Management of Hearing Loss in Children with Down Syndrome

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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 investigation 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.

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, & Husein, 2014), the effective management of otologic and audiologic issues is crucial. Common otologic findings in this population include small pinnae, 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, 2014; Diefendorf et al., 1995; Chin et al., 2014; Maurizi, Ottaviani, Paludetti, & Lugnarotti, 1985), 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.

To date, 17 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 dx 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

It is difficult to characterize all children with Down syndrome, as they present very differently. Some individuals have complicated birth histories and numerous comorbidities, yet go on to become successful hearing aid users while others do not. Thus, each patient must be considered on an individual basis with the goal of providing the best possible hearing.

Complicating factors that may delay HA fitting:
- Lack of compliance/cooperation during behavioral testing
- Delay in diagnosis due to presence of other medical issues
- Clinical focus on middle ear status without considering amplification needs

March-April, 2015: Completion of interviews with pediatric audiologists and otolaryngologists
April-May, 2015: Complete case reviews
May-August, 2015: Interview parents/family members
September, 2015: Present findings and recommendations to audiologists and otolaryngologists at UNC Hospitals

References


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