Chapter 8

Medical Home & EHDI: The Importance of Appropriate & Timely Screening, Diagnosis, Management, & Follow-Up

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In the early 1990s, a novel idea began sweeping the nation. The advent of a new generation of screening technologies made it possible to assess newborn babies' hearing and identify those who are deaf or hard of hearing (D/HH) in the first few months of life. Professionals familiar with early hearing screening technologies and the potential for markedly improved outcomes began advocating for universal newborn hearing screening (UNHS). Initial results from states that adopted UNHS were encouraging, and the idea gained increasing acceptance. In little more than a decade, dramatic state-by-state adoption has made UNHS the rule rather than the exception.

With the advent of UNHS, the frequency of hearing loss in newborns was discovered to be as high as 1 to 2 per 1,000 newborns—a number far greater than other conditions, such as phenylketonuria (PKU) or hypothyroidism, for which infants were already being screened. Instead of waiting for a delayed diagnosis at the typical age of 18-24 months or later, infants who are identified as D/HH started to benefit from options, such as early amplification and early intervention services, with subsequent language outcomes at or near the level of their hearing peers. Infants who are deaf should have early access to communication and language (oral or signed) through parent education and targeted intervention to ensure early communication skills and developmental progression. As a result, language and communication development could progress while the

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brain was growing and forming the complex synapses necessary for cognitive development. In many ways, UNHS opened up a world of opportunities to children who are D/HH.

Of course, along with early hearing screening comes a new responsibility. Infants who are identified as D/HH require timely retesting and follow-up, appropriate referral for diagnosis by professionals experienced in working with newborns, information about available communication and intervention (such as amplification of residual hearing, if desired by the family), and coordination with early intervention services to provide opportunities for optimal development. Who then should coordinate the myriad services needed by the infant who is D/HH? In the medical model of blood (spot) test screening for newborn metabolic disorders, the primary care physician has been the logical team leader for contacting families, assuring access to specialty care, providing follow-up, and reassessing compliance.

The Medical Home

The American Academy of Pediatrics (AAP) has been a leader in outlining how every child would benefit from having a well-defined "medical home." The concept, of course, is not as simple as having the name of a physician or the street address of a brick-and-mortar building. Instead, the term "medical home" is defined as an active process—a philosophy of care that emphasizes the role of the primary care physician in the care of ***all*** children, including children who have special needs. This physician serves as a focal point not only for the typical primary medical care of the child but also for the support of parents and family, the coordination of specialty medical care, the provision of referrals for various services, the assurance of timely follow-up and the medical interface for educational interventions.

In the rapidly evolving arena of newborn hearing screening and the availability of various interventions and communication modality options, however, few pediatricians and other pediatric clinicians can be expected to have a complete and up-to-date level of knowledge within this specialized area of care. Even highly motivated physicians express some trepidation at having the necessary knowledge and skill to provide expertise to families concerning their child's special needs related to hearing. Even at a frequency of 1 to 2 affected children in every 1,000 newborns, a primary care clinician might only care for a baby who is D/HH once or twice in a decade. Fortunately, there are numerous educational resources available to clinicians to assist them in learning more about caring for babies who are D/HH.

The Case for the Primary Care Physician

Infants who do not pass a newborn screening test and are subsequently diagnosed as being D/HH are referred for extensive pediatric audiology care and ideally are enrolled in early intervention to ensure opportunities to monitor and enhance developmental progress. Clearly, the initial stages of care include continuing evaluation with a number of sophisticated audiology testing protocols. Finding audiologists with experience working with infants becomes a paramount consideration. Evaluation by an otologist or otolaryngologist—physicians who specialize in diseases of the ear—is required to further assess the possible causes of hearing loss and to engage the family in conversation around the many possibilities available to them. If families are interested, other medical options, such as a genetics consultation and ophthalmological examination, may be important and should be considered.

Given that 90% of children who are D/HH have some residual hearing, amplification has increasingly become a cornerstone of treatment. Special expertise is required when fitting amplification for young children, as they cannot answer

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the examiner's questions or report about their own comfort or experience. Finally, early interventionists with skills in speech pathology, language acquisition, various communication modalities, and deaf education play an active role in assuring appropriate developmental outcomes.

It is important for the primary care physician to be the center of the medical home for several reasons. First, and perhaps foremost, the primary care physician is necessarily an active participant in the life of the family during a baby's first year. From initial evaluation in the hospital nursery through multiple visits assessing growth; monitoring developmental skills; and providing preventive care, such as immunizations, the primary care physician is an integral member of the healthcare team. This relationship provides the greatest opportunity for family support and education and a natural environment for follow-up and reassessment.

Perhaps equally important, frequent interactions emphasizing the importance of adequate communication and free exchange of documentation with all the members of the child's healthcare team provide many opportunities to coordinate the necessary sequence of care. It is critical that the primary care physician realize the importance of timely assessment and the earliest possible care plan. The AAP has been a leader in providing "just in time" information for those physicians who find themselves caring for an infant who is D/HH.

Photo courtesy of NCHAM

Furthermore, the primary care physician is positioned to reinforce the important messages delivered by the audiologist, otologist/otolaryngologist, and early interventionist. Such support may range from encouraging families to complete the recommended evaluation, emphasizing the importance of timing and prompt intervention, discussing communication options, assuring the completion of any recommended testing or imaging procedures, and reinforcing compliance with recommendations (e.g., daily use of hearing technology). The primary care physician must also be recruited to screen for risk factors for late onset or progressive hearing loss and encourage families to regularly monitor their child's hearing. Of course, the primary care physician must aggressively treat otitis media and retained middle ear fluid—conditions that might further compromise residual hearing.

Because primary care physicians have frequent interactions with infants and their families, especially during the first years of life, it is essential that they become familiar with the early hearing detection and intervention (EHDI) recommendations in order to be an active participant in providing care, guidance, and support to families.

Lastly, the need for coordination of care should not be underestimated. The primary care physician's role is critical not only in the timely completion of referrals but also in advocating for necessary services when the need for those services might be questioned by insurance providers. Furthermore, the collation of reports and opinions from multiple professionals under the single umbrella of

a medical home allows for a cross-check to assure that the efforts of various service providers are coordinated to achieve the best possible outcome for the child.

The Necessary Attributes

If providing the active participation known as medical home falls to the primary care physician, the attributes listed in *Table 1* should be brought by the physician to the care team, including the family, to assure success. As outlined by the AAP, this approach to primary care should include a number of features. Ultimately, the physician must have adequate knowledge to facilitate rather than obstruct the timely testing and appropriate care and resource provisions for these children and their families.

In this rapidly evolving field of knowledge, the physician will likely look to learn new information and seek out educational resources. The physician must work collegially, not only with physician peers and specialists, but also with audiologists, speech pathologists, early interventionists, and education professionals. Finally, the physician can participate as an advocate for the infant and family in assuring access to timely evaluation and prompt initiation of intervention, if desired, in the first few months of life.

Supporting the Medical Home

The medical home need not stand at the peak of a hierarchy of medical and educational services. Rather, those services should support and be supported by the primary care physician. Increasingly, models of "just-in-time" learning are being made available to physicians, such that the doctor might receive the latest up-to-date information whenever a child who is D/HH is newly identified in his or her practice. Audiologists and specialty physicians should also reach out to groups of primary care physicians to re-educate these colleagues about newer developments in the field of newborn hearing screening. Physician training programs will continue to need curriculum updates, such that the next generation of physicians may be adequately trained. Regional coordinators of state EHDI programs are in a unique position to personally assist the primary care physician in better understanding the newest developments in the field and the specific mechanisms of accessing services in their home state. Finally, the various professionals involved in the medical care and comprehensive early intervention of infants who are D/HH must provide timely information and feedback to the primary care physician about the child's ongoing care.

Through such efforts, each of the involved professionals will be supporting the true spirit of partnership and teamwork—thereby assuring the best possible outcome for children who are D/HH. It is a new era when being D/HH need not undermine a child's long-term development. It is a new era when a condition that previously might have been viewed as a disability need not interfere with a child's ability to reach his or her true potential. It ***is*** a new era, and the role of the medical home is increasingly recognized in supporting and advocating for the child who is D/HH.

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Table 1
Necessary Attributes

The care should be ***accessible***.	The care should be ***continuous***.	The care should be ***family centered***.
Only through the provision of timely appointments and prompt answers to the questions of family members and medical colleagues will successful identification and care plan development become a reality.	Though no physician will be available 24 hours a day, 7 days a week, a single physician should nevertheless be the point person for the family's ongoing needs.	When a child is identified as the "patient" with a special need, the educational needs of the family are critical, and the concerns and opinions of the parents must be honored. Through a model of parental education and shared decision making, the family becomes an integral part of increasing the success of the team.
The care should be ***coordinated*.**	**The care should be ***compassionate***.**	**The care must be ***culturally competent***.**
With participants in the fields of primary care, audiology, otology, early intervention, education, and sometimes subspecialties, such as developmental pediatrics, genetics, and audiology, coordination becomes paramount.	Every family envisions that their newborn child will be perfect in every way. When a child is identified as D/HH, families suddenly find themselves traveling along a rapidly moving roller coaster of evaluation and intervention. The physician must be engaged in supporting the family in their concurrent mourning and acceptance of this unexpected challenge to the health and well-being of their newest family member.	Increasingly, the importance of bridging cultural divides is recognized as integral to providing effective medical care. Addressing medical literacy and the family's cultural norms will be critical to assuring the most favorable outcomes for the child.