

*The following is a brief overview -- in the form of a Q&A -- of the physician's role in the follow-up of infant who did not pass newborn hearing screening. This information will be useful in efforts to gain a common knowledge base for moving forward.*

**Question: What should a primary care physician do if a newborn doesn't pass the hospital-based hearing screening?**

It is important to understand the prevalence of newborn hearing loss and the accuracy of initial hospital screening. If a newborn is never screened before hospital discharge, the child has one in 650 chance of having an undetected congenital hearing loss. But, depending on the screening test used, the child who fails the initial screening may have a risk of confirmed hearing loss as high as one in 10! It is reasonable to reassure families that their own child may be shown to have normal hearing, but it is critical that every newborn who fails initial screening return for follow-up screening and confirmatory testing if indicated. This may be a joint responsibility shared with the hospital and/or the audiology department, but nothing is as powerful as a phone call from the physician if for any reason the child has not returned in a timely manner.

**Question: When should the infant be rescreened?**

Infants should return promptly for rescreening – within the first few weeks of life. The infant should complete rescreening and confirmatory testing quickly so that if a hearing loss is identified, early intervention, including amplification with a hearing aid, can begin without delay. It is a reasonable goal to have every hard-of-hearing infant identified by two months of age. Any delay in this process deprives the developing brain of the auditory stimulus it requires for normal language development. (Even profoundly deaf newborns can benefit from very early introduction of sign language and may possibly benefit from amplification.) Also, rescreening the infant as an outpatient or proceeding with confirmatory testing requires a baby to be very quiet or asleep. Every month that passes decreases the likelihood that the infant will be in a quiet state at the time of the recheck appointment, and older infants may even require sedation just to perform the testing.

**Question: What does rescreening typically involve?**

Rescreening may initially be performed with the same screening techniques used in the hospital, either otoacoustic emission testing (OAE) or automated auditory brainstem response testing (AABR). These screening tests are "physiologic" in the nature of their measurement, rather than "behavioral" testing which requires the observation of a behavioral response in the infant.

**Question: What should parents be told if their newborn does not pass the screening test in the hospital?**

First, parents should be reassured. Most infants who do not pass the hospital-based test are eventually shown to have normal hearing. But parents should not be complacent – every baby who fails the initial screen must return for follow-up. If the child is subsequently identified as having a congenital hearing loss, parents should know that early intervention has been shown to have dramatic results, with subsequent language development at near-normal levels. Parents will never need to say, "If only I had known sooner."

**Question: What about the infant who only fails the screening in one ear?**

Even though unilateral hearing loss may be a less severe condition than bilateral hearing loss, these infants also deserve prompt follow-up. If a unilateral hearing loss is confirmed, the parents can be counseled about how to maximize the child's language development by being sure that the auditory stimuli are reaching the better ear effectively. But equally important, some of these children have progression of hearing loss in the ear

that initially passed screening. In addition to rescreening, any child with a confirmed unilateral hearing loss must be followed closely over time to assure that the condition does not evolve into a bilateral hearing loss.

**Question: What can parents expect in an audiologic evaluation if the infant does not pass the rescreen?**

Most importantly, the primary care physician should make sure that all follow-up and confirmatory testing is performed by pediatric audiologists with experience testing infants and fitting them with hearing aids, even if this requires extra travel for the infant and their family. You can determine who is a qualified pediatric audiologist by asking how many infants that provider has seen in that month, and whether he or she has expertise in using physiologic and behavioral methods to test hearing in infants. Neurologists also use Auditory Brainstem Responses to evaluate the integrity of the brainstem in specific neurological disorders. This assessment is inherently different than the pediatric audiologist's assessment using ABR to evaluate infant hearing; thus referral to a neurologist for an infant hearing assessment is not recommended.

Explain to parents that they can expect the audiologist to administer a battery of tests to assess the integrity of the auditory system from the outer ear through the inner ear and even the brain stem. This confirmatory testing is also performed using "physiologic" testing, typically including a standard diagnostic Auditory Brainstem Response (ABR, also known as BAER) along with additional OAE testing. The ABR may include different types of stimuli and assess the system for air conduction and bone conduction. Testing for middle function will also be done. The test battery may require several visits to accurately characterize the hearing loss.

Behavioral testing techniques are reliable after an infant is 6 to 12 months old – far too late to allow for early intervention and amplification, but they should become a part of the test battery used in the ongoing assessment on the infant with hearing loss.

**Question: If a child is confirmed to have congenital hearing loss, what else should the primary care physician do?**

Address family concerns. Families will need help understanding this new medical development. Work closely with the audiologist to better understand the nature and degree of the hearing loss. Pediatricians should interface with professionals in education to provide families with needed services for the infant with hearing loss.

Arrange for a complete evaluation by an otolaryngologist or otologist with experience working with infants and young children. Some children require further evaluation to assess the potential of progressive hearing loss, and all require the specialist to give medical clearance for the use of a hearing aid.

As we discover the increasing frequency of genetic syndromes among children with congenital hearing loss, it is recommended that these families also be referred to a medical geneticist with experience in the field of congenital hearing loss. Thirty percent of hearing loss is of uncertain etiology and there are more than 200 syndromic and non-syndromic forms of hearing loss that have been identified; about 20 percent will have associated clinical findings.

Every affected newborn should have a complete evaluation by a pediatric ophthalmologist, to assure that the visual stimuli to the brain are in no way compromised, and to assess for any associated eye anomalies or genetic syndromes with both visual and auditory impairment.

After hearing loss is confirmed, physicians need to be involved in the following:

- ▶ Coordinating services with the Individuals with Disabilities Education Act Part C agencies. Part C

agencies are responsible for Child Find and intervention for children with disabilities.

- ▶ Monitoring middle ear status to avoid further compromise to hearing.
- ▶ Monitoring developmental milestones because 30-40 percent of children with hearing loss will demonstrate multiple disabilities or delays.

**Question: What does audiologic habilitation mean? What does it entail?**

Audiologic habilitation includes hearing aid fitting. Hearing aid fitting proceeds when audiologic, otolaryngologic and other medical evaluations are in accord and the parent is in agreement with this recommendation. The fitting should take place in the first months of life; this is possible based on physiologic testing alone to provide maximum access to the acoustic features of speech in a safe listening range.

As physicians, we have all learned to respond quickly to an abnormal newborn screening test for PKU or hypothyroidism. It is time now for us to respond with similar urgency when a newborn does not pass the initial hearing screening test.