Chapter 11

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 with congenital hearing loss 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. Plus, instead of waiting for a delayed diagnosis at the typical age of 18-24 months or later, infants with hearing loss started to benefit from early amplification and intervention, with subsequent language outcomes at or near the level of their hearing peers. Even infants with profound hearing loss could have early access to language (oral or signed) through parental education and aggressive intervention. As a result, language development could progress while the brain was growing and forming the complex synapses necessary for intellectual development. In many ways, UNHS opened up a world of opportunities to children who are deaf or hard of hearing.

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Of course, along with early screening comes a new responsibility. Infants with hearing loss require timely retesting and follow-up, appropriate referral for diagnosis by professionals experienced in working with newborns, prompt intervention (typically including amplification of residual hearing), and coordination of early intervention services to maximize their potential. Who, then, should coordinate the myriad services needed by the infant who is deaf or hard of hearing? In the medical model of blood 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 with medical interventions.

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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 intervention, however, few pediatricians and family physicians can be expected to have a complete and up-to-date level of educational sophistication. 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 loss. Even at a frequency of 1 to 2 affected children in every 1,000 newborns, a primary care physician might only care for a new baby with hearing loss once or twice in a decade.

The Case for the Primary Care Physician

Infants who fail a newborn screening test and are subsequently diagnosed with hearing loss are referred for generally extensive pediatric audiology care and ideally are enrolled in early intervention. Clearly, the initial stages of care include continuing evaluation with a number of sophisticated audiology testing protocols, and 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, consider causes that might be amenable to surgery, and provide medical clearance for amplification or other hearing technology. Given that 90% of children with congenital hearing loss have some residual hearing, amplification with appropriate hearing aids is a cornerstone of treatment for most. Special expertise is required when fitting amplification for infants and young children, as they cannot answer the examiner's questions or report about their own comfort or experience. Finally, early interventionists with skills in speech pathology, language acquisition, 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

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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 with the primary care physician provide far too many opportunities for one professional to undo the good work of others providing services to the infant and family. A primary care physician with incomplete knowledge may not realize the importance of timely assessment and very early intervention. By recruiting and educating physicians to provide support rather than act as a barrier, early intervention can be accomplished—dramatically improving outcomes for every child with hearing loss.

Furthermore, the primary care physician is positioned to reinforce the important messages delivered by the audiologist, the otologist/otolaryngologist, and the early interventionist. Such support may range from encouraging families to complete the recommended evaluation, emphasizing the importance of timing and prompt intervention, assuring the completion of any recommended testing or imaging procedures, and reinforcing compliance with recommendations, such as daily use of hearing technology. The primary care physician must also be recruited to monitor for and aggressively treat otitis media and retained middle ear fluid—conditions that might further compromise residual hearing.

Photo courtesy of NCHAM

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 primary care physician 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 in Table 1 should be brought by the physician to the team 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 intervention for these children and their families.

In this rapidly evolving field of knowledge, the physician should also have an eagerness to learn new information and to seek out educational resources. The physician must be willing to work collegially, not only with physician peers and specialists, but also with audiologists, speech pathologists, early interventionists, and education professionals.

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 early intervention 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 intervention team.
The care should be ***coordinated***.	The care should be ***compassionate***.	The care should 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 hearing loss is diagnosed, 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 important to assuring the most favorable outcomes for the child.

Finally, the physician should have a willingness to participate as an advocate for the infant and family in assuring access to timely evaluation and prompt initiation of intervention 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 with hearing loss 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 Early Hearing Detection and Intervention (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 early education of infants with hearing loss must provide timely information and feedback to the primary care physician about the child's ongoing care.

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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 deaf or hard of hearing. It is a new era when congenital hearing loss 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 deaf or hard of hearing.