Bengal Journal of
Otolaryngology and Head Neck Surgery

Official Publication of The Association of Otolaryngologists of India, West Bengal

Volume 30 No. 3 - December, 2022

This journal is indexed in DOAJ, Index Copernicus, Google Scholar, OCLC WorldCat, WHO Hinari, OAIster, BASE, OAJI and EZB. It is also registered in the OAI database of conforming repositories. The articles are allotted DOI numbers and preserved with the LOCKSS program.

p-ISSN: 2395-2393
e-ISSN: 2395-2407
RNI No : 62551/95

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contd.

Bengal Journal of Otolaryngology and Head Neck Surgery
(Incorporating and directly descended from State Journal of Otolaryngology and Otolaryngology, Calcutta)

Published by
The Association of Otolaryngologists of India, West Bengal
CMC House, 91B Chittaranjan Avenue,
Kolkata - 700073,
West Bengal,
India.

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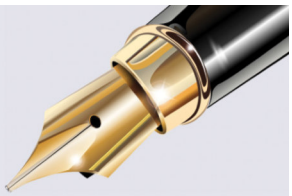

Auditory Neuropathy Spectrum Disorder (ANSD) is a group of hearing disorder in which the ear detects sound normally, but can't send it to the brain. People with ANSD may have normal hearing or hearing loss ranging from mild to severe, but always have difficulty with speech comprehension, more so with noisy environment and fast speech. The condition was originally termed as Auditory Neuropathy (AN). In 2008 the condition was renamed as Auditory Neuropathy Spectrum Disorder (ANSD). With better understanding of the disease, ANSD is diagnosed more frequently and now accounts for about 10% to 15% of cases of hearing loss. There are various etiological factors behind this disorder. These include familial, genetic mutations, premature birth, perinatal hypoxia, jaundice, low birth weight, dietary thiamine deficiency, some drugs, etc. Adults may also develop ANSD along with age-related hearing loss. Some people with neurological disorders like Charcot-Marie-Tooth syndrome and Friedreich's ataxia may also suffer from ANSD. ANSD is characterized by impaired or absent auditory brainstem responses with preserved otoacoustic emissions and/or cochlear microphonics. Some infants improve and start to hear and speak within a year or two. Other infants stay the same, while some get worse. Hearing aids and personal listening devices such as frequency modulation systems are helpful for some children and adults with ANSD. Previously it was believed that cochlear implants would not give any benefit in patients with ANSD. Currently, there are many reports of benefits of cochlear implants in children with severe or profound hearing loss with ANSD. It is interesting to know how the implant gives such successful results in patients suffering from ANSD. One of the reasons may be related to site of lesion. In most of the cases pathology involves either the inner hair cells or the synapse between the inner hair cells and the VIIIth cranial nerve rather than the nerve itself, particularly among young children. Even where the auditory nerve is the site of lesion, there is still an explanation regarding better outcome with cochlear implant. In case of compromised auditory nerve not able to adequately transmit a signal delivered acoustically, the discrete electrical pulses from the cochlear implant may increase or restore synchronous firing activity in the nerve. No tests are currently available, to determine whether an individual with ANSD

will benefit from cochlear implant or not. Some researchers have suggested that outcomes of cochlear implantation can be assessed on the waveform of transtympanic electrically evoked auditory brainstem responses. Robust responses suggest better performance.

With increasing awareness for early detection and treatment of neonatal hearing loss, Otorhinolaryngologists should keep their eyes open for diagnosis of ANSD. Children with progressive difficulty in speech and learning should also be screened for ANSD. More researches are needed for understanding variable course and effective treatment, predicting the outcome of treatment in ANSD particularly fruitfulness of cochlear implant.

Dr Chiranjib Das
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