Management of Hearing Loss in Children with Down Syndrome

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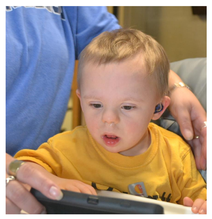
Purpose

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 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.

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, & Husein, 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, 2014; Diefendorf et al., 1995; Chin et al., 2014; Maurizi, Ottaviani, Paludetti, & Lungarotti, 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.


Clinical Questions

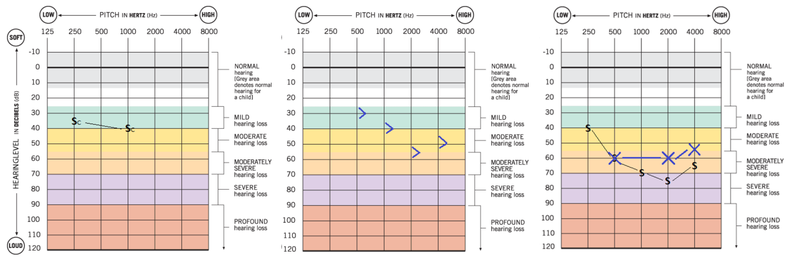
1. What are the challenges associated with medical and audiologic assessment and management of children with Down syndrome?
2. What factors determine whether or not a child with Down syndrome is fitted with amplification?
3. How are children with Down syndrome managed, audiologically, at UNC pediatric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

Case Example



First behavioral test
January, 2013
S = Soundfield c = One-time Response

Bone Conduction Responses
March, 2013

Most recent behavioral test
November, 2015
S = Soundfield X = Left Ear Response

Hearing History

- Diagnosed with bilateral mild to moderate sensorineural hearing loss at 6 weeks old via ABR
- Etiology of hearing loss: Connexin 26
- Fit with hearing aids at 2 months old
- Received 3 sets of PE tubes to date

Comorbidities

- VSD
- Jaundice
- Difficulty feeding after birth
- Positional plagiocephaly
- Low birth weight
- Sleep apnea

Challenges with Audiologic Diagnosis, Assessment, and Intervention

- Inconsistent use of hearing aids due to retention problems early in the process
  - Hearing aid use has improved over time
- Persistent fluid in middle ear space adding a conductive component to hearing loss
- Patient fatigue resulting in incomplete behavior testing results
- Patient often does not tolerate insert earphones leading to lack of ear specific information
- Small ear canals make visualization of tympanic membrane (eardrum) difficult
- Ear infection requiring numerous attempts to treat – precluded hearing aid use for ~1 month

Interview Outcomes

Pediatric Audiologists

Key factors considered regarding amplification (or not):

- Completeness of audiologic diagnosis
  - ABR, behavioral data, treatment of middle ear pathology
- Degree of hearing loss
- Parents' level of motivation and perception of need

Type of hearing loss does not impact decision to provide amplification to child given:

- Medical management must be completed
- Permanent nature of hearing loss must be confirmed
- Family involvement/motivation must be high
  - Ex: Fluctuating hearing loss with chronic conductive cases - must ensure that family will return for follow up

Reasons for not fitting a child with Down syndrome with hearing aids:

- Severe behavioral challenges will limit hearing aid use

Amount of hearing loss before recommending amplification:

- 25 dB PTA or greater

Otolaryngologists

Key factors considered when managing otologic care:

- Degree of hearing loss
- Status of middle ear/presence of OM

Reasons for delay of amplification

- Complexity of other medical issues (cardiac, breathing, etc.)

Opinion on amplification for children with Down syndrome

- Amplification should be provided to children with hearing loss that cannot be corrected surgically

Amount of hearing loss before recommending amplification:

- Outside of normal would qualify a child, with Down syndrome or not, for amplification

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 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

HA Users	HA Use Not Attempted	HA Use Attempted but Rejected
16	4	2

Type of hearing loss						
SNHL	CHL	Mixed	One CHL, one SNHL	One mixed HL, one CHL	Unspecified	No HL
9	5	2	1	1	3	1

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

Challenges associated with audiologic assessment and management:

- Stenotic ear canals precluding immittance measures
- Maintaining retention of hearing aids
- Draining ear canals
- Fluctuating nature of conductive components

Next Steps and Future Directions

- November-December, 2015: survey parents/family members
- Spring 2016: Summarize findings into manuscript
- Submit for publication

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