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

Dr. Deirdre Fitzgerald

MB BCh BAO BMed Sc IMRCSI DOHNS
Senior Registrar Otolaryngology
Royal Victoria Eye & Ear Hospital

TCGI Annual Conference, Naas, 13th April 2013
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

The hypothalamus and the pituitary in the brain control the normal secretion of thyroid hormones which in turn controls metabolism.
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
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

![Diagram showing the difference between infant and adult Eustachian tubes](image)
Ear & Hearing Problems

Conductive Hearing Loss

- Normal middle ear
- Otitis media
  - Ear drum
  - Auditory bones
  - Eustachian tube
  - 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

Mastoiditis is an infection of the Mastoid Air cells that are located in the Mastoid region of the Temporal Bone.
Ear & Hearing Problems

Sensoneural (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
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