Management of Hearing Loss in Children with Down Syndrome

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**Purpose**

When hearing loss occurs with Down syndrome, children are at increased risk for additional delays in speech, language, and cognitive development. The purpose of this research is to examine the audiologic and ENT management of children with Down syndrome in order to better understand the challenges associated with clinical management in this population and factors that lead to successful and timely intervention.

**Background**

The occurrence of hearing loss and related outer, middle, and inner ear anomalies in individuals with Down syndrome has been well documented in the literature (Diefendorf et al., 1995). As the most frequently occurring chromosomal abnormality present in about 1 in 700 live births (Chin, Khami, & Husman, 2014), the effective management of otologic and audiologic issues is crucial. Common otologic findings in this population include small pinna, stenotic ear canals, frequent cerumen impaction, otitis media with effusion, and in some cases sensorineural hearing loss (Chin et al., 2014). Abnormal nasopharyngeal development in children with Down syndrome leads to the higher susceptibility to upper respiratory tract infections observed when compared to the general population (Diefendorf et al., 1995). Aggressive management of middle ear disease in children with Down syndrome has shown favorable results (Shott et al., 2001). Considering the high occurrence of hearing loss reported in many studies (Laws & Hall, 1994; Diefendorf et al., 1995; Chin et al., 2014; Maurizi, Ottavai, Paladetti, & Lungarotti, 1981), it is likely that the development of speech and language in children with Down syndrome and resulting hearing deficits is negatively affected (Laws and Hall, 2014) and that amplification may provide significant benefits.

**Clinical Questions**

1. What are the challenges associated with medical and audiologic assessment and management of children with Down syndrome?
2. What factors determine whether or not a child with Down syndrome is fitted with amplification?
3. How are children with Down syndrome managed, audiologically, at UNC pediatric audiology?
4. How do parents perceive the benefits and challenges associated with hearing aid use?

**Methods**

This project has four components:

1. A series of structured interviews with pediatric audiologists and otolaryngologists is being conducted in a large tertiary care medical center (UNC Hospitals) to address the clinical questions above.
2. Case history information for children with Down syndrome followed at UNC Hospitals is being analyzed to determine medical and audiologic history and management.
3. Parents of children with Down syndrome who are using amplification will be surveyed to determine their impressions and recommendations.
4. Findings will be summarized and presented to the audiologists and otolaryngologists at UNC Hospitals.

**Results**

**Clinical Questions**

1. Diagnosis of bilateral mild to moderate sensorineural hearing loss at 6 weeks old via ABR
2. Otologic of hearing loss: Connexin 26
3. Fit with hearing aids at 2 months old
4. Received 3 sets of PE tubes to date

**Challenges with Audiologic Diagnosis, Assessment, and Intervention**

1. Inconsistent use of hearing aids due to retention problems early in the process.
2. Hearing aid use has improved over time.
3. Persistent fluid in middle ear space adding a conductive component to hearing loss.
4. Patient fatigue resulting in incomplete behavior testing results.
5. Patient often does not tolerate insert earphones leading to lack of ear specific information.
7. Ear infection requiring numerous attempts to treat – precluded hearing aid use for ~1 month.

**Case Studies**

To date, 22 cases of children with Down syndrome seen at UNC pediatric audiology have been reviewed. The following areas were identified for each case:

- Date of birth
- Birth History
- Comorbidities
- Age at diagnosis of hearing loss
- Number and dates of dB ABR testing: middle ear status at time of ABR
- Challenges experienced during audiologic assessment
- Type and degree of hearing loss
- Age at hearing aid fitting and outcomes
- Dates of PE tube placement and other ENT management

**References**


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