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Original Article

Evaluation of Hearing Handicap in Adults with Auditory Neuropathy Spectrum Disorder

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OBJECTIVE: The present study attempted to evaluate hearing handicap in adults with auditory neuropathy spectrum disorder (ANSD). The study also attempted to determine if gender, pure tone average, speech identification scores (SIS), and reported duration of hearing loss could predict the hearing handicap in adults with ANSD.

MATERIALS and METHODS: Hearing Handicap Inventory for Adults and Hearing Handicap Questionnaire were administered to 50 adults with ANSD.

RESULTS: Using both the scales, there was a significant hearing handicap in both the social and emotional domains in adults with ANSD. SIS was a good predictor of hearing handicap compared to other variables. The poor SIS can affect communication skills leading to higher degree of social handicap.

CONCLUSION: The ignorance regarding the exact cause for their hearing problems and lack of appropriate management strategies could lead to emotional problems in individuals with ANSD. However, further studies are essential for determining hearing handicap with the use of hearing aids and cochlear implants.

KEYWORDS: Adult, social problems, emotional adjustment, cochlear nerve

INTRODUCTION

Auditory neuropathy spectrum disorder (ANSD) can be defined as a clinical disorder in which the patient exhibits normal oto-acoustic emissions (OAE), but auditory brainstem response (ABR) is abnormal or absent [1-4]. The prevalence rate of ANSD in Western countries is reported to vary from 0.5% to 11% [5-10]. In the Indian population, Kumar and Jayaram [11] reported that1 in 183 were diagnosed as having ANSD among individuals with sensorineural hearing loss. The onset of ANSD symptoms usually falls into two different age groups. The symptoms may be exhibited in infancy and childhood or the symptoms may develop in adolescence or early adulthood [1, 11, 12].The majority of the reports from Western populations suggest that only one outof four ANSD patients are over the age of 10 years [1, 12-14]. In contrast, reports from the Indian population have shown that symptoms onset is mostly in adolescence (16 to 25 years) [11, 15, 16]. The speech recognition deficits in individuals with ANSD are good in a few individuals, but in majority of them they are out of proportion to their pure tone thresholds [4, 11, 17-19]. The site of lesion can be at the level of inner hair cells (IHCs), the IHC/auditory nerve fiber synapse, or demyelination of the auditory nerve [4, 20]. ANSD is a retro outer hair cell disorder that affects patients' communication abilities because of poor speech perception [1].

ANSDis a debilitating disorder affecting communication ability. The loss of communication skills leads to poor quality of life in adults with ANSD [21, 22]. Prabhu [14] reported that individuals with ANSD experience depression and anxiety because of their communication problems. The deterioration of communication among the peer group, psychological problems, social isolation, and poor scholastic performance can also lead to hearing handicap in individuals with ANSD. There is a dearth of literature that has attempted to determine hearing handicap and its possible predictors in a large group of adults with ANSD. Hence, it is essential to quantify the hearing handicap and to try to determine the predictors of hearing handicap based on demographic and audiological test results.

Hearing Handicap Inventory for Adults (HHIA) evaluates hearing handicap using 25 questions and has emotional and social/situational subscales [23]. This inventory is widely used to assess hearing handicap because it has high internal consistency and reliability [24, 25]. The

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Hearing Handicap Questionnaire (HHQ) evaluates the effect of hearing impairment on emotional handicap [26]. It assesses the emotional distress and social participation restriction caused by hearing problems [26]. Thus, administering these two questionnaires would help to determine the degree of handicap experienced by individuals with ANSD. The communication abilities in individuals with ANSD are reported to vary across gender, duration of hearing loss, pure tone thresholds, and unaided speech identification score (SIS) [27, 28]. Thus, this study also attempts to determine if demographic details (gender and duration of hearing loss) and audiological test results (audiometric thresholds and SIS) can predict hearing handicap in adults with ANSD. The specific objectives of the study were to determine the hearing handicap using the HHIA and HHQ in adults with ANSD. In addition, it also attempted to determine if gender, pure tone average (PTA), SIS, and reported duration of hearing loss can predict the hearing handicap in adults with ANSD.

MATERIALS and METHODS

Participants

A total of 50 adults with ANSD were considered for the study with 22 males and 28 females between the age ranges of 19 to 42 years (mean=24.3 years, SD=11.29 years). All of the participants had PTA thresholds at 500 Hz, 1 kHz, 2 kHz, and 4 kHz ranging from mild (26-40 dB HL), moderate (41-55 dB HL), moderately severe (56-70 dB HL), to severe (71-90 dB HL) degree of hearing loss [29]. They were diagnosed as having ANSD based on the presence of transient evoked otoacoustic emissions and a lack of ABR. They had no history or presence of middle ear pathology with A-type tympanogram [30] and they had no acoustic reflexes. The diagnosis of ANSD was confirmed by a neurologist. The reported duration of hearing loss ranged from 12 months to 180 months. A hearing aid trial was carried out, and the hearing aids were not beneficial in all of the participants of the study.

Procedure

Pure tone air conduction (AC) and bone conduction (BC) thresholds were estimated using a modified Hughson and Westlake procedure [31]. AC thresholds were obtained for pure tone frequencies from 250 Hz to 8 kHz and BC thresholds from 250 Hz to 4 kHz at octave frequencies. A 2-channel diagnostic audiometer was used to obtain AC and BCPTA thresholds and SIS. SIS using headphones were obtained for phonemically balanced words. Recorded word lists were routed from a personal computer through a 2-channel diagnostic audiometer at 40 dB SL (re: SRT). AGrason Stadler Inc. Tympstar (GSI-TS) was used for immittance testing. The better ear of the participant was tested to obtain tympanogram and acoustic reflexes for a probe tone frequency of 226 Hz. Acoustic reflexes were measured using 500, 1000, 2000 and 4000 Hz pure tones presented to both the ipsilateral and contralateral ears. An Otodynamics ILO v.6 OAE analyzer was used to obtain Transient Evoked Oto-acoustic Emissions (TEOAEs). After ensuring adequate probe fit, TEOAEs were measured for non-linear click trains presented at 80 dB pe SPL. Waveform reproducibility of more than 50% [32] and an overall signal to noise ratio of more than 3 dB SPL [33] for at least at two frequency bands were required for the presence of TEOAEs. A Biologic Navigator Pro (Bio-logic, Mundelein, IL) AEP system with ER 3A insert earphones was used to record ABR. Click-evoked ABR was recorded twice and replicated for 100 µsec click stimuli delivered at a repetition rate of 11.1 clicks/second at 90 dB nHL. The recording was obtained for a total of 1500 sweeps, and a filter setting of 100 Hz to 3000 Hz was used. ABR was considered as absent if peaks were not clearly identified in both recordings and lacked replication.

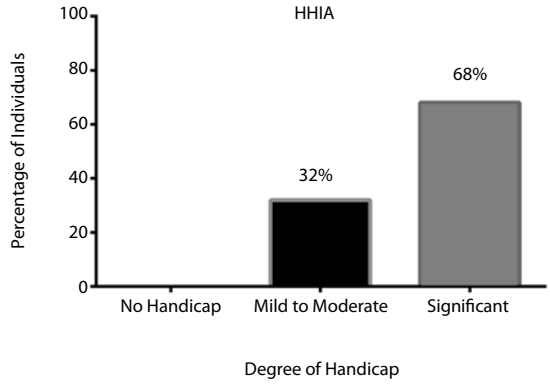

Figure 1. The percentage of individuals with different degrees of handicap on the HHIA.
HHIA: hearing handicap inventory in adults

The HHIA was administered according to the procedure suggested by Newman et al. [23]. The 25 questions in the inventory were administered, and scores for emotional scale, social scale, and total score were noted. The HHQ was also administered to all of the participants of the study. The test was administered according to the procedure suggested by Gatehouse and Noble [26]. All 12 items were administered, and the scores were calculated to determine the social restriction score, the emotional restriction score, and the global handicap score. The degree of handicap was determined based on the scores obtained on the HHIA and HHQ. Multiple regression analysis was performed to determine the predictors of hearing handicap for both questionnaires.

Ethical Considerations

Approval was obtained from the ethical approval committee of the institute, and the testing was done using non-invasive procedures. The objectives and procedures of the study were explained to the participants before evaluation, and informed consent was obtained prior to participation.

RESULTS

The results of the HHIA showed that the hearing handicap was mild to moderate in 16 out of 50 (32%) and significant in 32 out of 50 (68%) individuals with ANSD as shown in Figure 1. The mean handicap score for the HHIA was 54.9 with SD of 18.90.The results of subscales showed that 36 out of 50 (72%) had significant social handicap and 34 out of 50 (68%) had significant emotional handicap as shown in Figure 2.

The results of the HHQ also showed that the majority of the participants [35 out of 50 (70%)] had significant hearing handicap. It was also found that 36 out of 50 (72%) had a significant social restriction score and 33 out of 50 (66%) had a significant emotional distress score as shown in Figure 3.

Multiple regression analysis was done to determine if gender, duration of reported hearing loss, PTA, or SIS could predict the degree of hearing handicap. The regression model showed that the predictors explained 56% of the variance (R^2=0.56) for HHIA scores and 62% (R^2=0.62) for HHQ scores. In addition, it was found that SIS significantly

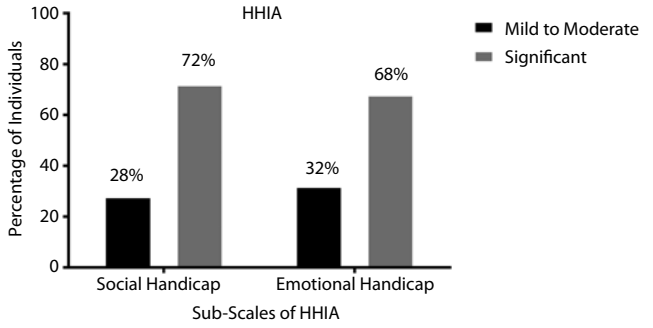

Figure 2. The percentage of individuals with handicap across the different subscales of the HHIA.
HHIA: hearing handicap inventory in adults

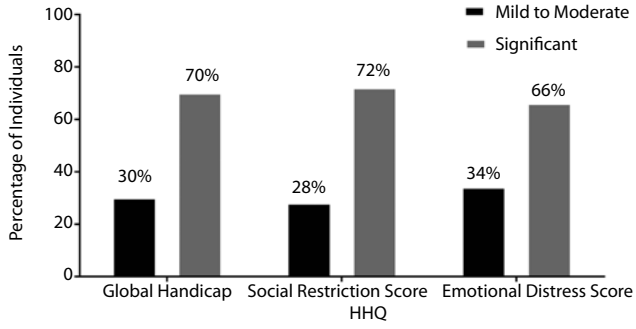

Figure 3. The percentage of individuals with different degrees of handicap on global, social restriction, and emotional distress scores on the HHQ.
HHQ: hearing handicap questionnairre

Table 1. Results of multiple regression analysis of global HHIA scores and HHQ scores with gender, duration of hearing loss, pure tone average (PTA), and speech identification scores (SIS) as the predictors

	HHIA			HHQ		
Independent Variable	**Pearson's Correlation**	**β**	**R^2**	**Pearson's Correlation**	**β**	**R^2**
SIS	−0.83*	0.73*	0.56	0.83*	0.76*	0.62
PTA	−0.05	0.01		−0.05	0.02	
Gender	−0.03	−0.11		−0.01	−0.07	
Duration of hearing loss	0.24	0.15		0.21	0.15	

*p<0.05

SIS: speech identification scores; PTA: pure tone average; HHIA: hearing handicap inventory in adults; HHQ: hearing handicap questionnairre

predicted the hearing handicap for HHIA (β=0.73, p<0.01) and HHQ (β=0.76, p<0.01). The significant negative correlation (r=−0.83, p<0.01) suggested that poorer SIS leads to larger hearing handicap. Thus, the results of the multiple regression analysis showed that SISwasa strong predictor of hearing handicap. It also suggested that PTA, gender, and duration of hearing loss were poor predictors of hearing handicap. The details of the multiple regression analysis are provided in Table 1.

DISCUSSION

This study showed that adults with ANSD experience significant hearing handicap. This study also showed that PTA, gender, and duration of hearing loss are not good indicators of hearing handicap in individuals with ANSD. However, SIS showed strong negative correlation with hearing handicap. The significant handicap was found in both the social and emotional domains of the HHIA and HHQ in the majority of the participants with poor SIS. Previous studies on ANSD report that individuals with poorer SIS have relatively more dys-synchrony [19, 34] leading to higher hearing handicap. Thus, the impaired communication ability leads to poor social interaction and isolation from peers and family negatively affecting their quality of life [21].

The ignorance regarding the exact cause for their hearing problems and lack of appropriate management strategies could lead to emotional problems in individuals with ANSD [14]. The early diagnosis and management of ANSD could lead to a reduction in handicap. Lima and Mantello [22] reported significant reduction in handicap score with appropriate hearing aid fitting. However, hearing aids are less beneficial in the majority of individuals with ANSD [1, 4, 27, 35-37]. Cochlear implant (CI)is reported to be beneficial in a few children with ANSD [1, 38, 39]. However, in a developing country like India, it is very difficult for patients to afford CI because the majority of patients with late-onset ANSD are from low socio-economic status [15]. Thus, hearing aids are the next viable option available for management of individuals with ANSD. Thus, at present the management options for late-onset ANSD are limited, especially in a developing country like India that cannot afford costly CI and frequency modulation devices [14]. There are other studies that contradict the use of Cland show that it might be of limited usefulness [40, 41]. In addition, there are very limited studies that have attempted to determine the usefulness of CI in adults with acquired ANSD. Thus, at present, there is a lack of appropriate management strategies that can benefit all of the individuals with ANSD. This could further aggravate their frustration and result insignificant hearing handicap. Thus, it is stressed that there is an urgent need for research on understanding the physiology of ANSD in detail and designing appropriate management strategies that are different from the traditional approaches to helping individuals with ANSD.

Limitations of the Study and Future Directions

The study was carried out using the English versions of the HHIA and HHQ, which are not standardized for the Indian population. There is a need for replicating the study using standardized questionnaires in the local language of individuals with ANSD. The changes in hearing handicap score with the use of hearing aids and CI should also be addressed. It is advised to assess hearing handicap in the sub-groups of ANSD patients who benefit from hearing aids. It is also essential to carry out qualitative studies using interviews or focus group discussions to better understand their difficulties and their needs.

CONCLUSION

The present study attempted to evaluate hearing handicap in adults with ANSD using the HHIA and HHQ. The study also attempted to determine if gender, PTA, SIS, and reported duration of hearing loss can predict the hearing handicap in adults with ANSD. The result of the study showed that using both the scales, there was a significant hearing handicap in both the social and emotional domains in adults with ANSD. The study also showed that SIS was a good predictor of hearing handicap. The poor SISaffect communication skills leading to a higher degree of handicap. However, further studies are essential for determining hearing handicap with the use of hearing aids and CI and for correlating ANSD with other audiological factors.

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of All India Institute of Speech and Hearing.

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

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