Answers to Audience Questions - WSPOS World Wide Webinars

WWW 24 – Season 2 – Infantile Nystagmus Syndrome - New Concepts In Diagnosis And Treatment


Larry Abel


Richard Hertle


Lionel Kowal


Helen Kuht


Shashikant Shetty


Mervyn Thomas


Dongsheng Yang

Richard Hertle (RH), Dongsheng Yang (DY), Larry Abel (LA), Helen Kuht (HK), Mervyn Thomas (MT), Lionel Kowal

1. Could the panel please list out any tricks for the differential diagnosis of infantile nystagmus syndrome and fusion maldevelopment nystagmus?

   RH: No trick necessary, associated infantile strabismus syndrome with “Latent” component to the nystagmus is more likely to be FMNS

   LA: If there is no strabismus, FMNS is extremely unlikely. If there is strabismus but nystagmus direction doesn’t change with occlusion, then it’s INS. If it does change with occlusion, then recording will tell you, as the waveforms are different between INS and FMNS.

   LK: Sometimes can be distinguished confidently:
   FMN always has strabismus, IN often doesn’t
   FMN always has smooth pursuit asymmetry, IN doesn’t
   FMN sometimes has torsional component on slit lamp exam, rare with IN
   Can be easily confused:
   FMN is always fast phase to fixing eye, IN can looks like this too esp if there is strabismus
   IN usually has a convergence null, FMN can seem to have this too (preference for fixation in aDduction)

   SS. In FMNS associated with esotropia common inpatients with DVD.Usually has ADDUCTION NULL IN FMNS .INS can have latent component and fixation will be in abduction.eye movement recordings will differentiate

2. Are there any comparable companies to GeneDx in Europe?

   HK: We are aware of the companies Blueprint Genetics and Asper Biogene, which are comparable to GeneDx. However, in Leicester we have not used either of these companies.

3. Are there any tips for OCT in kids with nystagmus?

   RH: Hand held OCT, dedicated and enthusiastic technician.

   MT: If you have a hand-held OCT, if possible, try to have a toy or a video (on a phone / iPad etc) available to keep the child’s interest and fixation whilst capturing the image. If the child is young, getting the parent to help stabilise their head can help too. If a null region has been identified, try taking the OCT with the child using their null region.

4. Do you send your patients for visual stimulation?

   DY: Yes, I do. However, it is very hard to get improvement if nystagmus is intense. I usually estimate visual acuity for patients using a software called automated nystagmus acuity function (ANAF published in BJO) and compare it with real visual acuity in that patient. If ANAF is better than the real visual acuity and there are no retinal disorders, I let

patients do visual stimulation. But I usually have to do surgery to reduce nystagmus first, and then do visual stimulation. I am running a special program called dark exposure research project that leads to fast vision improvement in many patients (some adult patients) after their nystagmus surgeries.

5. Do you use the opposing base prisms to reduce the anomalous head posture?

   RH: No, e.g., for a 20 degree face turn at least 10 Prism Diopters are needed in each spectacle lens and Fresnell Prisms degrade acuity.

   DY: No, I do not use the opposing base prisms to reduce the anomalous head posture. However, if the opposing base prisms can reduce nystagmus, they may help to reduce anomalous head posture.

   LA: Base-out prisms are used when INS damps with near fixation rather than in lateral gaze. If used in someone with accommodation, you need to add -1sph correction to account for the convergence-induced accommodation.

   LK: If a large horizontal or vertical head posture is dramatically reduced by 7^ BO OU [total 14^] = convergence null for distance fixation, this usually indicates IN or CAPAN

   SS we can try prisms in patients who have constant face turn less then 10 deg.patients who have nystagmus blockade in convergence then base out prisms can be tried with -1d lens

6. Do you send your patients to neurologist? Do you prescribe patches for children with nystagmus?
   RH: WRT Do you send your patients to neurologist?
   Only if there are associated systemic neuro signs or symptoms
   WRT Do you prescribe patches for children with nystagmus?
   Yes

   DY: I usually do eye movement recording first. If I see linear and pendular waveforms, I will send the patient to get MRI.
   I do not use patches for children with nystagmus because nystagmus increases with monocular view. I use neutral density filter to reduce the vision in the better eyes. In this way the increase in nystagmus is minimized and binocular visual function（stereopsis）can be improved.

   LK: WRT Do you send your patients to neurologist?
   If there is a hint that the nystagmus is not IN or FMN, or if the parents request it. This often generates an MRI; sometimes this shows PVL.
   WRT - Do you prescribe patches for children with nystagmus?
   If there is amblyopia

   SS If onset of nystagmus then MRI and referral if there is glioma.If there is saccadic oscillations or intrusions then to refer.patching is required if amblyopia is detected

7. What should we consider when planning surgery in cases where we cannot distinguish FMNS and INS for abnormal head position and strabismus?

   RH: See Attached article, which is part of an abundant literature on surgery algorithms for nystagmus (search pubmed)

   LK: Don’t do surgery unless you can confidently make the patient better.
   If the patient has FMN, straightening the patient will make the patient better.
   If you do not really know what is going on, you should not do surgery.

   SS CLINICALLY DIFF FMNS AND INS IS IMPORTANT.I fyou are not sure then you need to oberve

8. At what age do you recommend surgery?

   RH: 10-14 months (when child is on their feet, unless infantile strabismus accompanies diagnosis then in the first 6 mos of life)

DY: Usually, I recommend surgery when a kid is around 1 year old and the nystagmus interrupts vision development.

LK: When I can confidently improve the patient

SS. when I am confident when there is no PAN,SILENT NULL,DOUBLE NULL,PREFERABLY AFTER 4YRS OF AGE

9. How do you modify the Kestenbaum when one eye is esotropic? And what is your post amblyopia treatment?

   RH: WRT How do you modify the Kestenbaum when one eye is esotropic?
   See attached article
   WRT And what is your post amblyopia treatment?
   I do not treat amblyopia different in a patient with nystagmus.

   LK: WRT How do you modify the Kestenbaum when one eye is esotropic?
   Ask someone with experience for a surgical recipe for your specific patient
   WRT And what is your post amblyopia treatment?
   The usual amblyopia treatment

   SS FIRST ESTIMATE WHETER ESOTROPIC EYE IS THE FIXING EYE AND DETERMINE YOUR SURGERY

10. Do you have any experience with BOTOX for nystagmus for treating changing null points or in multiplanar head postures? Does adding BOTOX to surgery not touch too many muscles?

    RH: WRT Do you have any experience with BOTOX for nystagmus for treating changing null points or in multiplanar head postures?
    Yes
    WRT Does adding BOTOX to surgery not touch too many muscles?
    Botox is a temporizing treatment in acquired nystagmus and ineffective for infantile nystagmus.

    LK: Botox is not a reliable treatment

11. Dr. Yang: Do you do res and rec on vertical recti? What about the numbers ? Like Scott's procedure?

    DY: Yes, I do res and rec on vertical recti. The resection is about 5mm and recession is no more than 14 mm depending on axial length. Usually, res + rec on the vertical muscle has better effects in reducing nystagmus than on horizontal muscle. The surgery does not cause horizontal deviation.
    I do not think my procedure is the same as Scott's procedure. I do resection and recession on the same muscle to make sure that the muscle is recessed far enough to the back of equator.

12. How can we take visual acuity in all gazes?

    RH: Reposition the head and check acuity in the new head position(s)

    LK: The most important gaze positions for acuity are forced primary position, preferred head position, and the opposite- to- preferred

    SS PREFERABLY 9 GAZES 30 DEG IN ALL GAZES