In the name of God
Perilymphatic Fistula

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Perilymphatic Fistula

Definition

• Abnormal connection between fluid-filled inner ear & gas-filled middle ear

• First time recorded at 1897

• One of the most challenging topics in otolaryngology
**Etiology**

- **Congenital / Developmental**
- **Trauma**
  - **Direct**
    - Surgery
    - Blow
  - **Indirect**
    - Exertion (lift,...)
    - Blow
    - Barotrauma
Clinical Findings

- **Vestibular disturbances**
  - the most prevalent complaint
- **Hearing disturbances**
  - usually fluctuant
  - sudden loss
  - rapidly progressive
- **Tinnitus**
  - isolated is not seen
- **Aural fullness**

Most patients have combination of symptoms
Most children with sudden or progressive SNHL have suffered from **cochlear diseases** other than perilymphatic fistula

- hereditary progressive loss
- viral labyrinthitis
- autoimmune inner ear disease
- endolymphatic hydrops
Even in the situation of highest suspicion (sudden loss triggered by head trauma in a child with radiographically malformed inner ears) only a minority of patients demonstrate window fistulization. Many of these children have internal fistulization.
Perilymphatic Fistula

- Definition
- Etiology
- Clinical Finding
- Diagnosis

Diagnosis

- **History**
- **Physical Examination**
  - Fistula test
- **Paraclinic Examination**
  - Audiometry (PTA & SDS)
  - Electrocochleography (ECoG)
  - Electronystagmography (ENG)
- **Perilymph leak assay**
  - Volume of perilymph: 0.075 ml
  - Rarely dramatic leak
Perilymphatic Fistula

- Definition
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- Diagnosis

**Perilymph Leak Assay**

- **Exploration**
  - Seen by at least to surgical personnel
  - Shift in light reflex
  - Trendelenburg position
  - Valsalva
  - Internal Jugular vein compression
  - Drop of mineral oil

*Positive exploration rate: 55%*
Perilymphatic Fistula

- Definition
- Etiology
- Clinical Finding
- Diagnosis

Perilymph Leak Assay

- Exploration
- Endoscopy
  - first in 1967
  - Variability in observing fistula
    - Observer bias
    - Mistaken of fluid
    - Fluctuant nature of leak
Perilymphatic Fistula

- Definition
- Etiology
- Clinical Finding
- Diagnosis

Perilymph Leak Assay

- Exploration
- Endoscopy
- Fluorescein

  - Intratechal
    - Neurologic complications
  - Intravenous
    - Mixture of blood & perilymph
  - Labeled anesthetics
Perilymph Leaky Assay

- Exploration
- Endoscopy
- Fluorescein
- Electrophoresis

Protein concentration

- CSF: 20 mg/dL
- Perilymph: 200 mg/dL
- Serum: 7000 mg/dL

risk of dilution
Perilymphatic Fistula

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Perilymph Leak Assay

- Exploration
- Endoscopy
- Fluorescein
- Electrophoresis

- Protein concentration
- Specific protein
  - Beta 2 transferrin
  - Apolipoprotein D
  - Beta tace protein (PG D synthase)

must be pure & concentrated
Perilymphatic Fistula

- **Definition**
- **Etiology**
- **Clinical Finding**
- **Diagnosis**

**Perilymph Leak Assay**

- Exploration
- Endoscopy
- Fluorescein
- Electrophoresis
- Imaging (CT or MR)

- Intratechal contrast may be useful
- Pneumolabyrinth is highly suggestive
Intervention or Observation?
This is a question.
Medical

- Bed rest
- Head elevation
- Stool softeners
- Avoidance of straining, coughing, or sneezing

no reports of the percentage of response
Surgical

- **Grafting of the oval and round windows**
  - **Areolar tissue** coforms best to windows
  - **Perichondrium** is better than fat
  - **Fat** with Higher recurrence
  - **Fascia** may be disappeared

Improvement of adhesion with

Fibrin glue or Laser coagulation
Difficult outcome analysis because of range of complaints at initial

- Relief of vertigo: quite good (75 – 100%) although spontaneous recovery from vestibulopathy is common in pediatrics
- Relief of hearing loss: more frustrating
  - 50% of patients: improved hearing
  - 25% of patients: SRT>35 dB or SDS>80%
Vestibular symptoms are the most & Hearing loss are usually fluctuant and is sudden or rapidly progressive

Attention to more common DDx such as Ménière

There is not diagnostic study

Diagnosis is based on constellation of clinics & paraclinics & sometimes exploration

Can be surgical or medical, but superiority of either is not documented
• Special Thanks for Your Attention