CHALLENGING CASE: FAMILY RELATIONSHIPS AND ISSUES

Parental Request to Withhold a Hearing Test in a Newborn of Deaf Parents*

CASE

When the pediatrician went to examine Jenny at 1 day of age in the hospital, she was not surprised to find a healthy, full-term neonate with a normal physical examination, including a normal developmental assessment. She was responsive and robust with wide open eyes that engaged her mother and the pediatrician.

A review of the medical records revealed that both parents were significantly hearing-impaired. Jenny’s mother had a familial form of congenital neurosensory deafness. Both maternal grandparents had the same condition. Her mother learned to sign at an early age and completed a college degree. Jenny’s father acquired his deaf condition at 6 years of age after an episode of viral encephalitis. He communicated through sign language and was a successful administrator.

When the pediatrician suggested a hearing test (brainstem auditory evoked potential), Jenny’s mother, who was able to use spoken language as well as sign language, responded with a soft smile. “I don’t think that will be necessary. We will know if she can hear in a short time.”

Index terms: congenitally deaf, newborn hearing screening, parent consent.

Dr. Martin T. Stein

Screening for the presence of a disease in a pre-clinical state is common pediatric practice. It is the foundation of secondary prevention programs. At other times in contemporary medical practice, a variety of medical screening tests are requested by inquisitive parents either because of a concern about an inherited disorder, because of a particular or non-specific anxiety about a condition, or because it is simply available. Less often, a parent refuses a screening test recommended by a physician.

After more than two decades of pediatric practice, the situation encountered in the case under discussion was, at first, baffling. With an understanding that the mother carried an autosomal dominant form of congenital neurosensory hearing loss, the risk to the baby for inheriting the condition was 50%. Under the assumption that early detection and intervention with hearing aids would have a significant effect on language acquisition, didn’t I have an obligation to the child and her family to insist on the hearing test? Alternatively, an overzealous approach of insisting that the test be performed might incur the risk of alienating the parents. Finally, the refusal to test hearing seemed initially to be an unusual response in these knowledgeable parents. Why not discover the baby’s hearing status when the test was so simple and noninvasive?

In time, I gained an understanding about the parent’s thoughtful decision to withhold an audiological evaluation. The lessons learned about the parental decision-making process and about cultural imperatives that impact on medical decisions were important ones. To gain insight into the many questions raised by this case, two individuals with extensive experience among deaf children and families contributed commentaries. Dr. Steven Barnett is a physician in the Department of Family Medicine at the University of Rochester School of Medicine. He works with deaf patients and is interested in both clinical and cultural aspects of their health care. Professor Carol Padden is on the faculty in the Department of Communications at the University of California, San Diego. Congenitally deaf, Professor Padden has researched and written widely about unique cultural aspects within communities of deaf people.

Dr. Martin T. Stein, MD
Professor of Pediatrics
University of California, San Diego School of Medicine
San Diego, California

Dr. Steven Barnett

Although hearing loss in infants is relatively common (1 of 1000),1 it is often diagnosed late; the mean age at first detection of congenital hearing loss of moderate-to-profound intensity is 3 years.2 Some of the negative consequences of early hearing loss—speech and language delay with subsequent lower educational achievement3 and lower income and underemployment as adults4—are probably partially related to late diagnosis. With earlier detection, it is thought that some of these disadvantages can be minimized. Known risk factors can help identify half of the children with hearing loss.5 A risk factor for the child in this case is a family history of childhood hearing loss. Many physicians would agree that an infant with a hearing loss risk factor would benefit from a newborn hearing evaluation. Having parents who are deaf, who communicate in American Sign Language (ASL), and who decline a hearing evaluation all make the issues in this case more complex, and the best decision regarding hearing testing for this newborn becomes less clear.

Examination of the different perspectives may be

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helpful in understanding and resolving the disagreement over hearing testing. Both the physician, who recommends hearing tests, and the parents, who refuse them, have the same ultimate goal: a happy and healthy child. The differences in the approach to hearing testing may be a result of differences in the definitions of “happy and healthy” and of different perspectives on hearing testing.

For many people deafened early in life, deafness is not a disability. For them, being deaf is like being part of any other sociocultural minority community. The deaf community has its own language (ASL), social rules, theater, poetry, literature, and humor. ASL, recognized by linguists as a proper language and the third most commonly used language in the United States, is a rich and visual way for deaf people to communicate. The limitations of ASL in mainstream American culture are similar to those of other minority languages. Members of the deaf community primarily socialize and marry or partner with other deaf people. As is true within other sociocultural minority communities, there is a sense of shared experiences. For these deaf people, their deafness and language are a source of pride. They do not rate their health or their happiness as less because of their deafness. Many, if given the option, would not become hearing and would prefer to have children who, like them, are deaf. This is a different perspective from that of the physician, who is most likely hearing, and who was trained to think of deafness as a pathology.

Attitudes toward hearing testing probably also differ. Adults who have been deaf since childhood often have negative memories of childhood health care experiences. These memories may involve hearing testing, as well as hearing aid fittings, and even ear surgery. Deaf children often were forced to wear hearing aids that did not help them understand speech or that amplified sounds to a painful level. These interventions may all seem to be attempts to make the deaf child become a hearing individual. For deaf adults who view their deafness as an essential part of their person and as a connection to their cultural community, medical interventions to “cure” or to limit their deafness may be viewed as rejection. This rejection can be confusing to well-intentioned hearing physicians, for whom hearing tests are just another diagnostic test to detect disease.

In addition to learning about the parents’ perspective, it may be helpful for the physician in this case to examine the reasons for recommending a hearing test. The benefit of early identification of hearing loss is the opportunity to intervene in a way that minimizes delayed development of language. Language exposure for most young children is to speech. Language delay for children with hearing loss stems from the decreased access to the auditory (spoken) language in their environment. Interventions that increase language exposure, through hearing aids or speech therapy, are thought to help minimize the delay.

This language exposure scenario is true for the majority of families with infants who have hearing loss—90% of people with congenital hearing loss have parents with normal hearing and the family’s primary language is spoken. In families in which the parents are deaf and the primary language is a sign language, the scenario is different. In these families, the environmental language exposure is visual. Children in families with deaf parents who communicate in ASL have normal language development in ASL (as discussed by Lane et al. and noted by Petitto and Marentette). Infants in these environments talk earlier in sign language at the appropriate age.

How would hearing testing benefit an infant in an environment with visual language? There are three potential functional outcomes of hearing testing in this case. In one, the child is found to be deaf or so severely hard-of-hearing that hearing aids would be of no benefit. In this situation, the child is already in an environment conducive to language development. In a second, the child is found to have normal hearing, in which case she will learn ASL as a primary language, and like the children of non-English speaking immigrants to the United States, she will learn English outside of the family (and, in this case, from the hearing paternal grandparents). In the third possible situation, the child has a mild or moderate hearing loss for which amplification with hearing aids would be useful. As in the other two scenarios, this child will develop language appropriately in ASL, but hearing aids could potentially enhance the child’s ability to learn spoken English. In this situation, it is possible that hearing testing would be helpful, but it is important to realize that the benefits of early intervention for hearing loss on long-term functional and quality of life outcomes are unproven.

Before recommending hearing tests to determine whether the child is hard-of-hearing, it is helpful to have a sense of pretest probability. What is the hearing loss pattern in the maternal family? Does the mother have siblings, aunts, or uncles who are hard-of-hearing? Are there members of the paternal family with hearing loss (deaf or hard-of-hearing), which might indicate a genetic predisposition to infection-related hearing loss? By asking these questions, the physician not only gains valuable information about the infant’s potential for hearing loss but also establishes a relationship with the parents in a way that is culturally appropriate—within the deaf community, when meeting new people it is normal to inquire about deaf people in their family.

When discussing with parents the possibility of hearing testing for their infant, it is important for the physician to use culturally sensitive terms and phrases, such as “deaf, hard-of-hearing, or hearing” instead of “normal or abnormal”; “possibility” is neutral, whereas “risk” is negative; “speech delay” is more accurate than “language delay” with a child growing up normally learning ASL. The risk of pressing for a hearing evaluation against the parents’ wishes is alienating the family, which may limit all future health care interventions, including immunizations.

One of the goals of universal newborn hearing
screening is to identify the infants with hearing loss who have no known risk factors. This infant has an identified risk factor; knowing this, the child’s growth and development should be monitored by the parents and by the physician. Periodic developmental language testing is appropriate. The physician can ask parents about the child’s language use at home and can work with the sign language interpreter to assess the child’s vocabulary and sentence structure in the office (ASL and English syntax are different). If there is language delay in ASL, further testing may be warranted; deafness is sometimes associated with neurological problems. If the child is learning English, assessment of speech delay may be most accurate using other bilingual children as the norm. If speech is delayed, hearing testing may be appropriate.

Although there are recommendations for universal hearing screening of infants, these recommendations are probably based on the needs of deaf or hard-of-hearing infants who have hearing parents. The needs are probably different for deaf or hard-of-hearing infants with deaf parents. By understanding the reasons for hearing testing and by appreciating the perspectives of deaf parents, physicians can help keep families happy and healthy.

Steven Barnett, MD
Clinical Senior Instructor
Department of Family Medicine
University of Rochester School of Medicine
Rochester, New York

REFERENCES

Dr. Carol Padden
The question at hand for the pediatrician is when and how to approach parents of a deaf baby to determine whether the baby has inherited the parents’ deafness. It might seem obvious that testing should be done immediately after birth at the hospital, but some deaf parents will choose to wait. Most likely they have already considered the possibility of deafness before pregnancy. For these parents and others like them with inherited deafness, deafness is not a sudden or unexpected condition, but one which is deeply integrated into their cultural and social lives. Often when both parents are deaf, they are surrounded by family members and friends who are deaf. They do not view the condition as catastrophic or life-threatening.

Because there is a high rate of assortative mating (deaf by deaf) among deaf adults, there is a good likelihood that deaf adults may be carriers of several types of hereditary deafness. Without prenatal testing for most types of deafness, deaf parents look at the patterning of inheritance in their family to estimate their chances, mainly by informal means, and sometimes with professional genetics counseling. Indeed, deaf parents will want to know whether their child is deaf, but their need to know stems less from medical concern than from curiosity. Will the child be like themselves and become a part of their cultural community? What education should they plan for their children? Their families and friends in the community will share their curiosity and sense of anticipation.

So why wait? Some parents, as in this case, prefer to find out themselves. They want the discovery to be personal and intuitive rather than external and clinical. It sometimes may only take a few days after bringing the baby home from the hospital for them to detect the presence or absence of hearing loss. Deaf people are primed to observe and are conditioned to search for signs of hearing and deafness, such as a startled reaction to an abrupt sound or sleeping through a sudden loud noise. In this case, the parents seem to want to bond with the infant in the first few weeks and gradually come to realize the baby’s condition.

But this particular case should not be interpreted to suggest that deaf parents will always want to delay or decline immediate clinical diagnosis; some may even want the hospital to perform testing. Newborns’ responses can sometimes seem contradictory, and the parents may want a clinical diagnosis, particularly in cases in which the baby has partial hearing. And even when the parents are sure the baby is either deaf or hearing, they may still want confirmation later. The pediatrician can always ask in a follow-up examination whether the parents would like their home diagnosis to be confirmed. Here, the issue may not be whether to test, but when.

The lesson in this case may be that the same medical condition can have very different life interpretations; parents who know the condition intimately may have a different sense of urgency than those
who think of it as a clinical concern. These parents will appreciate the sensitivity of the pediatrician.

Carol A. Padden, PhD
Professor
Department of Communication
University of California, San Diego
San Diego, California

SUGGESTED READING

Dr. Martin T. Stein
Beginning with this Challenging Case, each case will be posted on the Journal of Developmental & Behavioral Pediatrics’ website: http://www.lww.com/DBP/

Participants on the web site are encouraged to comment on the case. Selected commentaries will be included in the published discussion in the Journal of Developmental & Behavioral Pediatrics. Through this format, we hope to encourage participation as well as discover new clinical insights or an alternative perspective on each case. Challenging Cases will be posted bimonthly on the 15th.

Several clinicians provided thoughtful commentaries that both supported Dr. Barnett and Professor Padden and offered alternative views. Excerpts from the website discussion follow:

Dr. William Oppenheim
The fact is, the mother knows more about what you are going to be looking for than you ever will. She has lived, loved, and been educated with the hearing loss, and she will undoubtedly know people in the deaf community who are knowledgeable about the right diagnosticians and ear specialists.

Dr. Debbie Sedberry
I agree with the previous comment that “mom knows best.” As a pediatrician, you’re going to be seeing this youngster within the next few weeks again anyway. Within a month or two, you and the parents will have a good sense as to whether or not this baby is hearing. Why push the parents at this point when you can gently bring it up again in a few weeks if it seems appropriate? I doubt that you will lose any significant degree of language development by waiting a short time (like 4 weeks) to get a sense of the infant’s hearing. If the baby is hearing normally, then you can begin to discuss with the family the challenges of raising a child with normal hearing. If not, you and the family can begin to discuss appropriate interventions.

Dr. Barbara Howard
This case involves a child at risk for hearing impairment with two hearing-impaired parents. Even though one parent was not deaf as a result of a hereditary problem, he was deaf from an early age. Therefore both parents may be biased toward having a hearing-impaired child and raising her in their culture. Medical opinions that it is “better” to be hearing are not always regarded as common sense by some members of that culture who seem to regard this as belittling to their personal value. Hearing impairment is typically not detected until the second half of the first year when vocalizations decrease or even later when speech and language development are impaired. Important development of cortical representations of language are missed if there is no hearing early on. In addition, the teaching of other forms of communication may not have been started if the impairment is unknown.

In this case, however, the parents presumably would be signing to the infant. No one might be vocalizing to the child even if she could hear, however. Thus her own choices about whether to speak, sign, or do both and which culture to be part of will be potentially curtailed by not identifying her hearing status at birth. I haven’t been in the position of conflict with parents about hearing status, but I think I would listen respectfully to their opinion about how they want to raise their child but then use the argument that their child’s options should not be curtailed by not determining the hearing status.

Dr. Henry Shapiro
Barbara Howard’s comments raise another issue. If the child is hearing-impaired, but only mildly or moderately, early amplification could make a dramatic difference. The case as posted implies that the mother would assume that her child would be severely or profoundly hearing-impaired. We do not have good research about the period between birth and 6 months, but evidence is mounting that there are outcome differences after 6 months. (Seventy-two deaf or hard-of-hearing children whose hearing losses were identified by 6 months of age were compared with 78 children whose hearing losses were identified after 6 months of age. All of the children received early intervention services within an age of 2 months after identification. Children whose hearing losses were identified by 6 months of age demonstrated significantly better language scores than did children identified after 6 months of age. [Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. Pediatrics 102(5):1161–1171, 1998] M.T.S.)

Dr. Sheila Gahagan
Most hearing-impaired people do not live in a world totally free of speaking people. Although the parents may favor a hearing-impaired culture, they usually have relatives and colleagues who hear and speak. Therefore, it is unlikely that the child would live in a world without any speech. I would imagine it would be discriminatory not to provide early intervention service to children of hearing-impaired parents, just as one would to children who are at risk for speech delay from any other etiology. The cul-
Dr. Martin Stein

By a few months of age, the parents reported that they knew that their child was hearing. An auditory brainstem evoked potential examination confirmed their observation. She is now 6 years old, thriving and academically advanced, with a capacity to communicate in both spoken language and sign language.
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