LET'S MAKE NYSTAGMUS GREAT AGAIN: A VIDEO TUTORIAL OF WIGGLES AN OPHTHALMOLOGIST MAY SEE



JERK nystagmus



PENDULAR nystagmus

Sally Letson Symposium, Ottawa 2018

davidsamuelzee@gmail.com

BEFORE WE START:

- Is NYSTAGMUS complex?
 - YES, highly complex, and it can be OVERWHELMING
- But can we extract from the anatomy and physiology easy to use, easy to remember, practical principles for our clinics?
 - YES, WITHOUT A DOUBT!
- Do we need fancy equipment and detailed quantitative analysis to evaluate nystagmus?
 - ABSOLUTELY NOT! The focused history and careful, ordered bedside exam usually points us to the diagnosis, and always directs us down the correct path for management. (Testing of course helps to confirm our diagnoses, and provides data for research BUT)
- HANG IN THERE! Rather than thinking that nystagmus takes you to Auguste Rodin's "Gates of Hell", you will receive "The Kiss" from your patients at the end of the day.





Auguste Rodin

A Road Map for Today

- Historical perspectives
- Organizing one's thoughts about nystagmus
- A flow chart for diagnosis
- The wave-form analysis
- Key anatomy
- Major types of jerk nystagmus: Brain stem and cerebellum
- Major types of pendular nystagmus: multiple sclerosis and ocular palatal tremor syndromes
- A few unusual types of nystagmus to recognize

Why do we need eye movements?

Sharp, detailed vision is possible only at the fovea, the small area at the center of the retina, and when images are held steady there.





How to think about nystagmus (without nightmares) (νυσταγμός, drowsiness, nodding, doze)

Eduard Hitzig (1871) first used the word *nystagmus* to describe the effects of galvanic stimulation on the mastoids behind the ears which produced a nystagmus with slow and quick phases.

The eye movements were likened to a fisherman's float drifting slowly in the water and then being snatched back



Life of Pi

Tatler and Wade, 2003



A NOTE ON OSCILLOPSIA (symptom):

- Illusory movement of the visual world: Usually with a back and forth, jerk, or wiggle sense
- Latin oscillo, to swing, + the Greek opsis, vision
- If with head <u>still</u>, usually due to a spontaneous nystagmus or other uncalled for eye movement
- If with head <u>moving</u>, usually related to an abnormality in the amplitude or direction of compensatory slow phases of the vestibulo-ocular reflex.

Ways to organize one's thoughts about nystagmus

- As a disorder of mechanisms that hold gaze steady
 - Vestibular balance
 - Gaze-holding networks (brainstem and cerebellum), neural integrators
 - Medial vestibular nuclei (MVN) /nucleus prepositus hypoglossi (NPH) in medulla (HORIZONTAL)
 - Interstitial nucleus of Cajal in midbrain (VERTICAL)
 - Cerebellar flocculus and paraflocculus
 - Mechanisms that keep eye movements <u>calibrated</u> (maladaptation and sensory deprivation (nystagmus of the blind))
 - <u>lon channel kinetics</u> that control membrane stability in the gaze holding networks (saccadic oscillations)
 - <u>Developmental abnormalities (motor and sensory, infantile (congenital) nystagmus, latent</u> nystagmus)
- Waveforms and pattern recognition (e.g., see-saw)

A Flow Chart to Aid Classification of Nystagmus

Is fixation impaired because of a *slow drift*, or an *intrusive saccade*, away from the target?



From Leigh and Zee, NEM, 5th edition, 2015

Waveforms of Nystagmus



POSITION

ЕΥЕ

Jerk or linear (vestibular)

Velocity decreasing

- acquired, gaze-evoked
- congenital, latent

Velocity increasing

- horizontal, usually congenital
- vertical, usually acquired

Pendular

(acquired or congenital)

KEY ANATOMY: Horizontal gaze-holding networks in medulla



KEY ANATOMY: Vertical gaze-holding networks in the midbrain



ANATOMY: Cerebellar flocculus and paraflocculus (tonsils)



Paraflocculus (Tonsils)

Flocculus

Velocity-increasing slow phase



- Velocity-increasing slow phases imply brainstem gaze-holding network (neural integrator) is <u>unstable</u>.
- DB Nystagmus will intensify in up-gaze or UB Nystagmus in down-gaze (anti-Alexander's Law) (recall Alexander's law; nystagmus intensity increases when looking in direction of the quick phase (e.g., peripheral vestibular nystagmus)).
- Velocity increasing slow phases in <u>acquired</u> lower-brainstem (e.g., deficiency of thiamine (b1) in Wernicke's encephalopathy) or in cerebellar floccular lesions.

Wernicke's Disease (lesions in the medial vestibular nuclei (MVN), part of the horizontal gaze-holding network AND mediates horizontal VOR)



Wernicke's Disease (lesions in the MVN and NPH): Clinical Points

- Wernicke's syndrome can present with ophthalmoplegia, nystagmus of virtually any type (horizontal, vertical), gaze palsies and internuclear ophthalmoplegia
- Upbeat changing with convergence to downbeat is common (caudal perihypoglossal nuclei (Roller and intercalatus))
- BILATERAL HORIZONTAL VESTIBULAR LOSS IS OFTEN ASSOCIATED.
- May show anti-Alexander's law (nystagmus more intense when looking in direction of slow phase)
- B1 deficiency occurs in the setting of MALNUTRITION due to alcoholism, chemotherapy, eating disorders (hyperemesis gravidarum, bullemia, anorexia nervosa), post gastric diversion, social isolation)
- Treat with IV 500mg B1

Upbeat nystagmus: effect of convergence



Clinical points

- This same pattern of nystagmus often occurs in acute Wernicke's syndrome.
- Spontaneous horizontal nystagmus that is suppressed with convergence is usually CONGENITAL (INFANTILE) NYSTAGMUS
- Spontaneous downbeat nystagmus that is enhanced with convergence or spontaneous upbeat nystagmus that is damped or converts to downbeat nystagmus with convergence is usually a CENTRAL VESTIBULAR NYSTAGMUS



Guillain-Mollaret Triangle (Dentate nucleus, Red nucleus, Inferior Olive)



PATIENTS WITH OCULOPALATAL TREMOR SYNDROME





Inferior olivary hypertrophy with increased signal in olive





Iron due to old hemorrhage in AVM

Whipple's Disease

- Cognitive Disorder
- Myoclonus
- Supranuclear ophthalmoplegia (vertical)
- Convergence pendular nystagmus
- Myorhythmia of masticatory muscles (and limbs)
- Uveitis
- Systemic (GI, joints, rash)
- Abnormal CSF
- Abnormal MRI
- PCR positive (blood and csf)

Congenital Nystagmus



Seesaw nystagmus

- Congenital and acquired forms (pendular)
- Achiasmatic Belgian sheep dogs and achiasmatic humans (also septo-optic dysplasia
- Acquired forms associated with third ventricle tumours (bitemporal hemianopia) and upper midbrain lesions
- Occasionally associated with palatal tremor
- Hemi-seesaw variant (jerk nystagmus): rostral midbrain-caudal thalamus

Nobody's Fool

