International Journal of Pediatric Otorhinolaryngology 79 (2015) 139–145

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Electrocochleography in children with auditory synaptopathy/neuropathy: Diagnostic findings and characteristic parameters

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ARTICLE INFO

Article history:
Received 21 July 2014
Received in revised form 17 November 2014
Accepted 21 November 2014
Available online 3 December 2014

Keywords:
Electrocochleography
Auditory synaptopathy
Auditory neuropathy
Cochlear microphonics
Compound action potential
Summating potential

ABSTRACT

Introduction: The early diagnosis of AS/AN in children remains challenging because it exclusively relies on the detection of OAE and/or CM, while ABR are pathologically changed or missing.

The aim of our study was to ensure the diagnosis of AS/AN, demarcate it to an outer hair cell damage and possibly differentiate between pre- and postsynaptic pathologies.

Methods: We retrospectively evaluated the transtympanic ECochG results of ten children with AS/AN and compared them to a matched group with SNHL and without any signs of AS/AN. We analyzed the thresholds, latencies and – as a new parameter – the amplitude ratio between CAP and SP.

Results: CM and SP thresholds were significantly lower than CAP thresholds in AS/AN patients and significantly lower than SP and CM thresholds in SNHL patients with comparable CAP thresholds. The CAP/SP ratio of amplitudes in SNHL children was more than three times (significantly) higher than in AS/AN children. The cutoff value was set at 1.0 in order to differentiate between both groups with a 80–90% sensitivity and specifity.

It was not possible to differentiate between a pre- and postsynaptic type of AS/AN in our collective.

Summary and conclusion: The ECochG can add valuable information for a precise differential diagnosis of AS/AN, especially in babyhood. We identified the CAP/SP ratio as a new parameter for differentiation between AS/AN and SNHL. When the CAP/SP ratio falls below 1.0, patients can be diagnosed AS/AN with high specificity and sensitivity. Significantly smaller SPL are needed to evoke SP and CM in the AS/AN group, thus showing the preserved hair cell function.

1. Introduction

Auditory synaptopathy/neuropathy (AS/AN) is characterized by absent or pathologically changed auditory brainstem responses (ABRs) in the presence of otoacoustic emissions (OAEs) and/or cochlear microphonics (CMs), which reflect the preserved cochlear hair cell activities. The term "auditory neuropathy" (AN), initially coined by Starr et al. in 1996 [1], was complemented by the term "auditory synaptopathy" (AS), since an isolated dysfunction of ribbon synapses of the inner hair cells may occur [2]. In this paper, the term AS/AN will therefore be used to specify both the synaptical and/or the neural disorders of the underlying pathology.

The pathophysiology of auditory synaptopathy is described as a dysfunction of the inner hair cells and their peripheral synaptic coding of sound. On the other hand, auditory neuropathy comprises the defective generation and propagation of action potentials in the auditory nerve. The pathophysiological correlate is a dyssynchrony in the activation of the auditory ganglion cells [2]. AS/AN is associated with risk factors affecting the auditory pathway that are caused more often by perinatal problems than by genetic disorders [3]. However, in the majority of cases, the underlying pathophysiological mechanisms remain unclear. The prevalence of AS/AN in children with profound hearing impairment is 8–11% and 0.25–094% within the group of children at risk for hearing impairment [4,5].

Affected patients present varying pure-tone thresholds ranging from normal hearing to profound hearing loss and from unilateral to bilateral sensorineural hearing loss (SNHL). Middle ear muscle reflexes are generally absent or thresholds are elevated [6], although some case reports indicate that this may not apply to all

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http://dx.doi.org/10.1016/j.ijporl.2014.11.025
0165-5876/

patients [7]. Especially in ambient noise, the word discrimination is typically disproportional poor compared to the pure-tone audiogram and the response to amplification with hearing aids is often poor [2,8,9].

Thanks to the ABR based universal newborn hearing screening programmes, babies and small children can now be diagnosed with AS/AN at a very early age. However, in this group of patients, an early and precise diagnosis of AS/AN remains challenging, since it exclusively relies on few parameters of the objective audiometry. As OAE may disappear throughout the course of the disease [1,10] and subjective audiometry has low evidence at that age, the diagnosis of AS/AN often relies on the detection of CM and ABR, which sometimes are difficult to assess and interpret. In the cases of severe-to-profound hearing loss and poor response to amplification with hearing aids, side-specific ECochG is used to ensure the diagnosis of AS/AN and verify the indication for a cochlear implantation [2]. In these cases, when the diagnosis is solely based on the detection of CM, more ECochG waveform parameters would be helpful to outline AS/AN to a delay of maturation of the auditory brainstem or to outer hair cell damage, as this may have implications for the outcome of cochlear implantation.

In latest investigations in children with AS/AN and a profound SNHL, McMahon et al. were able to differentiate between auditory synaptopathy and neuropathy (postsynaptic) by specific ECochG waveform patterns, thus identifying the pre- and postsynaptic site of lesions [11]. After cochlear implantation, patients with presynaptic lesion (AS) showed normal EABR morphology, whereas the EABR in patients with postsynaptic lesion (AN) were absent or pathologically changed. These results point to the importance of clinically reliable differentiation between auditory synaptopathy and auditory neuropathy in respect to the outcome of rehabilitation with hearing aids and/or CI.

In this study, we looked for additional diagnostic waveform parameters in ECochG, which may contribute to a reliable diagnosis of AS/AN, outline it to a delay in maturation of the auditory or an SNHL< and provide more information to differentiate between AS and AN.

2. Methods

2.1. Subjects

We included a group of 20 children (40 ears) who showed moderate-to-profound hearing loss in pure tone audiometry. A group of ten subjects ranging in age from ten months to eleven years (median = 38 months) showed a bilateral absence of or severe abnormalities in the ABRs with preserved CMs (Fig. 1) and was therefore diagnosed with AS/AN (group AS/AN). In 5 children of this group ABRs were missing and the other 5 children showed pathologically altered ABRs (mean threshold: 87 dB nHL; minimum: 80 dB nHL; maximum: 95 dB nHL). All 10 children within the AS/AN group presented preserved CMs in the auditory brainstem responses whereas OAE had been detected in only 6 children.

The other ten children aged from six months to nine years (median = 35.6 months) exhibited elevated ABR thresholds (mean threshold: 78.5 dB nHL; minimum: 55 dB nHL; maximum: 90 dB nHL), but clearly detectable ABR waveforms without signs of asynchrony, and were diagnosed with SNHL (group SNHL). OAE were not detectable in any child of the SNHL group.

All audiological studies were recorded at the Department of Otorhinolaryngology, University Hospital of Cologne. All patients showed a poor response to conventional hearing aids and were referred to ECochG to define CAP and frequency-specific CM thresholds in order to evaluate the indication for cochlear implantation.

2.2. AEP recordings

The Nicolet Bravo system (Viasys®, CareFusion, San Diego, California, USA) was used for ABR and ECochG recordings in all subjects. Clicks with a duration of 140 μs separated to condensation and rarefaction were monaurally presented at a repetition rate of 21.3 per second using ER-2A (Etymotic Research, Elk Grove Village, IL, USA) insert earphones at a maximum intensity of 135 dB

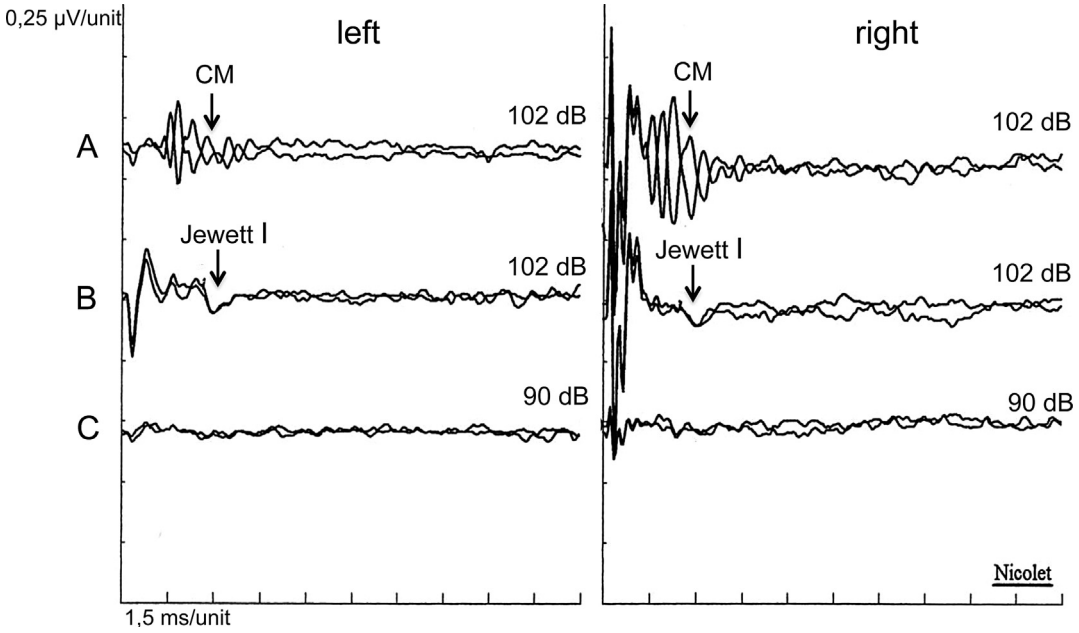

Fig. 1. Click-evoked ABRs in a child with AS/AN (A): superimposed averages to condensation and rarefaction clicks of 102 dB, showing phase-reversed CMs within the first 2–3 ms, which mask the intermixed ABR wave I; (B and C) ABR to alternating clicks; the phase-reversed CMs are cancelled during averaging and a rudimentary ABR wave I is revealed.