Audiologic Assessment and Management for Infants with Hearing Loss and Other Disabilities: Science, Clinical Practice and Creativity

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Introduction

Audiologists play a crucial role in early detection, diagnosis and intervention in infants and children with hearing deficits. A primary goal of their work is to maximize the use of the child’s residual auditory capacity through the fitting of amplification in order to facilitate the development of spoken language. This, in turn, can have a profound effect on the development of various aspects of children’s knowledge and abilities to interact with others in a meaningful way. To accomplish this goal, audiologists have the responsibility to use current technologies and to apply best practice throughout the diagnostic and amplification fitting process.

The need to provide a comprehensive and valid assessment of infants who have hearing loss is critical. In developing clinical assessment protocols, it is important to account for the fact that many infants and young children who are found to have hearing loss will have other coexisting conditions. For example, roughly one in every eight babies is born prematurely. These infants are at high risk of having hearing loss as well as other disabilities. Additional factors, such as the child’s health, cognitive status and functional needs will influence the approach taken in both audiologic assessment and intervention. Consequently, professionals have the responsibility to determine the type, degree and configuration of hearing loss of these infants in an accurate way, so that appropriate counseling with the parents, planning, fitting of hearing instruments, and communication development programming, can proceed in a timely manner. The following case study is presented to illustrate how the approach to audiologic assessment and intervention needed to be modified to work more effectively with an infant with multiple disabilities.

Birth History

GM and her twin sister were born prematurely at a gestational age of 29 weeks. She was 1250 grams at birth, had an APGAR of 5/7, perinatal asphyxia, respiratory problems and was in mechanical respiratory assistance for 15 days. In addition, GM was diagnosed to have hyperbilirubinemia and received phototherapy for this condition. She also experienced brain haemorrhage ten days following her birth. In total, GM spent 72 days in the neonatal intensive care unit (NICU) prior to discharge.

Audiometric Assessment

The most comprehensive audiological assessment has to yield information about the integrity of the auditory pathway, from the periphery through the central auditory system. Each measure in the test battery provides a unique piece of information about the status of the auditory system. It is only through such comprehensive assessment that the type, degree and configuration of a child’s hearing loss can be accurately determined.

GM underwent hearing screening when she was discharged from NICU. The first distortion product otoacoustic emission (OAE) results were negative and were positive for her twin sister. GM was referred for a second OAE screening one month later. The results of the second screening were again negative. Consequently, she was scheduled for a complete audiological assessment. At the same time, GM was referred to an ophthalmologist for an assessment of
Table 1. Air conduction tone-burst and click evoked ABR thresholds, in dB nHL, obtained at eight months of age.

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<thead>
<tr>
<th>Frequency (Hz)</th>
<th>500</th>
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<th>4000</th>
<th>Click ABR</th>
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her visual system. On the basis of this testing, GM was diagnosed to have bilateral hypoplasia of the optic nerve, nystagmus and convergent strabismus. The diagnosis of her visual impairment delayed the audiological assessment due to the resulting anxiety and stress that diagnosis of visual impairment generated in GM’s parents. As GM was considered high risk at birth, she was also given a neurological evaluation. No neurological difficulties were found.

At the age of eight months, GM was seen for a complete audiological assessment, beginning with auditory brainstem response (ABR) testing with sedation. The air conduction ABR thresholds obtained with both click and frequency specific tone burst stimuli are shown in table 1. These results are consistent with a moderate-to-severe hearing loss for both ears.

Click evoked ABR thresholds provide important information regarding the integrity of VIIIth nerve and brainstem auditory pathways, but they do not provide valid information concerning the configuration of the hearing loss as a function of frequency. For this reason, it is necessary to include frequency-specific ABR threshold measures in the audiological assessment with infants. In addition, when middle ear effusion is present, the degree of hearing sensory component of the hearing loss can be over-estimated. Therefore, it is important to take into consideration the middle ear status before interpretation of the ABR test results is made.

To this end, acoustic immittance testing was performed at the time of this initial audiologic assessment. The findings showed normal middle ear pressure and compliance with absent acoustic reflexes bilaterally.

Since children with multiple disabilities offer many different challenges, the importance of understanding and accounting for the complexity of each child's circumstances is crucial. Therefore, before proceeding with conditioned visual reinforcement audiology (VRA), GM’s ophthalmologist was consulted to learn more about her visual capabilities. He reported that she was unable to see further than 1.5 meters, and could only see in the lower quadrants. In addition, the ophthalmologist noted that GM would more easily see objects that are yellow and images with high contrast. Consequently, the VRA system set-up was adapted to GM’s capabilities, using only the two lower reinforcers. In addition, yellow toys were used.

Behavioral air and bone conduction thresholds were obtained across four sequential test sessions using insert earphone transducers and were consistent with a moderate sensory hearing loss in both ears (see table 2).

Data from Gravel and Wallace (2000) demonstrate that when assessing the hearing sensitivity of an infant born prematurely, it is important to use the infant’s corrected age, rather the chronological age, in predicting whether or not the infant might be conditioned successfully for VRA testing. In this case GM’s parents were pleased that their daughter could perform the test despite her visual impairment. On the basis of the complete audiological assessment, including both electrophysiological and behavioral evaluation procedures, the presence of a moderate sensory hearing loss bilaterally was confirmed.

### Hearing Instrument Fitting

Children with both hearing and visual impairments will likely have to rely more heavily on their hearing than those with hearing impairment alone. Further, amplification will be required to facilitate and enhance physical orientation and mobility skills – abilities that are essential for development. Thus, in cases such as the one reported here, it is even more critical that amplification be fitted with the greatest accuracy possible and that consistency in the auditory signals the child receives through amplification be maintained over time. This, of course, requires that the child’s hearing status, as well as the performance
Table 2. Air conduction VRA thresholds (dB HL) as a function of frequency (Hz), obtained with insert earphones at eight months of age.

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Table 3. RECD values (in dB) as a function of frequency (in Hz), for the left and right ears obtained at eight months of age.

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of hearing instruments, be monitored on a regular and frequent basis.

The measurement of the real-ear-to-coupler difference (RECD) is recommended as an important first step in pediatric hearing instrument fitting (Moodie, Seewald and Sinclair 1994; American Academy of Audiology 2003; see also Munro, Chapter Five in this volume). Measurements of GM’s RECD values were also obtained for both ears using an insert earphone transducer coupled to her custom earmolds. The RECD values obtained with GM at eight months of age are shown for each ear, as a function of frequency, in table 3. The values obtained were consistent with what might be expected for an eight-month-old infant. Note that, in the higher frequencies, the levels of sound that were measured within this infant’s ear canals exceeded the levels of sound measured in the coupler by as much as 16 dB. This important information was subsequently used to select the desired hearing instrument amplification characteristics for GM. In this way, the amplification that was selected for GM was individualized, by accounting for the unique acoustical characteristics of her occluded ear canals.

The Desired Sensation Level Method – DSL® version 4.1 formula (Seewald et al. 1997) was used to prescribe hearing instrument gain and output limiting characteristics for GM. The use of a systematic evidence-based approach that incorporates age-dependent variables into the computations is preferred when working with infants and young children (AAA 2003). Frequency-specific loudness discomfort levels (LDLs) should be obtained when children are old enough to provide reliable responses. However, when working with infants and young children, the DSL[i/o] algorithm provides predicted values for the purposes of electroacoustic fitting.

Table 4 shows DSL target values for the prescribed 2cc coupler gain characteristics across frequencies. The values shown in table 4 have been calculated using the infant’s individual RECD values as a function of frequency. The DSL target 2cc coupler output sound pressure level (OSPL) characteristics for GM are shown in table 5. These values were also calculated by the DSL® software system using GM’s measured RECD values as a function of frequency.

Verification

The measures performed during this step of the hearing instrument fitting process should allow the clinician to know that the fitted hearing instruments meet the target electroacoustic performance as closely as possible. First, the electroacoustic performance of the instrument should be matched, as closely as possible, to the prescribed 2cc coupler target values for gain and output limiting where the 2cc coupler values have been derived using an individualized real-ear-to-coupler difference. Second, the performance of the hearing instruments must be verified before they are
Table 4. 2cc coupler gain values (in dB) as a function of frequency (in Hz) prescribed by the DSL [i/o] algorithm for the eight month old infant.

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Table 5. 2cc coupler OSPL values (in dB SPL) as a function of frequency (in Hz) prescribed by the DSL [i/o] algorithm for the eight month old infant.

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fitted to the infant. The test signal used in the electroacoustic verification process should adequately represent the frequency, intensity and temporal aspects of speech. Modern advanced signal processing for noise reduction can interact with the test signals that are applied in electroacoustic verification. Therefore, the most accurate representation of the response of a modern DSP hearing instrument will be obtained through the use of modulated speech or speech-like test signals and by turning off the noise reduction signal processing during the verification measurement procedures (Scollie and Seewald 2002). In this case, electroacoustic verification procedures revealed that it was possible to match the DSL [i/o] 2cc coupler target performance for soft, average and loud speech inputs as well as output limiting to within ±5 dB across frequencies.

Validation

Validation is an ongoing process that is performed to ensure that the child’s auditory perception development with amplification proceeds according to expectations for a child with a given degree of hearing loss (Pediatric Working Group 1996). To this end, the Infant and Toddler Meaningful Auditory Integration Scale (IT MAIS) was used with GM’s parents to assess auditory behaviors in everyday situations after three months of hearing instrument use. The parents reported good acceptance of the hearing instruments, device bonding, and changes in vocal behavior associated with auditory stimulation. In addition, the Ling Six Sound testing revealed that she was able to detect, discriminate and identify all six sounds by audition alone. The PIP C10 is a word identification test used for Spanish language. On this test, GM’s score was 100% in an open set. GM’s voice is natural and she uses two-three phrases that are semantically correct. At age 27 months, GM’s vocabulary, phonological, expressive and receptive language development are age appropriate and similar to that of her normally hearing twin sister.

The on-going information obtained from GM’s auditory-verbal therapist (AVT) is critical in the validation of her auditory and speech production development with amplification. Such information can help the audiologist to know whether the ultimate goals of the hearing instrument fitting have been achieved. Her current management plan consists of regular audiological and monitoring of her communication development. When GM enters preschool, consideration for use of an FM system use will be made.

Hearing Instrument Orientation and Training

Parents and other family members or individuals who will assist in caring for the amplification system
should receive orientation, training, ongoing support and appropriate referral as needed from the audiologist. The orientation and training may take place over several appointments based on the family’s abilities to perform required tasks. The basic procedures and equipment for hearing instrument care, earmold insertion and removal, as well as basic troubleshooting procedures should be demonstrated and provided in written format to the family (American Academy of Audiology 2003).

Follow-up

Proper counseling, monitoring and follow-up are essential components of the hearing instrument fitting process. It is recommended that infants and young children be seen by an audiologist every three months during the first two years of hearing instrument use and at least every six months thereafter, if there are no concerns (Pediatric Working Group 1996). It is important to ensure that the child’s hearing is monitored. The frequency for counseling, monitoring and follow-up were increased in this particular case because of GM’s additional visual impairment and her fluctuating hearing thresholds due to her recurrent middle ear condition.

In hearing care work with infants and young children who have sensory hearing loss and who use amplification, routine follow-up procedures should include:

- otoscopy;
- audiometric evaluations including air and bone conduction audiometric and acoustic immittance measures;
- assessment of communication development;
- adjustment of the amplification based on updated information;
- electroacoustic measures of hearing instrument performance, listening checks, and examination of the earmold fit;
- measurement of the RECD each time that the earmold is replaced with hearing instrument performance adjusted accordingly, and finally;
- tests must be repeated whenever there are concerns raised by parents regarding the child’s performance with amplification.

Summary

It is hoped that this case study illustrates the approach to audiologic assessment that clinicians use when working with infants and young children needs to be sufficiently flexible and modified, when necessary, to account for the unique characteristics and capabilities of each child. Further, when working with young children, the electroacoustic characteristics of hearing instruments will need to be changed, sometimes frequently, over time to account for developmental changes in external ear acoustics, as well as temporary changes in hearing sensitivity resulting from middle ear conditions. Consequently, audiologic follow-up in children must be done frequently. The frequency of follow-up visits should be determined on a case-by-case basis. Each child has different needs and unique home and medical circumstances that will influence the decisions made by the audiologist. To take full advantage of early hearing screening programs, audiologists need to be current in their knowledge of best practice procedures, precise in applying their knowledge within the clinical environment and creative and flexible with each family with which they work.

Acknowledgment

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References
