International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Aided Performance in Individuals with Late-Onset Auditory Neuropathy Spectrum Disorder (ANSD) - A Pilot Study

Deepthi L[1], Dr. Satish Kumaraswamy[2], Nandhu S Mohan[3]

[1]Final Post Graduate Student, Dr M. V. Shetty College of Speech and Hearing, Malady Court, Kavoor, Mangalore-15, India
Corresponding Author Email: *lathadeepthi5[at]gmail.com*

[2]Ph. D. in Speech and Hearing, Professor and Principal, Dr M. V. Shetty College of Speech and Hearing, Malady Court, Kavoor, Mangalore-15, India
Email: *sat8378[at]yahoo.com*

Audiologist
Email: *smnandhu4803[at]gmail.com*

Abstract: ***Auditory neuropathy spectrum disorder ANSD is a condition characterized by distorted transmission of auditory signals from the inner ear to the auditory nerve and brainstem. In this disorder, outer hair cell function within the cochlea remains normal, while auditory nerve function is disrupted. The disruption is believed to result from desynchronized or reduced neural activity. The term ANSD was suggested considering the varied affected loci rather than a specific site of damage. This study aims to explore the aided speech perception in individuals with late-onset ANSD using different hearing aid configurations. The effect of amplification on speech perception, including the use of acoustic enhancement strategies, is investigated to provide appropriate management for this challenging condition.***

Keywords: Late-onset ANSD, acoustic enhancement strategies, hearing aid, aided speech perception

1. Introduction

Auditory neuropathy is a disorder where the transmission of auditory signals from the inner ear to the auditory nerve and auditory brainstem is distorted. Auditory neuropathy spectrum disorder (ANSD) is characterized by normal outer hair cell function within the cochlea; however, auditory nerve function is disrupted. Neurological and electrophysical evidence suggests that disrupted auditory nerve activity is due to desynchronized or reduced neural activity or both (Zeng, Kong, Michalewski and Starr, 2004). At first, the disorder was termed auditory neuropathy as most affected individuals reported to have associated peripheral neuropathy. Later, in view of the lesion restricting inner hair cells in some cases (Miyamoto, Kirk, Renshaw and Hussian, 1999), the term auditory dys-synchrony was suggested (Morlet, Berlinand Rose, 2002).

Sininger and Hayes (2008) suggested the term auditory neuropathy spectrum disorder (ANSD) considering that the damage is not confined to a particular site in most of these persons, rather there are different affected loci. Henceforth in this study, the condition will be uniformly referred to as ANSD.

The audiometric findings from the individuals with ANSD varied significantly, with behavioral thresholds ranging from normal to profound levels. Discrimination skills were also variable (Rance, David, Gary and Richard, 1999). The configuration of the audiograms is generally variable. The audiograms could be of rising configuration (Dowel, Alison and Field, 1999), flat and unusual configuration, and rising configuration with a peak at 2 kHz (Ajith & Jayaram, 2005). It was also noted that the individuals with ANSD with peaked audiograms showed better speech discrimination skills compared to individuals with audiograms of other configurations (Kumar & Jayaram, 2005; Jijo & Asha 2012).

Individuals with auditory neuropathy (AN) often suffer from temporal processing deficits causing speech perception difficulties and the speech perception abilities are disproportionate to their hearing thresholds (Narne & Vanaja, 2008). The impaired temporal resolution is likely to affect the perception of acoustic cues such as voice onset time, burst duration and formant transitions, thereby resulting in the poor perception of consonants, mainly the stops/ plosives (Bradlow, Chiathem & Kreaus, 2000).

Management of the speech perception difficulties of persons with ANSD is always a challenge to audiologists. Majority of the studies in the literature report no improvement, with only a few reporting minimal benefit with amplification in individuals with auditory neuropathy spectrum disorder (ANSD). Most of the studies regarding the effect of amplification on speech perception in ANSD have been carried out on children. Only a few investigations have studied the aspect of late-onset ANSD (Jijo and Asha, 2013).

Persons with ANSD have negligible benefits from conventional amplification devices. Although assistive listening devices like FM systems have proved to be beneficial compared to conventional hearing aid they do not address the primary psycho acoustical difficulties (temporal processing deficits) encountered by these individuals and hence their utility is expected to be limited to a few listening conditions only (Rance, Corben, DuBourg, King, and Delatycki, 2010).

Licensed Under Creative Commons Attribution CC BY

Acoustic enhancement strategies were found to improve speech perception in persons with ANSD. Independent studies have shown that both companding and envelope enhancement strategies are beneficial (Narne, Barman & Shachi, 2014; Kumar & Jayaram, 2011). Although the acoustic enhancement strategies have been found to show improvements, none of these techniques led to complete or near complete speech perception. The benefit derived is further reduced in the presence of background noise (Barman & Shachi, 2014). This means that persons with ANSD are less likely to be satisfied with management that involves only acoustic enhancement strategies.

Plomp (1988) found that conventional hearing aids with nonlinear amplitude compression reduces the amplitude fluctuation and in fact may deteriorate the speech perception in ANSD.

To evaluate the aided performance in individuals with ANSD, Jijo and Asha (2013) conducted a study on patients with late-onset hearing loss, by comparing their speech identification scores (SIS) with and without hearing aid. An improvement in speech identification with hearing aids was observed in 30% of the individuals investigated.

Zeng and Liu (2006) reported aided improvement of speech perception in 7 out of 9 adults with late onset ANSD. They found that binaural acoustic stimulation was significantly better than monaural acoustic stimulation using both clear and conversational speech.

Barman, Sujeet and Prashanth (2015) did a study to check the influence of number of channels in a hearing aid on speech perception with low-cut modification of amplification. Results showed that low-cut amplification with low number of channels was slightly better than standard amplification for aided speech perception.

Animesh and Prashanth (2017) analysed the effectiveness of low-cut modified amplification using receiver in the canal (RIC) and behind the ear (BTE) hearing aid in individuals with ANSD. They reported that there is significantly better performance with low-cut modified RIC when the performance is compared with low-cut modified BTE.

2. Methodology

Need of the study

In real-life conditions, human speech recognition (HSR) is often negatively affected by the influence of background noise, such as traffic noise or interfering speakers in multi-talker scenarios, by reverberation, or by impaired hearing. The limited benefit from hearing aids in individuals with ANSD could be because of inappropriate fitting of the device and conventional amplification could unnecessarily amplify frequencies which may not be used by individuals with ANSD. Exploring the effect of amplification on the speech perception of individuals with late-onset ANSD is important to provide appropriate management.

Aim of the study

Thus, the present study attempts to determine the difference in aided speech perception for a person with late-onset ANSD while using a hearing aid having high dynamic noise cancellation, adaptive digital fitting formula and AutoSence OS and with a hearing aid having maximum number of channels.

3. Method

The retrospective study was carried out by reviewing the clinical records of 28 patients with late-onset ANSD. The hearing aid selected are

Hearing aid 1 (HA1)-with maximum number of channels (48 channels).

Hearing aid 2 (HA 2)-with dynamic noise cancellation, adaptive digital fitting formula and AutoSence OS.

RIC hearing aids were selected - in accordance with hearing loss. Both hearing aids were programmed in accordance with the hearing loss.

The patient's Speech Identification Score (SIS)with and without background noise and self-evaluation of hearing aid using Shortened Hearing Aid Performance Inventory (SHAPI) was administered and the scores of both hearing aids were compared.

Inclusion Criteria:

The patients selected were having bilateral symmetrical sensor neural hearing loss ranging from moderate up to moderately severe hearing loss who were fitted with hearing aid bilaterally. The age range considered was from 18 to 60 years.

Exclusion Criteria:

Patients with a history of middle ear infections and neurologic conditions were excluded.

4. Result and Discussion

Table 1: Showing the mean, standard deviation, p-value and significance of performance with HA 1 and HA 2 in 28 subjects

	N	Mean	Standard Deviation	Paired t-test p-value	Significance
SHAPI HA 1	28	110.79	5.55	0.000	HS
SHAPI HA 2	28	85.71	4.05		
In the presence of noise HA 1	28	32.71	5.56	0.000	HS
In the presence of noise HA 2	28	43.57	5.59		
Without noise HA 1	28	43.57	5.26	0.000	HS
Without noise HA 2	28	48.71	5.23		

HS-Highly Significant

Licensed Under Creative Commons Attribution CC BY

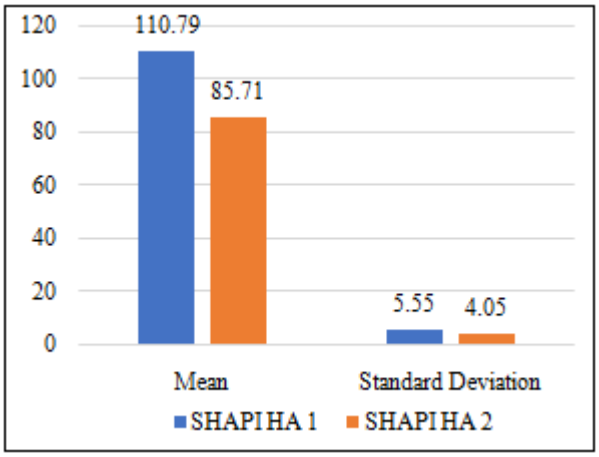

Figure 1: Showing the mean and standard deviation of SHAPI score with HA 1 and HA 2.

The high score in SHAPI indicative of poor performance of hearing aid.

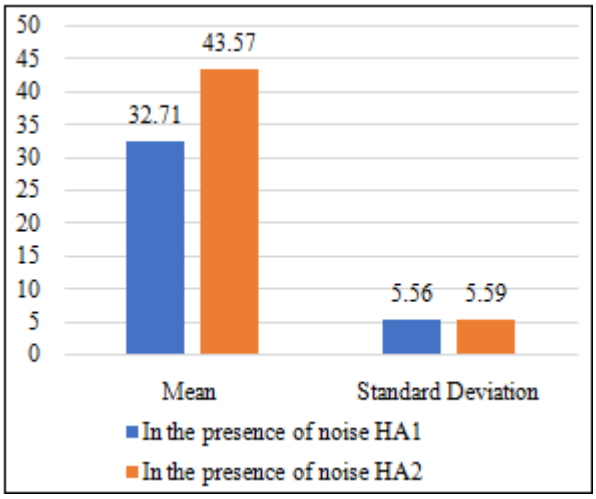

Figure 2: Showing the mean and standard deviation of HA 1 and HA 2 in the presence and absence of noise.

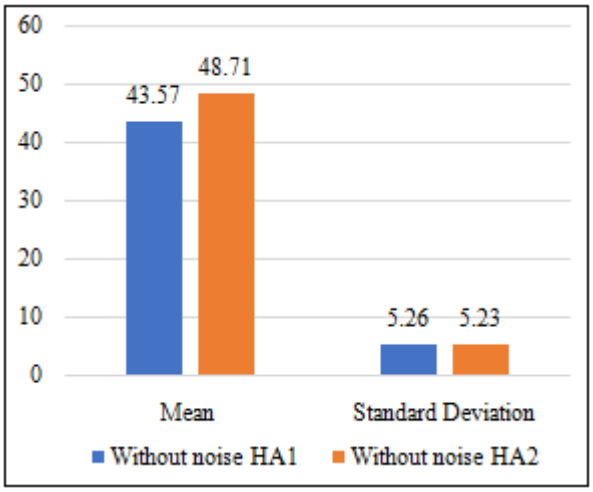

Figure 3: Showing the mean and standard deviation of HA 1 and HA 2 in the presence and absence of noise

From the result obtained on the comparison of performance with Hearing Aid 1 and Hearing Aid 2, which is shown in Table 1, Figure 1, Figure 2 and Figure 3 it is understood that a highly significant difference (P=0.000) in the mean and standard deviation was noticed.

5. Discussion

The current study aimed at finding the difference in aided speech perception for a person with late-onset ANSD while using a hearing aid having high dynamic noise cancellation, adaptive digital fitting formula and AutoSence OS and with a hearing aid having maximum number of channels. The patient's Speech Identification Score (SIS) with and without background noise is obtained and self-evaluation of hearing aid using Shortened Hearing Aid Performance Inventory (SHAPI) was administered and the scores of both hearing aids were compared. Significant improvement in the speech identification score with HA 2 was observed in the presence of noise. Reduced score in SHAPI of HA2 indicative of better performance.

6. Conclusion

For a person having late onset ANSD, the most important factor to consider while selecting hearing aid is those with better noise cancellation.

References

[1] American National Standards Institute. (1999). *American National Standard Maximum Permissible Ambient Noise Levels for Audiometric Test Rooms*. Standards Secretariat, Acoustical Society of America.

[2] Berlin, C. I., Hood, L. J., Morlet, T., Wilensky, D., Li, L., & Mattingly, K. R. (2010). Multi-site diagnosis and management of 260 patients with auditory neuropathy/dys-synchrony (auditory neuropathy spectrum disorder). *International Journal of Audiology, 49*, 30 - 43.

[3] Jijo, P. M., & Yathiraj, A. (2012). Audiological characteristics and duration of the disorder in individuals with auditory neuropathy spectrum disorder (ANSD)- A retrospective study. *Journal Indian Speech and Hearing Association, 26* (1), 17-26.

[4] Jijo, P. M. (2015). *The Effect of Intensity and Temporal Enhancement on Speech Perception in Individuals with Auditory Neuropathy Spectrum Disorder (ANSD)*. (Unpublished doctoral dissertation). University of Mysore, Mysuru.

[5] Kumar, U. A., & Jayaram, M. (2011). Speech perception in individuals with auditory dys-synchrony, *The Journal of Laryngology and Otology, 125*, 236-245.

[6] Lee, J. S., McPherson, B., Yuen, K. C., & Wong, L. L. (2001). Screening for auditoryneuropathy in a school for hearing impaired children. *International journal of pediatricotorhinolaryngology, 61* (1), 39-46.

[7] Maruthy. S., & Geetha, C. (2011). Audiovisual perception and processing in individuals with auditory dys-synchrony. Unpublished project funded by AIISH research fund submitted to AIISH, Mysore.

[8] Mathai, J. P., & Yathiraj, A. (2013). Audiological findings and aided performance in individuals with auditory neuropathy spectrum disorder (ANSD)- A retrospective study. *Journal of Hearing Science, 3* (1), 18-26.

Licensed Under Creative Commons Attribution CC BY

[9] Plomp, R. (1988). The negative effect of amplitude compression in multi channel hearing aids in the light of the modulation-transfer function. *The Journal of the Acoustical Society of America*, *83* (6), 2322-2327.

[10] Plomp, R. (1988). The negative effect of amplitude compression in multichannel hearing aids in the light of the modulation- transfer function. *The Journal of the Acoustical Society of America*, *83* (6), 2322-2327.

[11] Vanaja, C. S., & Manjula, P. (2004). LLR as a measure of benefit derived from hearing devices with auditory dys-synchrony. In *first conference on Auditory Neuropathy (*pp. 136-146). Department of Speech pathology and Audiology, National Institute of Mental Health and Neuro sciences

Licensed Under Creative Commons Attribution CC BY