

Chapter 6

Children Who Are Deaf or Hard of Hearing PLUS

Susan Wiley, PhD; Rachel St. John, MD, FAAP; & Candace Lindow-Davies

PLEASE NOTE: For the purposes of this chapter, the term “hearing loss” and “disability” are medical descriptors that may apply to newborns and infants who are deaf or hard of hearing at birth, as well as those who lose hearing at a later time. The authors fully recognize that individuals may not self-identify as having a disability and may prefer the terms “deaf” or “hard of hearing.” We support the inclusion of those terms when communicating with families, caregivers, and health professionals.

Early Hearing Detection and Intervention (EHDI) has made great advances in screening, early identification, and intervention for children who are deaf or hard of hearing (D/HH), which has helped to improve their language outcome (Kennedy et al., 2006; Moeller, 2000; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998). While this is encouraging, approximately 40% of children who are D/HH have medical or developmental difficulties (Gallaudet Research Institute, 2011) that can delay the age of identification and intervention for services, as well as require an expanded team to support the whole child. These children have been described as being “D/HH Plus” to indicate their needs while still keeping a positive developmental perspective for the future. When supporting families through the EHDI process, it is important to keep in mind that the complexity of these children can increase family stress.

Some of the likely reasons for a high rate of children who are D/HH Plus relate to some of the risk factors for hearing loss

(Joint Committee on Infant Hearing, 2007). These risk factors for hearing loss can overlap with risk factors for developmental delays (First & Palfrey, 1994), such as certain genetic syndromes, prematurity, congenital infections, and meningitis. These factors can impact broader brain development in a variety of developmental domains, as well as impact a child’s ability to acquire language. This is important to recognize, as often our first approach to understanding a child’s learning pattern is to focus on the effectiveness of amplification, intervention strategies, and family follow-through with day-to-day practice. However, there can be neurological contributions to a child’s slow rate of progress. Furthermore, children who are D/HH can have other reasons for developmental delays based on their family history or factors which are not related to hearing loss (such as lead exposure or family history of developmental or learning problems). It is important to keep in mind that a genetic cause of hearing loss does not protect a child from other factors that can impact

As children who are D/HH Plus can be more medically complex, it is important to be proactive in terms of monitoring.

health and development (Kenna et al., 2007; Wiley, Choo, Meinzen-Derr, Hilbert, & Greinwald, 2006).

To understand the types of issues that can exist in children who are D/HH, *Table 1* describes the rates of conditions among children who are D/HH (Gallaudet Research Institute, 2011; Nikolopoulos, Lioumi, Stamaataki, & O'Donoghue, 2006; Szymanski, Brice, Lam, & Hotto, 2012) as compared to rates within the general population (Boyle et al., 2011). There are challenges in understanding this data for children who are D/HH based on sampling, methods of study, and definitions and diagnostic clarity (particularly for specific learning disabilities and autism spectrum disorders).

As children who are D/HH Plus can be more medically complex, it is important to be proactive in terms of monitoring. While all children who are D/HH should have an ophthalmological exam, be seen by an otolaryngologist with knowledge in hearing loss, and be offered a genetics consultation (Joint Committee on

Infant Hearing, 2007), it is particularly important to provide ongoing surveillance of these children for vision and medical conditions. While a family may receive an identification of a genetic reason for their child's hearing status, the role of a geneticist is often broader than just focusing on hearing. There are practice parameters and considerations that would apply if children have an additional identification, such as an intellectual disability or autism spectrum disorder. Parents may also need to be counseled in terms of understanding that a genetics consultation is not a guarantee that a genetic reason for their child's hearing status will be found. Pediatric neurologists, developmental pediatricians, and pediatric medicine and rehabilitation specialists may play a role in children with cerebral palsy and vision impairment. In some children, it may be challenging to obtain a good vision exam. Children who have had brain-based problems can also have vision problems that are related more to how the brain processes what it sees rather than how the eye itself sees (cortical vision impairment).

Table 1
Rates of Conditions among Children Who Are D/HH

| Type of Disability | Rates Among Children Who Are D/HH | Rates in the General Population |
|-------------------------------------|-----------------------------------|---------------------------------|
| No disabilities | 60% | 86% |
| Cognitive (intellectual disability) | 8.3% | 0.71% |
| Cerebral palsy | -- | 0.3% |
| Blindness and vision impairment | 5.5% | 0.13% |
| ADHD | 5.4% | 5-10% |
| Specific learning disability* | 8% | 5-10% |
| Autism spectrum disorder | 7% | 1% |

*The Individuals with Disability Education Act of 2004 [34 CFR 300.8(c)(10)] indicates children cannot be identified with a specific learning disability (SLD) if the child's performance is the result of a "hearing disability" (U.S. Department of Education, 2004). However, many educators recognize the possibility of SLD presenting in children who are D/HH.

1-3-6 Rule

There have been some studies evaluating the 1-3-6 rule (screening by 1 month of age, identification by 3 months of age, and services by 6 months of age) for children who are medically complex. Understandably, infants with serious medical issues may experience a delay in hearing screening while those medical needs are being addressed. In a study by Chapman et al. (2011), the investigators linked statewide newborn hearing screening, birth defects registry, and birth certificate data with age of screening and identification of children who are D/HH (Virginia births 2001-2006). They found that children with isolated hearing loss were screened 25 days earlier and achieved identification >2.5 months earlier than children with complexities. They found a **greater risk of later screening or identification among preterm births and in children with multiple birth defects**. There was also an impact based on race and maternal education (black, non-Hispanic race, and maternal education ≤12 years).

The authors suggested some approaches to help reduce this gap, including completing diagnostic hearing assessments prior to discharge from the neonatal intensive care unit and using strategies for care coordination among children with medical complexities who do not pass hearing screening. This information points out the need for a broader system of care and care coordination for children who are D/HH Plus.

The Joint Committee for Infant Hearing has also recognized the unique needs of this group of children related to early intervention services (Joint Committee on Infant Hearing, 2013). In addition to the broad supports suggested in the supplement for all children who are D/HH as they relate to Part C programming, the committee included a goal (Goal 4) **that all children who are D/HH**

with additional disabilities and their families have access to specialists who have the professional qualification and specialized knowledge and skills to support and promote optimal developmental outcomes. Specifically, the JCIH encourages the knowledge and skills shown in *Table 2*.

Table 2 Professional Knowledge and Skills Encouraged by the JCIH

Monitoring

Monitoring and recognizing developmental concerns to design an effective intervention program.

Working

Working in teams with multiple specialists to support the best possible child and family outcomes.

Modifying

Modifying strategies to accommodate children's needs.

Empowering

Empowering families in their understanding of reports from specialists and providers.

Supporting

Supporting care coordination across all specialties that a child may need.

Recognizing

Recognizing the importance of potential expanded equipment needs (both communication and mobility devices).

Partnering

Partnering with families to prioritize a child's needs and services.

All children who are D/HH with additional disabilities and their families should have access to specialists who have the professional qualification and specialized knowledge and skills to support and promote optimal developmental outcomes.

The importance of true teamwork and collaboration cannot be understated for this group of children. Team members must understand their contributions as well as their limits and be active listeners of other disciplines—including listening to the significant input by parents and caregivers—in order to effectively adapt interventions for children who are D/HH Plus. ***Effective communication, paired with understanding the goals and priorities important to families, will help teams provide appropriate care and supports.*** Team members will benefit from being open to new ideas, being creative, and considering co-treatment approaches when appropriate. As the team establishes realistic goals with the family as the hub, relies on structured assessment and planning tools, and tries creative approaches and adaptations as needed, we may be able to strengthen the current services that children who are D/HH Plus receive.

The authors of this chapter have noted in their experience partnering with families with children who are D/HH Plus that it is easy for parents to feel alone in their experience. Often, their children do not “fit” into a traditional group where they can find support. These families are also often under significant time and financial pressure in caring for their child and may not be able to use available resources. Parents also may feel that their health care providers do not fully understand the emotional drain of caring for their children. Parent support resources are a critical part of parental well-being and success in caring for their children. These resources are different from state to state, and some areas may have resources that are relatively easy to access (i.e., the Texas Parent-to-Parent Program; www.txp2p.org), while other states do not.

The Internet can be a nonthreatening place to start looking for parent support resources, and parents will often naturally go there in an attempt to connect with other parents sharing their experience. It is helpful to recognize both the strengths and limitations of the Internet as a resource for families. Professionals can share websites

that may be of value for families, as well as encourage families to consider how website information may or may not apply to their child. Many families learn strategies and interventions from other families through the Internet. It is also helpful to provide guidance on whether these potential strategies and interventions are safe, effective, and appropriate considering the unique needs of their child. Encouraging families to inform you of what they are finding and being open to discussing them in a respectful and thoughtful manner can build strong partnerships with families.

It is all too easy to focus on things that children who are D/HH Plus cannot do rather than what they can. Dr. T. Jones from Gallaudet University has provided “rules” to guide instructions that are particularly related for this group of children (see *Table 3*).

Table 3 Dr. T. Jones “Rules” to Guide Instruction

- 1 **Focus on the donut, not the hole.**
- 2 **Celebrate successes, great or small.**
- 3 **If a dead man can do it, it is not an appropriate objective** (this is especially important when setting up learning and academic goals for children).
- 4 **Use meaningful contexts** to make concepts explicit (children are active participants in learning, and teaching is matched to them as unique individuals with unique needs).

We conclude this chapter with a quote and guidance from Candace Lindow-Davies of Minnesota Hands & Voices, who has a son who is D/HH Plus. She speaks beautifully about both the challenges and the rewards that having a child who is D/HH Plus brings to the table:

Effective communication, paired with understanding the goals and priorities important to families, will help teams provide appropriate care and supports.

D/HH Plus is meant to be a positive term, not in any way negative or insensitive to the child who has medical issues along with hearing loss.

- 1 Keep expectations high for children who are D/HH Plus. Often multiple diagnoses can lead some professionals to lower expectations for these children.
- 2 Seek additional training and experience with children who are D/HH Plus to help parents tweeze out the root cause of a particular issue. For example, “Is this a need related to being deaf or having autism?”
- 3 Assist families and professional collaborators to recognize that communication is critical to understanding other contributing challenges. If the child does not have adequate language development as a result of being D/HH, this will contribute to a child being misidentified as having learning disabilities, etc.
- 4 Help families adjust to multiple diagnoses. Some parents can easily accept some but have an increasing challenge with each additional diagnosis/label. It can be overwhelming and highly emotional each time a new concern or issue is raised.
- 5 Work with families to prioritize and coordinate care. Often families have numerous professionals and appointments to manage and could really benefit from a professional acting in the role of a “case manager.”
- 6 Support more research involving children who are D/HH Plus. Unfortunately, children who are D/HH Plus are often excluded from research. Admittedly these children are far from a homogeneous group, but it can be a bitter pill for a family to swallow that their child and family often cannot contribute to research that could make an impact. Furthermore, by not including children who are D/HH Plus in research, little can be gleaned about how to guide interventions and strategies for their progress.

“Candace’s Corner” for Families and Professionals

“D/HH Plus is meant to be a positive term, not in any way negative or insensitive to the child who has medical issues along with hearing loss. In fact, I see it as an “A+” or “B+,” meaning the child carries additional positive qualities, but it is a gift that needs to be carefully unwrapped. And it may not appear to be a gift when you first receive it. Time helps you appreciate, understand, and unfold the possibilities. And the “Plus” most often means the child and family has added responsibilities and requires additional expertise.”

- 7 Recognize that adulthood is a challenging prospect. Families may feel deeper anxiety about the future of a child who is D/HH Plus. This anxiety can increase over time, especially as transition approaches and “traditional” choices and life experiences for children are often not appropriate. For example, the reality of a teen not being able to drive a car, or the need for a young adult to go into a Transition Program after high school rather than going on to college like peers, can be very emotional. While this thought may seem beyond the reach of the early intervention system, transition is a life-long process. The EHDI system should strive to enhance coping and adaptive skills of all families and children served within the system.



Photo courtesy of NCHAM

References

- Boyle, C. A., Boulet, S., Schieve, L. A., Cohen, R. A., Blumberg, S. J., Yeargin-Allsopp, M., Visser, S., & Kogan, M. D. (2011). Trends in the prevalence of developmental disabilities in U.S. children, 1997-2008. *Pediatrics*, *127*, 1034-1042.
- Chapman, D. A., Stampfe, C. C., Bodurtha, J. N., Dodson, M., Pandya, A., Lynch, K. B., & Kirby, R. S. (2011). Impact of co-occurring birth defects on the timing of newborn hearing screening and diagnosis. *American Journal of Audiology*, *132*(20), 132-139.
- First, L. R., Palfrey, J. S. (1994). The infant or young child with a developmental delay. *New England Journal of Medicine*, *330*(7), 478-483.
- Gallaudet Research Institute. (2011). Regional and National Summary Report of Data from the 2009-2010 Annual Survey of Deaf and Hard of Hearing Children and Youth. Retrieved December 27, 2013, from http://research.gallaudet.edu/Demographics/2010_National_Summary.pdf
- Joint Committee on Infant Hearing. (2007). Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs. *Pediatrics*, *120*, 898-921.
- Joint Committee on Infant Hearing. (2013). Supplement to the JCIH 2007 position statement: Principles and guidelines for early intervention after confirmation that a child is deaf or hard of hearing. *Pediatrics*, *131*(4), e1324 -e1349.
- Kenna, M. A., Rehm, H. L., Robson, C. D., Frangulov, A., McCallum, J., Yaeger, D., Krantz, I. D. (2007, July 15). Additional clinical manifestations in children with sensorineural hearing loss and biallelic GJB2 mutations: Who should be offered GJB2 testing? *Am J Med Genet A*, *143A*(14),1560-6.
- Kennedy, C. R., McCann, D. C., Campbell, M. J., Law, C. M., Mullee, M., Petrou, S., Watkin, P., Worsfold, S., Yuen, H. M., & Stevenson, J. (2006). Language ability after early detection of permanent childhood hearing impairment. *New England Journal of Medicine* *18*;354(20), 2131-41.
- Moeller, M. P. (2000). Early intervention and language development in children who are deaf and hard of hearing. *Pediatrics*, *106*(3), e43.
- Nikolopoulos, T. P., Lioumi, D., Stamatakis, S., & O'Donoghue, G. M. (2006). Evidence-based overview of ophthalmic disorders in deaf children: A literature update. *Otology and Neurotology*, *27*(S1-24), Discussion S0.
- Szymanski, C. A., Brice, P. J., Lam, K. H., & Hotto, S. A. (2012). Deaf children with autism spectrum disorders. *J Autism Dev Disord*, *42*(10), 2027-2037. doi: 10.1007/s10803-012-1452-9.
- U.S. Department of Education. (2004). IDEA Building the Legacy: IDEA. Retrieved October 14, 2014, from <http://idea.ed.gov/explore/view/pl/root,dynamic,TopicalBrief,23>
- Wiley, S., Choo, D., Meinzen-Derr, J., Hilbert, L., & Greinwald, J. (2006). GJB2 mutations and additional disabilities in a pediatric cochlear implant population. *Int J Pediatr Otorhinolaryngol*, *70*(3), 493-500.
- Yoshinaga-Itano, C., Sedey, A. L., Coulter, D. K., & Mehl, A. L. (1998). Language of early- and later-identified children with hearing loss. *Pediatrics*, *102*(5), 1161-71.