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ORIGINAL ARTICLE

Evaluation of cochleo-vestibular functions in patients with auditory neuropathy

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Abstract *Background:* Auditory neuropathy (AN) is a specific hearing disorder with abnormal auditory neural responses in the presence of normal cochlear function. Affection of vestibular portion of cochleo-vestibular (VIII) nerve was only reported in few studies.

Objectives: To evaluate the cochleo-vestibular function in these patients and to detect any relationship between both cochlear and vestibular functions.

Method: The study was conducted on forty (AN) patients. All subjects were submitted to: full audiological test battery; history taking, clinical otological examination, basic audiological evaluation, Auditory Brainstem Response (ABR), Transient Evoked Otoacoustic Emissions (TEOAEs) and threshold equalizing test (TEN). Vestibular test battery; (vestibular evoked myogenic potential (VEMP) and VNG test battery).

Results: Audiological test battery confirmed the audiological criteria for auditory neuropathy reported in the literature. Results of this work showed that, there was preservation of TEOAE in all patients. Also, there was an increase in amplitude of CM in AN. Moreover, all AN patients showed presence of dead regions that started first in low frequencies and with increase number of dead regions the mid and high frequencies also affected. In addition to that the vestibular function tests showed that VEMP was preserved bilaterally in 15 patients, unilateral in 10 patients and absent in 15 patients. On the other side, VNG showed normal central vestibular system with unilateral weakness in 10 ears only.

Conclusion: Patients with auditory neuropathy could also have vestibular neuropathy. Vestibular neuropathy could be classified into three groups: superior vestibular neuropathy, inferior vestibular neuropathy and superior/inferior vestibular neuropathy.

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1. Introduction

Auditory neuropathy is a term used to describe specific hearing disorder with abnormal auditory neural responses in the presence of normal cochlear function. Auditory neuropathy (AN)

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first identified by Starr et al.[1] was then modified to Auditory Neuropathy/Dys-synchrony by Berlin et al.[2] Recently, NHS conference guidelines in 2008, proposed Auditory Neuropathy Spectrum Disorder (ANSD) as a term to describe auditory neuropathy.

The patients commonly have markedly reduced discrimination abilities in the presence of minimal loss of hearing sensitivity, especially in the background noise.[3] While this characteristic pattern of AN points to VIII cranial nerve affection, yet the specific sites and mechanisms of AN have not been exactly determined.

Most of reports in the literature discussed AN in terms of its impact on auditory pathway. For such purpose, many tests are in common use. These are auditory brain stem response (ABR), otoacoustic emissions (OAEs) and cochlear micro phonic (CM) measurements. Such tests denoted that the function of the cochlear nerve was disordered, whereas the cochlear outer hair cells were normal.[1] Also, threshold equalizing noise (TEN) test was developed for extensive evaluation of cochlear function.[4] Such test can give us more information as regards cochlear processing in patients with various forms of hearing loss.

Together with the cochlear nerve affection, it is highly probable that the vestibular nerve is involved as well.[5] Affection of vestibular portion of cochlea-vestibular (VIII) nerve was only reported in few studies. Kaga et al.[6] found absent caloric responses in patients with AN suffering from balance instability. Furthermore, Starr and Fujikawa[7] reported significant vestibular abnormalities in those patients in the form of absent or asymmetrical caloric response. Actually, the ordinary caloric test can only stimulate the lateral semicircular canal with subsequent evaluation of superior division of the vestibular nerve.

Measurements of vestibular evoked myogenic potentials (VEMP) could be utilized for separate evaluation of the inferior division of the vestibular nerve via saccular stimulation.[8] Utilizing this idea, Akdogan et al.[5] reported various forms of VEMP anomalies in children with AN. These studies had not concentrated on cochleo-vestibular correlations in AN patients. Accordingly, the functional integrity of the vestibular system as well as its involvement and its correlations with affection of auditory system in patients with AN need to be further examined.

2. Aims of the work

This work was designed to evaluate the cochleo-vestibular function in these patients and to detect any relationship between both cochlear and vestibular functions.

3. Subjects and method

The study group consisted of (40) patients diagnosed with auditory neuropathy whose ages ranged between 15 and 55 years. Diagnosis was based upon the following criteria[1]:

- Bilateral sensor neural hearing loss.
- Poor word discrimination scores pure tone audiometric thresholds.
- Absent or abnormal ABR results that were not correlated to pure tone audiometric thresholds.
- Preserved OAEs.

All participants (AN patients) in the current study were subjected to the following:

1. Full history taking.
2. Otological examination.
3. Basic audiological evaluation: using: (a) pure-tone audiometry using orbiter 922 (GM Otomtrix, Denmark): This included Air conduction and Bone-conduction. (b) Speech audiometry. (c) Immittancemetry using amplaid 724 (amplifon, Italy): including tympanometry and acoustic reflex threshold measurement.
4. TEOAE (Transient Evoked Otoacoustic Emission) using Otodynamic ILOV6 (Otodynamic LTO., UK). Stimuli were 80us rectangular clicks presented at a peak level of 80–90 dBpSPL. A total of 260 sweeps were averaged using a 500–6000 Hz bandpass. The level of TEOAE was spectrally and automatically determined at different frequency bands.
5. Auditory Brainstem Response (ABR) using intelligent hearing system evoked potential (smart EP, Miami, Florida). The stimuli were 100-usec broadband click with rarefaction polarity delivered at a rate of 19.3 to the subject ipsilaterally through headphone. The response was filtered between 200 and 3000 Hz, amplified 10.000 times, recorded over 10.24 ms time window, and 1024 sweeps were averaged for each run.
6. Cochlear micro phonic (CM) potential was done using intelligent hearing system evoked potential (smart EP, Miami, Florida). Recording using surface electrodes. Essentially, the same ABR testing protocol described before was used to record the CM except that the stimuli were delivered through insert earphone. Separate recording was done using three click polarities, namely, alternating, rarefaction and condensation. The CM recorded at 90 dBHL was considered present if the response reversed its polarity with change of the stimulus polarity.
7. Threshold equalizing noise (TEN HL) test for each ear was done using a compact disc.[4] The disk contains 8 tracks: the first track contains calibration tone, the other 7 tracks contain frequencies which are 500, 750, 1000, 1500, 2000, 3000, 4000 Hz and broad band noise.[4] The test was done using a two channel audiometer, since the levels of noise masker and the tone signal were separately controlled. The masked threshold was measured for each ear at each frequency. A dead region at a specific frequency is indicated by a masked threshold that is at least 10 dB above the absolute threshold and 10 dB above the nominal noise level per Equivalent Rectangular Bandwidth (ERB).[9]
8. Vestibular evoked myogenic potential (VEMP): using Intelligent hearing system evoked potential (smart EP, Miami, Florida).the stimuli were broad band click with rarefaction polarity, 100-usec duration, 125 sweeps with repetition rate 5 click/s. The recorded potentials were filtered through band pass filter 30–1500 Hz, with analysis time 50 ms and at 95 dBnHL intensity.
9. Video-nystagmography (VNG): the system used was Computerized Videonystagmography Ulmar version 0.1 (synapse, America): VNG test battery searching for spontaneous, gaze evoked, positional and positioning nystagmus was performed. Oculomotor test battery included Random Saccade test, Eye tracking and optokinetic tests. Bithermal