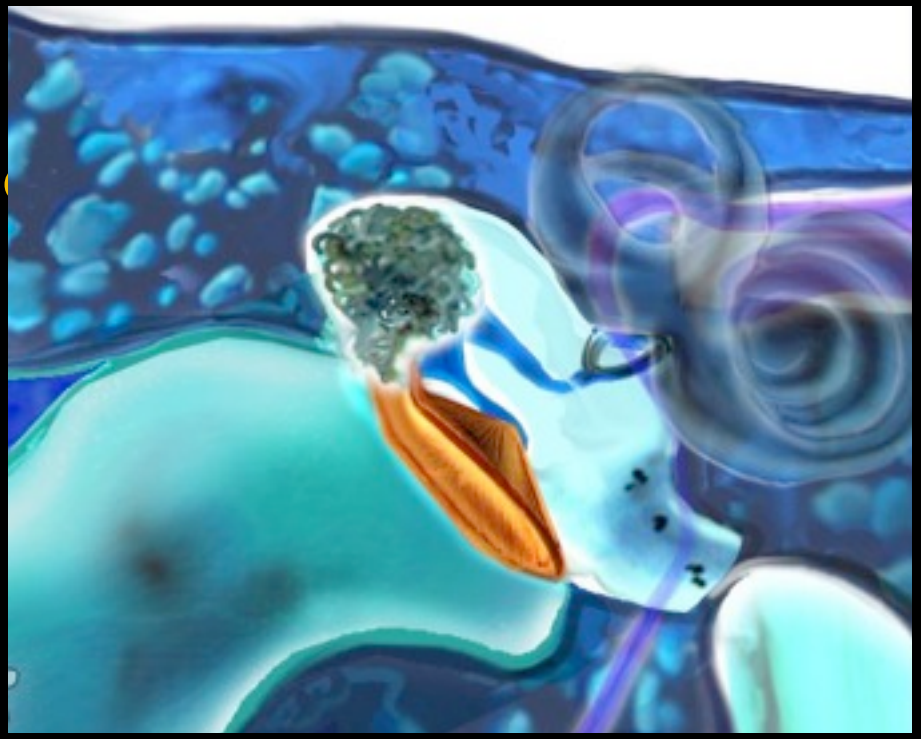
Cholesteatoma
in children



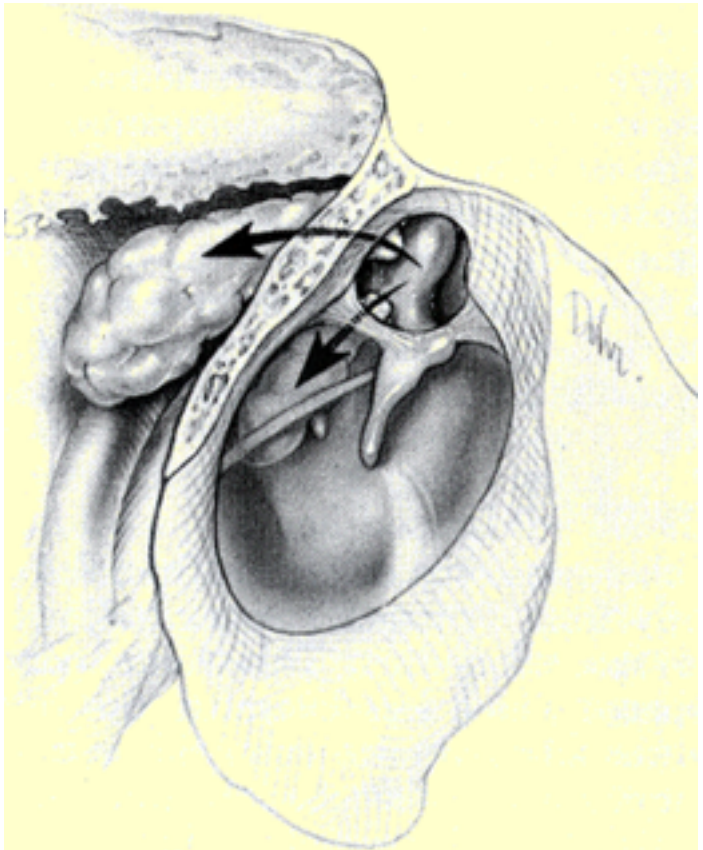
British Association of Paediatricians in Audiology
London Conference, Jan.2012

Matthew Clark FRCS (ORL-HNS)

Consultant Otologist
Gloucestershire Royal Hospital

Overview:

- Cholesteatoma and chronic otitis media – nomenclature
- Pathogenesis and sequelae
- Diagnosis and treatment
- Paediatric impact



Normal anatomy:

Pars Flaccida

Pars Tensa

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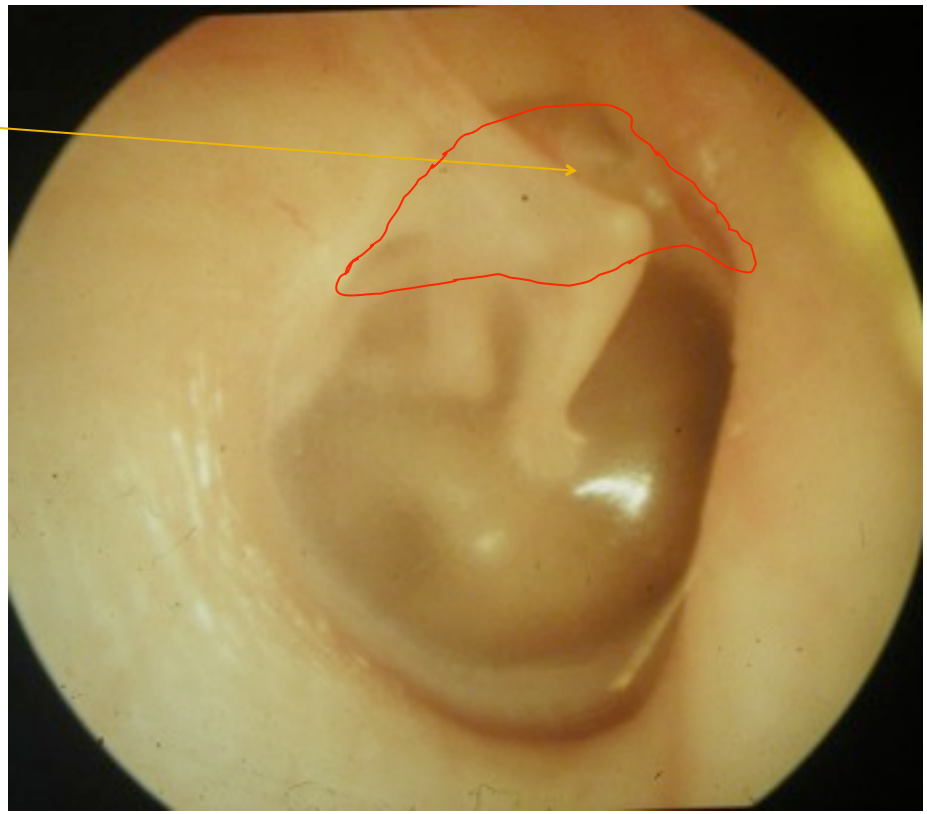

Pars Tensa

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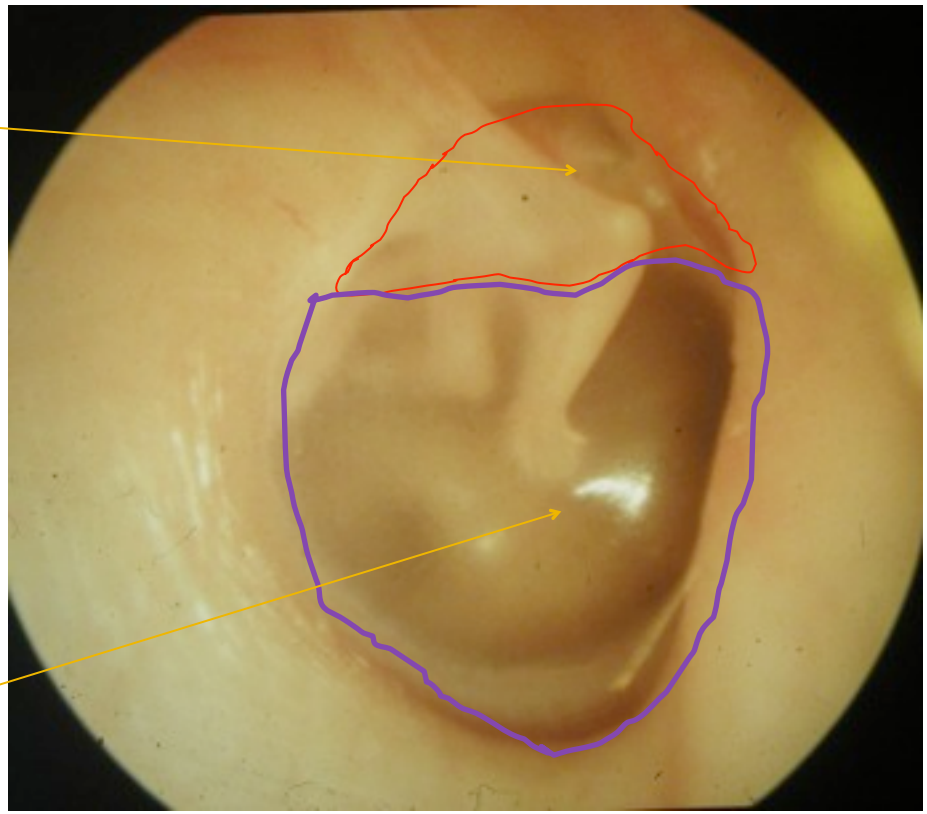

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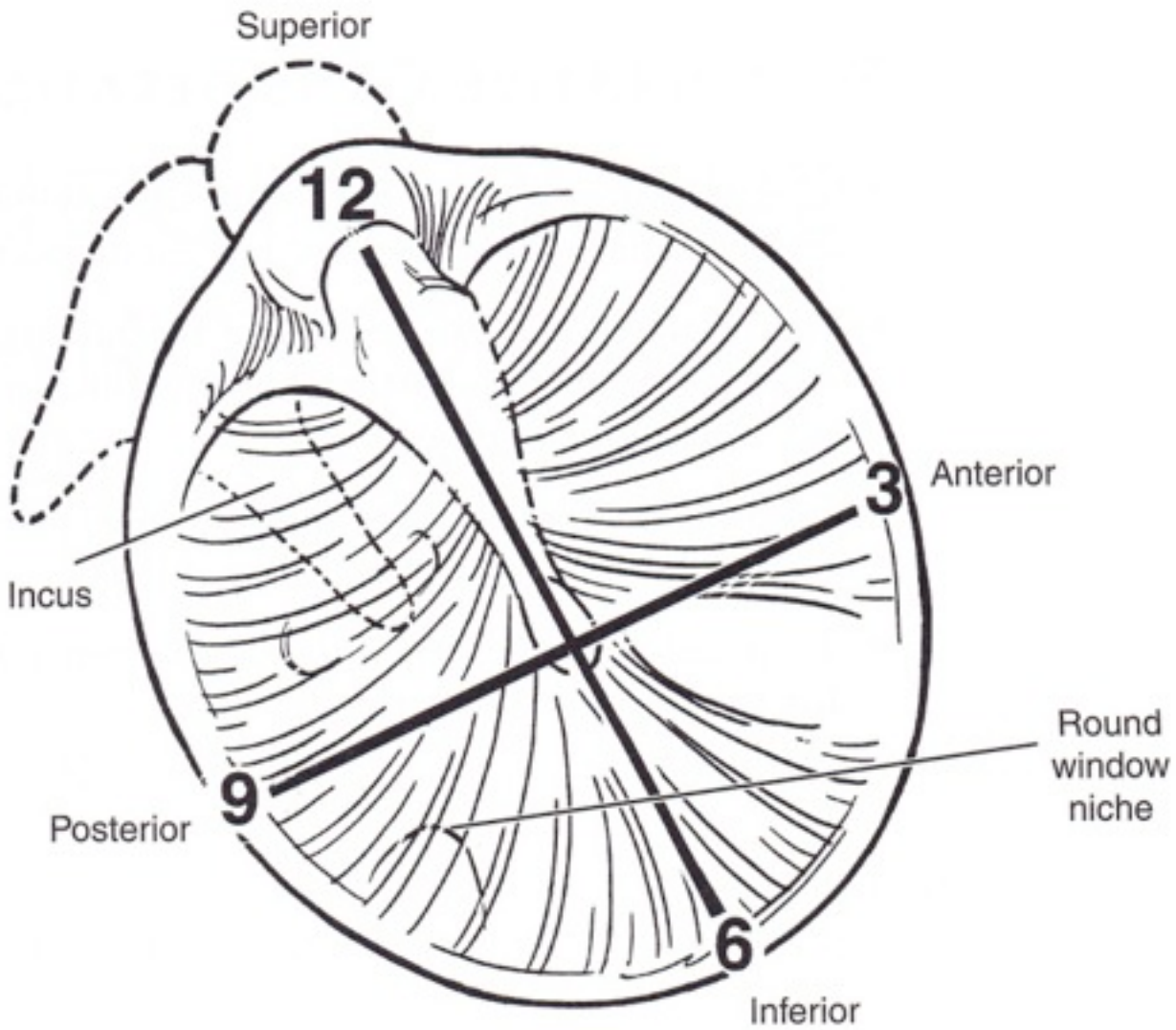

Anatomy...:

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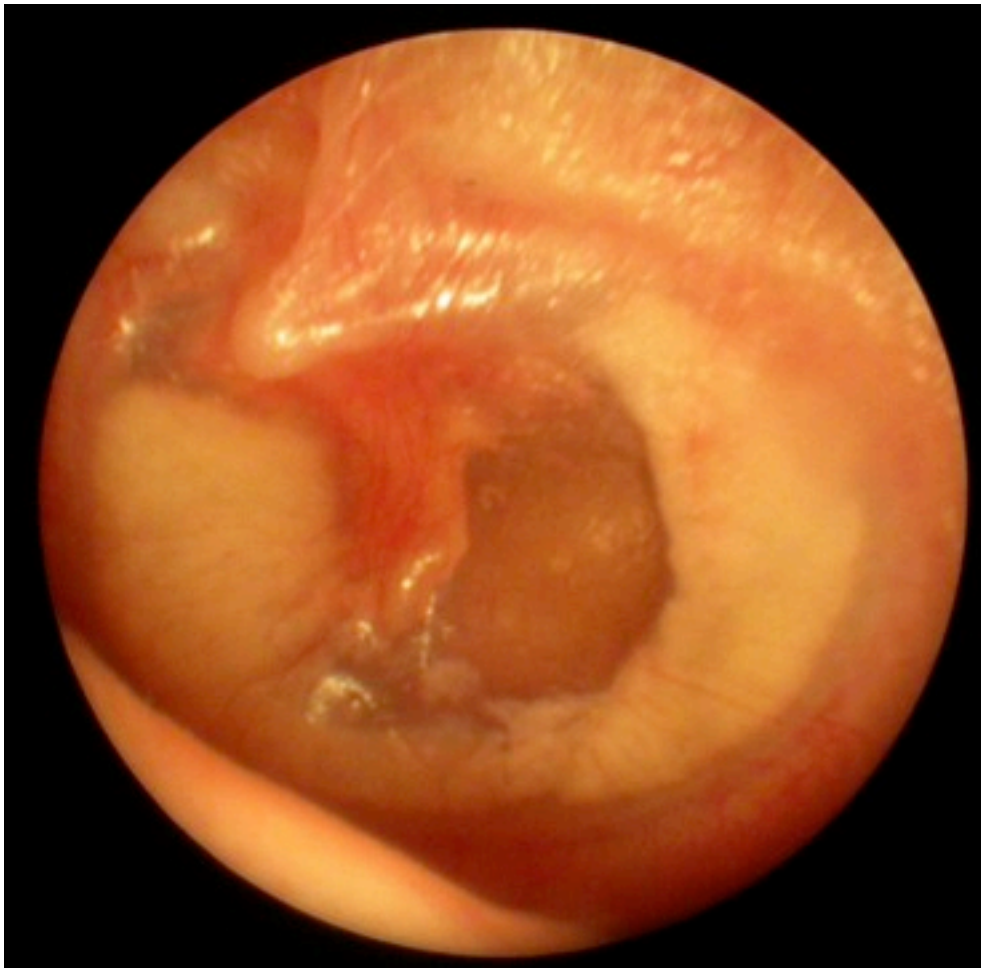
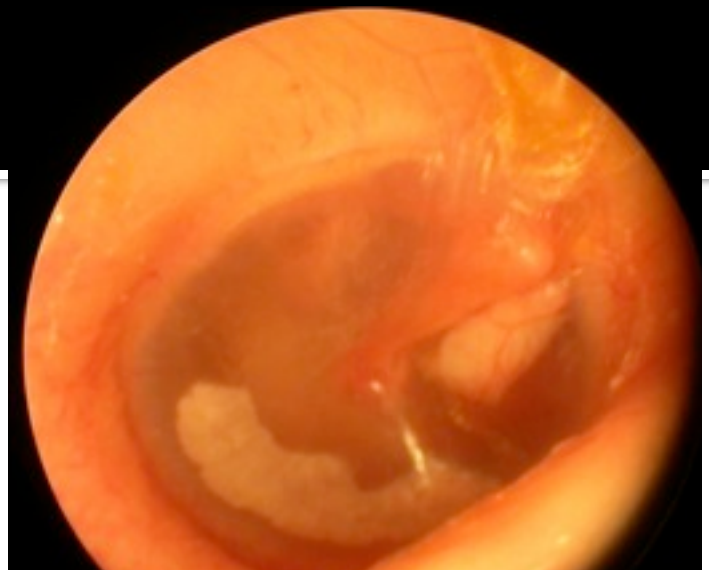
Anatomy...:

Otitis media classification:

- Acute or chronic
- Chronic:
  - Healed: tympanosclerosis, healed perforation
  - Inactive mucosal: dry perforation with ME mucosa non-inflamed
  - Active mucosal – perforation with mucopus from ME mucosa
  - Inactive squamous: retraction pocket not retaining debris

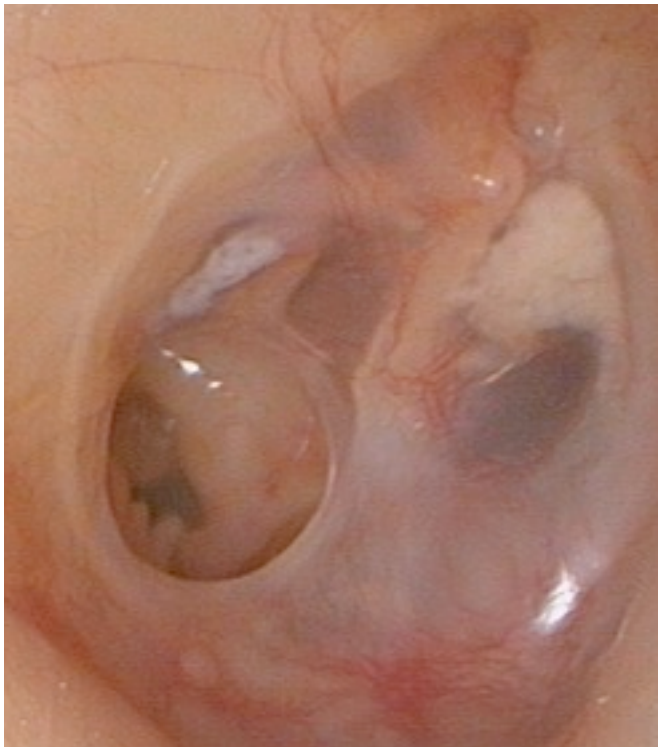
Healed COM:





Mucosal:

INACTIVE:



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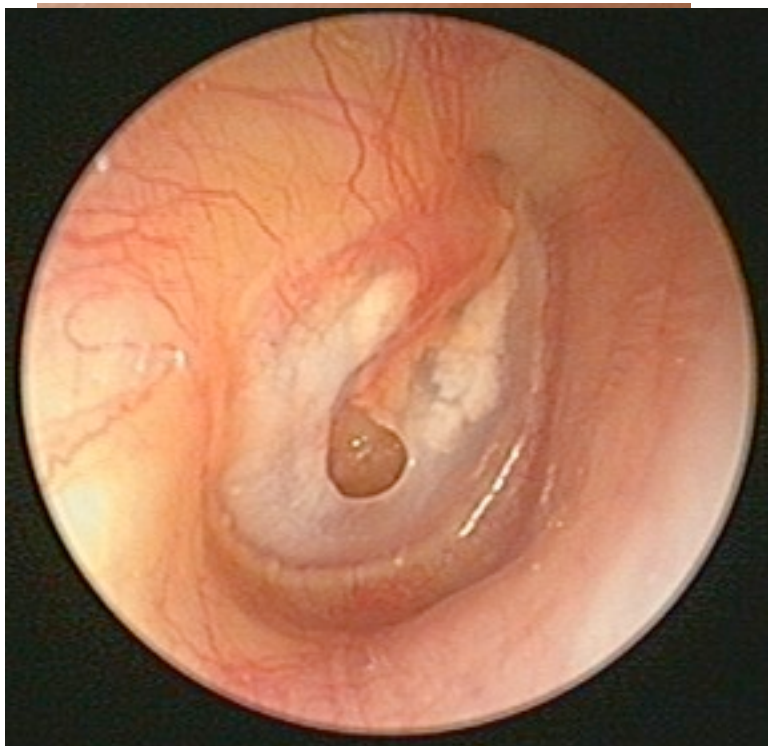

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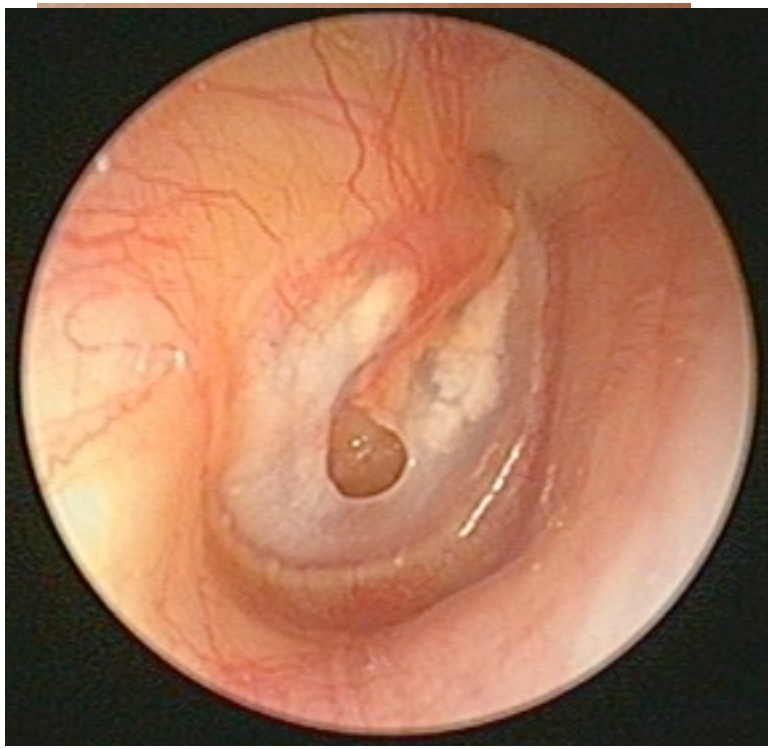

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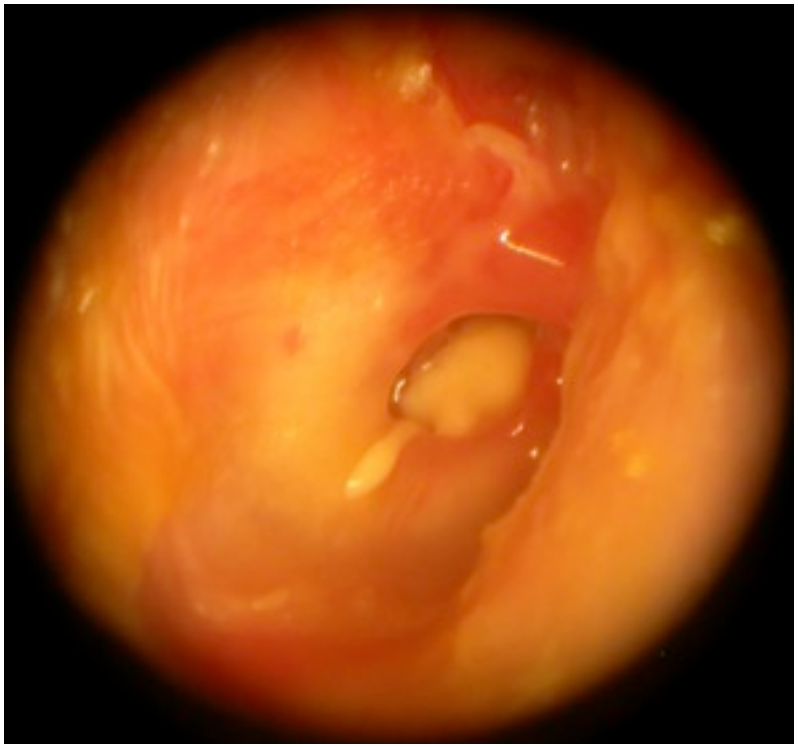

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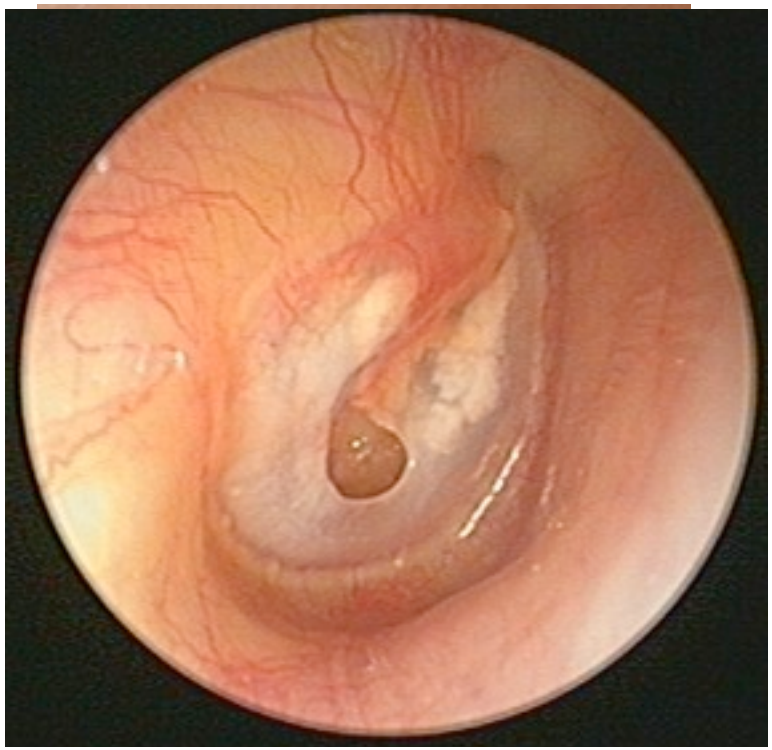
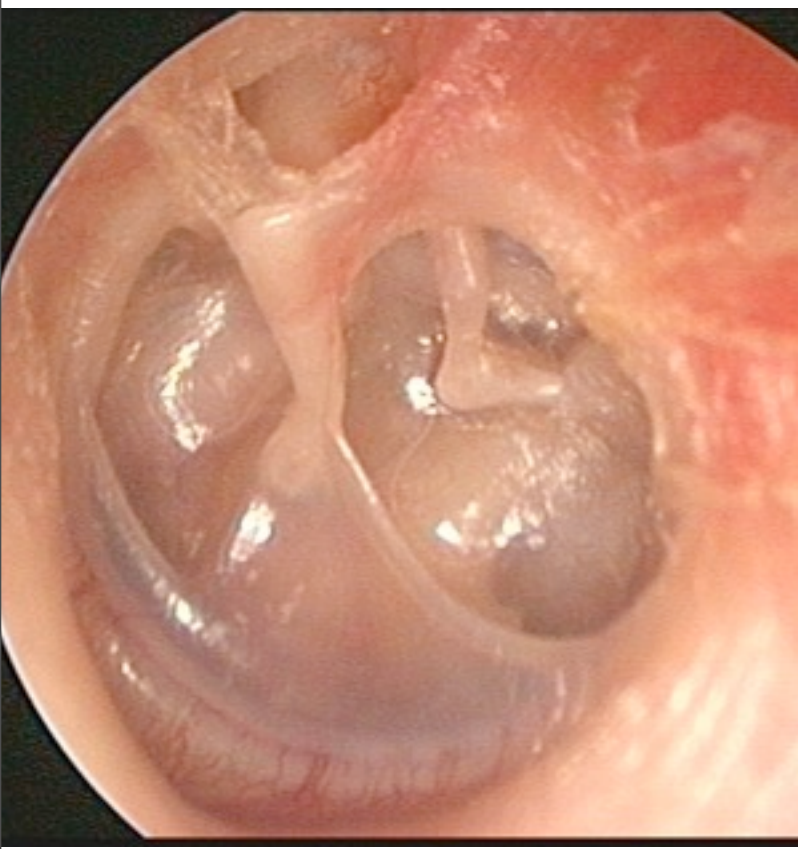


ACTIVE:

Inactive squamous:



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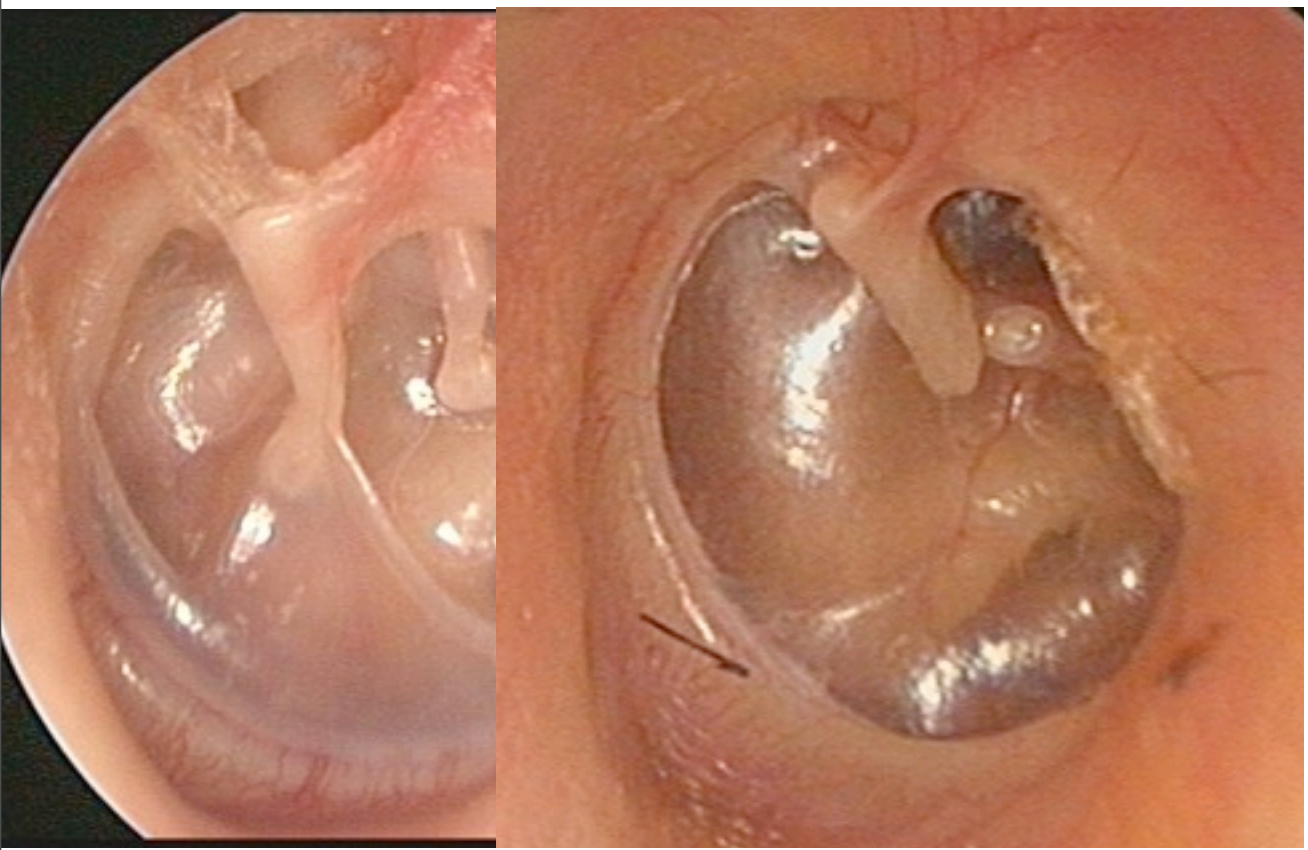

Inactive squamous:

Cholesteatoma:

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A 3-D epidermal structure exhibiting independent growth, replacing middle ear mucosa, reabsorbing underlying bone & tending to recur after removal.

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A 3-D epidermal structure exhibiting independent growth, replacing middle ear mucosa, reabsorbing underlying bone & tending to recur after removal.

Bad skin in the ME cleft

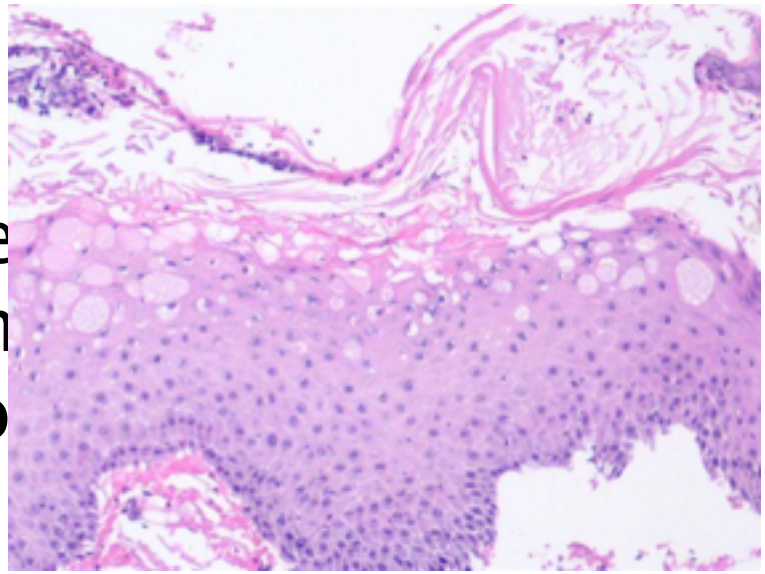
Incidence:

- Cholesteatoma: 5-15 per 100,000 population (children)
- COM effects up to 2% population
- Vigilance!

- Greater impact in the developing world
  - Uganda: >10% children have HL >31dB in better ear
  - Most due to COM
  - >40% childhood HL due to correctable

Pathogenesis:

- A defective wound-healing process
- Induction of a metaplastic e
- Pathologic collision of the h inflammatory response, no middle ear mucosa and a bacterial infection



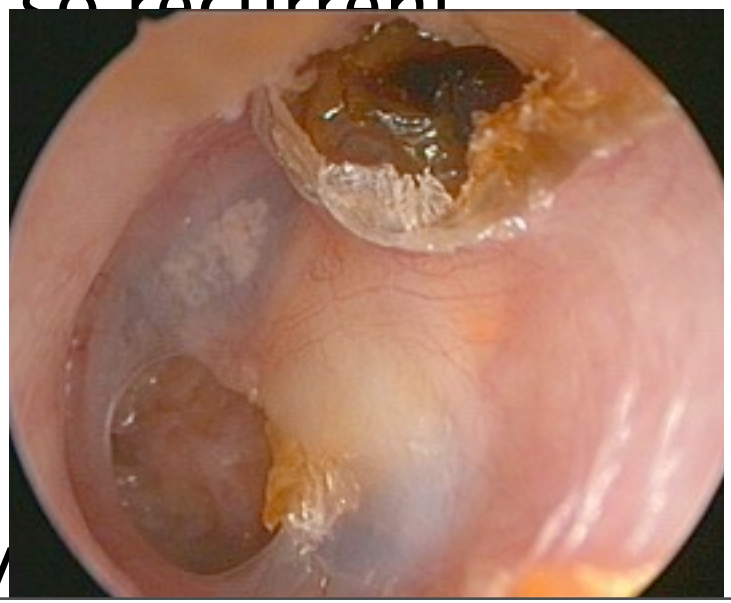
Why does this skin become invasive, migratory, hyperproliferative, aggressive and recidivistic?

What we know about cholesteatoma:

- Epithelium behaves more like wound-healing process than neoplasm
- Lack the genetic instability seen with malignant lesions
- Have hyperproliferative cells in all layers of epidermis: implies idiopathic response to both internal & external stimuli (cytokines released by inflammatory cells)
- Bacteria may incite and destabilise the squamous epithelium (hence it becomes proliferative, migratory & invasive)

What is the disease process?

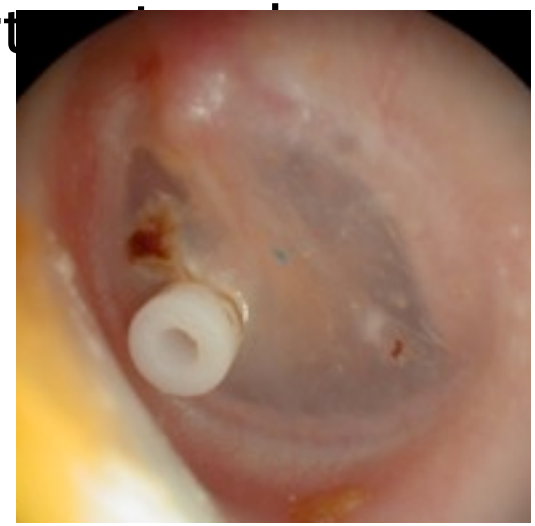
- Epithelial lined cyst containing desquamated keratin
- As exfoliation continues sac expands
- Dead cells/ keratin harbour bacteria (Pseudomonas, Coli bacilli), so recurrent infection/ putrefaction
- Promoted by moisture
- Granulation tissue between sac and underlying bone – inflammation leads to bony



Risk factors:

- Persistent/ refractory ME disease (& ETD)
- Australian study [2010]: ~46,000 children who had received grommets
  - 1% developed cholesteatoma (on av. 3.8 yrs later)
  - More Gs, greater risk
    - One set: 0.7%
    - 4 sets: 3.6%
- Some risk also linked to cleft palate, rural living, later age of 1st G

Adenoidectomy MAY be protective as may short
Gs



Types:

- Congenital
- Retraction pocket (90%+ of all)
- Implantation

Appear to be no molecular or cellular differences between the types

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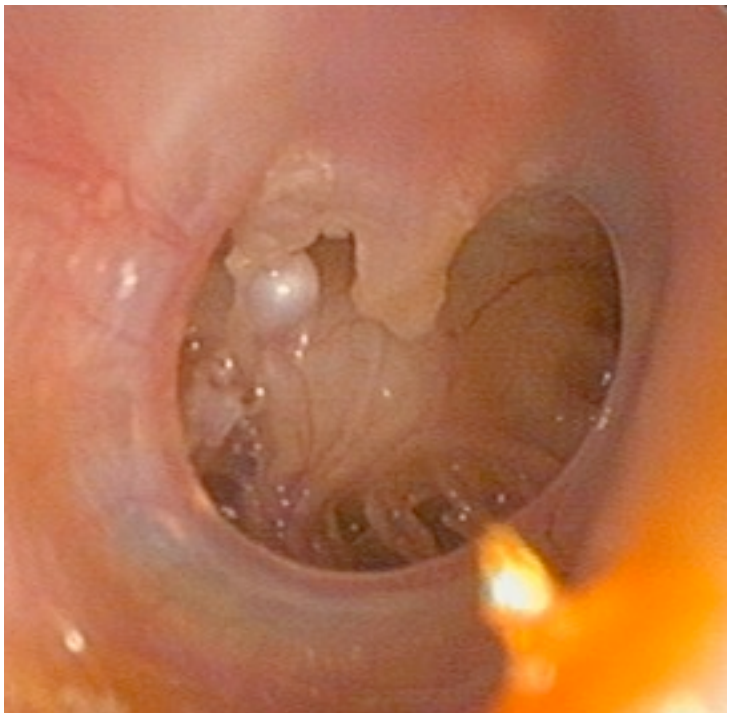
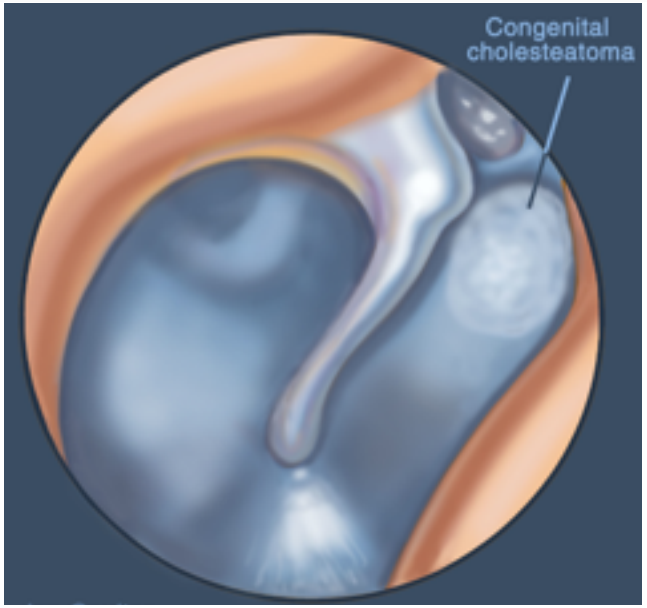


Congenital cholesteatoma:

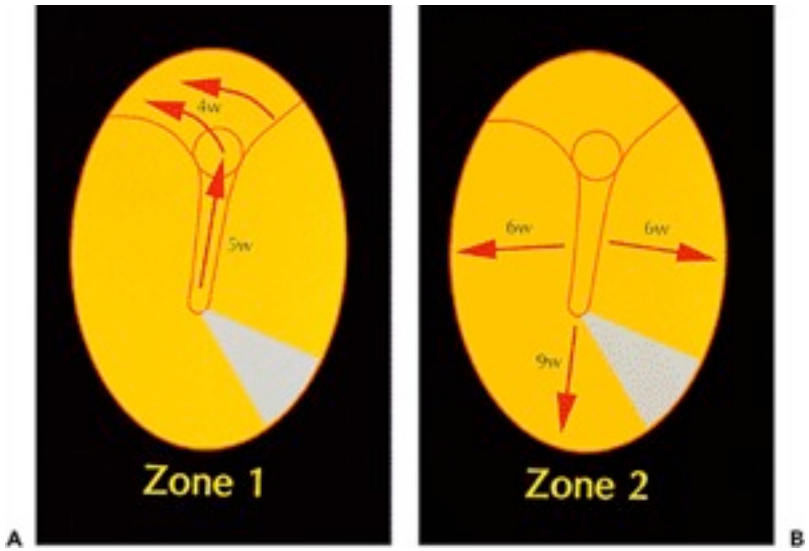


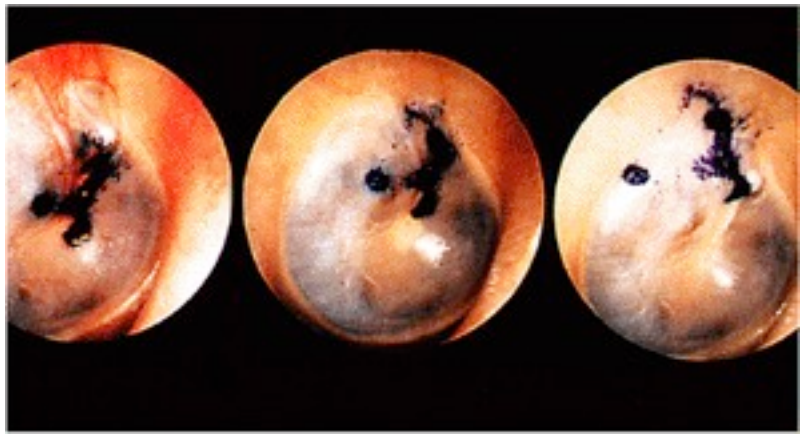
- Generally a disease of childre
- ~5% of total
- Emerge behind intact TM
- Anterior location/ peri-ET

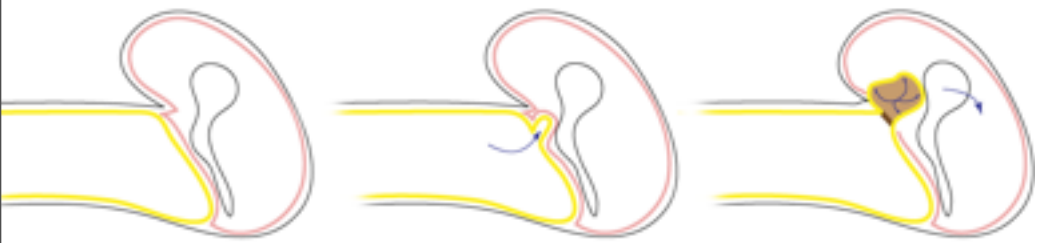
- Pathogenesis unclear:
  - Embryonic ectodermal rests – 1st pharyngeal pouch?
  - Squamous epithelium trapped during temporal bone embryogenesis
  - Metaplastic process secondary to ME inflammation?
- Widespread & 'aggressive'

‘Retraction pocket’ cholesteatoma:

- 90%+
- Originating from site of atelectasis, retraction
- Relationship with Eustachian tube dysfunction
- Epithelial migration fails to clear shed skin

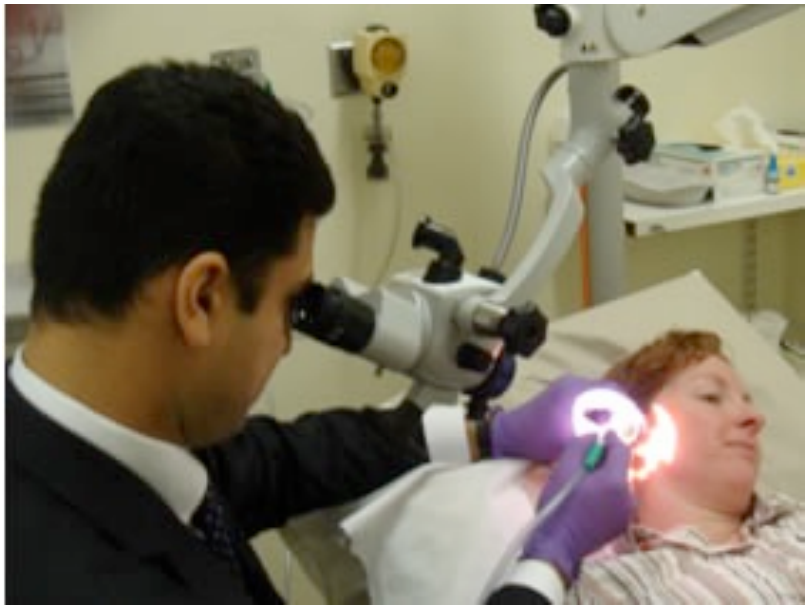






Diagnosis:

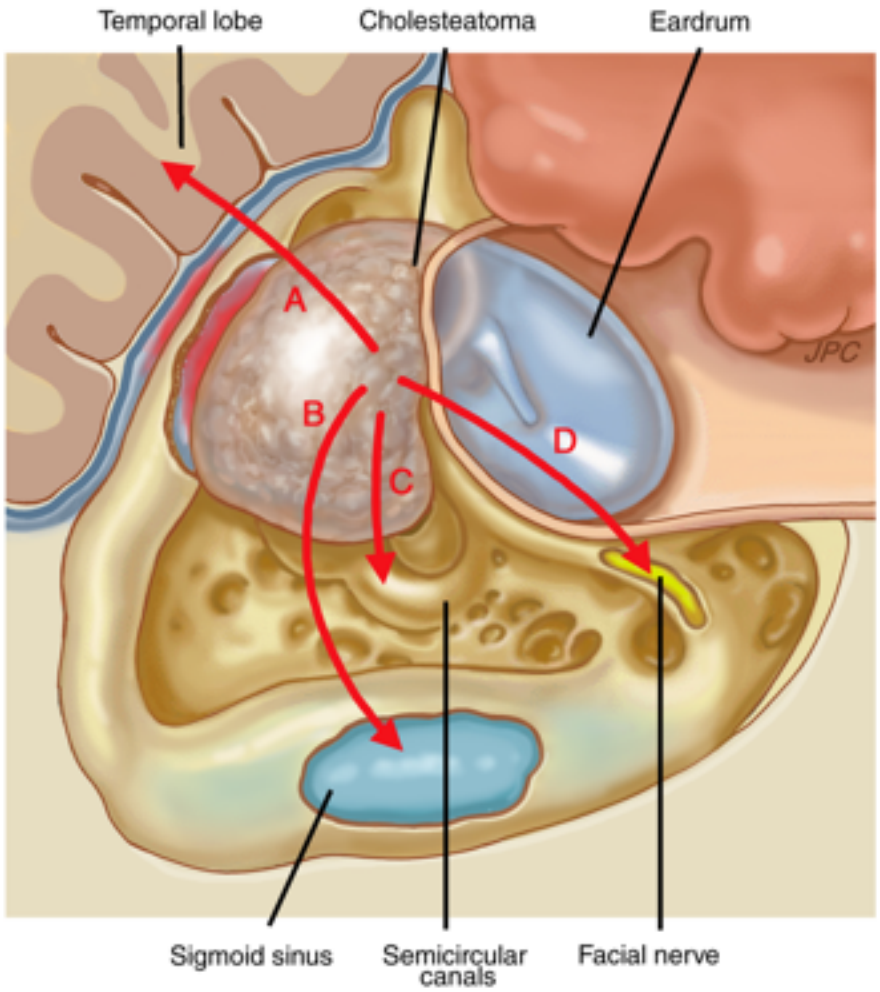
- History
- Otoscopy
- Audiology
- Radiology



Sequelae/ complications of

- Discharge
- Deafness
- Dizziness
- Droopy face
- Death

- AOM leads to more intratemporal and intracranial complications than COM



Discharge:

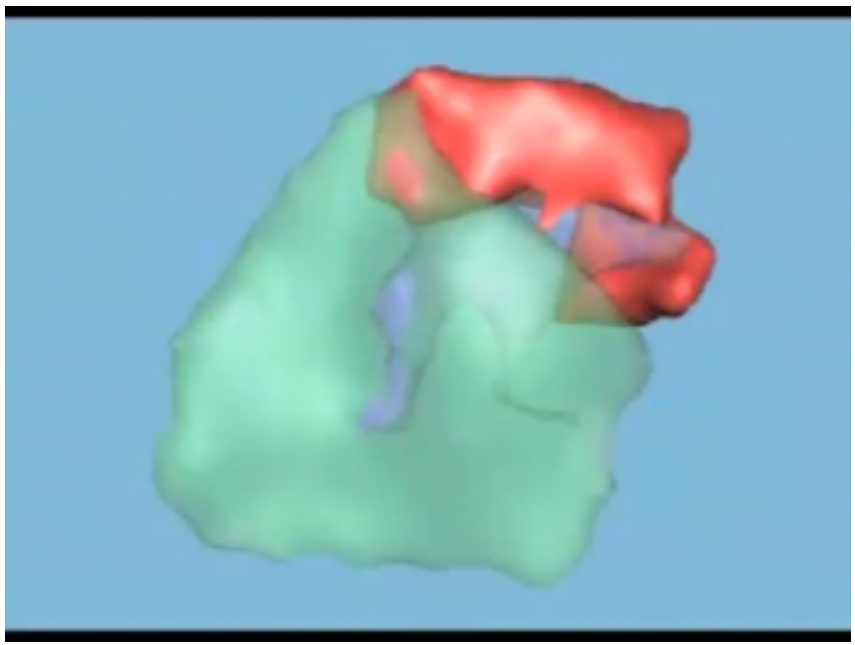
- Seropurulent from cholesteatoma
- Mucopurulent from middle ear mucosa infection

- Recurrent discharge unresponsive treatment: think CHOLESTEATOM

- ‘Wax’ on TM: think CHOLESTEATOMA

Deafness:

- May be none/ minimal
  - Disease transmits sound
- Conductive +/- sensorineu



Deafness:

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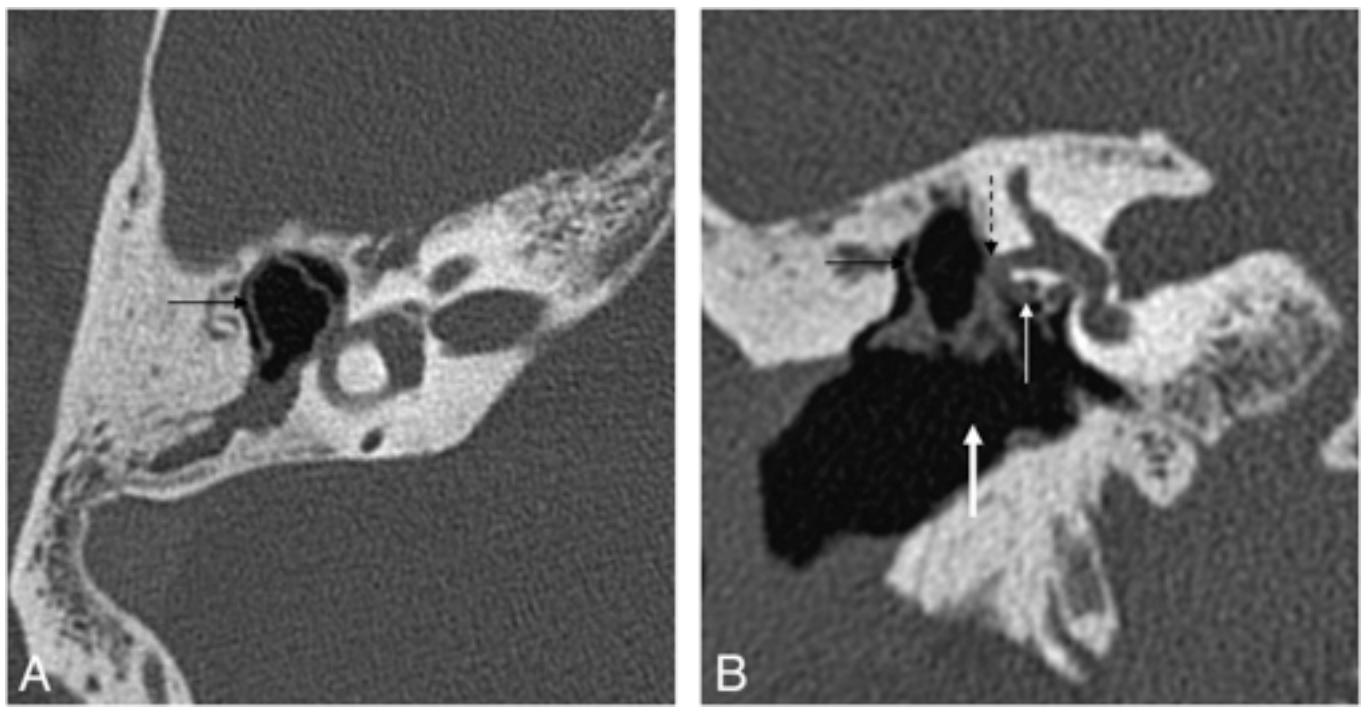
Dizziness:

- Fistulation of the lateral SCC
- Labyrinthitis: via RW membrane
- Exacerbations with infection



Facial palsy (droopy face)

- In ~10% population, part of the horizontal section of VII is dehiscent
- VII palsy more common in AOM than COM
- Taste disturbance

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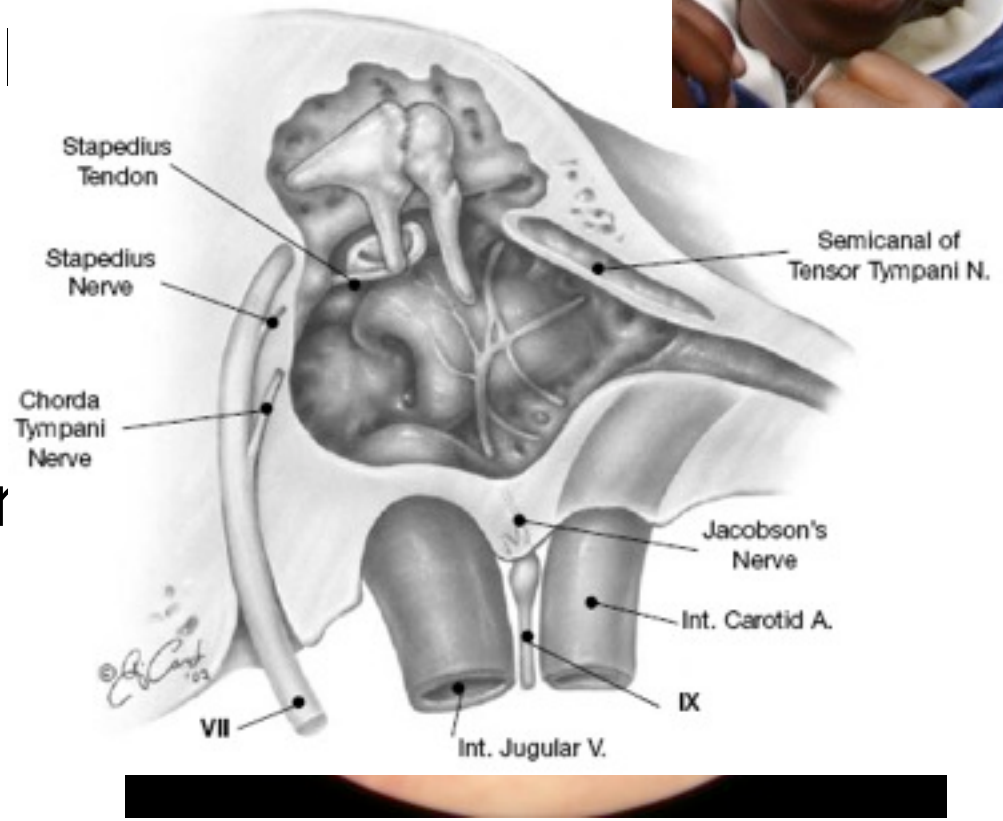

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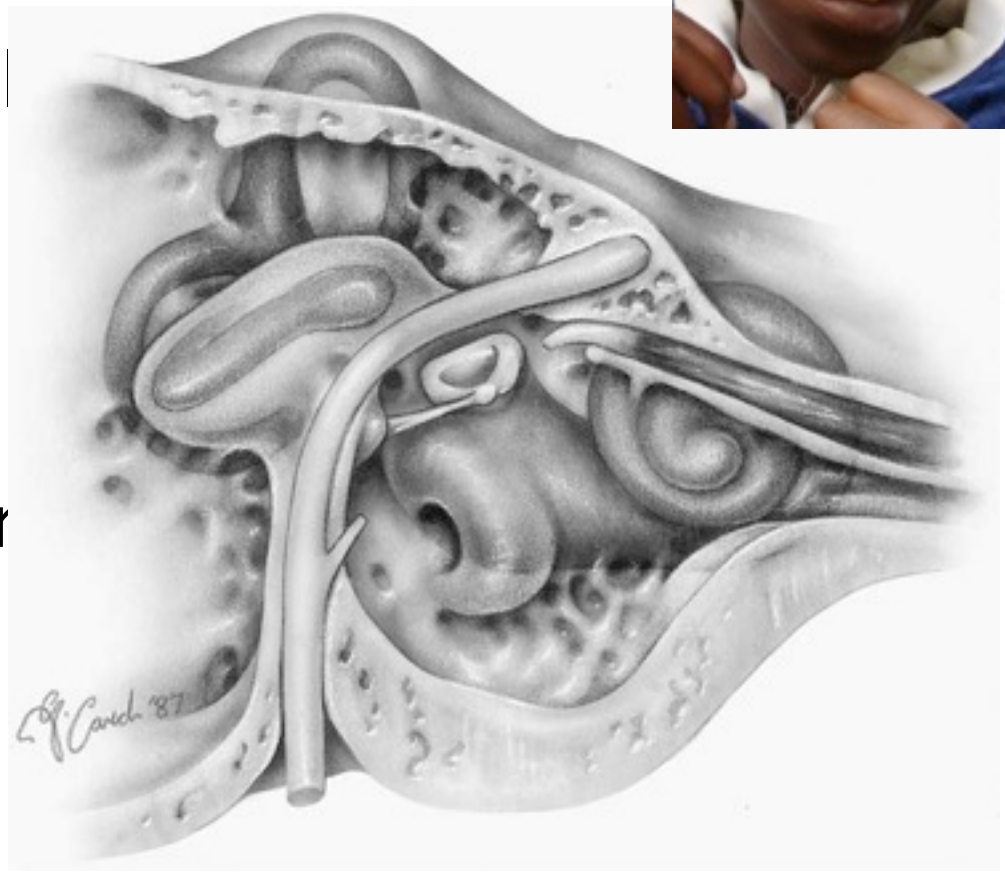


Death:

- Meningitis: AOM most common cause of bacterial meningitis, but also with COM
- Extradural, temporal lobe, peri-mastoid abscesses
  - Lifetime risk of developing otogenic intracranial abscess ~1:200
  - Lateral sinus thrombosis – thrombophlebitis can spread to cavernous sinus, cause septic emboli or cause septicaemia
- Petrositis with CN palsy (V, VI – Gradenigo's

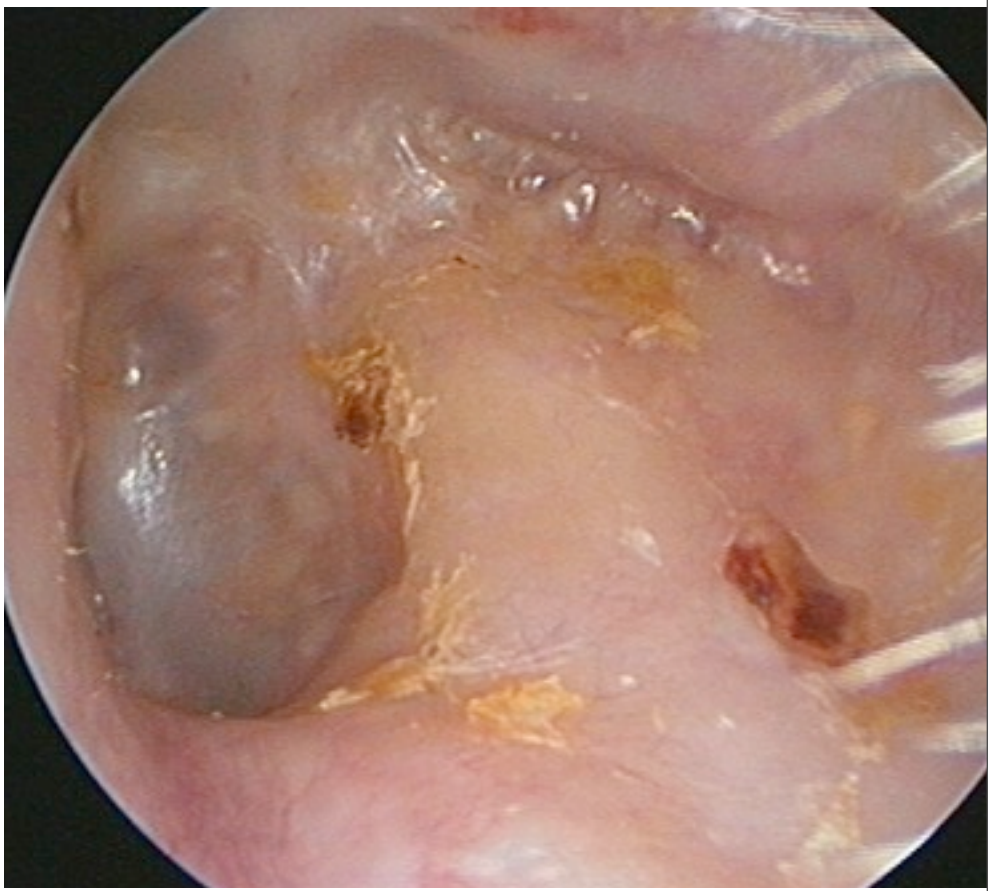
Treatment:

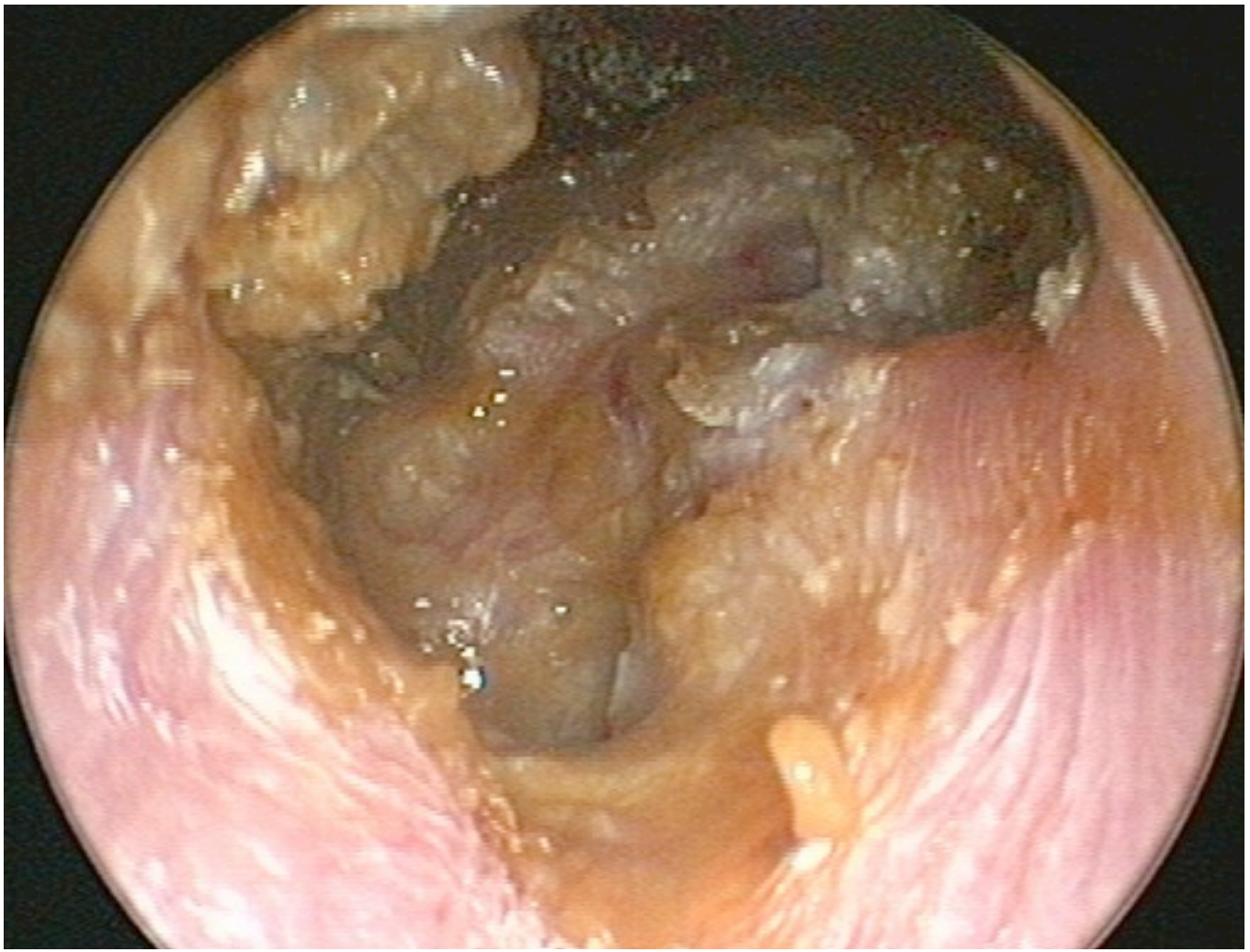
- Medical: aural toilet, topical medication – will NOT resolve cholesteatoma

- Surgical:
  - Aims:
    - Disease eradication (laser)
    - Hearing preservation or reconstruction
  - Approaches:
    - Canal-wall up (CAT)
    - Canal-wall down (modified radical

Canal-wall down:

- Mastoid cavity
  - Aural toilet
  - Water exclusion
  - Hearing reconstruction
  - Hearing aid fitting
- Avoid 2nd operation
- Better option if patient unlikely to come for f/u



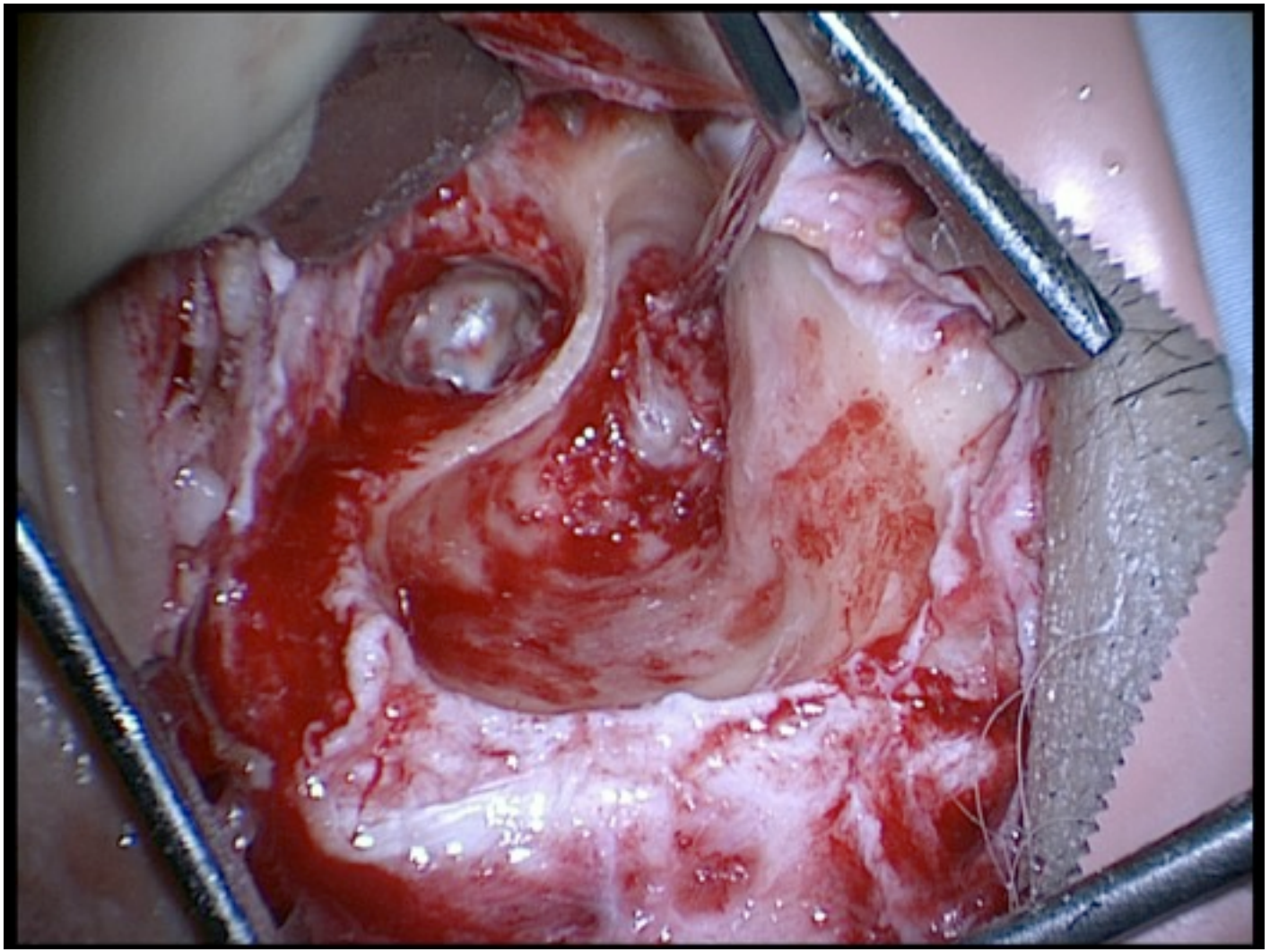


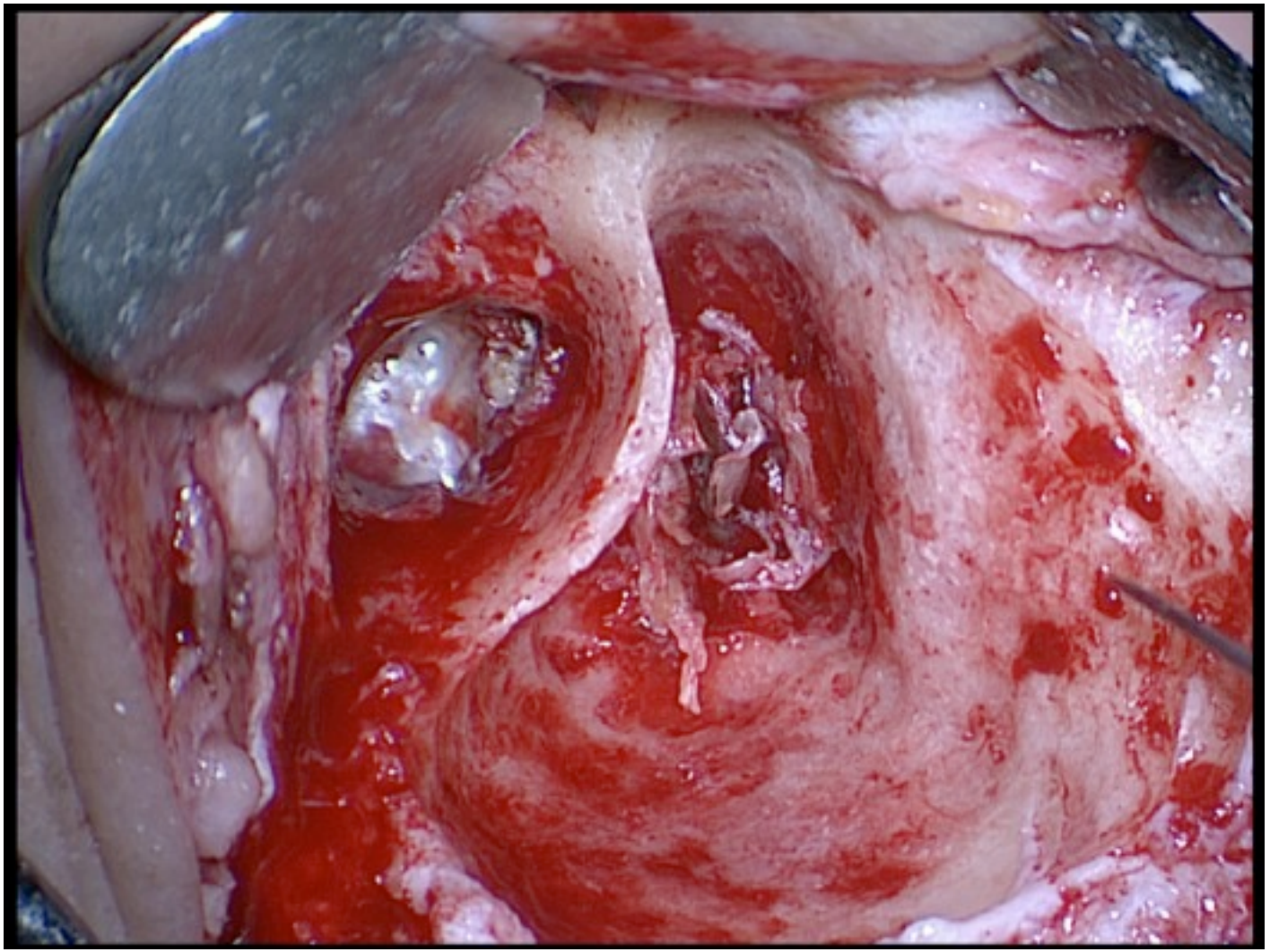
Canal-wall up:

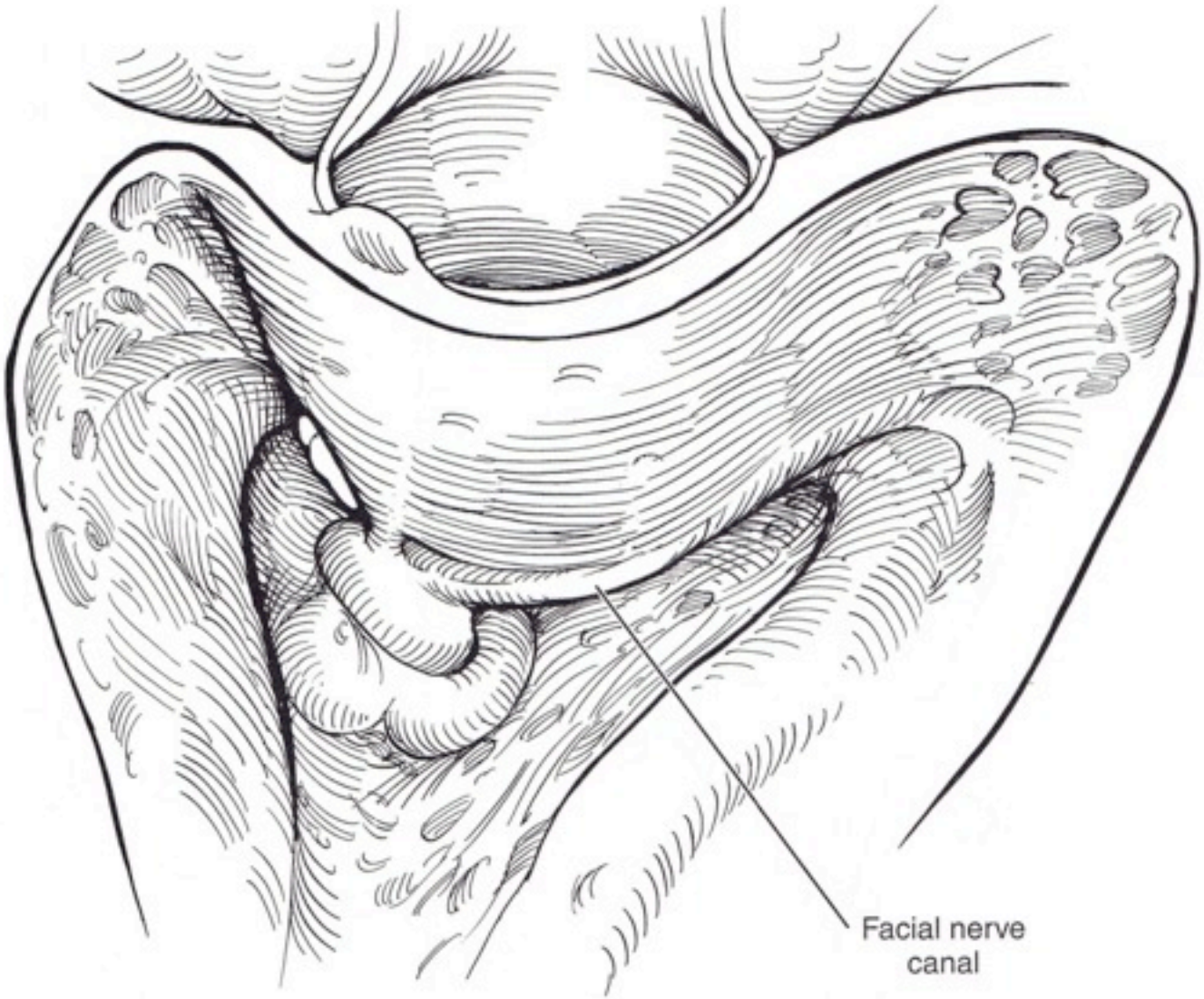
- Avoids water exclusions
- Allows for discharge from clinic
- Easier to help hearing (reconstruction or HA)

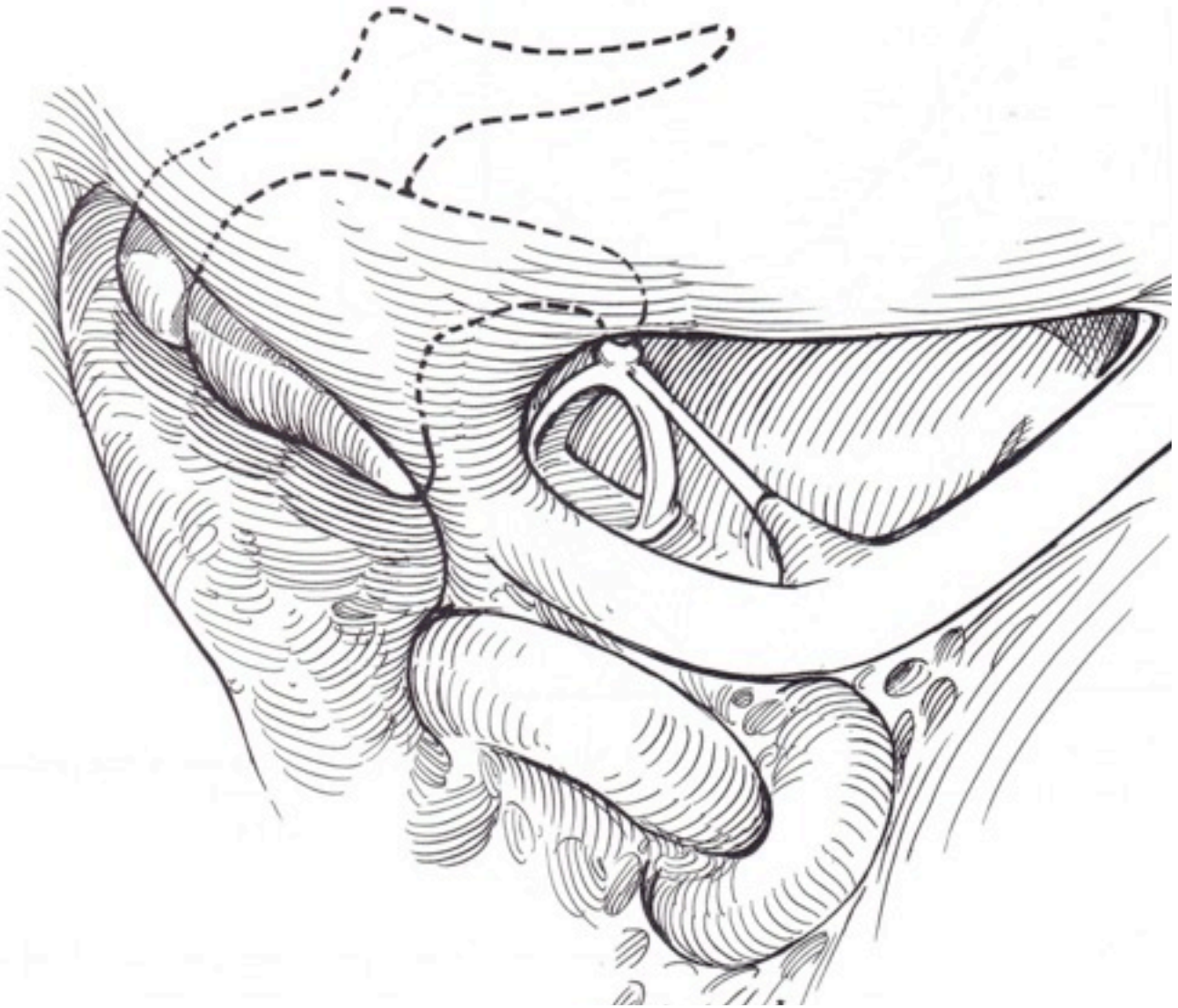
- Committed to 2nd operation

- Often ideal for paediatric population






Facial nerve
canal



Does it work?

- With CWU surgery, residual disease found <20%
- Hearing restoration from ossiculoplasty vary ~60-80%
  - But greater number now able to wear HA
- Disease specific quality of life measures (CES): surgery has significant positive impact on patient's life
  - Visits to doctor
  - Antibiotic requirement

Summary:

- Consider cholesteatoma when recurrent, offensive otorrhoea
- Serious. Life-threatening sequelae do exist, but are rare
- Surgical treatment usually required

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ANY QUESTIONS?