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both diagnostic evaluation and hearing instrument fitting in one facility, collaborative efforts among multiple programs and service providers are usually needed to ensure optimal management.

**Timely Referral and Initial Audiologic Assessment**

The Joint Committee on Infant Hearing (JCIH 2007) recommends that all infants who do not pass newborn hearing screening or subsequent rescreening begin appropriate audiologic and medical evaluations to confirm the presence of hearing loss before three months of age. In some screening models, infants who do not pass initially are re-screened within one month of hospital discharge; infants who do not pass are then referred for comprehensive evaluation. Other models provide comprehensive assessment immediately following referral from the hospital-based hearing screening. In either model it is important to establish protocols for prompt referral and comprehensive assessment. Unfortunately, it is not uncommon for us to see infants who failed the newborn screening only to be referred for multiple rescreenings that eventually resulted in a false negative outcome. In other cases the infant failed the re-screening but families were assured that the outcome was probably due to “debris in the ear canal or fluid in the middle ear.” Either outcome results in delayed identification and intervention. A critical component of any early hearing detection and intervention program (EHDI) is to ensure that information regarding a failed newborn screen is conveyed to families in a manner that increases the likelihood of the family returning for follow-up (JCIH 2007). Pediatricians and other service providers must be aware of the need for prompt referral to facilities where comprehensive assessment can be provided by pediatric audiologists who specialize in working with infants. Referrals made prior to three months of age, when the infant still sleeps several hours each day, will increase the likelihood of the infant sleeping naturally during the audiologic assessment and will often eliminate the need for sedation.

For the infant under six months of age, an essential goal of the initial comprehensive audiologic assessment is to obtain frequency-specific estimates of hearing thresholds for use as a starting point in hearing instrument fitting. Auditory brainstem response (ABR) evaluation using frequency-specific test stimuli (e.g. tone bursts) is useful in obtaining estimates of low, mid, and high frequency hearing sensitivity (Gorga, Reiland, Beauchaine and Jesteadt 1988; Stapells, Gravel and Martin 1995; Stapells 2000; Gorga et al. 2006). When hearing loss is evident, based on air-conducted test results, ABR assessment can be completed using bone-conducted stimuli. When the ABR is grossly abnormal or absent, it is essential that click-evoked ABR testing at a high intensity level be completed using both condensation and rarefaction stimuli to rule out auditory neuropathy/auditory dys-synchrony (AN/AD); (Rance 2005). The use of alternating polarity clicks alone will likely result in an incorrect diagnosis of profound hearing loss for infants with AN/AD.

Many of the infants seen in our clinic have already had one or more diagnostic ABRs before referral to our center. While some of these infants have undergone a comprehensive ABR evaluation that includes frequency-specific threshold estimation using air and bone-conducted stimuli, many will have had only an air-conducted ABR using click stimuli. Considering the variability known to occur in audiometric configurations it is essential that frequency-specific ABR measures be used to estimate thresholds for hearing instrument fitting. Audiologists who agree to perform diagnostic ABR evaluations on infants following referral from newborn hearing screening must have the ability to obtain frequency-specific thresholds using tone burst stimuli or to make referrals to a center where this can be provided. While an abnormal response using only click stimuli may confirm the presence of hearing loss, another test with frequency-specific stimuli will be needed before hearing aid fitting can be initiated. An incomplete initial study requiring a second test results in additional time, expense, and delays for the child and family.

Auditory Steady State Response (ASSR) is a newer electrophysiologic procedure being used in some clinics in addition to ABR during the comprehensive assessment. While recent studies have shown that both ABR and ASSR provide accurate frequency specific threshold information (Rance and Rickards 2002; Johnson and Brown 2005; Gorga et al. 2006), relatively few studies are available using ASSR on infants with hearing loss and, as of this writing, there is no way to determine if an infant has AN/AD if only ASSR is used. In addition, the criteria for determination of normal hearing are not as well defined for ASSR as they are with ABR. Care must be taken to avoid a diagnosis of hearing loss in a child with normal hearing when ASSR is used as the primary diagnostic tool (Johnson and Brown 2005). Clinicians involved with the comprehensive evaluation of hearing loss in infants should stay current with the latest re-
search on ASSR and other methods of evaluating hearing loss in infants. Although beyond the scope of this chapter, acoustic immittance measures and otoacoustic emissions are other essential components of the comprehensive audiologic test battery (see JCIH 2007).

Most infants under three months of age can be easily evaluated in natural sleep if families are prepared for the procedure ahead of time. Before arriving at the clinic, parents should be informed of the need to time the baby’s feeding prior to the appointment to increase the likelihood of the infant sleeping during the diagnostic assessment. Infants older than three months of age are likely to be awake several hours during the day and thus require sedation in order to complete the study. In our facility, ABRs on infants older than three months of age are often completed while the child is under sedation for radiologic procedures such as CT or MRI. Diagnostic ABR evaluations are also completed in the operating room in conjunction with other procedures such as placement of ventilation tubes or during cleft palate repair.

Otologic and Other Medical Examinations

Prior to hearing instrument fitting the infant should be seen for otologic examination and medical clearance. A comprehensive otologic evaluation by an otolaryngologist familiar with hearing loss in infants will include a complete medical history and ear examination (see also Chapter 5 in this volume). The otolaryngologist may also order radiologic studies including CT or MRI, laboratory studies, and electrocardiogram. Results obtained from radiologic studies in our clinic have resulted in the identification of a variety of problems including cochlear nerve deficiency, enlarged vestibular aqueducts, and cochlear malformations (Adunka et al. 2006; Buchman et al. 2006). These are all findings likely to have a significant effect on the treatment plan. For example, a child with enlarged vestibular aqueducts and the possibility of progressive hearing loss may require more frequent follow-up visits for reevaluation and counseling regarding prevention of head trauma. In cases of cochlear nerve deficiency, imaging studies may provide useful information to aid in decisions regarding cochlear implantation. The otolaryngologist may also suggest examination by an ophthalmologist, geneticist, or other specialists. At least one-third of children with hearing loss will have one or more challenges in addition to hearing loss that may affect audiologic management (Roush, Holcomb, Roush and Escolar 2004). A comprehensive evaluation by the geneticist may result in early identification of other medical problems requiring treatment. Some families may also find information provided by the geneticist helpful in understanding the etiology of their child’s hearing loss. During the weeks following initial diagnosis, learning more about the cause of their child’s hearing loss is of great importance to most families (Harrison and Roush 2001).

Good communication between the audiologist and otolaryngologist during the first year is essential not only during the initial assessment process but also as additional information about the infant’s hearing status is obtained. Changes in the child’s hearing thresholds, presence of otitis media, or severe-to-profound hearing loss with lack of benefit from amplification, are all reasons for a return visit to the otolaryngologist. Otitis media with effusion is highly prevalent in infants between six and eighteen months of age (Paradise et al. 1997) and if untreated may reduce the benefits of amplification. Communication between the audiologist, the otolaryngologist, and the family regarding management of middle ear problems increases the likelihood of optimal amplification. In addition to sharing reports of audiologic evaluations with the otolaryngologist, results should also be sent to the child’s primary care physician, a key individual who is likely to have an on-going relationship with the child and family. When kept well-informed about the child’s hearing status, the primary care physician can support the family and assist with audiologic recommendations and follow-up.

In a survey of parents of children with recently identified hearing loss, Harrison, Roush and Wallace (2003) reported that many families experience delays in fitting of amplification because of difficulties with scheduling appointments and/or the need for multiple appointments before hearing aid fitting is provided. In our center we have found that scheduling the infant to see an otolaryngologist on the same day as the comprehensive audiologic assessment reduces delays often associated with obtaining medical examination. If this is not possible, the audiologist should work with the otolaryngologist to facilitate timely access to medical review and clearance for hearing aids, so as not to unnecessarily delay the fitting of amplification.

Frequent visits are often necessary during the first few months following confirmation of hearing loss as the family returns for genetic evaluation, eye examination, radiologic studies, or other procedures. With good communication among clinic staff it is often possible to coordinate these appointments with return visits for ear impressions or earmold fitting.
Communicating with Families and Referral for Intervention Services

Sufficient time must be allotted on the day of diagnostic audiologic and otologic evaluations for discussing the results with the family and for taking ear impressions if the family is ready to proceed with amplification. It has been our experience that the majority of families choose to proceed with amplification immediately following comprehensive assessment; however, not all families respond to the diagnosis of hearing loss in the same way. Some families need more time before proceeding with amplification. It is important for audiologists and otolaryngologists to respect the family’s wishes with regard to the timeline for habilitation. We have found it helpful to offer parents the option of meeting with another family of an infant with similar degree of hearing loss. Several parents in our program have volunteered to meet with families of newly identified children to offer support and share information. In addition, parents in our state have organized regional parent support groups called HITCH-UP (Hearing Impaired Toddlers Have Unlimited Potential) that meet monthly to provide support and share information about strategies and resources families have found useful. Parents who attend these sessions report that the support group has been very beneficial to them as they learn about hearing loss and as their child adjusts to the use of hearing aids or cochlear implants.

A detailed review of habilitative options is beyond the scope of this chapter; however, it is important to keep in mind that comprehensive early intervention services are essential for infants with hearing loss; audiologists who work with infants must ensure that appropriate referrals are made. In North Carolina, following the confirmation of the hearing loss and with the family’s permission, results of the hearing evaluation are sent to a non-profit organization called Beginnings for Parents of Children who are Deaf or Hard of Hearing (www.beginningssvcs.com). Within a week after the diagnostic ABR, once hearing loss is confirmed, medical clearance for hearing aids has been received, and a return appointment for hearing instrument fitting is scheduled. In our clinic the goal is to schedule infants for hearing aid fitting approximately two weeks after the diagnostic ABR. Selection of hearing instruments for infants requires consideration of their unique needs. Electroacoustic flexibility is a key consideration when selecting hearing instruments since it is often necessary to proceed with limited information regarding the degree and configuration of hearing loss. With today’s digital hearing aids there are many instruments with multiple channels and the flexibility needed to fit a wide range of hearing losses. The size and style of the instrument are also factors that must be considered when selecting hearing aids for infants. Assuming the electroacoustic characteristics are appropriate, small, behind-the-ear instruments are preferred. Before ordering the device, it may be useful to
place a similar instrument behind the infant’s ear to ensure a good fit. There are many reasons why all-in-the-ear (ITE) style hearing aids are not appropriate for infants and young children, including the need to re-case hearing aids as the child grows, lack of FM compatibility, safety issues related to the hard acrylic casing in the ear canal, and the inability to use a loaner instrument when the child’s hearing aid is in need of repair. Other features that must be considered when working with infants include tamper resistant battery doors, a method to secure volume control settings, feedback management and FM compatibility. Tamper resistant doors must function in a way that prevents the infant from gaining access to the battery while allowing the hearing instrument to be easily turned on and off during insertion or removal from the infant’s ear. Ease of operation for the battery door is especially important for infants who will be in a day care setting with multiple caregivers. Protection of the volume control mechanism requires careful thought when providing amplification for infants. Although care is taken to adjust the hearing instrument using prescriptive formulas, there are times when a family may return home following a visit to the audiology clinic only to find that the infant is reacting negatively to loud sounds or that the instrument has excessive feedback that wasn’t obvious prior to the family’s departure from the clinic. In these instances and in cases where feedback occurs prior to the child’s next earmold fitting, it is useful for parents to have the option of reducing the volume control on a short-term basis until they can return to the audiology clinic for corrective action. In previous generations of hearing instruments this was accomplished using a volume control cover; however, in newer hearing instruments it is possible to deactivate the volume control. While this may be a useful feature in certain situations, when working with infants it is necessary to prevent the child from changing the volume control setting while allowing parents access. Feedback management should be accomplished using algorithms that reduce feedback with minimal reduction in the high frequency gain of the instrument.

Some features available in today’s hearing instruments, such as the ability to switch between omni and directional microphones and availability of multiple programs, may not be needed initially but may be useful as the child gets older. It is also important to keep in mind that some “automatic” features that may be appropriate for use with older children and adults, such as adaptive microphone technology and certain noise reduction algorithms, may be contraindicated for use with young children. While adults are able to judge when a feature is detrimental in a given listening situation, a young child is unable to control the selective use of these features. In addition, the listening behaviors and requirements of infants and toddlers are significantly different from those of adults (Stelmachowicz, Pittman, Hoover and Lewis 2001; Pittman, Stelmachowicz, Lewis and Hoover 2003). For example, the importance of “overhearing speech” to the communication development of infants and young children makes a hearing instrument with an omnidirectional microphone the best choice for infants and young children in most listening situations. For this reason, while many hearing instruments have multiple programs including some that sample sound in the environment and automatically switch between directional and omnidirectional modes, a single, basic omnidirectional program is usually sufficient during the initial stage of hearing instrument fitting for infants. Additionally, considerations in the selection and fitting of hearing aids for infants and children can be found in the American Academy of Audiology Pediatric Amplification Protocol (AAA 2003).

### Hearing Instrument Fitting and Verification

To ensure that speech is both audible and comfortable for the infant or child, hearing instrument performance must be verified and compared to a specific prescriptive fitting formula, e.g., DSL[i/o] v4.1 (Cornelise, Seewald and Jamieson 1995; Seewald 1995) or DSLv5 (Bagatto et al. 2005; Scollie et al. 2005; Seewald, Moodie, Scollie and Bagatto 2005). Although use of the manufacturer’s fitting software provides a good starting point for “pre-fitting” of hearing aids based on a prescriptive fitting formula, we often find that the actual performance of the hearing instrument does not match prescriptive targets for gain and output when verification measures are made. The manufacturer’s fitting software may allow the audiologist to select a prescriptive fitting formula (e.g. DSLv5, NAL NL-1), but at the present time there is significant variability among the various hearing instrument manufacturers. In a recent study conducted at the National Centre for Audiology in Canada, Seewald and colleagues (Seewald, Drake, Scollie, Moodie and Johnson 2006) compared five different manufacturer’s implementation of the same prescriptive method for varying degrees of hearing loss. They found that for the same hearing loss the recommended gain for average speech inputs varied by as much as 21dB and the prescribed out-
Hearing Instrument Orientation

Once RECDs have been measured and the hearing instrument performance has been verified, the audiologist provides the family with an orientation to the use of the hearing instruments. Family members are given an opportunity to practice inserting and removing the hearing aid and are provided instruction regarding care of the instrument and troubleshooting techniques. They are given a listening stethoset, battery tester, air blower, dehumidifier and retention strap. The family is also advised of the need for frequent earmold remakes during the first year. It is not unusual for an infant fitted with hearing aids at two months of age to require six to eight sets of earmolds in the first year.

Time spent educating the family regarding hearing aid retention issues, troubleshooting and repair, and follow-up, will not only help the family get off to a good start but will reduce the number of phone calls the audiologist receives during the first few weeks following hearing instrument fitting. A return appointment is scheduled three to four weeks following the initial hearing instrument fitting. During that visit the audiologist answers questions, checks on the progress of early intervention services, and discusses with the family the infant's auditory behavior. Due to the rapid growth of the ear canal in the first few months of life, it may also be necessary to take ear impressions again during this visit.

Families need to know that it is not uncommon for infants to learn to remove their hearing aids and of the ease with which they are able to get them into their mouths. While this is usually not an issue at two to three months of age, many families report problems with removal of hearing aids beginning at four to five months of age. Options for retaining the instruments include retention straps, “Huggie Aids” and double-side tape. If these techniques are unsuccessful, another option our families have found helpful on a short-term basis is the use of a lightweight cap that ties under the chin. While this is not an ideal solution it has been effective in cases where the parent, despite their best efforts, is unable to keep the hearing aids in place. With this short-term strategy and persistence by the family, most infants will successfully adapt to full-time hearing aid use within a few weeks.

Keeping hearing instruments in good working condition is always a challenge. This is especially true with infants and toddlers, who inevitably will have sticky fingers, play in the sand, etc. Clinics need to maintain a supply of loaner hearing instruments so that children have consistent use of binaural amplification when one instrument is in need of repair. If institutional funding is not available for this purpose it is often possible to find service organizations that are willing to purchase loaner instruments. The number of infants seen in our clinic has...
increased each year since the implementation of universal newborn hearing screening. This has resulted not only in additional appointments for diagnostic evaluations, but appointments needed for ear impressions and earmold fitting as well as time needed for troubleshooting the inevitable equipment problems that arise. In a busy practice it may be necessary to hire an assistant to help with minor equipment repairs, shipping, and the logistics of maintaining loaner hearing aids.

**Behavioral Audiometry**

Once the infant is sitting up and has reasonable head control, behavioral audiometry using visual reinforcement audiometry (VRA) is attempted. As with other developmental tasks, some infants are able to perform VRA at a younger age than others; however, most should be able to successfully perform the VRA procedure by 6–8 months developmental age (Widen 1990; Widen et al. 2000; Gravel and Wallace 2000). For a comprehensive clinical VRA protocol see Gravel (2000).

We recently reviewed data from the first 70 infants with bilateral sensory hearing loss fitted with hearing aids following referral from newborn hearing screening in our infant hearing program. For these infants, the median age at ABR testing was 2.6 months, the median age at hearing aid fitting was 3.9 months, and the median age when a frequency-specific behavioral audiogram was available for each ear (250–4000 Hz) was 8.5 months. Thus, frequency-specific estimates of hearing thresholds obtained from ABR allowed us to fit most infants with hearing aids at least four months earlier than would have been possible had we waited for thresholds from behavioral testing with visual reinforcement audiometry. It is important to remember, however, that electrophysiologic measures provide an estimate of behavioral thresholds and that actual measurement of hearing threshold levels should be completed as soon as possible to ensure accuracy in adjusting the hearing instruments. In addition, a complete audiogram obtained using VRA will provide the audiologist with additional information regarding the slope and configuration of the hearing loss. Finally, some children will have progressive hearing loss in the early months of life making behavioral confirmation of thresholds essential if the hearing instruments are to be fitted appropriately. Infants who are already accustomed to wearing hearing aids will tolerate the use of insert earphones attached to their custom earmolds more readily than if standard foam inserts are used. This allows us to obtain frequency and ear-specific audiometric thresholds as early as possible.

For young infants, more than one test session is generally required to obtain a complete audiogram (250–4000 Hz) for each ear. When updated hearing thresholds are obtained, the hearing instruments are re-adjusted to provide the best match to prescriptive targets for gain and output. It is also important to monitor the baby’s middle ear status using acoustic immittance measures during each follow-up visit and to schedule a return visit to the pediatrician or otolaryngologist for middle ear management when indicated.

While the VRA procedure is usually recommended for babies between the developmental ages of approximately six months to 2½ years, young children become increasingly more challenging to work with in the second year of life. For this reason, follow-up appointments to attempt behavioral assessment should be scheduled as soon as the baby appears to be developmentally able to perform the task. Once a complete audiogram has been obtained for each ear, it is possible to monitor changes in the child’s hearing thresholds during subsequent visits.

Sufficient time and staff are needed to perform VRA accurately. Although the VRA procedure can be accomplished using the parent as the test assistant, with young infants it is more efficient to have an experienced assistant in the sound booth while the baby sits in the parent’s lap. The likelihood of obtaining reliable thresholds in a young infant will also be increased by an appropriate room set up with the visual reinforcement unit located 90 degrees to one side of the infant. Once the infant understands the procedure it is usually possible to present the signal to either ear and condition a head turn response toward the reinforcer, even if the signal is presented to the opposite ear (see Gravel and Hood 1999).

Parental acceptance of the child’s hearing loss is always an important factor to be considered when working with young infants. Prior to universal newborn hearing screening it was usually the parent who first suspected hearing loss after observing their child’s lack of responsiveness to sound or delays in language development. With many infants now identified at birth, parents are required to accept the diagnosis of hearing loss with little or no evidence from direct observation. For most infants with severe or profound hearing loss, it is obvious to family members that the infant does not respond well to sound without amplification. For infants with milder degrees of hearing loss parents sometimes proceed with amplification initially but later question the need for hearing aid use as the baby becomes older and
responds to speech and environmental sounds without amplification. A comparison of unaided and aided detection thresholds in sound field using warbled pure tones or narrow band noise with the family in the sound booth can be a useful way of demonstrating the child’s improved detection of soft sounds. Another useful strategy is to have one parent sit with the audiologist in the control booth while the other parent is in the room with the infant. Family members are often impressed with the reliability of their infant’s responses during VRA assessment even at a young age. In addition, this provides an opportunity for the audiologist to educate the family about the hearing assessment process.

**FM systems, Cochlear Implants, and Follow-up Care**

While in past years FM systems were used primarily in educational settings, many families are now choosing to use personal FM systems to improve the signal-to-noise ratio in the home environment and during day-to-day activities such as riding in the car or going to a grocery store or playground. A good time to introduce FM systems is often around the first birthday when the child begins to walk and there is increasing distance between the parent and child. It is important that consistent full-time use of hearing aids be established prior to the introduction of an FM system. In addition, the appropriate use of FM must be demonstrated to the family.

Frequent return visits to the audiologist and otolaryngologist are needed in the first several months of life to define accurately the type and degree of hearing loss and to complete a full otologic work up. Once the medical assessment is completed, an audiogram has been obtained for each ear, and the infant has successfully adjusted to full-time hearing aid use, the frequency of return visits for follow-up can be decreased to approximately once every three months or when needed for hearing aid repair or earmold re-makes.

The management of children with severe and profound hearing loss requires special consideration. It is often incorrectly assumed that lack of response from ABR evaluation means the child has no residual hearing and will not receive benefit from amplification. While lack of response on current electrophysiologic testing (ABR or ASSR) usually indicates at least a severe hearing loss, there may be significant residual hearing particularly in the low and mid frequencies that will allow the infant to receive benefit from amplification in the early months of life, even if later the child is determined to be a candidate for cochlear implantation. Estimates of hearing thresholds based on electrophysiologic measures should be made for purposes of initial hearing instrument fitting, followed by confirmation of thresholds using VRA with insert earphones as soon as the child is able to perform the task. Families should be informed of the limitations of current electrophysiologic tests and made aware that behavioral audiometry with VRA is necessary to determine the degree and configuration of their child’s residual hearing.

It is vitally important that children with severe and profound hearing loss receive careful monitoring during the first few months following hearing instrument fitting to determine if communication goals are being met. If a child has been fitted with appropriate amplification at an early age, is receiving early intervention services, and is not making consistent progress in meeting communication goals with amplification during the first few months of life, referral to a cochlear implant team should be offered to the family. It is now common for children with profound hearing loss to receive cochlear implants by one year of age, but good communication between the family, audiologist, early intervention specialists, and otolaryngologist is needed to avoid delays in referral for consideration of cochlear implantation when amplification is not sufficient.

**Summary**

Successful management of hearing loss in infancy requires a team approach that involves the family in collaboration with the audiologist and other service providers. Essential components include prompt and accurate diagnosis followed by intervention based on the family’s preferences. Hearing levels must be carefully predicted using physiologic assessment procedures and initial fitting of amplification must be accomplished using an electroacoustic hearing aid selection fitting strategy appropriate for infants. Although accurate diagnosis and initial hearing instrument fitting are essential first steps in the habilitation of the infant with hearing loss, it is important to remember that a continuum of care is needed during the first year to ensure the best possible developmental outcomes. Follow-up visits will be needed to make new earmolds, to monitor middle ear status, to obtain ear and frequency-specific thresholds using behavioral audiometry, and to readjust hearing aids to en-
sure an optimal match to prescriptive targets. Finally, it must be recognized that in addition to providing technical services, the pediatric audiologist plays a key role in counselling, providing emotional support to families, connecting them with other families, and facilitating communication with service providers.

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**References**


