

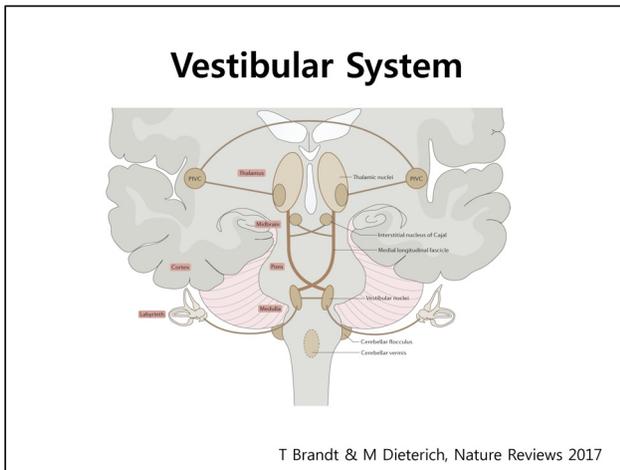
Nystagmus part 1. peripheral vestibular disorders



오 선 영
전북의대

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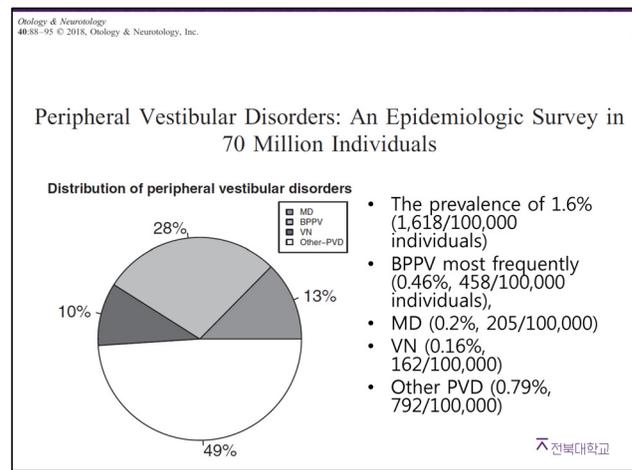
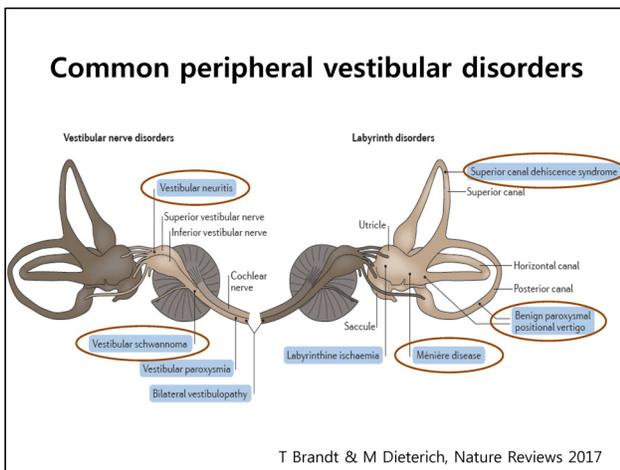
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Peripheral vestibular nystagmus

- Dysfunction of the end organ (semicircular canals, otoliths, vestibular nerve)
- 4 clinical settings**
 - acute, monophasic disorder : *vestibular neuronitis*
 - recurrent form of vestibular dysfunction associated with auditory symptoms (tinnitus and hearing loss) : *Ménière disease*
 - paroxysmal dysfunction of the vestibular system that produces vertigo in response to certain postures of the head : *benign paroxysmal positional vertigo (BPPV)*
 - toxic etiology (aminoglycosides, chemotherapeutics) : *bilateral vestibular hypofunction*

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Vestibular function tests

Tests of Semicircular Canal Function

- Head impulse tests (bedside, video)
- Caloric test
- Rotation test

Tests of Otoliths Function

- Subjective visual vertical (SVV)
- Fundus photo
- oVEMP
- cVEMP

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Peripheral vestibular nystagmus

Vestibular neuronitis

- Mixed horizontal-torsional nystagmus
- Follow Alexander's law
- Unidirectional
- Suppression by visual fixation
- Positive head thrust test

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Head impulse test (bedside)

Normal VN LT

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Video Head impulse test

Normal VN LT

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Peripheral vestibular nystagmus

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Peripheral vestibular nystagmus

LH LV LT

Without fixation With fixation

Rightward gaze Center Leftward gaze

10° 1s

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Spontaneous nystagmus; Direction

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Spontaneous nystagmus; Direction

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Spontaneous Nystagmus

Peripheral or Central nystagmus ?

	Peripheral	Central
Direction	Mixed torsional/horizontal	Pure vertical, torsional, or horizontal
Directionality	Unidirectional (Alexander's law)	Direction-changing
Effect of fixation	Suppression	No effect

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Case Meniere disease

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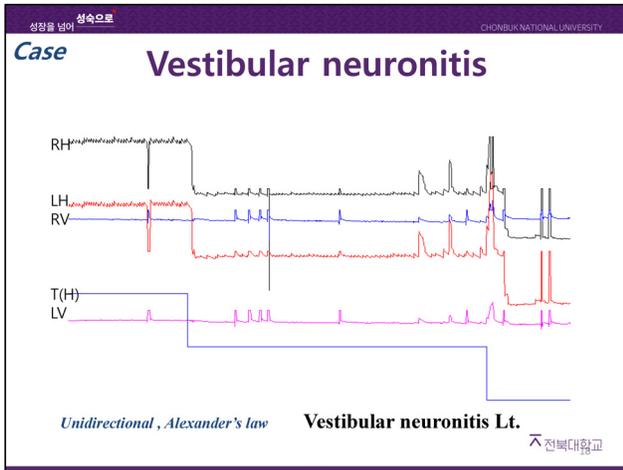
Case Vestibular neuronitis

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Case Vestibular neuronitis

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Direction of spontaneous nystagmus in acute vestibular syndrome

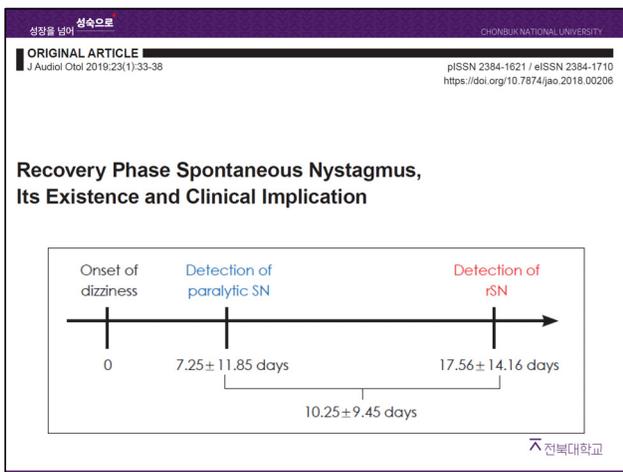
- Direction of the spontaneous nystagmus (SN) may change according to the stage of assessment

Table 1. Stage assessment

Stage of progress of vertigo	Direction of nystagmus		Head-shaking nystagmus	
	Spontaneous nystagmus		1st phase	2nd phase
Irritative	I	to affected side		
Paralytic	II	to intact side		
Central compensation	III-1	n.N.	to intact side	n.N.
	III-2		to intact side	n.N. to affected side
	III-3		n.N.	n.N. to intact side
	III-4		to affected side	to intact side
	III-5		to affected side	n.N.
Recovery	IV	to affected side		
Cure	V	n.N.	n.N.	n.N.

n.N.: no Nystagmus.

Matsuzaki M, Acta otolaryngol 2004;124:100-104

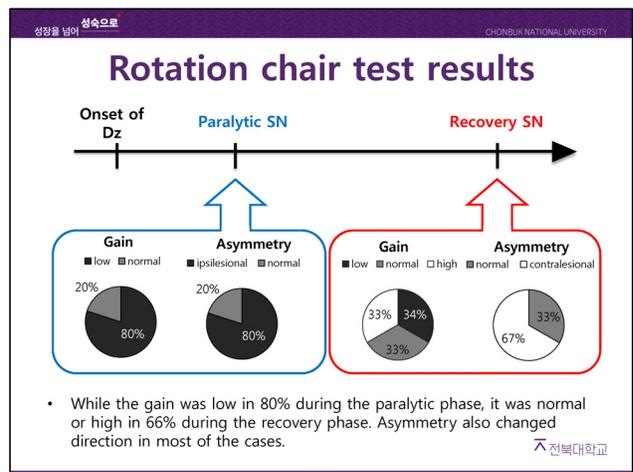
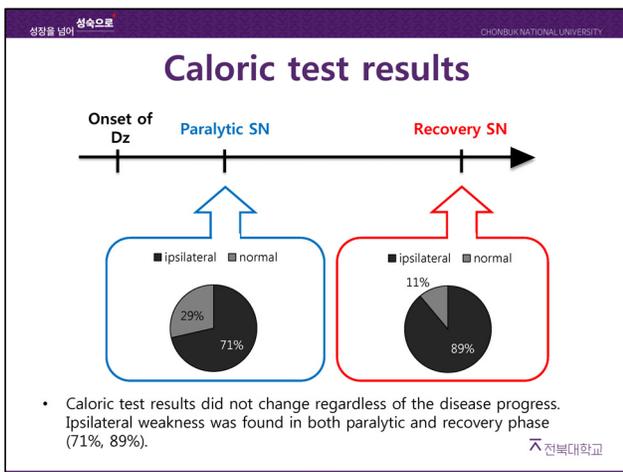


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SN and HSN

Sex/Age	Dx	Lesion side	SN		HSN	
			Paralytic	Recovery	Paralytic	Recovery
P01	M/64	SHLV Rt.	LB	RB	LB	-
P02	F/41	MD Rt.	LB	RB	LB	RB
P03	F/57	VN Rt.	LB	RB	LB	RB
P04	F/57	VN Rt.	LB	RB	LB	RB
P05	F/75	MD Rt.	LB	RB	LB	RB
P06	F/33	SHLV Rt.	LB	RB	LB	RB
P07	F/41	SHLV Rt.	LB	RB	LB	RB
P08	M/71	MD Rt.	LB	RB	LB	RB
P09	M/40	MD Rt.	LB	RB	LB	RB
P10	M/42	SHLV Lt.	RB	LB	RB	-
P11	M/85	SHLV Lt.	RB	LB	RB	LB
P12	F/45	VN Lt.	RB	LB	RB	LB
P13	F/64	VN Lt.	RB	LB	RB	LB
P14	F/56	VN Lt.	RB	LB	RB	LB
P15	F/51	MD Lt.	RB	LB	RB	LB
P16	F/60	MD Lt.	RB	LB	RB	LB

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Recovery phase SN in AVS

- Clinical manifestation of patients with rSN**
 - Direction of SN and HSN may be the same in most cases
 - Direction of SN may not change by head shaking
 - Mismatch in **side of lesion** between SHA and caloric test
 - SHA**: deviated to the contra-lesion side
 - Caloric**: weaker on the ipsi-lesion side
 - Mismatch in **VOR gain** between SHA and caloric test
 - SHA**: gain may be normal or abnormally high
 - Caloric**: may indicate unilateral low VOR gain
 - Hearing may be improved compared to the onset of attack

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Acute spontaneous vertigo; D/Dx.

→ Vestibular neuritis

+ unilateral hearing loss

→ Labyrinthitis,
→ Labyrinthine infarction,
→ Perilymph fistula
→ Meniere's syndrome

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Table 1. Differential Diagnosis of Common Causes of Acute, Prolonged Vertigo.^a

Cause	History	Physical Examination	Laboratory Testing†
Vestibular neuritis	Develops over a period of hours; resolves over a period of days; may follow an influenza-like illness	Spontaneous "peripheral" nystagmus; positive head-thrust test; imbalance	Electronystagmography: unilateral caloric hypoexcitability Audiography: normal Brain imaging: normal
Labyrinthitis	Develops over a period of minutes to hours; may be associated with systemic, ear, or meningeal infection	Same as for vestibular neuritis, but also unilateral hearing loss	Electronystagmography: unilateral caloric hypoexcitability Audiography: moderate-to-severe ipsilateral sensorineural hearing loss Brain imaging: normal
Labyrinthine infarction	Abrupt onset; previous vascular disease; may be associated with neurologic symptoms	Same as for vestibular neuritis, but also unilateral hearing loss; may be associated with neurologic signs	Electronystagmography: absence of a unilateral caloric response Audiography: severe ipsilateral sensorineural hearing loss Brain imaging: MRI may show silent brain infarcts
Perilymph fistula	Abrupt onset associated with head trauma, barotrauma, or sudden strain during heavy lifting, coughing, or sneezing; may be associated with chronic otitis with cholesteatoma	Same as for vestibular neuritis, but usually associated with unilateral hearing loss; possible perforation of the tympanic membrane; positive fistula test (vertigo and nystagmus induced by pressure in external ear canal)	Electronystagmography: unilateral caloric hypoexcitability Audiography: mild-to-moderate ipsilateral sensorineural hearing loss Brain imaging: CT of temporal bone may show erosion from cholesteatoma
Brain-stem and cerebellar infarction	Abrupt onset, history of transient ischemic attacks and vascular disease; usually associated with neurologic symptoms	Spontaneous "central" nystagmus; positive head-thrust test if entry zone of root of 8th nerve is involved; usually focal neurologic signs	Electronystagmography: unilateral caloric hypoexcitability if entry zone of root is involved Audiography: ipsilateral hearing loss if anterior inferior cerebellar artery is involved Brain imaging: MRI shows infarction in medulla, pons, or cerebellum

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Perilymph fistula

- Labyrinthine fistula - aberrant connection between the perilymph-filled inner ear and air-filled middle ear
- 3.6 to 13.9% of COM with cholesteatoma
- Lateral semicircular canal (mc)
- Positional vertigo, severe disequilibrium, and sensorineural hearing loss
- D/Dx BPPV or Meniere's disease

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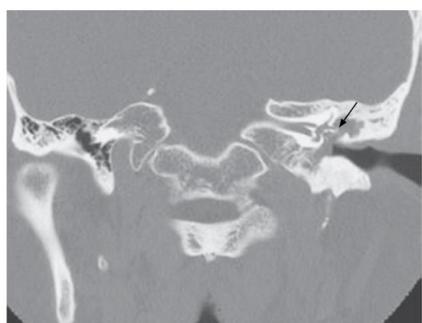
Case Perilymph fistula

- 42/M, severe nausea and vertigo by the application of pressure to the left external auditory canal.
- cholesteatoma
- Fistula test**
- a fistula between the air-filled middle ear and the fluid-filled inner ear.
- left-beating horizontal nystagmus → a bony fistula extending from the middle ear to the left lateral semicircular canal.

N ENGL J MED 366:4 NEJM.ORG JANUARY 26, 2012 전북대학교

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Case Perilymph fistula



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Case Perilymph fistula

- 61/F c HTN & DM
- ER visit, 2 days' dizziness of whirling nature accompanied by nausea and vomiting
- Loss of hearing Lt. for quite a long time, COM
- Spontaneous, right-beating horizontal nystagmus (6.4 d/s).
- Geotropic direction-changing horizontal nystagmus

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Case Perilymph fistula

A head turn to the right B head turn to the left
C head bent 30° forward in sitting D head flexed in supine position

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Case Perilymph fistula

A B

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Case Perilymph fistula

A B C D

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Case Perilymph fistula

A B

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Benign Paroxysmal Positional Vertigo (BPPV)

Historical Aspect

- Cupulolithiasis : Schuknecht, 1969
- Canalolithiasis : Hall et al. 1979
- Liberatory maneuver : Sermont, 1988
- Repositioning maneuver : Epley, 1992

A B

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Benign Paroxysmal Positional Vertigo (BPPV)

- Most common cause of paroxysmal vertigo and nystagmus with positional change
- prevalence 10.7 ~ 64.0 cases per 100,000
- lifetime prevalence of 2.4%
- the annual rate of recurrence, 15%
- Brief spinning sensations/vertigo
- ≤ 1 minute, induced by a change in head position
- typically develops when a patient gets in or out of bed, rolls over in bed, tilts the head back, or bends forward

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Ewald first law

AC, PC, HC, OD

Optical axis 23°

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Intorsion, Hypotropia

Excitatory, Inhibitory

PC, VIII, SO, IO, SR, IR, III, IV

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ANTERIOR CANAL EXCITATORY PROJECTIONS, POSTERIOR CANAL EXCITATORY PROJECTIONS, HORIZONTAL CANAL EXCITATORY PROJECTIONS

Right Eye, Right Ear, MLF, SV, LV, VI, MV, PH, XII, SR, SO, MR, LR, IO, IR

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Mechanisms; vestibular

LAC, RAC, LHC, RHC, LPC, RPC

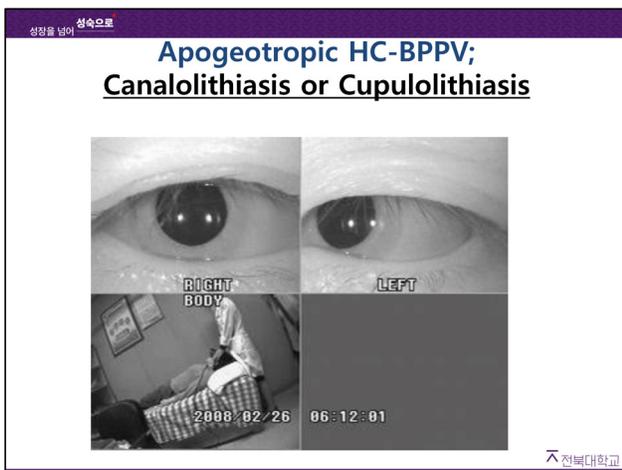
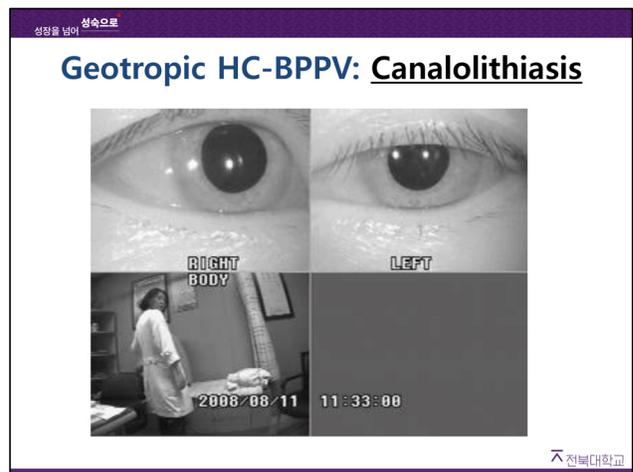
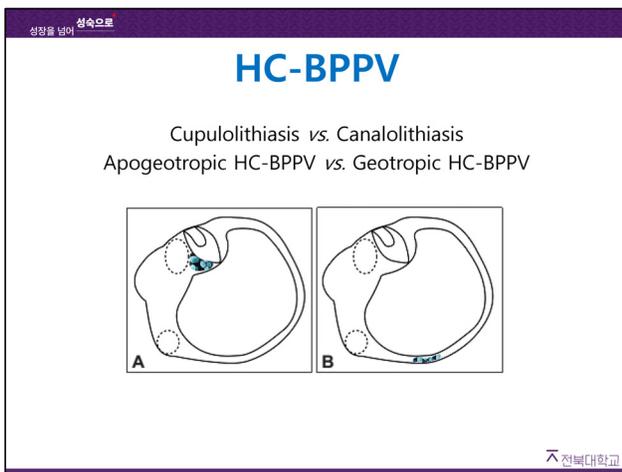
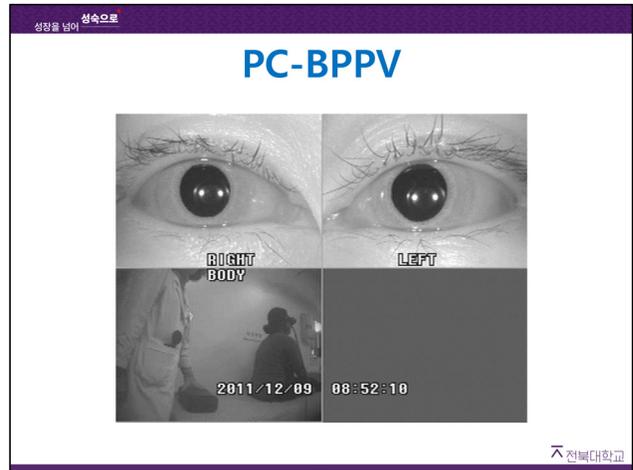
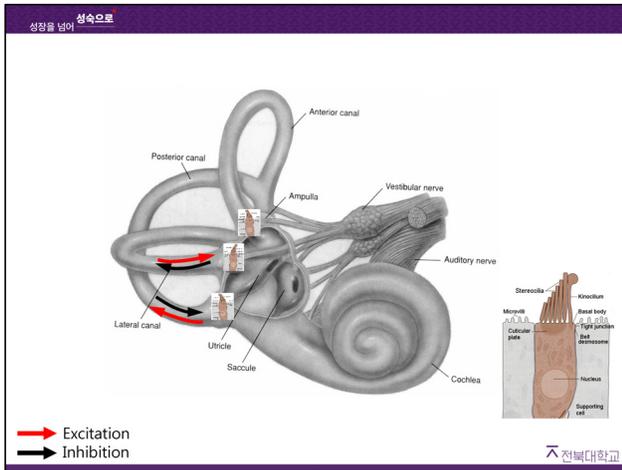
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Direction of Cupular Deflection, Head motion, Endolymph flow, Ampulla of SCC

Resting, Excitation, Inhibition

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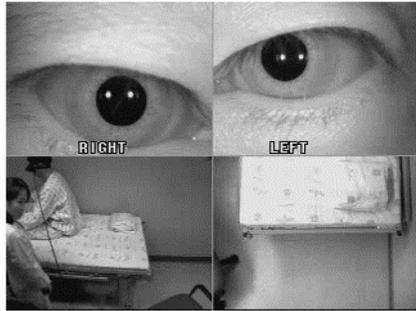
How to determine the affected side

	Lateralization	
	Geotropic	Apogeotropic
Intensity of nystagmus	Stronger side	Weaker side
Lying down nystagmus	Usually contralateral	Usually ipsilateral
Head bending nystagmus	Usually ipsilateral	Usually contralateral
Pseudo-spontaneous nystagmus	Usually contralateral	Usually ipsilateral
Null point	-	Present in lesion side
Reversal of nystagmus	Ipsilateral side	

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AC-BPPV



The image shows a 2x2 grid of photographs. The top row shows close-up views of a patient's eyes with nystagmus, labeled 'RIGHT' and 'LEFT'. The bottom row shows the patient sitting at a table in a clinical setting, with a whiteboard in the background.

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Repositioning maneuver for BPPV

```

    graph TD
      A[Positional Vertigo] --> B[Up and torsional Ny.]
      A --> C[Bidirectional Horizontal Ny.]
      B --> D[PC-BPPV]
      C --> E[Geotropic Ny.]
      C --> F[Apogeotropic Ny.]
      D --> G[Epley or mSemont]
      E --> H["LDN : healthy side  
HBN : affected side  
PSN : healthy side"]
      F --> I["LDN : affected side  
HBN : healthy side  
PSN : affected side"]
      H --> J[Barbecue or Gufoni]
      I --> K[Gufoni or Head-shaking]
    
```

Verifying ampullofugal direction of the otolithic mass !

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Canalith Jam of the Horizontal Semicircular Canal

Two mechanisms of spontaneous nystagmus in HSC-BPPV

- First, the HSC is anatomically tilted 30° upward
 - SN easily changed by head forward or backward
- Second, plugging of the endolymph with the otoconia
 - non fatiguing spontaneous horizontal nystagmus irrespective of the head position.

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Canalith Jam of the Horizontal Semicircular Canal

Case Report/Case Series

Persistent Spontaneous Nystagmus Following a Canalith Repositioning Procedure in Horizontal Semicircular Canal Benign Paroxysmal Positional Vertigo



The diagrams show two views of the horizontal semicircular canal. The left diagram shows a normal canal with otoconia (black dots) at the base. The right diagram shows a canalith jam where the otoconia have formed a solid mass (black arrow) at the base, blocking the ampulla. Labels include 'Utriculus', 'Crista', 'Gravity', and 'Complete impaction'. A 'Video' link is provided.

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Light Cupula

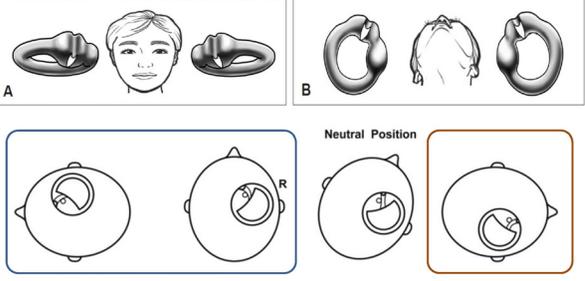
- Two types of geotropic nystagmus:

Transient type	Persistent type
Horizontal canalolithiasis (moving debris)	light cupula of the horizontal canal
	In a supine position, permanent horizontal nystagmus toward the intact side. In a neutral position, with the head turned 20° - 60° to the affected side, the nystagmus decreases and eventually stops. With greater head-turn, nystagmus toward the Earth lasts for more than 1 minute.

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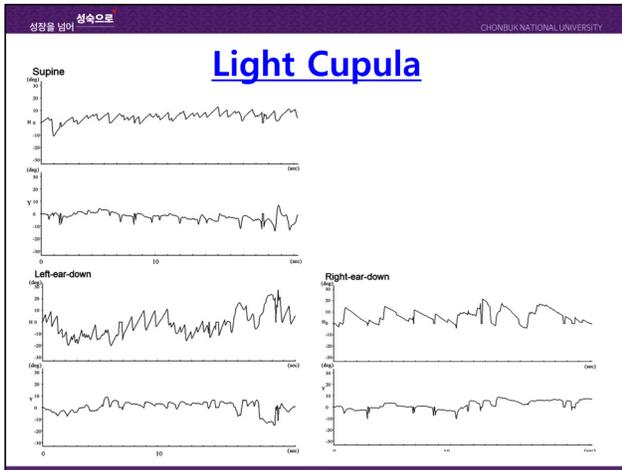
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Light Cupula



The diagrams illustrate the light cupula mechanism. Part A shows a normal cupula with a person's face. Part B shows a light cupula (a small mass on the cupula) causing nystagmus. Below, two diagrams show the 'Neutral Position' with 'Ampullofugal leftward nystagmus' and 'Ampullopetal rightward nystagmus'.

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Light Cupula

	LSCC canalolithiasis	Light cupula
Head-roll test		
Direction	Geotropic DCPN	Geotropic DCPN
Duration	Transient	Persistent
Latency	Yes	No
Null plane	No	Yes
Fatigability	Yes	No

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Superior Canal Dehiscence (SCD)

- A form of peripheral vestibulopathy in which absence of bone over part of the superior canal
- Diagnosis : history, examination, and confirmation of missing bone over the superior semicircular canal by high resolution CT.
- Common Sx : sound-induced vertigo (Tullio's phenomenon) and pressure/Valsalva-induced vertigo
- D/Dx : Meniere's, Lyme disease, congenital deafness, congenital syphilis, and perilymph fistula

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