Hearing Loss in Children with Down Syndrome

By Emily Nightengale, AuD

In 2011, the American Academy of Pediatrics published health care practice guidelines for children with Down syndrome that included monitoring for hearing loss. The guidelines recommend hearing evaluations every six months until school age, then annually through childhood. More frequent evaluations may be necessary or recommended should abnormal results be obtained or concerns for hearing arise (*Pediatrics.* 2011;128[2]:393). The guidelines were established since children with Down syndrome are at a higher risk for hearing loss as compared with their normally developing peers.

The anatomical ear structure of children with Down syndrome has characteristics that may predispose them to hearing deficits. They are more prone to conductive hearing loss secondary to cerumen impaction and middle ear pathologies, including, among others, middle ear effusion, acute otitis media, and eardrum perforations (*Otol Neurotol.* 2015;36[2]:348; *Eur Arch Otorhinolaryngol.* 2014;271[5]:863; *Otolaryngol Clin North Am.* 2012;45[3]:599). Otitis media with effusion has been reported in as many as 93 percent of children with Down syndrome (*Scott Med J.* 2011;56[2]:98). About 43-83 percent receive pressure equalization tubes for treatment of this recurrent and/or persistent issue (*Int J Pediatr Otorhinolaryngol.* 2001;61[3]:199). However, prior research indicates that surgical interventions for treatment of conductive hearing loss may not be as successful in this population (*Int J Pediatr Otorhinolaryngol.* 1999;49[2]:143; *Eur Arch Otorhinolaryngol.* 2014; *Otolaryngol Clin North Am.* 2012; *Am J Med Genet C Semin Med Genet.* 2006;142C[3]:131). Additionally, sensorineural hearing loss has been documented in the Down syndrome population, with computed tomography scans revealing structural abnormalities in the inner ear, such as narrow internal auditory canals and semicircular canal malformations (*Laryngoscope.* 2006;116[12]:2113; *Pediatr Radiol.* 2012;42[12]:1449).

DEVELOPMENTAL IMPLICATIONS

As with normally developing children, hearing loss is associated with poor language outcomes and can affect long-term development. Identification of hearing loss and awareness of its implications in children with Down syndrome is critical since this population may suffer from additional detrimental effects on cognitive abilities, such as poorer performance in sentence imitation, language comprehension, speed of word processing, and sensitivity to brief acoustic cues compared with age-matched peers with intellectual impairment (*Down Syndrome Research and Practice.* 1995;3[3]:75).

shutterstock/Monkey Business Images

Identification of hearing loss and awareness of its implications in children with Down syndrome is critical, since this population may suffer additional detrimental effects on cognitive abilities.

Current literature on this topic shows several limitations because of multiple reasons, including small participant

Dr. Nightengale is an audiologist at Children's Hospital Colorado. She specializes in pediatric diagnostic evaluations and management. She received her AuD degree from Rush University in Chicago, Illinois.

numbers, data collection spanning many years, variable disparities, no report of ethnic or race information, and limited information regarding audiologic interventions and treatment (*Res Dev Disabil.* 2013;34[7]:2251; *Otolaryngol Head Neck Surg.* 1979;87[3]:372; *Am J Ment Defic.* 1986;90[6]:636; *Chin Med J (Engl).* 2015;128[8]:1091).

To address these gaps, the Sie Center for Down Syndrome and the Audiology Department at the Children's Hospital Colorado formed a team to investigate hearing loss and hearing aid use in this patient population. The Sie Center for Down Syndrome is a multi-disciplinary clinic that provides quality care for children with Down syndrome and support to their families and caretakers. When the clinic opened in 2010, the center's medical director, Francis Hickey, MD, realized that many children were not receiving audiology services, despite the estimated 16-78 percent incidence of hearing loss in this population (*Res Dev Disabil.* 2013; *Otolaryngol Head Neck Surg.* 1979; *Am J Ment Defic.* 1986; *Chin Med J (Engl).* 2015). In the first year, he referred more than 244 children to the Bill Daniels' Center for Children's Hearing for evaluation and treatment.

STUDY RESULTS

Our study focused on four areas: confirmed permanent hearing loss, transient hearing loss, tympanometry results, and hearing aids/amplification (*Am J Audiol.* 2017;26[3]:301). Some 582 audiologic evaluations of 308 children with Down syndrome who received care in 2013 were analyzed. Information included results from audiometric testing, normal or abnormal findings, and testing related behaviors. Nearly 25 percent of patients were identified to have permanent hearing loss, with the majority having a permanent conductive hearing loss (38%) in bilateral ears (75%).

Surprisingly, the results indicated that middle ear effusion is not just an issue in childhood, but is a persistent problem from infancy through early adulthood, with an incidence that ranges from 37 to 69 percent.

Of note, sensorineural hearing loss and mixed hearing loss were documented in 34 and 28 percent of patients, respectively. There was no statistically significant difference in permanent hearing loss among ethnic groups (non-Hispanic/Latino versus Hispanic/Latino patients). Transient hearing loss secondary to middle ear effusion or otitis media was reported in at least 22 percent of children with Down syndrome. This is a conservative estimate, as this group consisted of children who had at least one normal and one abnormal evaluation in 2013. Again, there was no significant difference among ethnic groups for transient hearing loss. Due to the high incidence of transient hearing loss, tympanometry results across the pediatric age groups were investigated. Abnormal tympanograms were identified in nearly 40 percent of patients, and another 18 percent had results suggestive of patent pressure equalization tubes. Historical data revealed that 58.8 percent of participants had a history of ear tubes for otologic and/or audiologic treatment. Surprisingly, the results indicated that middle ear effusion is not just an issue in childhood, but is a persistent problem from infancy through early adulthood, with an incidence that ranges from 37 to 69 percent. Lastly, amplification devices were used by or recommended to 93 percent of patients with permanent hearing loss. Twelve children used hearing aids to treat recurrent transient hearing loss to receive consistent speech and environmental stimuli.

PREVALENCE AND TREATMENT OPTIONS

Study results clearly revealed a high prevalence of hearing loss (36%) in children with Down syndrome. They can experience permanent (conductive, sensorineural, and mixed hearing losses were identified in significant numbers) and transient hearing loss secondary to a middle ear disease. The study findings are consistent with previous literature and help support the published guidelines for management of hearing loss. Ethnicity was not a significant variable in terms of incidence of hearing loss. The study also found a high rate of abnormal tympanograms among pediatric patients. These were commonly reported from infancy to early adulthood, further emphasizing the need for ongoing monitoring and management. Otologic treatment, including placement of pressure equalization tubes, is the first step to improve the hearing and developmental outcomes of many of these children. If medical intervention is not an option (i.e., for those with sensorineural hearing loss), recommended, or accepted by parents/caregivers, then amplification is a viable option. This study found that nearly all the participants with permanent hearing loss either used hearing aids or received a recommendation to try amplification. Some participants without a confirmed permanent hearing loss were even treated with hearing aids.

A primary goal of pediatric audiology is to ensure that children have adequate hearing to meet their communication and learning needs. When a specific population is identified to have a high risk for hearing loss, close monitoring and management is recommended. Children with Down syndrome fall into this category. This study supports the need to educate physicians, parents, audiologists, and teachers on the importance of referrals, early identification of hearing loss, and treatment, including medical interventions and/or amplification. These action items are vital to support a child's linguistic, psychosocial, and cognitive development.

* *The author acknowledges the contributions of the following study co-authors: Patricia Yoon, MD; Kristy Wolter-Warmerdam, PhD ABD; Dee Daniels, RN CPNP; and Fran Hickey, MD, DO.*