CHAPTER

06 Electrophysiological Evaluations



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'Fast is fine, but accuracy is everything.'

~Wyatt Earp

Summary

This chapter will provide insights into the value of physiological diagnostic tools, including Auditory Brainstem Response (ABR) and Otoacoustic Emissions (OAE), for the paediatric population. It will offer guidance on the equipment and setup for optimal testing, tailoring the test battery to gather essential information and providing practical recommendations to simplify patient handling during testing. The insights provided in this chapter were acquired during several years of hands-on experience

Learning objectives

From this chapter, the reader will be able to:

1. Describe the clinical value of diagnostic OAEs and ABRs in paediatric hearing evaluations.
2. Demonstrate an understanding of the impact that the test environment and equipment setup can have on patient comfort, information-gathering efficiency and the accuracy of test results.
3. Understand the value of patient handling in a manner that delivers the most reliable and accurate results, without causing unnecessary harm or discomfort.
4. Demonstrate an understanding of how the different test results can form a cohesive clinical picture of the potential site(s) of dysfunction in each (unique) paediatric case.

Introduction

Hearing healthcare professionals (HHPs) are trusted by parents and caregivers to evaluate their child's hearing and provide them with answers and guidance. Understanding and respecting the fact that most parents will be very anxious by the time they enter a diagnostic clinical setting will highlight the gravity of the HHP's role in their child's life.

Physiological diagnostic tools like Auditory Brainstem Response (ABR) and Otoacoustic Emissions (OAE) offer valuable insights into the functioning of a young patient's auditory system. It allows the opportunity to cross-check various findings and physiologically confirm the presence or absence of a variety of dysfunctions, especially in 'difficult-to-test' populations. According to the clinical guidance document of the American Academy of Audiology (2020), 'difficult-to-test' is a term that refers to 'children with developmental delays or other conditions that affect communication, and cooperation'.

Physiological tests help to pinpoint the potential site of the lesion, provide guidance to do further testing when needed and support the HHP in decision-making regarding audiological management. This chapter will provide practical insights on optimising the test environment and patient set-up, tailoring the test battery for the specific patient population, prioritising

information gathering in an efficient manner and approaching these test procedures with respect and caution.

This chapter is divided into three sections:

- Electrophysiological equipment setup and environmental considerations
- Otoacoustic emissions (OAE)
- Auditory Brainstem Response (ABR) and Auditory Steady State Response (ASSR)

Electrophysiological Equipment Setup and Environmental Considerations

The accuracy of both OAE and ABR evaluations is heavily influenced by the test environment and equipment setup. The room location within the clinical setting will have a significant impact on successful testing when performing diagnostic OAEs and ABRs, and a quiet room with good acoustics will always be preferable. It is thus important to select a location that is further away from high levels of 'foot traffic' and general ambient noise (for example, the reception area, bathrooms and waiting rooms). Whereas a sound-treated room may be ideal for minimising background noise and reverberation, adding carpets and curtains is a simple way to improve the test environment for OAE and ABR measurement.

When conducting OAE measurement, the test space should be arranged in a way to facilitate easy movement between the equipment and the patient. The room should also allow for a comfortable test position for the patient and caregiver while the measurement is in progress. Seating options will differ based on the age of the patient. Smaller infants can be tested comfortably in their parents' arms or in their baby carrier, and a toddler might be more comfortable seated on their parent's lap or independently on a chair. Therefore, the physical space and size of the test room must be a consideration when selecting a room for OAE measurement.

The workspace for ABR measurements is usually carefully prepared to allow for easy access and optimal patient positioning. Parker, Slack and Skoe (2020) found that a simulated home environment delivered comparable quality ABR recordings compared to traditional laboratory settings. However, although this may be true for ABR, ambient noise levels might have a more pronounced impact on the integrity of DPOAE and TEOAE measures, as highlighted by Kanji (2019). Therefore, if the test system allows for both ABR and OAE testing, the ambient noise levels preferable for OAE measurements should be taken into consideration when choosing the location of the test room.

Another critical consideration, more so for ABR testing, is the electrical circuit characteristics of the clinical room. Electrical circuitry might influence the selection of a suitable room within a larger practice or hospital setting. Within a hospital setting, proximity to the radiology department, operating theatres, elevators, communication systems and anaesthesiology monitoring equipment may increase the likelihood of encountering troublesome artifacts on a regular basis (Stegeman and De Weerd 1990). According to Hall and Swanepoel (2010), accurate electrophysiological estimation of hearing thresholds is dependent on the ability to detect a signal in the presence of noise. Noise interferes with the recording in several ways and might arise from multiple varied sources, for example, airborne electrical or electromagnetic energy, electrical circuit interference, rectified radio frequency noise, physiological electrical artifact originating from the patient's nervous system and a myogenic (muscle) artifact (Hall and Swanepoel 2010). Although newer generation ABR equipment can filter out unwanted artefacts and noise, it is recommended to still make every effort to minimise interference where possible. Relying on these filtering technologies alone to improve waveform quality may result in 'smoothing' waveforms to an extent where analysis of the waveforms becomes very challenging and errors occur more frequently.

Various options are available to facilitate a comfortable patient position, even in small clinical spaces. Generally, the ABR clinical setting will require an examination bed, especially if sedation is being used. However, a comfortable couch or 'lazy boy' has proven to be useful in smaller spaces. In some cases, the test procedure can be performed while the patient is resting in a baby carrier or in the arms of the caregiver/parent. This is mainly to ensure that the patient is as comfortable as possible and to ensure that they remain asleep for the duration of the test. It is important to be cognisant of other physical comfort factors such as body temperature (too cold or too warm), movement restriction or even anxiety, as these factors



Treasures of Wisdom

Electrical environment testing

- Before selecting a clinical environment or performing a test in theatre, perform a 'baseline' environmental measure by placing the connected test electrodes in a glass of tap water and run a test. Monitor the EEG and rejection rate for signs of high levels of electrical interference in the test environment. The use of tap water ensures that the conduction medium (water) has ion flow. Bottled water does not have ion flow and will render this investigation unsuccessful.

- Never use an extension cord when performing ABR measurement – extension cords are notorious for creating high levels of electrical artifact.

may also influence the patient's ability to remain calm and quiet for the duration of the test.

Before any electrophysiological evaluation appointment, the equipment should be checked for optimal functioning, and all necessary consumables should be made easily accessible to facilitate quick and efficient testing. These steps should form part of the clinical 'biological checks' in general, and are non-negotiable. The last thing any clinician wants to face is faulty equipment or lack of consumables after their patient is already asleep and/or sedated.

Otoacoustic Emissions

OAEs were first described in the 1970s by David Kemp and are considered a well-established test procedure in the general diagnostic test battery. The presence of cochlear emissions was first hypothesised in the 1940s based on complex mathematical models of cochlear nonlinearity, but it was only truly proven in the late 1970s. Only then could low-intensity sounds that are produced by the cochlea could be successfully recorded using highly sensitive microphones (Campbell and Mullin 2023).

OAEs offer valuable insight into the outer hair cell functioning of the cochlea and are particularly helpful in understanding the overall clinical picture of young patients. To employ OAEs effectively in practice, HHPs must understand the mechanism by which OAEs are generated. According to Prieve and Lamson (2014), OAEs reflect the functioning of mechanical processes in the inner ear (specifically the outer hair cells) and are a byproduct of the cochlear amplifier rather than a reflection of the cochlea's sensory or neural functioning. According to Siegel and Kim (1982), OAEs may be present despite a severed acoustic nerve (CN VIII) and are typically absent in persons presenting with cochlear hearing loss (HL) (Siegel and Kim 1982).

OAE testing is a relatively simple test procedure, and technological advancements offer significantly improved ease of application and faster test speed. It is also relatively cost-effective and does not require multiple consumables to administer successfully. OAEs have shown tremendous diagnostic value since the early 1980s, and this technique has been enthusiastically embraced worldwide ever since.

There are four known types of OAEs, namely, spontaneous otoacoustic emissions (SOAEs), transient otoacoustic emissions (TEOAEs), distortion product otoacoustic emissions (DPOAEs) and sustained-frequency otoacoustic emissions (SFOAEs) (Campbell and Mullin

2023). Clinically today, two types of OAEs are mainly utilised in practice: TEOAEs and DPOAEs. The choice of the type of emission will depend on factors such as the patient's age range, the type of evaluation (screening vs diagnostic) or the target frequencies. The major differences between TEOAEs and DPOAEs are summarised in Table 6.1.

Table 6.1: Differences between TEOAEs and DPOAEs

TEOAEs	DPOAEs
• Mainly used in newborn hearing screening procedures	• Offers more frequency-specific information
• Most sensitive at test frequencies between 500 Hz and 4 kHz	• More sensitive to higher frequency regions (>4 kHz), which is particularly valuable for ototoxic monitoring
• More limited frequency specificity: TEOAE response is linked to a larger region of the cochlea. The detection and frequency content are therefore influenced by the status of the entire cochlea.	• Generally assessed as a function of primary tone frequencies. Acoustic distortion products (ADPs) are measured concurrently with the presentation of two pure-tone stimuli (for example, primaries) in one ear. When two primaries are presented to a healthy cochlea, it will result in the production of intermodulation distortion products that are mathematically correlated to the primaries' frequencies.
• Measured using a click (broader band) stimulus	• Measured using narrowband filtering focused on the frequency of interest

TEOAEs	DPOAEs
• Generally present in cases where behavioural hearing thresholds are better than 30 dB HL without functional or neural components • Generally present if cochlear sensitivity in the region of 2 kHz–4 kHz is approximately 20–40 dB HL • May be reflected on a graph or audiogram, although this is mainly system specific. TEOAE testing generates a waveform that represents emissions produced by outer hair cells at a specific frequency in response to a stimulus. A clearly defined waveform suggests normal functioning of the hair cells, while a weak or absent waveform suggests a possible cochlear dysfunction.	• Results reflected on a DP-gram that resembles an audiogram, but should not be interpreted as such • Can be measured in ears with greater levels of HL.

Adapted from Campbell and Mullin (2023), Prieve and Lamson (2014), Prieve and Fitzgerald (2009) and Tzanakakis et al. (2016).

There are two categories of OAE equipment on the market today: systems predominantly designed for screening OAEs, and systems designed for diagnostic OAE measurement. Screeners are focused on identifying the possibility of HL and do not offer detailed information about the degree and/or nature of an HL. Diagnostic OAEs, however, provide more frequency-specific information for each ear (Prieve and Lamson 2014). A more detailed outline of screening OAEs is presented in Chapter 4.

The interpretation of diagnostic OAEs is beyond the scope of this chapter. Information on the interpretation of OAEs can be found in Otoacoustic Emissions: Principles, Procedures, and Protocols (Dhar and Hall 2017).

Patient state and positioning in OAE testing

Diagnostic OAEs are very sensitive to noise generated internally from the patient. A quiet and calm patient will make measuring OAEs simpler, faster and more effective. With experience, reading a patient's body language and mood will become easier, which will make the planning and the timing of the test battery flow' more successfully. If a patient seems drowsy during the case history interview with the parents, you may forgo the case history conversation and proceed straight to the OAE measurement. The case history can be returned to at a later stage.

With newborns and young infants, feeding may be useful as a calming strategy to be able to successfully insert the probe tip and initiate measurement. Bear in mind that testing may need to be paused intermittently when sucking, burping, crying or cooing occurs. Keeping the patient calm is the main priority, not only clinically, but also relationally. The HHP should rely on the patient's parent/caregiver to suggest the most comfortable position for the procedure, for example, in their arms, on their lap, in their car seat or in a stroller. It is important to remember that establishing a trusting relationship with the patient and parents/caregivers is just as important as obtaining results. If a patient is physically too restless or upset to obtain reliable results, then a follow-up appointment may offer a second opportunity to try again. OAE measurement is remarkably adaptable when the HHP gains confidence in its application and optimal patient positioning is possible when HHPs get a little creative.

When performing OAEs in toddlers or older children, their ability and willingness to co-operate might improve (although not always in the case of patients with special needs and physical disabilities). Older children should be given the autonomy to choose a comfortable position for the test procedure, for example, sitting independently in a chair or holding onto a parent or caregiver's hand. Offering a form of distraction may also be helpful to keep them calm and quiet for the duration of the test (for example, a muted iPad or tablet, story books, picture books). Whereas performing OAEs earlier in a test battery may be helpful with newborns or young infants to capitalise on their restful state, fatigue may conversely be helpful with older children to keep them quiet and calm for the objective measures. Fatigue caused by behavioural testing (visual reinforcement audiometry and play audiometry) may even result in a quieter and calmer patient. Being flexible in your approach is critical. Figure 6.1 shows examples of patient positions for optimised OAE measurement.

Figure 6.1: (a)



Figure 6.1: (b)



Figure 6.1: (a) Positioning and distraction of a small baby using a small soft toy, (b) Independent positioning of an older toddler, with an 'unconventional' distraction tool (calculator)

Measurement and factors influencing OAE interpretation

The first step is to perform an otoscopic examination and to confirm a clear and intact outer ear, ear canal and tympanic membrane (see Chapter 5). This is generally followed by tympanometry to evaluate the middle ear's integrity. Patient factors like occluding earwax, middle ear effusion and the presence of grommets or ventilation tubes might influence the ability to measure emissions reliably, and therefore any external factors that may influence the test must be identified before starting the measurement. The stimulus that evokes OAEs must travel through the middle ear to the cochlea and back. Therefore, any factors influencing the outer or middle ear will impact an OAE measurement (Prieve and Lamson 2014). According to Sanches et al. (2024), the combination of diagnostic OAE measures and tympanometry is valuable for identifying and confirming changes in middle ear integrity as the measured OAE amplitudes are directly related to the integrity of the functioning middle ear and tympanic membrane. It should further be noted that successfully conducting an OAE measurement hinges on a secure probe fit. An unstable probe fit can result in poor repeatability, high noise levels and unreliable results.

Generally, sensitivity for OAEs is lower in the lower frequency and higher frequency ranges and internal and external noise may impact the validity of the measurement (Prieve and Lamson 2014). Therefore, limiting the test frequency range to a spectrum of 2 kHz – 4 kHz may be helpful if a patient is restless and co-operation is limited. Remember that some information is preferable to no information. HHLs are encouraged to perform at least two test runs to confirm repeatability and reliability. Recordings should present with a sufficiently low noise floor across the test frequency range to be accepted as valid and reliable.

The need for age-appropriate normative references has come under scrutiny as most diagnostic OAE systems in the market apply adult-normative ranges to their results by default. According to Blankenship et al. (2018), DPOAEs present with significant age-related differences, and these differences arise mainly from developmental changes that occur in the outer ear and middle ear at specific ages. Using adult normative references in paediatric DPOAE analysis is a risk for misdiagnosis. DPOAE amplitudes in children are generally larger and more robust compared to adult patients. HHPs can easily consider the DPOAE measurements of a paediatric patient that fall within an adult range as present, when they are in fact reduced or absent if referenced against paediatric norms. These normative ranges are generally pre-set on the equipment software and selecting the appropriate normative range for different age groups or even entering the correct date of birth is critical for accurate interpretation of test results.



Treasures of Wisdom

The perfect probe fit

- Select the appropriate size ear tip for the ear canal – too large or too small ear tips mayresult in leaking and an unstable fit.
- Insert the ear tip quickly and deeply – a fast and firm approach is best. However, be careful not to unnecessarily disturb a calm or sleeping patient.
- Ensure a snug and deep fit – the ear tip is the first line of defence in attenuating background noise. Also take care not to block the ear tip opening against the ear canal.
- Secure the probe clip or module to the patient's clothing or to a nearby item of clothing (parent/caregiver lapel) to avoid an unstable probe fit in the ear canal and noise resulting from friction.

Auditory Brainstem Response (ABR)

ABR was first introduced to the field of clinical audiology in the 1970s. This discovery has broken boundaries for the diagnosis and early intervention of the youngest infants and most difficult-to-test patients, which would have been impossible otherwise. Also known as Auditory Evoked Potentials (AEP), this measure provides a physiologic method of evaluating the integrity of the eighth cranial nerve (CN VIII) and estimating hearing thresholds electrophysiologically. The success of performing and interpreting ABRs relies heavily on the skill and knowledge of the clinician, particularly regarding the anatomical generators of each of the ABR's components (Hood 1998).

A normal ABR is generated by several anatomical and neural sites (Hood 1998):
Wave I – Distal portion of CN VIII
Wave II – Proximal portion of CN VIII
Wave III – Neurons in the cochlear nucleus
Wave IV – Third-order neurons of the superior olivary complex
Wave V – Lateral lemniscus and inferior colliculus

In this section, both neurological and threshold estimation techniques will be discussed. The interpretation of the ABR is beyond the scope of this chapter. However, information on the interpretation of ABRs can be found in the eHandbook of Auditory Evoked Responses: Principles, Procedures & Protocols by James W. Hall III (2015). Normative references for ABR testing are developmentally specific and HHPs are encouraged to review the normative data of several respected authors in the field of electrophysiology when analysing test results.

Patient state in ABRs

In an ideal world, all young patients are perfectly behaved, calm and quiet for as long as required to complete their electrophysiological tests. However, it is simply not reasonable to expect young patients to sit or lie perfectly still for ten minutes, let alone for the hour needed to conduct extensive ABR testing. Bioelectrical (physiological and myogenic) artifacts are among the most common artifacts HHPs encounter when performing auditory evoked potentials like ABRs (Stegeman and De Weerd 1990). To eliminate the probability of bioelectrical artifacts, the HHP may require the patient to sleep naturally or be sedated when needed.

Natural sleep ABRs

To facilitate an effective natural sleep ABR, it is recommended that the appointment be scheduled around the young patient's best nap time. Sleep deprivation may also help to fatigue the patient enough to fall asleep quickly and remain asleep for an extended period. The HHP should instruct the parent/caregiver to keep their childe up late the night before, wake them up early and keep them awake until they arrive at the test setting. It is also encouraged to perform the test preparations before the patient is allowed to fall asleep. Attempting to prepare the skin and placing electrodes once the patient is already asleep can wake the young patient up.

It is important to plan enough time for preparing the patient, performing the test (which may require the HHP to stop and start several times as the patient progresses through various sleep stages) and providing results, feedback and/or counselling. Natural sleep ABRs may sometimes require several attempts at various appointments to obtain a complete (reliable) test battery, depending on the patient's sleep quality and co-operation on any given day. Children over the age of one and a half years rarely sleep long and peacefully enough to obtain a complete (and reliable) ABR measure in one session, although there are always exceptions to the rule.

Sedated ABRs

If a patient is unable to sleep naturally or co-operate for the duration of an ABR evaluation, then sedation may be required. At this point, it is vital to stress that patient safety must always remain the highest priority, as there will always be risks associated with sedation. The HHP must always weigh the balance between potential sedation risks and the audiological information that needs to be acquired. If the risks outweigh the potential gains, then sedation should under no circumstances be considered an option, and an alternative solution must be explored. Furthermore, an HHP must never attempt a sedated ABR without the assistance and supervision of an anaesthetist, regardless of whether the test is conducted in a controlled setting like a theatre, hospital ward or practice room.

The type of sedation used for ABRs and other auditory evoked potentials may also impact the test reliability and validity. Conscious sedation is an excellent option for this procedure,

as it is a lighter type of sedation, where the patient maintains an open airway independently and can be aroused by physical or verbal stimulation (Hall 2007). Intravenous Propofol (also known as Diprivan) is a sedative drug generally used during conscious sedation procedures that typically produces a very modest delay in ABR inter-wave latencies (Hall 2007). This type of sedation has proven to be effective for patients older than three years of age. Hall (2007) also regards *Chloralhydrate* as a suitable conscious sedation option. Clinically, this option is more suitable for younger children (between the ages of one and two years) with a relatively stable daytime sleeping routine. *Chloralhydrate* is rarely effective when administered to children with body weight exceeding fifteen kilograms or when administered outside of their best nap time. Sleep deprivation increases the effectiveness of *Chloralhydrate*.

Certain sedative drugs are, however, contraindicated for auditory evoked potentials as they affect the latency, amplitude or in some cases even prevent potentials from generating altogether (Hall 2007). It is highly recommended that an experienced anaesthetist be consulted about the most suitable sedative drugs for electrophysiological evaluations. Lastly, test duration should be taken into consideartion with sedation. All sedation carries a level of risk to patients, and the longer they remain under sedation the more risk they are exposed to. A skilled and prepared clinician should be able to complete a comprehensive diagnostic ABR evaluation including neurological (click) ABR, threshold estimation testing (air-conduction and bone-conduction) and/or auditory steady-state response (ASSR) testing in under an hour.

Anatomical sites of electrode placement

Electrode placement and test parameter setup must be optimised to facilitate smooth transitioning between different test sets without unnecessarily disturbing the patient. The position of the electrodes is allocated via the international 10-20 system (Figure 6.2) which is typically used in auditory evoked potential measurement and clinical neurophysiology (Hall 2007). Four electrode sites are utilised for the above-mentioned test battery: Inverting electrodes (placed on the right and left earlobes), non-inverting electrodes (placed on Cz, which is located on the vertex or top middle of the head) and ground electrodes (placed on Fz or high on the forehead). The use of a jumper cable or ‘connector cable allows the tester to seamlessly alternate measurement between the two ears when obtaining responses and enables the tester to record two-channel recordings.

Figure 6.2: (a)

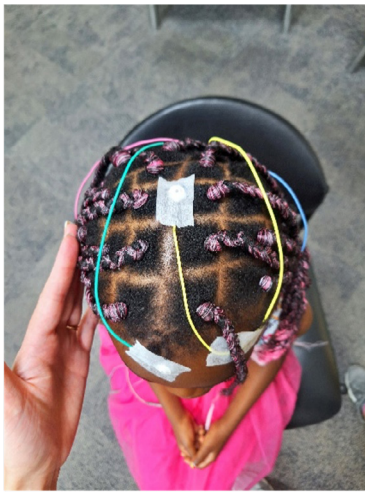


Figure 6.2: (b)

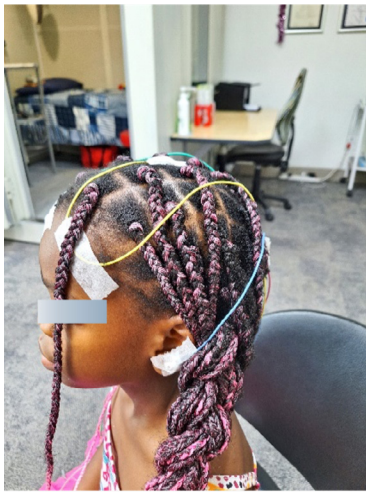


Figure 6.2: (c)



Figure 6.2: (a) Electrode placement and patient positioning non-inverting (yellow) an ground electrode (green) placement, (b) Left inverting electrode (blue) on the left ear-lobe, non-inverting electrode (yellow) and ground electrode (green), which is stabilised on the forehead, (c) Patient positioning in relation to headbox/pre-amplifier and insert earphones

The earlobes are considered the superior option for application of the inverting electrodes as they provide the tester with several distinct advantages, as described by Hall and Swanepoel (2010):

- Reduced post auricular muscle (PAM) artifact – if the electrode is placed directly over the postauricular muscle, it is particularly vulnerable to recording bother-some muscle artifact.
- Increased amplitude of wave I – an increase in amplitude of approximately 30% has been noted with electrode placement on the earlobe compared to the mastoid.
- Reduced stimulus artifact – with the placement of the inverting electrode on the earlobe, the bone conduction oscillator can be placed on the mastoid as normal without running the risk of significant electromagnetic stimulus artifact arising from placing the oscillator directly onto an electrode.

Skin preparation, and electrode and transducer placement

The skin must be carefully prepared for electrode placement. The anatomical sites are cleaned using a specialised granular gel or abrasive tape to remove any excess oils and dead skin cells from the surface where the electrode is placed. Care should be taken not to clean the anatomical sites too abrasively, resulting in serous drainage and/or bleeding, which will impact the impedance measurement. In addition, any excess granules should be removed from the anatomical sites before securing the electrodes, as this may also impact the impedance measurement.

When scooping conduction paste from its container, it is recommended that a small amount of the paste is transferred onto a prepared piece of gauze, using a plastic or wooden tongue depressor. Conduction paste should never be scooped directly from the container using an electrode, as conduction paste is known to be an excellent growth medium for microorganisms. Care should also be taken not to use too much electrode paste as protrusion of the paste increases the recording surface area and impacts the quality of the recording. The electrodes should then be secured to their designated anatomical sites using a small piece of micropore tape. A firm though gentle hand helps to secure the electrode for consistent and repeatable recordings and to prevent intermittent artifacts arising from an unstable electrode during testing. For optimal testing, the impedance should measure between 2 – 5 kΩ across the electrode range.

Earphones should be used where a patient's ear canal is available. If the patient ear canal is not available (as seen with atresia, microtia and/or anotia), then headphones might be considered. However, it should be noted that headphones can cause significant stimulus artefacts at high test intensities.

Test parameters

The test parameters for various auditory evoked potentials are system-specific and test-specific. Decisions regarding the selection of the test parameters should be based on the recommendations of respected authors in the field of auditory evoked potentials, manufacturer recommendations, HPP preferences and the test environment. It is important to highlight that any changes in the test parameters will have an impact on the tracings, analysis and interpretation of the test results. Normative references that have been determined for a

specific test system and environment, for specific auditory evoked potential evaluations, are no longer valid when any of the test setting parameters are altered and indeed may have a detrimental impact on the analysis of the test results.

When approaching an ABR evaluation, the tester should aim to answer two major questions:

1) Is CN VIII conducting sounds normally up to the midbrain level? (Neurological evaluation)
2) What are the estimated (electrophysiological) hearing thresholds? (Threshold estimation)

Neurological ABR evaluation

Once the patient is sleeping peacefully, either naturally or under sedation, and is appropriately prepared and positioned for the test, the HHP is advised to start with the neurological (click) ABR evaluation. This provides the opportunity to predict what is likely to be encountered later in the evaluation, for example, delayed absolute latencies are suggestive of possible conductive dysfunction, and the presence of cochlear microphonics is suggestive of possible Auditory Neuropathy Spectrum Disorder (ANSD) or retrocochlear dysfunction, etc. In this way, the next steps can be planned more efficiently, to obtain as much information in as little test time as possible.

The goal of the Neurological ABR should be to obtain waves I, III, and V. If the waveforms are unclear or poorly repeatable at first, an increase in the intensity may result in evoking these waveforms or responses. The use of a two-channel recording will also make analysis and interpretation simpler and more accurate by helping the HHP to confirm the presence of wave III and V. Changing the polarities (rarefaction, condensating or alternating) is also a useful and necessary tool to confirm the presence and legitimacy of a specific waveform. Simply repeating a single polarity is not enough to confirm the legitimacy of a waveform and some artifacts may repeat themselves quite convincingly if the HHP is not careful in their analysis.

An important rule when analysing Neurological ABRs is that with an increase in intensity, the amplitude of the waveforms should increase and the latency of the waveforms should decrease. Conversely, with the decrease in intensity, the amplitude of the waveforms should decrease and the latency of the waveforms should increase.

When analysing Neurological ABRs, it is critical to be aware of the effects that nerve maturity may have on the appearance of the waveforms. The major expected neuronal maturation occurs within the first two years of life, and interpeak latencies (IPL) decrease significantly during this time (Matin et al. 2021). Therefore, reviewing reputable age-appropriate normative data when analysing interpeak and absolute latencies is essential for an accurate diagnosis.

Threshold estimation ABR

Threshold estimation may follow the neurological evaluation. Tone burst ABR should be the preferred test method to physiologically estimate hearing thresholds, as it provides frequency-specific information and has proven reliable. Where possible, it is preferable to obtain both air- and bone-conduction estimated thresholds, since this provides information about the presence or absence of a conductive component and therefore also insight on the nature of the HL. As conductive pathologies are more prevalent in the paediatric population, a potential middle and/or outer ear influence will have to be considered with any intervention for a young patient.

With threshold estimation, the goal is to obtain a repeatable wave V up to the lowest intensity level upon which the wave V tracing disappears. Threshold estimation techniques using tone burst, CE-Chirp or even audiological click evaluations will require the HHP to use unique correction factors that are specific to their test equipment and test environment. To procure environment and system-specific correction factors, the HHP must acquire normative data for their clinical equipment and setting. This is achieved in the following ways:

- When acquiring a new test system, spend sufficient time customising the test settings and parameters (based on the recommendations of respected authors in the field of ABRs and manufacturer recommendations) for the specific testing needs.
- Perform biological hearing threshold estimations on at least twenty normal hearing individuals, for example, determine the lowest audible hearing level for each test frequency for twenty normal hearing individuals, specifically for each ear, using insert earphones.
- Calculate the average thresholds, at each frequency. The average will be accepted as the correction factor for each frequency. Never use the correction factors of

another HHP's system, as this will result in inaccurate interpretation and an unreliable diagnosis.

Threshold estimation hinges on the same principles as the neurological evaluation when considering the latency and amplitude of wave V. The absolute latency of wave V should increase or, at the very least, remain constant with the incremental decreases in intensity. The latency of wave V will never be earlier at a lower intensity compared to a louder intensity. These simple rules will guide the HHP in avoiding mistakes when analysing their waveforms.

Auditory Steady State Response (ASSR)

The ASSR was first introduced in 2001. Before that, the ABR was the only option for objective estimation of hearing thresholds. According to Hall and Swanepoel (2010), it is important for the HHP to understand that the ASSR does not refer to a single response but rather a series of responses that are generated at different locations along the auditory pathway. These responses are directly related to the stimulus characteristics presented to the ear (Hall and Swanepoel 2010).

One of the advantages of the ASSR is its maximum output levels. The ASSR can often produce stimulus levels at much louder intensities compared to the tone burst or click stimulus. This gives the HHP the advantage of being able to test more severe degrees of HL than would be possible through other ABR test techniques. Although ASSR is a convenient and relatively simple test technique to administer, it is important to be aware of certain limitations:

- ASSR should not be used as a standalone test and must be considered part of a comprehensive test battery.
- The ASSR only provides an estimated behavioural hearing threshold — it cannot distinguish between sensorineural, conductive, or mixed HL.
- The worse the degree of HL, the more accurate the estimated thresholds tend to be with ASSR.
- Minimal, sharply sloping and conductive types of HL tend to deliver more unreliable estimated hearing thresholds with ASSR.
- High levels of internal and external noise during ASSR testing may negatively impact the estimated hearing thresholds.
- Estimated hearing thresholds obtained in cases where ANSD and/or retrocochlear

pathology is suspected must be interpreted with great care as the auditory pathway is compromised. The thresholds may be better or worse than indicated.

The Electrophysiological Toolbox – Where OAEs and ABRs Come Together

If we simply consider the intricacies of normal hearing, then investigating abnormal hearing undoubtedly requires a comprehensive diagnostic toolbox in the hands of a skilled and experienced HHP. ANSD and other retrocochlear hearing conditions often present with strange clinical pictures that require an in-depth understanding of each portion of the auditory pathway. ANSD was first described by A. Starr and his colleagues in 1996 (De Siati et al. 2020). ANSD is characterised by the absence of repeatable ABR waveforms and/or altered waveforms and absent or elevated acoustic reflexes, in the presence of Cochlear Microphonics (CM) and/or OAEs (De Siati et al. 2020). Considering that ANSD accounts for up to 30% of all newborns discharged from neonatal ICU (NICU) who present with sensorineural hearing loss (SNHL) (De Siati, et al. 2020), the need for advanced and reliable electrophysiological testing in the paediatric audiology clinic becomes vital. As the outcomes for intervention in this special test population hinge directly on the correct diagnosis of the HL, the margin for error becomes ever smaller.

Correct diagnosis, and appropriate audiological intervention and counselling will have a powerful impact on the young patient's language acquisition, scholastic performance, and the family's expectations and acceptance of the HL. Neural maturation is also frequently overlooked when it comes to ABR analysis. With improvements in technology, our ability to diagnose younger children earlier has improved dramatically. Nevertheless, the push towards earlier detection may in some cases be harmful, particularly in high-risk paediatric patients (as defined by the American Joint Committee on Infant Hearing), 2019). According to Psarommatis et al. (2017), a significant number of high-risk infants with absent ABR waveforms showed complete or partial recovery at re-examination. They also found that the younger the infant was at the initial ABR evaluation, the higher the probability of a false-positive ABR finding (Psarommatis et al. 2017). The presence of OAEs was also found to be a strong predicting factor for the recovery of ABR waveforms despite the presence of multiple other risk factors (Psarommatis et al. 2017). Therefore, early cochlear implantation (generally before the age of one year) should be considered with great caution, especially for children fitting the ANSD

profile. Cochlear implantation should only be considered after obtaining stable and reliable electrophysiological and behavioural test results (Psarommatis et al. 2017).

Conclusion

Electrophysiological testing requires regular practice and constant learning. These physiologic evaluation tools are routinely utilised in the most vulnerable age groups where a potential error can have a huge impact on the trajectory of a child's life. A hard but necessary truth to acknowledge in your role as an HHP is that there is no room for error when it comes to children. Efficiency helps, but accuracy is everything.

Test your knowledge:

Based on the content covered answer the following questions:

1. Why are factors like timing, clinical environment and equipment setup and functioning considered critical for the success of electrophysiological testing?
2. What are the two major questions the tester wants answered when approaching an ABR evaluation, and why?
3. Why is a comprehensive electrophysiological test battery important when evaluating paediatric patients?

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