Comprehensive Audiological Management of Hearing Loss in Children, Including Mild and Unilateral Hearing Loss

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KEYWORDS

• Children with hearing loss • Amplification candidacy • Device selection
• Hearing aid verification • Outcome validation

KEY POINTS

- Timely diagnosis of childhood hearing loss should include prompt referrals (eg, medical, language) and recommendations for audiological intervention based on type, degree, and configuration of hearing loss.
- If a child is a candidate for amplification, the audiologist should work with the family to select a hearing technology, with consideration of device style, advanced features, and compatibility with assistive technology.
- Ongoing verification of amplification ensures that speech sounds amplified by the hearing aid are audible but not too loud, especially as a child grows and ear canal acoustics change.
- Audiologists should validate hearing aids using parental questionnaires and aided speech perception measures to assess the benefit of amplification for auditory and speech-language development.

Once hearing loss (HL) is confirmed, referrals for medical and language intervention should be made immediately. A full medical workup by a pediatric otolaryngologist is indicated anytime HL is diagnosed. This evaluation helps rule out transient conductive issues, ensures that a child's ears are healthy enough for hearing aids, and could reveal related conditions. The findings of a medical examination complement audiometric findings and may influence the technology recommendations. Specific recommendations for audiological intervention will depend on the type, degree, and

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configuration of HL. If an HL is permanent or unlikely to be resolved through medical management, amplification should be considered as part of the intervention whether the loss is sensorineural, conductive, or mixed. Any degree of HL can negatively impact spoken language development,[1,2] so amplification should be considered if a child has thresholds that are more than the normal levels for any frequencies that are important for understanding speech. Unilateral HL can impact access to binaural cues that are important for localization of sound and listening in noise, so children with mild to severe unilateral HL should also be considered candidates for amplification. Conventional audiometric testing in dB HL (decible hearing level) can produce threshold values that are enhanced by a child's small ear canal size, particularly for insert earphones.[3] In cases in which infants or children with small ear canals present with mild degrees of HL, audiologists may convert the dB HL audiogram to dB sound pressure level (SPL) to evaluate the effects of threshold elevation on speech audibility and weigh the potential audibility benefits of providing hearing aids.[4] Children with severe or profound degrees of HL might receive limited benefit from amplification and eventually be evaluated for cochlear implant candidacy.[5] However, hearing aids provided before cochlear implantation have demonstrated benefits for postimplantation outcomes,[6] so auditory stimulation via hearing aids is often recommended for children with severe to profound degrees of HL, even when the potential for long-term benefits are low.

SELECTION OF HEARING DEVICES

Once a child is determined to be a candidate for amplification, the audiologist and family work together to select hearing technology that is audiologically appropriate and flexible enough to accommodate a child's changing needs over time. Several clinical and nonclinical factors will influence what hearing aid they select.

STYLE

The most common style of hearing aid for infants and children is a behind-the-ear (BTE) hearing aid, coupled with an earmold. Different sizes of BTE models provide the audiologist with a wide range of power options to produce appropriate amplification for HL ranging from mild to severe. BTE devices can be reprogrammed to accommodate some progression in hearing sensitivity. When the pinna is substantial enough to support the weight and size of a BTE hearing aid, it should be considered the first choice for infants and children. As a child's ear grows, the earmold can be remade to ensure an adequate fit without having to replace the device. For older children and teens with normal low-frequency hearing, the audiologist and family might choose to pursue an open-fit BTE or receiver-in-the-ear device to reduce occlusion.

EARMOLDS

An earmold is a custom-fitted mold of the pinna and ear canal that connects with the BTE hearing aid via soft tubing and delivers amplified sound to the ear. Earmolds require that a set of ear impressions be taken and mailed (or scanned and sent) to an earmold fabrication laboratory. The audiologist makes these impressions by mixing and placing fast-setting material into the canal and concha. This process can take place under sedation if a child undergoes a sedated auditory brainstem response (ABR) to establish their level of HL. Earmolds come in several materials, and families are able to choose from a wide range of colors and designs. Soft earmold materials,

such as silicone, are currently the standard of care for pediatric patients, because they ensure a comfortable fit in a wide variety of environments and can be easily modified. Harder earmold materials like acrylic and vinyl are longer lasting than soft materials and can be a good option in cases of allergy to silicone.

Without a tightly fitting earmold, sound can leak out of the ear canal and reenter the device's microphones. This leakage creates acoustic feedback audible as a whistling sound to those in close proximity to a child with hearing aids. Excessive feedback should be addressed, because it indicates poor acoustic coupling that can lead to insufficient amplification. In the first year of life, a family can expect several visits to remake and fit new earmolds. If a child's ear has grown such that an earmold needs to be replaced, the audiologist should remeasure the child's ear canal acoustics using the real-ear-to-coupler-difference (RECD) procedure.

TECHNOLOGY LEVELS

Hearing aid manufacturers often market devices at several technological levels (at progressively higher cost), ranging from introductory devices to premium technology devices. Although the connotation of higher cost is often higher performance, research on premium hearing technology does not bear this out. The additional cost of premium-level technology is often not justified based on available research in children. **Table 1** contains a summary of available technology and the most recent evidence-based recommendations for their use.[7]

COMPATIBILITY

Children with HL often rely on assistive technology such as remote-microphone systems to supplement their amplification in acoustically adverse environments like classrooms. Families and audiologists must think ahead about features a child might need during the lifespan of their device (typically 3–5 years). For example, some hearing aids might not be compatible with the remote-microphone systems used in school settings. Although school districts are required to find solutions in such cases, careful selection at the time of fitting can prevent long periods of poor auditory access in the classroom due to incompatibility.

HEARING AID FUNDING

A final important factor in selecting a hearing device for a young child is funding. At the time of this writing, 17 US states have passed legislation that makes pediatric hearing aid insurance coverage mandatory; however, even in states with coverage mandates, there are exceptions. Other states have no such mandate and families can face steep out-of-pocket costs that can be a barrier to timely amplification.[8] State Medicaid programs cover amplification for children whose families meet qualification standards but place a heavy administrative burden on accessing these benefits or have limited funding. Families must use a provider who has enrolled in their state's Medicaid program and agreed to accept Medicaid rates for amplification services, which may force some providers not to provide care due to poor reimbursement rates. To fill gaps in pediatric hearing aid funding, hearing professionals can compile a reference of charitable and alternative funding mechanisms available within their state.

VERIFICATION

Once the appropriate device is selected, the hearing aid must be programmed and fit to the individual child. The fitting processes include teaching the child and their family

Table 1
Advanced hearing aid features and their recommendation status in current pediatric amplification guidelines[7]

Advanced Feature	Description	Considerations for Pediatric Use
Directional microphones	Digital processing strategies based on positional cues relative to hearing aid microphones meant to emphasize the amplification of sounds from the front	Careful consideration should be taken before activating directional microphones because of the importance of listening through overhearing or incidental learning for children. Automatic directionality is preferable to manual directionality
Feedback suppression	Algorithms that detect acoustic feedback and limit gain for high frequencies to resolve it	Feedback suppression should be activated in pediatric devices
Amplitude compression	Varies amplification based on the loudness of the incoming signal. Compression promotes the use of a listener's full range of hearing	Compression improves audibility for soft speech and maintains comfort for loud inputs

Data from American Academy of Audiology (2013). Clinical practice guidelines: Pediatric amplification. Reston, VA.

about daily use and care of the devices, as well as hearing aid verification. The goal of hearing aid verification is to ensure that speech sounds amplified by a hearing aid are audible, but not too loud, using objective acoustic measurements. If speech sounds are not audible, children who are learning spoken language might be at risk for receiving inadequate audibility and experiencing poorer language outcomes.[4,9] Audiologists must assess hearing aid fit relative to pediatric prescriptive targets and determine the audibility of speech at the initial fitting appointment and at regular intervals as the child grows (less than 1 year old: every 3 months; less than 3 years old: every 6 months; older than 3 years: every year). Verification should also be performed after any changes to a child's hearing thresholds, hearing aids, or earmolds. Verifying hearing aids helps ensure that speech remains audible as the child grows and ear canal acoustics change.

Audiologists perform hearing aid verification using a probe microphone system. This system evaluates sound levels produced by a hearing aid in response to various levels of speech input by measuring hearing aid output (**Fig. 1**). In children who can sit for multiple measurements, hearing aid output is measured directly in the ear canal using a small flexible probe tube microphone placed near the eardrum just beyond the hearing aid. Alternatively, clinicians can record the child's RECD, which is a quick measurement of a child's ear canal acoustics, and use those values across multiple measures to verify hearing aid output within a hearing aid test box. Age-related average values can also be used if a child's RECDs cannot be measured; however, average RECDs lack the specificity of individually measured values.

Performing probe microphone measurements allows audiologists to assess if hearing aid output meets levels prescribed by pediatric-fitting formulas (eg, desired sensation level) for soft, average, and loud speech inputs (eg, 55–75 dB SPL). These evidence-based formulas suggest optimal hearing aid output levels based on a child's

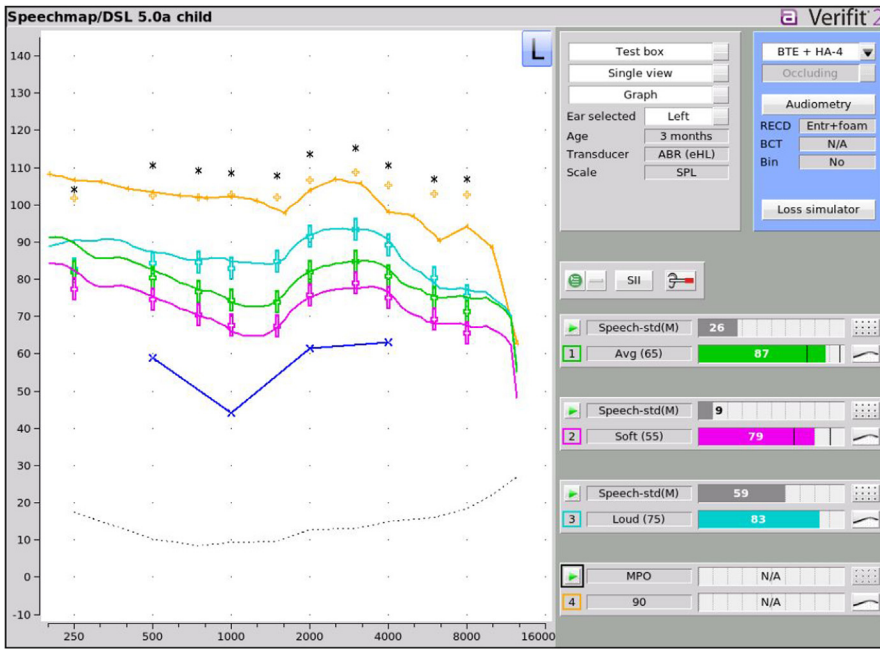

Fig. 1. A sound pressure level (SPL) o-gram for a child with mild to moderate hearing loss. Blue X symbols connected with a blue line represent the audiometric thresholds in dB SPL for the left ear. Crosses represent prescriptive targets for soft (pink), average (green), and loud (teal) input levels for speech. The orange line represents the maximum power output of the hearing aid. The unaided speech intelligibility indices (SIIs) for each input level are shown as gray bars in the legend on the right, and the aided SII for each input level is represented by the corresponding colored bar. The comparison of unaided and aided SIIs shows the change in audibility with amplification for each input level.

age and hearing levels. Fitting output to these targets can help achieve consistent speech audibility.[10] In addition, audiologists should ensure that the maximum power output of the hearing aid is not uncomfortably loud. Furthermore, advanced hearing aid features (see **Table 1**) can be verified using the probe microphone system or the coupler. Verification of these features should occur if these features are activated to avoid negative impact on audibility.

Clinicians also determine aided audibility of speech by looking at the speech intelligibility index (SII).[11] The SII quantifies the proportion of speech that is audible and useable by the listener, ranging from no access (SII = 0) to complete access to speech (SII = 1). The aided SII is calculated by summing the sensation level (ie, difference in hearing aid output levels relative to child's unaided hearing levels) of frequency bands that are weighted by the amount of speech information that each band provides. Although aided audibility is not a direct measure of speech understanding, it reflects the weighted proportion of speech signals that is audible. On average, children with better aided audibility have better language[12] and speech recognition abilities[13] than those with poorer audibility.

Measuring aided audibility helps inform clinicians, parents, and professionals of a child's strengths and challenges related to accessing speech (eg, ability to hear high-frequency phonemes like /s/; access to soft, distant speech). Monitoring is

particularly important for children with progressive HL, because hearing aid gain might need to be adjusted more frequently. If a child's SII is low or if the hearing aids fail to meet targets, clinicians should consider changes to a child's intervention that increase audibility, such as hearing aid reprogramming to meet prescriptive targets, fitting more powerful hearing aids, or cochlear implantation.

HEARING AID VALIDATION

In addition to hearing aid verification, progress with amplification needs to be documented through validation. Validation confirms that a child's communication needs are being met. Two types of validation are commonly recommended for children: parental questionnaires and aided speech perception assessment. Both provide important information about how a child is performing with hearing aids.[7]

PARENT QUESTIONNAIRES

Parent-report questionnaires can inform us about children's development in audition and spoken communication, both preintervention and postintervention. These questionnaires cover infancy through adolescence, although they are typically used in the birth to 3-year age range, when children are unable to self-report. Parents can complete questionnaires on their own or via an interview with a clinician. The LittlEARS[14] and the Parents' Evaluation of Aural/Oral Performance of Children (PEACH)[15] are two examples of valid and reliable questionnaires that ask parents to assess their child's functional auditory skills (eg, "Does your child react to his/her name?"). Repeated administration of these questionnaires can help clinicians track auditory skill development over time (see Ref.[16] for a review of parent-report tools).

AIDED SPEECH PERCEPTION ASSESSMENT

Aided speech perception assessment is another important validation tool that tells us how a child uses amplification to support listening and spoken language. Speech awareness can be assessed during infancy. Starting around 18 months, most children can participate in aided detection or discrimination tasks. The Ling Six Sound Test[17] is a commonly used assessment of a child's ability to detect or discriminate specific speech sounds that cover the long-term average speech spectrum. Speech recognition can be assessed with familiar words or sentences. If children are limited in their language production skills, they can be encouraged to point to real objects or pictures. Once a child has achieved ceiling levels on speech recognition in quiet, background noise can be added to the testing scenario. The addition of background noise offers a more ecologically valid approach to speech recognition, compared with listening in quiet.

Poor speech recognition performance can be due to several factors. Language and cognitive abilities can affect speech recognition, so children with cognitive-linguistic delays might have lower scores on a speech perception test than children who have stronger skills in these areas. Auditory access can also impact speech perception performance. If a child is demonstrating very low scores on an aided speech recognition test, and the test has been deemed appropriate for the child's language and cognitive level, the audiologist should use probe microphone measures to verify the audibility provided by the hearing aid. Because children show wide variation in speech perception skills, conducting assessments at each audiology appointment can assist in understanding the development of auditory skills in an individual child.

AIDED SOUND FIELD THRESHOLDS OR FUNCTIONAL GAIN

Another option for validating hearing aid fittings is to measure a listener's audiometric thresholds through a loudspeaker while wearing the hearing aids. This approach, known as aided sound field thresholds or functional gain, can tell us whether amplification leads to improvements in audiometric thresholds compared with an unaided audiogram. Although it might seem intuitive to validate amplification using aided sound field thresholds, there are numerous limitations to this approach that preclude the use of this procedure in contemporary practice. The signal processing in hearing aids affects the perception of the tones we use during an aided audiogram, leading to responses that are not reflective of speech audibility. Furthermore, being able to detect very soft sounds in a sound booth does not fully explain how a child is able to understand conversational level speech in real-life situations. The only time that aided sound field testing is appropriate for validation is for children who use bone conduction devices or cochlear implants. Both forms of hearing technology bypass the typical air conduction route for hearing and, therefore, preclude the measurement of aided speech audibility.

IMPACT OF HEARING AID USE ON LANGUAGE ACQUISITION

A child with typical hearing is expected to show steady growth in language skills over time, making 1 year of language growth over 12 months. Young children with HL can start off with delays compared with children with typical hearing, so they need to show faster language growth (more than 1 year of language growth over 12 months) to close that gap. For families that choose a listening and spoken language approach, hearing aids are a tool for closing that gap. A recent longitudinal study of 317 children with mild to severe HL found that increased hearing aid use had a positive effect on language growth rates. That is, children who wore hearing aids more often throughout the day displayed steeper change in language scores over time, whereas children who wore hearing aids less showed a flat trajectory.[9] More specifically, children who wore hearing aids 10 or more hours a day made more than a year's worth of language gains in a year's time, essentially closing the gap between themselves and children with typical hearing. In contrast, children who wore their hearing aids for less than 10 hours a day showed no change in their rate of language growth (ie, the gap between their language skills compared with average performance for same-aged children with typical hearing remained the same over time). Furthermore, amount of daily hearing aid use predicts language outcomes for children with HL, regardless of the degree of HL. In other words, children with mild HL showed as much benefit from hearing aids as children with severe HL.[18] These results provide strong evidence for the importance of consistent hearing aid use for achieving maximum benefits from auditory stimulation, particularly during important periods of early brain development.

DISCLOSURE

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CLINICS CARE POINTS

- Accurate and timely diagnosis of HL sets the stage for informed management recommendations, including amplification, if appropriate.

- Selection of hearing devices by the audiologist, child, and family should account for the individual child's age, audiological profile, and educational needs.
- Verification and validation of the fitted devices ensure that the child has access to audible speech of a variety of input levels and that the devices are providing benefit via auditory and speech perception outcomes.
- Consistent use of well-fit hearing aids facilitates auditory access, which leads to improved outcomes for children with HL.

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