

Perception of Burden by Families of Persons with Auditory Neuropathy Spectrum Disorders: A Pilot Study

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Abstract

Background: The heterogeneous nature of ANSD regarding onset, degree of hearing impairment, and non-availability of treatment options has made them more dependent on other family members than those with sensory neural hearing loss. Non-availability of treatment or rehabilitation measures would be a primary factor to add more burdens to the caretakers. No previous studies have explored the perception of burden by families in auditory neuropathy spectrum disorder. **Aim:** To assess the burden faced by the family, which has a constituent individual with ANSD. **Method:** The study used a cross-sectional design. Eleven patients with ANSD were selected through purposive sampling. The study was conducted at government run tertiary care neuroscience hospital, Bengaluru Karnataka. Tools used: Family burden interview schedule was used to assess the family burden among families of persons with ANSD. The study was approved by the IEC. **Results:** The results showed a severe burden in financial aspects, a moderate burden in family routine activities, family interaction and mental health of the family members. Mild burden was seen in family leisure activities and physical health of the family members. Most of them (10) have moderate subjective burdens. **Conclusion:** Moderate family burden was noticed in family mental health, routine activities, and family interaction. Severe burdens were noticed in the family's financial aspect. The age of the patient is positively correlated with the family burden. The psychosocial wellbeing of the family members will have a significant effect on the outcome of the rehabilitation measures.

Keywords: Auditory neuropathy, family burden, speech understanding difficulty, disability, hearing difficulties.

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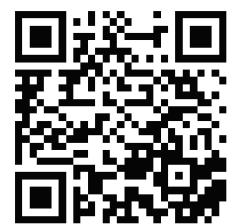
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Introduction

Hearing loss is considered to be a significant issue in public health worldwide. It is estimated that one in every ten people or 900 million will have hearing loss by 2050 (WHO, 2016). Many studies have tried to estimate the prevalence of auditory neuropathy spectrum disorder (ANSD). It remains uncertain till date, owing to the heterogeneity of ANSD patients. The prevalence of ANSD in individuals diagnosed to have hearing impairment ranged from less than 1% of hearing up to 10% (Moser T, 2006; Boudewyns A et al., 2006).

Auditory neuropathy spectrum disorder (ANSD) is a hearing disorder with unique physiological and perceptual consequences for the affected persons. It is a perplexing disorder with highly variable hearing thresholds, poor speech perception and poor co-correlation between hearing level and speech perception abilities. Adding to the difficulty is the questionable or no benefit from any available rehabilitation modes, including hearing aids. It results in communication breakdown, thus leading to social isolation and the exhibition of psychological symptoms in patients with ANSD. Disability in individuals with auditory neuropathy spectrum disorder is related to many life areas beyond their difficulty in understanding speech. It may develop secondary symptoms related to motor, cognition, social withdrawal, and so.

The burden perception among parents of hearing-impaired and intellectually challenged children is significant (Syed et al, 2020). Right from accepting the diagnosis of hearing impairment, coping with the diagnosis, understanding the impact is a stressful life occurrence with profound effects on parents and the family system with deep mental stress. Unlike hearing loss, ANSD presents with unique challenges. Majorly hearing loss occurs at a very early age and could be identified by its effect of speech and language impairment. The onset of ANSD on the other hand is mostly at the second decade of one's life, with reported incidental bias towards female (Yuvaraj P & Jayaram M, 2016). Late onset with exclusive speech understanding difficulties could often be misconstrued as behavioural

problems or poor scholastic performance. Unavailability of clear information and proper experts to diagnose ANSD, increases the family distress multi fold.

The heterogeneous nature of ANSD regarding onset, degree of hearing impairment, and non-availability of treatment options has made them more dependent on other family members than those with sensory neural hearing loss. Depending on the disease severity, behavioural impairment, and psychiatric symptoms, caregivers are at significant risk of mental and somatic health problems because of limitations in personal needs, occupational and social obligations, and financial burdens. Non-availability of treatment or rehabilitation measures would be a primary factor to add more burdens to the caretakers.

Rationale for the study

Studies have mostly focused on assessing the caretaker burden in hearing impairment. The real life issues faced by individuals with ANSD and their family members have received lesser focus. By the nature of the ANSD condition, the burden perceived by the family with a constituent ANSD member could be much higher. This study aims to assess the burden faced by the family, which has a constituent individual with ANSD.

Methods

The study used a cross-sectional design to assess the family burden in persons with auditory neuropathy (n=11). The study was conducted at the Out-patient Department of Speech Pathology and Audiology, NIMHANS, Bangalore, during the period of July 2022- December 2022. The Interview schedule was used to collect the data, and informed consent was taken from the patient's family members. A group of 11 family members who cared for a person with Auditory Neuropathy Spectrum Disorder gave consent to participate in the study. Diagnostic criteria: Individuals with auditory neuropathy spectrum disorder on the criteria for identification of auditory dys-synchrony, recommended by Starr, Sininger and Pratt (2000) which are as follows: subjects who (a) had preserved cochlear amplification, that is, who show otoacoustic emissions

(OAEs), (b) show no auditory brainstem responses (ABRs) or abnormal, if present, and (c) who show no acoustic reflexes.

Objective measures for diagnosing Auditory Neuropathy Spectrum Disorder: **Pure tone:** Air conduction and bone conduction pure tone thresholds for octave frequencies will be obtained using a calibrated clinical audiometer (GSI 61). A modified version of Hughson and Westlake procedure was followed using 5 dB step presentation size.

Testing will be carried out in a sound-treated room built as per ANSI (1991) standards for noise levels. **Speech audiometry:** Speech perception in quiet and noise will be done in a sound-treated audio room. Standardized test materials will be used to establish the speech recognition threshold (SRT) or speech detection threshold (SDT).

Immittance evaluation: Immittance evaluation for 226 Hz probe tone will be carried out with a calibrated middle ear analyzer (GSI Tympanometer). Ipsilateral and contralateral acoustic reflex thresholds will be recorded at 500 Hz, 1000 Hz, and 2000 Hz. **Recording of auditory evoked potentials:** Auditory brain stem responses (ABR) will be recorded using a standard protocol developed for ANSD.

The participants will be encouraged to rest and refrain from any activity during the procedure to avoid artifacts.

Recording of otoacoustic emissions: Distortion product otoacoustic emissions (DPOAEs) will be recorded to evaluate the functioning of the outer hair cells. The recordings will be done in a sound-treated room.

Inclusion and exclusion criteria: Family members of the individuals diagnosed as having ANSD. A family member who has at least lived with the patient for the past year and is aged between 25 and 60 years and those who speak Kannada or have a fluent reading of English, regardless of gender and education were considered.

Tools used: The 'Family burden interview schedule' (Pai and Kapur, 1981) was used to measure the objective and subjective burden on the family member of an individual having Auditory Neuropathy Spectrum Disorder. This schedule consists of 6 Domains with a total of 24 items (*Financial burden, Disruption of routine family activities, Disruption of family leisure, Disruption of family*

interaction, Effect on physical health of others, Effect on mental health of others). Family burden was rated on a three-point rating scale (Severe burden -2, Moderate burden -1 & No burden -0). After completing the interview, the subjective burden on the family as a whole was assessed, and a rating was given on a similar three-point scale. Each item was presented with 2-3 examples for a better understanding. Care was taken not to induce answers but to avoid interviewer bias. The interview was completed in one session, in which the evaluator read the questions to one of the family members and was asked to rate the actual burden for the items presented without hesitation using a three-point rating scale. Descriptive statistics such as mean, frequency and percentage were used to express the patient and clinical profiles. Ethical clearance obtained from Institute Ethics committee. Ref: NIMHANS/30th IEC (Beh.Sci & Neuro.Sci Div)/ 2021 dated 16.07.2021.

Results: Mean age of the persons having auditory neuropathy was 27.90 years (± 10.57). Out of 11 patients, seven were females, and four were males. The minimum age of the patient was 17 years, and the maximum was 49 years.

All the participants were late onset ANSD (> 12 years). Their average age at onset of illness was 17 years. Mean duration of illness was 11 years. The source of referral for six patients was self, three patients from private clinic and two from Government hospital. While five reported for hearing evaluation and another five for hearing disability certification. Three patients did not have any previous consultations. Four patients had previous consultation at Private Clinic and another four patients had audiology consultation at Government Hospital. While none of the patients were diagnosed as ANSD during previous consultation. Seven patients had undergone hearing assessment and the hearing thresholds ranged from mild to profound degree. The mean pure tone average (PTA) were 60.50 (± 21.38) dBHL and 58.68 (± 21.25) dBHL for right and left ear respectively.

Six patients never used any hearing aids, four patients tried using hearing aid and found it not to be beneficial. Those

who did not benefit from hearing aids, discontinued using it. One patient continued to use though not beneficial. Seven caregivers were males; two caregivers were females, and the other two patients, father and mother, were the caregivers. Four female patients had their father as their primary caregivers. The husband was the primary caregiver for two female patients, and the brother was the primary caregiver for another female patient.

Table 1: Socio-demographic Profile

Sl.No	Socio-demographic Profile	Categories	f(N=11)
1	Gender of the patient	Male	05
		Female	06
2	Gender of the Caregiver	Male	07
		Female	04
3	Caregiver relation	Father	04
		Mother	01
		Spouse	03
		Brother	01
		Both Parents	02

Table 2: Family Burden in Persons with Auditory Neuropathy

Sl.No	Family Burden in Auditory Neuropathy	Mean	S.D
01	Family Financial Burden	3.36	2.94
02	Family Routine Activities	2.54	1.86
03	Family Leisure Activities	0.72	1.10
04	Family Interaction	2.36	2.15
05	Physical Health of the family members	0.18	.40
06	Mental Health of the family members	1.63	.92
	Total Score	11.90	6.47

Table 2 shows a mean score of financial burden among the family members is 3.36, indicating severe burden, a moderate burden in family routine activities, mental health of the family members and family interaction. Mild burden present in family leisure activities and physical health of the family members. Most of them (10) have moderate subjective burdens. Only one family member reported severe subjective family burden. Family burden did not differ with degree of hearing loss. (F= .570, p=.653).

Table 3: Auditory Profile of the Patients

Sl.No	Auditory Profile	Categories	f (N=11)
1	SRT	Could not perform	09
		Could perform	02
2	SIS	Could not perform	09
		Could perform	02
3	SPIN	Could Not Perform	11
		Could Not Perform	11
4	OAE	Absent	01
		Present	10
5	ART	Absent	11
6	ABR	Absent	11
7	Degree of Hearing Loss	Mild	02
		Moderate	04
		Moderately Severe	03
		Profound	02

SRT- Speech Reception Threshold, SIS-Speech Identification Scores, SPIN- Speech Perception in Noise, ART- Acoustic Reflex Threshold and ABR- Auditory Brainstem Responses.

Table 3 shows the audiological profile of individuals identified to have auditory neuropathy spectrum disorder. The degree of hearing loss varied from mild to profound, where the majority of the patients had a moderate degree. Speech perception (SRT and SIS) was severely impaired in all individuals with ANSD. Only two patients could perform the speech perception task, while the remaining nine could not perform the task. None of the individuals could perform speech perception in noise (SPIN). Auditory brain stem responses (ABR) and Acoustic reflex (ART) were absent in all the patients.

Table 4: Correlation between Patient Age, Family Burden and Hearing Ability (PTA)

Variables	Correlation	PTA Right	PTA Left	Financial	Family Activity	Family Leisure	Family Interaction	Family Physical health	Family Mental Health	Family Subjective Burden
Patient Age	r	-.355	-.209	.233	.130	.854**	.685*	.659*	.027	.661*
	p	.284	.537	.491	.704	.001	.020	.028	.937	.027
†PTA Right Ear	r	1	.943**	-.098	-.055	-.209	-.316	-.206	-.112	-.138
	p		.0001	.775	.873	.537	.345	.544	.744	.686
†PTA Left Ear	r	.943**	1	-.297	-.258	-.205	-.258	-.261	-.115	-.110
	p	.000		.375	.443	.546	.445	.438	.737	.746

†PTA = pure-tone average

Table 4 shows the correlation between patient age, family burden and hearing ability. Patient age is positively correlated with burden in family leisure time, family interaction, physical health of the family members and subjective burden.

Discussion

The study aimed to explore the family burden among the family members of individuals with ANSD. The results of study revealed that the family leisure, family interaction and family physical health were severely affected. The family members of the persons with auditory neuropathy spectrum disorder showed moderate degree of family burden. The results showed that family interaction was severely affected by ANSD. One of the major contributing factors for this could be the difficulty in underrating speech. Speech identification abilities of individuals with auditory dys-synchrony have been well documented in the literature. It is the general opinion that speech identification scores of persons with auditory dys-synchrony are disproportionately poor to the degree of their hearing loss (Starr et al., 1996). In fact, poor speech identification scores disproportionate to the degree of hearing loss may be considered as the cardinal characteristic of this clinical population. In the current study, nine out of eleven patients showed zero discrimination ability. Even the other two patients had less than 50% score on speech discrimination task. The poor speech discrimination ability in ANSD might lead to a communication breakdown and withdrawal from a conversation with their family members. Family with constituent ANSD member can have psychological pathologies like shame, remorse, excessive load, feeling oneself to be caged, bitterness, isolationism and lack of control internally as well as externally (Gallagher S, 2008; Phillips AC; 2009 and Resch JA; 2010).

While the caretakers of hearing impaired also feel the burden (Syed IH, 2020), it could be far more for those with auditory neuropathy spectrum disorder. Individuals diagnosed to have hearing are corrected with appropriate hearing aids or cochlear implants. However, the general

opinion is that individuals with auditory dys-synchrony do not derive benefit from hearing aids (Berlin et al., 2003). There is irrefutable evidence that individuals with auditory dys-synchrony manifest severe problems in temporal processing. Conventional hearing aids either do not change temporal fluctuations of speech sounds (using linear amplification) or even reduce the fluctuations when a non-linear amplitude-compression circuit is employed. Therefore, conventional amplification is not beneficial for individuals with auditory dysynchrony. Secondly, individuals diagnosed to have hearing impairment are eligible for all the benefits from the government agencies. A retrospective design by Hemanth et al (2017) to investigate the effect of degree of hearing loss on speech identification scores (SISs) and aided improvement in individuals with sensorineural hearing loss (SNHL) and auditory neuropathy spectrum disorder (ANSD) concluded that present criteria (PWD Act, 2001) to issue disability certificate is suitable for individuals with SNHL, as there is a good agreement between their degree of hearing loss and speech understanding. In contrast, individuals with ANSD show severe speech understanding problems irrespective of their degree of hearing loss and do not fit the existing criteria. Lastly, Individuals with ANSD find it difficult in their schooling and also employment. All the above factors contribute to a significant family subjective burden.

Researchers are still in quest of effective treatments for people with auditory neuropathy spectrum disorder. While the options of hearing aids, cochlear implants and other technologies have been explored extensively, the outcomes of the studies have been limited (Berlin et al., 2003, Miyamoto et al. 1999). Family support at this juncture is of crucial importance for individuals with auditory neuropathy spectrum disorder. Most of the time the stress or burden of the family members is not considered, while suggesting communication strategies to persons with auditory neuropath spectrum disorder. Understanding the psychosocial wellbeing of the caretakers is an important factor for effective rehabilitation of those individuals with ANSD.

Limitations: Considering the prevalence of auditory neuropathy spectrum disorder, the current study was carried out on a smaller sample size.

Conclusion: Families with an ANSD member in it face a moderate family burden. Moderate family burden was noticed in family mental health, routine activities, and family interaction. Severe burdens were noticed in the family's financial aspect. The age of the patient is positively correlated with the family burden. The psychosocial wellbeing of the family members will have a significant effect on the outcome of the rehabilitation measures.

References

Berlin, C. I., Hood, L., Morlet, T., Rose, K., & Brashears, S. (2003). Auditory neuropathy/dys-synchrony: diagnosis and management. *Ment Retard Dev Disabil Res Rev*, 9(4), 225-231.

Boudewyns, A., Declau, F., van den Ende, J., Hofkens, A., Dirckx, S., & Van de Heyning, P. (2016). Auditory neuropathy spectrum disorder (ANSD) in referrals from neonatal hearing screening at a well-baby clinic. *European Journal of pediatrics*, 175(7), 993–1000.

Gallagher, S., Phillips, A. C., Oliver, C., & Carroll, D. (2008). Predictors of psychological morbidity in parents of children with intellectual disabilities. *Journal of pediatric psychology*, 33(10), 1129–1136.

Liu, Z., Heffernan, C., & Tan, J. (2020). Caregiver burden: A concept analysis. *International journal of nursing sciences*, 7(4), 438–445.

Miyamoto, R. T., Kirk, K. I., Renshaw, J., & Hussain, D. (1999). Cochlear implantation in auditory neuropathy. *Laryngoscope*, 109(2 Pt 1), 181-185.

Moser, T., & Starr, A. (2016). Auditory neuropathy--neural and synaptic mechanisms. *Nature reviews. Neurology*, 12(3), 135–149.

Pai, S., & Kapur, R. L. (1981). The burden on the family of a psychiatric patient: development of an interview schedule. *The British journal of psychiatry: the journal of mental science*, 138, 332–335.

Phillips, A. C., Gallagher, S., Hunt, K., Der, G., & Carroll, D. (2009). Symptoms of depression in non-routine caregivers: the role of caregiver strain and burden. *The British journal of clinical psychology*, 48(Pt 4), 335–346.

Resch, J. A., Mireles, G., Benz, M. R., Grenwelge, C., Peterson, R., & Zhang, D. (2010). Giving parents a voice: A qualitative study of the challenges experienced by parents of children with disabilities. *Rehabilitation psychology*, 55(2), 139–150.

Shetty, H., Mathai, J., & Kumar U, A. (2017). Disability certificate for individuals with hearing impairment – Time to rethink. *Indian Journal of Otology*, 23(1), 7–12.

Starr, A., Sininger, Y. S., & Pratt, H. (2000). The varieties of auditory neuropathy. *Journal of basic and clinical physiology and pharmacology*, 11(3), 215–230.

Syed, I. H., Awan, W. A., & Syeda, U. B. (2020). Caregiver Burden among Parents of Hearing Impaired and Intellectually Disabled Children in Pakistan. *Iranian journal of public health*, 49(2), 249–256.

World Health Organization (2016). Childhood hearing loss: strategies for prevention and care.

Yuvaraj, P., & Jayaram, M. (2016). Audiological Profile of Adult Persons with Auditory Neuropathy Spectrum Disorders. *Journal of audiology & otology*, 20(3), 158–167. parents a voice: A qualitative study of the challenges experienced by parents of children with disabilities. *Rehabilitation psychology*, 55(2), 139–150.