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Editorial

Auditory Neuropathy Spectrum Disorder: Clinical and Therapeutic Challenges

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Editorial

The auditory neuropathy (AN) or the auditory neuropathy spectrum disorder is a disease that is not yet well defined and do not have an accurate diagnosis [1,2].

It is believed to be a sensory disorder of the inner ear on its interface with the brain stem and/or in the auditory cortex. Several groups studying this issue still disagree with the diagnoses parameters as well as with the treatment and it is still a challenge for physicians to diagnosis AN [1,3,4].

The diagnosis accepted by the majority, is based in the analysis of complementary tests, auditory evoked potentials that connote an activity cochlear present and absence or severe abnormalities of neural function. Classically it is observed the presence of otoacoustic emissions and the lack of response in auditory brainstem response ABR, which would be a "paradox" [1,4,5].

However there are many situations and circumstances in which the diagnosis can be very difficult, such as in cases of patients that have a deafness and AN, in which the presence of emission otoacoustic would not be identified. In these cases we have to look for the research of cochlear microphonic [1,5,6].

It is also known that the above diagnostic tools are often insufficient to diagnose and a large investigation may be needed to diagnose, such as the genetic evaluation. The genetic evaluation still is a very difficult to access, because it has shown great variability and no consensus on what mutations are related to this disorder, beyond the classical mutations already described [1].

The common diagnostic parameters are cases in which the otoacoustic emissions are present with absent or abnormal ABR which is the typical case and unquestionable AN. When the otoacoustic emissions are absent and there is an suspicion of AN the cochlear microphonic is used to support the diagnosis [3,6].

Some more atypical and challenging cases would be those where the subjects have any clinical suspicious of AN and audiological evaluation finds pure tone threshold present (sometimes close to the normal references) with the absence of ABR, the otoacoustic emissions and cochlear microphonic which is classically not considered as AN. The clinical suspicious in these cases would be through to phenotype speech, behavior and development, personal, pregnancy and perinatal antecedents [2,3,6,7].

An immense diagnostic difficulty can be noted, which turns very hard the indication of a treatment, because the uncertainty of the diagnosis, and that is one reason why the treatment becomes even more complex [3,6,7].

Basically, the treatment consists in speech and language therapy and auditory training, developing the speech and understanding skills, and it is supported by conventional hearing aids and even by the cochlear implant when necessary [6,7,8].

The classical criteria to support cochlear implant as a treatment normally does not include AN as an indication, even in cases where there is AN with tone thresholds compatible with mild to moderate hearing loss, which is somewhat questionable [2,6,8].

After many uncertainties, many groups, mostly from US and Europe, were cutting edge and started submitting their patients with AN, that did not improve with "medical therapy" (therapy and/or appliances hearing), to cochlear implants [2,6,8].

This resulted in a change of concepts and paradigms and brought further discussions between the professionals involved. The discussions are still more complex, since most patients are the pediatric age group, and of course pre-lingual (without spoken language) [1,2].

The indication of cochlear implants for AN patients is discussed because it does not allow simple and objective assessments of the indication of cochlear implant as used in deaf patients with previous knowledge of spoken language (post- lingual) as is the case of speech perception test. This test is of fundamental importance indication and follow-up of patients with cochlear implant use is very limited pre-lingual patients [1,2,6,8].

Then used subjective rating scales that focus on earnings speech and hearing impairments. Most of these scales are subjective and its application often depends on the support of parents and involvement of the family, since most of the patients with AN are children and pre-lingual [6,8].

In conclusion, the medical literature needs more studies from all over the world to help professional understand better this situation and be able to get more support for these patients.

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