

AMPLIFICATION, IMPLANTS, AND FM SYSTEMS FOR INFANTS AND YOUNG CHILDREN WITH HEARING LOSS

Estimates of the prevalence of congenital/prelingual hearing loss in infants are 3/1000 for well-babies and 3/100 for neonatal intensive care (NICU) babies.¹ It is one of the most common birth defects in the U.S, occurring without respect to ethnic background, gender, or socioeconomic status.² Late identification and intervention of infant hearing loss has been linked to life-long deficits in speech and language development, socialemotional development, and academic progress.³⁻⁴ Left untreated, hearing loss has a broad based impact, leading to significant delays in cognitive, language, self-help and social/emotional development with an estimated lifetime cost of \$441,000 per person.⁵⁻⁶ The literature clearly demonstrates the role of early hearing detection and intervention (EHDI) programs in reducing the negative consequences of untreated congenital/prelingual hearing loss.⁷⁻¹¹ Optimal intervention strategies for the infant with any degree of hearing loss require that intervention begin as soon as there is confirmation of a permanent hearing loss to enhance the child's acquisition of developmentally appropriate language skills.⁹⁻¹¹

Timely decision making and parental involvement with this process facilitates language development, early reading skills, and social-emotional development.⁷ This familycentered philosophy recognizes that family members are the primary decision makers regarding their child's best interest.¹²⁻¹³ Effective decision-making skills contribute to increased confidence, parental empowerment, and self-efficacy leading to successful long-term outcomes for infants with HL.¹³⁻¹⁵

Pediatricians with an understanding of the need for early and timely parental decisions about technology play a key role in supporting informed family choices.¹⁶⁻¹⁷

What happens after diagnosis of hearing loss?

Learning that a child has a hearing loss can be an emotional process for parents and families. Most parents go through a natural grieving process that may or may not include denial.¹⁸⁻¹⁹ Some parents may procrastinate about making a decision related to communication options and may seek other opinions. Some parents need more time than others to reach the stage of acceptance. The family's coping strategies, family support system, and access to resources all play a role in the way that the family addresses intervention issues.²⁰⁻²¹ The information that parents are presented with and expected to absorb in a relatively short period of time is overwhelming and can be paralyzing. One of the most important early decisions that families face is about mode of communication. For families who are Deaf culture, sign language may be chosen as the primary means of communication.²² This is the native language and language of the home. Deaf culture families often feel that the child should not wear hearing aids until they are approaching school age. Family choices based on cultural values are recognized as valid decisions.

For families who use spoken language as the primary means of communication, **timeliness is everything**. Fitting technology and following up with training before the age of six months (early intervention) is critical.⁹ Timeliness is so important that **a decision to do 'something' as opposed to 'nothing'** is a good first step. Families who value spoken language as the desired outcome for their child do not have the luxury of time. Parents can view these early decisions as dynamic and open to change as new information becomes available.

Many families chose to use a combination of sign language and spoken language, placing value on 'any means of communication' as opposed to a 'specific mode of communication'. In order to make informed choices – parents need to know what the options are and why one option might be more appropriate than another for their family.⁹

Families value the expert opinion of a physician. Encouraging families to focus on the outcome they want for their child and to take the necessary steps to reach their goal is one way pediatricians can support the family through this process and facilitate timely intervention.²³

What types of hearing technology are used with infants and why?

There are three basic types of technology that can provide access for development of auditory skills. These include hearing aids, implants (cochlear and bone anchored), and FM systems. Technology must be coupled with appropriate auditory, speech, and/or language training to optimize outcomes.²²

Technology

Hearing aids and implants provide auditory stimulation and must be worn all waking hours. FM systems are worn in specific situations to provide auditory access across distance or in background noise.

Training

Training in auditory skills, speech, and language (regarded as training the brain) is needed for the child to develop age appropriate communication skills. When a child is very young (birth to 3 or 4 years of age), therapy is focused on teaching the parent how to provide auditory stimulation and a rich language environment. The focus of treatment changes to a child-centered approach when the child approaches school age.

Hearing Aids

Hearing aids can be fit as soon the hearing loss is diagnosed, as early as 2 to 3 weeks of age. Children fit with hearing aids prior to 6 months of age demonstrate language development skills commensurate to their peers.⁹⁻¹¹ Hearing aid options for infants are behind-the-ear (BTE) hearing aids with earmolds or bone conduction hearing aids for some children with anotia, atresia, microtia, or ossicular deformities resulting in permanent conductive hearing loss.

BTE hearing aids and earmolds are used with most infants and can be fit for any degree of hearing loss (mild, moderate, severe, or profound). BTE hearing aids are fit to the ear with a custom earmold, made after an impression of the ear is taken. During the first year of life, the earmold may need to be changed every month due to rapid growth of the ear canal. From ages 2 to 3, the earmold may need to be changed every 3 or 4 months.²⁴ Earmolds are changed every six months for children after they reach school-age. BTE hearing aids are practical and durable with a lower repair rate than other types of hearing aids. In-the-ear (ITE) hearing aids are not appropriate for infants and young children due to rapid changes of the ear canal during the first few years of life. Most children continue wearing BTE hearing aids until they reach adolescence.

A BTE hearing aid consists of a microphone, amplifier, speaker, and power supply. It is connected to the earmold with tubing. Digital hearing aids first became available in the early 1990's.²⁵ Although analog hearing aids are still manufactured by a few companies, the majority of BTE hearing aids are both programmable and digital. Programmable hearing aids can be attached to a computer enabling the audiologist to adjust the hearing aid for the child's hearing loss and make changes if the hearing loss gets worse.²⁴ The hearing aid program is adjusted to compensate for the intensity and frequency of sounds needed to provide audibility of speech. Although many hearing aid companies manufacture BTE hearing aids, those catering to the

needs of the pediatric population include Oticon (www.oticon.com), Phonak (www.phonak.com), Siemens (www.siemens-hearing.com), Unitron (www.unitronhearing.us), and Widex (www.widex.com).

Cochlear Implants

If a child has a severe or profound sensorineural hearing loss, the child may be a candidate for a cochlear implant²⁶, however; infants younger than 12 months of age cannot be implanted (FDA regulations). Children who are cochlear implant candidates wear BTE hearing aids until the implant surgery is scheduled.

A cochlear implant consists of the external component (transmitter and speech processor) and the internal component (receiver and electrode array). Cochlear implants bypass damaged portions of the ear and directly stimulate the auditory nerve. Signals generated by the implant are sent by way of the auditory nerve to the brain, which recognizes the signals as sound.

A cochlear implant is not a magic solution. The brain needs to be trained to interpret the signal.²² The brain requires more training for a cochlear implant than for hearing aids because there has been more auditory deprivation (hearing aids frequently do not provide audibility of the full range of speech sounds when fit to a severe or profound hearing loss). An infant getting a cochlear implant at 12 months of age puts that child at an auditory age equivalent of 0 – 3 months. It is possible to close the gap with a lot of hard work. Parents who choose this path need to understand that they are making a huge commitment in time and effort to achieve the outcome they desire.²²

There are three companies that manufacturer cochlear implants including Advanced Bionics (advancedbionics.com), Cochlear Americas (www.cochlearamericas.com), and MedEl (www.medel.com).

Bone Anchored Implant (Baha)

If a child has a condition resulting in a permanent conductive hearing loss, the child may be a candidate for a Baha implant²⁶; however, children younger than 5 years of age (current FDA regulations) cannot be implanted. [In Europe, it is routine to successfully implant a child at 2 years of age or when skull bones are 2.5 mm thick]. A Baha can be worn on a softband until the implant surgery is scheduled.

A Baha consists of an external component (sound processor and abutment) and an internal component (small titanium implant). This provides an alternative pathway for sound to reach the brain, via bone conduction instead of air conduction. Many permanent conductive hearing losses fall in the range of a mild or moderate loss. The Baha can be fit on the softband as soon as the hearing loss has been diagnosed, reducing the auditory deprivation experienced by the child.

The Baha is the only FDA approved osseointegrated hearing device and is manufactured by Cochlear Bone Anchored solutions (formerly: Entific Medical Systems) (www.entific.com), a subsidiary of Cochlear Ltd. (www.cochlear.com).

FM Systems

FM systems can be used with hearing aids, cochlear implants and Baha devices. An FM system consists of a transmitter and a receiver. The person talking wears the FM transmitter, the FM receiver is attached to the child's personal hearing device. Speech can be transmitted for up to 300 feet. FM systems can be used on implants and provide a solution for distance and background noise²².

FM systems may be used anytime the infant is not within 3 feet of a parent or care giver to compensate for the distance. For example, when a child is in a bassinette and parents are fixing

dinner and talking about what they are doing (incidental auditory stimulation and learning opportunity). The FM system is frequently used to overcome automobile background noise when the infant is behind the driver in a car seat. FM systems are used on family outings, in preschool environments, field trips, and classrooms.

Phonak (www.phonak.com), is the industry leader in FM systems.

How can pediatricians facilitate this process?

Early identification and intervention of hearing loss leads to better speech, language and learning outcomes for children. Understanding the need for timely action, technology options, and therapy required (parent-training and child-centered) can assist the physician in providing appropriate family support. The goal is to insure that the listening, language, learning, and literacy needs of all children are met so each child can achieve their full academic potential.

Support family choice

- Provide motivating family support
- Be familiar with the basic types of technology that provide access to auditory information
- Work with a pediatric audiologist who routinely provides services to the birth to 3 population, not all audiologists have this expertise
- Understand the importance of parent-training provided by a speech/language pathologist for parents of children with hearing loss (birth to 3)

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