

# *Evaluation of Hearing Loss in Childhood*

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## **1. Introduction**

Unidentified hearing loss in children, either congenital or acquired, may result in lifelong deficits in speech and language abilities<sup>1</sup> which, in turn, impact on communication skills, cognition, educational achievement, emotional development and psychosocial wellbeing.<sup>2</sup> Such delays may result in lower levels of education and employment in adulthood<sup>3</sup> and, consequently, increased overall lifetime cost that can exceed US\$1 million for those with profound hearing loss, including educational services, social services, and reduced work-productivity.<sup>4</sup> Cost is directly proportional to the severity of hearing loss and inversely related to language abilities.<sup>5</sup> Even children with mild or unilateral hearing loss may have lower academic skills.<sup>6</sup>

On the other hand, early detection and intervention within the critical period of development of speech, language and cognition improves the outcome<sup>1</sup> regardless of the magnitude of the hearing loss.<sup>7</sup> Therefore, it is recommended that all infants should be screened at no later than one month of age, and all infants with hearing impairment must be identified before three months of age. Infants with confirmed hearing loss should receive appropriate intervention at not later than six months of age.<sup>8</sup> Formerly, investigative data showed that parents of infants without risk factors first suspected hearing loss in those infants by the age of 15 months, and it was confirmed by 22 months of age.<sup>9</sup> Since the introduction of universal neonatal hearing screening, the average age of hearing loss detection has decreased to two to three months of age.<sup>10</sup>

Most children with congenital hearing loss are identified by the hearing screening. However, hearing loss in children may be of delayed onset, progressive or acquired. For that reason, if one or more risk indicators (**Table 1**) are present, a complete audiologic assessment must be performed at 24 to 30 months, regardless of the newborn screening results.<sup>8</sup>

**Table 1.** American Academy of Pediatrics Joint Committee on Infant Hearing Year 2007 Position Statement: Risk Indicators Associated With Permanent Congenital, Delayed-Onset\*, or Progressive Hearing Loss in Childhood

1. Caregiver concern* regarding hearing, speech, language or developmental delay.
2. Family history* of permanent childhood hearing loss.
3. Neonatal intensive care of more than five days or any of the following, regardless of length of stay: Extra-corporeal Membrane Oxygenation (ECMO),* assisted ventilation, exposure to ototoxic medications (gentamicin and tobramycin) or loop diuretics (furosemide), or hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as CMV,* herpes, rubella, syphilis, or toxoplasmosis.
5. Craniofacial anomalies including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.
6. Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss,* such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred, and Jervell and Lange-Nielsen.
8. Neurodegenerative disorders,* such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome.
9. Culture-positive postnatal infections associated with sensorineural hearing loss,* including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull / temporal bone fracture * that require hospitalization.
11. Chemotherapy*
12. Recurrent or persistent otitis media for at least three months

Source: Joint Committee on Infant Hearing Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics* 2007;120: 898–921

Besides, all infants should receive ongoing monitoring of age-appropriate auditory (**Table 2**) and communication skills (**Table 3**) as well as developmental milestones (**Table 4**), regardless of risk indicators or hearing screening outcomes.<sup>8</sup> Any parental concern about hearing or delay in communication must be taken seriously. Identification of any abnormality has to be proactive and consistent for early diagnosis. The regular surveillance of developmental milestones and auditory skills as well middle-ear status should be performed periodically. Speech and language present rapid evolution in the first three years of age so, it is appropriate to wait about one month or two, but no more than three months, to get an audiological assessment to rule out hearing loss.

**Table 2.** Guidelines for Children with Suspected Hearing Loss

Age (Months)	Normal Development
0–4	Should startle to loud sounds, quiet to mother’s voice, momentarily cease activity when sound is presented at a conversational level
5–6	Should correctly localize to sound presented in a horizontal plane, begin to imitate sounds in own speech repertoire or at least reciprocally vocalize with an adult
7–12	Should correctly localize to sound presented in any plane, should respond to name, even when spoken quietly
13–15	Should point toward an unexpected sound or to familiar objects or persons when asked
16–18	Should follow simple directions without gesture or other visual cues; can be trained to reach toward an interesting toy at midline when a sound is presented
19–24	Should point to body parts when asked; by 21 months can be trained to perform play audiometry

Source: Matkin ND. *Pediatr Rev.* 1984;6:151.

**Table 3.** Guidelines for Children with Abnormal Speech-Development

Age (months)	Referral Guidelines for Children with Speech-Delay
12	No differentiated babbling or vocal imitation
18	No use of single words
24	Single-word vocabulary of $\leq 10$ words
30	Fewer than 100 words; no evidence of 2-word combinations; unintelligible
36	Fewer than 200 words; no use of telegraphic sentences; clarity $< 50\%$
48	Fewer than 600 words; no use of single sentences; clarity $\leq 80\%$

Source: Matkin ND. *Pediatr Rev* 1984;6:151.

**Table 4.** Developmental Milestones for Communication and Language

Milestone	Average Age of Attainment (Months)	Developmental Implications
Smiles in response to face, voice	1.5	Child more active social participant
Monosyllabic babble	6	Experimentation with sound, tactile sense
Inhibits to “no”	7	Response to tone (nonverbal)
Follows 1-step command with gesture	7	Nonverbal communication
Follows 1-step command without gesture (e.g., “Give it to me”)	10	Verbal receptive language
Speaks first real word	12	Beginning of labeling
Speaks 4–6 words	15	Acquisition of object and personal names
Speaks 10–15 words	18	Acquisition of object and personal names
Speaks 2-word sentences (e.g., “Mommy shoe”)	19	Beginning grammaticization, corresponds with vocabulary of $\geq 50$ words

Modified from: Behrman RE, Jenson HB, Kliegman R, eds. *Nelson Textbook of Pediatrics*. 16th ed. Philadelphia, PA: Saunders; 2003.

The clues for suspicion of hearing loss are distinct among age-ranges in children. Until the child reaches age three, parents often report inattention, erratic response to sound, or speech delay. From three to six years of age the speech is not clear or is “twisted.” After six years of age the child has learning-difficulties. Delayed or atypical communication skills in childhood may be due mainly to hearing loss, aphasia, mental retardation, disorders within the spectrum of autism, and affective and behavioral disorders. A child with hearing impairment, unlike other communication disorders, prefers gestural communication and plays normally with other children from the same age-group.

Instead of informal hearing-screening such as parents reports or informal behavioral assessment in the physician’s office, formal audiological evaluation is the only way to attest to normal hearing. Assessment of hearing in childhood may employ the following methods:

A) Behavioral measures

- Behavioral observation without reinforcement: observation of gross changes in infants’ behavior in response to auditory signals from noisemakers such as drums, cymbals, bells, and rattles, among others. Indicated from birth to six months of age
- Visual reinforcement audiometry: localization of the sound source on a speaker or earphones, reinforced by an attractive visual stimulus. For young infants older than six months of age or with neurologic or psychiatric disorders.
- Conditioned-play audiometry: after three years of age
- Vocal audiometry

However, behavioral estimates of hearing sensitivity cannot always be obtained in young children, and besides, they usually do not assess each ear separately. Thus, the initial battery of audiological tests to confirm hearing loss in infants must include physiologic measures, followed by behavioral methods according to the child’s age.

B) Physiologic measures

- Otoacoustic emissions
- Tympanometry
- Stapedial reflex
- Auditory Brainstem Responses (ABR)
- Electrocochleography (ECoG)
- Auditory Steady-State Responses (ASSR)

## 2. Otoacoustic Emissions

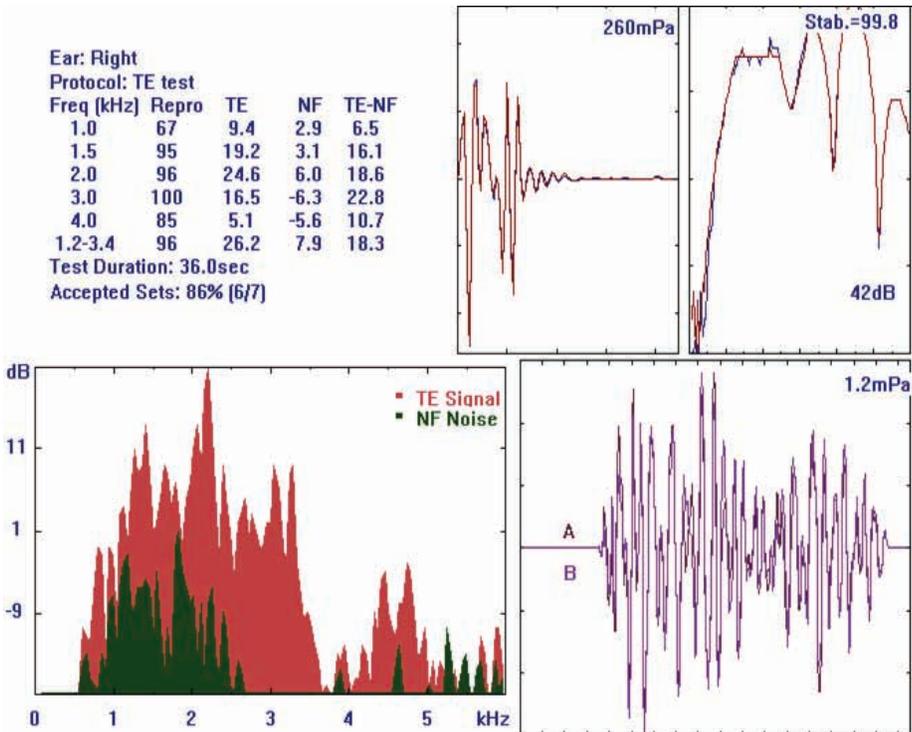
Otoacoustic emissions (OAE) are low-intensity acoustic signals registered in the external auditory canal,<sup>11</sup> generated by the non-linear mechanical activity of outer hair cells of the organ of Corti,<sup>12</sup> either spontaneously or in the presence of acoustic stimulation. Spontaneous OAEs are not considered a suitable clinical indicator of cochlear activity, since they are present in only 40-60% of subjects

with normal peripheral hearing.<sup>13</sup> Evoked OAEs are classified according to the type of stimulus used during measurement: stimulus frequency, transient, or distortion product.

### 2.1. Transient Evoked Otoacoustic Emissions (TEOAEs)

TEOAEs are responses collected a few milliseconds after acoustic stimulus presentation. The probe must contain a transducer to deliver a click or tone-burst stimulus and a microphone to detect the response, and its correct fitting into the ear canal is mandatory. Emissions can be evoked from the normal cochlea at most frequencies, depending on the broad spectrum of the stimulus.<sup>14</sup> Broadband clicks are usually used for measuring TEOAEs, which are registered in a time-length parameter. Typically, the high-frequency components of the response have shorter latency than lower frequencies. Responses to several stimuli are averaged to distinguish responses from the noise-floor, and in this way analysis of responses depends on the wave reproducibility, and on the signal-to-noise ratio (**Figure 1**).

**Figure 1.** “Pass” on the TEOAE test. The responses are robust and above the noise-floor, with high wave-reproducibility.



Extracted from authors' image-bank

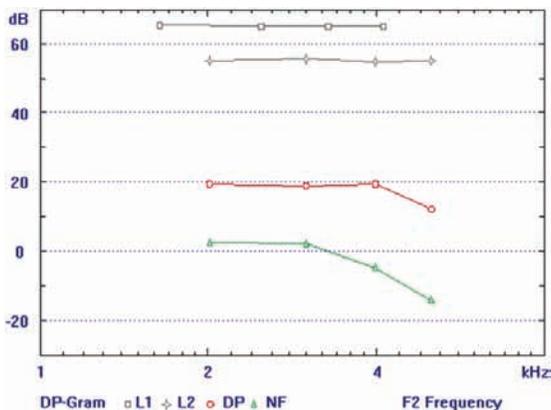
The TEOAE test is the most widely used OAE method for neonatal hearing screening, as it can be recorded in nearly all subjects with a hearing threshold 30-40 dB HL, or worse. Besides, TEOAEs obtained in newborns are uniform in the band-frequencies of 1 kHz to 5 kHz, with greater amplitude when compared to those for adult subjects.<sup>14</sup> Furthermore, this test is a fast, non-invasive, and objective method for auditory evaluation.<sup>15</sup> However, TEOAEs have limitations. In order to obtain accurate results, the recordings must be made in quiet environments. Background noise can significantly alter the false-positive rate results and test duration. Absent responses provide no information about hearing thresholds or type of hearing loss (conductive or sensorineural). Present responses cannot predict frequency-specific hearing levels from the spectrum of emissions.

## 2.2. Distortion-Product Otoacoustic Emissions (DPOAEs)

DPOAEs are elicited by the simultaneous presentation of two pure tones ( $f_1$  and  $f_2$ ), which are offered simultaneously to the cochlea and will generate different tones that can be measured as responses in the external ear canal. These tones, known as distortion-products, differ in frequency and amplitude from the primary pure tones and reflect the non-linear functioning of the cochlea. The largest distortion-products are obtained at the frequency of  $2f_1-f_2$ . For the measurement of DPOAEs, two separate channels for stimuli generation are necessary, and both must be electrically isolated to prevent artifacts.

The emissions can be measured and displayed in two forms. The DP-gram depicts the DPOAE amplitude as a function of stimulus frequency at a fixed loudness level (**Figure 2**), and the DP growth-rate (I/O function) plots growth of the DPOAE amplitude at a constant frequency and variable loudness levels of the pure tones.<sup>16</sup> Just as with TEOAEs, measurements of DPOAE are analyzed by means of the signal-to-noise ratio. The measured DPOAE level should be at least 6 dB above the noise-level.

**Figure 2.** DPOAE displayed in the DP-gram. Significant responses were recorded in the spectrum from 2 kHz to 5 kHz. (Extracted from authors' image-bank)



DPOAEs can be recorded in almost all ears with normal hearing and can also be measured in ears with mild to moderate sensorineural hearing loss.<sup>17</sup> Although the OAE is not a direct measure of hearing sensitivity, there is a quantitative correlation with the degree as well as the configuration of hearing loss, when it is mild to moderate.<sup>18</sup> Another advantage of this

method is the frequency specificity of the pure tone stimuli used in this test. The distortion-product, as well as the TEOAE, depends on optimal transmission through the middle ear, so it is important to determine middle ear status before interpreting findings.<sup>14</sup>

### **2.3. Interpretation of OAEs**

The presence of evoked OAEs denotes integrity and adequate function of outer hair cells, and therefore their absence may indicate abnormal cochlear function, consistent with hearing loss. However, their absence does not mean just alteration of cochlear function. Both the stimuli and the OAEs are transmitted through the middle ear and recorded in the external ear canal, so middle ear diseases or occlusion of the external ear canal can lead to abnormal responses.<sup>19</sup> Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells.

Regarding the external ear, even small quantities of cerumen or debris in the ear canal can block the probe-channels, with consequent inadequate stimulation. In newborns, especially during the first days of life, the OAE fail-rate is higher due to the presence of vernix caseosum in the external ear canal, amniotic fluid in the middle ear, and/or negative pressure in an unventilated middle ear cavity.<sup>19, 20</sup>

Adequate middle ear function is also a determinant factor in analysis of OAE, and several conditions like Eustachian tube dysfunction, otitis media with effusion, tympanic membrane perforation, or ossicular chain defects can interfere in its measurement.<sup>21</sup> The OAE test-result alone is not able to differentiate among abnormalities of the external ear, middle ear, or outer hair cells. In case of a negative test-result, other auditory tests should be performed to assess middle ear status (tympanometry, stapedial reflex measures) or to assess the inner ear and auditory pathways (ABR).

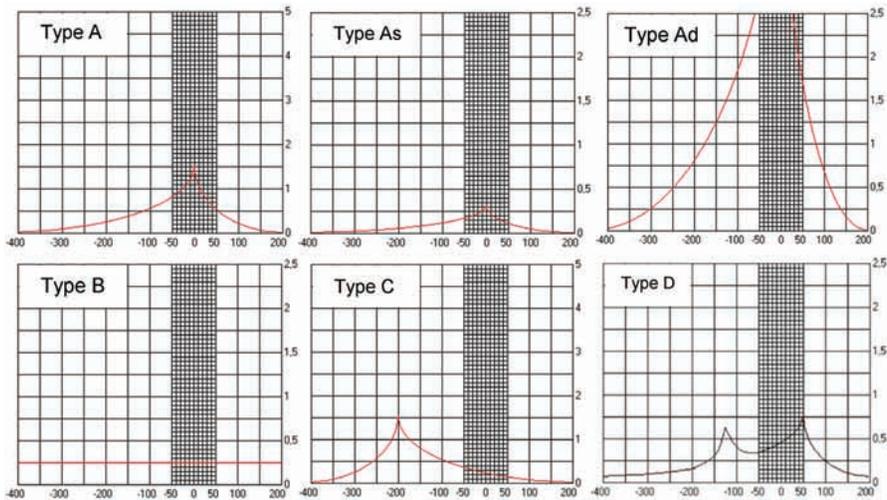
We should keep in mind that OAEs also do not assess the integrity of auditory pathways from the eighth nerve to the brainstem, and therefore will miss auditory neuropathy/dyssynchrony and other neural abnormalities.

### **3. Tympanometry**

Tympanometry evaluates variations in energy-transmission secondary to changes of pressure in the external ear canal, reflecting changes of the physical properties of the tympanic membrane, middle ear, and ossicular chain.<sup>22</sup> The equipment measures sound-pressure in the hermetically sealed external ear canal by the emission and reception of acoustic signals. During the examination, the tympanic membrane undergoes various degrees of positive and negative pressure, modifying its position and consequently absorption of the continuous emitted acoustic signal. Due to the low resonance frequency of the neonatal middle ear,<sup>23</sup> the use of probe-tones at 1000 Hz is recommended for patients up to the age of six months.<sup>24, 25</sup> Through this test, it is possible to verify whether the tympanic membrane shows normal movements and whether air-pressure is similar in the middle ear and the external ear canal, indicating proper functioning of the auditory tube and middle ear structures and integrity of the ossicular chain.

Based on the graphic representations of the tympanometry, it can be classified into the following types (**Figure 3**):

**Figure 3.** Tympanograms. **A:** normal Eustachian tube function with normal mobility of the tympanic membrane and middle ear structures; observed in ears with normal hearing or sensorineural hearing loss. **As:** stiffness of the tympanic membrane or ossicular chain; observed in ossicular chain fixation, otosclerosis, tympanosclerosis, and tympanic membrane thickening. **Ad:** high mobility of middle ear structures; seen in ossicular chain disruption or flaccid tympanic membrane. **B:** reduced compliance or minimal variation during the examination; observed in alterations of the external or middle ear such as cerumen obturans, otitis media (acute otitis media, otitis media with effusion, adhesive otitis media, cholesteatoma), some congenital malformations, and probe occlusion. **C:** negative pressure inside the middle ear; noted in episodes of Eustachian tube dysfunction and otitis media with reduced quantity of effusion. **D:** Double peak (“W” pattern); in newborns suggests low resonance frequency.



Extracted from authors' image-bank

Since this test is fast and easily performed, does not demand behavioral responses, and is unchanged during sleep or sedation, it is an important tool for auditory evaluation of infants, especially when they are referred for diagnostic audiological assessment following newborn hearing screening. Therefore, it is recommended in newborns who failed the otoacoustic emission test, because it helps in the differential diagnosis of sensorineural hearing loss and middle ear involvement, common in childhood.

#### 4. Stapedial Reflex

The stapedial reflex corresponds to the involuntary contraction of the stapedial muscle in response to high intensity sound stimulation. The contraction occurs bilaterally, even when the sound is presented unilaterally. So, the reflex can be obtained in the ipsilateral or contralateral ear.

The acoustic reflex pathway is composed of the tympanic membrane, middle ear, cochlea, vestibulocochlear nerve, brainstem, and facial nerve. The stapedial reflex testing allows for the gathering of information not only from the middle

ear but also from the auditory pathways of the brainstem. Thus, both afferent and efferent auditory pathways as well as the facial nerve must be working in order to observe the reflex. Besides complementing information obtained from tympanometry, the stapedial reflex test allows evaluation of the overall integrity of the peripheral and central auditory pathways.

During stapedial reflex threshold testing, the subject must remain quiet, because muscular artifacts may impair the accurate uptake of responses. Stimulation with pure tones in several frequencies and intensities may elicit contraction of the stapedial muscle, resulting in a change of compliance that can be detected by a probe in the external ear canal. Subjects with normal hearing present a stapedial reflex at about 70 to 95 dB HL above the hearing threshold. Abnormal stapedial reflexes are found in: 1) sensorineural or conductive hearing loss; 2) efferent impairment, such as middle ear disease or peripheral facial palsy; 3) brainstem injury; 4) auditory neuropathy/dyssynchrony (AN/AD). The stapedial reflex threshold may also predict hearing thresholds in young infants with normal middle ears, who are unable to undergo a behavioral assessment.<sup>26</sup>

## 5. Auditory Brainstem Responses

Auditory brainstem responses (ABR) are scalp-recorded electrical potentials elicited during the first 10 to 20 milliseconds following the onset of a transient acoustic stimulus. The responses are generated by the auditory nerve and brainstem<sup>27</sup> and are represented through waves, labeled sequentially in Roman numerals.

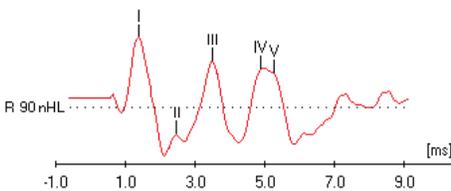
### 5.1. Click-evoked ABR

ABR are best generated with very brief stimuli presenting almost instantaneous onset. This rapid onset generates synchronous firing of numerous auditory neurons. At high intensities a wide region of the basilar membrane is activated, and hair cells of extensive frequency-ranges respond to this broadband stimulus. So no frequency specificity can be expected from this test.

The main purpose of click-ABR is to evaluate neural function, auditory brainstem integrity, and maturation of auditory pathways and to define the type of hearing loss (conductive, sensorial, neural). Click-ABR is also used with automated ABR systems for hearing-screenings in newborns.

Waves I and II are thought to represent activity from the cochlear nerve, wave III from the cochlear nucleus, and waves IV and V to represent brainstem activity at the lateral lemniscus up to the inferior colliculus<sup>28</sup> (**Figure 4**).

**Figure 4.** ABR waves (Extracted from authors' image-bank)

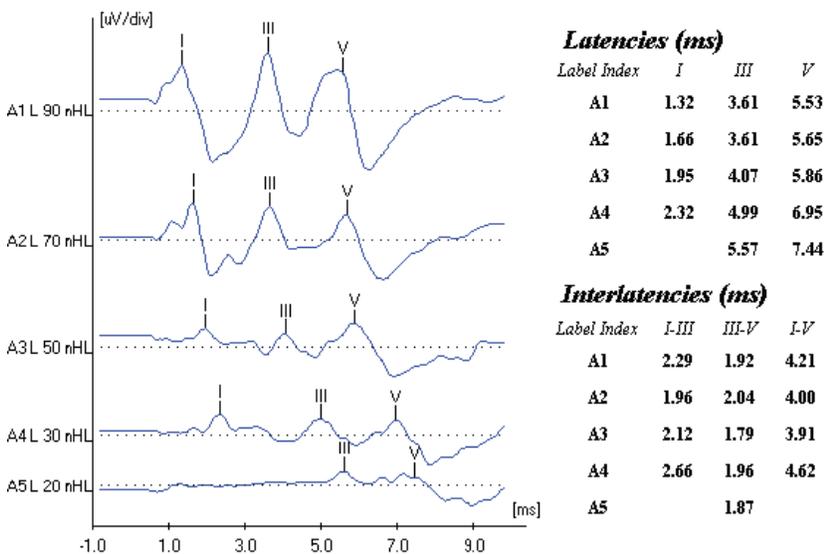


Latencies and interwave latencies are age-dependent, increased in newborns and infants. The cochlea may be fully functional by about 35 weeks of conceptional age, but ABR waveform is incomplete at birth and is

certainly immature for preterm infants. Therefore, estimation of conceptional age is crucial for appropriate interpretation of waveforms and latencies. Interwave latencies among waves I-V are prolonged (up to 5.0 ms) and decrease during the first months. Immature synaptic functioning, reduced axon diameters, and incomplete myelination of nerve fibers could explain all these alterations, as auditory brainstem pathways are undergoing maturation up to 18-24 months. After this age, the ABR is adult-like in latency and amplitude, as is shown in auditory developmental studies.<sup>29</sup> Even after the age of two years, developmental delays may show a prominent wave I and decreased amplitude of wave V, poor waveform morphology, and increased interwave intervals for waves I-V, I-III or III-V.

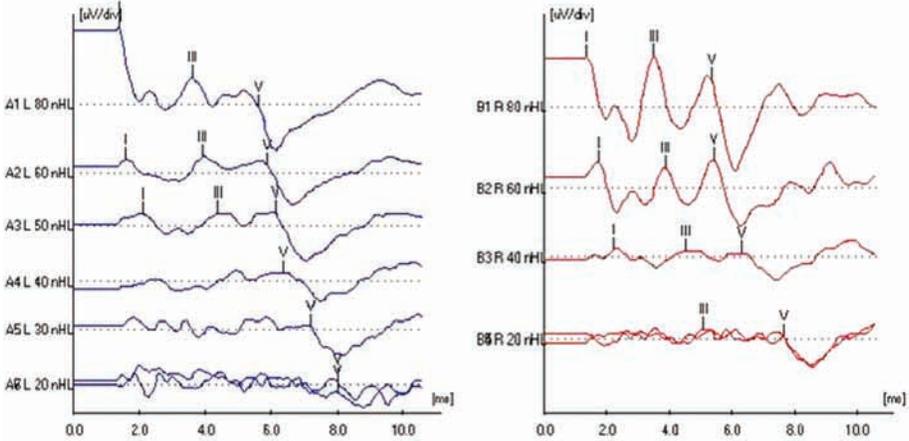
In subjects with normal hearing at high intensity levels, to 75 to 95 dB nHL, wave V latency is about 5.5 to 6.0 ms and increases about 2.0 ms at threshold level. Wave I latency is about 1.5 ms, wave III about 3.5 ms, and interwave latency values are approximately 2.0 ms. At lower intensities latencies of all waves increase, and wave I may be undetectable. Threshold is defined as the lowest intensity where a reliable wave V can be detected (**Figure 5**). At threshold, various frequencies may contribute to the response, but usually the best correlation is within the frequency range of 2-4 kHz. When the pure tone audiogram is normal or shows flat hearing loss, click-ABR thresholds are close to behavioral hearing thresholds, but in unusually shaped or sloping hearing loss, any frequency from 1 to 4 kHz may contribute to the threshold but may underestimate hearing loss<sup>30</sup> (**Figure 6**).

**Figure 5.** Normal click-ABR recorded from 90 dB HL to threshold (20 dB HL) from the left ear of a 12-month-old girl. Waves I, III, and V are well defined with age-appropriate latencies and interwave latencies.

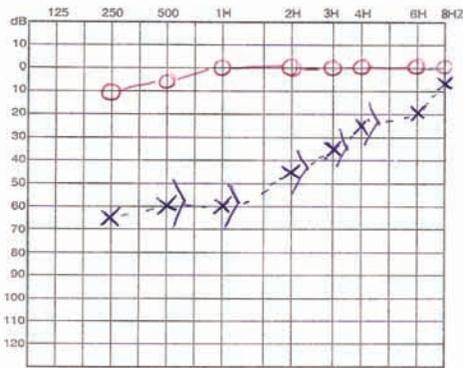


**Figure 6.** 7-year-old girl. Click-evoked ABR shows normal latencies and thresholds at 20 dB of hearing despite significant low- and middle-frequency sensorineural hearing loss in the left ear.

**D.B.M.L., 7 years old, female**



<i>Latencies (ms)</i>					<i>Interlatencies (ms)</i>			
<i>Label Index</i>	<i>I</i>	<i>II</i>	<i>III</i>	<i>IV</i>	<i>Label Index</i>	<i>I-III</i>	<i>III-V</i>	<i>I-V</i>
<b>Left</b>								
<b>A1</b>	1.37		3.58		<b>A1</b>	2.21	2.04	4.25
<b>A2</b>	1.58		3.91		<b>A2</b>	2.33	1.96	4.29
<b>Right</b>								
<b>B1</b>	1.33		3.50		<b>B1</b>	2.17	1.83	4.00
<b>B2</b>	1.75		3.87		<b>B2</b>	2.12	1.54	3.66

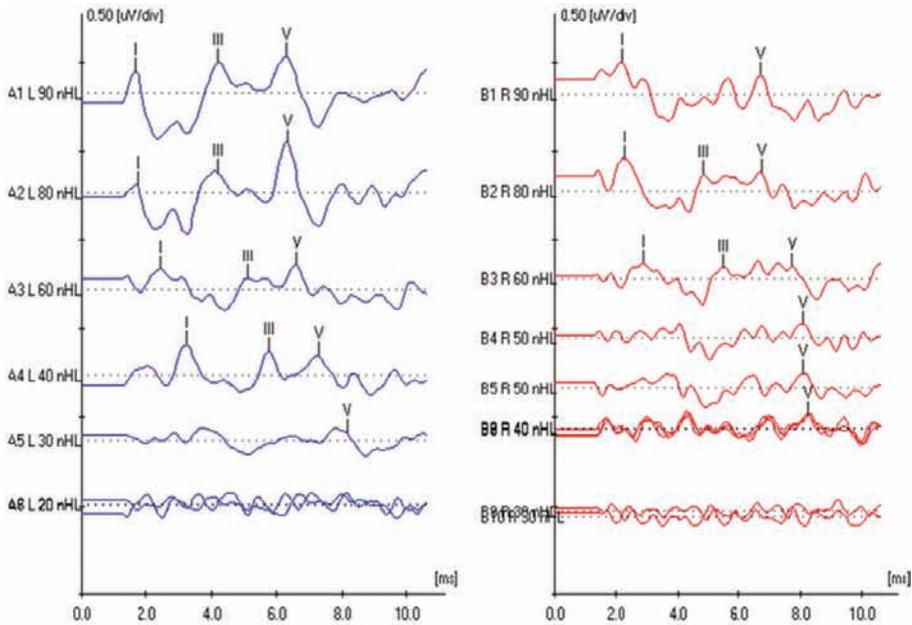


Extracted from authors' image-bank

In conductive hearing loss all latencies are increased, but the I-V interval is maintained. Poor wave forms may be seen, and threshold is elevated (**Figure 7**). On the other hand, an increased wave V latency and I-V interwave latency is the main alteration found in retrocochlear or brainstem dysfunction, associated or not with sensorineural hearing loss. In

these cases, wave I may be normal. Sensory hearing loss, with preserved neural function, usually shows normal latencies and interwave latencies but elevated thresholds.

**Figure 7.** Conductive hearing loss, right ear. Note that latencies in the right ear are increased, but interwave latencies are normal



**Latencies (ms)**

Label Index	I	II	III	IV	V
<b>Left</b>					
A1	1.67		4.21		6.29
A2	1.71		4.21		6.33
<b>Right</b>					
B1	2.21				6.70
B2	2.29		4.83		6.75

**Interlatencies (ms)**

Label Index	I-III	III-V	I-V
<b>Left</b>			
A1	2.54	2.08	4.62
A2	2.50	2.12	4.62
<b>Right</b>			
B1			4.50
B2	2.54	1.92	4.46

Extracted from authors' image-bank

**5.2. Frequency-specific ABR**

Frequency-specific responses are a keystone of diagnostic evaluation. Different tools are available like tone-burst ABR, Logon ABR, auditory steady state responses with various stimulus paradigms (AM / FM or exponential modulated pure tones). Tone-burst ABR is one of the most popular, widely available tests, included in the standard package of most manufacturers' equipment.

The stimulus is a gated sinusoid whose center frequency determines the nominal frequency, for example the 1 KHz tone-burst. There is some spread to

adjacent upper and lower frequencies, and non-linear gating functions are used to circumvent spectral energy splatter. Basically, this test is used to determine frequency-specific electrophysiologic thresholds at the main center frequencies (0.5, 1, 2 and 4 KHz). Therefore, wave V is followed from higher to lower intensities at each frequency, and like click-ABR testing, threshold is obtained at the lowest intensity where wave V is reproducible.

This test should not be used for high intensity neurologic testing. Whereas the 4 KHz tone-burst latencies are very close to click-ABR latencies, all lower tone-burst frequencies have longer latencies than those expected for clicks. Since frequency-specific waves contain responses of a reduced number of auditory fibers, amplitude of wave V is usually small. Sometimes averaging of more than 3000 to 4000 stimuli may be necessary to obtain a reliable threshold response.

Tone-burst thresholds are usually 10 dB worse than behavioral thresholds, but at 0.5 KHz this difference may be 15 to 20 dB. This is especially true for subjects with normal or near normal hearing. Differences between tone-burst and behavioral thresholds are smaller or may disappear as sensorineural hearing loss increases. In other words, tone-burst ABR predicts better frequency-specific thresholds in patients with moderate or severe hearing loss than in normal hearing subjects.

The main restriction to tone-burst ABR is the test duration. To date it has not been possible to record simultaneously responses from multiple frequency-specific stimuli in both ears (like ASSR), so each frequency has to be tested at various intensities down to the threshold in one ear, then the same procedure started in the other ear. Even experienced examiners, using age-appropriate stimulus rates and sufficient but not excessive averaging at each intensity, may need one to two hours to obtain all relevant four-frequency-threshold information from both ears. In young infants in natural sleep, sometimes a second session is necessary to complete the frequency-specific test. When older children are tested, sedation with chloral hydrate or midazolam may not assure perfect conditions for extended testing time. General light anesthesia is our first choice for children above six months of age. In most cases we are able to collect all relevant frequency-specific information during one testing session.

### **5.3. Bone-conduction ABR**

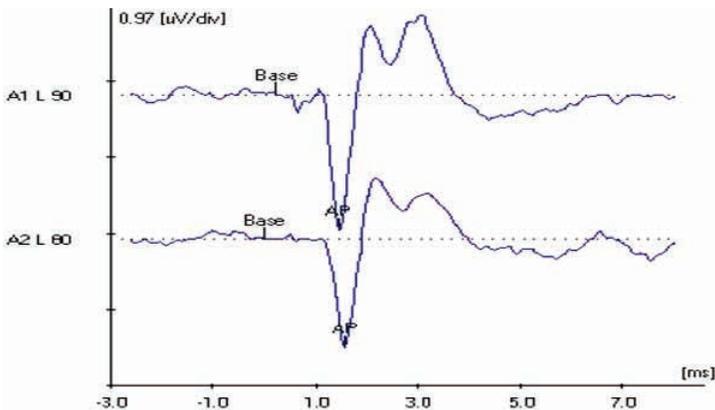
This test can be performed using click- or tone-burst stimuli. The stimulus is offered by a bone-vibrator placed on the mastoid, similarly to as it is in conventional audiometry. The main indications are occlusion or stenosis of the external ear canal, malformations of external or middle ear structures, and sometimes in cases of persistent otitis media with effusion. It is helpful to estimate the air-bone gap in conductive hearing loss. Therefore, this test should never be performed alone, but as an adjunct to conventional ABR air-conduction evaluation. When stenosis or atresia of the external ear canal is present, the more popular inserted earphones are not suitable for air-conduction testing. In this case, examiners should use the traditional TDH39 headphones to estimate air-conduction thresholds. Bone-conduction ABR provides important information to distinguish between pure

conductive or mixed hearing loss and is indispensable for rehabilitation of little patients, especially when a BAHA (bone-anchored hearing aid) is considered.

### 6. Electrocochleography

Electrocochleography (ECoChG) allows a near-field recording of electric activities of the cochlea and the auditory nerve. The major cochlear components are the cochlear microphonic (CM) and the summing potential (SP). The action potential (AP) represents electrical activity from the auditory nerve (equivalent to ABR wave I) (**Figure 8**).

**Figure 8.** Normal ECoChG



Extracted from authors' image-bank

Responses may be obtained by transtympanic recording (TT ECoChG), an invasive technique where the electrode is placed on the promontorium, or by non-invasive placement of the electrode on the tympanic membrane (TM ECoChG) or in the external auditory canal. The closer the electrode is to the cochlea, the larger are the response amplitudes. This makes the technique attractive when no reliable ABR can be obtained. In these cases, ECoChG may help to identify auditory thresholds and the site of lesion (middle ear, cochlea, or auditory nerve) defining the type of hearing loss: conductive, sensory or neural. ECoChG is especially helpful in children with persistent middle ear effusion and unreliable responses or very elevated thresholds on ABR, worse than expected for pure conductive hearing loss. During the procedure, the middle ear can be cleared by suction, the electrode is placed on the promontorium, and thresholds are estimated at the lowest intensity where an AP is reliably recorded. Frequency-specific responses can be obtained using tone-bursts, which is helpful for auditory rehabilitation or hearing aid fitting.

The major disadvantage of ECoChG is that it is best performed under general anesthesia, since the invasive TT ECoChG is the preferred technique in the pediatric age-group. Even for a comprehensive diagnostic ABR, children often

undergo general anesthesia, so in selected cases the ECochG measurement may be performed during the same session as the diagnostic ABR. If ABR does not display a reliable wave I, the ECochG may show a well defined wave I or AP, since near-field recording permits a substantially larger amplitude of wave I. This is even true for extratympanic recordings<sup>31</sup>.

When AN/AD is suspected, ECochG is a powerful tool to evaluate the site of the lesion.<sup>32,33</sup> Nowadays, this is one of the main indications of pediatric ECochG. Presence of the CM component and/or SP, in the absence of ABR, are positive findings for AN/AD.<sup>32</sup> The AP is usually absent (as in ABR wave I), but when it is present, the waveform is grossly abnormal. The presence of some cochlear hair-cell activity (CM and SP components) does not mean that hearing is preserved.

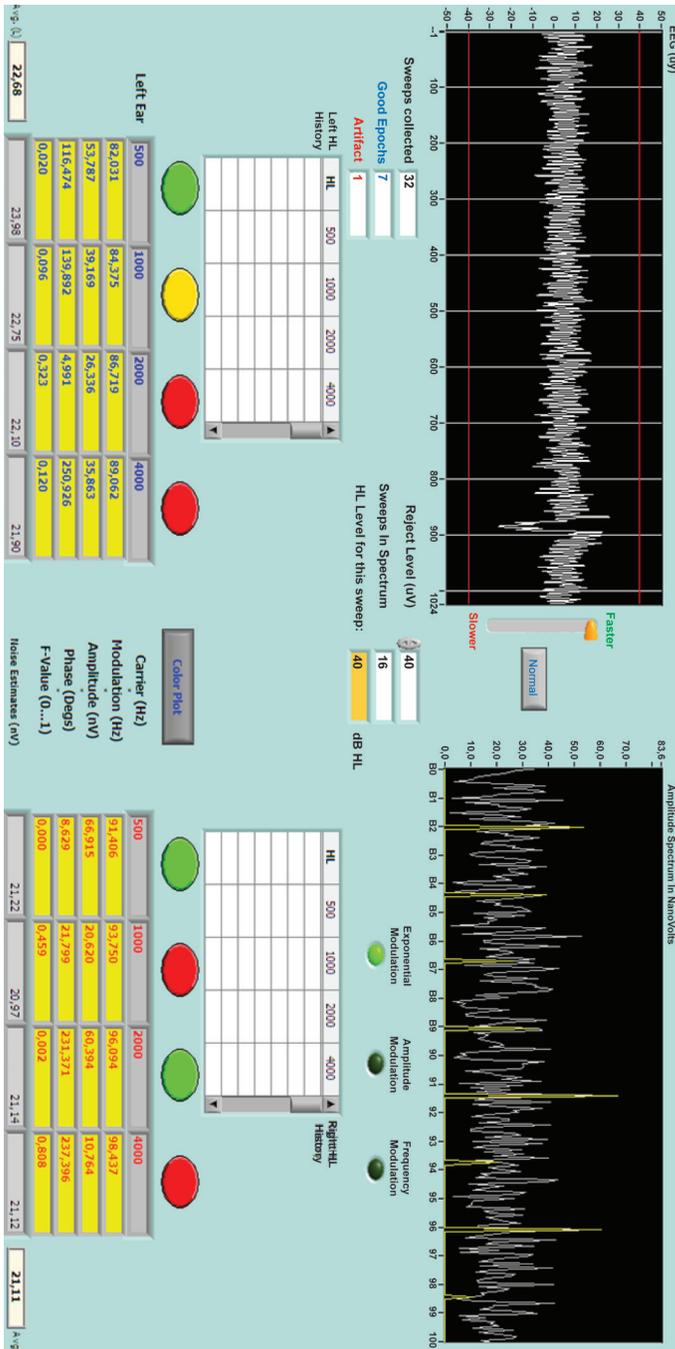
A combination of OAE, ECochG, and ABR is the best choice to adequately describe the site of lesion in AN/AD: outer hair cell or inner hair cell dysfunction, cochlear versus neural dysfunction.<sup>28</sup> This may be helpful for management options in these children. Caution should be exercised in very young children with presumed patterns of electrophysiologic AN/AD. At least some of them may show improvement with maturation of the auditory pathways, better hearing sensitivity, or audiometric contraindications for hearing aid fitting at subsequent examinations, so regular electrophysiologic and behavioral monitoring should be considered before other management steps (amplification, cochlear implants) are proposed to this very young age-group<sup>28,34</sup>.

## 7. Auditory Steady-State Responses

Auditory Steady-State Responses (ASSR) are scalp-recorded potentials evoked by continuous sinusoidal tones modulated in amplitude and/or frequency. Responses generated at a modulation frequency higher than 70 Hz seem to reflect auditory brainstem processes<sup>35</sup> and are used to obtain reliable responses in sleeping infants and children.<sup>36</sup> The response is phase-locked to the modulation envelope of the stimulus and is analyzed as a single peak that represents the periodicity in which the response protrudes from the electroencephalogram (EEG) spectrum. The presumed response can be extracted from background electrical activity (noise) of EEGs by means of statistical analysis (Fast Fourier Transform-FFT).<sup>37</sup>

The stimulus can be applied monaurally or binaurally, four frequencies at the same time, using either air- or bone-pathways (**Figure 9**). This multiple stimulus technique can decrease the time required to evaluate thresholds at multiple audiometric frequencies.

Figure 9. ASSR with binaural stimulation at multiple frequencies



Extracted from authors' image-bank

The ASSR can be used to predict hearing thresholds in infants and young children, with significant correlation with pure-tone behavioral thresholds.<sup>38</sup> The correlations increase with test- frequency and degree of hearing loss.<sup>39</sup> Besides, unlike the click-evoked ABR, the ASSR can provide threshold information at intensity levels up to 120 dB HL,<sup>36</sup> allowing investigation of residual hearing and differentiation between severe and profound hearing loss in young candidates for cochlear implants.<sup>39, 40</sup> Therefore, ASSR testing at elevated intensities allows a more appropriate hearing aid fitting. The absence of responses is indicative of profound hearing loss, with consequent poor results with a hearing aid.<sup>39</sup> In view of that, the ASSR is a unique tool in the assessment of young children before cochlear implant surgery.

As disadvantages, there are discrepancies between ASSR thresholds in normal hearing, conductive hearing loss and AN/AD. Electrical interferences and high EEG- noise can produce invalid results or underestimation of hearing-sensitivity. The test may also be affected by the state of arousal of the patient. Therefore, high artifact rejection levels and sedation are required in young children.

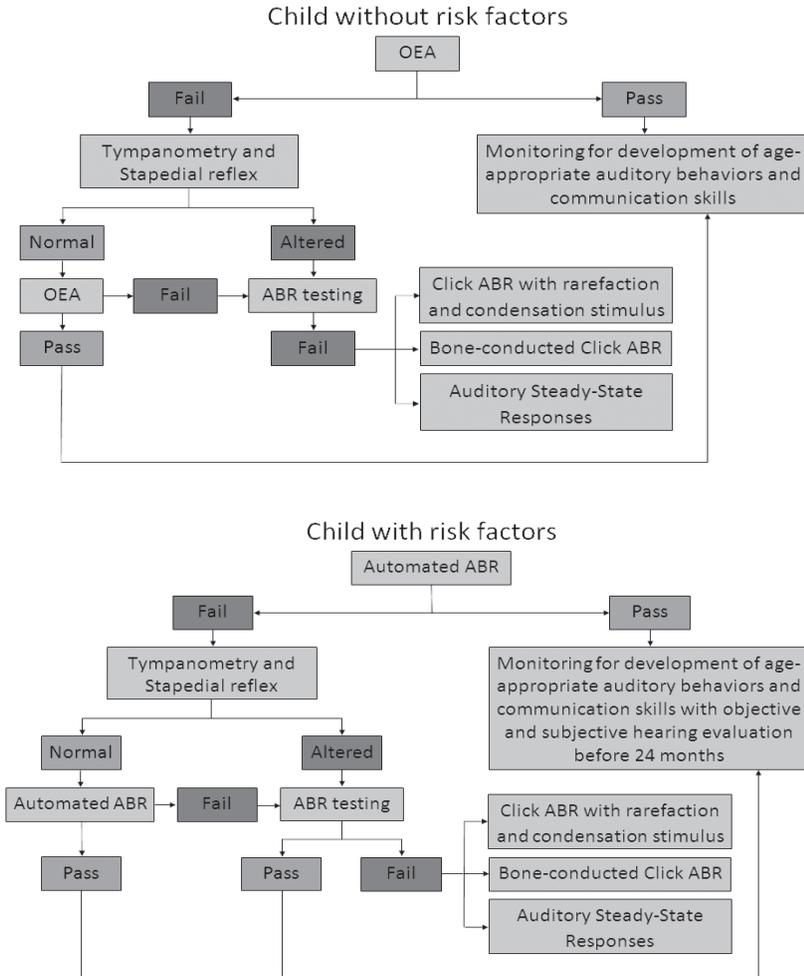
The ASSR is an important tool in the assessment of hearing in children, but its information is complementary and must be analyzed along with the results of ABR-testing.

## 8. Conclusion

It is important to remember that physiologic methods of auditory evaluation are not direct measures of hearing, but are nevertheless highly correlated with auditory status, especially peripheral auditory function. All the tests mentioned above reflect auditory status at the moment of examination. Therefore it is mandatory to perform sequential testing to evaluate maturation and development of auditory pathways. “Cross-checking” the results of otoacoustic emissions with tympanometry, stapedial reflex, click-evoked ABR, frequency-specific ABR findings (tone-burst or ASSR), and reliable behavioral tests is mandatory to confirm audiometric thresholds and predict hearing status, as they complement each other.

To assist in the screening and diagnosis of hearing loss in children without and with risk factors, algorithms (**Figure 10**) of the recommended approaches have been developed.

**Figure 10.** Algorithms of recommended approaches



Extracted from authors' image-bank

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