this – ? different mechanisms

'I AM WORSE THAN I USED TO BE'

- LMLN:
- Has lost binocularity and LN $\rightarrow$ MLN

- CN:
- new neurological lesion has caused deterioration,

or

- Stress – common as a temporary mechanism, rarely permanent
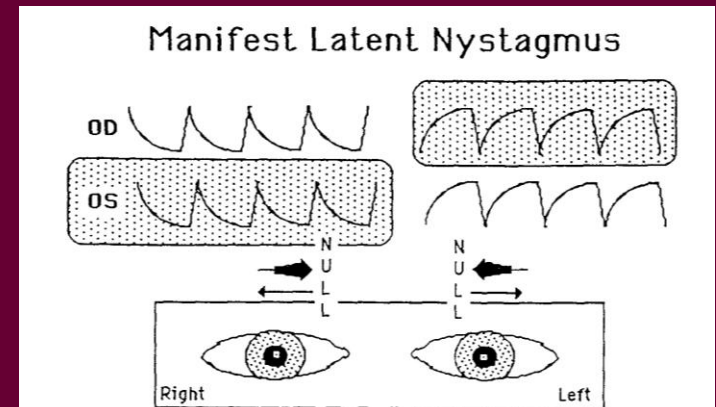
EMR features – usually diagnostic

CN

- Exponential increase in velocity in slow phase (unique)
- Fast phase brings eye back to foveation

LMLN

- Decreasing velocity in slow phase (GPN)
- Can be asymmetric in phase, amplitude or frequency

Clinical correlations : Dr Elaine Wong [Fellow]

- Chart Review of cN pts   n=84

- 60 with EMR

- 42 EMR & LK pre-EMR office diagnosis
  - 5 cases of PAN were excluded from analysis

Overview: office c.f. lab diagnosis

Clinical and EMR Correlation

Predictive Values of Clinical Features – CN

Predictive Values of Clinical Features – CN

Predictive Values of Clinical Features – LMLN

Predictive Values of Clinical Features – CN & LMLN

Conclusions :
Clinical and EMR Correlation

- Presence of convergence null – suggestive of CN
- Eccentric null and nystagmus to direction of gaze – less predictive
- No reliable clinical signs for LMLN

Do EMR!!!

No cardiologist would evaluate an arrhythmia without ECG

No neurologist would evaluate epilepsy without EEG

- Limitations exist! Can miss PAN!

- Gold standard in diagnosing types of cN

Does everyone with N need to be recorded?

- Not if you're absolutely certain about the diagnosis and have all the information you need for management

OR

- If you don't need to know e.g. insignificant face turn, good acuity,….

SURGERY IN cN : LMLN

- Straighten eyes perfectly
- ± 10 Δ not good enough: 0 Δ is needed
- Will convert MLN to LN
- Other surgeries for face turns and head tilts

SURGERY IN cN : CN

- Eccentric null

- Convergence null

- Null in primary position

- No definite null

SURGERY IN CN : Hertle's operation

- ANY surgery usually improves the waveform and may improve acuity even if main aim of surgery not accomplished eg residual face turn

- Why not try the most basic component of surgery – 'tenotomy – resuture' - and see if that helps the waveform?

- IT DOES

Eccentric null ≤ 20°   L face turn

Recess LMR 7mm & RLR 10 mm + tenotomy – resuture of the other two horizontal recti ~1 mm resection.

If a small duction/version paresis is not created then the head posture will usually return.

Eccentric null ≥ 25°   L face turn

Recess LMR 7mm , RLR 10 mm recess

Resect other 2 recti for total 17mm per eye.

If a small duction/version paresis is not created then the head posture will usually return.

- Prism adapt [no Fresnel] for max BOΔ

- BMR *x* mm based on max BO Δ + tenotomy – resuture the lateral recti

- Tenotomy – resuture the horizontal recti

SURGERY IN cN : PAN

- Look for convergence null

Prism adapt view BMR

or

- Tenotomy – resuture horizontal recti

SURGERY IN cN : warnings

- Albinism patients: have positive angle kappa & look divergent when they are straight

THE RECENT PAST AND THE IMMINENT FUTURE

- Increasing use of EMR to study the effects of different treatments and their combinations on our patients with cN will help us understand their condition better and plan more effective treatments

THANK YOU TO THOSE WHO TAUGHT & STILL TEACH ME ABOUT NYSTAGMUS: DRS. REINECKE, SPIELMAN, ABEL, DELLOSSO & HERTLE

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Lakota

Copper