

Front Page Feature

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Imagine....

Imagine—if you will—the excitement that surrounds the arrival of a new baby. While the pregnancy went as planned, soon after birth you notice your baby is breathing quickly and having difficulty breastfeeding. Suddenly, the baby is whisked away to the neonatal intensive care unit for further evaluation. When the doctor returns, the parents/caregivers are told about a condition called a [trachea-esophageal fistula](#); this is a condition whereby the wind pipe and the food pipe are connected, and the baby must be transferred to a pediatric hospital for urgent surgery. Though everything well with the surgery and recovery, the care team mentions that an ophthalmologist also examined the baby. The parents/caregivers are now being told that their baby has a [coloboma](#) – a condition where an area of the eye did not fully develop, and depending on the location of the coloboma, vision can be affected. Shortly after this news, the parents/caregivers receive the results that their baby requires further diagnostic testing by an audiologist to determine potential issues with hearing. As new parents, they are now faced with the possibility that their child will be both deaf or hard of hearing and visually impaired.

Consider another scenario where the birth hospital’s newborn hearing screening identified that a child is deaf or hard of hearing (D/HH). Everything goes smoothly with early identification, diagnostic testing, enrollment in early intervention, and good supports for language stimulation. But, something just does not seem quite right. Despite doing everything that was recommended, the child is not making progress in communication. The child is frustrated and often has tantrums. It is hard to know what the child wants and needs. While the parents/caregivers focus on what they can do for their child, they notice something is different when comparing their child to other children in a playgroup. They begin to wonder if they are doing something wrong.

You have just read the abbreviated journeys of a parent whose child has been diagnosed with CHARGE syndrome, and a parent of a child who was identified as deaf or hard of hearing with an autism spectrum disorder. These journeys are relatively common—various sources* estimate that approximately 40% of children who are D/HH also have other special healthcare needs. This experience is captured in the term deaf/hard of hearing **Plus**. Candace Lindow-Davies, from Hands & Voices, defines this in the following:

“Deaf/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. “

– Candace Lindow-Davies, Minnesota Hands & Voices

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The [Joint Committee on Infant Hearing](#) (JCIH) recognized the needs of children who are D/HH and whom also have other special healthcare needs. Within the [supplement on Early Intervention Practice](#), JCIH included specific recommendations related to the needs of this group of children. Guidelines specify that, “All children who are D/HH with additional disabilities and their families have access to specialists who have the professional qualifications and specialized knowledge and skills to support and promote optimal developmental outcomes.”

This goal can be challenging to accomplish. One barrier is in the early recognition that a child who is D/HH often needs a broader team approach beyond their deafness. Another barrier is determining what additional interventions are needed. Finally, working as part of a broad-based team creates challenges with regard to the focus on the condition, the child, and the family. While it is easy to see things from our distinct professional, clinical perspective, we must challenge ourselves to identify opportunities for [care coordination](#) and integrated care as part of a broader team.

Most importantly, it is crucial to integrate support for families throughout this journey. Many rare conditions do not have robust family networks, and this contributes to feelings of frustration and isolation. Even when there is a strong network available (eg, [The CHARGE Syndrome Foundation](#)), the multiple competing demands for therapies, medical visits, day-to-day support, and behavioral interventions make networking difficult and overwhelming.

You may be asking yourself, “What can I do?” The best thing is to **listen with the intent to understand** and **offer compassion**. Remind parents that their child’s unique learning pattern is not their fault as feelings of guilt are common. Link families with up-to-date and accurate information and resources. Celebrate the successes—big or small. Remember to “focus on the donut, not the hole.” The journey of recognizing and “unwrapping the gift” that every child possesses is important for us to embrace.

*Gallaudet Research Institute [GRI]. (2011). Regional and National Summary Report of data from the 2009-10 Annual Survey of Deaf and Hard of Hearing Children and Youth (pp. 1-12). Washington, D.C.: GRI, Gallaudet University.

Family Partnerships

The [National Family Association for Deaf-Blind](#) (NFADB) believes that individuals who are deaf-blind are valued members of society and are entitled to the same opportunities and choices as other members of their community. The mission of NFADB is to empower the voices of families of individuals who are deaf-blind and to advocate for their unique needs. To address the uniqueness of deaf-blindness, the NFADB works side by side with families, friends and professionals to provide the services listed below.

- [Train and support](#) family members as advocates for their child and family
- [Connect](#) families with similar interests and needs
- [Collaborate](#) with other organizations to ensure the voices of children and families are heard at the state and national levels
- Advise professionals researching best practices for educating, training, and assisting individuals who are deaf-blind

As the largest national nonprofit organization serving families of individuals who are deaf-blind, the primary goals of NFADB are to connect people, information, and resources.

Clinical Corner

Outcomes and Benefits of Pediatric Cochlear Implantation in Children with Additional Disabilities: A Review and Report of Family Influences on Outcomes

Abstract: The number of children with hearing loss with additional disabilities receiving cochlear implantation has increased dramatically over the past decade. However, little is known about their auditory and speech and language development following implantation. The purpose of this [review](#) is to evaluate the effects of cochlear implantation on the most common genetic and developmental disorders in children with hearing loss. Benefits of cochlear implantation for children with autism spectrum disorder, developmental delay, CHARGE syndrome, cerebral palsy, learning disorders, Usher syndrome, Waardenburg syndrome, and attention deficit/hyperactivity disorder are reviewed. Our review indicates that children with hearing loss and additional disabilities benefit from cochlear implantation, especially when implanted early. Thus, early interventions seem as important for these children as for deaf children without additional disabilities. Comparisons of outcomes across these disabilities indicate that children with little to no cognitive impairment (eg, Waardenburg syndrome, attention deficit hyperactivity disorder) have better outcomes than those with greater deficits in intellectual functioning (eg, autism, CHARGE syndrome). In addition, parents of children with hearing loss and additional disabilities report higher levels of parenting stress and greater child behavior problems than those without comorbid diagnoses. However, these parents are as sensitive when interacting with their children as parents with typically developing children using cochlear implantation. Given these results, it is critical to evaluate these children's developmental milestones to provide early implantation and intervention, appropriately counsel families regarding realistic expectations for the implant, and facilitate family adaptation.

Source: Ivette Cejas I, Hoffman M, Quittner A. Outcomes and benefits of pediatric cochlear implantation in children with additional disabilities: a review and report of family influences on outcomes. *Pediatric Health, Medicine and Therapeutics*. 2015(6):45–63.

Quality Improvement (QI) Buzz



Early Hearing Detection and Vocabulary of Children with Hearing Loss

A cross-sectional sample of 448 children with bilateral hearing loss, aged between eight and 39 months (with a mean age of 25.3 months), and who lived in 12 different states, participated in the [National Early Childhood Assessment Project: Deaf/Hard of Hearing Children](#) (NECAP). The productive vocabulary of these children was assessed by the [MacArthur-Bates Communicative Development Inventories](#) which samples a vocabulary of about 650 words. The study identified predictors of vocabulary outcomes of the children's most recent assessment in the first three years of life.

Findings from this study show that the combination of 6 factors accounted for 41% of the variance in vocabulary outcomes. Vocabulary quotients were significantly higher for children who met the Early Hearing Detection and Intervention (EHDI) guidelines (screen by 1 month, identify by 3 months, and early intervention by 6 months), were younger, had no additional disabilities, had mild to moderate hearing loss, had parents who were deaf or hard of hearing, and had mothers with higher levels of education.

Several of the variables are not amenable to Early Hearing Detection and Intervention (EHDI) system development; however, since each significant variable was an independent predictor, there is great significance regarding a system where each of the 1-3-6 goals are met. Health, amplification and educational interventions are unable to alter or improve the degree of hearing loss, the mothers' level of education, the hearing status of the parents, or whether the children have an additional disability. Currently, about 65% of the children identified through EHDI systems meet the 1-3-6 guidelines, and states have already demonstrated the ability to improve their follow-through statistics and increase the number of children who meet this benchmark.

Meeting each of the 1-3-6 guidelines is beneficial for all children, regardless of degree of hearing loss, mothers' level of education, hearing status of the parents, the presence or absence of an additional disability, and regardless of the home language of the family. The most important change for vocabulary outcomes of children who are deaf or hard of hearing is to increase the number of children who are captured by screening and follow the 1-3-6 guidelines. When children met only some of the benchmarks (eg, 1-3 but not 6, or 1-6 but not 3), their vocabulary outcomes did not differ from the children who missed both 3 & 6. All three groups had significantly lower vocabulary scores than the children who met each of the 1-3-6 benchmarks.

Vocabulary learning may be enhanced with system improvements that increase the number of children who meet the current early identification and intervention guidelines. Improvement of systems requires coordination and commitment of all professionals who interact with parents, beginning at the level of the infant hearing screening. Further support by physicians that infants return for follow-through for outpatient screening, diagnostic audiological assessment, and enrollment into appropriate early intervention services ensure completion of the process. In addition, intervention efforts need to focus on preventing widening delays with chronological age, assisting mothers with lower levels of education, and incorporating adults who are deaf/hard-of-hearing in the intervention process.

The message is simple and urgent. Professionals in EHDI need to do everything they can do to ensure that infants are not lost after the initial screen and the identification of hearing loss and enrollment into intervention are equally important. Additionally, early intervention systems need to develop improved strategies for supporting parents/families in helping their children learn and use new vocabulary words, especially after the age of two years.

Source: Yoshinaga-Itano C, Sedey AL, Wiggin M, et al. Early hearing detection and vocabulary of children with hearing loss. *Pediatrics*. 2017;140(2).

Medical Home Resources

Tools to Support Children and Youth with Special Health Care Needs and Their Families: Shared Plan of Care and Care Notebooks

For children and youth with special health care needs (CYSHCN) requiring multiple services and supports, the use of a shared plan of care facilitates implementation of key functions of the medical home model, including, but not limited to, comprehensive care coordination, communication, and patient- and family-centered care.

For children who are identified as Deaf Plus, it is important that care is coordinated. The [National Center for Medical Home Implementation](#), in collaboration with the National Academy for State Health Policy, created a [fact sheet](#) which outlines key components of a shared plan of care. The fact sheet discusses the role of families and state agencies to create, maintain, and support shared plans of care.

Another tool to consider is the [care notebook](#) which provides resources to assist families and caregivers in organizing and maintaining records of their child's services to ensure that care is comprehensive, coordinated, and family-centered. Examples of resources included in a care notebook follow: appointment logs, medical bill tracking forms, and a list of questions to ask clinicians.

And More....

Everybody Communicates: Toolkit for Accessing Communication Assessments, Funding, and Accommodations

Everybody communicates—whether using language, behavior, gestures, facial expressions, sounds, or other means. Created by the Autistic Self Advocacy Network, this [toolkit](#) is designed to help enhance effective communication for all individuals with communication-related developmental disabilities, including deafness or hearing loss. Findings have shown that children diagnosed as deaf or hard of hearing, and who receive early intervention, are more likely to have a significantly larger vocabulary than those diagnosed and receive intervention later.

Disclaimer: The information and opinions contained in this newsletter are compiled from various sources and represent a multitude of opinions and methodologies. They do not necessarily represent policy or recommendations from the American Academy of Pediatrics. For questions regarding content, or recommendations for future content, contact [Sandi Ring](#), Program Manager, Early Hearing Detection and Intervention, at the American Academy of Pediatrics.

The AAP EHDI program implementation staff send this e-newsletter to the Academy's EHDI Chapter Champions, other interested AAP members, state EHDI coordinators, and other stakeholders. For additional information on hearing screening, or to access tools and resources relevant to early hearing detection and intervention, click [here](#). If you would like to unsubscribe to this newsletter, contact Sandi Ring at sring@aap.org or 847/434-4738.