Management of Hearing Loss in Children with Down Syndrome

*Dani Warmund, B.S. (NC-LEND) *Jackson Roush, Ph.D., **Patricia Roush, Au.D., **Craig Buchman, M.D.

*Division of Speech and Hearing Sciences; Dept. of Allied Health Sciences; **Department of Otolaryngology

University of North Carolina School of Medicine, Chapel Hill , NC

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 interviewed to determine their impressions and recommendations.
4. Findings will be summarized and presented to the audiologists and otolaryngologists at UNC Hospitals.

Interviews

Pediatric Audiologists

1. What are key factors you consider when deciding whether or not to recommend amplification for a child with Down syndrome?
2. Are these considerations different when the child has a permanent sensorineural or conductive hearing loss vs. a chronic conductive hearing loss?
3. What are some reasons why you would not fit a child with Down syndrome with hearing aids?
4. How long would you wait before considering amplification for a young child with chronic conductive hearing loss?
5. How much hearing loss do you think a child with Down syndrome should have before you would consider recommending amplification?
6. In general, do you think we are providing amplification to children with Down syndrome in a timely fashion?
7. What has been your experience with families regarding acceptance of recommendations for amplification in children with Down syndrome?

Otolaryngologists

1. What factors do you consider when managing otologic care for children with Down syndrome?
2. Do you think children with Down syndrome who have permanent hearing loss receive amplification in a timely fashion?
3. What are your thoughts regarding amplification for children with Down syndrome when there is chronic conductive hearing loss that cannot be adequately managed with traditional treatments such as PE tubes?
4. How much hearing loss do you feel is necessary before you would recommend amplification for a child with Down syndrome? Do you feel this is different than the amount of hearing loss needed before considering amplification in a child without Down syndrome?
5. At what point do you think a child with Down syndrome and recurrent ear disease should be fitted with amplification?
6. What are the key points you discuss with families regarding management of middle ear problems in children with Down syndrome?
7. What factors do you consider when deciding whether or not to recommend tube placement for children with Down syndrome?

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
- 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

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 ongoing hearing loss that cannot be corrected with tubes

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, 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

Successful HA Users	HA Use Not Attempted	HA Use Attempted but Rejected
13	2	2

Type of hearing loss					
SNHL	CHL	Mixed	One CHL, one SNHL	One mixed HL, one CHL	Unspecified
9	3	0	1	1	3

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

Next Steps and Future Directions

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

- Chin, C. J., Khami, M. M., & Husein, M. (2014). A general review of the otolaryngologic manifestations of Down Syndrome. *International Journal of Pediatric Otorhinolaryngology,* 78, 899-904.
- Diefendorf, A. O., Bull, M. J., Casey-Harvey, D., Miyamoto, R. T., Pope, M. L., …, Wagner-Escobar, M. (1995). Down Syndrome: A multidisciplinary perspective. *Journal of the American Academy of Audiology*, 6, 39-46.
- Laws, G. & Hall, A. (2014). Early hearing loss and language abilities in children with Down syndrome. *International Journal of Communication Disorders*, 49 (3), 333-342.
- Maurizi, M., Ottaviani, F., Paludetti, G., & Lungarotti, S. (1985). Audiological findings in Down's children. *International Journal of Pediatric Otorhinolaryngology*, 9, 227-232.
- Shott, S. R., Joseph, A., & Heithaus, D. (2001). Hearing loss in children with Down syndrome. *International Journal of Pediatric Otorhinolaryngology*, 61, 199-205.

Acknowledgements

This work was sponsored, in part, by NC-LEND (Leadership Education in Neurodevelopmental and Related Disorders) through a grant from the U.S. Maternal and Child Health Bureau to the Carolina Institute for Developmental Disabilities, which provided funding for Ms. Warmund