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Children Who Are Deaf or Hard of Hearing With Additional Learning Needs

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Disclosure: Susan Wiley has no financial or nonfinancial relationships related to the content of this article.

Abstract

Additional learning problems are common in children who are deaf or hard of hearing (HoH). This higher rate of additional disabilities beyond the general population may be related to the overlap of causes for hearing loss, such as prematurity, that also can impact child development. Delayed identification of atypical learning strategies impacts appropriate interventions for all of a child's needs and, furthermore, may impact communication strategies, thus negatively impacting language outcomes. In this article, I outline some red flags for additional disabilities in children who are deaf or HoH. I will present an algorithm for thinking systematically through causes of slow language progress in children who are deaf or HoH to guide professionals who work with children. I will stress strategies to identify expanded team members and collaborate towards improved outcomes for children with different learning needs.

With the successful implementation of universal newborn hearing screening programs across the United States, we are seeing the benefit of early identification (Holster et al., 2009) and intervention more consistently across children with permanent hearing loss (PHL). More children are approaching language skills in the average range as compared to hearing peers (Yoshinaga-Itano, Baca, & Sedey, 2010; Vohr et al., 2012). Additionally, improvements in technology (i.e., hearing aids, cochlear implants) have given many children improved access to sound, prompting more families to pursue auditory/oral outcomes for their children. Early identification allows families more time to pursue communication options, as there is less urgency in making up for "lost time" that has been historically seen in late-identified, significantly language-delayed children.

However, with early identification comes a responsibility to understand the new trajectory of language development that is likely to occur for children benefiting from these improvements in identification and technology. We need to recognize when a child is not making adequate language progress and intervene appropriately. Because nearly 40% of children with PHL have an additional disability (Gallaudet Research Institute [GRI], 2008), it is imperative to recognize that not all children will respond to traditional therapeutic strategies for language development (see Table 1). When we assume a child is not making progress due to his or her hearing loss, we miss key opportunities to intervene differently at early ages in order to improve long-term communication skills. These children then miss the benefit from early

identification that their typically developing peers are receiving. The next advancement in the field of early intervention and educational interventions for children who are deaf or hard of hearing (HoH) should be striving to minimize this disparity in language and communication outcomes for this large group of children.

Table 1: Rates of Disabilities in children with hearing loss and the general population

Type of Disability	Hearing Loss GRI Data	General Population (Boyle et al., 2011)
No Additional Disability	60%	
Cognitive (Intellectual Disability)	9.8%	2.5%
Cerebral Palsy		0.3%
Blindness	3.9%	0.03%
ADHD	6.6%	5–10%
Specific Learning Disability	10.7%	5–10%
Other	12%	
Autism	1–4%*	1%

Individual learning styles can vary markedly among children. This is also the case for children who are deaf or HoH (Conway, Pisoni, Anaya, Karpicke, & Henning, 2011; Kritzer, 2009; Mellon, Ouellette, Greer, & Gates-Ulanet, 2009; Meronen & Ahonen, 2008; Zupan & Sussman, 2009). Although it is easy to make certain assumptions about children related to hearing loss (such as these children being visual learners), these assumptions may not hold for every child. Some children learn more through doing, using hands-on activities to understand concepts. Others learn from watching how others approach a problem. Some children need to understand the big picture (gestalt-style learning) before they are comfortable taking the steps towards putting the pieces together. Other students prefer a step-wise building of concepts towards the whole. Some children struggle with sequencing, attention, or working memory. All of these challenges in learning can affect language learning and subsequently we may see challenging behaviors, slow rates of progress, and frustration among children, families, educators, and therapists. Understanding the learning style of a child and his/her motivations is helpful in choosing therapy and education strategies for an individual child

Because there is a high rate of children who are deaf or HoH who learn differently, it is helpful to understand what children may be at risk for these challenges, monitor progress closely in order to intervene differently at young ages even in children without risk factors for learning challenges, and be creative in implementing interventions. Even if we are not certain of a specific diagnosis, we can intervene differently when we suspect something more than hearing may be interfering with development. It can be helpful to expand the intervention team to include professionals who may not have expertise in the field of deaf education/intervention, but rather have expertise in other areas in specific disabilities.

Who Is at Risk for Additional Disabilities?

Within the field of developmental disabilities, there are recognized factors that put children at higher risk for developmental and academic underachievement (Carey, Crocker, Elias, Feldman, & Coleman, 2009). These risk factors can be categorized in a number of ways, such as prenatal, peri-natal (around birth), and postnatal factors. Discussing the presence of risk factors with families can help alert them that we will be watching their child carefully to ensure we address all needs early in order to allow a child to meet their capabilities.

Prenatal and Perinatal Risk Factors

When we consider prenatal risk factors, there can be environmental exposures to the developing fetus and genetic factors that affect development. By taking a good pregnancy and birth history, we may uncover not only factors related to risks for hearing loss, but also risks for other problems which then allow us to have a higher suspicion that a child may experience atypical learning styles. This identification of risk factors put us on alert to observe development more carefully and address these issues proactively at earlier ages.

Prenatal risk factors include

- toxic exposures such as alcohol or lead (Chandramouli, Steer, Ellis, & Emond, 2009; Sen & Swaminathan 2007);
- uterine factors such as poor development of placenta, twin gestation (Refuerzo et al., 2010), pregnancy induced hypertension, gestational diabetes (Kowalczyk, Ircha, Zawodniak-Szałapska, Cypryk, & Wilczyński, 2002), and in-utero infections, such as cytomegalovirus,
- genetic risks such as syndromes or a family history of learning difficulties; or
- atypical embryologic development such as spina bifida or brain anomalies.

Peri-natal risk factors include

- birth asphyxia,
- prematurity,
- peri-natal infections (Group B Streptococcal infection, etc), and
- ABO incompatibility

Postnatal Risk Factors

A health history also can identify factors beyond the pregnancy and birth period that place children at risk for learning differences. Although audiologists and speech-language pathologists are not expected to understand all of a child's medical conditions, having an understanding of how medical issues may interfere with exposure to opportunities to learn, as well as their impact on how a child learns, can be helpful in the ongoing supports families may need for their child. By identifying areas that may affect learning, we can target interventions to try to modify the impact of these factors on child development.

Postnatal risk factors include

- environmental exposures such as tobacco smoke or lead (Chandramouli et al., 2009);
- failure to thrive or poor nutrition;
- infectious diseases such as meningitis or encephalitis;
- complex medical problems such as significant vision impairment, congenital heart disorders (Massaro, El-Dib, Glass, & Aly, 2008), and seizure disorders;
- trauma such as traumatic brain injury;
- emotional and/or physical abuse; and
- inappropriate environmental experiences

What About Normal Developmental Variation?

Sometimes we may think there is a developmental concern when the biggest problem is a poor fit between the environment and the child (Kiff, Lengua, & Zalewski, 2011). Children have temperamental variations that can impact how they respond to the environment. Some children are slow to warm, some children do better with routine and regularity, and others are

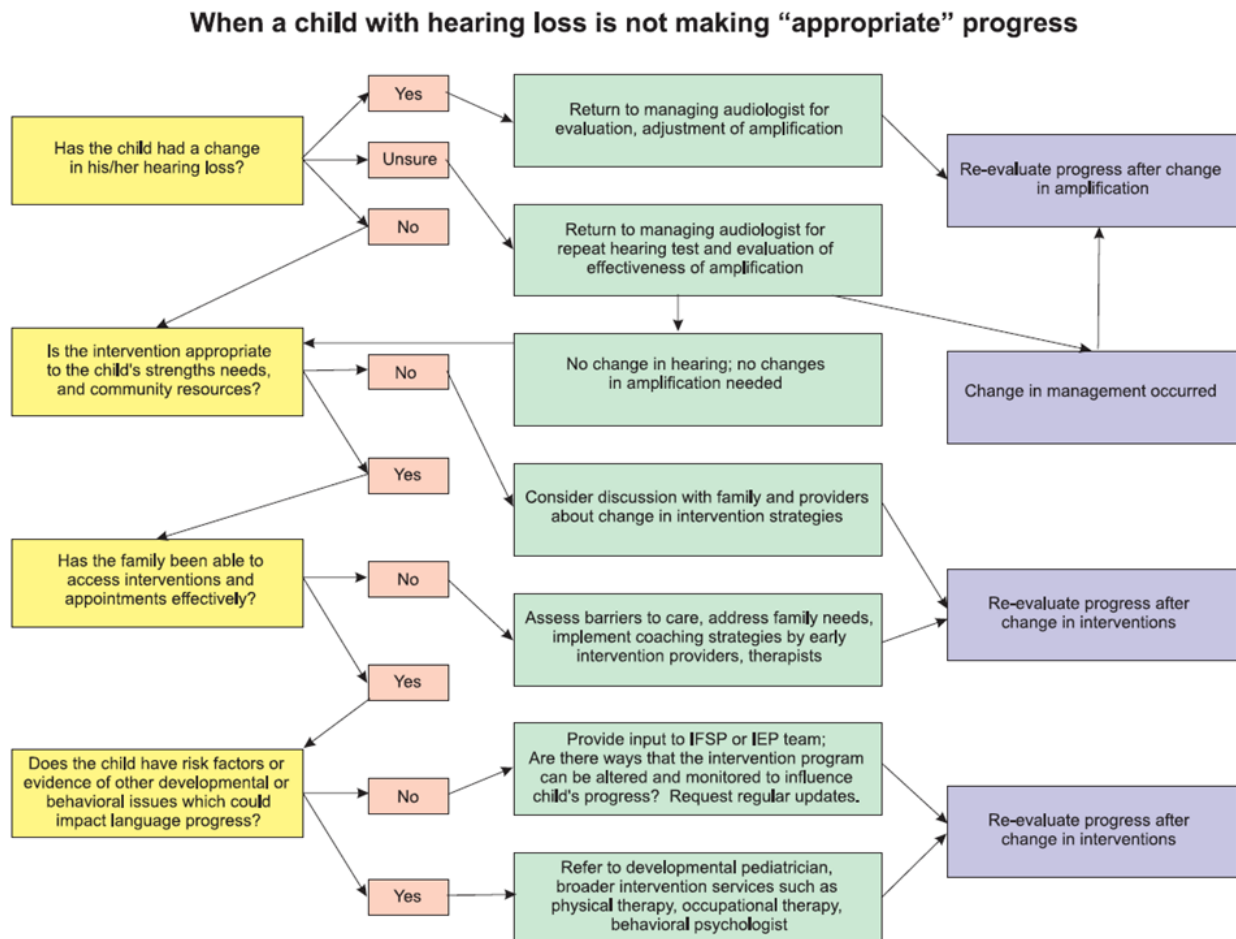
more able to perform in settings with low predictability and enjoy the changes that they may face. When we understand a child's temperament and how best to approach their needs, we tend to see children flourish and show their capabilities.

It is important to consider the psychosocial needs of children and families as well. Children grow up in the context of families. We often learn how to become parents from our experiences with our own parents. Families can face a variety of stressors and adjusting to the needs of a child who is deaf or HoH can add to the already existing needs of the family system (Meinzen-Derr, Lim, Choo, Buyniski, & Wiley, 2008). We also may face families from cultures different than our own. Cultural differences may affect how we understand and impact developmental differences. The National Center for Cultural Competence (<http://nccc.georgetown.edu/>) has a number of resources that can help clinicians improve their cultural competence. The most important consideration in working with all families is to make no assumptions and develop ways to ask about families' perspectives regarding their children's needs and strengths, as well as what they believe is causing the pattern we are seeing. When families feel supported and have trusting relationships with professionals, we can team together most effectively for improving child outcomes.

How Do We Monitor the Progress (or Notice the Signs) of an Atypical Learner?

Although risk factors can help us identify children with different learning needs, not all children who learn differently necessarily have one of these identifiable risk factors. This is important to remember, particularly when a child has a known cause of hearing loss that is not expected to carry risks for developmental problems. For example, children with connexin mutations (*GJB2 mutation*) initially were thought to be excellent candidates for cochlear implants as the genetic cause of hearing loss was isolated to the hearing (Dahl et al., 2003; Fukushima et al., 2002). However, later studies emerged that suggested that children with connexin mutations can have other findings that could impact developmental progress (Kenna et al., 2007; Wiley, Choo, Meinzen-Derr, Hilbert, & Greinwald, 2006). Having one identified genetic condition does not protect a child from the remaining genes and influences on development. Therefore, clinicians need to be observant of atypical learning in any child. Figure 1 shows an algorithm developed by Moeller and Wiley to guide professionals in understanding a child's individual and therapeutic characteristics, proceeding in a step-wise fashion towards determining where interventions may need to be altered or adapted in children who are deaf or HoH.

Figure 1: Algorithm When a Child Is Not Making Anticipated Progress



Motor development

In general, children who are deaf or HoH should have typical motor development (Lieberman, Volding, & Winnick, 2005), or have a good reason why they are not following typical motor patterns. The plausible reasons for delayed motor development in children who are deaf or HoH mainly encompass vestibular problems and vision concerns. Children also can have problems within the brain that impact motor development, such as those that can be found in children with cerebral palsy. However, clinicians would not expect motor delays based solely on hearing loss.

Red flags for motor development (Carey et al., 2009; Gerber, Wilkes, & Erdie-Lalena, 2010) include

- poor head and trunk control in early infancy,
- not walking by 15 months of age,
- frequent falls,
- developing a hand preference before the age of 1 year,
- not developing a hand preference by 2.5 years of age,
- difficulties crossing the mid-line by 4 years of age,
- immature grasp patterns, or

- poor sign production (if signing).

Vision Impairment

Children who are deaf or HoH are at higher risk for vision problems than children without hearing loss (Nikolopoulos, Lioumi, Stamataki, & O'Donoghue, 2006; Sharma, Ruschetta, & Chi, 2009). Certain conditions that cause hearing loss also impact the eye and subsequent vision. For example, some genetic syndromes are associated with a specific type of vision problem, such as coloboma in CHARGE syndrome, (Russell-Eggit, Blake, Taylor, & Wyse, 1990) and prematurity (Quinn et al., 2011) can put children at risk for retinopathy of prematurity with subsequent vision problems. Additionally, some infections such as cytomegalovirus (CMV) can cause retinal changes that affect vision (Anderson, Amos, Boppana, & Pass, 1996). In patients with Usher Syndrome, audiologists typically identify the hearing loss well before the vision impairment occurs (Friedman, Schultz, Ahmed, Tsilou, & Brewer, 2011). Some children have cortical vision impairment (Ospina, 2009), this means the eye itself is normal, but the brain does not process visual information typically. Children with brain-based problems are at higher risk for this condition and it is important to monitor for this possibility, particularly in children with conditions such as cerebral palsy or symptomatic CMV.

It is particularly important for audiologists to ensure that children receive good vision care while being on the lookout for concerns that indicate a child may have problems with vision (Joint Committee on Infant Hearing, 2007). Having an ophthalmologist knowledgeable in children and in vision problems that can co-occur with hearing loss can help identify concerns early. Additionally, engaging a vision specialist (educator) to determine a child's functional vision (rather than only using a report from an ophthalmologist to understand a child's vision) is critical in appropriate intervention planning. A good vision specialist will help determine appropriate lighting, contrasting colors, or pacing of presenting materials and what angle to optimize a child's ability to use their vision maximally for learning.

Conditions Associated with Vision Impairment include

- syndromes (Usher Syndrome, CHARGE, Waardenburg, etc.),
- infections causing retinopathy (CMV),
- retinopathy of prematurity, and
- brain-based abnormalities (septo-optic dysplasia, cerebral palsy, etc.).

Red Flags for Vision Impairment include

- poor visual regard,
- poor tracking (up or down),
- wiggling eyes,
- wandering eyes,
- head tilt,
- troubles with balance/clumsiness (running into walls),
- not seeing people from the side (getting hit by a ball coming from the side),
- night-vision difficulties, and
- poor acclimation going from dark to lighted environments (or vice versa).

Learning Disabilities

Although hearing loss typically has precluded the consideration of a specific learning disability by the definition in the Individuals with Disabilities Education Act (IDEA, 2004), it has become more apparent that children can, in fact, have both a hearing loss and a specific learning disability. It is challenging for professions to separate the impact of hearing and the impact of learning patterns on educational achievement. Professionals with a good

understanding of learning differences and how children who are deaf or HoH learn can help determine if a learning disability is also affecting a child's performance. The current educational system that includes with "primary" diagnosis labels to identify interventions can be a disservice to children with hearing loss and other co-existing problems if the primary label limits accessing appropriate services for all of a child's learning needs. It is beyond the scope of this article to provide sufficient background and identifying factors for learning differences.

Behavioral Difficulties

Behavioral challenges can occur in children who are deaf or HoH. It is critical to start with the concept of behavior as communication in this group of children. However, sometimes behaviors arise from other concerns, such as poor environment-child fit, a lack of developed coping strategies in the child, difficulties with attention span, activity level, sensory processing (Wiley & Moeller, 2007), anxiety, emotional lability, and aggressive behaviors. When we have children who are exhibiting behaviors that we are not expecting, it is important to obtain good information about the behavior itself (describe the behavior in very specific terms), understand the context in which the behavior is occurring (when does it occur), and try to understand the reasons (consequences of the behavior), both intended and unintended, that may keep a behavior occurring. Most behaviors have possible functions of attention, escape (or avoiding nonpreferred activities), tangible (getting something out of the behavior), or automatic (something internal that feels good). Understanding the purpose of a behavior helps us strategize to improve the behavior. The most challenging behavior is one that is coming from an internal motivation (such as stimulatory behaviors, which feel good). It may be challenging to effectively intervene, however, typically, we try to identify a competing behavior to replace the problematic behavior. Finding something that is equally motivating improves the success of replacing a nonpreferred behavior.

Identifying and implementing an effective behavioral intervention plan relies on good observations of behaviors and the environment in which they occur. Sometimes we need to pursue medication management in addition to behavioral interventions. However, without a strong behavioral program, the likelihood of sustained improvements for behavior when medications stop are unlikely. Medication alone is typically not the sole solution for children with challenging behaviors.

Communication Disorders

It is particularly challenging to determine if there is a language impairment that spans beyond the anticipated impact of hearing loss on language development. It has become evident that specific communication disorders such as apraxia of speech and autism spectrum disorders can occur in children who are deaf or HoH. Additionally, some children appear to have significant language processing difficulties that do not seem to occur merely because of their hearing loss. For these children, receptive language skills are well behind what is anticipated based on their learning or cognitive potential. This pattern is easier to recognize in children who have milder degrees of hearing loss and have early identification and effective interventions with supportive educational and home environments.

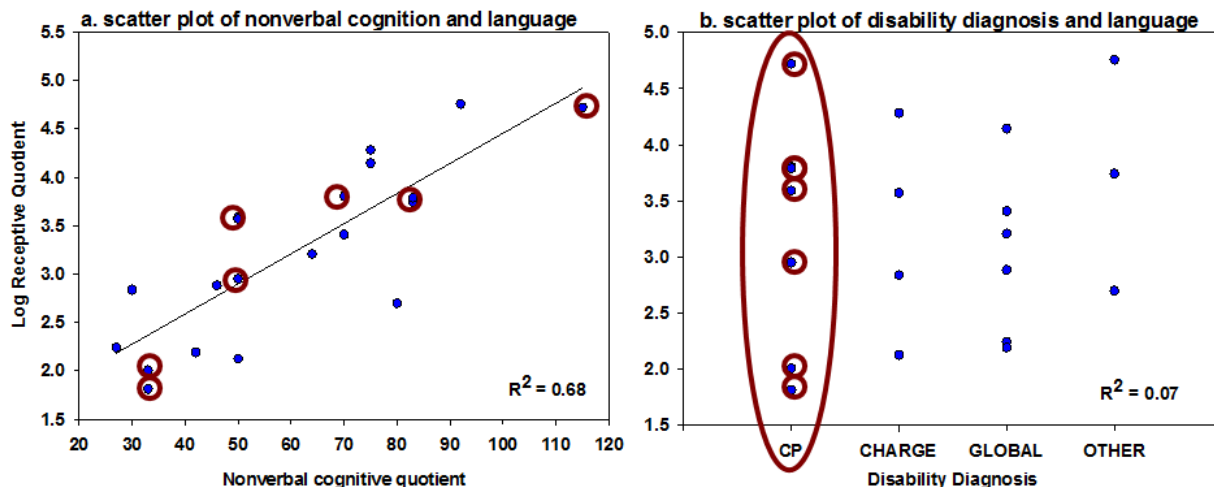
Of course, it is important to ensure we account for the impact of hearing loss on specified communication red flags. For example, some articulation problems can reflect the integrity of what a child is hearing with amplification. Also, if a child has severe-to-profound hearing loss, she or he may not respond to her or his name when called, which is a red flag for autism. However the child with hearing loss should respond to alternative ways to gain his or her attention. This is the reason that an integrated approach from professionals knowledgeable in the development of children who are deaf or HoH and professionals knowledgeable in disabilities can improve our likelihood of accuracy in determining if a problem indeed exists.

Intellectual Disability

Approximately 10% of children who are deaf or HoH have an intellectual disability (GRI, 2008). Cognitive abilities can impact the rate of a child's progress across all developmental

domains. Although intellectual capacity is not the only factor that impacts a child's language progress, it is strongly correlated with language outcomes (Meinzen-Derr, Wiley, Grether, & Choo, 2010). A specific developmental disability label such as cerebral palsy or a specific cause of an additional disability such as CHARGE syndrome is not particularly helpful in understanding a child's potential development due to the great heterogeneity within those disability categories (see Figure 2).

Figure 2: Comparison of Language Abilities By Cognitive Capability Vs. Disability Category



The graph on the left in Figure 2 plots language abilities by non-verbal problem-solving skills. This relationship shows that with increasing cognitive abilities, language abilities also increase. However, the graph on the right breaks out the data by disability label. This graph shows wide variability in language skills within a specific category. Furthermore, if you take the children with cerebral palsy (in the red highlighted circles) and clarify which points they are on the graph on the left, you can see that they follow a pattern based on their cognitive capabilities, rather than based on having a diagnosis of cerebral palsy. Having a valid evaluation of a child's cognitive capabilities (typically measured by nonverbal cognitive measures) can provide a better framework for understanding if a child is making the progress we would anticipate over time.

We Think There is a Problem

Talking to Families

When we suspect other learning challenges, it is important to explain to families why we suspect this and what our next steps should be. Describing what we usually tend to see in children with a similar degree of hearing loss can help guide the conversation. It is always difficult to bring up concerns, particularly when families are not aware their child is developing differently.

Talking to Professionals

When making referrals, it is important for clinicians to explain why they think the child's difficulties are not just due to a hearing loss, but may indicate a broader problem. When people outside the field of deafness are brought to the table, they may need to be educated about how children who are deaf or HoH typically learn and behave so that these individuals do not fall back on the same assumption that these problems are all related to hearing loss. By describing what you believe the hearing interventions should have accomplished and have and

have not done for a child, you can emphasize why the child needs further evaluation and a change in intervention. Discussing strategies that have already been implemented also help guide an understanding of what does and doesn't work.

When children are in early intervention programs or educational programs, audiologists can use these settings to broaden access to expertise in areas beyond hearing loss. If an audiologist thinks the child presents with issues related to motor development, he or she can seek out physical and/or occupational therapy to provide a structured evaluation. When behavioral challenges arise, psychologists can be helpful in identifying potential issues, as well as performing a functional behavioral analysis to better understand why a behavior is happening (the function of the behavior) and what may be keeping the behavior occurring (the consequences of the behavior) and identifying different strategies to improve behaviors (things we can change about our approach before the behavior occurs, what do to differently when the behavior occurs, and what we need to teach the child in order for them to respond more appropriately).

Sometimes, we need to expand our intervention team to include learning disability specialists. Although professionals in the field of deaf education can receive training on educational strategies, reaching out to experts in broader fields (such as specific learning disabilities, autism, technology experts, etc.), may allow them to better identify other educational strategies that can be adapted or used for children who are deaf or HoH with learning needs. Making a referral to a developmental-behavioral pediatrician or neurodevelopmental disabilities pediatrician can also help understand a child's learning profile.

Effective Teaming Towards Effective Intervention

Children who learn differently benefit from a strong team approach. Having a number of professionals perform observations in natural environments can serve as a foundation for thinking through how a child learns and responds to instruction. This foundation allows the team to develop creative ideas to test their effectiveness. Frequent monitoring of a child's progress allows the team to recognize when a child is having success with the intervention plan and when the plan needs to be altered.

Considerations for Family-to-Family Support

Identifying ways to promote family-to-family support is very important for all children, but poses particular challenges for the child who is deaf or HoH with an additional disability. Because some dual diagnoses are rather uncommon (autism and hearing loss or deaf-blindness), it may be unlikely for families to know another family with similar needs. The internet seems to be a common location for families to find a network.

Why it Matters

When we identify other issues, determine effective team partners in order to expand intervention strategies, and implement them effectively, we see the rewards of our efforts. When we see children who learn differently make progress and develop new skills, even if they are not at the same rate or level as other children of similar ages, it is very rewarding. Our goals and timelines to goals may differ, but it can be that much more exciting when we share with family's journeys towards each step forward!

Susan Wiley is a developmental pediatrician with a focused clinical and research interest in children who are deaf or hard of hearing with coexisting disabilities. She has evaluated more than 700 children who are deaf or hard of hearing. Her research interests have focused on understanding the language and functional progress of children who are deaf or hard of hearing with additional disabilities.

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