OBJECTIVE ASSESSMENT OF INFANT HEARING: ESSENTIAL FOR EARLY INTERVENTION

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Abstract

Substantial evidence supports the benefits of early intervention in infant hearing loss. Intervention can lead to the acquisition of effective communication skills and psychosocial development. Accurate diagnosis of infant hearing loss is not possible with conventional behavioral audiometry techniques. Objective auditory procedures are invaluable in the assessment of infant hearing because they do not rely on behavioral responses to sound and are unaffected by listener variables such as cognition, motivation, and language impairment.

Objective hearing procedures include aural immittance measures (tympanometry and acoustic reflexes), otoacoustic emissions (OAEs), electrocochleography (ECochG), auditory brainstem response (ABR), and the auditory steady state response (ASSR). Exclusive reliance on only one or two objective auditory measures often results in equivocal outcomes. Careful analysis of findings from a comprehensive objective auditory test battery can almost always yield a precise description of auditory status; it can often lead to accurate diagnosis of auditory dysfunction within weeks of birth. The key to meaningful analysis of findings from a test battery is the recognition of patterns associated with major auditory disorders. This is not a novel concept; it is simply the modern day version of the 40-year-old cross-check principle.

Key words: acoustic reflex • auditory neuropathy spectrum disorder (ANSD) • aural immittance measures • broadband noise (BBN) • chirp • LS (level-specific) chirp • EHDI (early hearing loss detection and identification) • sensory hearing loss • tympanogram • wideband reflectance/absorbance

EVALUACIÓN OBJETIVA DE LA AUDICIÓN EN LOS RECIÉN NACIDOS COMO BASE DE LA INTERVENCIÓN MÉDICA TEMPRANA

Resumen

Existen pruebas fundamentales que confirman los beneficios que resultan de la intervención médica temprana en caso de los recién nacidos con pérdida auditiva. Los resultados de la intervención médica son la adquisición de capacidad de comunicarse efectivamente y el desarrollo psicosocial. No es posible diagnosticar correctamente a los recién nacidos con pérdida auditiva usando las técnicas convencionales de audiometría por observación de comportamiento. Los métodos de estudio objetivos sirven como gran ayuda en la evaluación de la audición de los recién nacidos, ya que no se basan en las respuestas conductuales al sonido. Percepción, motivación y trastornos del lenguaje tampoco influyen en los resultados.

Los métodos de examen objetivos abarcan los estudios auditivos de impedancia (timpanometría y reflejo acústico), emisiones otoacústicas (OAE), electrococleografía (ECochG), respuesta auditiva provocada del tronco encefálico (ABR) y potenciales evocados auditivos de estado estable (ASSR). Apoyarse en uno o dos métodos de examen objetivos muy frecuentemente no da resultados claros. El análisis profundo de los resultados de una serie de exámenes casi siempre garantiza la descripción detallada del estado de audición; siempre lleva al diagnóstico preciso de las disfunciones auditivas durante las primeras semanas tras el nacimiento. El clave para el análisis profundo de la serie de exámenes es el reconocimiento de los patrones vinculados con las principales disfunciones auditivas. No es un concepto nuevo, es simplemente una versión moderna del principio comparativo de 40 años.

Palabras clave: reflejo acústico • trastorno en el espectro de neuropatía auditiva (ANSD) • estudios auditivos de impedancia • ruidos de banda ancha (BBN) • estímulos del tipo "chirp" • detección e intervención tempranas de la pérdida auditiva (EHDI) • pérdida auditiva sensorineural • timpanograma • reflectancia/absorbancia de banda ancha

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ОБЪЕКТИВНАЯ ОЦЕНКА СЛУХА У МЛАДЕНЦЕВ ОСНОВОЙ ДЛЯ РАННЕЙ МЕДИЦИНСКОЙ ИНТЕРВЕНЦИИ

Изложение

Существуют основные доказательства, подтверждающие пользу возникающую из ранней медицинской интервенции в случае младенцев с нарушением слуха. Медицинская интервенция ведет к приобретению способностей эффективной коммуникации и психосоциального развития. Правильный диагноз младенцев с нарушением слуха невозможен при использованию традиционных техник бихевиоральной аудиометрии. Объективные методы исследований очень полезные при оценке слуха младенцев, потому что они не заключаются в бихевиоральных реакциях на звук. На результат не имеет также влияния восприятие, побуждение, языковые нарушения.

Объективные методы исследований включают слуховые исследования импеданса (тимпанометрия и акустическая реакция), отоакустичуские эмиссии (ОАЭ), электрокохлеография (ЭкоГ), слуховые вызванные потенциалы ствола мозга (АБР) и слуховые вызванные потенциалы стабильного состояния (АССР). Полагание на одном или двух объективных методах исследований часто не дает четких результатов. Точный анализ результатов для всей батереи тестов почти всегда гарантирует точное описание состояния слуха, всегда ведет к точному диагнозу слуховых дисфункций во время первых недель после рождения. Ключом к тщательному анализу результатов батереи тестов является признание образцов связяных с главными нарушениями слуха. Это не является новой идеей – это просто современная версия сороколетного сравительного правила.

Ключевые слова: акустическая реакция • спектр нарушений слуховой невропатии (АНСД) • слуховые исследования импеданса • широкополосный шум (ББН) • стимул типа «хирп» • ранный диагноз и идентификация потери слуха (ЭХДИ) • сенсорная потеря слуха • тимпанограмма • широкополосное отражение/впитывающая способность;

OBIEKTYWNA OCENA SŁUCHU U NOWORODKÓW PODSTAWĄ DO WCZESNEJ INTERWENCJI MEDYCZNEJ

Streszczenie

Istnieją zasadnicze dowody potwierdzające korzyści wynikające z wczesnej interwencji medycznej w przypadku niemowląt z niedosłuchem. Interwencja medyczna prowadzi do nabycia umiejętności skutecznego komunikowania się i rozwoju psychospołecznego. Poprawna diagnoza u niemowląt z niedosłuchem nie jest możliwa przy użyciu konwencjonalnych technik audiometrii behawioralnej. Obiektywne metody badań są bardzo pomocne przy ocenie słuchu u noworodków ponieważ nie polegają na odpowiedziach behawioralnych na dźwięk. Na wynik nie mają także wpływu percepcja, motywacja, zaburzenia językowe.

Obiektywne metody badań obejmują słuchowe badania impedancyjne (tympanometria i odruch strzemiączkowy), emisje otoakustyczne (OAE), elektrokochleografia (ECochG), słuchowe potencjały wywołane pnia mózgu (ABR) i słuchowe potencjały wywołane stanu ustalonego (ASSR). Poleganie na jednym lub dwóch obiektywnych metodach badań często nie daje jasnych wyników. Dokładna analiza wyników dla całej baterii testów niemalże zawsze gwarantuje dokładny opis stanu słuchu; zawsze prowadzi do dokładnej diagnozy dysfunkcji słuchowych w przeciągu pierwszych tygodni po urodzeniu. Kluczem do wnikliwej analizy wyników baterii testów jest rozpoznanie wzorców związanych z głównymi zaburzeniami słuchu. Nie jest to nowa koncepcja, jest to po prostu nowoczesna wersja czterdziestoletniej zasady porównawczej.

Słowa kluczowe: odruch strzemiączkowy • spektrum zaburzeń neuropatii słuchowej (ANSD) • słuchowe badania impedancyjne • hałas szerokopasmowy (BBN) • bodźce typu „chirp" • wczesne wykrycie i identyfikacja niedosłuchu (EHDI) • niedosłuch odbiorczy • tympanogram • reflektancja/absorbancja szerokopasmowa

Introduction

Rationale for objective infant hearing assessment

The coordination of prompt identification, diagnosis, and management of hearing loss in children is now often referred to as Early Hearing Detection and Intervention, abbreviated EHDI. The "1–3–6 Plan" (or Principle) guides EHDI efforts. That is, hearing loss is detected before 1 month, diagnosis of hearing loss is complete within the first 3 months after birth, and intervention begins within 6 months. Substantial research evidence supports the benefits of early intervention for the acquisition of effective and efficient communication skills along with psychosocial development [1]. Rich, consistent, and reasonably normal auditory stimulation, beginning with the first 6 months after birth, drives nervous system development and takes full advantage of brain plasticity.

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Table 1. General clinical strengths and weaknesses of objective auditory measures available to clinical audiologists. Specific advantages associated with each measure are detailed in the text

• Do not require a behavioral response from the patient
• Results are not influenced by motivation
• Results are not influenced by cognitive status
• Results are generally not influenced by state of arousal
• Measurements can be made with patient sedated or anesthetized
• Results are not influenced by native language
• Patient is not required to follow detailed verbal instructions
• Motor status does not influence test results
• Measures provide information on regions of the auditory system from the middle ear to the cerebral cortex
• Generally high degree of sensitivity to auditory dysfunction
• In combination, provide site-specific information on auditory dysfunction
• Valid measures are possible from infants and young children
• Reasonable test time

The evolution of this rather accelerated schedule for EHDI essentially eliminates the role of behavioral hearing assessment in the initial diagnosis of hearing loss and the first habilitation efforts with hearing aids and other devices. Fortunately, objective auditory tests are available for early and accurate diagnosis of infant hearing loss. These include aural immittance measurements, otoacoustic emissions (OAEs), and auditory evoked responses such as auditory brainstem response (ABR), auditory steady state response (ASSR), electrocochleography (ECochG), and cortical auditory evoked responses like the auditory middle latency response (AMLR), the auditory late response (ALR), and the P300 response. The ALR is also referred to as the cortical auditory evoked potential (CAEP).

Cortical auditory evoked responses, and really all auditory evoked responses, appear sequentially following presentation of effective stimulation. In this discussion of objective auditory measures in infants and young children, it is relevant to point out that there is no clear and invariable distinction in the latency characteristics of the cortical auditory evoked responses. For example, the ALR and P300 responses occur within the same general post-stimulus analysis time. And, components of the AMLR when recorded from infants actually appear in the analysis time associated with the ALR for older children and adults.

Objective hearing procedures all share one major clinical advantage. They are not dependent on behavioral infant responses. The many general clinical strengths of objective hearing procedures are summarized in Table 1. This article reviews current application of four objective auditory assessment applied most often in the diagnosis of hearing loss in infants and young children, specifically: 1) aural immittance measures, 2) OAEs, 3), ABR, and 4) ASSR. The important diagnostic roles of two other categories of objective auditory measures (ECochG and cortical auditory evoked responses) are only mentioned in passing due to space constraints.

Other clinical applications

There are multiple clinical applications of objective auditory measures. We have already touched upon hearing screening and diagnosis of hearing loss in infants and young children. Although these applications are indeed crucial, others also play an important role in children and also in adults. Diagnosis of auditory neuropathy spectrum disorder (ANSD) is only possible with a combination of objective techniques, including ECochG. Objective measures, such as OAEs and tympanometry, offer the most efficient and accurate means of screening for hearing loss in pre-school and school age children. Objective auditory procedures permit prompt and unequivocal identification and diagnosis of false or exaggerated hearing loss and, therefore, timely and appropriate management of pediatric and adult patients. Finally, objective measures supplement behavioral diagnostic audiometry procedures in the assessment of auditory processing disorders, including those resulting from traumatic brain injury (TBI). Objective test data is particularly useful when analysis of behavioral findings is confounded due to listener variables such as cognition, motivation, and impairment of language (either native or specific). In short, objective test procedures are essential.

Historical perspective

Initial efforts to objectively assess hearing in children date back more than 60 years. In the 1950s, several teams of otolaryngologists and audiologists applied ECochG, which was used to estimate auditory thresholds in difficult-to-test children who had delayed speech and language and where hearing loss was strongly suspected (see reference 2 for review). The technique, however, was far from standard care because ECochG recording in children then required a surgeon to place an electrode close to the cochlea under general anesthesia.

In the 1960s, other groups of research-oriented audiologists and neurologists reported use of the auditory late cortical evoked response to estimate auditory thresholds in children who were unable to be assessed with behavioral audiometry. The good news was that anesthesia and surgical support was not needed for clinical measurement of cortical evoked responses, but the strategy depended heavily on patient cooperation and was very age-dependent. Children undergoing auditory late response measurement needed to be almost motionless, yet awake. Unfortunately, cortical auditory evoked response measurements required almost as much cooperation as behavioral audiometry. Until at least the mid-1970s, however, objective assessment of hearing in children using ECochG and cortical auditory evoked responses was available only in a relatively few major medical centers throughout the world.

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Table 2. Selected clinical strengths and weaknesses of aural immittance measures. All objectives measures share a number of clinical advantages, as summarized in Table 1 and described in detail in the text. Strengths of aural immittance measures that are discussed in the text are highlighted in bold

Strengths	Weaknesses
• Equipment is widely accessible • Clinically proven with over 40 years of clinical experience and research • **Normative data are available** • **Anatomy and physiology relatively well defined** • Relatively independent of developmental age or status • Brief test time • Relatively simple techniques • Useful as a screening technique • Measurement does not require sedation or anesthesia • High degree of sensitivity to middle ear dysfunction • Tympanometry provides information on middle ear mechanics • Detects and confirms perforation of the tympanic membrane • Detects and confirms patent ventilation tubes • **Acoustic reflex provides information on afferent auditory pathways** • **Acoustic reflex is sensitive to retro-cochlear auditory dysfunction** • **Acoustic reflex provides information on lower brainstem auditory pathways** • **Acoustic reflex provides information on 7th cranial (facial) nerve** • **Acoustic reflex objectively detects or rules out sensory hearing loss**	• Not a measure of "hearing" • Measurement requires an air-tight seal within external ear canal • Tympanometry only provides information on middle ear status • No information on higher brainstem auditory function • No information on cortical auditory function • No information on speech perception or understanding • Does not provide precise index of the degree of hearing loss • Acoustic reflex findings limited in patients with normal middle ear status

Cross-check principle: a 40-year perspective

The modern objective hearing test battery was introduced in the 1970s. Leading researchers, notably James Jerger, reported compelling evidence – in dozens of peer-reviewed publications from large-scale studies over varied patient populations – that objective auditory assessment was clinically valuable and necessary. Robert Galambos in the mid-1970s clearly demonstrated the unique contributions of ABR in newborn hearing screening and diagnosis of hearing loss in infants and young children [3].

In 1976, Dr Jerger and then clinic supervisor and PhD student Deborah Hayes first articulated the enduring 'cross-check principle'. In their classic *The Cross-Check Principle in Pediatric Audiometry*, Jerger & Hayes [4] (clearly illustrated with 5 case studies the limitations and pitfalls associated with exclusive reliance on behavioral test results. The authors made a strong case for the use of independent test procedures, principally aural immittance (impedance) measures and ABR to verify or "cross-check" the behavioral test results. Jerger and Hayes confidently state: "In summary, we believe that the unique limitations of conventional behavioral audiometry dictate the need for a 'test battery' approach. The key concept governing our assessment strategy is the cross-check principle. The basic operation of this principle is that no result be accepted until it is confirmed by an independent measure." [4, p. 620].

The objective test battery did not expand further until about 20 years later when OAE technology became available as a routine clinical procedure. Within the next decade, the Joint Committee on Infant Hearing in 2000 strongly recommended the routine application of ABRs elicited with frequency-specific tone burst stimulation and also bone conduction ABR measurement for auditory assessment of infants and young or difficult-to-test children. During the same period, the ASSR emerged as a clinically feasible technique for objective estimation of auditory thresholds, especially in children with severe to profound hearing impairment. More recently ECochG has resurfaced once again as a valuable clinical tool, this time in the diagnosis of children with suspected ANSD. We now have readily available – for use in patients of all ages – an assortment of objective techniques for early and accurate identification and diagnosis of every type and site of auditory dysfunction, from middle ear disorders to ANSD to central auditory processing disorders.

Thousands of journal articles, hundreds of book chapters, and even entire textbooks, describe in detail the functional anatomy of objective auditory tests and how the procedures are performed and findings are analyzed. This paper briefly reviews the advantages of four major objective auditory tests in the detection and diagnosis of infant hearing loss. It also highlights the unique contribution of each of these objective auditory procedures to the pediatric test battery.

Aural immittance measures

Making the most of middle ear measurements

Aural immittance measures are valuable clinically for a variety of reasons. They are quick, technically simple, easily recorded in persons of all ages without regard to developmental or cognitive status, and they have relatively high sensitivity and specificity to middle ear disorders. The many compelling clinical advantages specific to aural immittance measurements, particularly in pediatric patient populations, are summarized in Table 2.

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Research confirms that multi-frequency and multi-component techniques for tympanometry are more sensitive to low-impedance pathologies, such as tympanic membrane and ossicular chain abnormalities, than measurement of admittance recorded with a single low-frequency probe tone, usually 226 Hz. Nonetheless, audiologists typically rely on single component and single frequency tympanometry for detection and diagnosis of auditory dysfunction. Tympanometry and analysis of tympanogram findings is simpler when one component is recorded for one probe tone frequency. Most middle ear pathology in pediatric populations is detected and described with single-component and single-frequency tympanometry.

There are clear clinical indications for the use of high-frequency tympanometry in addition to low-frequency tympanometry in infants up to at least 4 months of age [5,6]. Aural immittance characteristics differ substantially for infants versus older children and adults. Specifically, in comparison to older persons the middle ears of infants have a higher resistance component for a low-frequency probe tone of 226 Hz. Ear canal volume measurements in infants under 6 months, however, should be conducted with a low-frequency probe tone.

Wideband reflectance/absorbance

Another approach to middle ear assessment, measurement of wideband reflectance or absorbance, offers potential advantages over conventional tympanometry, particularly for detection of pathology in neonates and young children [7]. The WMEP essentially involves simultaneous measurement of power reflectance, impedance, and admittance using either a broadband (chirp) stimulus or multiple sinusoidal stimuli over a relatively wide frequency range of about 250 Hz to 6000 Hz.

Test time for wideband reflectance or absorbance is less than 1 minute and measurements are made at ambient pressure or with induced ear canal pressure. An airtight seal between the probe and the ear canal wall is not required. Wideband reflectance or absorbance has considerable potential value for detection of auditory dysfunction in infants and older children. And measured in combination with OAEs using the same instrumentation, including a single probe, wideband reflectance or absorbance may result in an unusual and desirable combination of high sensitivity and high specificity for detection of middle ear disorders.

The diagnostically powerful acoustic stapedial reflex

Background. The acoustic stapedial reflex is one of several muscle responses to sound. It falls in the same general category as the post-auricular muscle response, the eyeblink reflex, and the startle response. Another middle ear muscle, the tensor tympani muscle, is involved in the startle response. Careful measurement of stapedial acoustic reflexes yields considerable information on the anatomical status of the auditory system, especially when recorded for four conditions, that is, measurement of ipsilateral and contralateral acoustic reflex activity using right and left ear stimulation.

The major pathways in the acoustic reflex arc can be divided anatomically into five general portions: 1) the middle ear, the cochlea, and the afferent pathway consisting of the 8th (auditory) cranial nerve on the side of the stimulus; 2) brainstem neurons within the cochlear nuclei; 3) for contralateral acoustic reflex measurement, the trapezoid body and medial superior olivary complex plus polysynaptic pathways including neurons within the reticular activating system; 4) an efferent pathway involving motor fibers within the 7th cranial nerve on the side of the probe; and 5) the stapedius muscle and middle ear on the side of the probe. The presence of acoustic reflexes is highly dependent on normal middle ear function. Most middle ear abnormalities obscure confident detection of acoustic reflexes, even relatively subtle disorders that are not associated with markedly abnormal tympanograms or a significant (≥10 dB) gap between air- and bone-conduction pure tone thresholds.

Identification of sensory hearing loss. Tympanometry has unquestionable value as a screening tool for the detection of middle ear abnormalities. However, hearing requires integrity of much more than the middle ear. The application of hearing loss estimation with acoustic reflex thresholds was first reported in the early 1970s [8]. Presuming normal middle ear function, acoustic reflexes permit quick, ear-specific objective differentiation of normal versus abnormal cochlear function. As clinical experience with acoustic reflex measurement accumulated, a direct relation emerged between hearing loss and acoustic reflex levels for noise signals. In particular, the acoustic reflex threshold for broadband noise (BBN) increases rather systematically with worsening pure tone thresholds for sensory hearing loss. In contrast, acoustic reflexes elicited with pure tone signals showed little change in threshold from normal hearing sensitivity through 50 or even 60 dB HL, a reflection of the loudness recruitment phenomenon.

In a study of 326 adult subjects with varying degrees of sensory hearing loss, Hall, Berry & Olsen [9] showed how the acoustic reflex threshold for a BBN signal presented in the contralateral condition could differentiate patients with a pure tone average <35 dB HL from those with a pure tone average ≥35 dB HL. Figure 1 illustrates the differential effect of sensory hearing loss on acoustic reflex thresholds elicited with tonal versus BBN signals. No subject with hearing loss (pure tone average >35 dB HL) had an acoustic reflex threshold for BBN of less than 85 dB SPL. The lower the acoustic reflex threshold for the BBN stimulus, the more likely hearing sensitivity is normal within the speech frequency region. Conversely, BBN acoustic reflex thresholds greater than 90 dB are invariably associated with sensory hearing loss.

The results from a study of acoustic reflex thresholds in neonates provides further support for the use of a BBN stimulus in objectively differentiating between normal hearing sensitivity versus sensory hearing loss. Kei [10] reported acoustic reflex threshold data collected with pure tone and BBN stimuli and a 1000 Hz probe tone in a group of 66 healthy newborn infants who had passed hearing screening. Acoustic reflexes were recorded under all stimulus conditions from all infants. The median acoustic reflex

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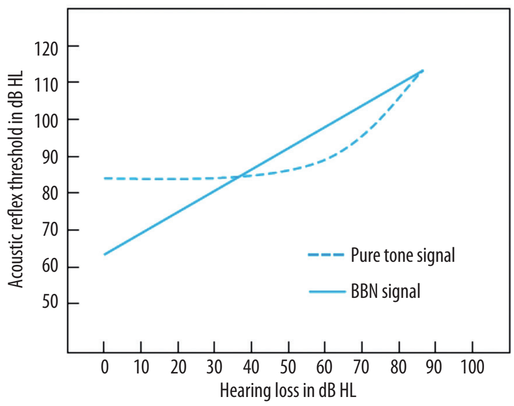

Figure 1. Acoustic reflex thresholds for pure tones (dashed line) and broadband noise stimuli (continuous line) as a function of hearing threshold levels

threshold in the normal hearing infant group was 55 dB HL for the BBN stimulus (with a range of 50–75 dB HL). These findings confirm that an acoustic reflex threshold of 75 dB HL or better is consistent with normal hearing sensitivity.

Diagnostic value of acoustic reflex patterns

Possible pathways and test conditions. A brief explanation of acoustic reflex conditions and patterns might be helpful [2]. In discussing acoustic reflex patterns, it is important to make a distinction between 'probe ear' and 'stimulus ear.' Tympanometry is performed with the probe ear. For ipsilateral reflexes, the probe ear and stimulus ear are one and the same. Acoustic immittance change – indicating the presence of an acoustic reflex – occurs in the same ear as the acoustic stimulation. The term uncrossed is also used for the ipsilateral test condition, as the acoustic reflex pathways do not cross the midline of the brainstem.

In the contralateral acoustic reflex condition, the stimulus is presented to the ear opposite the probe ear. Acoustic immittance change indicating the presence of an acoustic reflex occurs in the ear opposite the stimulation. The term crossed is interchangeable with contralateral, as the acoustic reflex pathways cross the midline of the brainstem via the trapezoid body and perhaps other decussating structures before coursing to the region of the motor nucleus of the 7th cranial (facial) nerve and then to the stapedius muscle via motor fibers within the 7th cranial nerve.

There are, then, four possible distinct and different measurement conditions in acoustic reflex measurement: 1) right ear ipsilateral; 2) left ear ipsilateral; 3) contralateral reflexes with the probe in the right ear and sound in the left; and 4) contralateral reflexes with the probe in the left ear and sound in the right. These four measurement conditions and normal findings for each are often shown graphically in a diagram like the one shown in Figure 2. An open box in the figure indicates the presence of normal acoustic reflexes with thresholds of ≤90 dB HL. A shaded box indicates abnormally elevated acoustic reflex thresholds, whereas a filled-in black box indicates that no acoustic reflex activity was detected in the test condition.

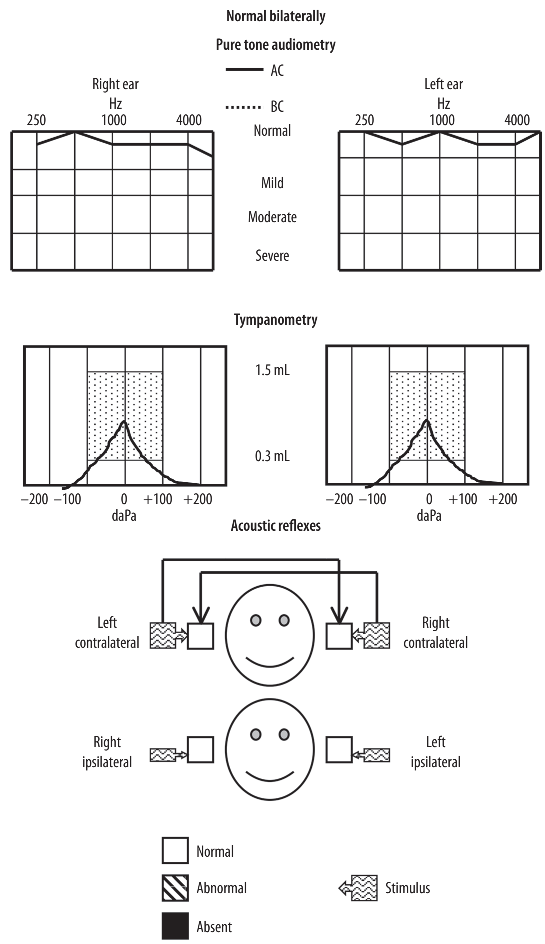

Figure 2. Plotted together are findings for pure tone audiometry (top), tympanometry (middle), and acoustic reflexes in the ipsilateral and contralateral conditions (bottom). Findings are for a person with normal hearing sensitivity

Combinations or patterns of findings for pure tone audiometry, tympanometry, and acoustic reflex recordings are generally related to likely clinical etiologies or diagnoses. The figures cited in the explanation below depict distinct patterns of acoustic reflex findings. In viewing these figures, and real world clinical findings, it is useful to first examine the findings for tympanometry to confirm or rule out middle ear disorder, followed by analysis of the acoustic reflex pattern for the four measurement conditions. The audiogram offers additional evidence of conductive hearing loss.

Vertical acoustic reflex pattern: mild conductive hearing loss. The vertical pattern is often encountered clinically, particularly in pediatric populations where middle ear disorders are commonplace. Figure 3 shows an example of the vertical acoustic reflex pattern. The tympanogram on the right ear is clearly abnormal, immediately alerting the clinician to the likelihood of a conductive hearing loss. Referring to the lower portion of the figure, acoustic reflexes are absent whenever the probe is in the right ear with middle ear dysfunction. Detection of a normal contralateral acoustic reflex with sound in the right

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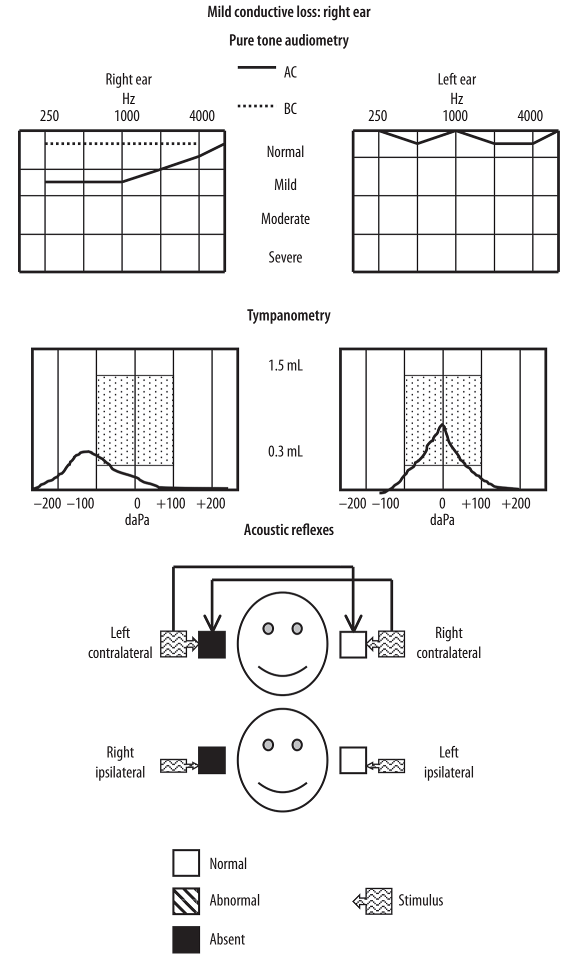

Figure 3. Integrated plot (as per Figure 2) for a patient with middle ear dysfunction and mild conductive hearing loss for the right ear. The combination of findings is sometimes referred to as the vertical acoustic reflex pattern

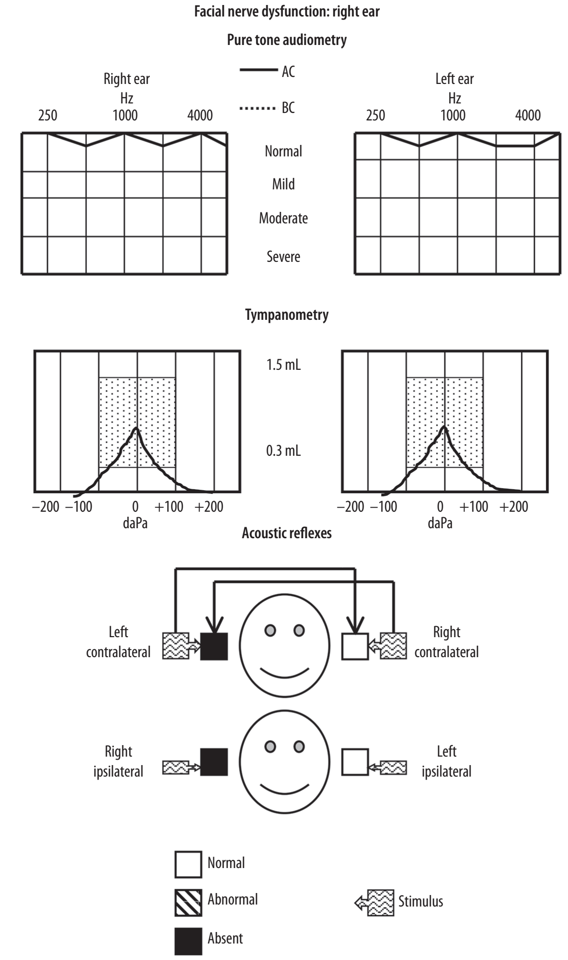

Figure 4. Integrated plot (as per Figure 2) for a patient with facial nerve dysfunction for the right ear in combination with normal middle ear function and normal hearing sensitivity bilaterally

ear and probe in the normal left ear confirms, even before reference to pure tone findings, that the conductive hearing loss is mild at most.

Greater conductive hearing loss for the right ear would result in elevation of the contralateral acoustic reflex measured with stimulation of the right ear and the probe in the left ear. A conductive hearing loss essentially reduces the effectiveness of the acoustic reflex stimulation by the magnitude of the air-bone gap. Since the acoustic reflex is normally activated with an intensity level of 85 dB HL, a conductive loss of 25 to 30 dB raises the contralateral acoustic reflex threshold for stimulation of the ear with conductive hearing loss to about 110 to 115 dB HL.

Keep in mind that, typically, no acoustic reflex can be measured with the probe in an ear with a middle ear disorder, even in conductive hearing loss associated with a very modest 5 to 10 dB air-bone gap. The audiogram showing a slight air-bone gap in the top portion of Figure 3 confirms the acoustic reflex pattern. Prediction of degree of conductive hearing loss from the acoustic reflex pattern is especially useful in infants and young children for whom pure tone audiometry is not yet possible.

Vertical acoustic reflex pattern: facial nerve disorder. Facial nerve disorder is a second explanation for the vertical pattern of acoustic reflex abnormality. The pattern arises because the facial nerve is the final efferent pathway to the stapedius muscle. Acoustic reflexes are abnormal and usually absent whenever the probe is in the affected ear, as illustrated in Figure 4. Two factors clearly distinguish this vertical pattern from the acoustic reflex pattern typical of mild conductive hearing loss illustrated earlier in Figure 3. The most obvious factor is normal tympanometry in facial nerve disorder, consistent with normal middle ear function. A normal audiogram, or at least no difference between air and bone conduction pure tone thresholds, also argues against middle ear disorder. Careful measurement of acoustic reflexes in the four test conditions permits identification of facial nerve disorder in patients of all ages, even infants and young children with syndromes or diseases that include as a sign facial nerve pathology and paralysis.

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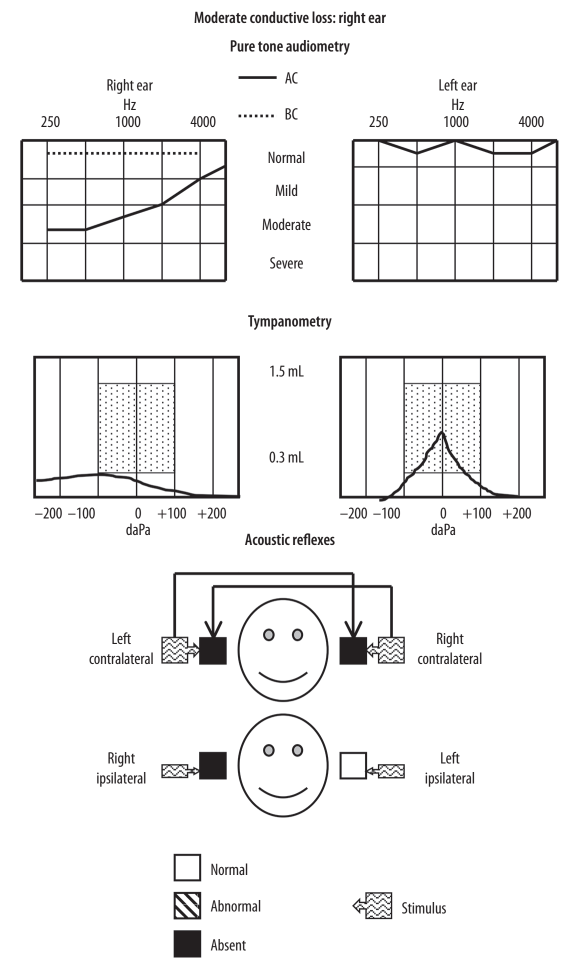

Figure 5. Integrated plot for a patient with middle ear dysfunction and moderate conductive hearing loss for the right ear. The combination is sometimes referred to as the 'inverted L' acoustic reflex pattern

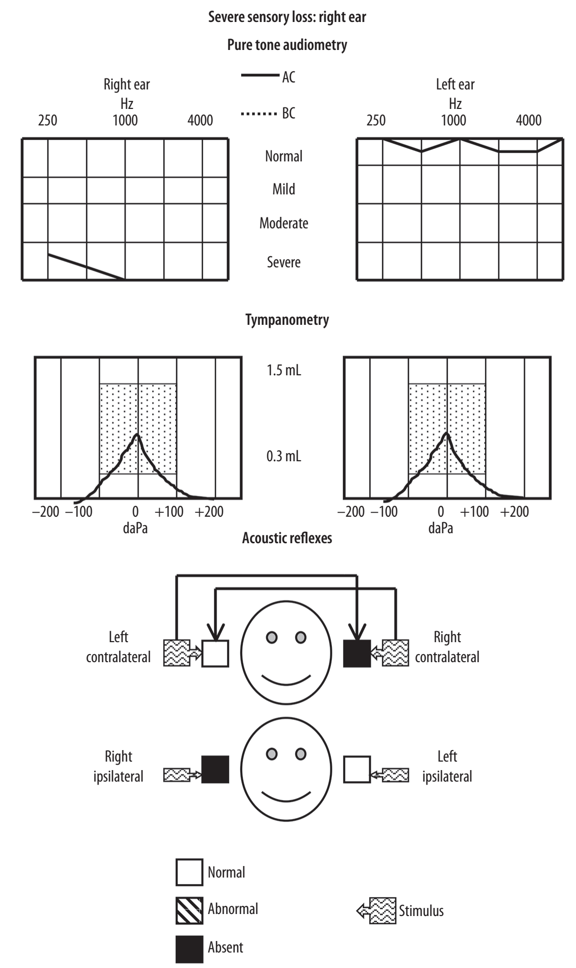

Figure 6. Integrated plot for a patient with severe sensory hearing loss on the right side. The combination is sometimes referred to as the diagonal acoustic reflex pattern

'Inverted L' acoustic reflex pattern: moderate conductive hearing loss. The 'inverted L' pattern for acoustic reflexes is really a vertical pattern with the addition of an abnormality in the contralateral acoustic reflex using stimulation of an ear with conductive hearing loss and the probe in the normal ear. This pattern is reflected in Figure 5. Almost any degree of conductive loss will produce some elevation of the contralateral acoustic reflex with sound stimulation in the conductive loss ear. Greater degrees of conductive loss and larger air-bone gaps are associated with progressive elevation of the acoustic reflex.

Diagonal acoustic reflex pattern: sensory disorder. When acoustic reflexes are abnormally elevated in threshold or absent with stimulation of one ear, the most likely explanation is a sensory hearing loss. The diagonal pattern is illustrated in Figure 6. The chances of detecting acoustic reflex activity decline as the degree of sensory hearing loss increases. Normal acoustic reflex findings are anticipated in mild and even moderate sensory hearing loss, reflecting the loudness recruitment phenomenon. Generally, acoustic reflexes for pure tone signals are recorded until the degree of loss exceeds about 60 dB HL. The presence of normal acoustic reflexes with the probe in each ear under at least one condition confirms normal middle ear function in both ears.

Diagonal acoustic reflex pattern: neural disorder. At first glance the diagnostic pattern seen in Figure 7 may appear similar to, perhaps indistinguishable from, the diagnostic pattern just illustrated in Figure 6. Close inspection of all available findings clearly differentiates the two patterns. The big difference is the degree of hearing loss. With neural auditory dysfunction secondary to an acoustic tumor such as a vestibular schwannoma, the diagonal acoustic reflex abnormality is often associated with only mild hearing loss. The neural pattern may also be suspected due to acoustic reflex decay.

'Inverted L' acoustic reflex pattern: neural disorder. A marked neural abnormality can produce the 'inverted L' pattern, described earlier for a severe conductive loss. Abnormality of the 8th cranial (acoustic) nerve affecting sound stimulation of the involved ear produces the diagonal component of the pattern. A large neoplasm compressing the brainstem as well as the 8th cranial nerve may also

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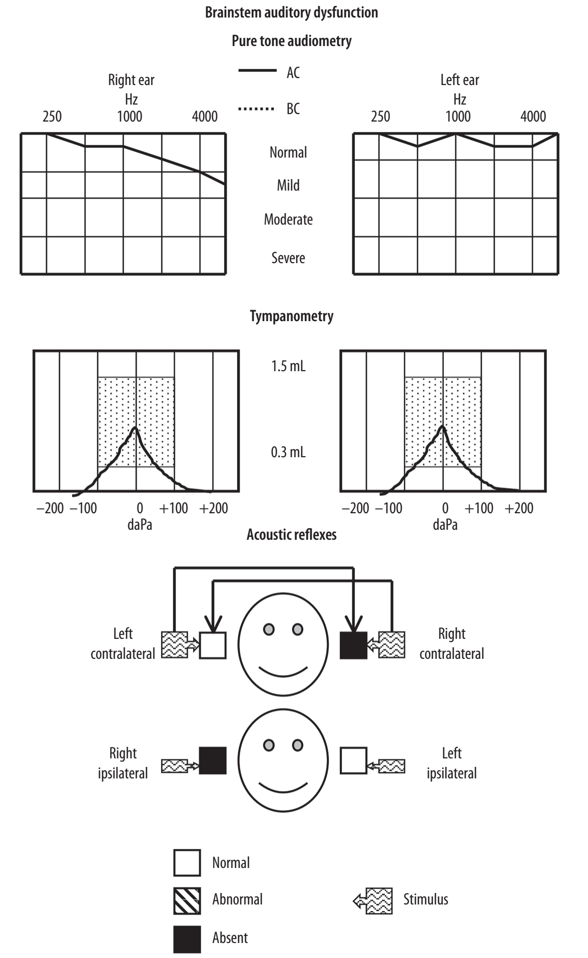

Figure 7. Integrated plot for a patient with neural hearing loss on the right side

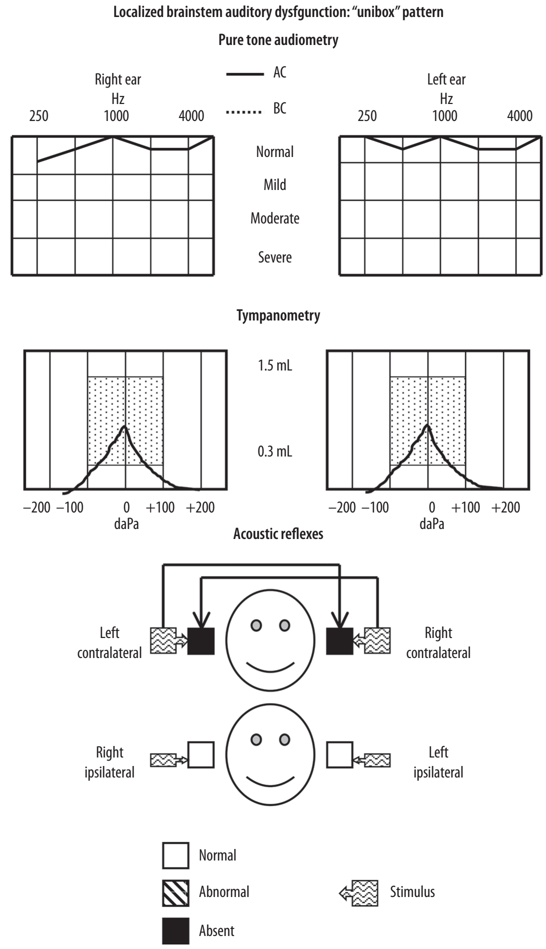

Figure 8. Integrated plot for a patient with brainstem auditory dysfunction. The combination is sometimes referred to as the horizontal acoustic reflex pattern

affect the crossed or contralateral acoustic pathways within the brainstem, with a resulting abnormality of both contralateral acoustic reflexes. The 'inverted L' neural pattern is consistent with a larger tumor involving the 8th cranial nerve and brainstem, whereas the diagonal neural pattern is found usually in smaller tumors affecting only the 8th cranial nerve. Two findings distinguish the inverted L acoustic reflex pattern for conductive hearing loss versus neural disorder. For the neural pattern, tympanometry is normal for the neural pattern and there is no evidence of an air-bone gap with pure tone audiometry.

Horizontal acoustic reflex pattern: brainstem disorder. A horizontal acoustic reflex pattern, as depicted in Figure 8, is encountered in patients with brainstem auditory dysfunction yet entirely normal peripheral auditory function. The presence of normal ipsilateral acoustic reflexes and normal tympanometry unequivocally rule out conductive hearing loss, sensory hearing loss, neural auditory dysfunction, and facial nerve disorder. The only appropriate anatomic explanation is brainstem auditory disorder.

Whenever the horizontal acoustic pattern is found clinically, it is very important to rule out technical problems and to verify that the appropriate stimulus intensity is being presented to each ear through the contralateral stimulus transducer. If supra-aural earphones are used for contralateral stimulation, collapsed ear canals must also be ruled out. The horizontal acoustic reflex pattern is a strong sign of brainstem auditory dysfunction in patients at risk for central auditory nervous system dysfunction, including those with head injury and suspected auditory processing disorder (APD).

Acoustic reflexes offer a completely objective auditory measure not influenced by multiple listener variables such as motivation, cognition, age, language, and attention that might compromise behavioral measures of auditory function. The horizontal acoustic reflex abnormality strongly suggests the need for a comprehensive assessment of central auditory function and, depending on the outcome of the assessment, otolaryngology and/or neurology referral.

'Uni-box' acoustic reflex pattern: brainstem disorder. Jerger et al. [11] first described a rare pattern of acoustic reflex findings. The pattern is characterized by an abnormality in only one contralateral acoustic reflex condition, as shown in Figure 9. All pathologic explanations, other than an isolated unilateral brainstem auditory abnormality,

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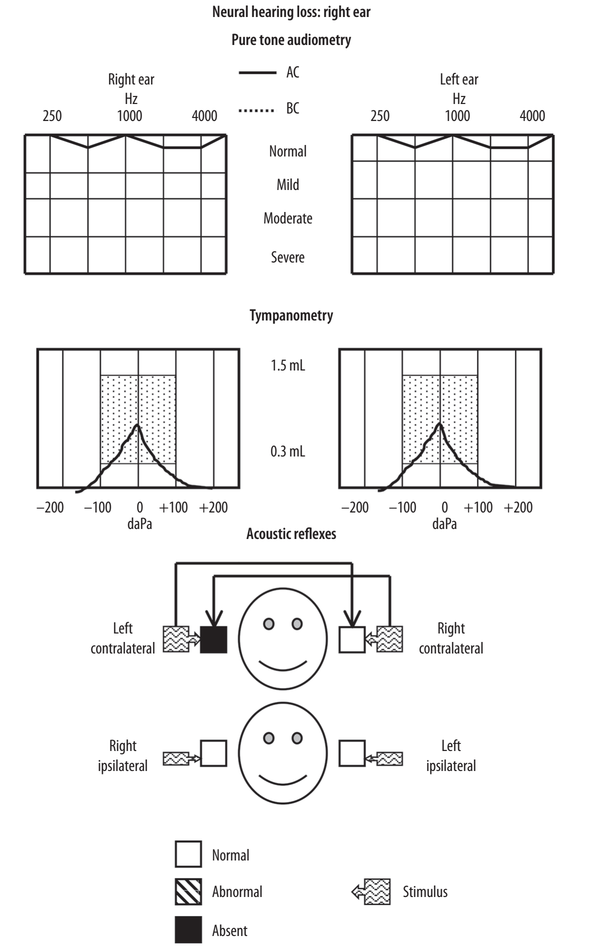

Figure 9. Integrated plot for a patient with localized brainstem auditory dysfunction. The combination is sometimes referred to as the 'unibox' acoustic reflex pattern

are convincingly ruled out due to the presence of normal acoustic reflexes in the other three acoustic reflex conditions, plus normal tympanograms and typically normal hearing sensitivity bilaterally. Observation of the uni-box acoustic reflex abnormality prompts a comprehensive assessment of central auditory function and, in many cases, otolaryngology and/or neurology referral.

Clinical efficiency of acoustic aural immittance measurements. The diagnostic efficiency of aural immittance measurements, including acoustic reflexes in the four test conditions, is unequalled in clinical audiology. With an investment of only a few minutes of test time and with equipment available in most audiology clinics, it is possible to objectively and sensitively identify facial nerve dysfunction and auditory dysfunction affecting a rather expansive region of the auditory system, from the middle ear to the lower brainstem. Patterns of findings for aural immittance measurements, made at the start of a hearing assessment and before behavioral hearing assessment, contribute to prompt and logical decisions on additional diagnostic testing and also appropriate patient management.

Otoacoustic emissions (OAEs)

Multiple evidence-based applications of OAEs

OAEs contribute importantly and in a truly unique way to the diagnosis of auditory dysfunction, even though they have essentially no value in defining the degree of hearing loss. Some of the many clinical applications of OAEs are listed in Table 3. Each of the applications listed in Table 3 is evidence-based. That is, research findings in support of the clinical application have been published in peer-reviewed journals. In terms of anatomic site sensitivity and specificity, in particular the detection and verification of outer hair cell dysfunction, OAEs have no rival in the hearing test battery. A full description of the generation and mechanisms of OAEs, OAE measurement and analysis, and the many evidence-based clinical applications of OAEs in children and adults is far beyond the scope of this brief review. A current review of the topic is available in a book devoted entirely to OAEs [12].

Simple guide to measurement and analysis

Pre-school hearing screening. Two selected clinical applications of OAEs are highlighted here. The first to be noted is the use of OAEs as a tool for hearing screening of pre-school children. There are many hundreds of publications describing newborn hearing screening with OAEs, but relatively little mention of their test performance or value in screening pre-school children [13,14]. Detection of hearing loss in children in the age range of 3 months to 5 years is just as critical as it is for newborn infants.

Table 3. Selected evidence-based applications of otoacoustic emissions (OAEs) for pediatric and adult patient populations (not in order of importance)

Pediatric	Adult
• Newborn hearing screening • Diagnosis of auditory dysfunction in infants and young children – Differentiation of site of auditory dysfunction – Identification of auditory neuropathy – False hearing loss • Monitoring ototoxicity • Pre-school and school screenings	• Diagnosis of cochlear versus neural auditory dysfunction • Identification of false and exaggerated hearing loss • Industrial and military hearing screening and conservation • Identification and monitoring of auditory dysfunction in noise/music exposure • Diagnosis and management of tinnitus and hyperacusis

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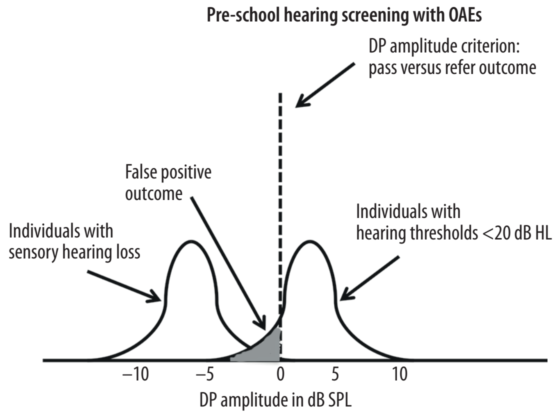

Figure 10. A decision criterion (Pass or Refer) when using DPOAE amplitude to screen pre-school children for hearing loss

There are four compelling reasons why screening is important for hearing loss after the neonatal period: 1) Hearing loss in young children can have a major negative impact on speech/language acquisition, social development, emotional and psychosocial status, and pre-academic skills, such as reading readiness; 2) Pre-school hearing screening permits detection of those young children with hearing loss who did not undergo hearing screening in the nursery before hospital discharge; 3) Screening during pre-school years detects progressive or delayed-onset cochlear hearing loss, a major contributor to the increased prevalence of hearing loss at school age; 4) Children who did not pass newborn screening and who were 'lost to follow-up' in EHDI programs are found with pre-school OAE screening; and 5) Ongoing screening efforts detect conductive hearing loss secondary to middle ear disease.

The limitations of pure tone hearing screening in the pre-school years are well appreciated, especially by audiologists and others who have attempted this challenging task. Drawbacks to pre-school hearing screening include poor reliability, the adverse effect of a variety of test actors (e.g., cognitive status of the child, noise in the test setting, and skill and experience of the tester), a sizable proportion of children who cannot be validly tested, and considerable test time [14].

In contrast, OAEs are quick, technically rather simple and, perhaps most importantly, not influenced by troublesome listener variables. Research indicates that OAEs stand up well to the traditional pure tone hearing-screening standard when certain assumptions are met [6,13,14],. Reliance on a simple signal-to-noise ratio (SNR) criterion for a pass or refer outcome is not sufficient. A DPOAE hearing screening test is enhanced with more rigorous criteria for a pass outcome, such as a SNR of >6 dB plus an absolute distortion product amplitude of >0 dB SPL.

This point is shown in Figure 10 with distributions for normal hearing versus hearing impaired persons as a function of DPOAE amplitude. DPOAE amplitude is plotted on the *x*-axis. Distributions of 'Pass' and 'Refer' outcomes are shown in relation to the criterion for a Pass outcome. Two criteria must be met for a Pass outcome: 1) A SNR of ≥6 dB confirming the presence of a DP; and 2) minimal DP amplitude of 0 dB, representing the lower limit of normal DP amplitude. The use of these combined criteria for concluding a Refer result detects almost 100% of ears with pure tone hearing thresholds of >20 dB HL.

OAEs in tele-audiology. Another exciting advance in clinical application is remote measurement of OAEs via tele-health techniques [15,16]. Investigation of newborn hearing screening with DPOAEs showed no difference in findings for on-site face-to-face hearing screening versus hearing screening remotely with tele-medicine technology. In other words, hearing screening with DPOAEs conducted remotely was validated against the conventional approach for hearing screening with these technologies. A trained technician or facilitator can perform hearing screening with an OAE device with immediate electronic storage to an Internet-accessible file. An audiologist then reviews OAE hearing screening outcomes remotely with an official reporting of the results. Asynchronous tele-audiology application of OAEs in hearing screening is inexpensive and highly efficient.

Auditory brainstem response (ABR)

Introduction to auditory evoked responses

The published literature on the multiple and varied applications of auditory evoked responses in clinical audiology is vast [2]. There is also remarkable accumulated clinical experience with electrophysiological responses elicited from the cochlea, auditory brainstem, and auditory cerebral cortex. ECochG was the first auditory evoked response to be discovered and applied clinically. Reports of the use of ECochG in the objective hearing assessment of difficult-to-test children date back to the 1960s. With the discovery of ABR, however, ECochG fell out of favor as an objective technique for auditory assessment. However, ECochG soon re-emerged as an electrophysiological procedure contributing to the diagnosis of Ménière's disease. In the 1980s, ECochG techniques were first applied in intra-operative neurophysiological monitoring during surgery that was apt to put the auditory system at risk. Most recently we have witnessed a resurgence of interest in ECochG as a critical test in the accurate diagnosis of ANSD, in particular the differentiation of pre-synaptic inner hair cell versus post-synaptic neural sites of auditory dysfunction [17]).

Auditory evoked responses are also useful in objectively evaluating function at the other end of the auditory system. Cortical auditory evoked responses were first recorded in 1939, less than 10 years after the discovery of ECochG. The literature on cortical auditory evoked responses, including the auditory middle latency response, the auditory late response, the auditory P300 response, and the mismatch negativity (MMN) response is even more extensive than for ECochG or ABR.

Many thousands of papers confirm the value of tonal and speech evoked cortical evoked responses in objective assessment of central auditory nervous system function in diverse patient populations, ranging from a variety

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of neuropsychiatric diseases to children and adults with suspected auditory processing disorders. One of the most recent and exciting applications of the auditory late response is objective evaluation of cortical functioning in infants with sensory hearing impairment and ANSD who are undergoing habilitation with hearing aids and/or cochlear implants. Readers are referred to original research publications and current textbooks [2, 18] for detailed information on cortical auditory evoked responses. The following review focuses exclusively on the ABR.

45 Years of ABR research and clinical application

In the 45 years since Jewett and Williston [19] discovered the ABR, effects of virtually every possible measurement parameter on the response have been investigated and described in the literature. Early studies of pediatric ABR application laid the foundation for the later emphasis on newborn hearing screening and for today's protocols for frequency-specific electrophysiological estimation of the audiogram using ABRs evoked with tone burst stimuli and the ASSR. Bone conduction ABR is now within the standard-of-care for hearing assessment of infants and young children [1,20]. The ABR is unrivaled as a powerful diagnostic tool for an objective assessment of infant hearing and the identification and diagnosis of auditory neuropathy. A short list of clinical applications of ABR also includes:

- Automated newborn hearing screening;
- Diagnosis of cochlear versus neural disorders in children and adults;
- Neurophysiological monitoring in the operating room;
- Neurophysiological monitoring of head-injured patients in the neuro-intensive care unit;
- Remote diagnosis of infant hearing loss via tele-audiology;
- Measurement of the neural representation of speech processing within the auditory brainstem.

A PubMed search (*www.nlm.nih.gov*) with the key words "auditory brainstem response" now produces over 12,200 articles. More than 300 articles were published annually from 1990 through 2010, but the number of papers on ABR has exceeded 400 per year since then. Admittedly, this vast literature consists of at least 4000 papers describing ABR in non-human animal species. Still, it would be reasonable to ask whether after 45 years and approximately 8000 reports of clinical ABR studies there is any new information that is worthy of publication. Multiple and varied lines of research contribute to the unabated volume of publications on ABR. Some of the articles describe application of the ABR in assessment of new clinical entities, often patients with rarely encountered diseases or genetic disorders.

Other investigators report technological advances in instrumentation that may lead to enhancement in ABR measurement or analysis. Remote ABR measurement via tele-audiology is now an option for provision of clinical services in regions where audiology expertise is lacking [21]. A sizeable proportion of recent publications are devoted to innovative stimuli for eliciting ABRs, including chirps and complex stimuli like speech. A substantial number of studies reported in the literature during the last decade are 'cover studies' conducted in developing countries or countries with emerging audiology and hearing research professions, such as India, China, Brazil, and Iran to name a few. These papers describe replications of early investigations, but with current ABR instrumentation and with data from much larger samples of subjects.

The review that follows highlights the recent use of chirp stimuli in clinical measurement of the ABR in infants and young children. It is excerpted from the *eHandbook of Auditory Evoked Responses* [2].

Chirp click stimulus

What is a chirp? As just noted, there is consensus that the ABR evoked with conventional click stimulation is dominated by activation of the basal region of the cochlea. Attempts to enhance the contribution of other regions of the cochlea to ABR generation include the creation of rather unique types of stimuli called 'chirps'. Chirps are sounds that sweep rapidly from low to high frequencies or vice versa. Upward chirps are applied in recording auditory evoked responses. The term chirp is derived from the sound that birds and some other animals produce. The *chirp stimulus* is designed mathematically "to produce simultaneous displacement maxima along the cochlear partition by compensating for frequency-dependent traveling-time differences" [22].

Since the 1980s, various authors have reported detailed technical descriptions and mathematical models for chirp stimuli for use in measurement of auditory evoked responses [2]. In theory, the chirp version of the click stimulus optimizes synchronization across a broad frequency region at high and low intensity levels, yielding a more robust ABR than the conventional click stimulus. A detailed explanation of the model of cochlear biomechanics and the mathematical functions important in the rationale for and generation of chirps is far beyond the scope of this discussion. The article authored by Fobel et al. [2] provides a useful source of background information on the topic.

Rationale for chirp stimuli. The overall physiological goal with chirp stimuli is to simultaneously activate a wide range of the cochlea from base to apex. This is achieved with temporal compensation for the traveling wave delay as it moves from the high to the low frequency portions of the cochlea. Estimations of the traveling wave delay are available from extensive analysis of the differences in the latency of wave V for ABRs evoked with high frequency versus low frequency tone burst stimulation, and also from systematic study with the derived-band technique for isolating contributions to the ABR of different frequency regions.

Dr Manny Don of the House Research Institute, and others, studied the effects of ipsilateral high pass masking on 'cochlear response times' associated with traveling wave distance and velocity along the basilar membrane [23]. Traveling wave time from higher frequency regions of the cochlea to lower frequency regions is approximately 5 ms. Factors influencing traveling wave times in the cochlea include stimulus intensity level, hearing loss, and subject age. Most early research on chirp stimuli was conducted with ABRs recorded at moderate to low intensity levels from normal hearing adult subjects.

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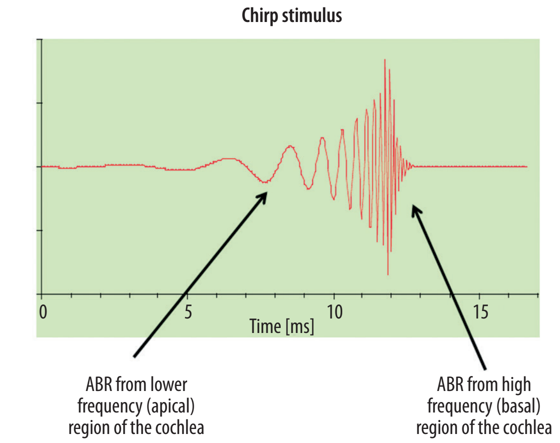

Figure 11. Chirp stimulus used to evoke ABR. The early portions of the waveform activate more apical regions of the cochlea and later portions activate more basal regions

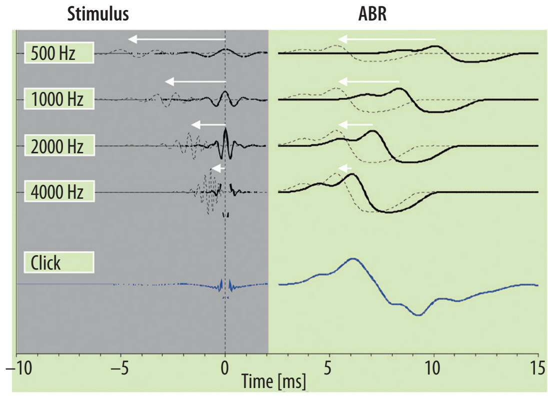

Figure 12. How temporal compensation leads to an enhancement of the amplitude of a chirp-evoked ABR

The following is a clinically oriented and admittedly oversimplified description of broadband chirp stimuli, or chirp versions of click stimuli that have been used to elicit ABRs since the pioneering studies of Jewett & Williston in the early 1970s. Tone burst versions of chirps are described later in the same chapter within a discussion of frequency-specific stimuli. Briefly, the spectrum of the chirp click stimulus, like the conventional click stimulus, includes energy across a wide frequency region. With chirp stimulation, however, lower frequency energy is presented earlier than higher frequency energy.

A click chirp stimulus is illustrated in Figure 11. Low frequency portions of the stimulus appear first with a progressive increase in time as frequency is increased. Rising or upward frequency chirp stimuli are mathematically designed to compensate temporally for travelling wave delays. Higher frequency energy in chirp stimuli is delayed relative to lower frequency energy. Low frequency energy is essentially given a 'head start' as it begins its journey to the distant apical region of the cochlea. Mid-frequency energy in the region of 1000 Hz is presented milliseconds later and high frequency energy is delivered last. Waves traveling to each of the frequency regions reach their cochlear destinations at the same time, harnessing synchronous activity from most of the cochlea, not just the high frequency portion.

Another way of considering the concept of click chirp stimulation is to describe the effect on ABR waveforms. Without the temporal compensation produced with chirp stimuli, ABR wave V latency for stimulation in the region around 500 Hz is about 5 ms longer than the latency for 4000 Hz stimulation. Delivering lower frequency stimulus energy to the cochlea about 5 ms earlier than higher frequency stimulus energy essentially produces a corresponding shift in wave V latency.

ABR wave V latency evoked with lower frequency stimulation occurs earlier than it typically does and at the same time as the ABR wave V for higher frequency stimulation. The overall effect is to evoke an ABR wave at the same latency for stimuli in each frequency region. Amplitude of the chirp-evoked ABR is enhanced with the addition of superimposed wave V components for stimuli within each of the frequency regions. This complex temporal compensation process is illustrated in Figure 12.

Larger amplitudes for ABR wave V is not a trivial goal. The significant clinical advantages of a larger wave V, often a doubling in amplitude and an increase in the ABR versus noise difference, include: 1) more confident identification of wave V near the minimum response level or threshold; 2) detection of an ABR at lower intensity levels for more accurate estimations of thresholds; and 3) decreased test time required for recording ABRs.

Factors influencing chirp-evoked ABR. As noted already, early research on chirps focused almost exclusively on data collected at low to moderate intensity levels in carefully controlled laboratory settings from normal hearing adults. Early and more recent clinical investigations in normal hearing infants and young children have consistently confirmed larger amplitudes for ABR wave V at low to moderate intensity levels [2].

There are well-appreciated effects of intensity level on the duration of the chirp stimulus and also on the cochlear mechanics and physiology underlying level-dependent changes in cochlear traveling wave delays. Mathematical formulas and models developed for low intensity chirp stimuli are not appropriate for higher intensity levels. Level-dependent variations in ABRs evoked with chirp stimuli pose a clinical problem. The amplitude of ABRs recorded at high intensity levels with chirp clicks developed for low-level stimulation is actually smaller than ABR amplitudes for conventional click stimuli. The clinical value of chirp stimuli in recording ABRs would certainly be diminished if it were limited to infants and young children with normal hearing or at most a mild hearing loss. The amplitude of ABR wave V tends to decrease as hearing loss increases. Therefore, one might argue that enhanced ABR amplitude with chirp stimulation would be of greatest value in patients with hearing loss.

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Table 4. Strengths and weakness of ASSR as a clinical tool

Advantages	Potential disadvantages
• Frequency-specific signals are employed for estimation of thresholds at audiometric frequencies from 250 to 8000 Hz • Frequency-specific auditory thresholds can be estimated with air conduction and bone conduction signals • Stimulus intensity levels as high as 120 dB HL can be used in eliciting frequency-specific thresholds. The ASSR is therefore useful for electrophysiological assessment of severe to profound degree of hearing loss in infants and young children • ASSR detection and analysis is automated and statistically based. Clinician experience in waveform analysis is not necessary • Clinical devices are available from multiple manufacturers	• ASSR recording requires a very quiet state of arousal. Movement artifact and interference may preclude testing or may invalidate results with overestimation of actual auditory threshold levels in young children who are not asleep. ASSR usually requires the patient sleep naturally or with sedation. Anesthesia is sometimes necessary for valid ASSR assessment of hearing sensitivity • The influence of deep sedation and anesthesia on the ASSR evoked by high modulation frequencies (e.g., >60 Hz) requires further investigation. Sedation and anesthesia invalidates threshold estimations for ASSR evoked with slow modulation frequencies (e.g., <60 Hz) • Modest discrepancies between ASSR thresholds and either behavioral and/or ABR thresholds are reported in the literature • Discrepancies between ASSR thresholds and behavioral thresholds are possible for patients with conductive hearing loss • Estimation of ear-specific thresholds with bone conduction signals requires the use of masking to the non-test ear. Unlike ABR, there is no biological marker for test ear with ASSR. • There is little site-specific information for patients with hearing loss since the ASSR waveform cannot be analyzed. ASSR cannot be used to differentiate sensory versus neural auditory dysfunction • Absence of ASSR does not differentiate between profound sensory hearing loss versus ANSD • In the USA there is no current procedural billing code specifically for ASSR

Level-specific (LS) chirps offer an option for obtaining the benefits of chirp stimulation at a variety of different intensity levels. LS chirps are based on a unique level-specific delay model [24]. In contrast to fixed chirps developed for use only at lower intensity levels, the duration of LS-chirps changes with stimulus level. The LS-chirps at each intensity level are based on a different delay model with the goal of eliciting the largest possible ABR amplitude. Intensity levels are calibrated using International Standards Organization reference values (dB p-p.e. RETSPL). The standard "specifies reference hearing threshold levels for tests signals of short duration applicable to the calibration of audiometric equipment where such signals are used."

Several additional comments about chirps are worth noting at this juncture. Chirp stimuli and their effectiveness in enhancing the amplitude of ABRs are dependent on specific mathematical formulas and models. Not all chirps are created the same way. The forgoing discussion focused mostly on CE-chirps developed by and described in the publications of Claus Elberling (CE) and colleagues who designed stimuli called 'CE-chirps'. It is reasonable and advisable to inquire about the development and clinical research evidence in support of chirps before applying them to ABR measurement from patients.

The second point has to do with clinical applications of chirp stimuli. The focus of this discussion has been the use of air conduction chirp stimuli in recording ABR in infants and young children. Chirp stimuli also appear to contribute to the additional applications of ABR, such as early detection of retrocochlear auditory dysfunction and bone conduction ABR. And chirp stimuli play a role in the measurement of ASSR and cortical evoked responses.

Role of ASSR in the pediatric test battery

An appreciation of the strengths and weakness of the ASSR, listed in Table 4, guides decisions on when it should be applied clinically and its role in the diagnostic process. Consistent with the cross-check principle and in common with behavioral hearing tests and other electrophysiological auditory procedures, ASSR should not be recorded in isolation but, rather, as a component in an appropriate test battery. The literature reveals papers describing diagnostic applications of ASSR in various pediatric populations in addition to estimation of auditory thresholds, including:

- Objective assessment of hearing aid gain in the sound field environment;
- Benefit from cochlear implantation;
- Diagnosis of ANSD;
- Detection and diagnosis of neurological auditory disorders;
- Assessment of auditory processing in dyslexia.

This is a good place to dispel two common misconceptions. ASSR and ABR are not competitive electrophysiological procedures. In other words, the clinical decision is not to record either tone burst ABR or ASSR. The two procedures are complementary. Diagnosis of hearing loss and plans for intervention are often based on results of some combination of ABR recordings and ASSR recordings in the same child, along with findings for other objective auditory tests. Also, clinical experience suggests that test time is equivalent for tone burst ABR and ASSR

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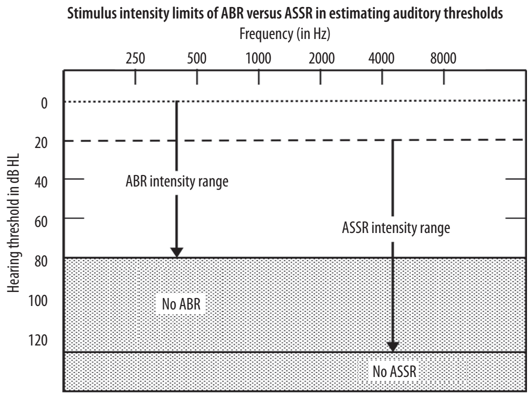

Figure 13. ASSR can be used to estimate auditory thresholds in patients who have severe to profound hearing loss which exceeds the intensity limits of ABR

measurement. Claims that tone burst ABR assessment is excessively time-consuming and ASSR requires relatively little test time are not supported with clinical research or experience. A skilled clinician can complete a frequency-specific ABR assessment for both ears in 30 minutes or less with a sleeping patient, a clear plan of action, a carefully constructed tone burst protocol including options like chirp stimuli, and consistently good use of test time.

The ASSR, like the ABR, offers an opportunity to estimate auditory thresholds in infants and young children who cannot be properly assessed with behavioral audiometry techniques. The ASSR has a distinct edge over behavioral audiometry, and in several respects even the ABR, in this clinically challenging patient population. One strong feature of the ASSR, in comparison to the ABR, is its capacity to define severe to profound hearing loss, that is, estimating hearing thresholds within the range of 80 to 120 dB HL, as illustrated in Figure 13. The limitation of ABR in defining the degree of severe-to-profound hearing loss >90 dB HL is well appreciated by clinicians and well documented in the literature.

ABR and ASSR can each contribute importantly and rather uniquely to diagnostic auditory assessment of children. However, it is important to keep in mind that neither the ABR nor the ASSR are actually tests of hearing. Each technique must be applied within an appropriate evidence-based test battery consistent with the cross-check principle [4] and with clinical guidelines for pediatric hearing assessment [1].

Chirp-evoked ASSR in infants

Papers are beginning to emerge describing comparison of chirp-evoked ASSR with hearing thresholds and with ABR thresholds [25]. Preliminary evidence suggests that chirp-evoked ASSRs are equivalent to (and perhaps superior to) conventional ASSR techniques for quick and accurate estimation of behavioral thresholds in adults and, most importantly, in infants and young children with normal hearing or hearing impairment [25,26]. Efficiency and accuracy of the chirp-evoked ASSR technique appears to be related to increased amplitude. One of the major clinical benefits of chirp stimuli is reduction in test time. For example, Mühler and colleagues [26] in a study with normal hearing and "mildly to moderately hearing impaired" adult subjects reported a mean time of 18.6 minutes for completion of ASSR for four test frequencies in both ears, using a "semiautomatic adaptive algorithm." Such brief test times open up the possibility of performing ASSR assessments in reasonably cooperative infants and young children who are sleeping naturally without the assistance of sedation or anesthesia.

Concluding comments

Each of the objective measures reviewed here offers certain compelling advantages for the auditory assessment of infants and young children. However, the diagnostic power of objective auditory tests is fully realized only when they are applied in combination. Careful analysis of findings for an objective auditory test battery almost always yields prompt and precise description of auditory status, and it often leads to accurate diagnosis of auditory dysfunction. The key to meaningful analysis of findings for a test battery is the recognition of patterns associated with major auditory disorders [27]. This is not a novel concept. It is simply the modern day implementation of the 40-year old cross-check principle.

Our review concludes with examination of selected patterns of auditory findings displayed in Table 5. Normal test findings are indicated with a minus symbol. The plus symbol is used to indicate abnormal findings, those that are positive for a disorder. In addition to the objective measures discussed in this review, the table includes representative findings also for ECochG and cortical auditory evoked responses. Even a cursory glance of information in the columns representing various auditory disorders in Table 5 confirms that none of the patterns is duplicated. Each pattern of findings is uniquely associated with a specific disorder.

Exclusive reliance on only one or two objective auditory measures often results in equivocal outcome. That is, the patient's diagnosis is not clear, and their hearing loss could be due to one of multiple disorders. With an appropriately complete test battery, however, auditory disorders can be confidently differentiated. An example might be useful to clarify this statement. Let us assume a 3-month-old infant undergoes follow-up diagnostic auditory assessment after failing neonatal hearing screening. The patient is a graduate of the intensive care nursery whose history includes management with several potentially ototoxic drugs plus neurological risk factors such as premature birth and asphyxia. Concerns include the possibility of cochlear hearing loss, neural disorder (e.g., a form of ANSD), or possibly central auditory nervous system dysfunction. Findings for common objective tests, such as acoustic reflexes, OAEs, ABR, and perhaps ECochG clearly point to the most likely auditory disorder.

In summary, the application of a complete objective test battery is the most effective and efficient strategy for prompt and accurate diagnosis of auditory dysfunction and hearing loss in infants and young children. Objective auditory assessment is essential for a successful EHDI program. Hearing assessment with a collection of objective auditory tests defines the standard of care in pediatric audiology.

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Table 5. Patterns of objective test findings in selected types of auditory disorders

Test	Auditory disorder						
	Normal	Middle Ear	Cochlear		ANSD	CNS	APD
			OHC	IHC			
Aural immittance							
Tympanometry	–	+	–	–	–	–	–
Acoustic reflexes*	–	+	±	±	+	+	±
OAEs	–	+	+	–	–	–	–
ABR: click stimuli							
Air conduction	–	+	±	±	+	–	–
Bone conduction	–	–	±	±	+	–	–
ABR: tone burst stimuli	–	±	±	±	±	–	–
ASSR	–	±	±	±	±	–	±
ECochG							
CM	–	+	+	–	–	–	–
SP	–	+	–	+	±	–	–
AP	–	+	±	±	+	–	–
Cortical auditory responses	–	±	–	–	±	+	+

* Acoustic reflexes elicited with broadband noise (BBN). Key to symbols: – = normal; + = abnormal; ± = variable.

References:

1. American Academy of Pediatrics, Joint Committee on Infant Hearing. Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs. Pediatrics; 120: 898–921.
2. Hall JW III. eHandbook of auditory evoked responses. Amazon.com, Kindle Direct Publishing, 2015 (*http://www.amazon.com/dp/B0145G2FFM*).
3. Galambos R, Hecox K. Clinical applications of the auditory brainstem response. Otolaryngol Clin North Am, 1978; 11: 709–22.
4. Jerger JF, Hayes D. The cross-check principle in pediatric audiometry. Archiv Otolaryngol, 1976; 102: 614–20
5. Hall JW III. Aural immittance measures in audiology: More useful now than ever before. Hear J, 2010; 63: 15
6. Hall JW III, Swanepoel D. Objective assessment of hearing. San Diego, Plural Publishing, 2010.
7. Feeney MP, Hunter LL, Kei J, Lilly DJ, Margolis RH, Nakajima HH, Neely ST et al. Consensus Statement: Eriksholm workshop on wideband absorbance measurements of the middle ear. Ear Hear, 2013; Suppl. 1.
8. Jerger J, Burney P, Mauldin L, Crump B. Predicting hearing loss from the acoustic reflex. J Speech Hear Disord, 1974; 39: 11–22.
9. Hall JW III, Berry GA, Olson K. Identification of serious hearing loss with acoustic reflex data: Clinical experience with some new guidelines. Scand Audiol, 1982; 11: 251–55.
10. Kei J. Acoustic stapedial reflexes in healthy neonates: Normative data and test–retest reliability. J Am Acad Audiol, 2012; 23: 46–56.
11. Jerger JF, Jerger S and Hall JW III. A new acoustic reflex pattern. Archiv Otolaryngol, 1979; 105: 24–28.
12. Dhar S, Hall JW III. Otoacoustic emissions: Principles, procedures & protocols. San Diego: Plural Publishing, 2011
13. Kreisman BM, Bevilacqua E, Day K, Kreisman NV, Hall JW III. Preschool hearing screenings: A comparison of distortion product otoacoustic emission and pure tone protocols. J Ed Audiol, 2013; 19: 49–57.
14. Hall JW III. Effective and efficient preschool hearing screening: Essential for successful early hearing detection and intervention (EHDI). Journal of Early Hearing Detection and Intervention, 2016; 1: 1–12.
15. Swanepoel D, Hall JW III. A systematic review of telehealth applications in audiology. Telemedicine and e-Health, 2010; 16: 181–200.
16. Swanepoel D, Clark JL, Koekmoer D, Hall JW III, Krumm M, Ferrari DV, McPherson B et al. Telehealth in audiology: The need and potential to reach underserved communities. Int J Audiol, 2010; 49: 195–202.
17. McMahon CM, Patuzzi RB, Gibson WP, Sanli H. Frequency-specific electrocochleography indicates that presynaptic and postsynaptic mechanisms of auditory neuropathy exist. Ear Hear, 2008; 29: 314–25.
18. Picton TW. Human auditory evoked potentials. San Diego: Plural Publishing, 2011
19. Jewett, DL, Williston JS. Auditory evoked far fields averaged from the scalp of humans. Brain, 1971; 4: 681–96

DOI: 10.17430/897777

20. NHSP. Newborn Hearing Screening Program. "Guidance for Auditory Brainstem Response testing in babies", 2013 *http://www.thebsa.org.uk/wpcontent/uploads/2014/08/NHSP_ABR-neonate_2014.pdf*.
21. Ramkumar V, Hall JW III, Nagarajan R, Shankarnarayan VC, Kumaravelu S. Tele-ABR using a satellite connection in a mobile van for newborn hearing testing. J Telemed Telecare, 2013; 19: 233–37.
22. Fobel O, Dau T. Searching for the optimal stimulus eliciting auditory brainstem responses in humans. J Acoust Soc Am, 2004; 116: 2213–22.
23. Don M, Ponton C, Eggermont J, Masuda A. Auditory brainstem response (ABR) peak amplitude variability reflects individual differences in cochlear response times. J Acoust Soc Am, 1994; 96: 3476–91.
24. Kristensen SGB, Elberling C. Auditory brainstem responses to level specific chirps in normal-hearing adults. J Am Acad Audiol, 2012; 23: 712–21.
25. Venail F, Artaud JP, Blanchet C. Refining the audiological assessment in children using narrow band CE-chirp-evoked auditory steady state responses. Int J Audiol, 2015; 54: 106–13.
26. Mühler R, Mentzel K, Verhey J. Fast hearing-threshold estimation using multiple auditory steady-state responses with narrow-band chirps and adaptive stimulus patterns. ScientificWorldJournal, 2012; 2012: 192178.
27. Hall JW III. Introduction to audiology today. Boston, Pearson Educational, 2015

DOI: 10.17430/897777