Evaluation, Diagnosis and Management of Vestibular and Balance Dysfunction in Children

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AAOHNS Instructional Course Materials

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Introduction

1. comprehensive evaluation

2. screening for vestibular & balance function

3. clinical populations and cases
Comprehensive Evaluation of Vestibular and Balance Function
Embryology of the Inner Ear

Week Gestation

- 10: Bony Vestibule
- 12: Membranous labyrinth
- 20: Beginning of brain myelination
- 32: Vestibular receptors fully responsive (can elicit Moro reflex)
- Birth: Vestibular nerve myelination
Vestibular Responsiveness

Infants < 6 months cannot suppress their VOR

Absence of VOR by age 10 months is abnormal
History

#1. What does it feel like?

- sense of motion
- lightheadedness/presyncope/syncope
- aura

Is this vertigo?

- Anxiety / migraine
- Orthostasis in teenage girls
- Temporal lobe seizure
#2. How long does it last?

<table>
<thead>
<tr>
<th>Condition</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>BPPV</td>
<td>seconds-minutes</td>
</tr>
<tr>
<td>TIA / migraine</td>
<td>hours</td>
</tr>
<tr>
<td>Meniere’s</td>
<td>days-weeks</td>
</tr>
<tr>
<td>Labyrinthitis / VN</td>
<td></td>
</tr>
</tbody>
</table>
History

#3. How many attacks of vertigo have there been?

One prolonged: VN, labyrinthitis, infarct

Several: Meniere’s, TIA, migraine

Many: BPPV
History

#5. What sets it off?

- position change
- change in pressure/straining
- head trauma

- BPPV
- PLF / hydrops / SCD
- EVA
#6. What makes it worse?

- moving or keeping still
- rolling over in bed

Vertigo *always* worse with movement

rolling over = BPPV
History

#7. What else happens at the same time?

tinnitus, fullness, hearing loss
dysarthria, diplopia, paresthesia
cranial nerve weakness

headache, paroxysmal torticollis

sweating, palp, dyspnea, chest tightness

Hydrops

VBI

Skull base / intracranial lesion

Migraine / BRVC

Panic / orthostasis
#8. What is the background history?

- otologic disease
  - SNHL (syndromic / non-syndromic) / ototoxic medications
  - vascular disease (congenital cardiopulmonary disease, von Hippel-Lindau)
  - FH neoplasms (NF-2, Gorlin’s syndrome, Costello syndrome etc.)

<table>
<thead>
<tr>
<th>Labyrinthitis / fistula / BPPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital / Acquired vest. Hypofunction</td>
</tr>
<tr>
<td>Intracranial vascular lesion (hemangioblastoma)</td>
</tr>
<tr>
<td>Cerebellar lesions (NF, medulloblastoma)</td>
</tr>
</tbody>
</table>
#8. What is the background history?

- Anxiety / depression
- Motion intolerance
- FH balance disorders
- Autoimmune disease
- Seizure history
- Ophthalmologic disease

- Panic attacks
- Migraine
- Periodic ataxia, migraine, hereditary vestibulopathy
- Autoimmune inner ear disease
- Temporal lobe seizures
- Oculomotor anomaly, amblyopia, acuity, depth perception
Physical Exam

- static testing
  - spontaneous and gaze evoked nystagmus
  - hyperventilation
  -Valsalva
  -tragal compression

- dynamic testing
  - Dix-hallpike maneuver
  -head thrust maneuver
  -post- headshake nystagmus
  -dynamic visual acuity
  -per-rotatory nystagmus
  -VOR suppression

- vestibulospinal testing
  - pastpointing
  -fukuda stepping test
  -romberg
  -gait/one foot standing
Pediatric Physical Exam

- Head and Neck Exam
- Tragal Compression (fistula sign)
- CN II-XII
- Motor Power Upper/Lower Limbs
- Dysdiadokinesia (rapid alternating movements)
- Dysmetria (pass pointing)
- VOR Suppression
- EOM (spontaneous/gaze nystagmus)
- Saccadic eye movements
- Vergence/fixation
- Head thrust testing
- Post head shake nystagmus

- Per/Post Rotatory nystagmus
- Dynamic Visual Acuity
- Rhomberg (modified)
- Unterberger (Fukuda Stepping test)
- Static/Dynamic Balance (BOT-2, one foot standing, running)
- Dix-Hallpike
- Hyperventilation

**Audiometric Assessment**
(reflexes, SRS)
Vestibular end-organ testing
Head thrust testing
aka
Halmagyi test
aka
Head impulse test (HIT)
VOR SUPPRESSION
FUKUDA STEP TEST

AKA

UNTERBERGER TEST
DYNAMIC VISUAL ACUITY TESTING
PER-ROTATORY NYSTAGMUS
AND
POST-ROTATORY NYSTAGMUS
HEAD SHAKE NYSTAGMUS
Balance Assessment

Expensive & Complicated

vs.

Cheap & Easy
Gait Analysis

- Dynamic balance kinetics and kinematics
- Position / movement CM walking straight line (60 Hz data collection)
  - Self selected speed for 9 meters
Computerized Dynamic Posturography

- Limits of stability using 8 standard trials
- Reaction time, movement velocity, endpoint excursion, max excursion, directional control, measures of sway
Balance Assessment
Cheap & Easy

Subjective

- Rhomberg
  (tandem Rhomberg)
- 1 foot standing
  (eyes open/closed)
- run around the clinic
Balance Assessment
Cheap & Easy

Subjective

- Rhomberg
  (tandem Rhomberg)
- 1 foot standing
  (eyes open/closed)
- run around the clinic
Balance Assessment
Cheap & Easy

More objective

- Peabody Motor Subset (1 – 7 years)
  - Gross Motor Scale
  - Fine Motor Scale
- Bruininks-Oseretsky (4 – 21 years)
  Test of Motor Proficiency
# Measuring Balance

**Bruininks-Oseretsky (BOT2)**

## Balance Subtest

<table>
<thead>
<tr>
<th>Balance subtest Items</th>
<th>Max. Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Standing with feet apart on a line</td>
<td>Eyes Open 10 sec.</td>
</tr>
<tr>
<td></td>
<td>Eyes Closed 10 sec.</td>
</tr>
<tr>
<td>2. Walking forward on a line</td>
<td>6 steps</td>
</tr>
<tr>
<td>3. Standing on one leg on a line</td>
<td>Eyes Open 10 sec.</td>
</tr>
<tr>
<td></td>
<td>Eyes Closed 10 sec.</td>
</tr>
<tr>
<td>4. Walking forward heel to toe on a line</td>
<td>6 steps</td>
</tr>
<tr>
<td>5. Standing on one leg on a balance beam</td>
<td>Eyes Open 10 sec.</td>
</tr>
<tr>
<td></td>
<td>Eyes Closed 10 sec.</td>
</tr>
</tbody>
</table>
Balance Assessment
The Bare Minimum

<table>
<thead>
<tr>
<th>One Leg Standing</th>
<th>Sensitivity</th>
<th>Specificity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eyes Open &lt; 8 seconds</td>
<td>100%</td>
<td>49%</td>
</tr>
<tr>
<td>Eyes Closed &lt; 4 seconds</td>
<td>90%</td>
<td>84%</td>
</tr>
</tbody>
</table>
Vestibular end-organ testing

- lateral canal function
  - caloric
  - rotational chair
    - low and high frequency (0.5 to 5 Hz)
  - video head impulse test goggles

- saccular function
  - cervical vestibular evoked myogenic potentials (VEMP)

- utricular function
  - ocular VEMP
Lateral Canal Function
Lateral Canal Function - Caloric

**Bithermal Calorics**
- Low-frequency (0.01 Hz)
- Assess laterality

**Making it Child-Proof**
- VNG vs. ENG
- Air vs. Water Calorics
- Adequate alerting
- Wax and middle ear fluid
- Kids enjoy being dizzy
- Appropriate targets
VNG / ENG

- Other measures
  - spontaneous, positional, positioning nystagmus
  - smooth pursuit tracking
  - saccadic eye movements
  - optokinetic nystagmus
Lateral Canal Function - Rotation

- Low to high frequency (0.1 to 15.0 Hz)
- Higher frequency than calorics (0.01 Hz)
- Physiologic (head/body rotation)
- Phase, Gain and Symmetry of the VOR
- Limited assessment of laterality
- Subject to compensation
Rotational Chair Testing
Making it Child-Proof

- Better tolerated than calorics
- Small children/infants sit in parents lap/car seat
- Use VNG vs. ENG vs. Scleral Coil
Lateral Canal Function
vHIT testing

- high frequency horizontal canal function
- assesses laterality
- alternative to rotary testing
Lateral Canal Function
vHIT testing
Saccular Function
Cervical VEMP (cVemp)
Saccular Function - cVEMP
(Vestibular Evoked Myogenic Potential)

- Myogenic response of Sternocleidomastoid m.
- Tests saccule & inferior vestibular nerve
- P1 = 13 ms
- N1 = 23 ms
Saccular Function - cVEMP
(Vestibular Evoked Myogenic Potential)

- 500 Hz tone burst (75-95 dB HL air or 66 dB pip bone)
- Tonic SCM contraction = EMG 50-250 microvolts
- 80-100 samples averaged
cVEMP (Making it Child-Proof)

- Latency differences by age

<table>
<thead>
<tr>
<th>Latency Range (ms)</th>
<th>P1</th>
<th>N1</th>
</tr>
</thead>
<tbody>
<tr>
<td>8.3 – 14.4</td>
<td>14.8 – 21.9</td>
<td></td>
</tr>
</tbody>
</table>

- Active response
- Video feedback
Utricular Function

Ocular VEMP (oVemp)
Utricular Function

- Ocular Vemp (oVemp)
- Subjective Visual Vertical
Easy pediatric vestibular screen

1. motor milestones
2. standing on one foot
3. head thrust testing
# 1. Motor Milestones

## Red Flags

<table>
<thead>
<tr>
<th>Motor Milestone</th>
<th>Timeframe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absence of head control</td>
<td>4 months</td>
</tr>
<tr>
<td>Unable to sit unsupported</td>
<td>7-9 months</td>
</tr>
<tr>
<td>Unable to crawl/bottom shuffle</td>
<td>12 months</td>
</tr>
<tr>
<td>Not attempting to walk</td>
<td>18 months</td>
</tr>
</tbody>
</table>
2. Dynamic balance: 1 foot standing

<table>
<thead>
<tr>
<th>Age</th>
<th>Duration (sec)</th>
<th>1 foot standing</th>
</tr>
</thead>
<tbody>
<tr>
<td>30 months</td>
<td>1</td>
<td>(briefly)</td>
</tr>
<tr>
<td>36 months</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>4 years</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>5 years</td>
<td>10</td>
<td></td>
</tr>
</tbody>
</table>
3. Lateral Canal Function: Head Thrust Testing
Easy pediatric vestibular screen

1. motor milestones
2. standing on one foot
3. head thrust testing
Kids who don’t know they are dizzy

Vestibular and Balance Disorders in Deafness
Differential Diagnosis: Dizziness with hearing loss

- Acute onset
  - Viral neuronitis/labyrinthitis
  - Autoimmune disease
  - Cogan’s syndrome
  - EVA, other anatomical variants
- Otitis media
- Labyrinthine concussion with or without fracture
- Lyme Disease
- Whiplash
Differential Diagnosis: Dizziness with hearing loss

- Subacute/chronic onset
  - Meniere’s Disease/Endolymphatic Hydrops
  - Ototoxicity
  - Perilymphatic fistula
  - Superior semicircular canal dehiscence
  - EVA

- HIV, Lyme disease
  - Syphilis
  - Otitis media
  - Usher Syndrome
  - Schwannoma
Deafness and Vestibular Function

- 232 children
- representative etiology

- Abnormal Cochlea: 21%
- Meningitis: 7%
- Other: 13%
- Connexin: 26 (17%)
- Unknown: 42%
Deafness and Vestibular Function

- 232 children
- representative etiology

70% have dysfunction
20-40% complete areflexia
Deafness and Balance

BOT-2 Scale Score

<table>
<thead>
<tr>
<th></th>
<th>norms</th>
<th>implant</th>
</tr>
</thead>
<tbody>
<tr>
<td>p-value</td>
<td>0.0001</td>
<td></td>
</tr>
</tbody>
</table>

* Indicates statistical significance.
Deafness and Balance

* $p = 0.0001$

Mean 4.4 years
Impact of Etiology on Balance
There are Consequences

vestibular & balance dysfunction

α

risk of CI failure

Bilateral Areflexia

Odds ratio: 7.6

Implant failure
Vestibular and Balance Disorders

Kids who know they are dizzy
Differential Diagnosis: Dizziness without Hearing Loss

- Acute:
  - Benign Paroxysmal Positional Vertigo (BPPV)
  - Migraine variants
  - Postural orthostatic tachycardia syndrome (POTS)
  - Epileptic vertigo (esp. post-traumatic)
  - Vestibular neuronitis
Differential Diagnosis: Dizziness without Hearing Loss

- Chronic Dizziness
  - Chiari Malformation
  - Multiple sclerosis
  - Ocular abnormalities
  - Post-concussive
  - Familial periodic cerebellar ataxia (types 1-7)
  - Vertebro-basilar insufficiency
### Table 1. Distribution of Diagnoses in 132 Children Evaluated for Vestibular and Balance Disorders

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Peripheral vestibulopathy</td>
<td>39</td>
<td>29.5</td>
</tr>
<tr>
<td>Migraine/benign recurrent vertigo of childhood</td>
<td>32</td>
<td>24.2</td>
</tr>
<tr>
<td>Motor/developmental delay</td>
<td>14</td>
<td>10.6</td>
</tr>
<tr>
<td>Traumatic brain injury</td>
<td>13</td>
<td>9.8</td>
</tr>
<tr>
<td>Central nervous system structural lesion</td>
<td>12</td>
<td>9.1</td>
</tr>
<tr>
<td>Behavioral/psychogenic</td>
<td>8</td>
<td>6.1</td>
</tr>
<tr>
<td>Idiopathic/normal</td>
<td>4</td>
<td>3.0</td>
</tr>
<tr>
<td>Movement disorder/neurodegenerative</td>
<td>4</td>
<td>3.0</td>
</tr>
<tr>
<td>Encephalopathy</td>
<td>2</td>
<td>1.5</td>
</tr>
<tr>
<td>Vascular</td>
<td>2</td>
<td>1.5</td>
</tr>
<tr>
<td>Peripheral neuropathy</td>
<td>1</td>
<td>0.8</td>
</tr>
<tr>
<td>Oculomotor abnormality</td>
<td>1</td>
<td>0.8</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>132</td>
<td>99.9</td>
</tr>
</tbody>
</table>

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**Comprehensive Vestibular and Balance Testing in the Dizzy Pediatric Population**

Robert C. O’Reilly, Lowell Greywoode, Thierry Morlet, Freeman Miller, John Henley, Chris Church, Jeffrey Campbell
<table>
<thead>
<tr>
<th>Condition</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inner ear malformations (EVA and third window)</td>
<td>9</td>
</tr>
<tr>
<td>Sensorineural hearing loss (excluding EVA)</td>
<td>4</td>
</tr>
<tr>
<td>Benign paroxysmal peripheral vertigo</td>
<td>3</td>
</tr>
<tr>
<td>Ménière’s disease</td>
<td>3</td>
</tr>
<tr>
<td>Otitis media</td>
<td>3</td>
</tr>
<tr>
<td>Progressive cochleovestibulopathy</td>
<td>2</td>
</tr>
<tr>
<td>Cytomegalovirus</td>
<td>2</td>
</tr>
<tr>
<td>Temporal bone fracture</td>
<td>2</td>
</tr>
<tr>
<td>Congenital vestibular hypofunction</td>
<td>2</td>
</tr>
<tr>
<td>Post–cochlear implantation</td>
<td>2</td>
</tr>
<tr>
<td>Vestibulopathy NOS</td>
<td>2</td>
</tr>
<tr>
<td>Vestibular neuritis</td>
<td>2</td>
</tr>
<tr>
<td>Autoimmune inner ear disease</td>
<td>2</td>
</tr>
<tr>
<td>Viral labyrinthitis</td>
<td>1</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>39</strong></td>
</tr>
</tbody>
</table>

**Comprehensive vestibular and balance testing in the dizzy Pediatric Population**

Robert C. O’Reilly, Lowell Greywoode, Thierry Morlet, Freeman Miller, John Henley, Chris Church, Jeffrey Campbell
### Table 3. Distribution of Individual Diagnoses in Children With Migraine Disease (24.2% of the Total Population)

<table>
<thead>
<tr>
<th>Condition</th>
<th>n</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine equivalent</td>
<td>16</td>
</tr>
<tr>
<td>Migraine</td>
<td>7</td>
</tr>
<tr>
<td>Benign recurrent vertigo of childhood</td>
<td>6</td>
</tr>
<tr>
<td>Paroxysmal torticollis</td>
<td>3</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>32</td>
</tr>
</tbody>
</table>

Comprehensive Vestibular and Balance Testing in the Dizzy Pediatric Population

Robert C. O'Reilly, Lowell Greywoode, Thierry Morlet, Freeman Miller, John Henley, Chris Church, Jeffrey Campbell
Tips in the Diagnosis of Vestibular Migraine in Children

- prevalence
  - 2-10% school aged children
- in children with normal otoscopic findings, vertigo is commonly caused by migraine and migraine equivalents: benign paroxysmal vertigo (BPVC) of childhood
Childhood Migraine

Benign Paroxysmal Vertigo of Childhood (BPVC) (Benign Recurrent Vertigo of Childhood (BRVC))

- ICHD-II R1: “childhood periodic syndromes that are commonly precursors of migraine”
  - Presents at age 1-4 yrs.
  - Most common cause of dizziness at this age
  - Nystagmus, nausea, emesis, diaphoresis, torticollis
  - Brief duration
  - Resolution in 1-2 years
  - “Classic” migraine later in life
BPVC
Benign Paroxysmal Vertigo of Childhood

Diagnostic Criteria - Basser Criteria (1964)

- sudden brief attacks of vertigo (sec. to min.)
- before school age
- accompanied by
  - nystagmus
  - pallor
  - nausea
  - phonophobia
  - photophobia
Childhood Migraine

- Migraine with Aura
  - 14-30% have headache with aura (more than adults)
  - Occurs in older children
  - Often different than adults:
    - headache bilateral
    - late afternoon onset
    - < 2 hour duration.
Results

- 39/240 (16%)
  - BPVC (10)
  - migraine with aura (29)
    - Diagnosed by Peds Neurology and Neurotology
- mean age at testing: 11.3 yrs (± 4.7)
- 23 Female; 16 Male
- 10 BRVC (mean age 4.3) (1 child < 2 years)
- 29 Migraine with Aura (mean age 13.5)
- MRI: all normal
Results

- **Audiometry**
  - 3 abnormal (1 bilateral profound) in migraine group

- **Tympanometry**
  - 10% abnormal (40% of BRVC): Type C or PE tubes

- **DPOAEs**
  - 1 abnormal (migraine pt. with profound SNHL)

- **VEMP**
  - 28 tested: all present and normal

- **Rotational Chair Testing**
  - 37 tested: all essentially normal (high gain/single frequency asymmetry)
Results

- **VNG**
  - 7/28 migraine: “central” (positional nystagmus or oculomotor abnormality)
  - 5/10 BRVC: normal

- **Gross Motor Development**
  - 53 % abnormal (40% BRVC/ 58% migraine)
  - “below average” strength, agility, coordination

- **Gait**
  - 67% full assessment / 33% videos
  - 51% abnormal (40% BRVC/ 55% migraine)

- **Posturography**
  - 35% abnormal
  - Slow reaction time / increased postural sway

- **BMI**
  - 35% overweight or obese
    - 57% OW/OB at least 1 Gait/Gross Motor/CDP abnormality
    - 86% NW – at least 1 Gait/Gross Motor/CDP abnormality
Summary

1. vestibular/balance deficits common in deafness

2. screening for vestibular dysfunction is feasible

3. the differential is broad
Cases
Case Presentation 1

- 16 year old boy
- 8 month history of visual blurring and instability
  - Began while an inpatient
    - Treated medically for crohn’s related bowel perforation
    - 2 week course of ampicillin/gentamycin/flagyl
  - Unstable / Unable to play usual sports (curling)
  - No acute vertiginous episodes
  - No audiologic complaints
Case Presentation 1

Clinical Assessment

- saccades noted on head thrust (Halmagyi) testing bilaterally
- visual acuity testing
  - 20/10 static visual acuity testing
  - 20/100 dynamic visual acuity
- no evidence of post rotatory nystagmus
- dynamic instability with eyes closed
Case Presentation 1

Ancillary Testing

- Caloric – no response to ice water
- cVEMP – absent
- Rotary Chair with scleral coil – reduced gain
Case Presentation 1

Diagnosis – Bilateral Vestibular Loss – ototoxicity

- Gentamycin induced vestibulotoxicity
- Genetic work-up negative
  - Mutations in mitochondrial RNA (MTRNR1/MTTS)
    Fishel-Ghodsian et al, 1997; Gardner et al, 1997
- Vestibular rehabilitation program (physiotherapy)
Case Presentation 2

- 2.5 year old girl
- 3 month history of episodic vertigo
  - sudden onset
  - ‘the house is shaking’
  - child would close eyes and want to be held
  - 1-2 minutes in duration
  - loss of appetite, occasional emesis
  - following acute episode child would want to go to bed
  - paroxysmal torticollis as an infant
Case Presentation 2

- frequency: 3-5 episodes / month
- initial episodes in head hanging position
- subsequent episodes while sitting and running
- child began avoiding head hanging position
Case Presentation 2

- **Clinical Exam**
  - Head and Neck exam WNL
  - EOM Normal
  - Halmagyi Normal
  - Ø Post head shake Nystagmus
  - Gait / Rhomberg Normal
  - Dix Hallpike Normal
  - Dynamic Visual Acuity Normal (LEA Symbols Chart)
  - Cranial Nerves / Cerebellar Exam normal
Case Presentation 2

- Audiometry
  - Normal PTA
  - Normal tympanometry and middle ear pressure
- ENG Caloric – Warm water – equal
- VEMP Normal bilateral
- Rotational Chair (0.5 to 5 Hz) Normal gains bilaterally
- MRI - Normal
Case Presentation 2 : BPVC
Benign Paroxysmal Vertigo of Childhood

Diagnostic Criteria - Basser Criteria (1964)

- sudden brief attacks of vertigo (sec. to min.)
- before school age
- accompanied by
  - nystagmus
  - pallor
  - nausea
  - phonophobia
  - photophobia
Case Presentation 3

- 15 year old boy suffered concussive injury following football tackle
- Episodic vertigo
  - Lasting seconds
  - While rolling onto right side or looking up and right
  - Frequency 3 – 4 times per day
Case Presentation 3

Clinical Exam
- Head and Neck exam WNL
- EOM Normal
- Halmagyi Normal
- Ø Post head shake Nystagmus
- Dynamic Visual Acuity Normal (Sloan Letters Chart)
- Gait / Rhomberg Normal
- Dix Hallpike – symptomatic vertigo with classic geotropic rotary nystagmus - right head hanging position
- Cranial Nerves / Cerebellar Exam normal
Case Presentation 3

- Audiometry
  - Profound SNHL - Right
  - Normal tympanometry and middle ear pressure
- ENG Caloric – equal
- VEMP Normal bilateral
- Rotational Chair (0.5 to 5 Hz) Normal gains bilaterally
- CT - Normal
Case Presentation 3

**DIAGNOSIS** – Post-traumatic Benign Paroxysmal Positional Vertigo (BPPV)

**TREATMENT**

- Multiple attempts at repositioning with ongoing recurrence of symptoms over 18 months
- Posterior semicircular canal occlusion
- No further symptoms
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AAOHNS Instructional Course Materials

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