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EDITORIAL

Auditory neuropathy and auditory neuropathy spectrum disorders

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Auditory Neuropathy Spectrum Disorders (ANSD) are new classification which was proposed in 2008 by Colorado Children's Hospital Group and defined as normal otoacoustic emissions and absent ABRs in newborn. In our long term follow up study, hearing of ANSD are changed into three types. Type I is normal OAE and normal ABR (normal hearing), Type II is absent OAE and absent ABR (profound sensoryneural hearing loss), Type III is normal OAE and absent ABR (true auditory neuropathy). However, still complications of vestibular problems in ANSD are not known so far.

Historically, in the same year of 1996, a new type of bilateral hearing disorder was reported by Kaga et al. and Starr et al. in different journals. Auditory nerve disease paper was published in the *Scandinavian Audiology* by Dr. Kaga and Auditory Neuropathy paper was published in *Brain*. At present, these different terms are considered to be identical in pathophygiology.

The auditory nerve disease or auditory neuropathy is a disorder characterized by mild-to-moderate pure-tone hearing loss, poor speech discrimination, absent ABR but normal cochlear outer hair cell function revealed by normal OAE and –SP of Electrocochleography.

A variety of processes and etiologies are thought to be involved in the pathophygiology. Most of the reports in the literature discuss auditory profiles and gene mutation of *OTOF* or *OPA1* of patients only but not pathophygiology.

Auditory nerve components consist of cochlear nerve, superior vestibular nerve and inferior vestibular nerve. My question is which nerve of these auditory nerve components is involved in AN? We reported our results of auditory and

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vestibular system assessment of our adult patients of auditory nerve disease. Our study revealed: the age of onset is common during the period of teenage or later. Half of patients had different neurological episodes such as cerebellar infarction, blindness, spino cerebellar ataxia and virus cerebellitis. All of pure-tone audiograms show a low-frequency loss with rising slope pattern, the severity of which ranged from mild-to-moderate degree. All of speech audiometry shows that the maximum speech discriminations in all patients are below 50% except one patient.

The auditory evoked response revealed common results of normal DPOAE, normal summating potentials of Electrocochleography and absence of ABRs.

Meanwhile, caloric test and damped rotation chair test can examine functions of lateral semicircular canals, superior vestibular nerve and oculomotor system in brainstem. On the other hand, Vestibular Evoked Myogenic Potentials (VEMP) is a new face of vestibular function test for otolith organs inferior vestibular nerve.

I show three cases with different results of vestibular examination, Case 1 shows loss of caloric reaction and absence of VEMP. Then both superior and inferior vestibular nerves must be involved. Case 2 shows normal caloric reaction and normal VEMP. Then both superior and inferior vestibular nerves must be normal in left side. Case 3 shows normal caloric reaction and damped rotation chair test. However, VEMP is lost. Then, in this case, superior vestibular nerve is intact but the function of inferior vestibular nerve must be damaged.

I functionally classified vestibular test results into three types. Type 1 is both caloric and VEMP are normal. Type 2 is caloric test is normal but VEMP is abnormal. Type 3 is both caloric and VEMP are abnormal.

However, auditory and vestibular system of ANSD should be more intensely studied because of unknown pathophysiology in developmental age.