

Differential Diagnosis of Auditory Neuropathy/Dys-Synchrony and (Central) Auditory Processing Disorders

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Biography

Thierry Morlet, PhD is the Head of the Auditory Physiology and Psychoacoustics Laboratory at A.I. DuPont Hospital for Children (Wilmington, DE). Dr. Morlet earned his Ph.D. in the neuroscience program at the University of Lyon in France.



Abstract:

The similarities between auditory thresholds, tympanometry and otoacoustic emission responses in some children with auditory neuropathy and children with auditory processing disorders can make differential diagnosis a challenge for these children. A full test battery, including tympanometry, acoustic reflexes, otoacoustic emissions, and auditory brainstem response is required in order to determine an appropriate diagnosis. This article describes auditory neuropathy/dys-synchrony and the test results that differentiate auditory neuropathy/dys-synchrony from auditory processing disorders.

Definition of Auditory Neuropathy/Dys-Synchrony

Auditory Neuropathy/Dys-Synchrony is a term used to describe a condition found in some patients who show normal outer hair cell function (as evidenced by presence of normal otoacoustic emissions (OAEs) and/or cochlear microphonic) and absent or abnormal auditory brainstem responses (ABRs). This condition was first suspected more than 20 years ago and confirmed after OAEs became widely used (Kemp, 1978), because it was discovered that some hearing-impaired patients who were expected to have no outer hair cell function had normal OAEs. The disorder was eventually named auditory neuropathy in 1996 (Starr et al., 1996) and renamed Auditory Neuropathy/Dys-Synchrony (AN/AD) in 2001 (Berlin et al., 2001).

The main characteristics of patients with AN/AD are (Berlin et al., 2002):

- absent middle ear muscle reflexes (MEMRs)
- absent or abnormal ABRs
- present cochlear microphonic (CM)
- present OAEs
- pure tone audiogram varying from normal to severe/profound
- speech reception in quiet varying from excellent to poor
- speech reception in noise generally poor
- absent masking level difference
- no suppression of otoacoustic emissions

Incidence and Etiology of AN/AD

At least 1 in 10 patients with sensorineural hearing loss shows desynchronized ABRs with normal OAEs and/or large cochlear microphonics (Rance et al., 1999; Berlin et al., 2000; Lee et al., 2001; Sininger, 2002). Most cases of AN/AD are bilateral but it is possible to have AN/AD in only one ear.

The etiology of AN/AD is not entirely understood yet. However, the site of lesion appears to be either the inner hair cells, the synapses between the inner hair cells or the auditory nerve. Several risk factors, such as neonatal hyperbilirubinemia, anoxia and infectious disease, are suspected. A genetic factor is assumed in about 40% of patients with AN/AD. Approximately 50% of patients have no defined etiology (Starr et al., 2000). Some patients with AN/AD may have other disorders in addition to AN/AD while others may not. Starr et al. (2000) identified a peripheral neuropathy in about 40% of their patients with AN/AD.

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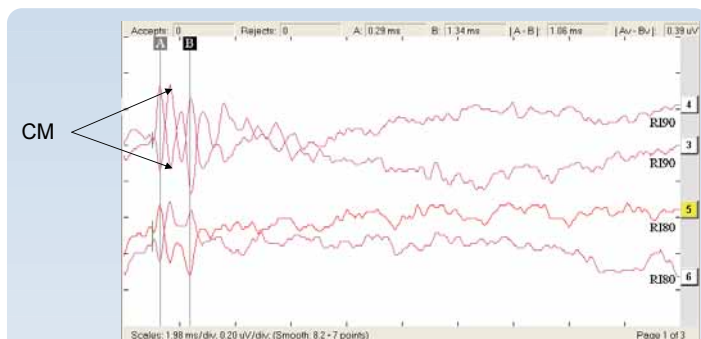


Figure 1: Typical ABR recording in a patient with AN/AD. The use of positive and negative polarity clicks reveals the CM and the absence of neural response. Contrary to neural responses, the CM does not shift in latency when the intensity of the stimulus is decreased.

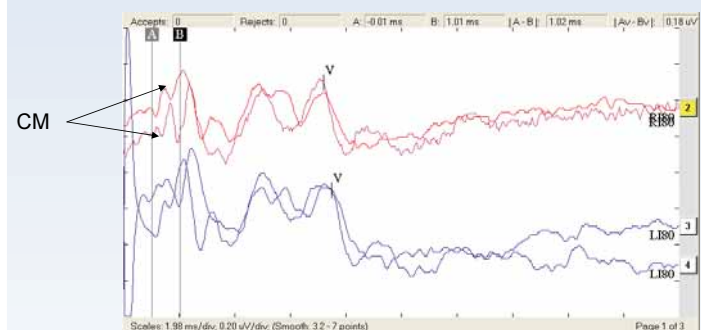


Figure 2: ABR responses in a normal patient. The use of positive and negative polarity clicks reveals the neural response because it doesn't reverse. A short (normal) CM can be noticed at the beginning of the tracing, just before Wave I.

Diagnosis of AN/AD

The diagnosis of AN/AD is done with a battery of tests which consists primarily of OAE, ABR, and middle ear muscle reflex (MEMR) testing. Since AN/AD is characterized by normal outer hair cell function and abnormal function in the region of the inner hair cells and/or auditory nerve, the test battery must be sensitive to both cochlear and auditory nerve function in order to differentiate AN/AD from the more common form of cochlear hearing loss. Most individuals with AN/AD have intact OAEs because the outer hair cells function normally. In some patients, however, OAEs are absent or disappear in time. This makes the diagnosis less obvious. The disappearance of OAEs in some patients with AN/AD is not well understood. In some cases, it is thought that a prolonged use of powerful hearing aids is responsible for the damage to outer hair cells. In other cases, it is hypothesized that the outer hair cells, in addition to the inner hair cells, can be affected by this disorder.

Fortunately, outer hair cell function can also be assessed by identifying the presence of the cochlear microphonic (CM) in ABR tracings. Appropriate identification of the CM will help in evaluating outer hair cell function in patients who have lost or never had OAEs and it will help evaluate outer hair cell function in cases of middle ear effusion when the recording of OAEs is not possible. The ABR of individuals with AN/AD is unlike the ABR seen in other types of hearing problems (see Figures 1 and 2). Because not everyone with

AN/AD has normal OAEs, it is very important that the ABR is done in a specific way that separates the cochlear from the neural response. The way to separate these responses is to use two click polarities for the stimulus: one positive polarity (condensation) click and one negative polarity (rarefaction) click. The cochlear part of the response, or CM, occurs early in the recording and reverses depending on the polarity of the click stimulus, whereas the neural response occurs later and does not reverse regardless of the stimulus polarity. This is the only way that an ABR test result can properly be used to differentiate AN/AD from profound sensorineural deafness.

Additionally, the MEMR responses in patients with AN/AD are typically abnormal or absent because good neural synchrony is necessary to obtain normal MEMR.

Hearing characteristics of patients with AN/AD

Some patients with AN/AD have little to no sound awareness while others appear to have normal hearing with the exception of difficulty hearing in noisy environments. Zeng et al. (1999) showed that in patients with AN/AD, poor speech recognition is due to a severe impairment of the temporal processing abilities. Unlike the more common form of permanent hearing loss that is caused by outer hair cell loss, in cases of AN/AD, the ABR and audiogram results do not provide the clinician with information as to the severity of hearing impairment. As mentioned above, the hearing levels on the audiogram can range from normal hearing to a profound hearing loss (Berlin et al., 2003a). These thresholds indicate the patient's level of sound awareness but do not reflect the severity of the patient's auditory timing problem.

About 7% of children with AN/AD will develop language normally and will start speaking within 1 year to 18 months (Berlin et al., 2003b), despite their abnormal ABR results. Many will show normal hearing in quiet environments but will have a lot of trouble understanding speech in the presence of background noise. These children can often be misdiagnosed as having a (central) auditory processing disorder (APD).

(Central) Auditory Processing Disorders

APDs may be broadly defined as a deficit in the processing of information that is specific to the auditory modality, despite normal pure tone hearing sensitivity (Jerger and Musiek, 2000). APDs may manifest as a deficit in sound localization, discrimination, pattern recognition, temporal processing and poor performance in the presence of competing or degraded acoustic signals (ASHA, 1996). Approximately 5% of school-aged children have some type of APD (Musiek et al., 1990). APDs are often associated with other listening and learning deficits, such as specific language impairment and dyslexia. APDs may also be associated with the presence of neurological conditions in a few cases (such as tumors), delayed maturation of the central auditory pathways, and developmental abnormalities (Bamiou et al., 2001).

Comparison of APD and AN/AD

A hallmark of an APD is the audiometrically normal-hearing child's failure to hear well in the presence of competing speech or background noise (e.g., Bellis, 1996; Chermak et al., 1999; Bamiou et al., 2001; Chermak, 2002; Muchnik et al., 2004). Interestingly, some children with AN/AD show similar hearing behavior as children with an APD, in that there is somewhat appropriate development of speech and language, normal understanding of speech in quiet, but difficulty understanding speech in noisy environments. Therefore, both categories of children will present with a normal pure tone audiogram, normal OAEs and similar speech discrimination scores in quiet and in noise.

Although the two disorders share the above-mentioned characteristics, children with AN/AD will present with absent MEMRs (in rare cases, MEMRs can be present but elevated) and absent or abnormal ABR waveforms, whereas children with an APD have normal MEMR thresholds and normal ABR responses to a click stimulus. It is therefore important to include MEMR testing for all children suspected of having an APD. If MEMRs are absent or elevated, an ABR recorded with both click polarities to rule out AN/AD will then be necessary.

Children with AN/AD who are able to develop speech and language without specific intervention do not represent the majority of subjects with AN/AD. However, numerous newborn screening programs are still OAE-based (this is mostly the case in well-baby units). This means that the majority of children with AN/AD will not be flagged for follow-up by newborn screening. Among them are those who do not have any sound awareness and those who cannot develop speech and language on their own without specific intervention. These children will at least be referred for further testing once it has become apparent to the parents and/or pediatrician that there is a hearing problem. However, children who do not show major delays in the first few years of life will be completely missed. Some of them will later be inappropriately diagnosed with an APD (or other type of learning disability) if a hearing evaluation includes only the pure tone audiogram, word recognition in quiet, tympanometry, and OAE measures. This is why a pre-audiometric triage including tympanometry, MEMRs and OAEs is strongly recommended for all children seen in consultation (Berlin et al., 2003a).

	AN/AD	APD
Tympanogram	normal	normal
MEMR	abnormal or absent	present
OAE	present or absent (over time)	present
ABR	abnormal or absent	normal
Pure-tone thresholds	normal to severe/profound	normal
Word recognition (quiet)	excellent to poor	excellent
Word recognition (noise)	poor	fair to poor

Comparison of test results for AN/AD and APD

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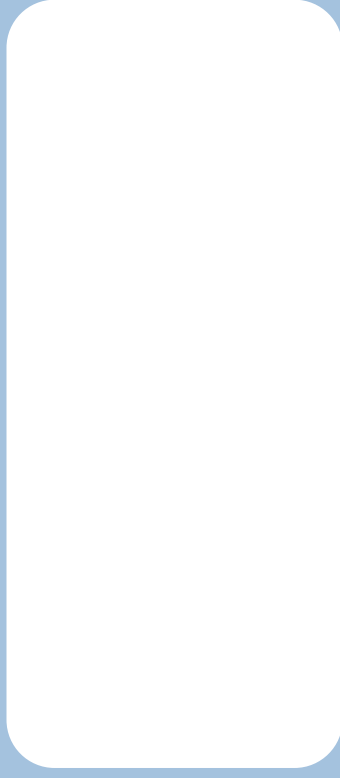
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