

# NYSTAGMUS MADE SIMPLE

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## I. INTRODUCTION

Nystagmus may be defined as a to and fro movement of the eyes, where the first movement is a slow movement. This is to be differentiated from the to and fro movements of saccadic intrusions where both the first and second movement are relatively fast saccades. The presence of nystagmus in a patient can be an important diagnostic clue that can lead to the proper localization of disease within the central vestibular pathways, the peripheral vestibular system or the afferent visual pathways. Various forms of congenital nystagmus (often termed infantile nystagmus) should be differentiated from acquired forms. The qualities of nystagmus including the type of movement (jerk or pendular), direction (up, down, horizontal, torsional), inter-eye variation, provocations and associated neurological symptoms and signs, should be elicited to best understand the type of nystagmus, proper localization and treatment. Treatment of the underlying condition may be supplemented with both tried and true and emerging medical and surgical therapies aimed at ameliorating the nystagmus and associated oscillopsia. This syllabus will briefly review the neuro-anatomical pathways crucial to the understanding of nystagmus, and then will summarize its major forms, their localization, associated conditions and treatment. Waveforms and characteristics of nystagmus have already been reviewed.

## II. NEUROANATOMICAL UNDERPINNINGS

- **Brainstem control of saccades**
  - The initiation of fast eye movements meant to direct the a target onto the fovea (saccades) occurs when the ocular motor neurons (of cranial nerves III, IV and VI) fire at a high rate known as a pulse, which encodes the velocity of the movement. The maintenance of the achieved eye position in eccentric gaze is governed by a lower frequency firing rate of those neurons know as a step.
  - The discharge frequency of the motor neurons during the pulse determines the speed of the saccade. For horizontal saccades, this pulse is controlled by the parapontine reticular formation (PPRF), whose premotor excitatory burst neurons stimulate the neurons of the abducens nucleus. For vertical saccades, the premotor excitatory neurons lie in the rostral interstitial nucleus of the medial longitudinal fasciculus. (riMLF)
  - The discharge frequency of the motor neurons during the step determines the amplitude of the saccade, i.e. the position in which the eye is maintained. This is calculated by performing an integral of the velocity of the saccade (pulse) over time, to obtain the distance. This calculation is performed by *the neural integrator*, which for vertical eye movements is the interstitial nucleus of Cajal in the midbrain, and for horizontal movements is composed of the medial vestibular nucleus and nucleus prepositus hypoglossi (NPH), enhanced by a positive feedback loop provided by the flocculus.<sup>1</sup>
- **Vestibular system**
  - The vestibular system is responsible for compensatory movement of the eyes in response to body and head movements, in order to maintain foveation. Its sensory organ is the labyrinth in the inner ear, which is a series of motion detectors in which the movement of endolymph stimulates hair cells to encode the speed and direction of self-motion.
  - Rotational movements are detected by the three semicircular canals. The horizontal, posterior and anterior semicircular canals.
    - i. The horizontal semicircular canal (HSCC) is stimulated by ipsilateral lateral head movements. Its signal travels along the vestibular branch of the 8<sup>th</sup> nerve to the

- ipsilateral MVN, from which a signal travels to the contralateral abducens nucleus, resulting in contralateral compensatory gaze.
  - ii. The posterior semicircular canal (PSCC) is stimulated by head movements backwards and clockwise (from the patient's perspective), and result in compensatory downward and counterclockwise eye movements. The signal is again mediated by the MVN, this time with output to the riMLF, INC and ocular motor nuclei governing vertical and torsional eye movements.
  - iii. The anterior semicircular canal (ASCC) is stimulated by head movements forwards and clockwise (from the patient's perspective), and result in compensatory upward and counterclockwise eye movements. Like the PSCC, the signal is mediated by the MVN, with output to the riMLF, INC and ocular motor nuclei governing vertical and torsional eye movements.
- Linear movements are detected by the utricle and saccule.
  - i. The saccule detects vertical linear movement of the body. Compensatory eye movements are governed by the vertical gaze centers of the midbrain.
  - ii. The utricle detects horizontal linear movements of the body. Compensatory eye movements are governed by the horizontal gaze centers (Abducens nucleus and targets)
  - iii. The utricle also detects body tilt and governs a response known as the ocular tilt reaction (opposite torsion of the head and both eyes, and infraduction of the contralateral and supraduction of the ipsilateral eye). The responsible pathway courses through the vestibular nerve, MVN, ascends and decussates in the pons and travels up the medial longitudinal fasciculus to synapse in the contralateral INC, riMLF, third and fourth nuclei. When this system is disrupted, the resulting imbalance between one side and the other results in a pathological ocular tilt reaction, with an ipsilateral hypotropia and ocular torsion. This condition which results in vertical diplopia, is known as a skew.

### III. VESTIBULAR NYSTAGMUS

- **Types:** Nystagmus may result from disruption of either the central vestibular system (brainstem or cerebellum) or peripheral vestibular system (labyrinth or vestibular nerve). Differentiating the two in cases of acute vertigo with nystagmus is a crucial step in determining the differential diagnosis and thus the emergent nature of the work up, i.e. whether or not an urgent MRI is required. Although both central and peripheral are examples of jerk nystagmus, the patterns of eye movement and associated features may be used to differentiate the two types. The following features suggest central vestibular nystagmus: gaze-changing, pure upbeat, pure downbeat, pure torsional.
- **Central vestibular nystagmus**
  - i. Patterns
    1. Direction changing nystagmus – upbeat in upgaze, right-beat in right gaze, left beat in left gaze, with or without downbeat in downgaze
    2. Pure upbeat nystagmus (UBN) –poorly localizing but tends to reflect disease of the midbrain or paramedian medulla.
    3. Pure downbeat nystagmus (DBN) – localizes to the cervicomedullary junction (CMJ). Disruption of a normal, flocculus-derived inhibition of tone from the ASCC may be contributory. This disinhibition of the ASCC results in an upward drift, followed by a corrective downward saccade, together comprising the two components of DBN.
    4. Pure torsional nystagmus – localizes to medulla or INC.
  - ii. Causes: Etiologies include brainstem tumors, stroke, hemorrhage or demyelination as well as certain medications such as anti-epileptic drugs. DBN, in particular, may result from Chiari malformations with the resultant compression of the CMJ.
  - iii. Treatment –
    1. Treatment aimed at the underlying cause is often effective; for example, steroids for demyelination may eradicate the nystagmus.

2. 4 aminopyridine (4-AP) is a non-selective blocker of voltage-gated potassium channels, which increases Purkinje cell excitability in the flocculus, thus restoring the normal inhibition of vertical drift resulting in upbeat and downbeat nystagmus.<sup>2</sup>
- **Peripheral vestibular nystagmus**
    - I. Features – Peripheral vestibular nystagmus is ameliorated by fixation. It is often accompanied by tinnitus and/or hearing loss when the cochlea is involved. Nausea and vomiting are often associated and the vertigo is typically intense.
    - II. Types
      - Complete unilateral injury –
        - Pattern – A combined horizontal (with a contralateral jerk) and torsional jerk nystagmus (with the top of the eye jerking away from the affected side). A loss of horizontal tone leads to ipsilateral drift and therefore a contralateral jerk nystagmus. Loss of the ASCC and PSCC sensitivity leads to a torsional drift so that the top of the eye moves in the direction of the affected side and the resultant jerk nystagmus moves the top of the eye towards the other side. The vertical effects of loss of the ASCC and PSCC cancel each other out.
        - Etiology – vestibular nerve injury, injection of gentamicin
      - Horizontal canal dysfunction –
        - Pattern – contralateral jerk
        - Etiology – labyrinthitis, vestibular schwannoma
      - Benign paroxysmal positional vertigo (BPPV) – otoliths are calcium carbonate crystals that weight down the hair cells within the utricle during linear movement along the XY plane or during head tilt to weigh them down and allow continuous recognition of head tilt or linear movement. In BPPV, some of these crystals detach, either due to trauma or age, and fall into one of the semicircular canals, typically the PSCC. Episodes are brief (seconds to minutes) and provoked by head movements.
        - Posterior semicircular canal BPPV (PSCC BPPV) –
          - Pattern - results in a combined upbeat and torsional nystagmus (the eye jerks up and top of the eye jerks torsionally towards the affected ear).
          - Diagnosis – Diagnosis of PSCC BPPV can be confirmed by performing the Dix Hall Pike maneuver (DHP), in which the head is turned towards the suspected affected side and the patient moves from a sitting up position to supine, with the turned head extended beyond the back of the table. The head turn puts the PSCC in the plane of the back. Vertigo and the combined upbeat-torsional nystagmus typically ensues.
          - Treatment –
            - Epley maneuver<sup>3</sup> – the patient is sitting up and their head is turned towards the affected side. They are quickly lied back just as in the DHP. After 30 seconds, the head is turned straight up, then to the unaffected side, then turned further towards the ground (on the unaffected side) and then they are sat up. The otoliths are hopefully thread out of the PSCC. IN double blind sham-controlled trials, patients undergoing the Epley maneuver were between 22-37 times as likely to experience resolution of BPPV.<sup>4</sup>
            - Sémont maneuver<sup>5</sup> – the patient is sat up and then the whole body and head are moved laterally, away from the affected ear, until the patient is lying on their side.
        - Horizontal semicircular canal BPPV (HSCC BPPV) – the otoliths fall into the HSCC in about 5% of patients.

- Pattern – HSCC is overactive so this results in a horizontal jerk nystagmus towards the affected ear.
- Diagnosis – The DHP may be negative, in which case it is best diagnosed with the supine roll test (SRT or Pagnini-McClure test<sup>6</sup>) where the patient is lied straight back. The head is then turned to the right and the patient is observed for nystagmus, and the same is done to the left. Because the HSCC is U shaped and the ampulla with the hair cells is located at one end, the effect of turning the head is different depending on whether the otoliths are in the first arm of the U (posterior) or second arm of the U (anterior).
  - *Geotropic HSCC BPPV* – Geotropic means that during the SRT, the nystagmus always beats towards the ground. (to the right when the head is turned to the right, and to the left when the head is turned to the left) The side to which the head turned which produces a more intense nystagmus is the affected side. Geotropic nystagmus happens when the otoliths fall from the utricle into the posterior (first) arm of the curved PSCC. This is because the ipsilateral head turn pulls the otoliths farther into the canal, stimulating the cupula and leading to a contralateral drift of the eyes, and a secondary ipsilateral jerk component. The contralateral head turn pulls the otoliths away from the ampulla of the affected top ear, leading to a relative under-action on that side. Thus there is a drift towards the affected ear and a secondary contralateral jerk component so that again, the eyes jerk towards the ground.
  - *Apogeotropic HSCC BPPV* – Apogeotropic means that during the SRT, the nystagmus jerks away from the ground, i.e. rightward in left head turn and leftward in right head turn. This occurs in the rare case of the otoliths making their way to the anterior (second) arm of the HSCC. Now an ipsilateral head turn will pull the otoliths away from the ampulla, inhibiting the HSCC, producing a drift towards that side and a contralateral jerk. When the head is turned towards the unaffected ear, the otoliths now fall towards the ampulla and the eyes drift away from the affected ear with a secondary jerk back towards the affected ear, i.e. away from the ground. In Apogeotropic HSCC BPPV, the side with the *less* intense nystagmus is the affected side.
- Treatment –
  - Barbeque (Lempert) roll maneuver<sup>7</sup> – start the patient lying down with the head turned to the affected side. The head is then turned 90 degrees to face up, then to the unaffected side, then turned to face prone and then finally the original position. Each position is held for the duration of any observed nystagmus. The otoliths are hopefully threaded out of the HSCC.
  - Gufoni maneuver<sup>8</sup> for geotropic –the patient is sitting up and is quickly moved so that they are lying on their unaffected side. After 2 minutes, their head is turned 45<sup>0</sup> towards the ground, kept there for 2 minutes and then they are sat up. This may sometimes convert the patient to a PSCC BPPV if the otoliths fall into the PSCC.

- Gufoni for Apogeotropic - the patient is sitting up and is quickly moved so that they are lying on their affected side. After 2 minutes, their head is turned 45° away from the ground, kept there for 2 minutes and then they are sat up.
- Anterior semicircular canal BPPV (ASCC BPPV) –rare (3% of BPPV)
  - Pattern – results in a downbeat nystagmus, often with a subtle torsional component.
  - Treatment –
    - Reverse Epley maneuver – Start on the unaffected side
    - Yacovino maneuver – patient lies supine with head hanging 30 degrees in extension; then the head is moved 30 degrees in flexion, and finally the patient sits with the head 30 degrees forward.<sup>9</sup>

III. ACQUIRED PENDULAR NYSTAGMUS (APN) - APN is typically a constant nystagmus which produces significant oscillopsia.

▪ **Acquired Pendular Nystagmus, isolated**

- Pattern -
  - May have horizontal, vertical and torsional components
  - Horizontal and vertical may be in phase producing diagonal nystagmus
  - Horizontal and vertical may be out of phase producing elliptical nystagmus.
  - APN may be disconjugate so that the directions are different in each eye
  - APN may be dissociated so that the amplitude may be different in each eye.
  - Momentarily suppressed by saccades.
  - May be suppressed by convergence
- Localization – APN is felt to be secondary to involvement of the neural integrators (MVN, NPH and flocculus for horizontal and INC for vertical and torsional).<sup>10</sup>
- Etiologies –
  - Classically results from demyelination, i.e. multiple sclerosis
  - Leukodystrophy - Pelizaeus-Merzbacher
  - Toluene abuse<sup>11</sup>
  - Spinocerebellar ataxia
  - Whipple's disease
  - May be mimicked by unrecognized congenital pendular nystagmus
- Treatments –
  - Medical treatments are aimed at antagonizing NMDA receptors and restoring control of the neural integrator.<sup>12</sup>
  - Memantine
  - Gabapentin
  - Baclofen –GABA<sub>B</sub> agonist; only works for vertical APN.
  - Base out prisms – elicits convergence
  - Botulin toxin injections into the extraocular muscles.
- **Oculopalatal Myoclonus (OPM) – a type of APN**
  - Pattern – A vertical or vertical-torsional pendular nystagmus of 1-3 Hz is coupled with a vertical tremor of the palate with the same frequency. There may be simultaneous movements of facial muscles. APN accentuated by closing the eyes and persists during REM sleep. The palatal movements are asymptomatic so the practitioner must remember to look for it.

- Etiology – Disruption of the triangle of Mollaret, which describes a network connecting the red nucleus, inferior olive, and dentate nucleus of the cerebellum.<sup>13</sup> Often a pontine hemorrhage or cavernous malformation is responsible. Loss of inhibition of the inferior olive leads to olivary hypertrophy. The neurons of the inferior olive, which can act as pacemaker cells, begin to form connexin junctions and become electrochemically coupled, resulting in a periodic coordinated firing.
- Treatment
  - Variable results with medical treatments such as phenytoin and valproic acid.
  - Disinsertion of the inferior and superior recti of both eyes may ameliorate the nystagmus and oscillopsia.<sup>14</sup>

#### IV. Other Nystagmus syndromes with localizing value.

##### ▪ **Periodic Alternating Nystagmus (PAN)**

- Pattern – A non-positional, horizontal jerk nystagmus which switches directions spontaneously, with a nystagmus-free interval between right and left beating nystagmus. The period is approximately 2 minutes.
- Localization – typically reflects cerebellar disease, particularly the nodulus and uvula. These structures contribute to the inhibition of the “velocity storage” of the medial vestibular nucleus (MVN) for the horizontal VOR. That is, the degree of compensatory contralateral eye movement per degree of head movement is controlled. Upon disruption of this pathway, horizontal drift ensues with secondary contralateral saccades leading to jerk nystagmus. This inhibition utilizes GABA<sub>B</sub> receptors.
- Treatment – Exquisitely responsive to baclofen, which enhances GABAergic tone at the MVN.

##### ▪ **See Saw Nystagmus**

- Pattern – A pendular nystagmus where one eye elevates and intorts while the other depresses and extorts followed by the opposite movements. The pattern is best observed while looking at the bridge of the patient’s nose.
- Localization – suprasellar region, often in association with chiasmal compression and bitemporal hemianopia.
- Etiology –
  - Suprasellar masses such as craniopharyngioma
  - Brainstem strokes
  - In association with loss of crossing fibers in the optic chiasm.<sup>15</sup>

##### ▪ **Hemi See Saw Nystagmus**

- Pattern - A jerk form of See Saw nystagmus.
- Localization – Junction of midbrain and thalamus. Likely due to disruption of the INC leading to a pathological ocular tilt reaction, with the riMLF producing a corrective torsional and vertical saccade. Alternatively, it may localize to the lateral medulla.<sup>16</sup>

#### V. PEDIATRIC NYSTAGMUS SYNDROMES

##### ▪ **Infantile Nystagmus Syndrome (INS)<sup>17</sup>**

- Epidemiology – INS typically emerges between 2-6 months of age. and affects approximately 1/1000 children, affecting males more frequently.
- Pattern – Horizontal in all fields of gaze (uniplanar)
  - INS is binocular and conjugate. It is typically pendular in primary gaze and jerk in lateral gaze. At an early age, pendular waveforms predominate.

- A null point or null zone is often present and describes a field of gaze in which the amplitude of the nystagmus is minimized. This will often result in the patient taking on a head turn to allow them to be in at the null point while looking straight ahead.
- Suppressed by convergence
- The slow phase of the jerk components of INS are typically accelerating, which differentiates them from those of most forms of vestibular nystagmus. This is only measurable using nystagmography (magnetic search coils, for example).
- The direction of the optokinetic nystagmus (OKN) is characteristically reversed.
- Etiology –
  - INS is felt to result from an instability of visual fixation, either due to a problem with motor system (“motor INS”) or secondary to poor vision (“sensory INS”). In either case, the eyes drift away from the intended target of gaze, which provokes either a corrective fast movement (to generate a *jerk* pattern of nystagmus) or a corrective slow movement (to generate a *pendular* pattern of nystagmus).
  - It is theorized that INS, similar to acquired pendular nystagmus, results from a leaky neural integrator.<sup>18</sup>
- Causes
  - Idiopathic
    - Including X-linked idiopathic INS, with FRMD1 mutation.
  - Optic nerve hypoplasia
  - Oculocutaneous albinism (autosomal recessive), ocular albinism (X-linked), often associated with foveal hypoplasia. Trans-illumination defects of the iris may be seen and faulty decussation of optic nerve fibers in the optic chiasm may occur.
  - Retinal dystrophies (achromatopsia, Leber’s congenital amaurosis, congenital stationary night blindness, cone dystrophies)
  - Peroxisomal disorders
- Treatment
  - Contact lenses<sup>19</sup> – through afferent feedback
  - Prisms – to bring the null point into primary gaze and reduces head turn<sup>20</sup>
  - Strabismus surgery
    - Kestenbaum procedure (i.e. resection of horizontal recti) – approximates null point to primary gaze and reduces head turn.<sup>21</sup>
    - To create divergence and force convergence
  - May respond to NMDA receptor antagonists (memantine or gabapentin)<sup>22</sup>
- **Latent Nystagmus**
  - Pattern
    - Latent nystagmus (LN) is a jerk nystagmus in the horizontal plane that only occurs when one eye is covered. The eyes jerk in the direction of the uncovered eye. The slow phase may have a linear or decelerating slope
    - Manifest latent nystagmus (MLN) is a form of LN in which the nystagmus occurs even with both eyes uncovered, because the patient suppresses one eye (as if it is covered) and the other is used for fixation.

- LN / MLN is typically associated with a congenital esotropia and amblyopia.
- Etiology
  - A precise localization of LN/MLN has not been clearly elucidated. However, it has been proposed that LN results from the emergence of monocularly-driven OKNs in the setting of loss of binocularity due to strabismus. With one eye fixating, this monocularly-driven OKN occurs.<sup>23</sup>
- Treatment
  - Four muscle tenotomy and reattachment has been shown to improve nystagmus waveforms and patient quality of life.<sup>24</sup>
- **Spasmus Nutans**
  - Triad includes:
    - Head tilt
    - Pendular nystagmus
    - Head nodding
      - 3Hz, 180° out of phase with nystagmus
  - Pattern of Nystagmus
    - Pendular
    - High amplitude, low frequency (like shimmering)
    - Dissociated, may even be monocular
    - Disconjugate
  - Etiology: Felt to reflect a delay in calibration of eye movements.
  - Prognosis: Tends to remit within 3 years.
  - Mimickers: optic nerve gliomas, retinal dystrophies. Spontaneous resolution of the SN does not preclude diagnosis of an associated optic pathway glioma.<sup>25</sup>

## VI. MIMICKERS OF NYSTAGMUS

- **Convergence retraction “nystagmus”**
  - Found as part of the dorsal midbrain (Parinaud) syndrome
  - Attempted upgaze in the setting of upgaze paresis results in compensatory activation of the remaining extraocular muscles, resulting in repeated globe retraction, and due to disproportionate medial rectus strength, convergence. There is no pathological drift of the eyes and therefore this is not a true nystagmus.
- **Voluntary “nystagmus”**
  - Often observed in children seeking to entertain, this phenomenon is actually a series of voluntary saccades.
  - One tell-tale sign includes an inability of the patient to maintain the demonstration for too long.

## VII. PHYSIOLOGIC FORMS OF NYSTAGMUS

- **End-gaze nystagmus**
  - A few beats of nystagmus in the direction of extreme (beyond 45 degrees) lateral gaze is physiologic and likely results from a limitation of neural integrator function. Asymmetric end gaze nystagmus should prompt a search for a lateralizing lesion.
- **Rotationally induced**
  - Normal VOR: With head flexion, the HSCC become aligned with the horizontal plane. Spinning the patient in a chair will then produce a drift away from the direction they are turning and a jerk towards the direction of movement. The eyes should be closed to avoid a competing OKN.



- After-nystagmus: when the patient opens their eyes after being spun, one will see a transient nystagmus in the direction opposite of the previous VOR. This is due to a temporary inhibition of the previously stimulated canal when the patient's spinning is stopped.
- **Caloric-induced**
  - Cold water infusion into the outer ear canal will impact convection currents in the HSCC, inhibiting its function. The eyes will drift toward the affected ear, followed by a cortically-mediated contralateral saccade (jerk). The test is used to determine brainstem and cortical function following cardiac arrest. If the brainstem is intact but the cortex has suffered pervasive hypoxic injury, the eyes will drift to the ear with the cold water but there will be no jerk phase. If the brainstem is also severely injured, there will be no VOR at all and the eyes will be unaffected. Examination of the tympanic membrane should confirm that it is intact before performing this.
  - Warm water infusion will stimulate the HSCC, leading to a drift away from the ear, and a corrective saccade back towards it. (jerk nystagmus towards the ear.)
- **Optokinetic nystagmus**
  - Passing successive stripes (typically on a strip or drum) past a patient should lead to ipsilateral smooth pursuit as the patient follows a stripe, followed by a contraversive saccade as the eyes move back to pick up the next stimulus. This optokinetic nystagmus (OKN) has localizing value, since the smooth pursuit is governed by the parietal lobe ipsilateral to its direction, and the saccade is governed by the frontal eye field contralateral to its direction.<sup>26</sup> (thus an OKN in one direction is controlled by the cortex ipsilateral to the movement of the stripes.) OKNs are involuntary in patients with count fingers vision or better; it is therefore a good test to perform when one suspects that severe vision loss is nonorganic. The horizontal OKN response appears to depend on the function of the nucleus of the optic tract (NOT)<sup>27</sup>.

#### VIII.CONCLUSION:

Nystagmus comes in a myriad number of forms, each signifying a different localization in the nervous system. The prompt recognition of acquired nystagmus, and its differentiation from congenital forms of nystagmus can help the neurologist localize and diagnosis disease and direct treatment appropriately. Numerable medical and surgical options have emerged for various forms of nystagmus which can help reduce oscillopsia, improve visual acuity and enhance patients' quality of life.

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