Bell's Palsy and Hearing Loss

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Otologic Problems

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Great Website

UTMB Galveston - Otolaryngology Department – Dr. Quinn Grand Rounds

ENT resident powerpoint presentations – very detailed

Much better than wikepedia

http://www.utmb.edu/otoref/gmrs/gmrsindex.html
Anatomy

Otologic Problems
How often do you see them??  Underestimate
Bell’s Palsy/Facial paralysis – Rare (diagnosis)
Hearing Loss – common – sudden (diagnosis)
Otitis – common – OME, OE, TMJ, Qtips
Otorrhea – common – OME, OE, sweat
Vertigo – common – ear or not (dizzy)
Aural Fullness – common – ETD, Meniere’s
Tinnitus – common, SNHL, pulsatile

Otologic Urgencies/Emergencies
How often do you see them??
Sudden Hearing Loss
Vertigo – (Stroke)
Facial paralysis (Stroke, AOM) – Bell’s not urgency – but if surgery to be considered – quick referral
Massive Bleeding from the ear
Meningitis/abscess from ear infection or CSF leak
Acute decompensation from Tumor
Suicidal Tinnitus
**Otologic (Mis)Diagnoses**

Sudden Hearing Loss – not OME – TM appearance, tuning fork
ETD – in obese adults – likely to be CSF or NP mass
BPPV or Meniere’s - Vertigo from Stroke
Bell’s Palsy - Facial paralysis from Stroke, AOM, cancer – parotid/skin
Otolgia – AOM – no fever, no bulging TM, no hearing loss – no need for antibiotics

**Otologic History**

Hearing Loss
Otalgia
Otorrhea
Vertigo
Aural Fullness
Tinnitus
Facial Paralysis

**Facial Paralysis**

Onset
Progression
Unilateral or Bilateral (Melkerson-Rosenthal, Lyme)
Rash (Lyme Disease)
Cholesteatoma
Parotid Mass
Ramsey Hunt Syndrome
MOE
Acoustic Neuroma
Bell’s Palsy – only after ruling out other causes
Facial Nerve

Anatomy

- Contains 7,000-10,000 fibers
- Nuclei
  - Somatic – Motor
  - Taste – Tractus solitarius
  - Secretomotor – Superior salivatory
- Segments
  - Intracranial (cisternal)
  - Meatal
  - Labyrinthine
  - Petrosal
  - Mastoid
  - Extratemporal

House Brackman grading system

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Normal facial function</td>
</tr>
<tr>
<td>II</td>
<td>Mild – Slight synkinesis/weakness</td>
</tr>
<tr>
<td>III</td>
<td>Moderate – Complete eye closure, noticeable synkinesis, slight forehead movement</td>
</tr>
<tr>
<td>IV</td>
<td>Moderately Severe – Incomplete eye closure, symmetry at rest, no forehead movement</td>
</tr>
<tr>
<td>V</td>
<td>Severe – Asymmetry at rest, barely noticeable motion</td>
</tr>
<tr>
<td>VI</td>
<td>Total – No movement</td>
</tr>
</tbody>
</table>
Bell’s Palsy

• Facial paralysis
  – Acute onset, limited duration, minimal symptoms, spontaneous recovery
  – Idiopathic in past
  – Diagnosis of exclusion
  – Most common diagnosis of acute facial paralysis

Pathophysiology

• HSV viral reactivation leading to damage of facial nerve
  – Neuropraxia – no axonal discontinuity
  – Axonotmesis
    • Wallerian degeneration (distal to lesion)
    • Axonoplasmic disruption, endoneurial sheaths intact
  – Neurotmesis
    • Wallerian degeneration (distal to lesion)
    • Axon disrupted, loss of tubules, support cells destroyed
Evaluation

- Careful history – timing
  - Associated symptoms (pain, dysgeusia)
  - SNHL, vesicles, severe pain
  - Trauma, acute or chronic OM, recurrent exposure
- Physical exam
- Audiometry
- CT/MRI/other
- Topographic
- Electrophysiology

Electrophysiology

- ENoG (electroneuronography)
  - Most accurate, objective
  - Records summation potential (CAP)
  - Degree of degeneration is directly proportional to amplitudes of measured potentials
  - Done after Wallerian degeneration starts (3-4 days)
  - Compare each day

Electrophysiology

- ENoG
  - Essen (1977) – over 90% degeneration on ENoG prognosis worsens
    - 90-97%: 30% recovery fully
    - 98-99%: 14% recovery fully
    - 100%: none recovered fully
  - Fisch (1981)
    - 50% with 95-100% degeneration by 14 days have poor recovery
    - High likelihood of further degeneration if reaches 90%
    - Thus, if ENoG reaches 90% within 2 weeks: 50-50 recovery
Medical Management

• Eye protection

• Steroids
  – Stankiewitz (1987) – no efficacy
  – Austin (1993) – randomized, double blind, placebo controlled study
    • Improvement in grade with prednisone
    • All with prednisone (House 1-2)
    • 17% without House 3 (statistically significant)
    • Trend towards denervation protection

Medical Management

• Antivirals
  – Adour (1996) – double blind
    • Only 20% progressed to complete paralysis
    • Acyclovir had less degrees of facial weakness
    • Acyclovir had lower incidence of House 3-5

• Conclusions

Medical Management

• Rapid Referral to Otologist (not general ENT)

• Me – in case patient interested and could benefit from surgery
Surgical Management

- Gantz (1999) – multi-institutional review
  - Assess if patients with degeneration over 90% within 14 days would benefit
  - Middle cranial fossa (meatal foramen to tympanic segment)
  - If conductive block not identified (6%) – transmastoid added
  - 92% with surgery recovered to House 1-2
  - 45% without surgery to House 1-2

Herpes Zoster Oticus: Symptoms

- Early: burning pain in one ear, headache, malaise and fever
- Late (3 to 7 days): vesicles, facial paralysis
- Others – lyme disease, ...

Hearing Loss

Conductive
Sensorineural
Mixed

Who has and uses a tuning fork?????
Very inexpensive
Very helpful
Testing Hearing

Tuning forks  ******************
  Weber – toward conductive, away from sensorineural
  Rinne  - AC > BC normal, sensorineural
          - BC > AC conductive

Pure-tone audiometry
Spondaic word threshold
Monosyllable word discrimination
Masking
Auditory Brainstem Response
Otoacoustic Emissions
Tympanometry configurations

## Conductive

External ear
Tympanic membrane
Ossicles
Middle ear space

## External ear

Atresia
Tumor
Infection – acute, chronic
Foreign Body
AOE: Mild to Moderate Stage

- Progressive infection
- Symptoms
  - Pain
  - Increased pruritus
- Signs
  - Erythema
  - Increasing edema
  - Canal debris, discharge

AOE: Severe Stage

- Severe pain, worse with ear movement
- Signs
  - Lumen obliteration
  - Purulent otorrhea
  - Involvement of periauricular soft tissue

AOE: Treatment

- Most common pathogens: *P. aeruginosa* and *S. aureus*
- Four principles
  - Frequent canal cleaning
  - Topical antibiotics
  - Pain control
  - Instructions for prevention
Otomycosis: Signs

- Canal erythema
- Mild edema
- White, gray or black fungal debris

Otomycosis: Treatment

- Thorough cleaning and drying of canal
- Topical antifungals

Necrotizing External Otitis (NEO)

- Potentially lethal infection of EAC and surrounding structures
- Typically seen in diabetics and immunocompromised patients
- *Pseudomonas aeruginosa* is the usual culprit
**NEO: Symptoms**

- Poorly controlled diabetic with h/o OE
- Deep-seated aural pain
- Chronic otorrhea
- Aural fullness

**NEO: Signs**

- Inflammation and granulation
- Purulent secretions
- Occluded canal and obscured TM
- Cranial nerve involvement

**NEO: Diagnosis**

- Clinical findings
- Laboratory evidence
- Imaging
- Physician’s suspicion
- Cohen and Friedman – criteria from review
NEO: Treatment

- Intravenous antibiotics for at least 4 weeks
  - with serial gallium scans monthly
- Local canal debridement until healed
- Pain control
- Use of topical agents controversial
- Hyperbaric oxygen experimental
- Surgical debridement for refractory cases

NEO: Mortality

- Death rate essentially unchanged despite newer antibiotics (37% to 23%)
- Higher with multiple cranial neuropathies (60%)
- Recurrence not uncommon (9% to 27%)
- May recur up to 12 months after treatment

Tympanic Membrane - Middle Ear

Retraction
Perforation
Cholesteatoma (primary, acquired)
Trauma
Ossicular Discontinuity
Ossicular Fixation
Otosclerosis
Tumors (vascular, neural)
Congenital Ossicular Problems
Surgical decision
Otitis Media - Definition

Inflammation of the middle ear
May also involve inflammation of mastoid, petrous apex, and perilabyrinthine air cells

Otitis Media - Classification

• Acute OM - rapid onset of signs & sx, < 3 wk course
• Subacute OM - 3 wks to 3 mos
• Chronic OM - 3 mos or longer

OM - Epidemiology

• Increasing incidence?
• Increases after newborn period
• 2/3 with AOM by one year of age
• 1/2 with >3 episodes by three years
• most common in 6 - 11 mos
• Day care – increase risk
• Smoke exposure – increase risk
OM - Medical Conditions

- Cleft palate
  - decreases after repair
- Craniofacial disorders
  - Treacher-Collins
- Down’s syndrome
- Ciliary dysfunction
- Immune dysfunction
  - AIDS
  - steroids, chemo
  - IgG deficiency
- Obstruction
  - NG tubes
  - NT intubation
  - adenoids
  - malignancy

Microbiology

- PCN-resistant Strep
  - 1979 - 1.8%
  - 1992 - 41%
  - Altered PCN-binding proteins
  - Lysis defective
  - Age, day-care, and previous tx
- H. flu and M. catarrhalis
  - beta-lactamase production
  - All M. catarrhalis +
  - 45-50% H. flu

Diagnosis

- Pneumatic otoscopy is gold standard
  - Color - opaque, yellow, blue, red, pink
  - Position - bulging, retracted
  - Mobility - normal, hypomobile, neg pressure
  - Assoc pathology - perfs, cholesteatoma, retraction pockets
- Head & neck exam
Treatment - AOM

• Adults and older children - observation?
• Antibiotics - consider drug resistance patterns
  – Amoxil - not for β lactamase +
  – TMP-SMT - not for group A strep
  – Need high middle ear concentrations

Antibiotics – up to date ??

• First line
  – Amoxil - 60-90 mg/kg divided tid
  – Cefin - β lactam stable
  – Augmentin - β lactam stable
  – Bactrim, Pediazole

• Second line
  – Augmentin
  – Cefin
  – Rocephin
  – Macrolides - Zithromax, Biaxin

Treatment - Recurrent AOM

• Chemoprophylaxis – thumbs down
  – Sulfisoxazole, amoxicillin, ampicillin, pen
    – less efficacy for intermittent prophylaxis
• Myringotomy and tube insertion
  – decreased # and severity of AOM
  – otorrhea and other complications
  – may require prophylaxis if severe
• Adenoidectomy
  – 28% and 35% fewer episodes of AOM at first and second years
Treatment - OME

- MEE > 3 mos or assoc hearing loss, vertigo, frequency, ME pathology, discomfort
- Antibiotics
  - shown to be of benefit, 75% PCR + bacterial DNA
- Antibiotics + steroid
  - 23% improvement compared to abx alone
  - prednisone 1 mg/kg day x 7 days
  - varicella?
- Myringotomy & tympanostomy +/- adenoidectomy

Tympanostomy tube insertion

- Unresponsive OME >3 mos bil, or >6 mos uni, sooner if assoc hearing problems
- Recurrent MEE with excessive cumulative duration
- Recurrent AOM - >3/6 mos or >4/12 mos
- Eustachian tube dysfunction
- Suppurative complication

Complications of ETD, AOM

- Intratemporal
  - hearing loss
  - TM perforation
  - CSDM
  - retraction pockets
  - cholesteatoma
  - mastoiditis
  - petrosis
  - labyrinthitis
  - adhesive OM
  - tympanosclerosis
  - ossicular discontinuity and fixation
  - facial paralysis
  - cholesterol granuloma
  - necrotizing OE

- Intracranial
  - meningitis
  - extradural abscess
  - subdural empyema
  - focal encephalitis
  - brain abscess
  - lateral sinus thrombosis
  - otitic hydrocephalus
Cholesteatoma - Introduction

- Keratin-producing squamous epithelium in the middle ear, mastoid or petrous apex
- Johannes Müller (1838) coined the term
- “a pearly tumor of fat…among sheets of polyhedral cells”
- Exhibits independent growth, replaces mucosa, resorbs bone

Primary acquired cholesteatoma

[Image of primary acquired cholesteatoma]

Primary acquired cholesteatoma

[Image of primary acquired cholesteatoma]
Evaluation

- Physical Examination
  - Otomicroscopy
  - Posterosuperior retraction pocket with squam
  - Granulation from diseased bone
  - Aural polyps
  - Pneumatic otoscopy – positive fistula response suggests erosion into labyrinth
  - Cultures should be obtained in infected ears

Evaluation

- Audiology – usually conductive loss, may vary greatly; confirm with tuning forks
- Imaging
  - CT temporal bone – definitely obtain for revision cases, complications of chronic suppurative otitis media, sensorineural hearing loss, vestibular symptoms, other complications of cholesteatoma

Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Noise</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Hereditary</td>
<td>Drugs</td>
</tr>
<tr>
<td>- Toxic</td>
<td>Aminoglycosides</td>
</tr>
<tr>
<td>- Infectious</td>
<td>Loop Diuretics</td>
</tr>
<tr>
<td>- Spontaneous</td>
<td>Platinums</td>
</tr>
<tr>
<td>Vascular</td>
<td>NSAIDS</td>
</tr>
<tr>
<td>Viral</td>
<td>Autoimmune</td>
</tr>
<tr>
<td>Bacterial</td>
<td>Iatrogenic</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Tumors</td>
</tr>
<tr>
<td>Trauma</td>
<td></td>
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</table>
**Intro**

- Hearing loss affects 28 million Americans
- Sensorineural Hearing loss 17 million
- People with hearing loss suffer emotional, social and communicative dysfunction
- Only to increase with aging population
- Can’t hear, can’t understand (related ??)

**Waardenburg Syndrome**

![Image](image_url)

**FIG. 89-18. Waardenburg syndrome. This mother and daughter have Waardenburg syndrome type I. Both have hearing loss and dystopia canthorum. The child also has heterochromia irides.**

**Branchio-oto-renal Syndrome**

![Image](image_url)

**FIG. 89-19. Branchio-oto-renal syndrome. This 3 year old boy has bilateral supernumerary thumbs, microtia and pre-auricular pits, and anemia.**
# Usher Syndrome

- Prevalence of 3.5 per 100,000 population
- Affects 16,000 deaf and blind persons in the US
- Syndrome characterized by SNHL and retinitis pigmentosa
- Three subtypes divided based on severity of progression of the hearing loss and extent of vestibular involvement
  - Type 1: congenital bilateral profound HL and absent vestibular function
  - Type 2: moderate losses and normal vestibular function
  - Type 3: progressive HL and variable vestibular function
- Linkage analysis reveals 5 genes for type 1, 2 for type 2, and 1 for type 3
- Ophthalmologic evaluation essential, subnormal ERG patterns have been observed by age 2 to 3

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# Chronic NIHL

- Defining Characteristics

<table>
<thead>
<tr>
<th>Year</th>
<th>Frequency (Hz)</th>
<th>NITS (dB)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-2</td>
<td>500-2000</td>
<td>15</td>
</tr>
<tr>
<td>3-9</td>
<td>2000-4000</td>
<td>20</td>
</tr>
<tr>
<td>10-19</td>
<td>4000-8000</td>
<td>25</td>
</tr>
<tr>
<td>20-29</td>
<td>8000-12000</td>
<td>30</td>
</tr>
<tr>
<td>30-39</td>
<td>12000-20000</td>
<td>35</td>
</tr>
</tbody>
</table>

- Aminoglycosides
  - Streptomycin, kanamycin, neomycin, amikacin, gentamicin, tobramycin, sisomycin, netilmicin
  - Enter into inner ear by unknown mechanism
    - Secreted into the perilymph by spiral ligament
    - Diffuse through round window membrane
  - Eliminated by kidney
Loop Diuretics

- Ethacrinic acid, furosemide, bumetaside
- Clinically (6-7%)
  - Usually tinnitus, temporary and reversible
    SNHL, rare vertigo within minutes
  - High doses can cause permanent SNHL
  - Highest risk– coadministration of
    aminoglycosides

Salicylates and NSAIDS

- Most common OTC drugs in US
- Mechanism
  - Normal histology (no hair cell loss)
  - Decreased blood flow, decreased enzymes
- Clinically
  - Tonal, high frequency tinnitus (7-9 kHz)
  - Reversible mild to moderate SNHL (usually
    high frequency)– rarely permanent

Antineoplastic Agents

- Cisplatin
  - Incidence is high (62%-81%)
  - Pathologically
    - Outer hair cell degeneration
  - Clinically
    - Bilateral symmetric SNHL, usually high frequency–
      not reversible, cumulative
    - Risks factors– age extremes, cranial irradiation, high
      dose therapy, high cumulative dose
Conventional Hearing Aids

- Only 10-20% use hearing aids
- 4.5 million hearing aid users
- 12% who have them don’t wear them
- Only 58% “very satisfied”

BTE

- Attached to earmold
- Powerful
- Wide range of HL
- Can have open earmolds
- Less repair problems
- Less feedback
- Telecoils

ITE

- Fits in concha
- Microphone at ear canal level
- Mild to severe HL
ITC

• Only face sticks out into concha
• Wide range of HL
• Microphone at opening of ear canal
• Takes advantage of most of the auricle

CIC

• Hidden in canal
• Full effect of auricle
• Receiver very close to tympanic membrane so requires less amplification

Cochlear Implant

• Inner coil is placed in a cavity created in the skull during surgery
• Electrode array is placed through a cochleostomy into the scala tympani
• Multichannel, multielectrode implants take advantage of the tonotopic organization of the cochlea
How the Cochlear Implant Works

1. Microphone
2. Sound Processor
3. Cord
4. External Receiver
5. Internal Receiver
6. Electrode Array
7. Hearing Nerve

Pictures provided by Advanced Bionics

Who is a candidate?

• Adults:
  – Could be the person in your office really struggling with hearing aids
  – Keeps getting fit with aids, really expensive, not working really well
  – If patient is really focused on your face while communicating (lip-reading) – perhaps candidate
  – Increases dramatically with age
  – Don’t want audiologists making medical decisions for patients
  – Don’t want PCP making surgical decisions for patients
  – PLEASE REFER
Current FDA Guidelines

- **Adults:**
  - Bilateral severe to profound sensorineural hearing loss
  - No functional benefit from appropriately fit hearing aids (i.e., fit using real-ear measurements)
  - 50% or less scored on sentence recognition tests for the ear to be implanted and 60% or less in the non-implanted ear
  - Realistic expectations
  - Willingness to comply with the follow up schedule

- **Children:** 12 months to 17 years
  - Bilateral severe to profound sensorineural hearing loss
  - No functional benefit from appropriately fit hearing aids (i.e., fit using real-ear measurements)
  - Lack of progress in the development of auditory skills and spoken language with INTENSIVE auditory training
  - High motivation and realistic expectations from the family
  - Other existing medical conditions that will not interfere with the cochlear implant procedure

Who is a Candidate for a Cochlear Implant?

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Current CI Manufacturer’s

- Advanced Bionics Corporation
  - www.advancedbionics.com
- Cochlear Americas
  - www.cochlear.com
- Med El Corporation
  - www.medel.com/US

Advanced Bionics
Harmony BTE Sound Processor

Advanced Bionics
HiRes 90K
Cochlear Americas
Nucleus 5 Sound Processor

Freedom BTE and Bodyworn Controller
No more BTE/BW processor with the Freedom

Med El Maestro

The New MAESTRO™ Cochlear Implant System
Now available in twelve sensational colors.
AB – Listening Check

Now what?

- Surgery
  - 2-3 hour procedure
  - typically scheduled as outpatient

- Programming
  - Initial stimulation 3 weeks after surgery
  - 1 month, 3 month, 6 month, 12 month
  - After first year, return every 6-12 months
**Sudden SNHL**

- **Definition**: 30 dB or greater SNHL over at least three contiguous audiological frequencies occurring within 3 days or less

**Introduction**

- Suggested causes of Idiopathic SNHL (ISNHL)
  - Viral infections
  - Autoimmune
  - Vascular compromise
History

- Time course
- Associated symptoms
  - Vertigo/dizziness
  - Aural fullness
  - Tinnitus
- Ototoxic drug use
- Symptoms of URTIs
- H/O head trauma, straining, sneezing, nose blowing, intense noise exposure
- H/O flying or SCUBA diving

History

- PMH:
  - Autoimmune disorders
  - Vascular disease
  - Malignancies
  - Neurologic conditions
  - Hypercoagulable states
    - Sickle cell disease (African Americans)
- PSH: stapedectomy or other otologic surgeries

Physical Exam

- Complete H&N exam in everyone
  - Ear: r/o effusions, cholesteatoma, cerumen impaction
  - Weber/Rinne
  - Neurologic exam – cerebellar findings
    - Tandem gait
    - Romberg
    - Nose to finger, heel to shin
  - Vestibular – Dix-Hallpike test
Diagnostic Testing

- Audiogram
  - Pure tone
  - Speech discrimination
  - Tympanometry
  - Stapedial reflex
- Laboratory testing
  - CBC
  - ESR
  - RPR, VDRL
  - Lymphocyte transformation test
  - Western blot for antibodies to 68 KD protein

Diagnostic Testing

- MRI:
  - Rule out cerebellopontine angle tumors
  - Multiple sclerosis
  - Ischemic changes
- 13% of patients with acoustic tumors present with SHL
- 23% may recover hearing

Viral

- Current belief – viral cochleitis causes the majority of cases of ISSNHL
- 1983 – Wilson and colleagues
  - Viral seroconversion rates greater in patients with ISSNHL (63%) compared to control (40%)
    - Influenza B
    - Mumps
    - Rubeola
    - VZV
Treatment

- Therapy for ISSNHL is controversial
- Difficult to study
  - High spontaneous recovery rate
  - Low incidence
  - Makes validation of empiric treatment modalities difficult

Treatment

- Proposed treatment modalities
  - Anti-inflammatory – steroids, cytotoxic agents
  - Diuretics
  - Antiviral agents
  - Vasodilators
  - Volume expanders/hemodilutors
  - Defibrinogenators
  - *** Hypaque/Dextran (UIHC) ****

Conclusion

- SHL is devastating to patients
- Frustrating for physicians to dx and tx
- Thorough H&P
- Rule out treatable cause
- Directed labs, Audiogram MRI
- Discuss risks, benefits, and alternatives of treatment with the patient
- Treat the disorder aggressively
- Rehabilitate those whose hearing does not improve
- Follow patients for development of associated diseases and for contralateral ear disease
"Dizziness"

- Presyncopal faintness
- Loss of balance
- Light-headedness
- Psychologic disorders
- Vestibular disease

Dizziness

- BPPV
  - Acute, latent, and fatigable vertigo
  - Can occur any time following injury
  - Dix Hallpike
  - Epley Maneuver

BPPV - Epley

[Diagram: Steps for Epley Maneuver]
Meniere’s Disease

- Torok
  - 60-80% remission with all txs
- Silverstein
  - 59% remission at 2 years and 70% remission at 8 years in untreated group
- Ruckenstein
  - vestibular suppressants only group effect
  - other tx nonspecific and no better than placebo

CSF Otorrhea and Rhinorrhea

- Temporal bone Fractures are the most common cause of CSF Otorrhea
- Beta-2-transferrin
- HRCT

CSF Otorrhea and Rhinorrhea

- Management
  - Conservative therapy
  - Antibiotics
  - Surgery
CSF Rhinorrhea

- History and PE
- Unilateral watery rhinorrhea
- Increases with valsalva and posture
- May see leak/encephalocele with endoscope
- Collect fluid

Spontaneous CSF Otorrhea

- Beta-2-transferrin
- HRCT
- CT cisternogram
- MR cisternogram
- Surgical repair

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