

Turner's Syndrome and its Ear, Nose & Throat Manifestations



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Introduction

Turner's Syndrome

- 1/2500 female live births
- Stature & gonads almost universally affected
- Many other organ systems affected to varying degrees and at different stages of life

Introduction

Turner's Syndrome

Multi-disciplinary approach to management
is therefore essential



Diagnosis

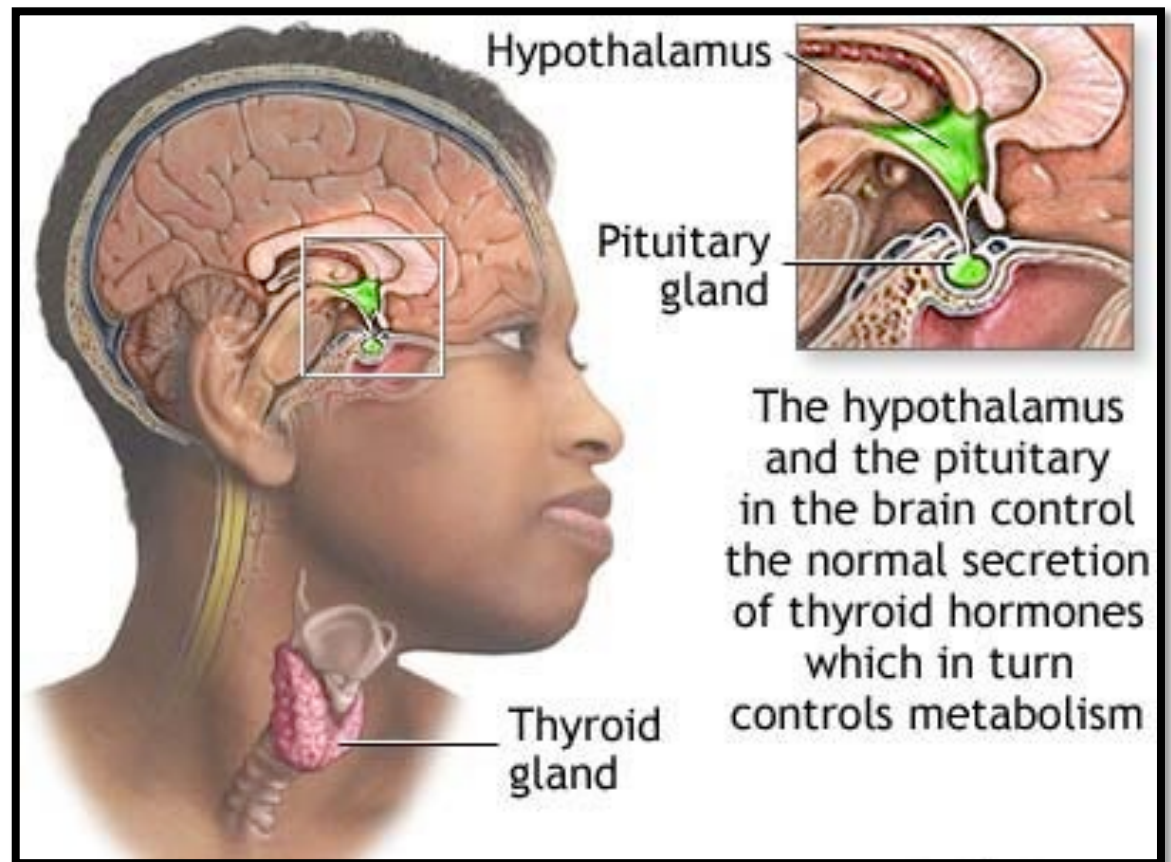
- Diagnosis of Turner syndrome: - in utero
 - newborn period
 - late childhood
 - adulthood



THYROID PROBLEMS

Hypothyroidism

Thyroid Function



Hypothyroidism

Thyroid Function

- Autoimmune thyroid disease increases in frequency with age in Turner syndrome (*Hashimoto's thyroiditis*)
- 50% antibody positivity
- 25-30% hypothyroid as adults

Hypothyroidism

Thyroid Function

- Thyroid (TPO) autoantibodies should be measured every 5 years
- If positive, TSH should be measured annually
- Thyroid replacement therapy daily

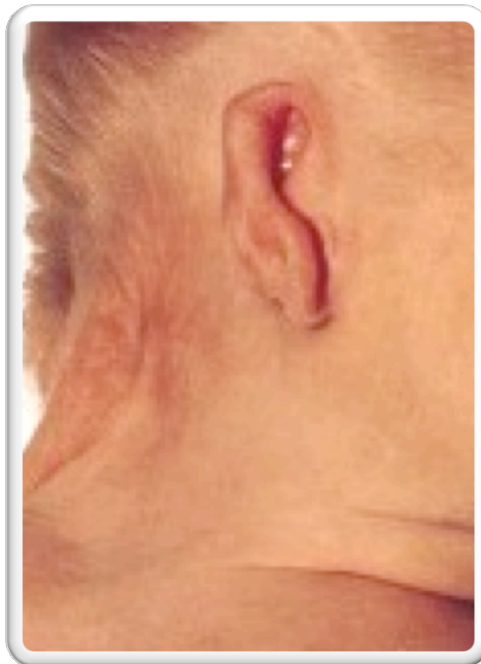


EARS

Ear & Hearing Problems

External Ear Malformations & Otoplasty

- May be associated with malformations



Ear & Hearing Problems

External Ear Malformations & Otoplasty

- Keloid scarring more common in Turner syndrome

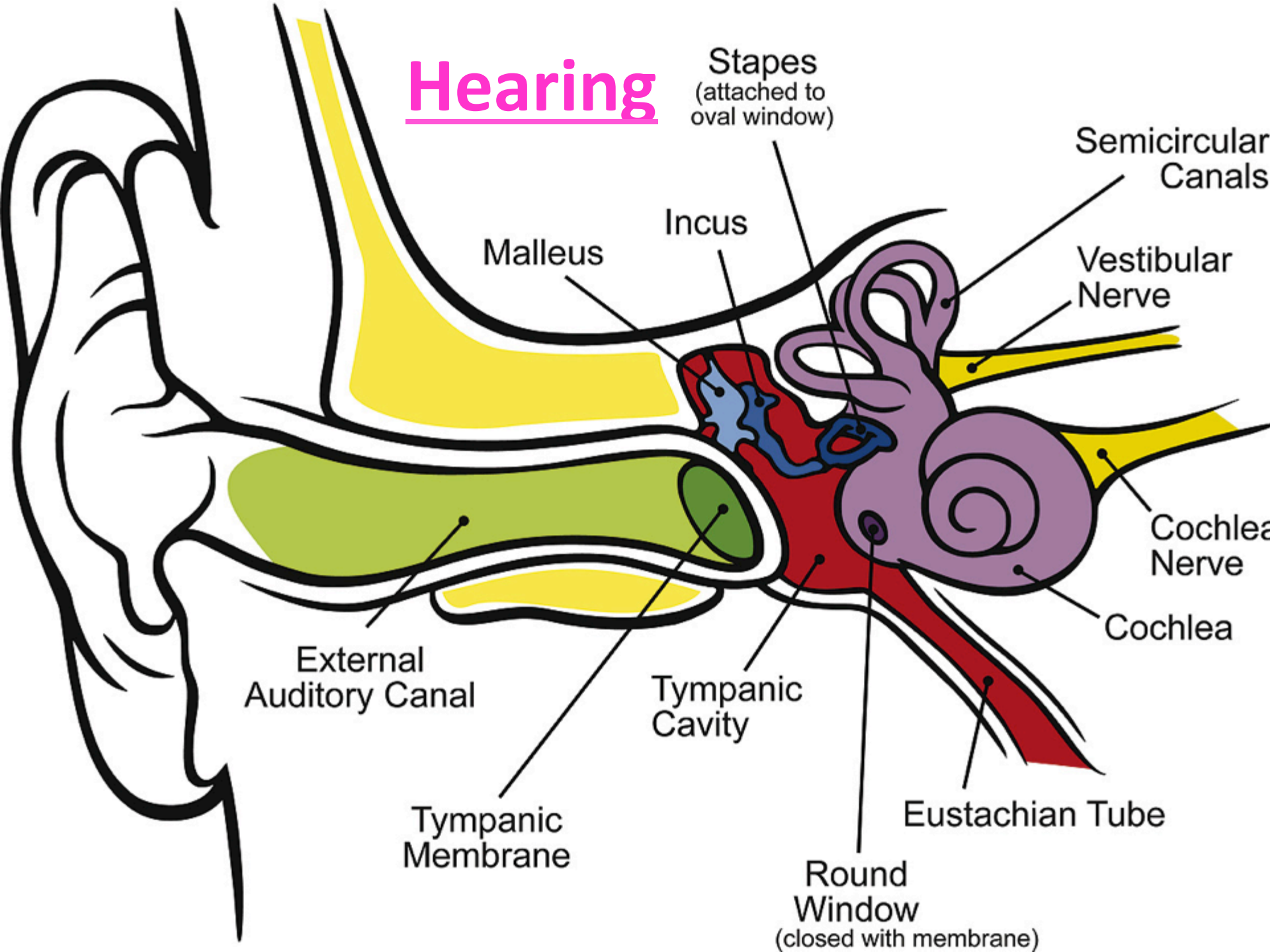


Ear & Hearing Problems

Hearing Loss

Up to 15% of adults with Turner syndrome experience clinical hearing loss

Hearing



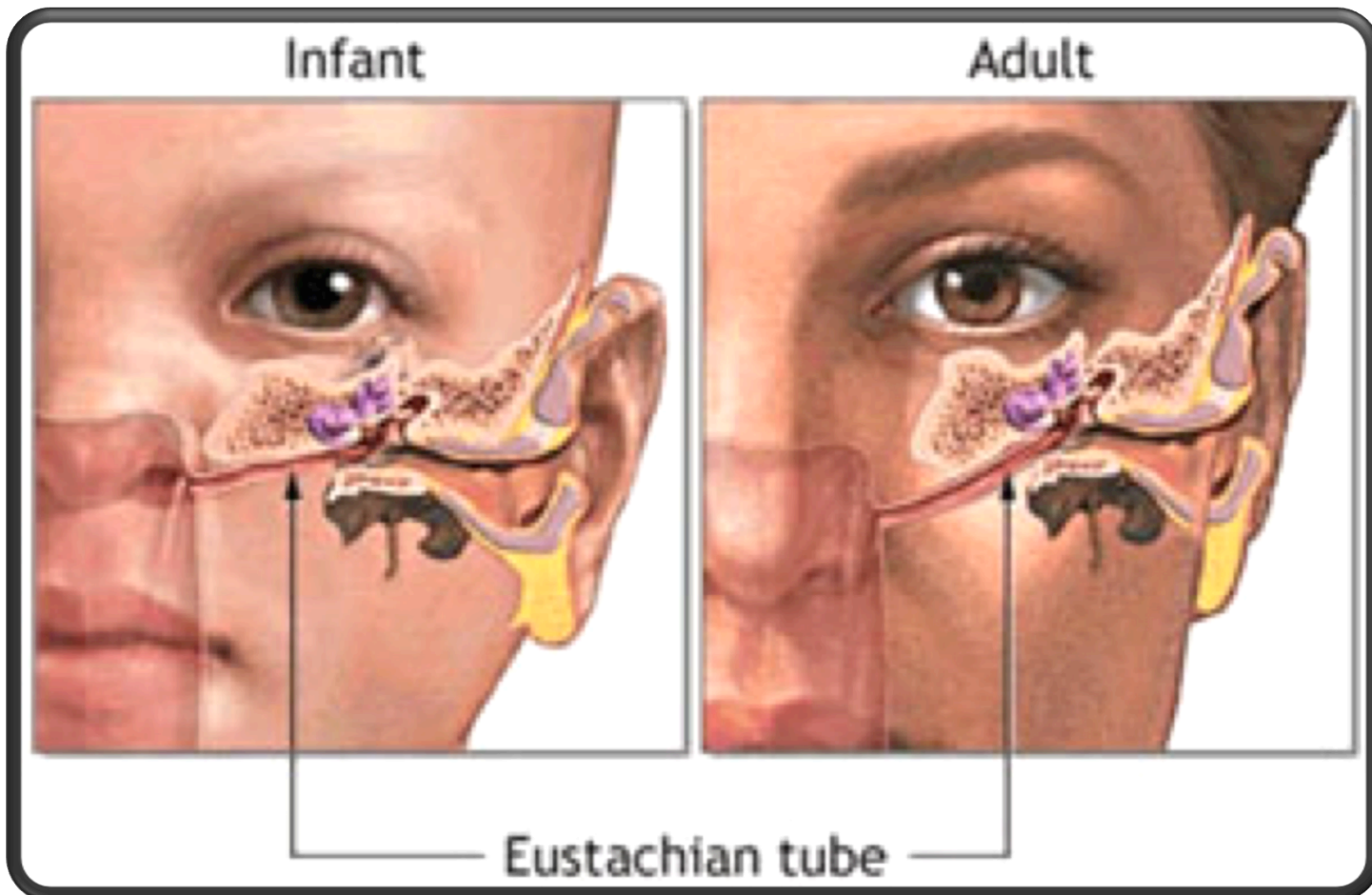
Ear & Hearing Problems

Conductive Hearing Loss

- Common
- Secondary to recurrent Otitis Media (OME)
- Peak OME: ages 1 – 6 years

Ear & Hearing Problems

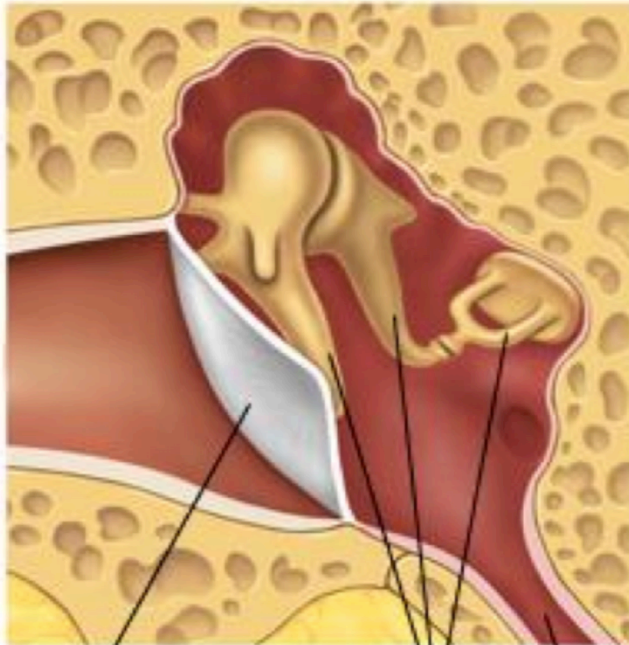
Conductive Hearing Loss



Ear & Hearing Problems

Conductive Hearing Loss

Normal middle ear

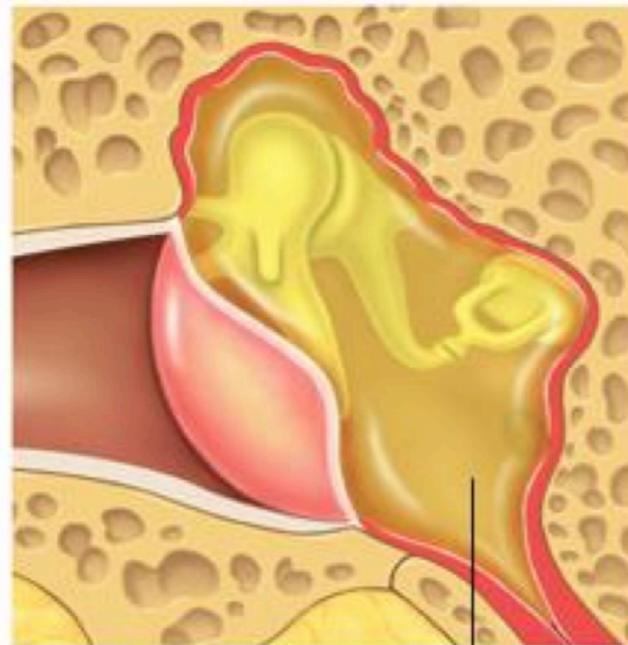


Ear drum

Auditory bones

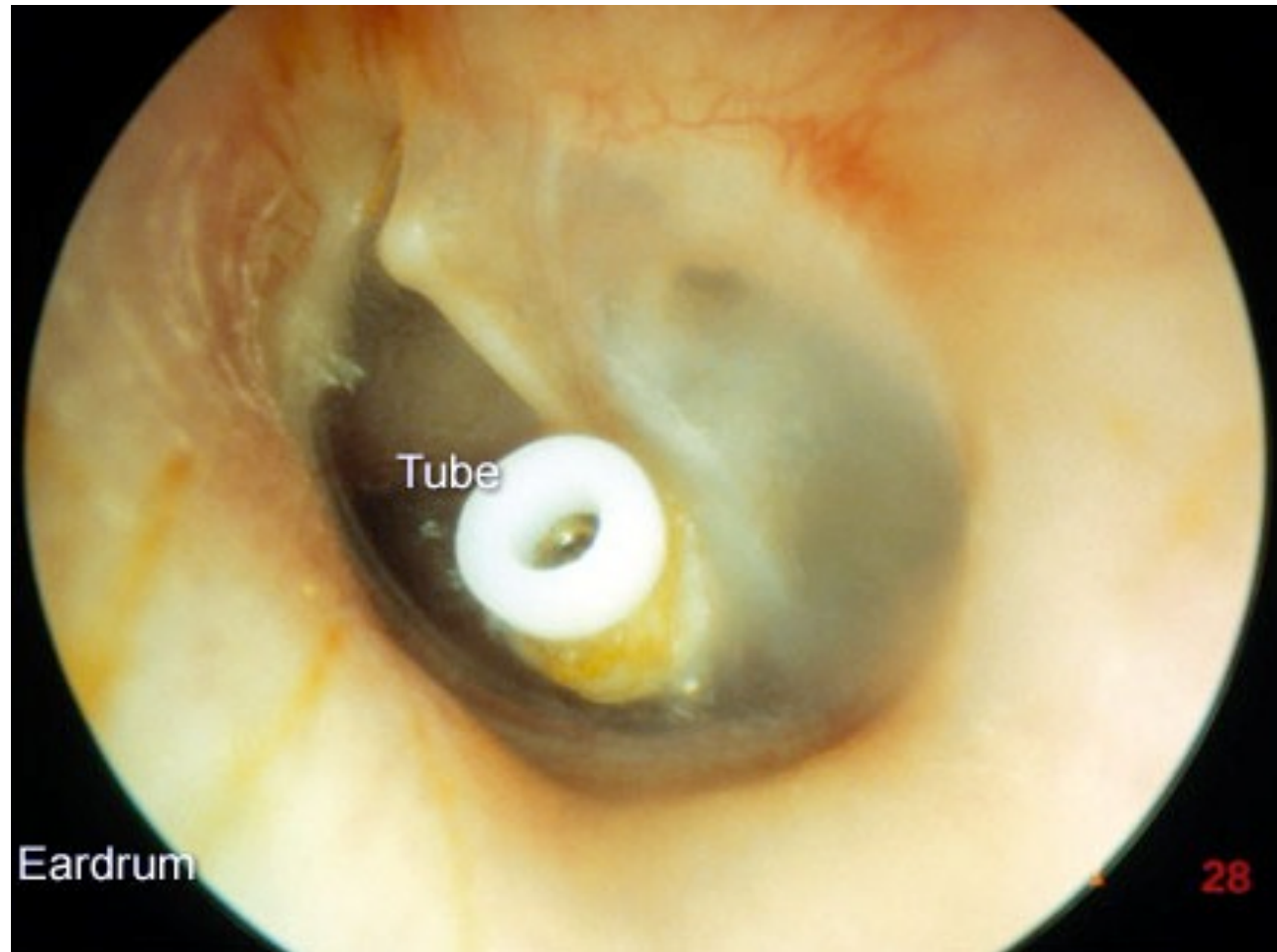
Eustachian tube

Otitis media



**Infected fluid
in middle ear**

Grommet



Ear & Hearing Problems

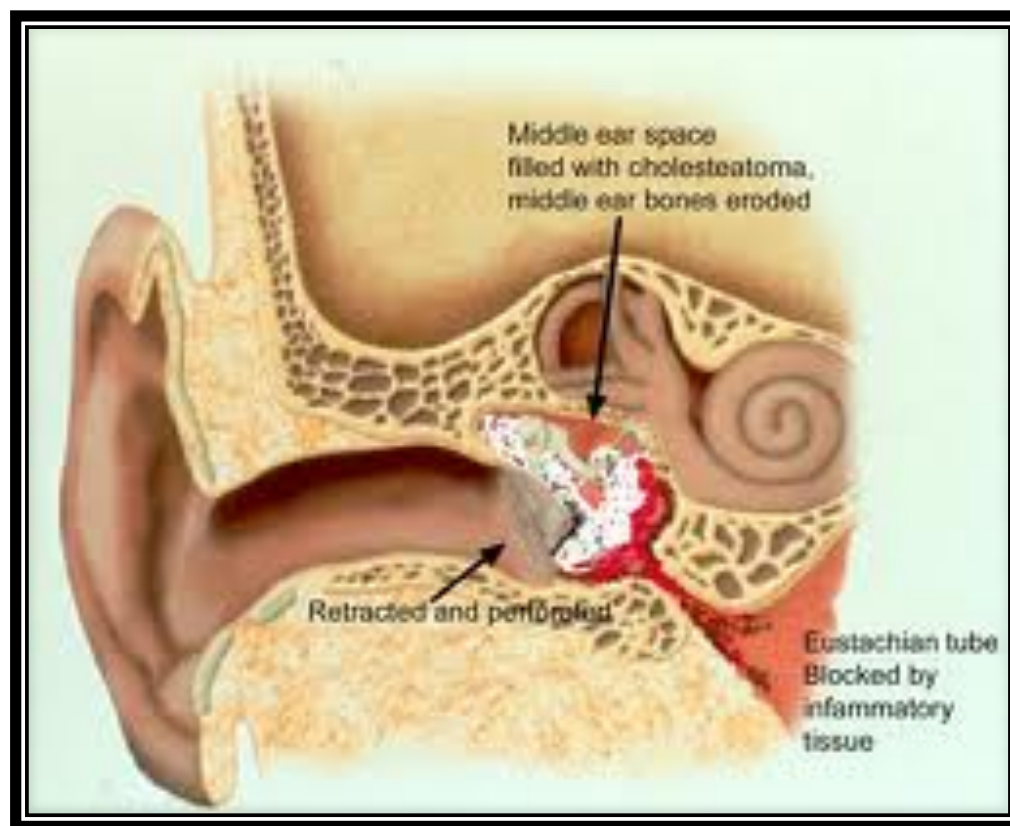
Conductive Hearing Loss

■ Monitoring & treatment necessary to prevent:

- Mastoiditis
- Cholesteatoma
- Deafness

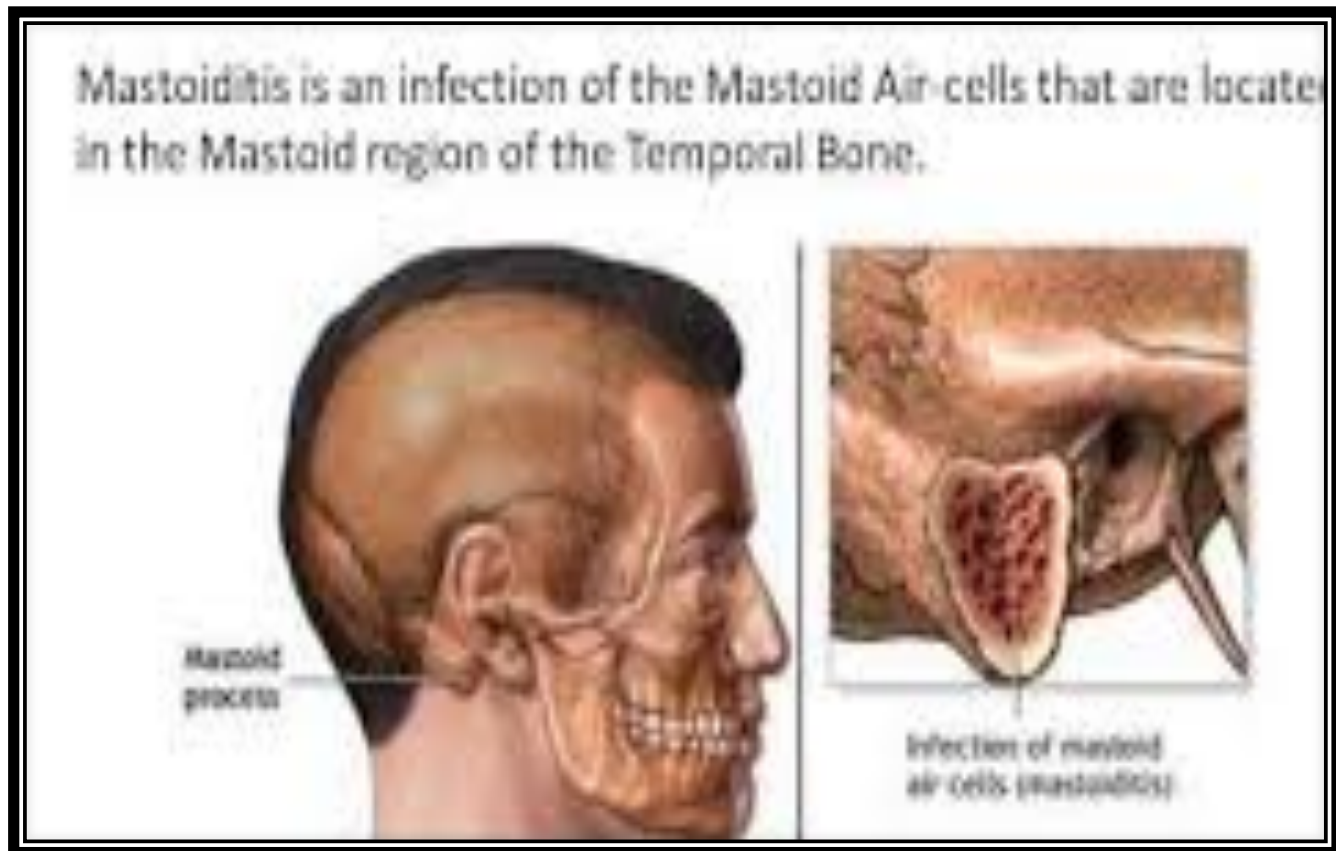
Ear & Hearing Problems

Cholesteatoma



Ear & Hearing Problems

Conductive Hearing Loss



Ear & Hearing Problems

Sensorineural (Nerve) Deafness

- Progressive sensorineural hearing loss
- 50-90% of women with Turner syndrome >30 y.o.
- Detectable by age 6
- Overt hearing loss sometimes apparent in childhood
- Hearing Aids / Other devices

Ear & Hearing Problems

Speech & Language Difficulties

- Speech problems may be secondary to hearing disorders
- Audiology, ENT, Speech Pathologist referrals
- Clinical evidence of hearing loss often only appears in adulthood → review is advised throughout life

Thank You