Problem with baby’s hearing? An intervention checklist

It’s time to make sure infants with positive screens for hearing loss get the follow-up treatment they need—and deserve. This tool can help.

**PRACTICE RECOMMENDATIONS**

- Check hearing screening results for all newborns in your practice. **B**
- Refer all newborns who fail screening for audiological and medical evaluation and diagnosis before 3 months of age. **B**
- Refer infants with diagnosed hearing loss for early intervention services no later than 6 months of age. **B**
- Educate families about services and resources available to them and their hearing-impaired child. **B**

**Strength of recommendation (SOR)**

- **A** Good-quality patient-oriented evidence
- **B** Inconsistent or limited-quality patient-oriented evidence
- **C** Consensus, usual practice, opinion, disease-oriented evidence, case series

Congenital permanent hearing loss occurs in about 3 of every 1000 births.¹ Undiagnosed hearing loss can result in speech-language, academic, social, and other developmental delays. Until about 20 years ago, most children with hearing loss were not diagnosed until about 3 years of age.² By that age, opportunities for effective intervention to help these children develop communication skills were often delayed, and many children remained seriously disabled.

In this enlightened age, when newborn hearing screening is nearly universal (92%), the prospects for children with hearing impairments are brighter—but not as bright as they could be.³ That’s because more than half the newborns with positive screens are lost to follow-up.³ Too many remain “lost,” without a diagnosis or access to services, until they show up at school without the language skills they need to keep up, academically or socially, with their classmates.

The medical home can help

The American Academy of Pediatrics (AAP) and the Health Resources Services Administration aggressively promote the concept of the medical home as the best locus for coordinating the care of children with special needs, and the American Academy of Family Physicians (AAFP) has endorsed the medical home concept.⁴ According to the AAFP’s Joint Statement of Principles, the medical home is responsible for coordinating care across all elements of the health care system and the patient’s community.⁴ Most physicians in a recent survey believed the medical home should be responsible for coordinating services and guiding families in the development of intervention plans for children with hearing loss.⁵ Family physicians who provide a medical home for infants and young children are in an ideal position to ensure that children with hearing loss are not lost to follow-up and that they receive the services they need to lead healthy lives.

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What follow-up entails
According to the 2007 Position Paper of the Joint Committee on Infant Hearing, a body composed of representatives from the AAP, the American Academy of Audiology, the American Speech-Language-Hearing Association, and other professional organizations concerned with hearing loss, an effective program to mitigate the impact of hearing loss should follow this timetable6:

- By 1 month of age, all infants should receive hearing screening. (Of note: In 2008, the US Preventive Services Task Force issued a B recommendation for universal hearing screening of all newborns.)
- By 3 months of age, hearing loss should be diagnosed.
- Within 1 month of diagnosis, hearing aids should be fitted for infants whose parents choose hearing aids.
- As soon as possible after diagnosis—but no later than 6 months of age—infants with confirmed, permanent hearing loss should receive early intervention services.

Intervention services should include medical and surgical evaluation, evaluation for hearing aids, and then cochlear implants for those with severe-to-profound hearing loss who do not benefit from hearing aids. Communication assessment and therapy should also be considered. The goal of intervention is to help infants with hearing loss develop communication competence, social skills, emotional well-being, and positive self-esteem.6

The CLINICAL TOOL provides a detailed overview of the early hearing detection and intervention (EHDI) process outlined by the Joint Committee and a checklist of roles and responsibilities for physicians serving as medical homes for children with hearing loss and their families. Unfortunately, many primary caregivers report that their medical training did not prepare them to guide families through this process.4-10 This article is intended to provide the additional information caregivers have requested to help them meet these obligations.

Getting an early start: Newborn screening
Ideally, intervention should begin before a child is born. When parents come in for prenatal visits, talk to them about the importance of newborn hearing screening. Tell them to expect that their baby will be screened at the birth hospital or at an outpatient audiology facility, and that this screening should be done by the time the baby is 1 month of age.

Tell parents their baby’s hearing will be tested with automated screening equipment that measures otoacoustic emissions from the baby’s ears or auditory brainstem response (ABR) to sound, both measurements that correlate with a child’s hearing and auditory behavior. Infants in well-baby nurseries can be screened by either technology. Infants in neonatal intensive care units (NICUs) for more than 5 days should be screened with ABR technology, which is better able to pick up neural losses.3 NICU babies frequently are at higher risk for neural hearing loss including auditory neuropathy/dyssynchrony, a condition that may account for about 8% of pediatric hearing losses annually.3

Be sure to explain to expectant couples that a positive screen indicates only that a problem may exist. It is not equivalent to a diagnosis of hearing loss, which is usually made by an audiologist.

The medical home’s role. The medical home should make sure that infants who fail an initial or secondary hospital screening and those who were missed or born outside the hospital are referred for outpatient screening. Newborns who fail initial screening should have both ears rescreened before hospital discharge, even if only one ear had failed previously.6 Additionally, any infant readmitted to the hospital within the first month of life who has a condition associated with potential hearing loss should be rescreened before discharge, preferably with an ABR.6 Risk factors associated with permanent-congenital, delayed-onset, or progressive hearing loss in children are listed in the TABLE.

Reporting screening results
Screening results are useful only if they are transmitted to caregivers and families. Breakdowns in transmission are a persistent problem for the early hearing detection and intervention process.5 The process is facilitated when hospitals make sure that all ba-
## CLINICAL TOOL

### The early hearing detection and intervention process

<table>
<thead>
<tr>
<th>Prenatal period</th>
<th>Birth – 1 month</th>
<th>By 3 months</th>
<th>No later than 6 months</th>
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<tbody>
<tr>
<td>Education for parents about the importance of newborn hearing screening and the distinction between screening and diagnosis</td>
<td>Hearing screening for all infants in hospital or at audiologist outpatient facility</td>
<td>Infants with positive screens are diagnosed by an audiologist with auditory brainstem response testing</td>
<td>Physician and parents monitor developmental milestones and experience with hearing aids</td>
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<td></td>
<td>Auditory brainstem response screening for all NICU infants ≥5 days of age</td>
<td>Hearing loss is ruled out or confirmed</td>
<td>Meeting with family and early intervention personnel to develop an individualized family service plan</td>
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<td>Rescreening for all infants with risk factors who are hospitalized within 1 month of discharge</td>
<td>Audiologist shares results with family, medical home, and the early hearing detection and intervention coordinator</td>
<td>Individualized family service plan is in place, as mandated by federal law under the Individuals with Disabilities Education Act</td>
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<td></td>
<td>Infants are linked to a medical home before discharge by the hospital</td>
<td>Families are counseled regarding diagnosis and follow-up and given educational materials</td>
<td>Early intervention services are instituted in accordance with the individualized family service plan</td>
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<td></td>
<td>Screening results are given to families and to the baby’s medical home</td>
<td>Audiologist recommends treatment in the medical home or referral to an ear, nose, and throat specialist for medical evaluation and treatment</td>
<td>Together, audiologist and family develop expectations for hearing aids</td>
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<td>Families are counseled about screening results and follow-up</td>
<td>Audiologist and/or physician provides referrals for genetic counseling and ophthalmologic consultation</td>
<td>If hearing aids are unsuccessful, families are counseled about cochlear implants</td>
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<td>Audiologist alerts parents and the medical home that child may need hearing aids and early intervention services</td>
<td>If family wishes, audiologist or medical home makes a referral to a cochlear implant team</td>
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<td>Audiologist or physician counsels parents on the test results, treatments, and communication options: aural-oral, total communication, and sign language</td>
<td>Medical clearance and insurance authorizations are obtained for cochlear implants</td>
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<td></td>
<td>Reports from all involved providers and agencies are transmitted to state early hearing detection and intervention coordinator</td>
</tr>
</tbody>
</table>

### Checklist for family physicians

- Encourage all families to have their baby’s hearing screened
- Explain screening procedures
- Assess for family history of hearing loss
- Provide informational materials about newborn hearing screening
- Answer family’s questions about newborn screening or hearing loss
- Review screening results
- Make referrals for outpatient screening and audiolotogic diagnostic evaluation by an audiologist
- Ensure all infants hospitalized after discharge are rescreened
- Assess risk factors for hearing loss, including congenital and delayed-onset types
- Ensure that screening results have been transmitted to state early hearing detection and intervention coordinator
- Review audiologic diagnostic evaluation results
- Ensure that an audiologic reevaluation has been completed
- Review findings from otolaryngology and audiologic consultations with family
- Encourage family to comply with professionals’ recommendations and stress importance of keeping appointments
- Refer for genetic counseling and ophthalmologic consultation
- Provide families with preliminary information on amplification and communication options
- Coordinate early intervention services
- Confer with audiologist on child’s progress with hearing aids and consideration of cochlear implants
- Provide families with basic information about cochlear implants
- Counsel families about the risks and benefits of cochlear implants
- Make referral to cochlear implant team
- Encourage families to comply with professionals’ recommendations

NICU, neonatal intensive care unit.

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abies and their families are linked to a medical home at the time of discharge.

Hospital personnel should also be responsible for providing screening results to families in a face-to-face meeting. Medical home providers should review screening results again with parents and answer any questions that might have arisen after the initial hospital stay. Screening results should be given to parents in a sensitive manner, and patient education materials should be provided in parents’ native language, written at an appropriate reading level. Because hospital staff may not be trained to do this properly, it is important that medical home providers oversee the process and address any parental concerns.2

Screening results should also be reported by the birth hospital to the state’s EHDI coordinator, part of the national tracking and surveillance system funded by the Centers for Disease Control and Prevention to ensure that all children with hearing loss achieve communication and social skills commensurate with their cognitive abilities.11 You can find contact information for your state coordinator at www.cdc.gov/ncbddd/ehdi/documents/EHDI_contact.pdf.

**Diagnosing hearing loss**

When an infant who fails newborn screening testing arrives at the medical home, the urgent next step is to make sure he or she is referred to a pediatric audiologist for a complete audiologic diagnostic evaluation. Advising parents to “wait and see” is not appropriate; researchers have identified that response as a major obstacle to successful follow-up.2 The audiologic evaluation should be done by the time the baby is 3 months of age and should be performed by a pediatric audiologist specializing in diagnosis and management of young children with hearing loss.4-6

Tell parents that, with their approval, the audiologist may fit the baby with hearing aids as early as this visit, as amplification can help even very young infants hear all sounds in the environment, particularly spoken language.

**Ongoing monitoring**

Family practices serving as medical homes should continue to monitor children who pass their newborn screening but have high-risk factors for delayed-onset hearing loss. Those factors are listed in the Table. Refer children at higher risk for an audiologic diagnostic evaluation by 24 to 30 months of age.6 Follow-up on parental concerns about infant hearing or speech and monitor infants’ developmental milestones, auditory skills, and middle ear status using the AAP’s pediatric periodicity schedule.3 Conduct global developmen-
tal screenings at 9, 18, and 24 to 30 months of age, and refer for speech-language-hearing evaluations when appropriate.⁶

**The medical home as central referral point**

The medical home is a central referral point for the complex needs of children with hearing loss. The physician and all other providers involved in the child’s care should report results of diagnostic evaluations to state EHDI coordinators. The medical home’s continued involvement includes medical clearance for hearing aids, additional consultations, and screenings as necessary to help children receive needed services and keep them from being lost to the system.⁵,⁹

Referral for genetic consultation is important, because about half of all autosomal recessive sensorineural hearing losses that are not part of a syndrome are caused by mutations in the Connexin 26 GJB2 gene.¹²,¹³ Referral to a pediatric ophthalmologist is similarly important, to identify deficits in visual acuity that frequently co-occur with hearing loss, especially in preterm infants.⁶ Results of these consultations can assist the physician in guiding families through the intervention process. Coordination of care among multiple providers is essential. When a family physician’s practice serves as a medical home for a child with hearing loss, the physician should oversee and coordinate the efforts of all stakeholders in the EHDI process; make referrals to, and receive reports from, all providers involved in diagnosis and treatment; and ensure that relevant information is shared. During all phases of the process, the role of the family physician is to encourage families

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**TABLE**

Risk indicators associated with hearing loss in childhood

- Caregiver concerns* about hearing, speech, language, or developmental delay
- Family history* of permanent childhood hearing loss
- Neonatal intensive care for ≥5 days, or with any of the following:
  - Extracorporeal membrane oxygenation*
  - Assisted ventilation
  - Exposure to ototoxic medications such as gentamicin and tobramycin or loop diuretics such as furosemide
  - Hyperbilirubinemia requiring exchange transfusion
- In utero infections such as cytomegalovirus, * herpes, rubella, syphilis, and toxoplasmosis
- Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
- Syndromes associated with hearing loss or progressive or late-onset hearing loss, * such as neurofibromatosis and osteopetrosis, as well as Usher, Waardenburg, Alport, Pendred, or Jervell and Lange-Nielsen syndromes
- Neurodegenerative disorders* such as Hunter syndrome, or sensory motor neuropathies such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
- Culture-positive postnatal infections associated with sensorineural hearing loss, * including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
- Head trauma, especially basal skull/temporal bone fractures* that require hospitalization
- Chemotherapy*

*These indicators are of greater concern for delayed-onset hearing loss.

**Source:** American Academy of Pediatrics, Joint Committee on Infant Hearing. Pediatrics. 2007.⁶

Tell parents that a positive screen indicates that there may be a problem; it is not the same as a diagnosis of hearing loss.
to comply with professionals’ recommendations and to stress the importance of making and keeping scheduled appointments.

Making plans for intervention
Families with children who have any degree of permanent hearing loss in one or both ears are entitled to early intervention services.6 In most states, these services are provided by a multidisciplinary team at no or low cost through a federal grant program. Services can be home- or center-based. They may include, as needed, education for the affected child and family; physical, speech/language, and occupational therapy; and social work and psychotherapy services. The team works with the family to develop an individualized family service plan to document and guide the early intervention process.14

Families with a hearing-impaired child have a range of options to choose from in their search for an approach that is best for the child and most acceptable to them. Communication options span a continuum from emphasis on sign language as used by the deaf community to a variety of oral-aural approaches designed to lead to spoken language. Parental choices are influenced most heavily by the child’s success with hearing aids. Parents need unbiased, culturally sensitive counseling about all available communication options and hearing technologies, so they can make informed choices for their children.6

Answering parents’ questions
To ensure that parents have appropriate expectations for what auditory technology can do for their child, they need to receive information about traditional digital signal processing hearing aids, osseointegrated hearing implant systems (also known as bone-anchored hearing aids), and cochlear implants. They need to know that a trial period will probably be necessary to determine whether hearing aids are appropriate or if cochlear implantation should be considered. Cost of hearing aids and cochlear implants is a serious concern for parents, and for many, it presents a major barrier to obtaining optimal care.2

Many insurance plans cover cochlear implants, but require that children be at least 12 months of age, have bilateral profound sensorineural hearing loss (or severe-to-profound sensorineural hearing loss for those ≥24 months of age), receive minimal benefit from hearing aids, be enrolled in auditory rehabilitative therapy, and possess no medical contraindications.15 Hearing aids and cochlear implants may also be covered for children enrolled in Medicaid or the State Children’s Health Insurance Program.16 However, according to a recent evaluation of hearing screening programs nationwide, public and private insurance policies almost never provide adequate coverage for hearing services.2

As parents consider the options, be sure they are aware that some children younger than 12 months of age are receiving cochlear implants (sometimes in both ears), that stimulation of both ears is being recommended at earlier ages, and that it is also common for children to use a hearing aid in one ear and a cochlear implant in the other.17,18 Distinguishing among hearing loss types and knowing which treatment options are most effective helps physicians counsel families appropriately about making the best decisions for their children. Some physicians have expressed uncertainties about these issues and have requested additional information on this topic.19 Audiologists can help physicians obtain this information and help them to better counsel families about these options.

References


