

A child in my practice has been identified with a permanent hearing loss. Now what do I do?

Johnny was screened as part of the universal newborn hearing screening program in your state and did not pass. You referred Johnny for a diagnostic audiologic evaluation and the report back informs you that he has a moderately severe sensorineural hearing loss. His mother comes in to follow-up on these results and wants your opinion on some things that the audiologist recommended.

Although this is not a daily occurrence in your office practice, you realize that providing the right guidance and care is very important for Johnny and his family. Providing psychosocial support for the family and implementing medical and therapeutic/educational interventions are key to ensuring the best outcomes possible for Johnny.

It is not just about hearing; it is also about brain development and prevention of delays. Early and prompt intervention is critical. Hearing loss is considered a developmental emergency. As such, the health care provider must support the family and ensure that there is no delay in beginning early intervention once hearing loss has been confirmed. This might include early fitting of amplification, early use of sign language, or both. The process of completing medical referrals should never delay prompt intervention once hearing loss is confirmed.

Developmentally, the needs of children with hearing loss are often served by **three types of services**: 1) Audiologists knowledgeable in pediatric testing and amplification, 2) Early Intervention Providers, and 3) Speech-language pathologists.

Because hearing loss affects brain development, the developmental and behavioral care of children with hearing loss is also important for primary care providers to address. All children with hearing loss should have a managing audiologist who assists in monitoring the status of their hearing and effectiveness of amplification strategies. Young children often need frequent visits to the **pediatric audiologist**.

Referral to **Early Intervention Services** as soon as a diagnosis of hearing loss is made can empower families in meeting their child's communication and developmental needs and decrease the negative impact of hearing loss on a child's language and cognitive development. There may be many different types of expertise among early intervention providers. It is important to have access to professionals who can provide unbiased information about communication modes, provide appropriate family support, and provide expertise in the communication mode the family chooses.

Some families access private therapies beyond those provided by Early Intervention Services. **Speech-language pathologists** can monitor speech and language development and provide families with support and strategies to encourage language development for their child with a hearing loss. Speech-language pathologists may have a variety of skill sets. These skill sets (ie, sign language, auditory-oral focus, cued speech, etc) should support a family's choices for their child's communication development.

Medically, the **primary goals** are: 1) identification of an etiology of the hearing loss to help the family anticipate their child's needs as well as those of siblings, and 2) preventing or diminishing

the impact of secondary medical concerns (ie, vision health, additive effect of middle ear disease on hearing levels).

To attain these medical goals, there are **three subspecialty referrals** that are considered standard of care.

- An otolaryngologist knowledgeable in pediatric hearing loss is involved in providing clearance for hearing aid use, undertakes a number of medical tests to determine an etiology of the hearing loss, and monitors children who might be candidates for the technology of a cochlear implant.
- A geneticist knowledgeable in pediatric hearing loss assists in determining genetic causes of hearing loss, including syndromic and nonsyndromic. Geneticists provide information to families about recurrence risk and risks of hearing loss to siblings and extended family members.
- An ophthalmologist knowledgeable in eye conditions associated with pediatric hearing loss can help identify conditions that can affect vision in children with hearing loss.

These referrals can be helpful in the initial evaluation and work-up of a child with sensorineural hearing loss; however, monitoring of medical conditions needs to continue at subsequent health maintenance and other illness-related visits.

Particularly relevant to children with hearing loss is aggressively managing **middle ear disease**. Although there are clinical protocols for the management of middle ear disease in typically developing children, a child with hearing loss requires a different approach. Even small changes in hearing related to middle ear disease can substantially decrease the impact of amplification on language development and behavior. Children who use hearing aids may not be able to tolerate their amplification during an ear infection. Persistent fluid may prolong the time of ineffective amplification. Referral to the otolaryngologist for these findings is recommended.

It is also important to follow typical anticipatory care for children with hearing loss, specifically **regular vision screening and immunizations**. Vision health is particularly important for children who have hearing loss. Additionally, children with inner ear abnormalities are at higher risk of acquiring meningitis. Children who are being considered for a cochlear implant should receive their full primary series and booster immunizations against pneumococcal disease and *Haemophilus influenzae* type b.

As Johnny's primary care provider, you play a **pivotal role** in monitoring health conditions that can affect his progress and in supporting and empowering his family in your role as a medical home, a coordinator of the variety of medical and educational systems and providers that promote Johnny's overall health and well-being.

Reference:

Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*. 2007;120(4):898-921.