

Case Report

The impact of high gain conventional hearing aid on OAEs in a case of auditory neuropathy/ dys-synchrony

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Abstract. Aim of the study is to focus on the impact of high gain hearing aid on outer hair cells' functioning as reflected by otoacoustic emission (OAE) changes. A child diagnosed as bilateral moderate -moderately severe sensori neural hearing loss with primary auditory neuropathy / dys-synchrony (AN/ AD) was referred to AYJNIHH (ERC) after being fitted with high gain hearing aid. She had used the hearing aid for less than a year. Audiological evaluation at AYJNIHH (ERC) was indicative of moderate to moderately severe hearing loss with absence of distortion product otoacoustic emissions (DPOAE). She was recommended to discontinue the use of the hearing aid for a couple of weeks. Auditory brainstem response (ABR) and OAE was administered after 14 days. No identifiable peak V was obtained in both the ears however the case had emissions on DPOAE. She was fitted a mild class hearing aid and included for speech language therapy. Conservative trials with amplification devices, diligent monitoring of amplification output levels and OAEs along with possibilities of a cochlear implant was to be explored during therapy. The primary reason behind deterioration in DPOAE may be temporary physiological changes resulting in temporary threshold shift. Therefore, high quality, low gain, wide dynamic range compression hearing aids may be the probable cause of return back of OAEs following two weeks without amplification. Amplification may contribute to loss of OAEs and the risk from use of amplification must be weighed against the benefits of amplification provided to the client.

Key words: Otoacoustic emissions, auditory neuropathy, outer hair cells', ABR, temporary threshold shift.

1. Introduction

In the early 1980s, Davis and Hirsh (1), Worthington and Peters (2), and Lenhardt (3) were among the first to publish case accounts of people with absent ABR with normal and near normal hearing thresholds. In these 20 years, this constellation of results has been given a variety of names, including paradoxical, brainstem auditory processing syndrome, central auditory dysfunction, and neural synchrony disorder. Most recently researchers and clinicians have proffered and accepted the term auditory neuropathy, although whether the condition is, in fact, a neuropathy is not yet established (4).

Auditory neuropathy/ dys-synchrony (a "sub set" of sensory neural disorders) is a broader term which describes a range of disorders affecting afferent neural activity in the peripheral and central auditory pathways (5, 6). The term 'dys-synchrony' signifies the concept of inability to temporally synchronize the neural activity (6) resulting in restriction in perceptual limit for particular auditory parameters, for example, temporal resolution and speech understanding (7, 8). By definition, patients with this disorder have normal otoacoustic emissions (OAEs) and cochlear microphonic (CM), but an absence of or severely abnormal ABR (4). Early estimates of AN/AD ranged from 0.5 to 1.3% of the clinical population and 12 to 14% of those who would otherwise have been thought to have a severe- to-profound cochlear hearing loss (4). Other clinical characteristics appear to vary. Tone thresholds can range from normal or near-normal sensitivity to severe impairment. Impaired auditory processing skills typically are reported, especially in noisy environments. Acoustic reflexes are

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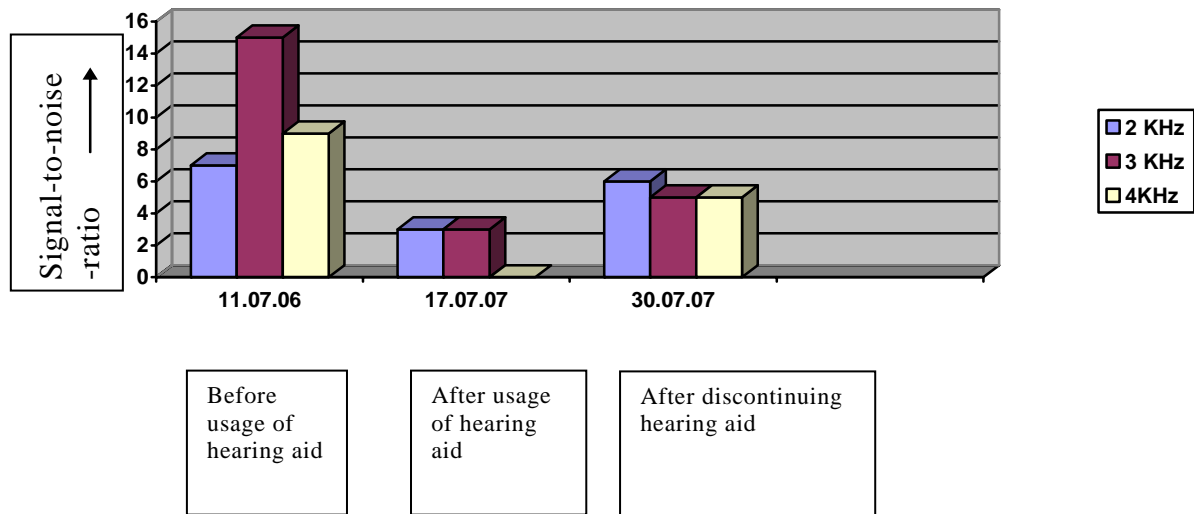


Fig. 1. Shows the changes in signal-to-noise-ratios in distortion product otoacoustic emission in right ear.

Table 1. Shows the changes in signal to noise ratios in distortion product otoacoustic emission in right ear

Frequency	Signal-to-noise ratio on 11.07.06	Signal-to-noise ratio on 17.07.07	Signal-to-noise ratio on 30.07.07
2KHz	7 (pass)	3 (refer)	6 (pass)
3KHz	15 (pass)	3 (refer)	5 (pass)
4KHz	9 (pass)	0 (refer)	5 (pass)

absent. Some cases are transient or intermittent; others change little over time and may even worsen. Auditory dys-synchrony can occur in the absence of any apparent medical problem; with a variety of other symptoms and conditions, or it can be associated with conditions, such as infectious processes (e.g. mumps), immune disorders, and various genetic and syndromal conditions (9).

It also has been shown to occur with diffuse neonatal insults such as anoxia, hyperbilirubinemia, and acidosis (6), as well as transiently with fever (10). Management of children with AN/AD has been a controversial issue. Some children benefit with conventional hearing aids while others with severe impairments in detection of speech sounds or severe temporal processing problems, have fallen within candidature for cochlear implants (11).

Off late studies indicates careful selection of amplification as over amplification may damage outer hair cells function (12). Hence there is need to document case study which would help in

understanding the various aspects to be considered before prescribing amplification device.

In this report, we aimed to focus on the audiological profile of a child with auditory neuropathy and the impact of conventional hearing aid on outer hair cells' functioning as reflected by otoacoustic emissions.

2. Methods

A child aged 2 years, female with chief complaint of poor hearing acuity and limited vocabulary was studied. Her problem was noticed at the age of 1 year by the parents. The case is the first and single issue of her parents. Medical history reported that mother suffered from malnutrition during pregnancy and the child suffered from parathyroid at the age of 2 years. There were history of delayed language development and delayed development of motor milestones. The case went to private audiology set-up dated on 10.07.06 where an audiological

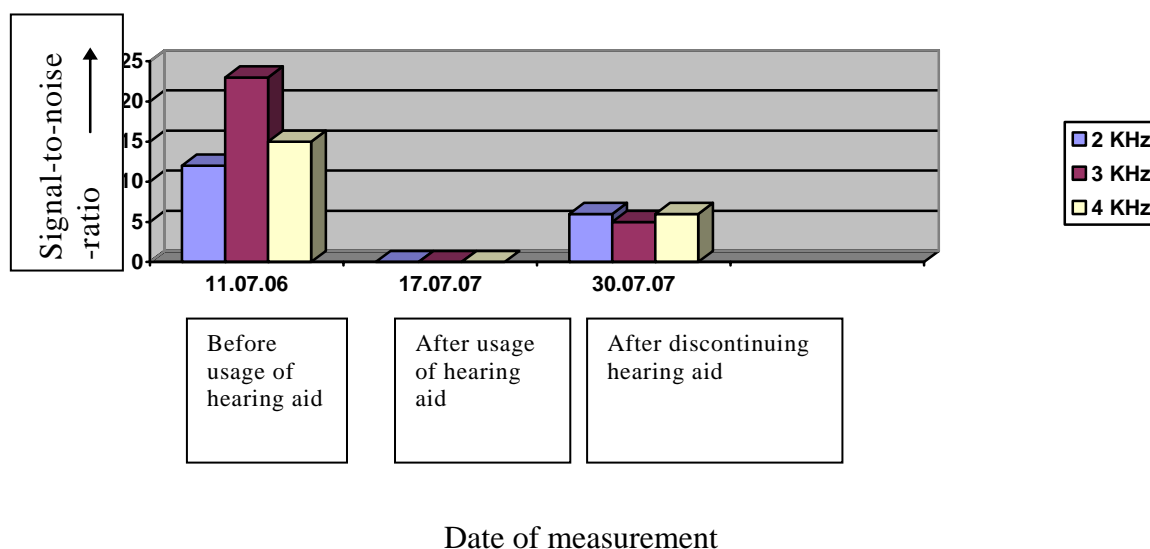


Fig. 2. Shows the changes in signal to noise ratios in distortion product otoacoustic emission in left ear.

Table 2. Shows the changes in signal to noise ratios in distortion product otoacoustic emission in left ear

Frequency	Signal-to-noise ratio on 11.07.06	Signal-to-noise ratio on 17.07.07	Signal-to-noise ratio on 30.07.07
2KHz	12 (pass)	0 (refer)	6 (pass)
3KHz	23 (pass)	0 (refer)	5 (pass)
4KHz	15 (pass)	0 (refer)	6 (pass)

test battery was administered. Behavioural observation audiometry revealed bilateral moderately severe hearing loss. on auditory

brainstem response audiometry using RMS BERA Mark-II no identifiable peak V was obtained at 110 dB n HL in both the ears. The case 'passed' bilaterally on distortion product otoacoustic emissions (DPOAE). DPOAE was obtained using Oto Read Database software version 7.70.1. The pass criteria for signal to noise ratio is 5 dB or more than 5 dB. Neurologist recommended the case for magnetic resonance imaging (MRI) and CT scan. According to the report of neurologist, there was a bilateral moderate to severe delay in central auditory pathway conduction and the case was diagnosed as auditory desynchrony. She was recommended for hearing aid trial, auditory training and speech language therapy. After completion of hearing aid trial the case was fitted

with a hearing aid pseudo bilaterally which provides the full on gain of 65dB SPL and corresponds to a strong class hearing aid. . The aid was used for less than one (exact duration could not be specified by mother) year.

This case was referred to AYJNIHH, ERC for the purpose of speech and language therapy. Reevaluation at AYJNIHH (ERC) on behavioural observation audiometry revealed moderately severe to severe hearing loss with bilateral 'A' type tympanogram and absence of acoustic reflexes in both the ears. Auditory steady state responses indicate thresholds of 84dB HLcg, 104 dB HLcg, 87dB HLcg and 81dB HLcg for .5, 1, 2 and 4 KHz respectively in the left ear and 84dB HLcg, 104dB HLcg, 97dB HLcg and 81 dBHLcg for .5, 1, 2, 4 KHz respectively in the right ear.

No abnormality was detected in ear, nose and throat evaluation. Psychological evaluation revealed average developmental progress. The

case who had “Passed” on DPOAE previously was “Refer” in DPOAE bilaterally on using MAICO ERO Scan on 17.07.07. Auditory brainstem response was administered on the same day. No identifiable peak V obtained at 96 dB n HL in both the ears using tubular insert phone on Nicolet compass Meridian. The case was diagnosed as having moderately severe to severe hearing loss and recommended to discontinue the use of hearing aid for two weeks following which reevaluations were to be done.

Auditory brainstem response and otoacoustic emission were administered after 13 days (on 30.07.07). No identifiable peak V was obtained at 96 dB n HL in both the ears, behavioural observation audiometry revealed moderately severe to severe hearing loss with ‘A’ type tympanogram in both the ears. Acoustic reflexes were absent in all frequencies ipsilaterally and contralaterally and the case was passed in DPOAE using the same instrument (MAICO ERO Scan) with a signal to noise ratio of 5dB and above (Fig 1,2 and Table 1, 2). No changes were noticed on the ASSR thresholds.

3. Discussion

Many audiologists have the impression that hearing aids can not help, hearing aid may hinder perception, and that higher intensities of sound may damage an apparently intact cochlea in a case with AN/AD. In the present study, distortion product otoacoustic emissions were absent after one year of hearing aid use returned back following a couple of week without amplification (table: 1 and table: 2). The hearing aid issue is not straight forward because many patients have elevated thresholds that might be improved by amplification (4). OAEs may have been weak or absent following prolonged hearing aid use, but the OAES return and became clear and robust following a week without amplification (4). It has been shown that OAEs may deteriorate in some children with auditory neuropathy (13, 14) without any change in pure-tone sensitivity. If inappropriately monitored there may be considerable risk that children with auditory neuropathy have inadequate amplification for their hearing needs. Hood (15) recommended: ‘high quality, low gain, wide dynamic range compression hearing aids’. The reason behind this methodology appears to be the sparing of outer hair cells by preventing temporary or permanent threshold shift owing to noise exposure from hearing aids. This may be the probable cause of return back of OAEs following a week without amplification (4). This approach

is intended to minimize any deleterious effects of amplification on otoacoustic emissions until the importance of maintaining otoacoustic emissions in these patients is better understood. If hearing aids are tried, frequent monitoring of otoacoustic emissions for either temporary or permanent effects on OAEs should be a part of the management programs. The prevailing treatment seems to be conservative trials with amplification devices, with diligent monitoring of amplification output levels and otoacoustic emissions along with utmost consideration of patient and parent feedback regarding benefits (4). It is not clear, however, that the outer hair cells that generate the OAEs or CMs provide any functional benefit in terms of sensitivity or frequency discrimination in those who have auditory neuropathy (16). At a functional level, the excessive masking contributes directly to the extreme difficulty of understanding speech in noise because the perceptual signal to noise ratio would be much lower than the physical signal to noise ratio in participants with auditory neuropathy (17). Innovative hearing aid that can convert conversational speech into clear speech can be beneficial (17). At preset, cochlear implant appears to be the treatment of choice for participants with auditory neuropathy (18). If the site of lesion is in the inner hair cells or the synaptic transmission, then the cochlear implant ought to be an effective treatment, as it bypasses both inner hair cell and the synapse to directly stimulate the auditory nerve fibers with electric currents (17). If the site of lesion is related to nerve damage, then the cochlear implant can still be effective because electric stimulation provides much more synchronized neural firing than acoustic stimulation, thus possibly overcoming at least partially if not fully, the neural desynchrony problem (19). If auditory neuropathy involves extensive loss of neurons, then the cochlear implant would be less effective (20); FM systems or other assistive technology may be appropriate. FM system can improve signal to noise ratio by 15 dB (21). In addition, methods of communicating and teaching can effect signal acquisition in a top-down manner. Thus, instrumental modifications often are indicated (21).

4. Conclusion

This study is clearly warranted that amplification may contribute to loss of OAEs and the risk from use of amplification must be weighted against the benefits (if any) of amplification provided to the client (16). The

alternative modes other than conventional hearing aid should also be taken into consideration like changing the communication environment to improve access to auditory signals, FM systems to improve signal to noise ratio, assistive technology, cochlear implant etc. Central resources training to improve higher-order meta-linguistic, meta-cognitive and related skills to compensate for the auditory deficit should be opted. Remediation of the specific auditory deficits through targeted auditory training activities, which may include computer based programs or other specific stimulation activities also should be taken into consideration. The children whose speech perception is most disabled by the auditory neuropathy can not cope with audition based education systems. These children should move to visual based programs like sign language where they could succeed more readily.

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