

Hearing Loss & Audiologic Management of the Patient with Turner Syndrome

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Introduction

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Newborn Hearing Screening

According to NIH: 2-3 of every 1,000 children born in the U.S. are born with hearing loss

Congenital Hearing Loss is one of the most common birth defects

- Appx. 50% of all cases of congenital HL are genetic
- Appx. 70% of all cases of hereditary HL are nonsyndromic and the other 30% are syndromic

Universal Newborn Hearing Screening = every infant screened prior to hospital discharge receives proper follow up

- Identified by 1 month
- Diagnosed by 3 months
- Intervention by 6 months

How We Hear 101

The ear is divided into 3 parts that lead up to the brain

- The External Ear: sound travels down the ear canal and strikes the ear drum causing it to vibrate
- The Middle Ear: Vibrations from the ear drum cause the ossicles to vibrate which then creates movement of the fluid in the inner ear
- The Inner Ear: the movement of the fluid in the inner ear cause changes in tiny hair cells. Movement of hair cells sends electrical signals from the inner ear to the auditory nerve which leads to the brain.

The brain then interprets these electrical signals as sounds

Degree and Types of Hearing Loss

Hearing loss is described in terms of degree of severity:

- Normal:
 - o 0-15dB for children
 - o 0-25dB for adults

Conductive Hearing Loss:

- Problems within the outer and/or middle ear
- Bone conduction hearing is within the normal range
- Air conduction hearing can range from mild to moderate-severe

Sensorineural Hearing Loss:

- Problems within the inner ear
- Bone conduction and air conduction are approximately equal and can range from mild to profound

Mixed Hearing Loss:

- Problems within the outer and/or middle ear as well as inner ear
- Hearing loss by both bone conduction and air conduction
- Hearing loss via air conduction is greater than hearing loss via bone conduction

Tympanometry

Assessment of the function of the ear drum (tympanic membrane)

Classification:

- Type A – WNL
- Type B – Flat (fluid, cerumen, TM perf)
- Type C – Retracted/negative pressure
- Type Ad – Hypermobility
- Type As – Stiffness

Turner's Syndrome

More than 80% of patients with TS have ear malformations, including low set ears, elongated ears, cup-shaped ears and thick ear lobes.

Conductive Hearing Loss

- Ear infections and otitis media (fluid in the middle ear) are common in childhood because of the position of the Eustachian Tube early in life.
- Children with TS are at even greater risk for middle ear problems and are thought to be pre-disposed to middle ear problems due to ear malformations and abnormalities
- Some persistent middle ear problems can lead to sequelae such as TM perforations, drainage and the development of cholesteatomas.

Sensorineural Hearing Loss

- Studies shows there can be either a progressive SNHL with a midrange bilateral symmetric dip in the 1.5-2.0kHz region or a high frequency downward sloping SNHL
- Most common onset for a patient with TS is during the 2nd and 3rd decades of life but may be present as early as 6 years old.
- This hearing loss may begin in childhood and worsen over time, more than 90% of women with TS in their forties have some degree of HL and more than 25% of them need hearing aids.

Management

Obtain a baseline audiologic evaluation at the time of diagnosis of Turner's Syndrome and follow-up annually

Heightened surveillance for middle ear problems, especially in childhood. Follow-up with ORL specialist for medical management and audiologist to monitor hearing.

If a hearing loss is identified – hearing aids may be recommend dependent on the type and severity of the hearing loss

- If patient is of school age, an FM system may also be appropriate in academic settings.

Academic/Education Support

Speech and Language Intervention

Tips for Healthy Ears and Hearing for All

Refraining from Q-tip use on yourself or child

Hearing Conservation to prevent noise induced hearing loss or additional hearing loss or additional hearing loss from noise exposure

- Ear plugs should be utilized in high noise environments
- Reducing the volume and limiting time listening to personal audio devices.

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