Clinical Correlation for hearing and vestibular systems

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Anatomy/Clinical Correlation
Step by step
- External ear
- Middle ear
- Inner ear
- Vestibular and cochlear nerve
- Central structures

External Ear
- Focuses sound
- Accumulates Wax
- Place to put ear-rings

Common things that can go wrong with the external ear
- Blockages
  - Wax
  - Foreign bodies
  - Infection
- Holes
  - Mastoid cavity

Ear Wax

Foreign Bodies
- 12 year old boy – “I put a bug in my ear and nothing came out”
- Otoscope shows insect stuck to drum
- Much worse –
  - Beans
  - Beads
  - Pebbles
Otitis Externa

- Swimmer's ear
- Painful condition
- Treated with antibiotic/steroid ear drops
- Best to use non-ototoxic drop

Common things that go wrong with the external ear

- Ear drum problems
  - Holes (perforations)
    - Tubes
    - Trauma
  - Infections (e.g. bullous myringitis, herpes)
  - Scarring/stiffening (tympanosclerosis)

Q-tip injury (Perforation)

- Ouch!
- Temporary hearing loss and pain
- About 10db loss with perforation (0db=perfect, 100db=deaf)

Wake up stuff

- Q tips are actually rarely a cause of perforations
- Pencils, safety pins, are more dangerous
- Japanese culture does things differently — "mimikaki" used by loved ones to clean out wax from partner's ears.

http://japundit.com/archives/category/mimikaki/

Chronic otitis media

- Usually perforation
- Reduced hearing
- Drainage

Ear drums in middle ear disease

- Bullous Myringitis
- Ventilation tube
Middle Ear -- anatomy

- Mechanical Transformer for sound
- Full of air
- Pops on airplanes and elevators
- Stapedius and tensor tympani

Things that can go wrong in the middle ear

- Blockages
  - Infections
  - Otosclerosis
  - Tumors
- Holes
  - Perilymph fistula
  - Cholesteatoma
- Disconnections
  - Ossicular chain discontinuity

Conductive Hearing Loss
Sign of middle ear disease

- Moderately common
- Sound is not transmitted into inner ear.
- Diagnosis via air-bone gap.
- Hearing aids work well
- Treatment is often available that will restore hearing.

Case example of conductive hearing loss (Otosclerosis)

30 Y/O WM with low frequency, slowly progressive bilateral hearing loss over years. Hearing was initially easily corrected with hearing aid. Now is finding it difficult to follow conversations.
**Otosclerosis: summary**

- Physiology -- bone stiffens ossicular chain.
- Reduces "air" conduction.
- NEW: Genetic link --
  - osteogenesis imperfecta and measles
- Diagnosis: Audiogram shows low frequency conductive loss with "air-bone gap".
- Make sure it isn't just wax or fluid.

- Treatment: Watchful waiting, hearing aid, stapes surgery.

**Glomus Tumor: another conductive problem**

**Diving injuries**

**Perilymph Fistula**

- Both middle and inner ear
  - Blowout due to pressure (Scuba), explosion, trauma
  - Dizziness and hearing loss
  - Fix with a patch

**Inner Ear Disorders**

**Membranous Labyrinth**

MRI of inner ear
Superior Canal Dehiscence
- Opening between top of superior canal and dura
- Tullio’s phenomenon
- Pressure sensitivity
- Valsalva produces nystagmus

Disorders of the canal mechanics
- Cogan’s syndrome
  - Autoimmune inner ear disease
  - High sed rate, interstitial keratitis, deafness
  - Canal is plugged by fibrous tissue
- Canal Fistula
  - Opening in the canal = SCD
- BPPV

BPPV Mechanisms
- canithiasis (animation)

Brandt - Daroff Exercises: Method
- 30 sec each position
- 5 cycles/set (10 minutes total)
- 3 sets/day, 42 sets in total
- 2 weeks of daily exercises

Epley Maneuver (a.k.a. CRP)
- For classic (PC) BPPV
- Move debris out of posterior canal into vestibule
- Keep debris there for 2 days till it is dissolved (sleep sitting up)

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Epley Maneuver: Method

Epley Maneuver: (animation)

Sleep semi-recumbent for 2 nights after the Epley Maneuver

Results of Epley Maneuver

- Most authors report 75% cure from a single treatment, and >95% from repeated treatments.¹
- About 38% of patients recover with no treatment or sham treatment by 2 weeks.² ³
- The Epley usually is the treatment of choice for classic BPPV.


Vestibular Hair Cells

Disorders of Vestibular Hair Cells

- Otoxicity – aminoglycosides selectively damage vestibular hair cells
48 YO Airline stewardess developed an ingrown toe-nail infection. She underwent a course of gentamicin and vancomycin. 12 days after starting therapy she developed imbalance. 21 days after starting, she was "staggering like a drunk person". Meclizine was prescribed. Gentamicin was stopped on day 29. One year later, the patient had persistent imbalance, visual symptoms, and had not returned to work. Hearing is normal. She presently does volunteer work.

Bedside diagnosis of Gentamicin toxicity

- Romberg – unable ECTR.
- DIE test
  - Distance vision with head still
  - Distance vision with head moving (horizontal or vertical, 1-2 Hz)
    - Normal: 0-2 lines change.
    - Abnormal: 4-7 lines drop with movement

Gentamicin toxicity

- Causes permanent imbalance and oscillopsia
- Hair cells do not regenerate
- No medical treatment is available
- Physical therapy is helpful to promote compensation

Otolith disorders

- Not much is known
- Tilt disorders?
- Rocking?
Otolith assessment

- VEMP – vestibular evoked myogenic potential. Only available test of otolith function (saccule)

Cochlear hair cells

- Arranged in a tonotopic spiral
- High frequencies at base
- Spiral Ganglion wound around cochlea

Direction of view
From medial

Otoacoustic Emissions (OAE)

- Test of outer hair cells (“outies”)
- Stimulate ear with sound
- Listen to ear and average it’s response
- Good for malingering
- Good for ototoxicity

VEMP

Endolymph and Perilymph

Hair cell disorders cause sensorineural hearing loss

- Noise induced hearing loss
- Age related hearing loss
- Meniere’s disease
- Ototoxicity (e.g. Aspirin)
- Vascular
- Autoimmune inner ear disease
Bedside Examination of hearing

- Comprehension of speech – “eh, what was that?”
- Tuning forks: classic tests (512 is best)
  - Rinne: Bone > Air? Conductive pattern
  - Weber: hear tone better on one side or another
- Rubbed fingers, watch, whisper (high freq)
  - Screen for hearing loss
  - Older people nearly always have trouble with higher frequencies.

Cochlear Hearing loss

- Bone and Air conduction reduced
- Commensurate reduction speech comprehension
- Normal tympanogram
- Normal acoustic reflex
- Recruitment and/or diplacusis may occur

Meniere’s Disease

Low frequency sensorineural hearing loss is typical of early Meniere’s disease

Case

- 45 year old woman, speech pathologist
- Troubled by spells of dizziness, nausea, vomiting, gradually increasing hearing loss
- Many years of symptoms

Meniere’s Disease

Hydrops

Normal
Meniere’s disease

- Eventually people with Meniere’s disease go deaf on their “bad” ear.
- Meniere’s disease is a chronic disease.

Treatment of Meniere’s

- Medical – salt restriction, diuretic, vestibular suppressants
- Surgical – various destructive options
  - Gentamicin injection is currently favored
  - Labyrinthectomy if deaf
  - Vestibular neuromyectomy if gentamicin fails
- Strange devices and placebos
  - Meniett

Case

- Grandma is brought in by her daughter
- She recently moved in with her daughter’s family, and she turns up the TV too high for the kids.
- When you talk to her, she smiles and asks you to repeat, while closely watching your lips

Mild Presbyacusis

Grandma needs a hearing aid

Sensorineural Hearing Loss

- Most common - 23% of population > 65
- Diagnosis made by hearing loss but no air-bone gap
- Normal tympanogram
- Hearing aids may not work
- Treatments are mainly aimed at preventing further damage and making do with what is left.
Case
- Local commodities trader having trouble understanding people yelling things at him in the "pit"
- Also has developed ringing in his ears

Noise induced hearing loss
- Sensorineural hearing subtype
- Usually a notch pattern
- Either very loud noise (explosion) or repetitive
- Preventable with hearing protection

Eighth Cranial Nerve

Things that can go wrong with the ganglia or the nerve
- Vestibular neuronitis
- Sudden hearing loss (the viral type)
- Acoustic neuroma
- Microvascular compression

Vestibular neuritis
- Sudden severe vertigo
- Lasts for several weeks
- Usually vomiting/imbalance
- Attributed to herpes virus (herpes-1)
- Symptomatic treatment (initially)
- Recently shown that steroids are helpful
- May use PT after 2 months to encourage compensation

Sudden hearing loss (SHL)
- Sudden unilateral deafness
- May be vascular or viral (mostly viral)
- About 50% recover by 6 weeks
- No compelling evidence for steroids but most use them anyway
- Hearing aid if don’t recover
Case

- ML - 21 year old man
- Gradually increasing hearing loss on right side
- Developed a headache and had a CT scan done in ER
- Huge tumor was seen

Acoustic Neuroma

Treatment of Acoustic Neuroma

- Watchful waiting (about 25%)
- Operative removal (about 50%) – losing ground
- Gamma Knife (about 25%) – gaining ground because effective and noninvasive

Microvascular compression and vestibular neuralgia

- Irritation of vestibular nerve
- Quick spins
- Motion sensitivity
- May follow 8th nerve surgery
- Wastebasket syndrome in some cases?

Diagnosis of MVC

- Normal ENG/Audiogram
- Abnormal BAER or VEMP
- May have nystagmus on hyperventilation
- Response to Trileptal
- 3D CISS MRI

Central Vestibular Wiring
Vestibular Nucleus Commissures

Push-Pull organization
Bilateral symmetry provides redundancy
Vestibular nucleus lesions – “perverted nystagmus”

Central balance disorders

- Brainstem
- Cerebellum

Physical therapy may be helpful but prognosis is poorer and recovery may be much slower than in peripheral injuries.

Things that can go wrong with the brainstem or cerebellum

- Multiple sclerosis
- Stroke
- Tumor/AVM
- Chiari malformation

Multiple Sclerosis

- No single pattern
- Multiple lesions distributed in time and space

Posterior Fossa stroke

- 50 year old surgeon developed vertigo and unsteadiness
- Continued to operate for a week before seeking medical attention but wife wouldn’t let him drive.
- PICA stroke seen on MRI

You can’t be sure from your exam that patient doesn’t have a stroke

- Cerebellar/brainstem disease can be indistinguishable from peripheral vestibular disorders
Wallenberg’s Syndrome
Lateral Medullary

- PICA territory
- Supplies lateral 50% of medulla and inferior 1/3 cerebellum
- Ipsilateral cerebellar signs
- Vertigo (vestibular nucleus)
- Contralateral pain and temp (STT)
- Dysphagia/Dysphonia (Nucleus ambiguus)
- Ptosis/Miosis

AICA syndrome

- AICA supplies pons, cerebellum, 8th nerve
- Facial weakness
- Vertigo/hearing loss
- Incoordination

Vascular supply of Labyrinth

SCA Syndrome

- SCA supplies superior cerebellum, midbrain
- Ataxia and diplopia

Hemorrhagic cerebellar stroke

- Causes
  - Hypertension, tumors, trauma
- Differences from ischemic stroke
  - Much more dangerous
  - Can swell and compress brainstem
  - Surgery is common to decompress
Hemorrhagic cerebellar stroke

- Signs/Symptoms
  - Ipsilateral or diffuse cerebellar signs
  - Occipital headache
  - Signs of increased ICP
    - Projectile vomiting
    - Confusion

Chiari Malformation: Diagnosis

- Cerebellar tonsils herniate downward
- Adult onset
- Straining or coughing produces headache or fainting
- Unsteadiness
- Nystagmus

Oculomotor signs in Chiari

- Downbeating nystagmus (1/3 Chiari)
- Face paraesthesis (Broca's)
- Alternating skew deviation
- Various odd central nystagmus
  - Divergence nystagmus
  - Central positional nystagmus
  - Valsalva induced nystagmus

Chiari Malformation: Treatment

- Watchful waiting (about 90%)
- Suboccipital decompression

Central Auditory Pathways

Ventral cochlear nucleus pathway traverses SO – determines direction

Dorsal cochlear nucleus pathway – determines quality of sound

Output from cochlear nucleus ascends brainstem, synapse in inferior colliculus, terminate in auditory cortex
Central Auditory Disorders
- Transient brainstem deafness
- Speech impaired greater than pure tones
- Auditory hallucinations
- Normal pure tone and bone
- Normal tympanogram
- Unable to understand speech in noise or competing messages

Inferior colliculus lesions
- Presented with pure word deafness – can hear sounds, but not understand speech
- HIV positive
- CN8 lymphoma
- Bilateral IC lesions

Auditory Cortex Lesion
- Older man
- Heard rock music in middle of night
- Went next door to ask them to turn it off
- Everyone was asleep

Cortical Deafness

Cases
We are almost done

Case 1
- A 30 year old Chicago Park District Worker came in because of dizziness.
- He fell off of a truck two years ago, hit his head, and now he becomes very dizzy whenever he lies down.
- He has been sleeping in a chair for two years.
- What is the most likely reason for his positional vertigo?

Lessink et al., 2005. Sudden deafness from Stroke. Neurology, 64(10), p1817
Case 2

- A 55 year old worker at a local steel mill comes in because of dizziness and hearing disturbance. Every month or two, he has an attack in which one ear gets plugged up, he gets a loud roaring noise, and everything starts to spin for several hours.
- He is concerned that he may be unsafe to work.
- What is the most likely cause of his episodic symptoms?

Case 3

- A 60 year old diabetic was on peritoneal dialysis while waiting for a kidney transplant. She got peritonitis and was given an antibiotic to put into the dialysate. After 2 weeks, her balance became much worse and she noticed that she couldn't see very well when her head was moving.
- What is the diagnosis? Treatment?

Case 4

- A 75 year old diabetic awoke one morning dizzy.
- Accompanying the vertigo was a droopy eye lid, and clumsiness on one side. There was also dysphagia and a hoarse voice. There were no hearing symptoms.
- What artery is the most likely one to cause this stroke?