Summary

Auditory neuropathy (AN) is a hearing disorder characterized by preservation of outer hair cell function as indicated by the presence of oto- acoustic emissions (OAE) and/or cochlear microphonics (CM), with absent auditory brainstem responses (ABRs). Individuals with AN can have various degrees of hearing loss, with generally disproportionately poor speech understanding. The disorder can affect individuals of all ages **(Starr et al., 1996).**

AN can accompany abnormalities of both pre-synaptic (e.g., IHCs; **Starr et al., 2004**) and postsynaptic (e.g., auditory nerve; **Starr et al., 2003**; **Rodrigues-Ballesteros et al., 2003**) functions. Electrocochleography (ECochG) could be used as a tool to identify both post-synaptic neural and pre-synaptic receptor abnormalities in AN reflecting specific cochlear physiological deficits accounting for disrupted auditory nerve activities. Individuals with disorders of the inner hair cells or their synapses appear especially likely to benefit from cochlear implant, as there are sufficient numbers of auditory nerve fibers and ganglion cells present capable of responding to electrical stimulation **(Santarelli et al., 2006).** Having studies of pre- and post-synaptic responses to sound might better predict success in auditory neuropathy patients. This will provide more accuracy in diagnosis and management of patients with auditory neuropathy.

30 patients with auditory neuropathy were included in this study. They were divided into 2 sub-groups: the first sub-group (15 children: 4-15 years old) and the second sub-group (15 adults: 16-40 years old). Also, this study included 30 subjects with normal threshold and latency of compound action potential served as controls for comparison with the patients with auditory neuropathy. All patients were submitted to complete general and audiological assessment, including ECochG. This study was conducted in Audiology Unit, Ain-Shams University, Cairo, Egypt and Audiology Unit, Fayoum University, Fayoum, Egypt.

ECochG done in all auditory neuropathy patients revealed one of the following forms:

Presence of receptor summating potential (SP) without neural compound action potential (CAP) consistent with pre-synaptic disorder of

inner hair cells and their synapses with auditory nerve terminals, presence of both SP and CAP consistent with post-synaptic disorder of auditory nerve and absence of both SP and CAP consistent with combined pre-synaptic and post-synaptic disorder.

The first two forms were previously described by **Santarelli et al. (2008).** 16.7% of patients in the present study showed absence of both SP and CAP. Correlation between the previous forms and duration of hearing loss in the current study revealed that as the duration of hearing loss was prolonged, the probability for obtaining the third form was increased. It was considered a combined pre-synaptic and post-synaptic disorder.

CAPs were identified in only 43.3% of patients in the present study. A combination of reduced neural input and desynchrony in auditory neuropathy patients would have profound effects on the formation of a detectable compound action potential **(Zeng et al., 1992 & 2006).** The prolonged ECochG potentials found in many of the patients in the present study is consistent with temporal dispersion of neural activities evoked by acoustic stimulation.

The results of our study indicate that:

1-Auditory neuropathy may be clinically presented as early onset and late onset forms with different audiological profile, etiological factors and prognosis.

2- The etiological factors of auditory neuropathy are variable. Hyperbilirubinemia was the most prevalent risk factor in this study.

3- Cochlear microphonic is superior to OAE's in identification of auditory neuropathy, especially in children.

4- Electrocochleography is most essential in identification of the site of lesion in auditory neuropathy.

5- A combined pre-synaptic and post-synaptic disorder occurs as the duration of hearing loss prolongs.

6- The post-synaptic type of auditory neuropathy is associated with mild and moderate degrees of hearing loss. On the other hand, the pre-synaptic type of auditory neuropathy is commonly associated with severe degrees of hearing loss