

OTOSCLEROSIS

Example Case Scenarios

Case 1

A 30 year old female presented with progressive deafness in left ear. There is also H/O tinnitus in left ear. On otoscopic examination, the tympanic membrane was normal. Rinne's was negative, Weber's was towards the left. There is H/O hearing loss in her mother in child bearing age.

Case 2

A 35 years old female presented with progressive hearing loss in the right ear, also associated with tinnitus. There is a history of hearing loss in her mother during child bearing age. She noticed her hearing loss during last pregnancy. Tympanic membrane is normal. Rinne is negative, Weber is lateralized to right ear. Patient is complaining of

mild hearing loss in the left ear also. Audiometry shows conductive hearing loss.

Case 3

A 30 year old female with a family history of hearing loss developed hearing loss during pregnancy. It is bilateral and slowly progressive. Audiometry showed conductive hearing loss.

Case 4

A 53 years old female presented with progressive left sided hearing loss and tinnitus. Hearing loss worsened during her pregnancy. Tympanic membrane is normal & PTA showed conductive hearing loss.

Case 5

A 30-year-old female presented with bilateral progressive hearing loss for the last five years after her first pregnancy. Hearing loss is also associated with tinnitus. On examination both tympanic membranes are normal. Rinne's is negative.

Most Probable Diagnosis

Otosclerosis

A classic cause of progressive conductive hearing loss in young females with a normal tympanic membrane and positive family history.

Why this diagnosis fits the cases

Common exam clues present:

- Young to middle-aged female
- Hearing worsens during pregnancy
- Positive family history
- Progressive hearing loss
- Tinnitus common
- Tympanic membrane normal on otoscopy
- Tuning fork:
 - Rinne's negative → Conductive loss

- Weber lateralized to affected ear
 - Audiometry → Conductive hearing loss
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What is Otosclerosis?

A disease of abnormal bone remodeling in the otic capsule → causes stapes fixation → impaired sound transmission to inner ear.

Types of Otosclerosis

| Type | Pathology | Hearing Loss Type |
|------------------------|------------------------------|--------------------------|
| Stapedial otosclerosis | Stapes footplate fixation | Conductive (most common) |
| Cochlear otosclerosis | Cochlear capsule involvement | Sensorineural |

| | | |
|------------|---------------|--------------------|
| Mixed type | Both involved | Mixed hearing loss |
|------------|---------------|--------------------|



Differential Diagnosis

Other causes of conductive hearing loss with normal TM:

- Serous otitis media
 - Adhesive otitis media
 - Tympanosclerosis
 - Ossicular discontinuity
 - Attic fixation of head of malleus
 - Congenital stapes fixation
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History Features



Demographics

- Female predominance
- Age: 20-40 years

- Worse during pregnancy
 - Positive family history (~50%)
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Hearing Symptoms

- Bilateral, progressive hearing loss
 - Usually conductive
 - May become:
 - Sensorineural
 - Mixed
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Associated Symptoms

- Tinnitus → ~80%
 - Vertigo → ~25%
 - Due to endolymphatic hydrops → Association with Ménière's disease
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Speech Changes

- Monotonous speech
 - Soft but well-modulated
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Special Symptom — *Paracusis Willisii*

Patient hears better in noisy surroundings
(Because others speak louder)

Examination

Otoscopy

- Tympanic membrane:
 - Normal OR
 - Pink hue → *Flamingo tint*
 - Schwartz sign → Reddish glow due to hypervascularity
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Eustachian Tube

- Normal function
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Tuning Fork Tests

| Test | Finding | Meaning |
|---------|-----------------------------|-----------------|
| Rinne's | Negative (BC > AC) | Conductive loss |
| Weber's | Lateralized to affected ear | Conductive loss |

Investigations

Pure Tone Audiometry (PTA)

- Conductive or mixed hearing loss
 - Carhart's notch → Dip in bone conduction at 2 kHz
 - Normal speech discrimination
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Tympanometry (Impedance Audiometry)

- Decreased compliance
 - Type As curve
 - Absent stapedial reflex
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HRCT Temporal Bone

- Shows immature otosclerotic bone foci
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Exploratory Tympanotomy

- Direct palpation shows: → Stapes fixation
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Investigation Flowchart

Patient with progressive hearing loss → Tympanic membrane normal → Tuning fork tests show conductive loss pattern → Pure Tone Audiometry (Conductive/mixed loss + Carhart's notch present) → Tympanometry (↓)

Compliance + Absent stapedial reflex) → HRCT Temporal bone (if needed) → Diagnosis: Otosclerosis

Treatment

Medical Management

- Sodium fluoride → Slows progression
 - Hearing aids → For non-surgical candidates
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Surgical Management (Treatment of Choice)

Goal: Restore ossicular mobility using prosthesis

| Procedure | Description |
|--------------|--|
| Stapedectomy | Removal of stapes + prosthesis placement |
| Stapedotomy | Small fenestra in footplate + prosthesis (preferred) |

Treatment Flowchart

Confirmed Otosclerosis → Assess severity & patient fitness → Mild / Not fit for surgery

- Hearing aids
- ± Sodium fluoride

→ Fit for surgery

- Stapedotomy (preferred)
- Stapedectomy (alternative)

Exam Pearls

- ★ Young female + pregnancy-associated hearing loss
- ★ Normal TM + Conductive loss = Think otosclerosis
- ★ Paracusis Willisii is classic
- ★ Carhart's notch at 2 kHz
- ★ Absent stapedial reflex = Key finding
- ★ Surgery with prosthesis = Definitive treatment

-> The End <-