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A Study on Diagnosis of Auditory Neuropathy Spectrum Disorder in Adults and Comparison of Its Management with Hearing Aids Versus Cochlear Implantation

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ABSTRACT

This study was done at two super specialty hospitals with the purpose of identifying the best diagnostic methods and management options for Auditory Neuropathy Spectrum Disorders (ANSD) in adults. In the management part, we compared the benefits of hearing aids versus cochlear implantation in adult subjects with ANSD. This study was conducted between August 2014 and July 2020. Out of a large number of patients who came to the department, 30 patients who were willing for the study and fulfilled the criteria of ANSD were taken as subjects. Average age of the subjects was 29 years. Specific risk factors which are common in infantile ANSDs were not seen in adult subjects. Pure tone audiometry showed sensory neural hearing loss varying from mild to profound in severity and varying configurations. The word discrimination scores varied from 0% to 70%. Ipsilateral and contralateral Acoustic reflexes were absent in both ears. Brain stem evoked responses showed absent/abnormal peaks at maximum stimulus presentation levels. Cochlear microphonics was present in 80% of individuals and DPOAEs were present in 70% of individuals. Only 30% showed benefit after hearing aid use. 4 individuals underwent cochlear implantation and they showed improvement in hearing threshold, speech discrimination, and reception skills. Thus benefits of hearing aid are limited in late onset ANSDs but they show considerable benefit with cochlear implantation. Audiological investigations such as PTA, Speech audiometry, Immittance Audiometry, OAE & ABR plays the key role in diagnosis of ANSD. A large sample size study and long term follow up is needed to assess the exact outcome of various management options in adults with ANSD.

Keywords:
ANSD - Auditory Neuropathy Spectrum Disorder,
DPOAE – Distortion Product Otoacoustic emissions
OAE – Oto Acoustic Emissions.
ABR - Auditory brainstem response,
PTA- Pure Tone Audiometry
cochlear microphonics, hearing aids, speech discrimination, cochlear implantation
SNHL – Sensory Neural Hearing Loss
ANSI – American National Standards Institute

INTRODUCTION

The term "Auditory Neuropathy" and "Auditory Dysynchrony" is a form of hearing impairment in which cochlear amplification function is normal but nerve conduction in the auditory pathway is

disordered(1, 2). It is a clinical condition characterized by sensorineural hearing loss with poor speech discrimination (SNHL), auditory brainstem response (ABR) is absent but otoacoustic emissions (OAE) are present. Several investigators have described the various aspects of infantile Auditory Neuropathy Spectrum Disorders (ANSD) including the diagnostic and management options but very limited studies are conducted in adults (3,4,5). In the present study we have tried to explore the assessment and management options for adults with ANSDs.

AIMS AND OBJECTIVES

In the current scenario only few studies are reported to discuss the diagnostic and rehabilitation aspects of ANSDs in adults. There are no definite criteria for diagnosing ANSD in adults and controversies exist in the management aspect of this disorder. The present study aims to find the best diagnostic criteria and also management option in adults with ANSD by comparing the benefits of hearing aids versus cochlear implantation.

MATERIALS & METHODS

This study was conducted at two Super specialty hospitals in north Kerala, India. The 30 subjects participated in this study were selected from a large number of Individuals who came to the departments with complaint of reduced hearing from August 2014 to July 2020. All the 30 subjects fulfilled the criteria for ANSD and were willing to participate in this study. Children below 3 years of age, subjects with conductive or mixed hearing loss and subjects with deficient speech were excluded in this study. All Audiological investigations were carried out from a sound proof audiology room with ANSI standards. Calibrated two channel GSI Audiostar pro & GSI 61digital diagnostic audiometer was used for Pure tone Audiometry, Speech audiometry and special tests (tone decay test). Intelligent Hearing System (IHS) was used to record DPOAEs, Cochlear microphonics and ABR. Calibrated GSI Tympstar and Inventis Flute Immittance Audiometer was used to conduct Immittance Audiometry.

Procedure

The study was divided into two phases

I)Assessment Phase

Assessment begins with detailed case history followed by Audiological investigations such as Pure tone Audiometry, Speech audiometry, Special test such as Supra Threshold Adaptation Test (STAT), immittance audiometry, OAEs, Cochlear microphonics and ABR.

In history part, we collected information about pre, peri and postnatal history, speech discrimination difficulties in quiet and noisy situation, music perception and telephonic conversation difficulties. In pure tone audiometry, the degree and type of hearing loss and also the audiometric pattern was assessed. After pure tone audiometry, the individuals identified with pure SNHL underwent further investigation with speech audiometry. Speech discrimination and speech reception thresholds were measured in speech audiometry. Individuals with poor speech discrimination and poor speech reception threshold were further evaluated with ABR and DPOAEs. The final study group was made with those who had SNHL, poor speech discrimination, abnormal ABR, cochlear microphonics present and robust DPOAE. Majority of the individuals had history of difficulty in understanding speech (50%), sound localization (20%), and decreased hearing in noisy environment (20%). Some individuals (10%) had problem with discriminating male and female voices.

Pure tone audiometry findings : Audiograms with increased thresholds at low frequencies is a usual finding in both adults and children with auditory neuropathy(6). Pure tone audiometry showed sensory neural hearing loss varying from mild to profound in severity. Among these 70% of the individuals showed more

loss in low & mid frequencies with better responses in higher frequencies and remaining 25% showed a flat audiometric configuration. In 50% of ears the threshold was less than 50dB, and remaining 50% showed a threshold greater than 50dB. 80% individuals showed fluctuation in hearing loss while evaluating in regular intervals and remaining 20% showed a constant pattern. In 40% ears audiometric pattern was symmetrical and remaining 60% ears showed asymmetrical one.

Speech audiometry: Speech reception threshold and discrimination scores were not correlating with pure tone threshold and 40% individuals showed difficulty in tolerating loud sounds. The word discrimination scores varied from 0% to 70%. 40% of individuals showed scores in between 50% to 70% and in 30% individuals, the scores were between 10-50%. The remaining 30% showed less than 10% scores.

Supra Threshold Adaptation Test: 40% individuals showed positive tone decay and in remaining 60% tone decay was absent. As the degree of hearing loss increased, most of the individuals felt difficulty in tolerating loud sounds at higher sensational levels.

Immittance audiometry: All individuals had normal tympanograms bilaterally, with both ipsilateral and contralateral reflexes being absent in all frequencies. Abnormal or absent middle ear reflexes are a common finding in both children and adults with ANSD(6).

Cochlear microphonics: The cochlear microphonics is an alternating potential generated by the polarization and depolarization of the cochlear hair cells. The cochlear microphonics reflects the integrity of outer hair cells and thus plays a significant role in the identification of ANSDs (6). In our study cochlear microphonics were present in 80% of individuals out of which 70% of individuals showed long ringing cochlear microphonics which was measured by altering the phase of the signal. In 30% individuals cochlear microphonics were absent.

Brain stem evoked responses audiometry: ABR peaks were bilaterally absent in 70% individuals at maximum stimulus presentation levels (99dBNHL) and 30% individuals showed poor waveform morphology and replicability. This is in agreement with studies reported in literature (1,7,8). An absence or severe abnormality of the ABR at maximum presentation levels in ears with SNHL is consistent with significant cochlear and neural damage. If there is a significant cochlear damage, cochlear microphonics is expected to be absent.

Otoacoustic emissions: In our study, DPOAEs were present in 70% individuals and showed higher amplitude in emissions especially at mid frequencies compared to low and high frequencies. OAE response reflects the function of cochlear amplifier and outer hair cells. Presence of otoacoustic emission in ears with absent ABR is therefore suggestive of auditory neuropathy(10). In remaining 30% individuals even though DPOAES were absent cochlear microphonics, the diagnostic indicator of ANSD was present.

II) Management Phase

Management phase included rehabilitation of 30 individuals (17 females, 13 males; age <40 years) diagnosed with ANSD using hearing aid and Cochlear implant. The individuals who were satisfying the diagnostic criteria of ANSD were given a trial of hearing aid for 3 months with regular follow up. During the follow up, speech reception, discrimination abilities and tolerance problems were assessed.

All the subjects diagnosed with ANSD were given hearing aids for trial in both ears for three months time and advised to use hearing aids continuously and regularly for

one month period of time. Those individuals who were not benefitted with hearing aids were given the option of cochlear implantation and they were under regular follow up. During the follow up visits, speech production, speech discrimination, music perception and telephonic conversation were assessed.

Hearing aids in ANSD

After binaural hearing aid fitting, all the subjects were asked to use it regularly initially for a period of 3 months. Regular follow up visits were planned and they were asked to come for hearing aid reprogramming if they have any issues with amplification. The improvement in speech clarity with hearing aid amplification was limited and the acceptance of amplification was poor in most individuals. Improvement in speech perception were minimal. 30% showed benefit after hearing aid use. They reported good sound awareness and aided speech discrimination at a satisfactory level. They did not show any tolerance problems at higher sensation levels. Despite of periodic hearing aid programming and follow up, 70% individuals showed limited improvement in aided thresholds. They also exhibited tolerance problems at higher sensation levels. In them, 40% of individuals showed no benefit with amplification devices due to discomfort issues and 30% removed hearing aids after a relatively short period following poor improvement in auditory skills.

Cochlear implantation in ANSD

Individuals with poor speech discrimination and no benefit from hearing aids were considered for cochlear implantation. Four individuals underwent cochlear implantation and they showed improvement in aided threshold and received auditory sensations to electrical current at normal presentation levels. Hence their aided audiogram showed thresholds within speech banana range. They also exhibited good performance in speech discrimination tasks. Music perception of 3 individuals also improved gradually following implantation and they could use telephone effectively. Postoperative speech production skills were similar to normal adults. Neural response imaging or telemetry was present in three individuals.

RESULTS AND DISCUSSION

This study aimed to find out the best diagnostic and management options for adults with ANSD. Various researches show the prevalence rates of ANSD range from 0.23 % to 15% in individuals with hearing loss (9,10,11). In our study we observed a prevalence rate of 1% which was similar to other studies. We couldn't find any exact etiological factors which lead to ANSD in adults. Sininger and Oba (2001) carried out a survey in adult and pediatric individuals and found that auditory neuropathy occurred without associated risk factors in 27% individuals (8). In our study, we could not find any definite etiological factors similar to that of infantile variety. We noticed a family history of hearing loss in 5 individuals which may be due to a genetic cause for the problem. It needs further evaluation and investigations for confirmation.

The results of audiological evaluation were similar with the studies reported in literature (4,5,7). It is more likely that in individuals with auditory neuropathy, the auditory nerve is not able to provide sufficiently high or synchronized rates of discharge to activate the motor neurons of the stapedius muscle (10,11) and hence stapedial reflexes are absent. Also they show normal outer hair cell function in the cochlea and hence robust DPOAEs could be measured. However eighth nerve that carries electrical signals to the brain has responses that are dyssynchronous (2,12,13). This can be attributed to the absence of ABR in individuals with ANSD. Hearing aid trial was given to all individuals, but only a minority showed improvement in symptoms while others had no improvement or even worsening of symptoms after a follow up of 3 months. The problems that were observed

during follow up included poor improvement in signal clarity, poor acceptance of amplification and speech perception, discomfort issues, and some individuals removed hearing aids after a relatively short period following poor improvement in auditory behaviors. Limited benefit in hearing aid performance can be attributed to poor speech discrimination abilities which in turn are the result of dys-synchronous firing of auditory nerve fibers.

Only 4 individuals were ready to accept the option of cochlear implantation. Main reasons were financial concerns and concerns about losing the residual hearing in case Cochlear Implants were not found beneficial after surgery. All those individuals who underwent cochlear implantation are on regular follow up till date. In all individuals improvement was noted in hearing threshold, speech reception and discrimination skills. Berlin et al studied the evaluation and management of auditory neuropathy in both infants and adults and reported improvement in 85% of the individuals which were implanted. (5)

CONCLUSION

From this study we conclude that Audiological investigations such as Pure Tone Audiometry, Immittance Audiometry, Oto Acoustic Emissions and Auditory Brainstem Response play the key role in diagnosis of Auditory Neuropathy Spectrum Disorder. The benefits of hearing aids are limited in adults with ANSD but good improvement was seen in patients who had undergone cochlear implantation.

Limitations And Future Directions

Many patients diagnosed with ANSD were not willing for Cochlear Implantation due to financial concerns and also fear of losing their residual hearing in case the cochlear implant was found not beneficial after the surgery. Other nerve conduction studies like Promontory Stimulation Test were not used to differentiate between pre & post synaptic ANSD. We got only 4 individuals who were ready for Cochlear Implantation. A study with large sample size and long follow up is needed to assess the exact outcome of various management options in adults with ANSD. Since there is much controversy regarding the effectiveness and reliability of Promontory Stimulation Test, development of any another reliable diagnostic tool to differentiate between pre & post synaptic ANSD will play a key role in pre-operative assessment for cochlear implantation in all individuals with SNHL.

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REFERENCES

1. Starr A, Picton TW, Sininger Y, Hood LJ, Berlin CI. Auditory neuropathy. Brain. 1996;119(3):741-53.
2. Berlin C, Hood L, Rose K. On renaming auditory neuropathy as auditory dys-synchrony. Audiology Today. 2001; 13(6):15-7.
3. Berlin CI, Hood L, Morlet T, Rose K, Brashears S. Auditory neuropathy/ dys-synchrony: Diagnosis and management. Mental retardation and developmental disabilities research reviews. 2003;9(4):225-31.
4. Deltenre P, Mansbach A-L, Bozet C, Christiaens F, Barthelemy P, Paulissen D, et al. Auditory neuropathy with preserved cochlear microphonics and secondary loss of otoacoustic emissions. Audiology. 1999;38(4):187-95.
5. Berlin CI, Hood LJ, Morlet T, Wilensky D, Li L, Mattingly KR, et al. Multi-site diagnosis and management of 260 patients with Auditory Neuropathy/Dys-synchrony (Auditory Neuropathy Spectrum Disorder*). International journal of audiology. 2010; 49(1):30-43.
6. Rance G. Auditory neuropathy/dys-synchrony and its perceptual consequences. Trends in Amplification. 2005;9(1):1-43.
7. Rance G, Beer DE, Cone-Wesson B, Shepherd RK, Dowell RC, King AM, et al. Clinical findings for a group of infants and young children with auditory neuropathy. Ear and hearing. 1999;20(3):238.

8. Sininger Y, Oba S. Patients with auditory neuropathy: who are they and what can they hear. Auditory neuropathy: A new perspective on hearing disorders. 2001:15-35.
9. Chance P, Fischbeck K. Molecular genetics of Charcot-Marie-Tooth disease and related neuropathies. Human molecular genetics. 1994;3(suppl 1):1503-7.
10. Doyle KJ, Sininger Y, Starr A. Auditory neuropathy in childhood. The laryngoscope. 1998;108(9):1374-7.
11. Berlin CI, Li L, Hood LJ, Morlet T, Rose K, Brashears S, editors. Auditory neuropathy/ dys-synchrony: after the diagnosis, then what? Seminars in hearing; 2002: Copyright© 2002 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel.:+ 1 (212) 584-4662.
12. Rapin I, Gravel J. "Auditory neuropathy": physiologic and pathologic evidence calls for more diagnostic specificity. International journal of pediatric otorhinolaryngology. 2003;67(7):707-28.
13. Lee JS, McPherson B, Yuen KC, Wong LL. Screening for auditory neuropathy in a school for hearing impaired children. International journal of pediatric otorhinolaryngology. 2001;61(1):39-46.

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