



Screening, Diagnosis, and Management of Hearing Loss in Children with Significant Cognitive Disabilities



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Developmental Disabilities in Children who are Deaf or Hard of Hearing

- Developmental disabilities (DD) affect 40% to 56% of children with permanent hearing loss (HL) (Guardino, 2008; Gallaudet Research Institute, 2013)
- The most prevalent developmental disabilities to co-occur with HL (Van Naarden Braun, 2015) include:
 - Intellectual Disability (23%)
 - Cerebral Palsy (10%)
 - Developmental Delay (10%)
 - Learning Disability (10%)
 - Autism Spectrum Disorder (7%)
 - Vision Impairment (5%)
- The prevalence of HL+DD is increasing (Musyoka, 2017)
- HL+DD can affect communication, learning, socialization and other developmental domains (Caron & Rutter, 1993)

When Hearing Loss occurs with Developmental Disabilities the Hearing Loss is likely to be *Under-identified*, and even when diagnosed, *Under-served*

- Undiagnosed hearing loss in children who have HL+DD ranges from 23% (Evenhuis, 2001) to 75% (Hey et al., 2013)
- Nearly 50% of students with the most severe intellectual and physical disabilities and a known hearing loss do not use hearing aids, and among those who do, fewer than 25% use an FM system in the classroom (Erickson & Quick, 2016)

Why is Hearing Loss Under-identified and Under-served in Children with Developmental Disabilities?

- Overshadowing of hearing loss by other conditions or diagnoses (Erickson & Quick, 2016)
- Perceptions of limited academic potential (Erickson & Quick, 2016)
- Challenges associated with audiologic assessment (Roush et al, 2004)
- Challenges associated with measuring outcomes/benefits of hearing aids and cochlear implants (Wiley et al, 2005; Archbold et al, 2015)

Children with “Significant Cognitive Disabilities” often require specialized audiology procedures

ABR with sedation

ABR under general anesthesia

Adapted cochlear implant evaluation

- ‘Significant cognitive disabilities’ occur in approximately 1% of the school-age population and include the most severe intellectual and physical disabilities (U.S. Department of Education)
- Many of these children require specialized clinical procedures for evaluation of hearing and assessment of benefit from hearing aids or cochlear implants

What can be done to improve Screening, Diagnosis, and Management of Hearing Loss in Children with Significant Cognitive Disabilities?

- Ensure there is comprehensive and systematic review of the child’s hearing history and audiologic management
- Provide support and technical assistance to parents and school personnel
- Determine if some of the child’s audiologic management can be provided in the school setting

Audiology and Hearing-Related Services that can be Provided in the Educational Setting

- Otoscopy
- Physiologic Measures
 - Otoacoustic Emissions
 - Acoustic Immittance
- Behavioral Measures
 - Conditioned Play Audiometry
- Hearing-Related Functional Outcome Measures
- Orientation to care and use of hearing technology for school personnel
- Ongoing communication with the child’s clinic-based audiology service providers

Limitations of School-Based Audiology Services for Children with Significant Cognitive Disabilities

- Pediatric audiologist/s must be on site
- Instrumentation must be transported to/from the school
- ABR and VRA are unfeasible in most educational settings
- ENT/medical personnel not on site
- Less control over acoustic environment
- Family members not likely to be present

Potential Advantages of School-Based Audiology Services for Children with Significant Cognitive Disabilities

- Environment is familiar to the child
- SLPs, OTs, special educators, and others who know the child are available to assist with assessment and evaluation of aided benefit
- Transportation is less complicated for non-ambulatory children
- Greater flexibility in scheduling

Comprehensive Review of Hearing History and Audiologic Management for Children with Significant Cognitive Disabilities Includes:

- Outcome of newborn hearing screening obtained via statewide database
- Consideration of technology used for hearing screening (ABR/OAE) and whether ANSD has been ruled out
- Results of any diagnostic audiology evaluations and recommendations
- Current status and next steps for audiologic follow-up

Future Directions

- Development of a review protocol for systematic analysis of EHDl outcomes in children with developmental disabilities
- Application of the above protocol to ~130 children with significant cognitive disabilities in a self-contained educational setting in North Carolina
- Continued field testing of selected school-based audiology services for children with severe cognitive disabilities
- Development of a hearing-related functional outcome measure for children with developmental disabilities including those with significant cognitive disabilities
- Ongoing efforts to raise awareness of hearing loss in children with developmental disabilities

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Acknowledgements

We are grateful to the US Maternal and Child Health Bureau (MCHB) and the Health Resources and Services Administration (HRSA) Universal Newborn Hearing Screening Program for funding a Pediatric Audiology Supplement to the North Carolina LEND Program that enables us to provide specialized training to these and other graduate students at UNC.