

# Nystagmus part 2. central vestibular disorders



최 정 윤

서울의대

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## Contents

- I. Nystagmus from central vestibular system:
- II. Nystagmus from abnormalities of the visual system
- III. Nystagmus from gaze-holding system
- IV. Nystagmus from vergence eye motion
- V. Congenital nystagmus

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## Nystagmus from central vestibular system:

- 1) DBN
- 2) UBN
- 3) Torsional nystagmus
- 4) Horizontal nystagmus
- 5) Seesaw/Hemi-seesaw
- 6) Periodic alternating nystagmus / Windmill nystagmus

## Nystagmus from central vestibular system:

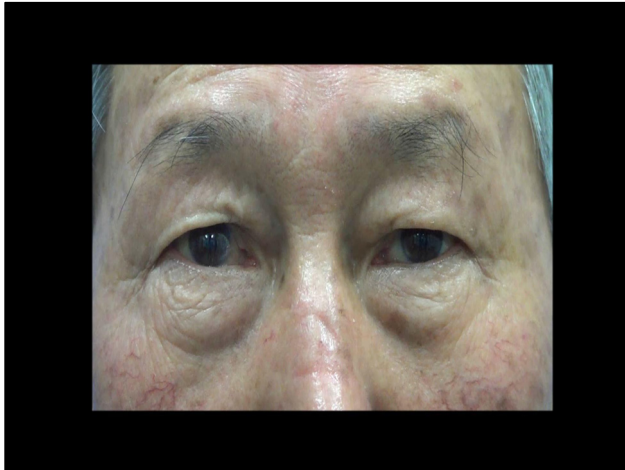
### 1) DBN

Best evoked on looking **down and laterally**

Slow phases may have **linear-, increasing-, or decreasing-velocity** waveforms

May be precipitated or exacerbated or changed in direction, by altering head position, vigorous head-shaking (horizontal or vertical), or hyperventilation

Cerebellar degeneration,<sup>280,74</sup> including familial episodic ataxia,<sup>74,75</sup> paraneoplastic degeneration,<sup>2,75,76</sup> and multiple system atrophy<sup>77</sup>  
Craniovertebral anomalies, including Arnold-Chiari malformation, Paget's disease, basilar invagination, and syringobulbia<sup>284,522,530</sup>  
Infarction of brainstem or cerebellum<sup>27,531,532</sup>  
Rotational vertebral artery syndrome<sup>284</sup>  
Dolichoectasia of the vertebralbasilar artery<sup>527,542</sup> or compression of the vertebral artery<sup>73</sup>  
Multiple sclerosis<sup>27,56,564</sup>  
Cerebellar tumor, including hemangioblastoma<sup>265</sup>  
Encephalitis<sup>28</sup>  
Head trauma<sup>27</sup>  
Increased intracranial pressure and hydrocephalus<sup>528,532</sup>  
Toxic-metabolic  
Anticonvulsant medication<sup>27,34,77,133,347,543</sup>  
Lithium intoxication<sup>542,546,752</sup>  
Alcohol intoxication<sup>73</sup> and induced cerebellar degeneration<sup>74</sup>  
Wernicke's encephalopathy<sup>280,428</sup>  
Magnesium depletion<sup>531,568</sup>  
Amiodarone<sup>27</sup>  
Opoids<sup>73</sup>  
Deficiency of vitamin B12<sup>40</sup> or thiamine<sup>64</sup>  
Tobacco abuse<sup>40</sup>  
Heat stroke<sup>564</sup>  
Tetanus<sup>522</sup>  
Congenital<sup>28,57</sup>  
Transient finding in infants<sup>282,542,747</sup>  
Idiopathic form<sup>53</sup>



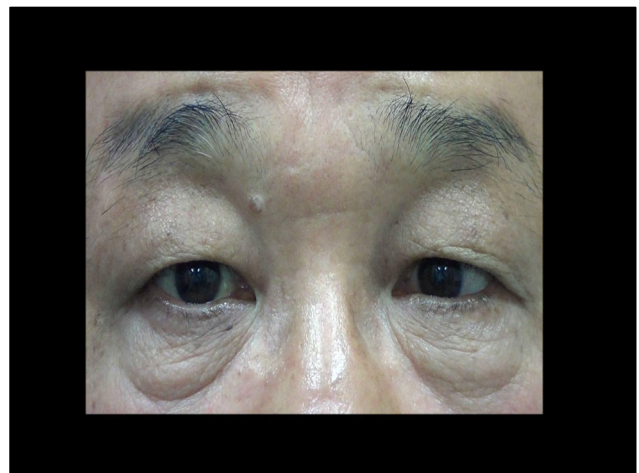
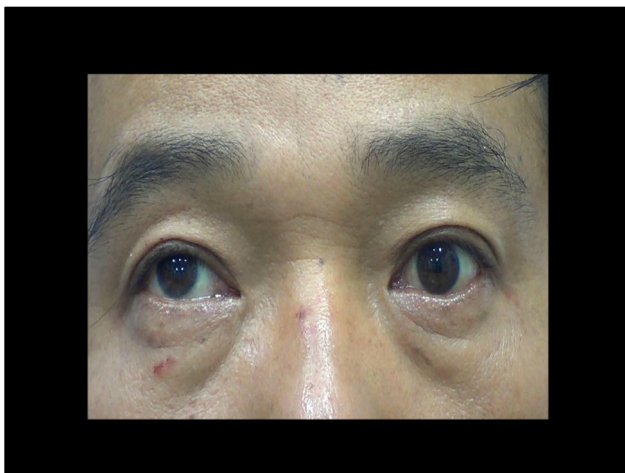
### Nystagmus from central vestibular system:

#### 2) UBN

Present in center position; usually increases on looking up

Slow phases may have linear-, increasing-, or decreasing-velocity waveforms

Infarction of medulla<sup>42,114,126,165,166,167</sup> or cerebellum and superior cerebellar peduncle<sup>73,74</sup>  
 Wernicke's encephalopathy<sup>130,131,166b,744</sup>  
 Multiple sclerosis<sup>126,161,168</sup>  
 Tumors of the medulla,<sup>129,130</sup> cerebellum,<sup>119,167b</sup> or midbrain<sup>164,169</sup>  
 Cerebellar degenerations<sup>161,162</sup> or anomalies<sup>167c</sup>  
 Brainstem encephalitis<sup>125</sup>  
 Creutzfeldt-Jacob disease<sup>72a</sup>  
 Behcet's syndrome<sup>161</sup>  
 Meningitis<sup>123</sup>  
 Leber's congenital amaurosis and other congenital disorders of the anterior visual pathways<sup>162</sup>  
 Thalamic arteriovenous malformation<sup>163</sup>  
 Congenital<sup>133</sup>  
 Organophosphate poisoning<sup>166</sup>  
 Tobacco<sup>167</sup>  
 Associated with middle ear disease<sup>169</sup>  
 Transient finding in infants<sup>142</sup>



### Nystagmus from central vestibular system:

#### 3) Torsional nystagmus

Torsional jerk nystagmus (minimal vertical or horizontal components) present with eye close to central position

Slow phases may have linear-, increasing-, or decreasing-velocity waveforms

Poorly suppressed by visual fixation of a distant target

Often occurs in association with ocular tilt reaction or unilateral internuclear ophthalmoplegia

Syringobulbia, with or without syringomyelia and Arnold-Chiari malformation<sup>70</sup>  
 Brainstem stroke (e.g., Wallenberg's syndrome)<sup>161</sup>  
 Arteriovenous malformation in the brainstem<sup>161,169</sup> or middle cerebellar peduncle<sup>132</sup>  
 Brainstem tumor<sup>153</sup>  
 Multiple sclerosis<sup>153</sup>  
 Oculopalatal tremor ("myoclonus")<sup>153</sup>  
 Head trauma<sup>153</sup>  
 Congenital

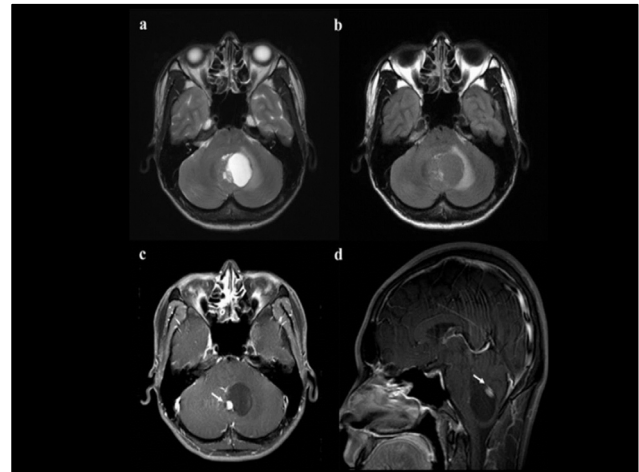


### Nystagmus from central vestibular system:

#### 4) Horizontal nystagmus

Central vestibular disturbances occasionally cause nystagmus that is horizontal (when the eyes are close to central position).

Patients with horizontal nystagmus that is present in the central position should be always be observed for a period of several minutes to exclude the PAN.



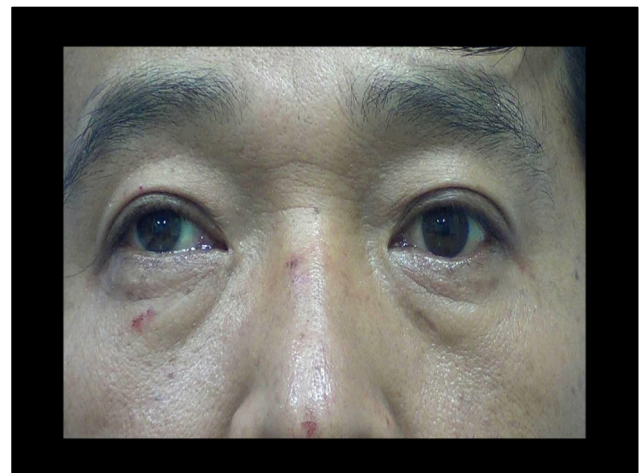
### Nystagmus from central vestibular system:

#### 5) Seesaw / Hemi-seesaw nystagmus

Waveform may be pendular (seesaw); or jerk (hemi-seesaw), in which the slow phase corresponds to one half-cycle

Hemi-seesaw form associated with ocular tilt reaction and other manifestations of otolithic imbalance

Pendular seesaw form associated with bitemporal hemianopia, chiasmal disorders, visual loss



### Nystagmus from central vestibular system:

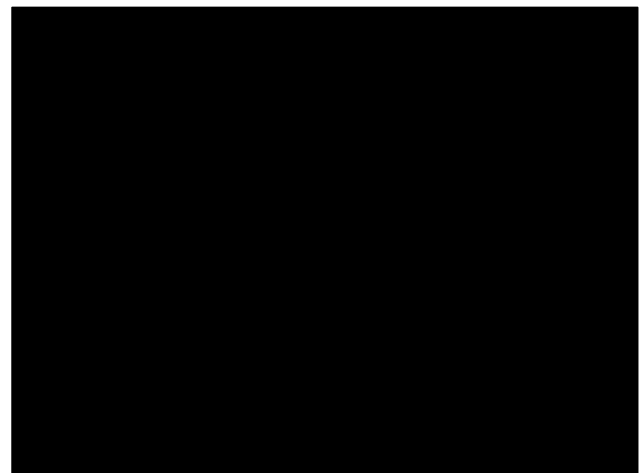
#### 6) Periodic alternating nystagmus

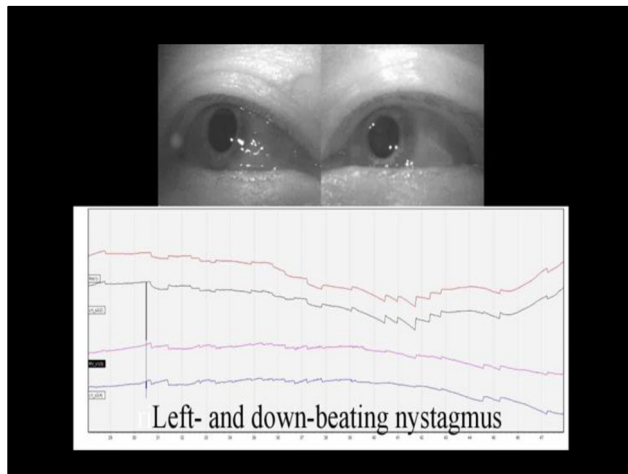
Horizontal nystagmus, reverses direction approximately every 90-120 seconds

Nystagmus cycle is usually little affected by visual fixation

Vestibular stimuli, such as head rotations, can change or transiently stop nystagmus

Downbeat nystagmus and square wave jerks may become more obvious in the brief null period when the horizontal nystagmus wanes and then reverses





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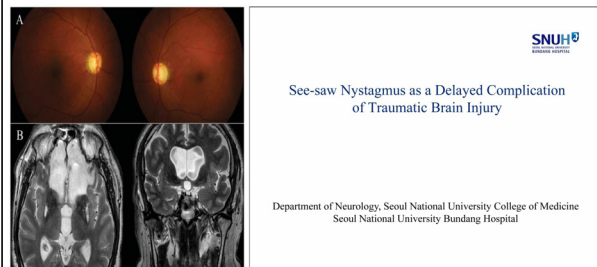
## Nystagmus from abnormalities of the visual system:

### Localization:

- |  |                                |
|--|--------------------------------|
| Eye or optic tract,  | 1) Seesaw nystagmus            |
| Optic chiasm,  | 2) Pursuit paretic nystagmus   |
| Posterior cortical area,                                   | 3) Acquired Pendular Nystagmus |
| Cortico-ponto-cerebellar<br>or Olivocerebellar projections |                                |

## Nystagmus from abnormalities of the visual system:

### 1) Seesaw nystagmus



## Nystagmus from abnormalities of the visual system:

### 2) Pursuit paretic nystagmus



## Nystagmus from abnormalities of the visual system:

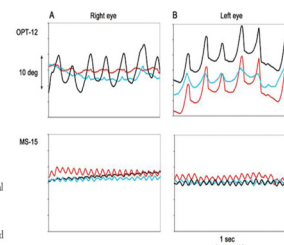
### 3) Acquired Pendular Nystagmus

#### IN ASSOCIATION WITH DEMYELINATING DISEASES

- Frequency 2-8 Hz (typically 3-4 Hz)
- Generally greater amplitude in the eye with poorer vision
- Internuclear ophthalmoplegia commonly associated
- May have an associated upbeat component

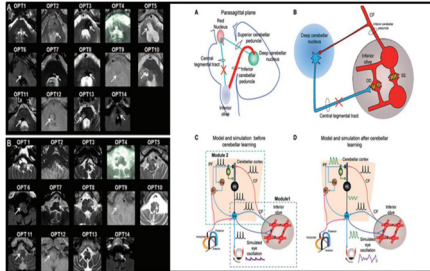
#### SYNDROME OF OCULOPALATAL TREMOR

- Frequency 1-3 Hz (typically 2 Hz)
- May be vertical (with bilateral lesions) or disconjugate vertical-torsional
- Accentuated by eyelid closure
- Movements of palate and other branchial muscles may be synchronized



## Nystagmus from abnormalities of the visual system:

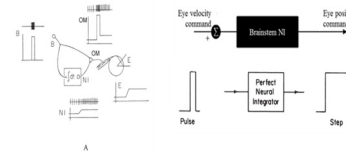
### 3) Acquired Pendular Nystagmus



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## Nystagmus from gaze-holding system:

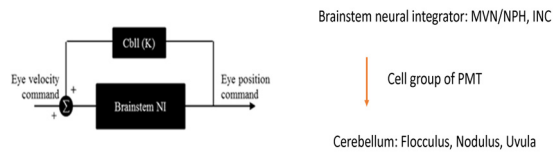


Normal subjects do not have "perfect" neural integrators.

➢ Any biological system is subject to a certain amount of noise.

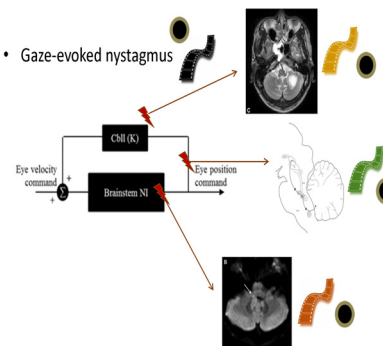


## Nystagmus from gaze-holding system:



## Nystagmus from gaze-holding system:

- Gaze-evoked nystagmus





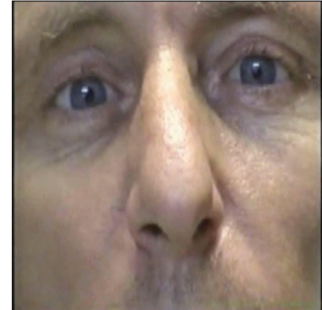
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## Nystagmus from vergence eye motion

Convergent-Divergent pendular oscillations

Convergence-retraction nystagmus



## Nystagmus from vergence eye motion

Convergent-Divergent pendular oscillations

Convergence-retraction nystagmus



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## Congenital nystagmus

- 1) Infantile nystagmus syndrome
- 2) Latent nystagmus and Fusional maldevelopment nystagmus
- 3) Spasmus nutans

## Congenital nystagmus

### 1) Infantile nystagmus syndrome

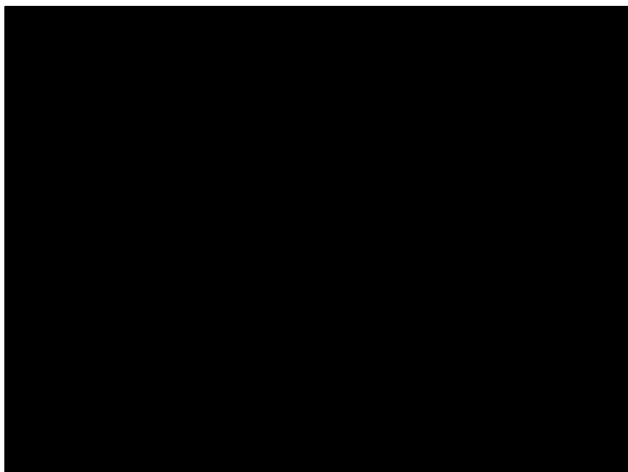
Present since infancy

Usually conjugate, horizontal; smaller torsional or vertical components

Pendular or increasing-velocity waveforms punctuated by foveation periods, during which eyes are transiently still and aimed at the object of interest

Accentuated by visual attention or arousal

Null zone



## Congenital nystagmus

### 2) Latent nystagmus (fusional maldevelopment nystagmus)

Present since infancy; associated with strabismus and lack of binocular vision

Conjugate, horizontal nystagmus beating away from covered eye

Slow phases may have linear-, or decreasing-velocity waveforms

Smooth pursuit asymmetry, depending on viewing eye and on-going nystagmus

Associated with dissociated vertical deviation (eye under cover deviates up)



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