Otology seminar
Perilymphatic fistula

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Perilymphatic space

- Space between bony and membranous portion of labyrinth
Perilymphatic space

- Perilymphatic space of **semicircular canal** --continuous with perilymphatic space of **vestibule**
- -continuous with perilymphatic space of **scala vestibule**
- -continuous with perilymphatic space of **scala tympani**(at helicotrema)
- All perilymphatic space open into each other
Perilymph

- Extracellular fluid
- Scala tympani and scala vestibuli
- Major cation: Na
Perilymph

- Osmolarity of perilymph: similar to CSF
- Source: controversial
  1. From CSF, enter cochlea via cochlea aqueduct
  2. Produced locally in cochlea by ionic or ultrafiltration mechanism
Several actual or potential dehiscences

1. **Oval window**: footplate → middle ear
2. **Round window**: secondary tympanic membrane
3. **Fissula ante fenestram** and **fossula post fenestram**: two small extension of the perilymphatic space from vestibule toward middle ear cavity

4. **Vestibular aqueduct**: from vestibule to posterior cranial fossa, transmit endolymphatic duct and accompany vein

5. **Cochlear aqueduct** (perilymphatic duct)
perilymphatic fistula

- **Definition**: abnormal opening in fluid filled inner ear
- **Frequency**: unknown
- **Mortality/Morbidity**: children → recurrent meningitis
- Acute or chronic perilymph fistulas affect quality of life.
- **Sex**: Prevalence is higher in females than in males.
- **Age**: young children → congenital abnormalities otherwise, no age specific.
Pathophysiology

- Perilymphatic space connect to subarachnoid space through cochlear aqueduct
- Pressure changes in subarachnoid space → communicated to inner ear
- Lifting, straining, coughing, sneezing → increase CSF pressure → increase inner ear pressure → tear of oval window or round window
Pathophysiology

- Barotrauma, compression trauma of ear, Valsalva maneuver, sneezing → increase middle ear pressure → ruptures or tears of round window or oval window
Pathophysiology

- Mechanism: Perilymph loss → hearing loss: unclear
- Decompression of perilymph space → secondary endolymphatic hydrops → develop symptoms of endolymphatic hydrops (Meniere’s disease)
Fistula types and usual causes:

1. **Window fistulae**
   - **Oval window type**
     - Stapedectomy surgery (for otosclerosis)
     - Head trauma or barotrauma (pressure injury)
   - Acoustic trauma

2. **Round window type**
   - Barotrauma -- SCUBA diving, airplane pressurization
   - Congenital malformations (such as Mondini)
2. **Canal type**
   - Cholesteatoma (chronic infection, Magliulo et al, 1997)
   - Superior canal dehiscence (congenital weakness in bone combined with head trauma, Minor 2000)
   - Head trauma
3. **Other otic capsule**, not in canal (congenital weakness in bone, neither located in the canal or window areas)
Stapes surgery

- 1960s PLFs that developed following stapedectomy procedures
- Loss of perilymph following stapedectomy could produce hearing loss, disequilibrium, and tinnitus
- Incidence: <0.1%
Stapes surgery

- Nystagmus after stapes surgery
- 13 patient: toward operated ear (3), toward opposite ear (2), operated ear → opposite ear (2), operated ear → opposite ear (2), no nystagmus (4)
- 1. inner ear damage by operation
- 2. postoperative perilymphatic fistula
- 3. floating footplate
- 4. stimulation of hair cells by high potassium ion in the perilymph due to blood flow into the inner ear.

Cholesteatoma

- Fistula formation into membrane labyrinth
- Labyrinthine fistula: 0.3 per 100000
- Symptoms: subjective hearing loss (90%), otorrhea (65%), dizziness (50%)
- Most common fistula: lateral semicircular canal (Swartz 1984)
- Can also occur at round and oval window
- Diagnosis: HRCT
Cholesteatoma

- HRCT
Cholesteatoma

- Cholesteatoma eroding into the labyrinth

should be left in place until the remainder of procedure is nearly complete
Superior semicircular canal dehiscence syndrome

- Positive Tullio and Hennebert’s sign
- Anatomical defect of sup canal at HRCT
- Minor et al 1998
- Etiology: failure to develop normal thickness of bone overly sup canal or bone disrupt (trauma)
- Clinical: Tullio phenomenon, Hennebert’s sign, hyperacusis
Superior semicircular canal dehiscence syndrome

- Diagnosis: HRCT 0.5mm
- Treatment:
  - avoid loud noise
  - tympanotomy
  - surgical repair:
    - middle fossa craniotomy
    - transamastoid superior canal occlusion
Clinical manifestation

- **Hearing loss**: Fluctuating sensorineural hearing loss that may be sudden or progressive
- **Tinnitus**
- **Aural fullness**
- **Vestibular symptoms**
  - Vertigo, with or without head position changes
  - Dysequilibrium
- **Motion intolerance**
- **Nausea and vomiting**
- **Disorganization of memory and concentration**
- **Perceptual disorganization in complex surroundings such as crowds or traffic**
Tests recommended when fistula is strongly suspected:

- fistula test
- Valsalva test
- Audiometry
- ECOG
- ENG
- Temporal bone CT scan, high resolution
- MRI scan
- VEMP (Vestibular evoked myogenic potentials)
Fistula test (Hennebert sign)

- Fistula test: pressure each ear canal with small rubber bulb → nystagmus
- Positive test: suggestive for PLF
- **Window fistula**: little nystagmus
- **Superior canal dehiscence**:
  - strong nystagmus
- Low sensitivity and specificity
Audiometry

- May show sensorineural hearing loss
- Superior canal dehiscence: may show bone conduction score better than air (conductive hyperacusis)
ECoG

- useful in the diagnosis of both Méniere disease and PLF.
- Both conditions: elevated SP/AP ratio.
- The mechanism increases in PLF is controversial.
- Some authors: increase in SP/AP ratio in patients with PLF may be the consequence of a decrease in the AP.
Guinea pig studies: increases in the SP/AP ratio in guinea pigs with artificially induced PLFs.

Increase in the SP/AP ratio during stapedectomy. The creation of a control hole in the footplate → NO change in the SP/AP ratio

A change is noted only after some perilymph has been removed from the vestibule.

ECoG

- individuals with surgically proven PLFs who have elevated SP/AP ratios preoperatively, the SP/AP ratio reliably returns to normal after surgical repair of the fistula.

Dynamic platform posturography

- Pressure applied to external ear canal → tympanic membrane → middle ear → inner ear → abnormal sway
Perilymph labeling method

- **Intrathecal or intravenous fluorescein**
  - Fluorescence was not evident in perilymph when complete hemostasis was obtained. Poe DS et al: Intravenous fluorescein for detection of perilymphatic fistulas. Am J Otol. 1993 Jan;14(1):51-5.

- **Beta-2 transferrin:**
Perilymph labeling method

- **Apo D and apo J**
- Two high-density lipoprotein-associated apolipoproteins—apo D and apo J—at concentrations 1 to 2 orders of magnitude higher in terms of total protein.
- The functional significance of the two apolipoproteins is not known.

Middle ear endoscopy

- rapid examination tool with which to verify certain areas of the middle ear anatomy, but it is of limited value for ruling out the presence of PLF.


MRI

- not the best test for fistulae
- showing other possibly confounding problems such as tumors or multiple sclerosis plaques.
HRCT

- very accurate in identifying canal fistulae (Fuse et al, 1996)
- at least 1 mm resolution
- congenital abnormalities of the cochlea, vestibule, and vestibular aqueduct may also be documented
HRCT

- Air in the labyrinth (pneumolabyrinth) → most convincing finding of fistula.
- Middle ear effusions → suggestive of fistula.
- Variants in the stapes structure → congenital fistula at the level of the oval window.
HRCT

- pneumolabyrinth
Medical therapy

- Some tears of the inner ear membranes probably heal without surgical intervention
- bed rest
- elevation of the head of the bed
- use of stool softeners
- avoidance of Valsalva maneuver
- sedation.
Patients with fistula should avoid

- Lifting
- Straining
- Bending over
- Popping the ears
- Forceful nose blowing
- Air pressure changes such as due to air travel
- High speed elevators
- Scuba diving
- Loud noises (such as your own singing or a musical instrument)
Operation

- The definitive treatment of PLF is **surgical exploration with grafting of the fistula.**
- The timing of surgical exploration is controversial.
Operation

- under local or general anesthesia.
- Tympanomeatal flap is designed, incised, and elevated.
- Curetting away the posterior bony overhang (scutum) to permit adequate visualization of the round and oval window niche.
- Carefully observed for the accumulation of clear fluid.
Operation

- Autogenous tissue grafts are placed directly over the leak.
- If no actual leak → footplate and round window are grafted prophylactically.
- Some surgeons do not graft unless an active leak is visualized.
- Other surgeons graft routinely, even if no leak can be detected by visual inspection.
Operation

- Adipose tissue originally was used but its use resulted in an unacceptably high rate of recurrent fistula.
- Fascia or perichondrium now is used.
- Some authors use fibrin glue; others do not.
Complications from PLF repair

- Tympanic membrane perforations: 1-2% of patients.
- Postoperative hearing loss
  - risk of severe-to-profound hearing loss: Mondini dysplasia
- Alteration of taste (chorda tympani injury)
Follow-up care

- seen again 1-3 weeks postoperatively
- A follow-up audiogram should be obtained at 6 weeks
- another follow-up audiogram should be obtained at 6 months.
- Beyond 6 months, follow-up care is determined by the patient's condition.
Outcome and prognosis

- 86 patients who underwent exploratory tympanotomy for suggested PLF. Active PLF: 35, all patients: oval and round window grafts.

- Improvement

- 68% of patients with surgically confirmed PLFs
- 29% who did not demonstrate active PLF.

- Improvement in hearing: 18.7%

Outcome and prognosis

- 49% of treated ears improved in terms of auditory function, but only 23% improved to serviceable hearing levels, and 11% had continued hearing deterioration.

- Hearing improvement in 17% of patients, stabilization of hearing in 67%, and continued progression in 17%.

- Improvement of vestibular systems in 83-94% of patients.
Outcome and prognosis

- Surgical exploration is highly effective for vestibular symptomatology, but its effect on hearing loss is less predictable.
Conclusion

- Perilymph fistulae are difficult to diagnose

- Congenital anomalies of the inner ear or middle ear should be considered in children with PLF

- Surgical exploration is mainstay of treatment and is effective for vestibular symptomatology