

Original Article

Activity Limitation and Participation Restriction in Adults with Auditory Neuropathy Spectrum Disorder

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Abstract

Background: Auditory Neuropathy Spectrum Disorder (ANSD) is a hearing disorder which may be characterized by normal or near normal hearing, or hearing loss ranging between mild to severe; but always associated with pronounced difficulty in speech recognition. This difficulty in speech recognition can affect communication in all situations, subsequently causing activity limitation and participation restriction in the affected individual. The current study aims to study the extent of activity limitation and participation restriction in adults with ANSD using a clinician developed tool. **Material and Methods:** Data was obtained from a sample of 23 adults with bilateral ANSD ranging in age between 18 and 52 years. The hearing status of the participants ranged from normal hearing sensitivity to severe hearing loss. A tool was developed based on ICF framework, including body functions, activities and participation and environmental factors. It consists of 83 statements spanning ten domains. Each statement was rated using a 5-point scale i.e. always, often, sometimes, seldom and never to indicate the frequency with which the participant experienced that phenomenon. **Results:** Every adult who participated in the study faced activity limitation and participation restriction in all domains. However severe difficulty was faced in domains of sensory functions and pain, learning, communication and domestic life. Moderate problem was experienced in domains of mental functions, general tasks and demands, interpersonal relations and interactions, major life areas, community and social life and supports and relationships. **Conclusions:** Results imply considerable activity limitation and participation restriction in adults with ANSD. Such information can help the clinician to address specific problems during intervention; especially considering that patients with ANSD do not get adequate benefit from amplification devices.

Key words: ANSD, Activity limitation, Participation restriction, ICF

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INTRODUCTION

I Auditory Neuropathy Spectrum Disorder (ANSD) is a condition characterized by normal or near normal cochlear hair cell function and absent or abnormal auditory nerve function. Difficulty hearing in noise, fluctuating hearing, and speech perception performance not predicted by level of residual hearing have been reported to be key symptoms associated with ANSD.^{1,2} The hearing threshold of the person may range anywhere between normal hearing to profound hearing loss. The person with ANSD can hear sounds but has difficulty perceiving speech signals. Oto-acoustic

emissions are present in these cases in any degree of hearing loss. The site of lesion in these cases lies either in the inner hair cells, the synapse between inner hair cells and auditory nerve or in the ascending auditory nerve. Problems faced by individuals with ANSD are primarily due to communication breakdown influencing overall quality of life. Due to the severe difficulty in speech perception associated with it, ANSD could have various effects on the individual affected by it as well as on the family members of the affected person. The impact of ANSD on the person may affect day-to-day functioning,

mental state, social behavior, communication, education, work performance, economic state, emotional state of person, etc.; thus, affecting overall quality of life. The family may also go through emotional problems or may be affected economically, etc. As hearing loss in general and ANSD in particular, may be associated with impact on several areas of functioning of an individual, assessment of this impact needs a holistic tool. The World Health Organization (WHO) developed a classification known as The International Classification of Functioning, Disability and Health (ICF) in 2001³ for application to various aspects of health. The ICF focusses on the impact of an impairment on the individual. It provides a description of situations with reference to human functioning and its restrictions and serves as a framework to organize this information. It structures the information in a meaningful, interrelated and easily accessible way. ICF presents conception of health and disability in a new light. It includes health domains and health related domains and consists of two lists: one which consists of body structure and functions, and a second list which includes domains of activities and participation and further includes a list of environmental factors, since an individual's functioning and disability occur in a particular context. The ICF is a multipurpose classification. It is constructed to study various disciplines and different aspects of the individual. The aim of the classification is to be very specific but also interrelated at the same time. It can be used at the individual, institutional, and social level. It can be used as a research tool or clinical tool, social policy tool and an educational tool. The effects of ANSD could be pronounced as individuals with this disorder have a severe problem in perceiving speech and most of them do not get benefit with hearing aid. Thus, they may have severe activity limitation and participation restriction. The ICF framework could be effectively applied to study the activity limitation and participation restriction in adults with ANSD which would help us to know the effects of the disorder on the affected individual. Many studies have reported on quality of life or activity limitation and participation restriction in individuals with pre-lingual or post-lingual hearing impairment,^{4,5,6,7} but none have reported it in adults with ANSD. Thus, the current study is

taken up to study the effect of ANSD on activity and participation in adults using a clinician developed tool based on the ICF framework.

MATERIAL AND METHODS

The study used a Survey design. Clearance was obtained for the study protocol from the Ethics Committee of the AYJNISHD, Mumbai. All procedures were in strict adherence to the approved protocol and the study was conducted between September 2014 and February 2015 at the institute.

Participants: Data was obtained from a sample of 23 adults with ANSD who were diagnosed on a test battery comprising of pure tone audiometry, speech audiometry, Oto-Acoustic Emissions, and Auditory Brainstem Response. A diagnosis of ANSD was made when the tests showed a presence of oto-acoustic emissions and absence of Auditory brainstem response. Participants with retro-cochlear hearing loss due to other causes were excluded. The average age of the participants was 32.1 years and range was between 18 and 52 years. There were 12 females and 11 male participants. The hearing status of the participants ranged from normal hearing sensitivity to severe hearing loss and had ANSD bilaterally. The demographic details of the 23 participants are shown in Table 1.

Tool: For development of the study tool, ICF core sets for hearing loss published by WHO⁸ were reviewed. Statements covering body functions, activities and participation and environmental factors were prepared. These statements denoted problems faced by the individual in the various areas of functioning. As specified by ICF, a five-point rating scale viz. never, seldom, sometimes, often, always was used to denote the severity of the problem faced by the person. The qualifiers for the scale have been derived from the ICF 0-4, wherein the response 'never' was scored as 0, 'seldom' was scored as 1, 'sometimes' as 2, 'often' as 3, and 'always' was scored as 4. Thus, for every statement, the maximum attainable score is 4 and minimum score is 0. The greater the score the greater is activity limitation and participation restriction. 'Not applicable' was also provided as one of the options for each statement. The number of statements collated to form the tool was 125. This was validated by five ASLPs familiar with ICF. Each of the five respondents rated each statement as appropriate and not

appropriate for and after this process, the final tool consisted of 83 statements, covering ten domains as shown in Table 2. The tool was then translated into two local languages – Marathi and Hindi by three native speakers of these languages and back translated by three other native speakers. The translated version that was the closest to the original was retained for the respective language.

Procedure: Detailed case history was acquired from each participant and informed written consent was obtained. 19 of the 23 participants answered the questionnaire through face-to-face interaction with the researcher while 4 of them answered and returned it via e-mail.

Table 1: Details of participants

No.	Sex	Age	Left ear status	Right ear status	Hearing aid use	Occupation	Age of onset
1	F	52	Moderate	Moderately severe	Yes	Housewife	15 years
2	F	25	Normal	Normal	No	Housewife	Childhood
3	F	35	Normal	Normal	No	Housewife	Childhood
4	F	19	Moderate	Moderate	Yes	Student	Childhood
5	F	36	Moderately severe	Moderately severe	No	Business	10 years
6	F	20	Moderate	Moderate	No	Student	Childhood
7	M	21	Moderate	Severe	No	Student	Childhood
8	M	43	Moderate	Moderately severe	Yes	Shopkeeper	12 years
9	M	44	Moderately severe	Severe	Yes	Police	14 years
10	F	36	Moderately severe	Moderate	Yes	Housewife	Childhood
11	F	18	Moderate	Moderately severe	Yes	Student	Childhood
12	F	34	Moderate	Moderate	No	Housewife	Childhood
13	M	44	Moderate	Moderately severe	Yes	Clerk	15 years
14	M	36	Moderate	Moderately severe	Yes	Vegetable vendor	Childhood
15	F	19	Moderate	Mild	No	Student	Childhood
16	M	21.6	Moderate	Moderately severe	No	Farmer	Childhood
17	M	33	Severe	Moderate	No	Not working	Childhood
18	M	40	Moderate	Mild	Yes	Shopkeeper	20 years
19	F	30	Mild	Normal	No	Business	Childhood
20	M	38	Moderately severe	Moderate	Yes	Watchman	10 years
21	M	35	Moderate	Moderate	No	Service	15 years
22	F	35	Mild	Moderate	No	Service	Childhood
23	M	24	Moderate	Moderately severe	No	Student	Childhood

Table 2: ICF Domains included in the tool

Domain number	Name of domain	No of Item	Example	Max score
1.	Mental functions	10	I lack self- confidence since I have the hearing problem	40
2.	Sensory function and pain	8	I find it difficult to locate the source of sound.	32
3.	Learning	8	I cannot understand the lecture or seminar which I am attending as I have hearing problem.	32
4.	General tasks and Demands	4	I find it difficult to hear the alerting signals while I am driving a vehicle or when I am walking on the road.	16
5.	Communication	15	I feel left out of conversations due to my hearing problem.	60
6.	Domestic life	2	I have difficulty communicating in situations such as renting a home, or buying anything from a shop.	8
7.	Interpersonal interactions and Relationships	8	I find it difficult to maintain my relationship with friends, and relatives because of my hearing problem.	32
8.	Major life areas	11	I was fired from my job because of my hearing problem	44
9.	Community and social life	2	I avoid social or community gatherings like weddings, birthday parties, religious functions, etc. due to my hearing problem	8
10.	Supports and relationships	15	People tend to avoid me or boycott me because of my hearing problem.	60

RESULTS:

The frequency distribution for the median ratings for the ten domains is shown in Table 3. It can be seen from the Table 3 that all participants experienced activity limitation and participation restriction in learning, communication and domestic life. Percentage of participants reporting considerable problems was the highest for domains of learning, general tasks and demands,

communication, domestic life, interpersonal interactions and relationships and major life areas. Descriptive statistics were obtained by calculating medians and percentile scores obtained by the participants in each domain. Results obtained are given in Table 4 for each of the ten domains. Analysis indicates that participants have a Moderate problem in the domains of Mental Functions, General tasks and Demands, Interpersonal interactions and

relationships, Major life areas, Community and social life and Supports and relationships whereas severe problem was observed in the areas of Sensory functions and pain, Learning, Communication and Domestic life. From the above findings, it can be seen that more activity limitation and participation restriction

is seen in areas of communication, learning, sensory functions and pain and domestic life as compared to other domains. The quartiles and the median scores for the various domains are depicted in Figure 1.

Table 3: Frequency distribution of the five ratings for the ten domains

	Always	Often	Sometimes	Seldom	Never
Mental functions	13.48	32.61	31.30	12.17	10.43
Sensory functions and pain	23.37	32.07	15.22	11.96	17.39
Learning	38.59	41.85	15.22	3.80	0
General tasks and demands	13.75	30	46.25	5	5
Communication	33.04	31.27	29.79	5.01	0
Domestic life	13.04	41.30	32.61	13.04	0
Interpersonal interactions and relationships	10.33	35.33	36.41	10.33	7.61
Major life areas	8.41	24.77	54.21	4.67	7.94
Community and social life	4.35	28.26	32.61	15.22	19.57
Support and relationships	3.72	17.03	35.91	18.58	24.77

Table 4: Median score for each domain

Domain	Maximum possible score	Median score of participants
Mental functions	40	22
Sensory Functions and Pain	32	18
Learning	32	25
General tasks and demands	16	14
Communication	60	43
Domestic life	8	5
Interpersonal interactions and relationships	32	19
Major life areas	44	20
Community and social life	8	3
Supports and Relationships	60	32

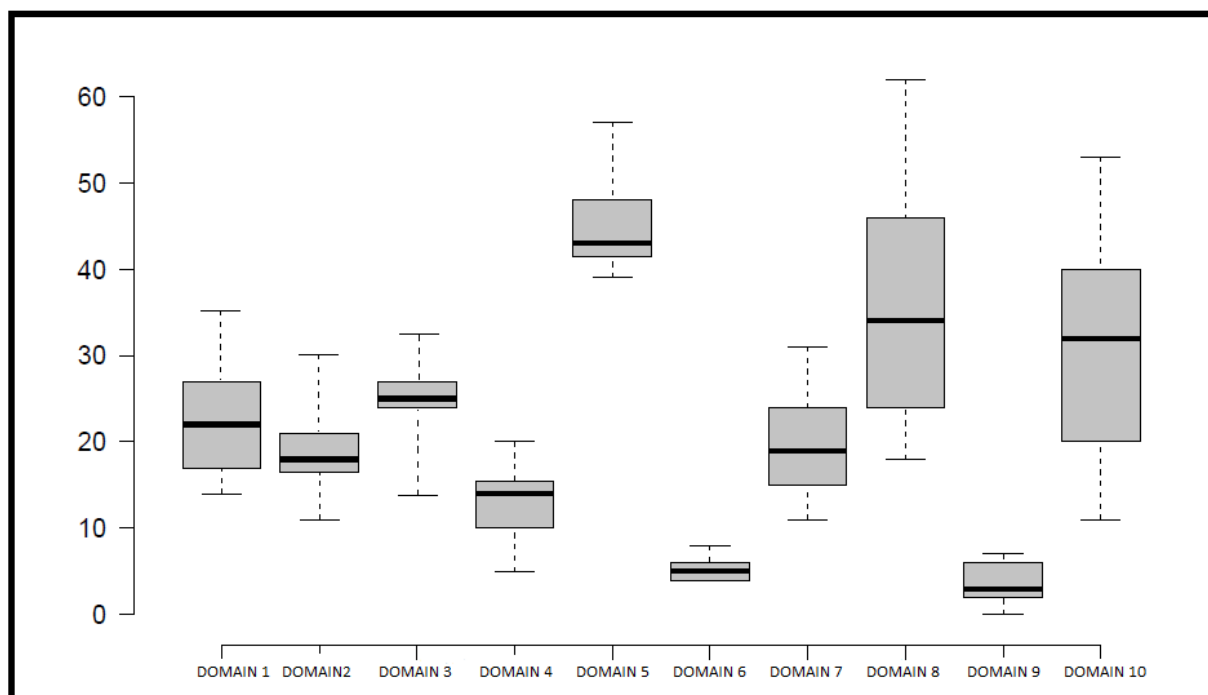


Figure 1: Graphical representation of scores for the various domains

DISCUSSION:

For Mental Functions, the maximum rating was observed for “often” indicating that most of the participants perceived activity limitation and participation restriction in this domain where skills like attention or feelings like self-confidence, distress, anxiety, etc. are probed. It can be presumed that persons with ANSD may face such feelings primarily due to the severe difficulty in understanding speech of others and inability to participate in conversation in important situations. Association between psychological health and hearing loss has been studied by several authors^{9,10} who report increased feelings of loneliness and distress, increasing significantly with an increase in the degree of hearing loss, primarily due to more of the speech signals being missed. More than 50% participants perceived significant difficulty in the domain Sensory functions and pain, which looks into aspects like localization, difficulty in hearing sounds, and differentiating the sounds. This is expected as except two of the participants, others had a significant degree of hearing loss, making it difficult for them to respond to auditory signals and locate or discriminate them. The domain of Learning considers the aspect of perceiving and understanding of speech sounds for learning or understanding tasks. The maximum rating was observed for ‘often’, indicating that most of the participants perceived activity

limitation and participation restriction in the domain of learning. Six of the 23 participants were students and experienced marked difficulty understanding speech during lectures or verbal instruction during class. Participants also reported difficulty ‘sometimes’ in the domain of General tasks and demands. This domain looks into the area of taking up responsibilities or following rules. Statements referring to tasks such as responding to alerting signals at home or while walking on the road were included in this domain. Considerable difficulty in this domain is also primarily owing to the loss of hearing sensitivity making it difficult to access auditory signals important in completing general tasks. The Communication domain considers various aspects of being able to communicate, or using some kind of cues or speech reading to understand what is said, or problems during discussion etc. Most participants reported considerable difficulty in this domain. The results suggest that the participants faced a lot of problem in communication and this correlates with a similar study done by Rathna Kumar¹¹ which studied the quality of life in 41 subjects with ANSD, having moderate to profound hearing loss using Hearing Handicap Inventory for Adults (HHIA). This study highlighted the communication difficulty faced by the individuals having ANSD. The domain of Domestic life looks at the areas where we require hearing in daily

living. Here, the maximum rating was observed for 'often'. A similar study was performed on older adults by Dalton, Cruickshank, Klein and Wiley¹² to study the impact of hearing loss on daily life of older adults. Communication difficulties were assessed using HHIE questionnaire while the problems faced in activities of daily living was assessed by instrumental ADL. It was found that people having moderate to severe hearing loss reported more difficulty in activities of daily living. The study correlates with findings of the present study as majority of the participants in this study have moderate to severe degree of hearing loss with a marked difficulty in understanding speech. The domain that looks into the interpersonal relationships and interactions tapped the difficulties experienced in social interactions. Most participants provided a rating of 'sometimes'. In social interactions, it is possible that the communication partners are accommodative and go out of their way to compensate for the difficulties faced by the person with ANSD, thereby not reflecting severe difficulties in such interactions. For the domain that looks at the major areas of life like work or education the maximum rating was observed for 'sometimes'. Helvik, Krokstad, Tambs¹³ assessed possible consequence of measured hearing impairment and perceived hearing difficulty for early retirement. They reported that young and middle aged men and women who had low frequency loss had a high risk of early retirement, whereas no independent effect was found in mid frequency and high frequency losses. The above study highlights that hearing impairment affects the work efficacy of the adults and leads us to expect similar impact on adults with ANSD. The work-related problems were also studied in a study done by Brennan, Gombac, Sleightholm⁴ who performed a survey to study Participation Restriction and Activity Limitation due to hearing loss. The survey included people who were completely deaf, deafened, hard of hearing or had some hearing loss. It examines the areas of education, employment, computer usage, and aids and assistive devices, for those with a hearing limitation. The results showed that educational attainment decreased due to the hearing limitation, and the limitation also affected the career choice. In case of employment the subjects faced problem of limitation in their ability to work, discrimination at workplace and problems in advancement in workplace. Some people also were unemployed because of their problem. Similarly, persons with ANSD who have severe trouble with speech

perception would face similar or more problems in education and work.

The domain that looks at social and community life obtained a maximum rating of 'sometimes'. Here again, it could be possible that many of the participants, especially those who were housewives tried not to participate in social and community events. They avoided such situations and hence provided a rating of 'sometimes' referring only to the limited instances when they participated in such situations. The domain assessing supports and relationships looks at the attitude of society as self-perceived by the person with ANSD. For this, most participants rated 'sometimes'. Negative attitude of significant others was reported by many participants with ANSD in this study.

CONCLUSIONS

Based on obtained results it can be concluded that every adult who participated in this study faced activity limitation and participation restriction in all the domains. However severe activity limitation and participation restriction was faced in domains of sensory functions and pain, learning, communication and domestic life. Though the results of the study cannot be generalized to a larger population of individuals with ANSD, they help in gaining a better understanding of this specialized population of individuals so that we can better attend to their needs and appropriately assess outcomes. One of the major challenges faced by the clinician and the therapist is the rehabilitation of persons with Auditory Neuropathy Spectrum Disorder because these individuals are not only difficult to diagnose but it is also difficult to understand in what areas they are facing more problems. The results of the present study have provided insight into specific areas in which adults with ANSD may face severe problems. This may help in implementing new therapeutic strategies that may optimally improve the quality of life of people with ANSD.

LIMITATIONS

Like all qualitative research, this study has some limitations. The sample is small, and local. No assumptions regarding generalization can be made. The reports are totally interview based which may have led to overstating or understating the problems faced by the individuals. Effect of different co-variables like age, degree of hearing loss, use of amplification device, etc. was not

analyzed. Age range was very wide and participants were not equally distributed across equal age intervals. No standardized test was used to measure activity limitation and participation restriction.

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