Acute Dizziness with Hearing Loss

Autoimmune Inner Ear Disease:
- Unilateral or bilateral
- Vertigo, lightheadedness, ataxia, motion intolerance
- unilateral/bilateral/fluctuating/progressive SNHL
- fluctuating tinnitus and aural fullness occur present
- symptoms progress over weeks to months
- diagnosis primarily clinical
- symptoms may improve with steroids
- associated w/ systemic autoimmune diseases (15-30%), rarely endocrine disease and/or recurrent fevers

Tests: NO DEFINITIVE TESTS. Lymphocyte transformation test (controversial), Mitogen inhibition test (technical difficulties), Circulating Immune Complexes, ESR, CRP, ANA, RF, TSH, anti-microsomal antibodies, anti-gliadin antibodies (Celiac disease), HLA testing.
Others: FTA-ABS (syphilis), HIV, Hb A1C, and Lyme titers

Cogan’s Syndrome:
- rare inflammatory disease
- ocular: nonsyphilitic ocular interstitial keratitis, scleritis, episcleritis, iritis or choroiditis
- audiovestibular: sudden SNHL, vestibular impairment
  - fluctuating hearing and vestibular function
  - variable vestibular impairment (SCCs, otolith organs)
  - may appear before or after ocular symptoms (up to 2 yr delay)
  - w/o steroids, SNHL may progress
- systemic: pericarditis, vasculitis, fevers, arthralgias, myalgias
- some theories that vaccinations and infections may trigger

Enlarged Vestibular Aqueduct (EVA):
- hearing loss ranges from mild to profound, fluctuating, step-wise progressive or sudden and can be either Mixed or SNHL
- ? progression of HL from minor head trauma
- vestibular dysfunction ranges from mild imbalance (clumsiness) to vertigo
- unilateral or bilateral
- diagnosis: axial CT or T2 MRI - mid point of vestibular aqueduct >1.5 mm (Valvassori and Clemis – traditional definition.); Newer criteria: > 1.0 mm at midpoint, > 2.0 mm at operculum (Vijayasekaran)
- third window effect?
- Histologically – thin walled endolymphatic sacs which are missing the rugal folds and perisaccular loose vascular tissue seen in normal endolymphatic sacs
- Possible association b/w EVA and Meniere’s.

Tests: VEMPS – lower thresholds, higher amplitudes;

ETD and Middle ear effusion:
- Most common causes of vestibular disturbances in children
• Parents report clumsiness, awkwardness, unsteadiness, frequent falls
• Fluid visible on exam, pneumatic otoscopy w/ limited TM mobility
• Balance disturbance equally likely w/ unilateral effusions as bilateral effusions
• Hearing loss (Conductive)

Tests: ENG – no abnormalities on OKN and Pursuit testing; Spontaneous unidirectional horizontal nystagmus in 1/3, positional nystagmus in 17.5%

HIV:
• Congenital infection
  • Chronic or acute vertigo, ataxia, hearing loss (CHL or SNHL), tinnitus
    o Hearing loss can be unilateral or bilateral, sudden or progressive
    o ABR: increased I-III latencies
  • ENG: both central and peripheral findings
    o Pursuit abnormalities
    o Weakness on caloric
• Etiology – unknown
  o temporal bone studies have shown viral-like particles and inclusion bodies in the hair cells of the vestibular end organs
  o ? if result of antiviral therapy

Labyrinthine Concussion:
• follows head injury (temporo-parietal and occipito-parietal areas)
• headaches, vertigo, tilting sensation, dizziness, nausea/vomiting, visual disturbances, unsteadiness (sway to affected side), irritability, personality changes, cognitive impairments, sleep disturbances
• Symptoms present for several days – subside over 4-6 wks
• Recurrent attack of vertigo (5-10 sec) w/ nausea
• Can have positional nystagmus on Dix-Hallpike testing (BPPV)
• Normal hearing
• May have balance problems for many years as seen w/ tandem gait and Romberg w/ eyes closed

Tests:
  Dix-Hallpike – positive test for affected ear
  Normal rotational chair

Lyme Disease:
• Spirochete Borrelia burgdorferi (BB) spread by tick bite
• First Stage: Erythema chronic migrans
• Second Stage (acute disseminated stage):
  o 3-24 months after infection
  o Cardiac abnormalities (eg AV block)
  o Arthritis/arthritis
• Third Stage (“neuroborreliosis” – infection of CNS)
  o 6 months – 2yrs after tick bite
  o Peripheral neuropathies
  o Encephalitis, chronic meningoencephalitis
Rheumatoid arthritis
Chronic dermatologic d/o (acrodermatitis chronica atrophicans)
Headache, fatigue, arthritis fevers, malaise
Cranial Nerve abnormalities are seen in 5-10%
  - Facial N Paralysis 80% of CN abnormalities; can be bilateral
  - CN VIII: Vertigo, Unexplained SNHL, Tinnitus, aural fullness
  - CN III (oculomotor) and VI (abducens)
CT normal; MRI – may be normal or may show focal brain inflammation (w/ predilection for white matter) – in cases of encephalitis.
IgG Antibodies to BB elevated in CSF and serum (past or present infection); CSF pleocytosis and elevated protein indicate active infection. “titers may be negative in up to 1/3 of cases” – Controversial. Current thinking is that “seronegative Lyme” is very rare;
Tests: From one case report only: ENG – saccadic dysmetria, increased latency; Smooth pursuits – normal; Calorics – normal;

Meniere’s Disease/ Endolymphatic Hydrops:
  - Meniere’s is rare in childhood
  - Recurrent paroxysmal vertigo, SNHL (fluctuating to progressive), aural fullness, tinnitus, n/v and headache
  - HL may be high frequency (unlike adults) initially
  - FH may be significant for Meniere’s
  - Hydrops may also be seen with congenital CMV and syphilis;
    - Congenital CMV – vestibular symptoms may not be present for many years; alternatively – a child may have delay in walking
    - Congenital CMV – vestibular function may be normal, may have unilateral or bilateral peripheral hypofunction
Tests: Calorics – Unilateral weakness (if one ear affected)
Rotational Chair – asymmetry
ECOG – SP:AP ratio >0.45 (seen in 2/3 of pts w/ Meniere’s)

Otitis Media (acute) and Complications: see Miyamoto paper

Ototoxicity:
  - Shown to or alleged to produce vestibular ototoxicity: aminoglycosides, loop diuretics, salicylates, quinidine, barbiturates, various antibiotics (minocycline, erythromycin, polymixin, chloramphenicol) and certain chemotherapeutic agents (cisplatin, nitrogen, mustard.)
  - Affects both VOR and VSR
  - High-freq SNHL (destroys outer hair cells in basal turn of cochlea)
  - Disequilibrium, ataxic gait, stumble easily, lose balance w/ quick turns
  - Pts may navigate by holding onto wall or furniture
  - Oscillopsia w/ severe damage – secondary to loss of VOR; these pts have balance problems when trying to walk in darkness or on uneven surfaces
Tests: Rotational Chair (VOR) – reduced gains, reduced time constants; CDP – falls on SOT 6
Perilymph Fistula:
- direct injury to labyrinth or barotrauma
- imbalance, intermittent dizziness, or vertigo
- progressive SNHL
- tinnitus
- nystagmus
- unilateral or bilateral
- Positive fistula test
- CT may have congenital abnormalities (eg Mondini)
- Definitive diagnosis: MEE

Superior Canal Dehiscence Syndrome:
- Vertigo and oscillopsia induced by loud sounds (Tulio phenomenon), pressure changes in the EAC (Hennebert’s sing) or Valsalva
- Vertical oscillopsia, unsteadiness
- c/o autophonia, hearing pulse or their own eyes move
- vertical-torsional eye movements evoked by sound or pressure stimuli
- Apparent CHL – mimicking otosclerosis: air-bone gaps are greatest at lower frequencies and bone conduction thresholds “better” than 0 dB
- HRCT – can see dehiscence
Tests: VEMPS – low threshold, high amplitude

Temporal Bone Fractures:
- Transverse fractures may violate IAC, SCCs, vestibule, cochlea or FN
- Hearing loss (usually immediate and severe but can be progressive if PLF or pt develops endolymphatic hydrops)
- theoretical mechanisms of injury:
  - Avulsion of CN VII
  - Disruption of membranous labyrinth
  - Vascular vasospasm, thrombosis or hemorrhage
  - Disruption of endosteum of round or oval window resulting in PLF
  - Fracture across vestibular aqueduct – occlusion and secondary endolymphatic hydrops
- Vertigo, nausea/vomiting
- Spontaneous nystagmus
- HRCT

Usher’s Syndrome:
- Retinitis pigmentosa, bilateral SNHL, ± Vestibular dysfunction, ataxia
- AR
- High degree of clinic heterogeneity
- Type I: profound congenital HL, Vestibular dysfunction, retinal degeneration beginning in childhood
  - Type IB: peripheral vestibular dysfunction, absence of VOR

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• Type II: mod to severe congenital HL, normal vestibular function, late onset retinitis pigmentosa
• Type III: progressive HL
• Ophthalmologists – Electroretinography

Tests: Type I: ENG, Rotational Chair – absence of post-rotary nystagmus

Vestibular Neuronitis and Labyrinthitis:
• Sudden, unilateral, peripheral vestibular deficit
• vertigo – lasts for few days; unsteadiness may persist
• onset frequently after a URI
• acute hearing loss (labyrinthitis only)
• horizontal nystagmus beating towards healthy ear
• viral etiology – HSV, measles, mumps
• predominantly damages superior branch of VN
• utricular dysfunction seen in both
• main difference: saccule loss primarily w/ labyrinthitis

Tests: Calorics – decreased response affected ear
Rotational chair – greater h-VOR asymmetry in labyrinthitis than neuritis
VEMP – absent in approximately 1/3 of pts
SVH – deviated from horizontal by the affected side was down; Three months later, this deviation was greater if VEMPS were absent

Waardenburg’s Syndrome:
• Vestibular complaints in 72% of Type I and II
• Vertigo, imbalance or dizziness
• AD with variable penetrance
• Type I: Hearing loss, dystopia canthorum (lateral displacement of inner canthi), hypertrichosis of eyebrows, white forelock, skin pigmentary changes, heterochromia irides,
• Type II: same as Type I except dystopia canthorum
• Of those subjects w/ vestibular c/o: 77% had abnormal vestibulo-ocular tests (calorics, vestibular autorotation), 57% had abnormal vestibulo-spinal function tests, and 59% had abnormal ECOG (SP:AP > .40)
• CT: enlargement of the vestibular aqueduct and the upper vestibule, narrowing of the internal auditory canal porus, and hypoplasia of the modiolus

Whiplash Injury:
• Vertigo, dizziness
• Tinnitus
• ± Hearing loss
• Visual disturbances
• rapid flexion-extension of head results in cervical muscles spasm, impairing blood flow through the vertebro-basilar circulation

Tests: Positional nystagmus w/ head hyperextended; COR gains increased (likely secondary to reduced neck movements.)
ENG findings: spontaneous, positional nystagmus, gaze nystagmus, disturbances of smooth pursuits, and optokinetics. VOR abnormalities on both rotational chair and calorics. SVV abnormalities and saccadic impairments may be seen up to 1 yr out.

References:


Acute Dizziness without Hearing Loss

Benign Paroxysmal Positional Vertigo (BPPV):
- Classic description is of the posterior canal
- Sudden attacks of vertigo, nausea and vomiting triggered by head movements
- Dix-Hallpike maneuver: latency of few seconds before onset of rotary nystagmus. Nystagmus is of limited duration and fatigues w/ repeated provocation
- Uncommon in children

Migraines/Migraine Variants:
- Most common etiology of pediatric dizziness
- Paroxysmal ischemia of the vestibular nuclei
- Infants: Paroxysmal Torticollis
  - Avg age 18 mo
  - Ischemia of Inferior VN
  - w/ or w/o pallor, vomiting or behavioral changes
  - Episodic torticollis lasts between 4 hrs and 4 days
  - Frequency and duration of episodes decline as child gets older
- Young Children: Benign Paroxysmal Vertigo of Childhood (BPV)
  - Begins before age 5 and resolves within 2 yrs of onset
  - Rotary vertigo lasting seconds to minutes (1-5 min)
  - Autonimic signs/symptoms: pallor, nausea, vomiting, sweating
  - No headaches, tinnitus or hypoacusis
  - Sometimes fear, inconsolable crying
  - Not associated w/ head position
  - Resolves spontaneously and may occur several times/wk
- Older Children: Paroxymal Vertigo/Migraine Variant:
  - Mean age of onset 8 yrs
  - Vertigo lasting minutes
  - Pallor, nausea, feeling unwell, noise intolerance, light intolerance, anorexia, and vomiting
  - Positive FH of migraines
  - Some also have other migraine related symptoms: abdominal migraines, recurrent limb pain, cyclic vomiting
  - Triggers: fatigue, stress, missing a meal, travel, climate, bright lights, lack of sleep
  - symptoms often improve or resolve w/ sleep
- Older Children and Adolescents: Vertebrobasilar migraine/Basilar A. Migraine:
  - vertigo, tinnitus, decreased hearing, ataxia, dysarthria, visual symptoms, diplopia, paresthesias, headaches, decreased level of consciousness

Postural Orthostatic Tachycardia Syndrome (POTS):
- Symptoms: near syncope, lightheadedness, vertigo, TIAs, tachycardia, weakness, fatigue, exercise intolerance
- symptoms caused by transient alterations in autonomic tone that produce neurocardiogenic hypotension and bradycardia
• Children and adolescents
Tests:  Tilt table testing

Post-traumatic Epilepsy/ Epileptic Vertigo:
• 5-7% of closed heard injuries
• damage to superior temporal lobe, middle temporal gyri and supramarginal and angular gyri
• vertigo or dizziness
• normal hearing
• central vestibular system: frontal, parietal and temporal lobes; these areas are multisensory integration units rather than pure sensory cortex
• Epileptic discharges may present as vertigo
• Seizure may stem from pursuit eye movements rather than vestibular dysfunction
Tests:
EEG – abnormal; may need to do in sleep deprived state (document temporo-parietal spikes)
ENG – directional preponderance ± labyrinthine preponderance

Vestibular Neuronitis
See vestibular labyrinthitis in previous section

References:
Chronic dizziness with hearing loss

Autoimmune:
See acute dizziness with hearing loss section

Otitis Media (Complications):
See acute dizziness with hearing loss section

Ototoxicity:
See acute dizziness with hearing loss section

Acoustic Neuroma (Retrocochlear Lesions):
• Most common posterior fossa tumor in children
• Associated with NF II -- Diagnostic criteria: (from Miyamoto)
  o Bilateral vestibular schwannomas
  o Parent, sibling or child w/ NF2 plus
    ▪ Unilateral vestibular schwannoma
    ▪ Meningioma, glioma, neurofibroma or posterior subcapsular lens opacity
  o Unilateral vestibular schwannoma plus one in second entry above
  o Multiple meningiomas plus unilateral vestibular schwannoma, glioma, neurofibroma, schwannoma, posterior subcapsular lens opacity, cerebral calcifications
• Asymmetrical hearing loss – high-freq SNHL
• Speech Discrim disproportionate to PTA
• ABR: abnormal waveform or wave V latency ≥ 0.2 msec
• MRI w/ Gad – detection of tumors as small as 2 mm

References:
Chronic dizziness without hearing loss

Chiari I Malformation:
- Common clinical manifestations: torticollis, opisthotonus (titanic spasm of back muscles – trunk is arched forward, head and lower limbs backward), headaches, vocal cord paralysis, apnea, swallowing difficulties, ataxia,
- Variable vestibular symptoms and signs: oscillopsia, diplopia, blurred vision and vertigo
- Horizontal or downbeat (most common) nystagmus, jerky pursuit, Abducens palsy, increased blind spot; other types of nystagmus may also be seen
- The result of oculomotor dysfunction and central vestibular dysfunction (including vestibulocerebellum)
- Diagnosis: MRI – herniation of cerebellar tonsils
- Significant improvement and/or resolution of symptoms w/ posterior fossa decompression

Multiple Sclerosis:
- Progressive Demyelinating disease characterized by recurrent episodes of demyelination separated by time and space
- Course and severity – highly variable
- Mean age of onset - 28 yrs
- Women > Men
- Through course of disease can affect vestibulo-spinal tract
- Disequilibrium, balance dysfunction, dizziness, ataxia, nausea/vomiting, double vision, loss of vision, Intranuclear ophthalmoplegia, nystagmus
- Variable other neurologic symptoms
- MRI – white matter changes
Tests: VEMP – prolonged p13 and N23 latencies; SVV – many will have large deviations even w/o vestibular symptoms;

Ocular Abnormality:
- In one study, 5% of 523 children referred for vestibular testing had only an ocular disorder
- Sx usually present in children > 6yrs (activities involving prolonged attention)
- Sx at end of day or w/ fatigue, w/ prolonged use of computers/televisions, sometimes related to walking or rapid head movements
- Sx: sensation of rotation (vertigo), displacement of the environment, rolling, headaches, nausea/vomiting,
- Diagnoses: Refractive errors (myopia, hyperopia or astigmatism), amblyopia (unilateral loss of vision without an organic lesion), latent strabismus, convergence insufficiency

Post-concussive:
See labyrinthine concussion in acute dizziness with hearing loss section
Familial Periodic Cerebellar Ataxia/Vertigo, Episodic Ataxia Type 2:

- AD – variable clinical phenotype b/w and within families
- Bouts of vertigo, ataxia, dysarthria, nystagmus, nausea, vomiting and fatigue.
- Lasts ½ hour to 2 hrs
- About half of pts have associated migraine headaches
- Some have hemiplegia, epilepsy
- Triggers: emotional stress, exercise; occasionally: heat, coffee, small amts of alcohol, bright sunshine
- Symptoms relieved by sleep
- Onset before age 20 (mean age 12
- Neuro exam – normal to cerebellar ataxia
- Mutation in CACNA1A gene on chromosome 19p13 (sequence analysis – diagnostic)
- MRI: normal to atrophy of cerebellar vermis
- May be due to defect in cerebella pH homeostasis

Tests: Oculomotor studies: interictal gaze-evoked, rebound and downbeat nystagmus; abnormal saccades, ocular fixation and postural stability; may see square-wave jerks with lateral gaze, hypometric saccades, and both horizontal and vertical components during diagonal saccades. Reduced saccadic peak velocity may be earliest manifestation.

Vertebro-basilar insufficiency:

- Lateral medullary infarction (Wallenberg’s Syndrome)/cerebellar infarction
  - Occlusion of vertebral a or PICA or one of it’s branches
- Can also occur secondary to arterial dissection
- Vertigo, ataxia, dysarthria and headache
- Diverse types of nystagmus: horizontal, horizonto-rotary, torsional, see-saw and others have been described.
  - Wallenberg: Eyes open, horizontal nystagmus beats away from side of lesion
- Klippel-Feil – recurrent vertebrobasilar embolism


References:


Bedside Exam

- Basic head and neck exam
- Cranial nerves
- Ears
  - Fistula test (symptoms and nystagmus with pneumatic otoscopy)
  - Tuning forks
- Eyes
  - EOM (30° R and L)
  - Spontaneous nystagmus – w/ and w/o vision
    - Pt fixates on target about 4 ft away – look for nystagmus
    - W/ VNG googles – monitor for nystagmus
- Head Impulse Test (Halmagyi head thrust)
  - Gently grasp pts head on both sides and tilt forward 30° (LSCC coplanar w/ ground)
  - Ask pt to focus on your nose as examiner turns head 15-20 degrees to one side then other rapidly and abruptly (>2000 deg/sec) – observe eyes
    - Normal – eyes move almost exactly 180° out of phase w/ head
    - Peripheral vestibular d/o on left side – head turned to left, eyes do a catch up saccade to the right
- Head Shake
  - Pt wears VNG or Frenzels (eliminate visual fixation)
  - Pt shakes his/her head back and forth for 10-30 cycles – look for post-head shake nystagmus (transient nystagmus)
    - Peripheral lesions – fast phase of nystagmus toward contralesional ear
    - Central lesions – prolonged, vertical or disconjugate nystagmus
- Valsalva-induced nystagmus
  - VNG or Frenzels
  - Valsalva 1: increase air pressure in sinuses and ME: pt takes a deep breath, pinches nose and closes mouth tightly – then blows as if equalizing pressure in the ears when descending in an airplane
  - Valsalva 2: increase venous pressure in cranium: strain against a closed glottis and lips as if pressuring lungs to stabilize trunk while lifting a hvy weight
  - Nystagmus seen w/ Arnold-Chiari malformation, PLF and SCCD
    - Fast phase towards site of lesion
- Cerebellar Signs
- Dix-Hallpike
- Gait
- Tandem gait
- Romberg
- Fukuda Stepping Test
  - Pts feet should be shoulder width apart, eyes closed and arms outstretched.
  - Have pt march in place – bringing knees as high as poss
  - Peripheral vestibular d/o: Turn 45° toward site of lesion