

Potential Pitfalls in the Audiological Assessment of Infants and Young Children

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Introduction

Progress made over the last several years in early identification of hearing loss through implementation of newborn hearing screening (NHS) programs has created a unique opportunity for audiologists to provide amplification to children with permanent hearing loss (PHL) very early in life. Along with this long-awaited opportunity comes the daunting responsibility for accurately determining the type, degree and configuration of an infant's hearing loss so that appropriate counseling of parents, fitting of hearing aids, management planning and follow-up can proceed in a timely manner. Also critically important is to confirm the presence of normal hearing in a baby who failed NHS (a false-positive outcome), so that parental anxieties can be alleviated and unnecessary interventions avoided.

Key to this process is the comprehensive audiological assessment. Current otoacoustic, physiological, electrophysiological and behavioral technologies and procedures have provided the pediatric audiologist with an assessment armamentarium that can yield information about the integrity of the auditory pathway from the periphery through the central auditory system. Unfortunately, as clinicians are learning rapidly, there are numerous 'pitfalls' that must be anticipated when evaluating the hearing status of infants and very young children and when monitoring children with confirmed PHL over time. Unknowingly encountering an audiological pitfall can deleteriously impact the validity of the audiological assessment, the primary building block of a

sound foundation for optimal amplification device provision.

This chapter will address some of the potential pitfalls that audiologists might encounter when assessing the hearing of infants and young children. Three types of errors that could be made in audiological diagnosis will be described. Case studies will illustrate when clinicians might encounter audiological pitfalls, why the pitfalls occur, and comment on how clinicians might avoid these obstacles to accurate, timely and appropriate service provision.

Background

The Year 2000 Joint Committee on Infant Hearing Position Statement Principles and Guidelines (JCIH 2000) state that all infants should have access to hearing screening in the neonatal period regardless of risk status (i.e., universal newborn hearing screening). The goal for infants who fail NHS is to begin appropriate audiological and medical evaluations to confirm the presence of hearing loss before 3 months of age, and to begin early intervention services by age 6 months. JCIH 2000 encourages the development of comprehensive audiological assessment strategies to confirm the existence of PHL, delineate its characteristics, and monitor its stability in the long term. Clearly the goals specified by JCIH 2000 require that audiological assessments be not only comprehensive, but provided in a timely manner.

In response to the proliferation of NHS programs and local, state and federal support for the practice, the US Preventative Services Task Force (USPSTF) an agency of the Association for Healthcare Research and Quality (AHRQ) issued its newest *Recommendation on Newborn Hearing Screening* in October of 2001. In its previous statement on NHS issued in

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1995, the task force recommended targeted newborn hearing screening; that is, the testing of only infants with risk indicators (primarily babies cared for in neonatal intensive care [NICU] or special care baby [SCBU] units). The USPSTF following an extensive review of the published literature on NHS formulated the new 2001 Recommendation. The Task Force evidence review followed formal guidelines and the rating system for study quality developed by the AHRQ (formerly, the Agency for Healthcare Policy and Research: AHCPR). Based on its review, the USPSTF concluded that there was currently insufficient evidence (an “I” rating) to recommend for or against the practice of providing hearing screening to all newborns before hospital discharge (i.e., universal screening). Thus, the Task Force recommended that universal NHS be neither discontinued, nor endorsed as public health policy. The summary of the evidence supporting their recommendation on NHS was published recently in the *Journal of the American Medical Association* (Thompson et al. 2001).

Relevant to the topic of this chapter, the Thompson et al. (2001) summary stated that diagnosis and management decisions regarding newborns that failed the in-hospital hearing screening were based primarily on the results of a diagnostic auditory brainstem response (ABR) evaluation. This ABR assessment, they noted, was usually completed when infants were between 1 and 6 months of age. The Task Force did find evidence that the initial ABR assessment resulted in the provision of early intervention services to most infants with PHL. However, the USPSTF also found evidence suggesting that the initial diagnostic ABR might, in some cases, overestimate the number of infants with PHL.

In support of this conclusion, the USPSTF report cited the well-known Wessex Trial (Kennedy 1999) conducted in England. In that trial, infants referred by the participating hospitals following NHS received their first diagnostic ABR evaluation between the ages of 2 to 3 months. Of 158 infants who received ABR assessment, 27 were determined to have bilateral PHL. However, as the USPSTF summary noted, in two cases (7.4% of infants), the initial diagnosis of PHL was incorrect. Later (at 4 and 10 months of age) both children had normal hearing sensitivity confirmed. The USPSTF review also referred to the results of the Rhode Island Hearing Assessment Project (Vohr, Carty, Moore and Letourneau 1998) for the years 1993–1996. Five infants (29%) of the 17 initially diagnosed with mod-

erate PHL were later found to have only mild hearing impairments.

Relevant to our profession, several conclusions of the USPSTF regarding audiological assessment of infants referred by NHS programs are worth highlighting. First, the USPSTF found no evidence supporting the reliability of diagnostic audiological assessment procedures (including behavioral test methods) for use with this population. Second, the Task Force concluded that “the intermediate diagnostic standard determining PHL is imperfect; in expert hands, as many as 7% of infants diagnosed as having PHL may eventually prove to have normal hearing”. Third, the USPSTF commented, “the frequency of [audiological] misdiagnosis in everyday practice settings has not yet been studied” (Thompson et al. 2001, p. 2006).

The USPSTF also examined evidence suggesting delayed confirmation of hearing loss for some children following UNHS. Using data from the Wessex trial (1998), the Task Force report concluded that universal NHS resulted in statistically significant increased referral rates for audiological assessment of infants with PHL by 6 months of age versus infants who were not screened before hospital discharge. However, the USPSTF summary noted that the age of intervention for screened infants with PHL (10 months) in the Wessex trial was not statistically different than the age of intervention for unscreened babies with hearing loss who participated in the study (Thompson et al. 2001). It is important to remember however, that for decades behavioral hearing screening using the Home Health Visitor Distraction Test has been routinely provided to all infants in England at about 9 months of age. Consequently, the finding of ‘no difference’ in age of intervention for screened and unscreened infants may be unique to that country and therefore, the result may not generalize to the US or to other countries for which routine hearing screening later in infancy is not a component of pediatric health care provision.

The Thompson et al. (2001) evidence summary also referenced the 1995–1997 outcome data reported by the New York State Newborn Hearing Screening Demonstration Project (NYNHSDP) (Dalzell et al. 2000). The NYNHSDP found that the median age of confirmation of bilateral PHL was 5 months for 81% (29 of 36) of infants who were subsequently fit with amplification. NICU infants had their hearing loss confirmed later than infants with PHL from the well-baby nursery. PHLs of mild/moderate degree were

diagnosed later (median = 3.5 months) than PHLs of severe/profound degree (median = 2.0 months). For about 20% of infants however, the confirmation of type and degree of PHL took months longer. The reasons for the delays in confirmation of PHL were attributed to: (1) *eventual diagnosis of mild hearing loss*; (2) parental non-compliance with follow-up; (3) illness of the child; (4) presence of other developmental delays; and, (5) “*audiological uncertainty*” regarding test results (Dalzell et al. 2000).

Types of Audiological Misdiagnosis

There are several categories of audiological misdiagnosis that may occur when evaluating the hearing status of infants and young children. Likely, the most common type of audiological misdiagnosis occurs when a hearing loss is identified correctly, but an incorrect conclusion regarding the type or degree of the impairment is made. This type of audiological misdiagnosis will be referred to as a misidentification. The second type of audiological misdiagnosis is the false-positive outcome. It is the error of diagnosing a child as having PHL when normal hearing exists. The third type of audiological misdiagnosis, the false-negative outcome, is arguably the most serious. Following the audiological evaluation, the child with a true PHL is determined to have normal hearing.

Why should pediatric audiologists be concerned and continually vigilant over possibly committing one of these three types of audiological misdiagnoses? It is because the potential consequences of such errors may be detrimental to the child and family in the short and long term. Audiological misdiagnosis can result in: (1) a delay of varying length in the confirmation of the child’s true hearing status; (2) a delay in the referral of a child for a potentially treatable medical condition; (3) a delay in referral of the child and family for beneficial early intervention services; (4) the provision of inappropriate management/therapies (e.g., audiological, medical, surgical, prosthetic, educational, communication); (5) parental anxiety, confusion, and loss of confidence in recommendations made by the clinician subsequent to the discovery of the audiological misdiagnosis; (6) unnecessary expenditure of resources by the family (e.g., emotional, time, monetary) and by the system (e.g., expertise, services, financial); and, (7) reduced confidence in the profes-

sional capabilities of audiologists by others (e.g., policy makers, legislators, agencies, physicians, educators).

Potential Audiological Pitfalls

Considering the broad diversity and complexity of cases regularly referred to pediatric audiologists, likely there are overall relatively few cases of audiological misdiagnosis. As reviewed previously, however, because the consequences of such clinical errors can so negatively impact the child and family, the audiologist must adopt current technologies and test procedures that reduce the possibility of encountering audiological pitfalls during the evaluation process.

The following eight cases were selected to illustrate the three types of audiological misdiagnoses delineated above. Each case is overviewed, the clinical impression following the audiological assessment, as well as the child’s actual audiological diagnosis are provided. Next the audiological pitfalls encountered are delineated. This is followed by specific comments about the test procedures or techniques that could have been used, or modified, to avoid the pitfalls and the resultant audiological misdiagnosis.

One mechanism for quality assurance and continuing education in the medical setting is ‘Clinical Grand Rounds’. A regular presentation in Grand Rounds is known as ‘M and M’ (morbidity and mortality), a session during which clinicians present and openly discuss difficult cases and the circumstances that may have led to a medical misdiagnosis. The presentations are informative and serve to raise awareness about potential pitfalls, thereby supporting quality clinical practice. The section that follows should be thought of as a ‘Clinical Grand Rounds’. It is intended to support consistent, quality pediatric audiological service delivery by vigilant and informed professionals. It is not meant to in any way to diminish the high quality of the audiological assessments currently provided to the majority of infants and young children evaluated by pediatric audiologists. Although we may continually strive to achieve it, none of us is the perfect clinician: even the ‘experts’ can make clinical errors. Therefore, with the intent of promoting continued quality assurance in audiological service delivery to all children and their families, the following cases and comments are presented.

Type I Audiological Misdiagnosis: Misidentification

Case 1: *Temporary conductive hearing loss secondary to the presence of middle ear effusion is diagnosed as sensorineural.*

Overview. An infant 3 months of age was referred for audiologic assessment from a NHS program. The baby spent two months in the NICU. Air-conducted ABR threshold recordings in both ears were elevated, consistent with a loss of moderate-severe degree. No otoacoustic emissions (OAEs) were evoked in either ear. The tympanogram was recorded using a standard 220-Hz probe-tone frequency and a 'type A' pattern (Jerger 1970) resulted.

Clinical Impression. Moderate-severe bilateral sensorineural hearing loss (SNHL).

Actual Audiological Status. Conductive hearing loss secondary to middle ear effusion.

Audiological Pitfalls.

1. Failure to record bone-conduction ABR thresholds
2. Use of a standard 220-Hz probe frequency for tympanometry.
3. Misinterpretation of the reason for absent OAEs

Comments. Conductive hearing loss associated with middle ear effusion (MEE) or otitis media with effusion (OME) is a common condition of early childhood. The prevalence of MEE in the well-baby nursery has been estimated at 3% (Pestalozza 1984), 9% (Doyle, Burgraaff, Fugikawa, Kim and Macarthur 1997), and 11% (Chang, Vohr, Norton and Lekas 1993) in various studies. In the NICU/SCBU, the prevalence of MEE appears to be higher with estimates at 7% (Smurzynski et al. 1993), to as high as 21% (Pestalozza 1984), 29% (Sutton, Gleadle and Rowe 1996), and 30% (Balkany, Berman, Simmons and Jafek 1978).

Later in infancy, the point-prevalence of OME examined prospectively in a large cohort, increased monthly from to .33 to .48 in infants 3 to 8 months of age from urban environments (Paradise et al. 1997). In 5- to 16-month-olds, the percentage of children with MEE has been estimated to be between 29% and 64% as determined from the prospective study of cohorts in North Carolina and New York (Gravel and Wallace 2000; Gravel et al. in review). Gravel (1989)

reported conductive hearing loss associated with MEE was present during one-half of routine well-baby visits in 6- to 12-month-old infants from low-income families. Widen and her colleagues (2000) reported that in the audiological follow-up of a large cohort (> 3000) 8- to 12-month-old infants, 55% of parents reported their babies had had OME, and 30% of babies actually had MEE on the day of their audiological assessment.

Tympanometry findings often may be invalid in young infants yielding false-negative results (suggest normal middle ear function with MEE present) when a standard 220-Hz probe-tone frequency is used for assessment (e.g., Paradise, Smith and Bluestone 1976). However, high-frequency (660/678 Hz and 1000 Hz) probe tones appear to be reasonably sensitive and specific for tympanometry in infants younger than 4 months of age (Shurin, Pelton and Finkelstein 1977; Marchant et al. 1986; McKinley, Grose and Roush 1997; Baldwin 2000). Recently, the protocol developed by the UK Working Group (2000) on Tympanometry in Neonates Under 4 Months (<http://www.unhs.org.uk>) recommended the inclusion of single-component admittance measurement using a 660/678 Hz or 1000 Hz probe-tone frequency be included in the audiological test battery for infants 4 months of age and younger. In addition, the high-frequency probe is recommended for the measurement of the acoustic middle ear muscle reflex (MEMR). The Recommendations also include a method for interpreting the tympanogram results. (Note: various Working Groups have had responsibility for developing evidence-based uniform test protocols for the audiological assessment of infants referred by universal NHS programs currently being phased-in throughout the United Kingdom.)

The ABR is currently the single most useful technique for estimating auditory sensitivity in infants and young children who cannot be reliably assessed with conditioned behavioral test procedures. In infants and young children (1 week to 8 years) with normal hearing or sensorineural hearing loss, the frequency-specific ABR threshold and behavioral threshold are in good agreement (within 15 dB in 83% and 20 dB in 93% of cases), with correlations $\geq .94$ for 500, 2000 and 4000 Hz (Stapells, Gravel and Martin 1995).

However in some cases, clinical experience suggests that the ABR may overestimate the actual degree of hearing loss when MEE is present (Stapells, Gravel and Bernstein 1987; Stapells 1989). Figure 1

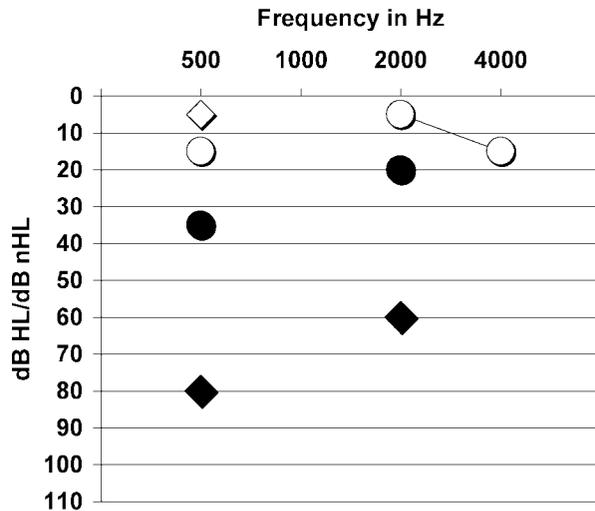


Figure 1. ABR and behavioral test results obtained from a 9-month-old with otitis media with effusion. Filled diamonds = air-conduction (AC) auditory brainstem response thresholds (ABR), open diamonds = bone-conduction (BC) ABR threshold; filled circles = AC behavioral thresholds; open circles = BC behavioral thresholds. Behavioral responses are plotted in dB HL and ABR responses in dB nHL. Figure adapted from Stapells 1989.

from a case described by Stapells (1989) depicts the 'audiogram' (behavioral and ABR thresholds) of a 9-month-old infant who had known normal cochlear function. Note that the ABR air-conduction thresholds greatly overestimate behavioral thresholds at the same nominal test frequencies. However, observe that the behavioral and ABR bone-conduction thresholds are similar and both provide evidence of a significant air-bone gap associated, in this baby, with OME. The mechanism(s) contributing to this disparity are not clear. It is stressed that this does not happen in every child with MEE. However, the possibility of such an outcome supports the need for inclusion of bone-conduction ABR in the assessment of infants who exhibit elevated air-conduction thresholds, regardless of the tympanometric findings (e.g., Gravel and Hood 1998). Stapells (2000) has provided an excellent review of bone-conducted tonal ABR and provided normative threshold values that are useful in clinical practice. Gorga and Neely (Chapter 4 in this volume) also provide a review of other factors that may influence the accuracy of the ABR when using the procedure to estimate auditory sensitivity.

If middle ear effusion is present, it is unwise to postpone the ABR evaluation until MEE/OME has resolved. Davis and colleagues (1997) have reported that this practice can result in significantly delayed confirmation of underlying sensorineural hearing loss in some children. To avoid months of delay in confirmation of a PHL that is masked by the presence of middle ear effusion, bone-conduction ABR should be considered a routine component of the comprehensive audiological assessment.

Case 2: SNHL is correctly identified in a child; the degree of the hearing loss is misdiagnosed as greater than the actual impairment.

Overview. A click-ABR was completed on a 4-month old infant referred because of parental concern over lack of response to auditory stimuli. No responses were recorded at the maximum output (106 dB nHL) of the test equipment. The child had normal tympanograms and absent acoustic MEMRs. At the time of the initial test, OAE technology was not part of clinical protocols. The parents were told that the child had "no hearing". The baby was seen at another facility that was able to provide air- and bone-conduction frequency-specific ABR and conditioned behavioral testing (Visual Reinforcement Audiometry: VRA). Results indicated good agreement between ABR and behavioral thresholds in low- and high-frequency regions in both ears. Bone-conduction ABR and behavioral thresholds, tympanometry and acoustic MEMR results were consistent with SNHL. The infant clearly had useable residual hearing and was immediately fit with binaural amplification (Gravel, Kurtzberg, Stapells, Vaughan and Wallace 1989).

Initial Clinical Impression. "No hearing" in either ear.

Actual Audiologic Status. Moderate-severe low-frequency SNHL sloping to hearing loss of severe to profound degree in the high frequencies bilaterally.

Audiological Pitfalls.

1. Interpretation of an absent click-ABR as indicative of "no hearing".
2. Failure to obtain a frequency-specific ABR to examine low-frequency hearing (500 Hz) and better estimate thresholds in high-frequency regions (at 2000 and 4000 Hz).
3. Failure to obtain ear-specific air-conduction and bone-conduction behavioral responses.

Comments. The need to provide a complete audiologic assessment of infants with severe and profound SNHL is critical particularly since the age of cochlear implant candidacy has decreased in the US to 12 months of age. Based on click-ABR assessment alone, an infant's residual hearing cannot be adequately delineated. Ear-specific behavioral and frequency-specific ABR and behavioral threshold assessments are important in determining appropriate intervention strategies. It is important to ensure that conventional amplification has been selected to maximize the infant's ability to use his/her hearing for the development of auditory skills. Now this is accomplished through threshold-based prescriptive hearing aid fitting procedures. Only when it is clear that sufficient experiences with well-fit amplification have been provided can informed decisions regarding cochlear implantation be made by parents and professionals.

Case 3: *A stable sensorineural hearing loss is misidentified as progressive*

Overview. Moodie, Sinclair, Fisk and Seewald (2000) reported the case of an 8-month-old infant with severe to profound sensorineural hearing loss whose audiologic assessment using VRA was completed using insert earphones. The child's threshold responses were specified in dB HL; specifically, the audiometer dial setting necessary to elicit reliable threshold behavioral head-turn responses at 500, 1000, 2000 and 4000 Hz. Subsequently, in follow-up, the audiological assessment of the same baby at 14 months of age using insert earphones demonstrated a decrease in hearing threshold sensitivity at 1000, 2000 and 4000 Hz (by 15, 20 and 20 dB HL, respectively).

Clinical Impression. Progressive high-frequency hearing loss

Actual Audiological Status. Stable hearing loss; the poorer dB HL thresholds measured audiometrically can be attributed to changes in the child's external ear acoustics with maturation.

Audiological Pitfalls.

- 1 Failure to consider the effect of changes in external ear geometry on thresholds obtained using insert earphones.
- 2 Failure to convert behavioral thresholds measured in dB HL to dB SPL using correction factors (the child's own real-ear-to-coupler difference values) measured at each age.

Comments. Seewald and Scollie (1999) and Moodie et al. (2000) remind us that the output of an insert earphone, calibrated in a standard 2cc coupler (approximating the volume of the external ear of an adult) will yield different dB SPL values when measured at the eardrum of infant than an adult. Indeed, the dB HL value on the audiometer dial required to produce the same dB SPL value at the eardrum for an infant and an adult can differ significantly, particularly in high-frequency regions. The same is true for children. Moodie et al. (2000) reported that in the case of the infant described above, the predicted differences in SPL for the same HL audiometer dial setting for the infant at 8 months compared to the same infant at 14 months were on the order of 6, 12, 12, and 17 dB less at 500, 1000, 2000 and 4000 Hz, respectively.

Infant dB SPL values as measured at the eardrum are affected by the natural acoustic increase in SPL resulting from the smaller residual volume of the infant ear than the adult. Importantly, changes in the size of the infant's ear canal with maturation could affect measured audiometric threshold values: increasing the dB HL audiometer dial setting is needed to compensate for the increase in the size of the external ear canal. Said differently, a greater dB HL dial setting is needed to achieve the same dB SPL value at the eardrum. Without consideration of the change in the external ear characteristics with maturation, the audiologist might erroneously attribute poorer HL thresholds to progressive hearing loss, when in fact, no change in threshold sensitivity for dB SPL at the eardrum exists.

Using a procedure described by Seewald and Scollie (1999), each child's real-ear-to-coupler difference (RECD) values across the speech frequency range are regularly ascertained during early childhood in order to individualize the hearing aid fitting by revising amplification targets particularly following the refabrication of the child's custom earmolds. During the first year of life, this usually occurs frequently, sometimes once every one or two months; see figure 5 below and the chapter by Beauchaine (see Chapter 8 in this volume). If the individual RECD values for an infant cannot be obtained, average values are available. However, it is optimal to measure the child's own RECD rather than rely on average values.

Seewald and Scollie (1999) recommend using the RECD values for audiometric purposes. The same individually measured RECDs measured for setting

amplification targets are applied to the child's dB HL audiometric thresholds. Therefore dB HL thresholds are converted to dB SPL values; given the use of the child's own RECD values, the result best approximates dB SPL thresholds if measured at the eardrum. Briefly, in the Seewald and Scollie method, the audiological assessment is undertaken using the child's own earmolds instead of the insert earphone/foam tip for test signal delivery. With the nub and tubing of the insert earphone coupled to the child's own custom earmold tubing, behavioral thresholds (using VRA, for example) are assessed. Thresholds are recorded in dB HL dial setting and subsequently converted to dB SPL values by applying the child's measured RECD at each nominal test frequency. Over time, records used for monitoring the child's hearing sensitivity are maintained in dB SPL. Unlike dB HL values, the derived dB SPL measures approximate the actual dB SPL needed to reach threshold if those values could be measured at the eardrum.

Monitoring hearing over time in this manner could prevent the audiologist from the pitfall of erroneously counseling parents that their infant's hearing loss was worsening. Obviously, this needless and unwarranted anxiety is undesirable. Further, unnecessary audiological (changes in output of the hearing aid), medical (drug therapies) and surgical (e.g., fistula exploration) procedures could result because of such an audiological misidentification.

Case 4: *An auditory disorder is identified; the type of auditory impairment is misdiagnosed.*

Overview. A toddler who was cared for in the NICU in the newborn period was referred for audiological assessment because of lack of response to sound. The baby had not received hearing screening in the newborn period. The infant was tested behaviorally using VRA and a severe hearing loss was identified. Tympanograms were considered to be normal in both ears and acoustic MEMRs were absent. To confirm the hearing loss, the child received an ABR evaluation. An air-conduction click-ABR was completed using alternating polarity clicks. The infant was tested in natural sleep because the facility could not perform sedated ABR testing. Results of the ABR were interpreted as the presence of wave I, as well as later waves at stimulation intensities of 90 and 80 dB nHL, but not at 70 dB nHL. Only one run was recorded in each ear at each stimulation intensity

before the child awoke. Again the tympanograms were considered normal. OAE assessment was not completed, since the test was not available at the facility and the audiologist felt it unnecessary given the agreement among behavioral, ABR and acoustic MEMR results.

Clinical Impression. Severe bilateral sensorineural hearing loss.

Actual Audiological Status. Auditory neuropathy/auditory dys-synchrony.

Audiological Pitfalls.

1. Failure to examine click-ABR with separate replicated runs of opposite polarity (condensation and rarefaction).
2. Failure to complete evoked otoacoustic emissions evaluation.

Comments. Although an auditory disorder was identified (a severe bilateral sensorineural hearing loss), the actual disorder (auditory neuropathy/auditory dys-synchrony) was misidentified. Auditory neuropathy or auditory dys-synchrony (AN/ADS) is an auditory disorder that has recently received a great deal of attention in the literature. In classic cases, the individual with AN/ADS has behavioral thresholds that may range from normal to profound hearing loss. The ABR is atypical or often absent. Acoustic MEMRs are absent. Notably however, OAEs are often present and sometimes particularly robust. Children with AN/ADS usually have severe difficulty understanding speech, more so than their loss of hearing sensitivity as measured audiometrically would suggest. Usually the use of a visual communication system (notably, Cued Speech) rather than auditory-based communication facilitates the acquisition of language. Children with AD/ANS may or may not benefit from conventional amplification and there is evidence that children with AN/ADS may benefit from use of a cochlear implant.

Two major pitfalls were encountered in this case: first, the failure to evaluate the infant using evoked otoacoustic emissions technology and, second, failure to collect sufficient ABR data so as to ensure the conclusions reached regarding the type of hearing loss were valid. Insufficient audiological assessment data led the audiologist to form an incorrect impression of the type of auditory disorder the child was experiencing. Had all components of the test battery been completed, the incongruity of the results would have been readily apparent.

In this case, evoked otoacoustic emissions testing would have revealed the presence of OAEs. This finding would have been inconsistent with a sensorineural (cochlear) hearing loss of severe degree, as in severe SNHL, OAEs would be absent. The pitfall was to have assumed based on behavioral, ABR and acoustic MEMR results that OAE testing was redundant.

Failure to replicate ABR waveforms using separate runs of opposite click-polarity (condensation and rarefaction) rather than alternating the click polarity within the same run resulted in incorrect interpretation of the electrophysiological findings. Had the full test protocol been completed, the audiologist would have recognized that the response interpreted as wave I of the ABR (a neural response) was, in fact, the cochlear microphonic (CM). Characteristically, the CM follows the polarity of the click stimulus. Therefore, separate runs of condensation and rarefaction clicks reveal a response whose polarity reflects the polarity of the clicks. If the response considered to be wave I reverses in polarity it is indeed the CM. Note that using alternating polarity clicks to derive an ABR essentially cancels the cochlear response. Moreover, if with decreases in stimulus intensity, the presumed wave I remains at the same latency, the response is not neural. Interpretation of one run of an ABR without replication is dangerous and can lead to incorrect conclusions regarding the presence of ABR waveforms.

Had the full ABR protocol been completed, there would have been clinical clues that the infant's auditory disorder was not sensorineural in type. Since the complete ABR protocol could not be completed at that session (infant awoke), the baby should have been brought in for one or more tests until all information had been collected and the ABR results were considered to be complete and reliable.

The difference in the approach to parent counseling, conventional amplification fitting, medical referral, communication approach and early intervention planning are greatly different for children with AD/ANS versus those with peripheral sensorineural hearing loss.

It is important to note that NHS programs that use only OAE technology will fail to identify most infants with AN/ADS, as OAEs are generally present. NHS using ABR screening technology would result in a screening failure (see Sininger, Chapter 15 in this volume, and Sininger and Oba 2001 for a complete review).

Type II Audiological Misdiagnosis: False-Positive

Case 5: *Normal hearing sensitivity diagnosed as sensorineural hearing loss.*

Overview. Behavioral testing of a 7-month-old premature infant was undertaken using the VRA technique. The audiologist had difficulty conditioning the infant; results were considered to be of only fair reliability and minimal response levels (MRLs) were obtained at elevated levels for speech and 500-Hz, 2000-Hz and 4000-Hz signals in the sound field. The tympanograms were consistent with normal middle ear function in both ears.

Clinical Impression. Possible mild-moderate hearing loss.

Audiological Pitfalls.

- 1 Assuming by the chronological age of the infant that VRA testing would be successful.
- 2 Failure to crosscheck behavioral test results with otoacoustic and physiologic tests, specifically acoustic MEMR measures and OAEs.
- 3 Failure to refer for ABR assessment since hearing loss was suspected and behavioral responses were considered of only fair reliability.

Comments. VRA is a useful procedure for the behavioral assessment of infants, beginning at about 6 months (developmental) age. Figure 2 presents data from Widen (1990), Gravel and Wallace (2000) and Widen et al. (2000) on the success rate of VRA as a function of corrected age in months. The Widen 1990 and Gravel and Wallace 2000 studies used a computerized VRA procedure; the Widen et al. 2000 data were collected using a traditional clinical procedure that followed a rigorous test protocol that included scoring of responses during control trial intervals. Note the importance of considering the developmental age of the child with regard to interpreting the validity of the VRA task. For infants aged 5 months corrected age, there is only a fair probability (50–75%) that VRA testing will be successful. By 6-months, however, there is an 80% probability that babies will be successfully tested using VRA. For the vast majority (> 86%) of infants between 7 months and 24 months of age, the VRA procedure is successful.

When assessing the hearing sensitivity of an infant born prematurely, it is important to use the

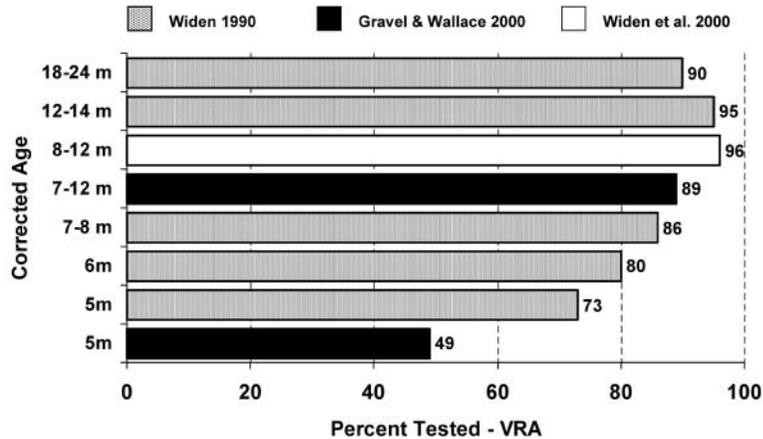


Figure 2. Percent-success rate for VRA test procedure according to age in months (m). Ages are corrected for prematurity. Data from three studies are displayed: gray bars are from Widen 1990, the filled bars from Gravel and Wallace 2000, and the open bar from Widen et al. 2000.

infant's corrected age rather than the chronological age in predicting whether or not the infant might condition for VRA. Therefore, if an infant aged 7 months is being evaluated and the infant was born 2 months prematurely, there is a lower probability that the infant will condition for VRA than if the baby were a full-term 7-month-old (7 months – 2 months premature = corrected age of 5 months).

When assessing young children with developmental delay, it is also important to have an estimate of cognitive function in order to determine whether the child is developmentally appropriate for the procedure. Data from Widen (1990) demonstrate that using developmental age (estimated using a standard scale of development), infants functioning below 5 months of age have about a 50% probability and those below 4 months, a 20% probability of being successfully tested with VRA.

Type III Audiological Misdiagnosis: False-Negative

Case 6: *Severe to Profound sensorineural hearing loss diagnosed as normal hearing because of unreliable behavioral testing.*

Overview. Behavioral assessment of hearing was completed on a healthy 9-month-old infant who was born at a hospital that did not offer NHS. The mother was encouraged to seek audiological follow-up by the pediatrician because of her concern about the infant's

inconsistent responses to sound. The baby was tested using the VRA procedure. Two audiologists tested the infant: one outside the test booth controlled stimulus presentations; the other, in the booth with the infant, served as the distracter. A 60 dB HL speech stimulus was used to shape the head-turn response. The infant was trained to turn by pairing the stimulus with the illumination and activation of the visual reinforcers. The parent and distracter were both able to hear the test signals. Once the toddler was making head-turns, minimal response levels were obtained for speech and tonal signals from 500 through 4000 Hz at 20 dB HL. Normal tympanograms were recorded in both ears.

The mother was counseled that the child had normal hearing. One year later, with no development of communication

and the parents still concerned about the child's lack of responses to sound, an ABR was completed along with OAEs and acoustic MEMR testing. All measures were consistent with severe to profound SNHL; conditioned behavioral testing supported the audiological diagnosis.

Clinical Impression. Based on the behavioral responses and the normal tympanograms, the child was considered to have normal hearing sensitivity in at least one ear.

Actual Audiological Status. Severe-to-profound sensorineural hearing loss

Audiological Pitfalls.

1. Shaping the head-turn response using an inaudible stimulus and therefore reinforcing random, non-contingent head turns (false-positive responses).
2. Failure to include catch trials in the test protocol which would have provided evidence of false-positive responding.
3. Use of biased observers (both audiologists and parent).
4. Failure to crosscheck behavioral outcomes with electrophysiological, physiological and otoacoustic measures.

Comments. The case of a false-negative audiological assessment is arguably the most significant audiological misdiagnosis for a child and family because of the frequently great delay in the confirmation of

hearing loss. Berlin and Hood (1993) reported on such a false-negative case. In their report, the infant received an ABR at 7 months and based on the electrophysiologic test was determined (correctly) to have a severe-to-profound hearing loss. The baby was fit with hearing aids but made little progress with the acquisition of auditory-oral communication. Behavioral testing was undertaken, both observational techniques and ‘conditioned’ response procedures completed using sound field test signals were thought to be consistent with hearing thresholds in the 25–35 dB HL range. Interestingly (although apparently overlooked by the audiologist), sound field aided responses were *poorer* than unaided. Based on these results (and the aided-unaided incongruity) it was concluded that the child had a central processing disorder. The hearing aids were removed and subsequent behavioral tests were all consistent with normal hearing. Again, with no progress being made, the child was referred at the age of 3.5 years for another ABR. Results (consistent with the first ABR assessment) supported the presence of severe to profound SNHL. Appropriate intervention was begun immediately and the child made good progress.

Adopting several important practices in behavioral assessment are important to avoiding the pitfalls of VRA assessment that are primarily related to the infant not being under stimulus control and observer bias. Nozza (1999) has stated that in visual reinforcement audiometry “teaching babies the head-turn response is easy. The hard part is teaching them when *not* to turn” (See Chapter 3 in this volume). When testing an infant using VRA:

- (1) Follow a specific test protocol that is evidence based.
- (2) Include control (silent) trials and evaluate (quantify) the false-positive response rate to determine the reliability of assessment
- (3) Add ‘objectivity’ to VRA (Widen 1993) using a rigorous, rule-based protocol or computer-assisted VRA procedure that is specifically designed to reduce bias (through the use of blinded observers).

Case 7: *Severe to Profound sensorineural hearing loss diagnosed as normal hearing because of incorrect interpretation of ABR results.*

Overview. In the last volume from the International Conference Proceedings, the author reported

a case that bears reiterating in this chapter on audiological pitfalls (Gravel 2000).

A full-term 6-month-old infant was seen at one clinical audiology facility for behavioral assessment because of the mother’s concern over the infant’s lack of response to sound. The baby was successfully conditioned for VRA assessment and behavioral results suggested a severe to profound hearing loss. Behavioral testing was thought to be reliable. The facility did not have frequency-specific ABR available and so the infant was referred to another facility for the test. ABR recordings were completed using tonal stimuli and an ascending procedure. Responses at 500 and 2000 Hz were considered to be normal. No responses were collected at intensities greater than used to obtain the ‘normal’ responses.

Clinical Impression. Based on the ABR, the child was thought to have normal auditory sensitivity in both ears. Over the course of several years, the child was seen for repeat testing at various facilities as there was continued lack of response to sound and no development of spoken communication. At each visit to an audiologist, the mother provided the clinician with the results of the ABR assessment. Subsequently, every behavioral test the child received following the ABR resulted in normal threshold responses. No other tests were administered except tympanometry. The child began learning a formal sign system for communication and was diagnosed as being ‘cortically deaf’. At 4 years of age, the child received another ABR evaluation that was consistent with the 6-month behavioral results. Otoacoustic emissions assessment and acoustic MEMR reflex testing were consistent with SNHL.

Actual Audiological Status. Severe to profound sensorineural hearing loss.

Audiological Pitfalls.

1. Inappropriate interpretation of ABR waveforms.
2. Failure to replicate responses.
3. Failure to complete full ABR protocol (threshold and supra-threshold assessment).
4. Failure to assess the child with OAEs and acoustic MEMR measures.
5. Biased behavioral assessments

Comments. Lack of an ABR protocol that included replication of waveforms using intensities at and above estimated threshold levels among other factors resulted in an inaccurate interpretation of the ABR recordings. In this case, the misinterpretation of

the ABR recordings as ‘normal’ undoubtedly biased subsequent behavioral audiometric test results. Moreover, the bias for a ‘normal’ outcome was so strong that otoacoustic and acoustic MEMR testing was not completed. In the presence of normal tympanograms, had clinicians applied these routine acoustic and physiological crosschecks of the behavioral test, incongruous findings may have been identified. It is important to remember that tester bias can arise from multiple sources and result in false-negative audiological diagnosis

Case 8: *Normal hearing concluded based on electrophysiological (ABR) testing alone.*

Overview. A case reported by Gravel and colleagues (Gravel, Kurtzberg, Stapells, Vaughan and Wallace 1989) serves to illustrate the consequences of over interpretation of electrophysiological findings. Air-conduction threshold ABR was completed on an infant in the NICU who experienced severe perinatal asphyxia. The results revealed threshold responses recorded bilaterally to 20 dB nHL. (Fortunately, obligatory cortical evoked potentials were also recorded and showed no response to any stimuli). Otoacoustic emissions were not available for use in the test battery at the time of test. However, we would speculate that this infant would have displayed normal cochlear emissions. Later when the infant was evaluated using behavioral observation techniques to examine global hearing status, there were no functional responses to sound.

Clinical Impression. Given the results of the ABR assessment, the clinician might conclude that this infant had normal hearing.

Actual Audiological Status. While the infant displayed normal auditory pathway integrity to the level of the brainstem, the cortical auditory evoked potentials revealed severe cortical damage secondary to perinatal asphyxia. The infant displayed no functional responses to sound.

Potential Audiological Pitfall. Interpreting normal ABR threshold responses as normal hearing (in the global sense).

Comments. While behavioral observation audiometry may have limited value for determining audiometric thresholds, observation of auditory behaviors in response to sound are useful in gaining insight into functional hearing (Diefendorf and Gravel 1996; Gravel 2000). Examining behavioral orienting responses from infants during the neonatal

period may even provide insight into neurodevelopment (Diefendorf and Gravel 1996). Behavioral observations of auditory abilities should be used as a means of crosschecking (validating) the results of electrophysiological assessment. However, using unconditioned behavioral response procedures for specifying the degree and configuration of hearing loss for the purposes of in-depth audiological assessment or hearing aid fitting should be avoided. Observing the responses of an infant to various sounds and determining whether or not the baby’s responses are age appropriate (Northern and Downs 1984) is important regardless of the outcome of the electrophysiological assessment (or OAE testing, when the measure is used alone). It is recommended that some behavioral examination of auditory function be included in the pediatric audiological test battery regardless of the child’s age or the outcome of electrophysiological tests (Diefendorf and Gravel 1996).

Avoiding Audiological Pitfalls with a Test Battery Approach

Throughout the cases presented here, the overriding pitfall was the failure of audiologists to administer a battery tests and examine the outcomes of the measures for agreement. As demonstrated by these cases, failure to use the full test battery and seek accord among the results can lead to all three forms of audiological diagnostic errors (misidentification, false-positive and false-negative outcomes).

Figure 3 presents a schematic of the approach to pediatric assessment that has been advocated by many (e.g., Gravel and Hood 1998). A comprehensive test battery should include multiple assessment procedures including behavioral and physiological tests (ABR, otoacoustic emissions, and acoustic MEMRs). As these cases have demonstrated, it is unwise to determine the hearing status of an infant based on the results of one test alone or on a limited test battery. Examples of the pitfalls that can be encountered when using only a single assessment procedure (such as VRA or ABR) were demonstrated in the previous cases. Although no specific case example was presented, the cautions raised apply to the interpretation of evoked otoacoustic emissions results as well.

Recall the classic study of distortion product otoacoustic emissions (DPOAEs) by Gorga and colleagues (1997) that found overlap in the distributions

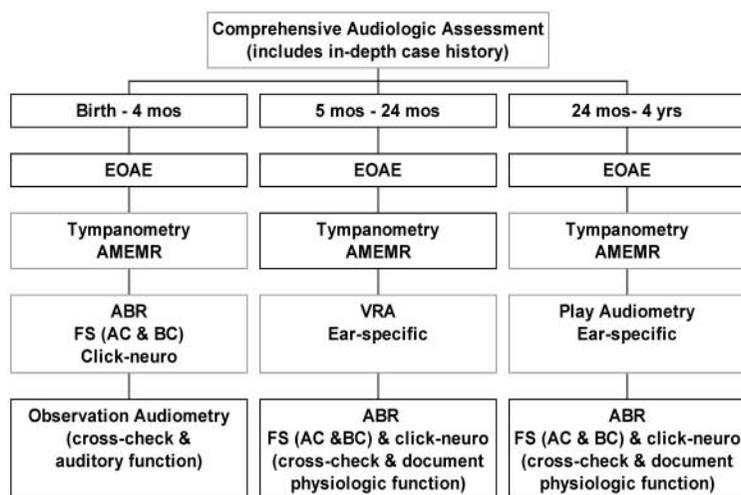


Figure 3. Audiological assessment battery recommended for use with neonates and young infants (birth – 4 months), infants and toddlers (5 months to 24 months) and young children (24 months to 4 years). Adapted from Gravel and Hood 1998.

of response properties from normal ears and ears with SNHL. The authors reported a relation between DPOAEs and audiometric threshold and demonstrated that generally, DPOAEs can be used to predict the magnitude of hearing losses less than about 60 dB HL. However, the authors stressed that it was not possible to select a criterion for which no errors occurred. Thus other measures (threshold ABR and/or behavioral audiometry), in addition to DPOAE, are needed to differentiate normal hearing from mild SNHL and mild hearing loss from moderate impairments.

Other tenets of the test battery approach include the following:

1. Follow test protocols that are evidence-based and time efficient.
2. Provide a comprehensive audiological test battery during the initial diagnostic evaluation.
3. Complete the entire test battery of behavioral and physiologic measures regardless of age.
4. Repeat all tests whenever there are concerns raised by parents regarding previous findings, or when the original audiological diagnosis is incomplete or incongruous with the child's present status.
5. Crosscheck behavioral results with electrophysiological tests (Jerger and Hayes 1976), and crosscheck electrophysiological results with behavioral measures of hearing status.

6. Expect more challenges in the audiological assessment of infants with developmental disabilities.
7. Obtain a complete case history, current medical status report, and query auditory responses, communication development and the overall impressions of parents; compare the audiological assessment results with the background data obtained.

An important principle of pediatric audiology is that no test in the battery be considered redundant. In these days of cost considerations and time constraints, adhering to this principle is sometimes a challenge. The audiologist should appreciate that each measure comprising the test battery yields a unique piece of information that no other test in the battery provides in the determination of auditory system integrity. If one adheres to this principle,

then every infant referred for comprehensive audiological assessment should receive the entire battery of tests before an audiological diagnosis is made. It is only then that the type, degree and configuration of a child's hearing loss can be determined or the unambiguous conclusion of normal hearing be drawn.

Are We Prepared?

A quality audiological assessment of infants and young children requires: (1) resources (within and outside the facility); (2) appropriate test equipment; (3) personnel trained and experienced in the use of current test technologies, procedures and protocols; (4) sufficient time to complete the audiological assessment, completely review the findings and answer the questions of parents; and as discussed, (5) adherence to the test battery approach to audiological assessment. Given the overwhelming increase in early identification programs, the question arises as to whether or not we are prepared to provide such comprehensive audiological assessments to infants identified as at risk for hearing loss through newborn hearing screening programs.

To address the issue of 'preparedness', a survey of 25 audiological facilities in New York City was completed (Gravel 1998). A questionnaire was sent to audiologist-heads of service. When no response was forthcoming, inquiries were made by telephone. All

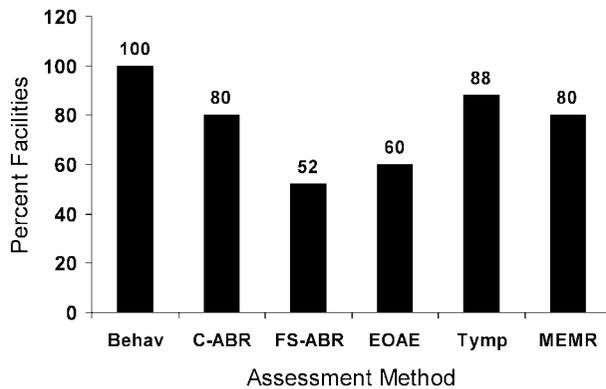


Figure 4. Results of a survey of 25 audiology facilities in New York City depicting the percentage of facilities reporting the use of various audiological tests with infants 12 months of age and younger. Behav = behavioral tests; C-ABR = click-ABR threshold assessment; FS-ABR = frequency-specific ABR threshold assessment; EOAE = evoked otoacoustic emissions; tymp = tympanometry; MEMR = acoustic middle ear muscle reflex assessment. (Gravel 1998)

respondents said that they provided audiological evaluations to infants 12 months of age and younger. Questions were asked regarding the availability and use of audiological assessment procedures and current technologies. Figure 4 presents the responses from the 25 facilities.

Note that all (100%) indicated that behavioral testing of infants 12 months and younger was routinely completed. However at the time of the survey, only 52% had the technology to provide frequency-specific ABR assessment and only 60% used otoacoustic emissions. Surprisingly, 88% of respondents said that middle ear function was examined in this age group through tympanometry and only 80% routinely determined acoustic MEMR thresholds. Given the approach to the audiological assessment of infants and young children advocated here, the findings of the survey presented above (although now three years old) give us pause as to the preparedness of audiologists and facilities to meet the current challenges of pediatric evaluation.

In their published recommendations, the Pediatric Working Group (formed following the October 1994 Conference on Amplification for Children with Auditory Deficits), stated that: “audiologists should have experience with the assessment of infants and children with hearing loss and the knowledge and equipment necessary for use with current pediatric assessment methods.” The Pediatric Working Group

further recommended that: “facilities that lack the expertise or equipment for assessing infants and children should establish consortial arrangements with those that do” (Bess et al. 1996). The notion of audiological referral facilities that enter a consortium agreement seems a reasonable solution when the entire audiological test battery cannot be completed at one facility. Regardless of how it is accomplished, however, it is stressed that the entire battery needs to be completed before ‘audiological certainty’ is reached.

Figure 5 presents an idealized schedule of audiological follow-up that might occur during the first year of life for an infant with hearing loss identified through newborn hearing screening (Gravel 2000). The interface of ongoing audiological assessments with the selection and fitting of amplification (including earmold changes and RECD measurements), and the initiation of early intervention are also displayed. Obviously multiple visits are required in order to: (1) completely characterize the audiogram (ear-specific thresholds from 250 through 6000 Hz); (2) provide audiological information for the refinement of the hearing aid fitting; (3) monitor the status of the hearing loss (detecting progression or fluctuation); (4) make appropriate referrals for medical management; and, (5) plan and validate intervention strategies. The figure emphasizes the time and resources required in order to deliver quality audiological services to infants with hearing loss and their families in the first year of life.

Summary and Conclusions

The need to provide accurate, reliable and comprehensive audiological assessments of infants and young children will become more important as early identification programs increase. The possibility of audiological misdiagnoses of various types exists because of audiological pitfalls that may be encountered in the clinical evaluation of hearing. Many audiological pitfalls can be avoided when the clinician adopts a test battery approach to audiological assessment (Gravel and Hood 1998), makes use of the crosscheck principle (Jerger and Hayes 1976), and continually examines the results for congruence among the measures. As caring and committed professionals, we must be vigilant about the possible audiological pitfalls that could be encountered during the confirmation of a child’s hearing loss, as well as

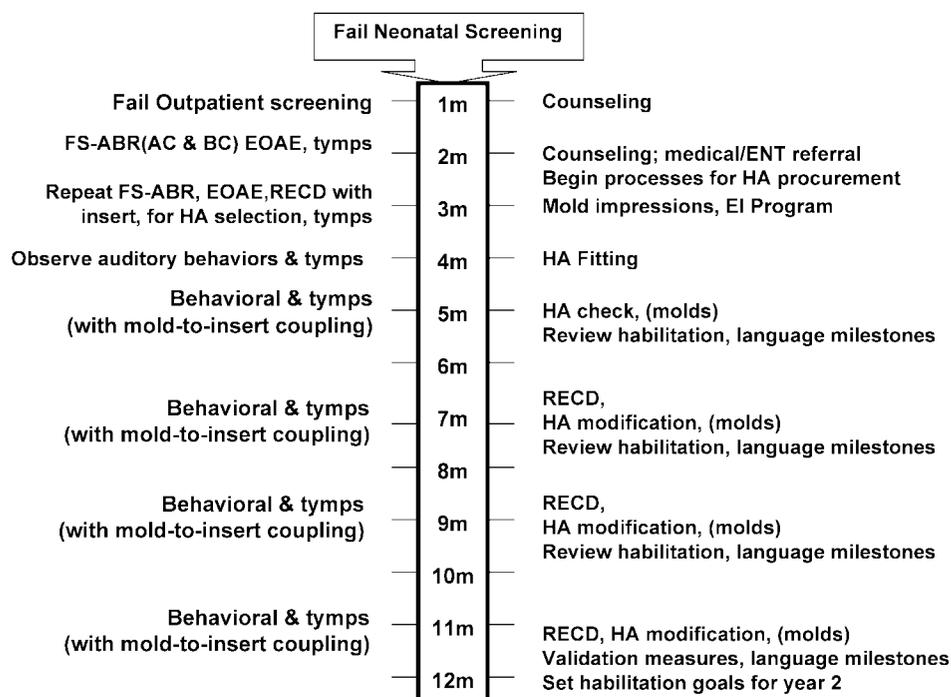


Figure 5. Schematic of the audiological follow-up of an infant in the first year of life following early identification of a permanent hearing loss through newborn hearing screening. On the left of the timeline (depicted in months [m]) are audiological follow-up measures; on the right, hearing aid (HA) selection and fitting including earmold fabrication (molds) and the measurement of the real-ear-to-coupler difference (RECD), leading to early intervention (EI). Audiological tests include: behavioral testing, tympanometry (tymps), evoked otoacoustic emissions (EOAE), frequency-specific ABR threshold assessment (FS-ABR) for air- (AC) and bone-conducted (BC) stimuli. Refer to the text for an explanation of mold-to-insert coupling.

during the audiological monitoring of that loss in the longer term. Our apprehension about committing an audiological misdiagnosis is justified based on our awareness of the potentially adverse consequences of such an error for the child and the family. Indeed, evidence now suggests that years of inappropriate intervention for some children may not be completely surmountable (Yoshinaga-Itano, Sedey, Coulter and Mehl 1998; Moeller 2000).

Today, there are still too few clinicians who have specific training or direct experiences with the audiological assessment of very young infants. Most professional training programs provide minimal academic preparation or direct clinical experiences specifically with the very young pediatric population. This has become a significant training and continuing education challenge for our profession.

Consider that in the not too distant past, audiologists would have expected, on average, to confirm the presences of moderate permanent hearing loss in a typically developing child by about 24 months of age

(Harrison and Roush 1996). Because of the increasing availability of NHS programs, we can now confirm the presence of a similar degree of congenital PHL by 5 months of age regardless of the infant's risk status, and by about 3 months of age for non-risk infants (Dalzell et al. 2000). Clearly, the age of the population that now requires timely and comprehensive audiological assessment to characterize the type, degree and configuration of the hearing loss, or to confirm the presence of normal hearing, has changed markedly in a relatively short period of time. Therefore, the requisite skills and knowledge of the pediatric audiologist must keep pace in order to best serve this very young group of children. If as professionals, we lack the formal training or educational background necessary to provide optimal audiological services to this young population, our responsibility is to acquire such skills and information rapidly. Only this will allow the pediatric audiologist to meet these exciting new challenges while avoiding the potential pitfalls that may be encountered in the comprehensive, on-going

audiological assessment of infants and young children with hearing loss.

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