Auditory System Development and Dysfunction: What Do We Really Know about Childhood Hearing Loss?

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Audiologists must have a thorough understanding of normal auditory system function in order to diagnose hearing impairment and dysfunction. This is particularly true for the pediatric audiologist who works with a very young patient who may provide limited behavioral information. This background knowledge should include familiarity with auditory anatomy and physiology, psychoacoustics, and speech perception. The information comes to audiologists from a wide range of both human and animal studies, from studies of developing and mature auditory systems, and finally, from studies of both normally functioning systems and systems with auditory pathology. The academic background of audiologists provides this information from the first undergraduate class through the final graduate courses. However, in everyday practice, it is the working view of what constitutes an impaired auditory system that determines what audiologists actually do.

The purpose of this article is to provide a framework for audiologists to guide their work with infants and children who may have hearing loss. An impaired auditory system may be characterized differently by the use of different criteria for dysfunction. Four different types of criteria will be discussed. These criteria include: a threshold criterion, a disease/dysfunction criterion, an audibility criterion, and an expanded audibility criterion. These criteria may be applied individually or together, depending upon the patient and his or her needs and presenting problems. In addition, the effects of auditory development on the use of these criteria in practice will be reviewed.

WHAT IS AN IMPAIRED AUDITORY SYSTEM?

The answer to this question will differ, depending upon the questioner and his or her frame of reference. The focus of an anatomist will be structural; the focus of a physiologist will be functional in relationship to the structures being monitored. The audiologist's view of an impaired auditory system will determine which diagnostic and rehabilitative procedures are used and why. In general, an audiologist might characterize an impaired auditory system as one that: 1) cannot detect sound at levels we have determined to be normal; 2) has one or more sites of lesion that interfere with auditory function; 3) does not have the entire speech spectrum available for processing; and 4) has difficulty functioning in a noisy environment. Each of these four characteristics of the impaired system also may be associated with one of the criteria for impairment noted above — a threshold criterion, a disease/dysfunction criterion, an audibility criterion, and an expanded audibility criterion, respectively.

Threshold Criterion

Measuring hearing that is normal or abnormal is one of the longest traditions in audiology. Pediatric audiologists attempt to determine threshold behaviorally for infants as young as five months of age with techniques such as Visual Reinforcement Audiometry (VRA) (Moore et al, 1977; Wilson and Thompson, 1984). Below five months, tech-
Techniques such as Behavioral Observation Audiometry (BOA) have been used; however, this approach has poor reliability within infants and great variability in results obtained in comparison of infants (Wilson and Thompson, 1984; Northern and Downs, 1991; Hayes and Northern, 1996). For infants from the neonatal period through the toddler stage, the use of auditory brainstem response (ABR) testing has provided a good estimate of behavioral threshold (Gorga, 1989; Werner et al, 1993). The use of ABR for threshold estimates is particularly important for infants when behavioral tests are unreliable in early infancy.

When applying this criterion, audiologists divide threshold into descriptive categories with threshold ranges in dB HL re ANSI (1989) specified. For example, Bess and Humes (1995) list the following categorization scheme: normal hearing between 0 and 25 dB HL, mild hearing loss between 26 and 45 dB HL, moderate hearing loss between 46-55 dB HL, moderately severe hearing loss between 56-70 dB HL, severe hearing loss between 71-90 dB HL, and profound hearing loss greater than 91 dB HL. More recently, Gelfand (1997) described a slightly different taxonomy: normal hearing 15 dB HL or less, slight hearing loss from 16 to 25 dB HL, mild hearing loss between 26 and 40 dB HL, moderate hearing loss between 41 and 55 dB HL; the remaining categories and hearing level descriptors remain the same as those proposed by Bess and Humes. For those who work with children with profound hearing loss, other auditory categories may be added. Boothroyd (1993) suggests three additional categories: those with considerable residual hearing or capacity (thresholds generally range between 90 to 100 dB HL), those with little residual hearing or moderate auditory capacity (thresholds generally range between 101 to 120 dB HL), and those with no residual hearing or auditory capacity (thresholds are 120 dB HL or greater).

The application of such categorization schemes has proved to be useful descriptively and is widely accepted. However, because each category is delineated with upper and lower numerical values that are contiguous, and the widest range of hearing levels is covered, the audiologist might assume a certain linearity of the hearing loss range. That is, one might infer that the severity of hearing dysfunction progresses in an orderly fashion, with each 10 dB of additional hearing loss causing approximately the same decrease in auditory function, regardless of the degree of hearing loss. However, Van Tasell (1993) has suggested that such a linear view of hearing loss may be misleading. She reviewed the results of two sets of studies — one addressing the selection of appropriate gain characteristics for adult listeners with different degrees of hearing loss (Byrne et al, 1990, 1991) and the other from animal research concerning the function of outer hair cells (OHC) in the process of normal and abnormal hearing (Patuzzi et al, 1989; Norton, 1992). Results of both sets of studies suggest that a hearing loss of 60 dB of cochlear origin is associated with some significant changes in behavior and function. Adults with sensorineural hearing loss of 60 dB or greater prefer more gain from their amplification than do adults with milder hearing losses (Byrne et al, 1990, 1991). Animals with hearing loss up to 60 dB have damage to the OHC only; beyond 60 dB HL, there is likely to be damage to the inner hair cells (IHC) as well (Norton, 1992). Thus, Van Tasell suggested that the function or dysfunction of the IHC causes a significant change in auditory capacity. Van Tasell further indicated that hearing loss greater than 60 dB is fundamentally different from hearing loss in the mild-to-moderate range and is likely to require changes in the types of prescriptive strategies used to select amplification. Alternatively, changes in the signal processing schemes themselves, such as the extraction or encoding of selected speech features, might be necessary, as described by Faulkner et al (1990) and Faulkner et al (1992).

Based on Van Tasell’s (1993) review of auditory function in relationship to amplification, audiologists may need to revise their application of a threshold criterion for evaluating children, especially if the evaluation leads to the selection of hearing aids. That is, the identification of hearing loss in excess of 60 dB HL should alert audiologists to the need for more careful evaluation of auditory function beyond simple detection.

Disease/Dysfunction Criterion

Another traditional approach taken by audiologists is to identify the site of lesion of a hearing loss by using a combination of behavioral and physiological measures. By careful use of audiometric testing, immittance testing, measurement of auditory brainstem responses (ABR) and otoacoustic emissions (OAEs), audiologists are able to determine the site of auditory dysfunction as the middle ear, cochlea, eighth nerve, or auditory brainstem with a reasonable degree of certainty.
(Champlin, 1996; Martin and Clark, 1996; Gelfand, 1997).

Before the introduction of OAEs, threshold criteria and disease/dysfunction criteria were essentially compatible in the assessment of infants with potential hearing losses. That is, the inability to measure an ABR was highly correlated with the measurement of a significantly elevated auditory threshold measured behaviorally. Both of these test results were considered evidence that there was dysfunction along the auditory pathway with the greatest likelihood of cochlear pathology. Recently, a new pathology has been identified that questions this type of assumption. Auditory neuropathy has been described as a condition in which there is the combination of grossly abnormal auditory evoked potentials with the presence of normal or near-normal behavioral thresholds (Starr et al, 1991; Sininger et al, 1995) or with the presence of normal OAEs (Stein et al, 1996). It would appear to be insufficient, based on the current study of auditory neuropathy, to use either a threshold or a disease/dysfunction criterion alone to evaluate infants and children.

Audibility Criterion

In recent years, the importance of the audibility of the speech spectrum for infants and children to develop normal speech and oral language skills has been emphasized repeatedly (Seewald et al, 1996; Stelmachowicz et al, 1996). In using this audibility criterion, audiologists have changed their concern from the detection thresholds of the infant or child per se to the impact that threshold loss has on the availability of speech spectrum cues to the listener. One metric, the Articulation Index (AI), is a numerical score that varies from 0 to 1.0 and is based specifically on the audibility of the spectrum of speech (ANSI, 1969; French and Steinberg, 1947). The AI has been used by many investigators to provide an indication of speech recognition ability in listeners with hearing losses (Dubno et al, 1989; Humes et al, 1986; Pavlovic et al, 1986).

Van Tasell (1993) discussed the relationship of AI with the estimated intelligibility of connected discourse. She reviewed the results of a study by Fabry and Van Tasell (1990), in which subjects with normal hearing listened in a variety of noise-masked conditions that resulted in a range of calculated AI values. The results of this experiment showed that the intelligibility of connected discourse increased rapidly with increasing AI. That is, the performance intensity function (AI as a function of rated intelligibility) had a particularly steep slope, so that small changes in AI resulted in large changes in rated intelligibility. This suggests that increasing the audibility of even a portion of the speech spectrum may have a very significant impact on perceived intelligibility. Interestingly, when the AI was calculated to be as low as 0.3, subjects judged the speech passages to be 100% intelligible, because of the highly contextual nature of this particular speech signal. That is, listeners did not need the entire speech spectrum to be audible for them to understand the meaning of the passages overall. When AI was reduced to 0.2, rated speech intelligibility decreased to 75%.

Figure 1 compares the available audible speech spectrum for two listeners. The left panel shows audibility for a listener with normal hearing, shown by the lower curve. The right panel shows audibility for a listener with a high frequency hearing loss, again shown by the lower curve. In both panels, the upper curve represents the speech peak levels and the hatched area represents the audible speech spectrum. The listener in the left panel has an AI of 1.0— all of the speech spectrum is audible. The listener in the right panel has an AI of 0.5— much of the speech spectrum above 1500 Hz is inaudible. According to the results of Fabry and Van Tasell (1990), the listener with the high-frequency hearing loss would be likely to rate connected passages as entirely intelligible, although the same listener would make a number of errors in a word recognition task with lower linguistic redundancy.

Van Tasell (1993) suggests that the AI, and thus the audibility approach, is appropriate for hearing loss of 60 dB or less. Once hearing loss exceeds 60 dB, other factors, such as broadened auditory filters, may influence speech recognition ability in addition to audibility (Rosen and Four-
Thus, audiologists should consider the application of the audibility criterion as an appropriate one for working with infants and children with hearing loss up to 60 dB HL. Beyond 60 dB HL, the increase in threshold, which suggests a dysfunction of IHC as well as OHC, will affect audibility in ways beyond those predicted by the AI model.

**Expanded Audibility Criterion**

Audiologists have long been concerned with the effects of background noise on speech recognition. Van Tasell (1993) demonstrated that the reduced recognition of speech in noise is directly related to the audibility of the speech spectrum. In Figure 2 the left panel shows the audibility of the speech spectrum for the same normal listener shown in the left panel of Figure 1. This time, the normal listener is asked to recognize speech in the presence of a spectrally shaped background noise at a signal-to-noise ratio of +15 dB. Once again, the hatched area represents the audible speech area. For this condition, the listener's AI was reduced to 0.5, a considerable decrease. However, if that same listener were presented with connected discourse and asked to rate its intelligibility, the rating would still be 100%, because one does not require total audibility to understand highly contextual connected discourse (Fabry and Van Tasell, 1990). In comparison, the listener with sensorineural hearing loss, shown in the right panel, shows a similar drop in AI, from 0.5 in quiet to 0.2 in noise. However, an AI of 0.2 would result in an estimated intelligibility rating of only 75%, a significant decrement in intelligibility. As Van Tasell (1993) explains, "...noise affects the hearing-impaired listener more drastically because he or she is already operating with a reduced-redundancy speech signal in quiet" (p. 36). In summary, for equal values of AI, noise-masked listeners with normal hearing and listeners with sensorineural hearing loss would demonstrate the same speech recognition performance. The problem with the hearing-impaired listener's performance in noise is that his/her AI will be poorer than the normal listener's AI in the same noise condition because his/her AI in quiet already was reduced as a result of the hearing loss.

Audibility in quiet does not provide the complete picture of speech recognition for listeners with sensorineural hearing loss. An expanded audibility criterion is necessary to account for the effects of noise. Because few real world listening situations are quiet, the application of the expanded audibility criterion is frequently necessary.

**WHAT ARE THE EFFECTS OF DEVELOPMENT?**

The impaired auditory system of an infant or child has all of the problems noted above for the adult system. In addition, in recent years, research in the area of developmental psychoacoustics and physiology has shown that the auditory systems of infants and children are developing rapidly and are different from the mature auditory system. A number of studies from a variety of laboratories, using different behavioral assessment techniques (VRA, forced-choice head turning, observer-based psychoacoustic procedures) have shown that the detection thresholds of infants are greater than adult thresholds by as much as 15 to 30 dB at 3 months of age and are still 10-15 dB greater than adult values at 6 months of age (Nozza and Wilson, 1984; Olsho et al, 1987; Olsho et al, 1988; Trehub et al, 1991; Werner, 1996). Differences in threshold are observed in children, particularly in the low frequency range, as late as 8 years of age (Elliott and Katz, 1980; Trehub et al, 1988). Developmental differences between infants, children, and adults also are observed for masking patterns (Nozza and Wilson, 1984; Schneider et al, 1989; Allen and Wightman, 1994), temporal resolution (Wightman et al, 1989), and frequency resolution (Olsho et al, 1982; Olsho, 1984; Allen et al, 1989; Allen and Wightman, 1992). In all these areas, infants and children have poorer discrimination abilities than do adults. Although nonsensory factors, such as attention and memory, must be accounted for in models of developmental psychoacoustics, it is clear that there is a real difference in auditory ability and capacity be-
between the developing infant and child and the mature adult (Werner, 1992, 1996).

There are developmental differences observed for speech stimuli as well as for non-speech stimuli. In studies of developmental speech perception, children’s speech discrimination abilities improve over time, even up to 10 years of age (Sussman and Carney, 1989; Sussman, 1993; Elliott and Hammer, 1993). Infant speech perception research, across different laboratories and language groups, has demonstrated that during the first year of life, children become sensitized to the subtle auditory cues in the speech of their linguistic community (Kuhl, 1991; Werker, 1991; Kuhl, 1994; Jusczyk, 1995). In particular, infants are able to make finer discriminations for individual speech sounds or groups of sounds when the phonetic content comes from the language they hear around them. Jusczyk et al (1993) also have demonstrated that infants prefer to listen to speech sound combinations that occur in their native language (i.e., native phonotactics), in contrast to combinations from other languages.

These behavioral differences also are accompanied by physiological differences between developing and mature systems. Immittance data from infants, particularly from multifrequency tympanometry, show a clear pattern of maturation from the neonatal period through later infancy, in which the tympanic membrane and ear canal walls appear more mobile than in adults (Paradise et al, 1976; Holte et al, 1991). These differences may be accounted for by the anatomical and physiological maturation of the outer and middle ear (Keefe and Levi, 1996). Differences in ear canal volume continue to be observed through 6 years of age for children with tympanostomy tubes postoperatively, possibly reflecting the process of pneumatization of the mastoid bone (Shanks et al, 1992). Similar developmental changes are observed for the measurement of ABRs from typically developing infants for both click and frequency specific tone bursts (Gorga et al, 1989; Werner et al, 1993; Jacobson and Hall, 1994; Stapells et al, 1994). Differences are observed in waveform morphology and latency from the neonatal to later infancy stages. In the measurement of OAEs, particularly for transient evoked otoacoustic emissions, larger response amplitudes and more high frequency components in the spectrum of the emission are observed for neonates and infants than for adults (Widen, 1997). Over the course of development, responses approach those of adults, although differences were observed for children as old as 13 years of age (Norton and Widen, 1990; Norton and Harrison, 1993).

WHAT IS THE COMBINED EFFECT OF AUDITORY IMPAIRMENT AND DEVELOPMENT?

A child’s impaired auditory system is defined by a threshold measure behaviorally that is really a minimum response level (Wilson and Thompson, 1984). Consequently, the application of a simple threshold criterion to determine hearing loss has some inherent error. The physiological measures used (e.g., immittance, ABR, OAEs), also are affected by the development of auditory structure and function, complicating the application of straightforward disease/dysfunction criteria in the diagnosis of hearing loss. The child with hearing loss must try to learn the phonetic cues of his or her linguistic environment with a limited speech spectrum. The application of a straightforward audibility criterion, using a method such as the AI, assumes that once the speech spectrum is available completely, the listener can use it for speech recognition. This assumption may not hold for children initially. Phonetic learning, the first stage in oral language acquisition, can begin only when the spectrum of speech is audible. However, phonetic learning is a lengthy process and continues throughout childhood, even for children with normal hearing (Elliott and Hammer, 1993; Sussman and Carney, 1989; Sussman, 1993). The calculated AI based on the fitting of appropriate amplification represents a long-term goal AI rather than an immediate index of audibility. Any application of threshold or audibility criteria should be made with the reservation that hearing losses greater than 60 dB HL reflect different types of cochlear dysfunction than do milder hearing losses and may have particular interactions with development that are as yet unknown (Van Tasell, 1993). Information about changes in AI for children as a function of the addition of background noise is scarce. At present, research directed at the effect of noise on children’s speech perception with normal and impaired hearing suggests that children may be more susceptible to the effects of noise than adults, particularly after some auditory deprivation (Gravel and Wallace, 1992). Further research is indicated for us to understand the interaction of audibility and developmental processes.

Each pediatric audiologist should examine his or her assumptions about the threshold, disease/dysfunction, and audibility criteria that she or he
uses when assessing children and fitting them with amplification. Changes in auditory and speech perception skills should be assessed in light of normal developmental processes as well as an understanding of auditory dysfunction.

REFERENCES


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