PROBLEMS AND LIFE EFFECTS EXPERIENCED BY INDIVIDUALS WITH AUDITORY NEUROPATHY SPECTRUM DISORDER: AN EXPLORATORY STUDY USING THE ICF CLASSIFICATION

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CERTIFICATE

This is to certify that this dissertation entitled **"Problems and Life Effects Experienced by Individuals with Auditory Neuropathy Spectrum Disorder: An Exploratory Study Using the ICF Classification"** is the bonafide work submitted in part of fulfillment for the degree of Master of Science (Audiology) of the student with Registration Number: **20AUD011**. This has been carried out under the guidance of a faculty of this institute and has not been submitted earlier to any other university for the award of any other Diploma or Degree.

Mysuru,

August, 2022

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Dedicated to

Acha and Amma

For their endless love, support, and encouragement

&

To all those individuals with ANSD*,*

hoping this study will help them one day!

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ABSTRACT

Auditory Neuropathy Spectrum Disorder is a condition with diversities, including an array of audiological and non-audiological manifestations. The overall impact of Auditory neuropathy spectrum disorder (ANSD) can be understood only through systematic profiling of its auditory and non-auditory effects. The conventional audiological test battery assesses only the former and largely ignores the latter's effects. Employing the International Classification for functioning disability and health (ICF), this study aimed to profile the overall impact of ANSD and correlate them with conventional auditory measures.

The study sample included 30 adults with Auditory Neuropathy. The data was collected from all the participants using two open-ended questions: a Problem effects Question (PQ) and a life-effects question (LEQ). A content analysis approach was used to link the responses to ICF categories. There were 125 responses related to PQ and 180 for LEQ, with body function and activity limitation being the most affected domains in PQ and LEQ, respectively. The functions most commonly affected under each domain is discussed. No correlations were found between the ICF-based responses and audiological measures such as Pure Tone Audiometry (PTA) and Speech Identification Scores (SIS), signifying the need for profiling the heterogeneity of ANSD using internationally established tools like ICF.

Keywords: Auditory Neuropathy Spectrum Disorder (ANSD), International classification for functioning disability and health (ICF), Open-ended Questions, Problem Question (PQ), Life effect Question (LEQ), Activity Limitation participation restriction, Body functions.

Chapter 1

INTRODUCTION

The term Auditory Neuropathy Spectrum Disorder (ANSD) is utilized to denote a condition in an individual who presents with an Auditory Brainstem Response waveform that is absent or abnormal and the presence of Otoacoustic Emission and/or Cochlear Microphonics (Berlin et al., 2010; Rance & Barker, 2009) The prevalence of ANSD in general, varies 10% among newborns (Sininger, 2002), from 1-to 10% in schools (Foerst et al., 2006); Cheng et al., 2005), and 40% in Neonatal intensive care unit (NICU) graduates(Rea & Gibson, 2003)with hearing impairment. Individuals with ANSD can be categorized as early-onset (Berlin et al., 2010; Starr et al., 2000a; Uus et al., 2011), wherein the characteristics of the disorder are presented by an individual during infancy and childhood or late-onset, wherein the typical characteristics of the condition are not exhibited until adolescence, or early adulthood (Kumar & Jayaram, 2006; Narne et al., 2014; Prabhu & Avilala, 2012). Detailed analysis of clinical features suggests that ANSD varies in diverse dimensions, including the cause and age of onset, presence of peripheral neuropathy, and behavioral and physiological measures of auditory function. (Starr et al., 2000).

Individuals with ANSD also exhibit an array of deficits such as that of temporal processing including temporal integration (Zeng et al., 2005), gap detection (Michalewski et al., 2005), temporal modulation detection (Kumar & Jayaram, 2005; Narne, 2013), pitch processing including poor pitch discrimination at low frequencies (i.e., less than 4 kHz) (De Siati et al., 2020; Rance et al., 2004), and perception of binaural beats, spatial perception including interaural time differences (Zeng et al., 2005) and speech perception in noise

(Kraus, Bradlow, Cheatham, Cunningham, King, Koch, Nicol, Mcgee, et al., 2000). Although speech understanding is the major defecit, ANSD individuals express other covert aspects that take a toll on their day-to-day functioning in the dimensions of social, emotional, psychological, economic aspects (Sonawane & Nandurkar, 2017). Sonawane & Nandurkar, (2017) studied the effect of ANSD on Activity limitation and Participation restriction on 23 adults using a validated 5- point rating scale. According to the findings, the majority of participants had issues with learning, general tasks and demands, domestic life, communication, interpersonal interactions, and relationships, in addition to other significant life domains.

Treating children with ANSD requires an individualized plan as well as providing family support for those children with ANSD(Stredler-Brown,(2002). Functionality of an individual with ANSD, especially in those who had a late onset, is reported to be affected in terms of carrying out their civic responsibilities, role in occupational setting and activities of socialization. The majority of the participants also reported a lack of sense of independence and restriction in the quantity of recreation (Sebastian et al., 2015). The disorder is also found to be associated with impairment of body functions such as tinnitus (Prabhu, 2019), vertigo (50.9%), imbalance (15.8%), headache (12.3 %), nausea (7%), vision problems (14 %) (Prabhu & Jamuar, 2017), type I diabetes (Rance et al., 2014) and deprivation of hearing (Sebastian et al., 2015). The environmental factors such as the presence of noise in the auditory scene often affects the perception of speech in individuals with auditory dysynchrony. Therefore, the use of assistive listening devices like FM systems becomes essential for an enhanced quality of speech understanding in these individuals irrespective of whether they have normal hearing or are users of hearing aid or cochlear implant, especially in the educational set- ups (Gökdoğan et al., 2016) The manifestation of these aspects most likely compromises the individual's performance at school, work, and other public places. A study conducted by Prabhu,(2018) on individuals with late-onset ANSD revealed that depression, anxiety, and stress were prevalent among them, irrespective of the severity of hearing impairment. Hence, the basis of rehabilitation should also not be confined to physiological problems, but the psychological problems should also be addressed(Prabhu, 2017a). Also, a significant amount of influence is exerted on the family members and friends as well.

1.1 Need for the study

Although, individuals with ANSD exhibit diverse manifestations of the condition, the conventional assessment of their difficulties is confined only to audiological profiling. Review of literature shows that the auditory perceptual difficulties in them is investigated using battery of behavioural such as Pure tone audiometry (Berlin et al., 2010; Narne et al., 2014), Speech identification scores, (Narne et al., 2014) Psychoacoustic tests such as Gap detection test (Michalewski et al., 2005) and Speech perception in noise (Narne et al., 2014). In addition, electrophysiological assessments (Narne et al., 2014; Sanyelbhaa Talaat et al., 2009; Vignesh et al., 2016) link the pathophysiological evidences to origins in neural synchrony at brainstem (Narne et al., 2014; Vignesh et al., 2016), cortical regions (Abdeltawwab, 2014; Gabr, 2011; Kraus, Bradlow, Cheatham, Cunningham, King, Koch, Nicol, Mcgee, et al., 2000; Narne et al., 2014; Zeng & Liu, 2006) or complex processes like identification and classification (Apeksha & Kumar, 2020, 2017). However, these studies fail to recognize the toll that

ANSD individuals face in their personal and emotional lives (Prabhu, 2016, 2017; Sonawane & Nandurkar, 2017), with deleterious impact of the disorder on their quality of life. A complete profile of the limitations in ANSD individuals on everyday listening situations can be understood by using perceptual measures using questionnaires such as Hearing Handicap Inventory for adults (HHIA,Ventry & Weinstein, 1983), Hearing Handicap Questionnaire (HHQ, (Gatehouse & Noble, 2009), Depression Anxiety Stress Scales (DASS,Lovibond & Lovibond, 1995), Satisfaction with Amplification in Daily Life (SADL, de Carvalho et al., 2015), Parent's Perspective Questionnaire (PPQ, Tokat et al., 2019), Quality of Life Scale(QOLS, Sebastian et al., 2015) along with those developed for the purpose of specific studies (Ehrmann-Müller et al., 2020; Sonawane & Nandurkar, 2017). However, these studies were conducted using questionnaires in English and other foreign languages, which have not been standardized for the Indian population. The extent to which the participants inferred the appropriate meaning of the different questions in these perceptual measures is uncertain unless carried out in their respective local languages (P. Prabhu, 2017a). The appropriateness of the questions in these scales may not apply to all types of populations (that may vary in geography, ethnicity, culture, etc.) and hence cannot capture an accurate picture of the individual or situation. Hence, the use of perceptual closed-ended questionnaires limits the choices ANSD individuals can report based on the ratings used, the questions asked, and the choices offered. Open-ended questionnaires can be advantageous as they give the participants freedom to express all their limitations and is devoid of experimental bias (Lormore& Stephens, 1994; Stephens & Pyykkö, 2011). The use of an internationally renowned classification to then code the keywords of open-ended responses can be an invaluable tool for obtaining a wholesome view of the difficulties of individuals with ANSD.

The International Classification of Functioning, Disability, and Heath (ICF) is multifaceted(World Health Organisation WHO.,2001) with codes spanning different domains of the impact of any condition on the individual's general and emotional health status. It is a dynamic tool that utilizes different codes to depict the adverse outcomes of different non-fatal health conditions. The major components of the classification include Body structure, Body function, Activity limitation, and Participation restriction, Environmental factors, and Personal factors, which play a significant role in realizing the impact of several health conditions as well as disabilities (Üstün et al., 2003). The utility of this tool in addressing the overall impact of auditory disorders such as Hearing impairment (Helvik et al., 2006), including Single-sided deafness (Durisala et al., 2017), Tinnitus (Manchaiah et al., 2018), Meniere's disease (Levo et al., 2010) as well as other vestibular system related disorders namely Benign Paroxysmal Positional Vertigo (BPPV) and Vestibular Migraine (Mueller et al., 2012) has been documented in Literature. ANSD is one such auditory condition, wherein the understanding of the disorder as a whole is relatively less recognized and spoken about. Only one notable case study gives instances of affected emotional and mental well-being in individuals with ANSD (Helvik et al., 2006; P Prabhu, 2016).

Given the scanty literature dealing with the overall perceptual profiling of the ANSD in everyday situations and the ever-increasing need to understand the same, the current study aims to utilize ICF to shed light on the hardships that individuals with ANSD experience on a daily basis, which are otherwise ignored mainly in auditory tests. The study's specific objectives were to explore the overall effects of ANSD problems on body function, activity limitation, environmental function, and personality domains of ICF Classification.

1.2 Aim of the study

The current study aimed to profile the problems and life effects experienced by individuals with Auditory Neuropathy Spectrum Disorder using the ICF classification.

1.3 Objectives of the study

- To document the perceptual correlates (auditory & non-auditory effects) of ANSD using ICF classification, specifically for the domains: body function, body structures, activities, participation, contextual factors, environmental factors, and personal factors.
- To quantify and highlight the most affected domains (Body function, Body structure, Activity limitation, and Participation restriction, Environmental factors, and Personal factors) of ICF classification in individuals with ANSD, separately for problems and life effects questions..
- To profile the clinical correlates of ANSD on pure-tone audiometry, and speech audiometry (Speech recognition thresholds/speech identification scores) and to correlate it with problems coded using ICF classification.

Chapter 2

REVIEW OF LITERATURE

2.1 Auditory neuropathy spectrum disorder (ANSD) Definition

Auditory neuropathy spectrum disorder (ANSD) is reported as a condition in which an individual's otoacoustic emissions (OAE) are present or were present, together with their auditory brainstem responses (ABR) being abnormal or absent (Berlin et al., 2010; Rance & Barker, 2009). Altered ABR is seen primarily at a higher stimulation rate, suggesting an asynchronous firing of auditory nerve fibers. However, the presence of OAE/cochlear microphonics (CM) is a reflectance of preserved cochlear amplification. Along with these discoveries, Berlin and his colleagues(2003) recommended that normal tympanometry and absent acoustic reflexes of the middle ear muscle (which is indicative of an asynchronous auditory nerve firing) can also powerfully suggest "auditory dys-synchrony," which was the term proposed as an alternate to auditory neuropathy.

Starr et al. in the year (1996) proposed the term "Auditory neuropathy" to define their ten patients who had acquired hearing loss with normally functioning cochlea. All these patients had peripheral neuropathy, which was developed later in life. In order to confirm their diagnosis, they considered the pre- neural responses (cochlear microphonic or otoacoustic emissions) present with an absent auditory brainstem evoked response. They came to the conclusive terminology of ANSD with normal outer hair cells functioning, with impaired auditory nerve firing. The usage of the term ANSD was recommended by a panel of professionals in Como, Italy, during the New-born Hearing screening in the year 2007.

On several occasions, when the cochlear microphonics (CM) are present along with an abnormal or absent ABR with or without OAE, abnormalities also can be suspected as ANSD (e.g., (Berlin et al., 1993; Deltenre et al., 1999; Rance et al., 1999; Starr et al., 1996) To accurately diagnose ANSD, obtaining Auditory brainstem responses in condensation and rarefaction polarities helps distinguish CM from neural responses (Berlin et al., 2003). Berlin et al. (2003) reported in their study about variations among ANSD patients in terms of the presence and absence of different test results, presenting symptoms, and underlying neurophysiology.

2.2 Prevalence

The prevalence of ANSD varies across different age groups. Epidemiological studies concerned with the prevalence of ANSD in children were very diverse across the evaluated population and across studies (Sanyelbhaa Talaat et al., 2009; Vignesh et al., 2014). The available studies on the prevalence of ANSD in the pediatric population range from 5.1 to 15 per 100 detected cases of sensorineural hearing loss (Madden et al., 2002). It was thought to be an uncommon disorder initially; however, current prevalence data proved that 7 to 10% of children with a permanent hearing impairment are cases of ANSD (Madden et al., 2002; Rance, 2005). Researchers proposed that 1 in each 200 hearing impaired children below the age of 3.5 years (0.5%) had audiological findings, which are enough to diagnose them as ANSD patients ((Davis & Hirsh, 1976 Cone-Wesson & Rance, 2000). However, Berlin and his colleagues (1999) estimated that ANSD is comparatively more in children with permanent sensorineural hearing loss (4%).

Kirkim and his co-workers prepared a retrospective chart in 2008 based on universal newborn hearing screening program (UNHSP) outcomes for three years of the Western Anatolian region of Turkey, suggested the prevalence of ANSD as 15.38 % in 65 neonates with unilateral or bilateral sensorineural hearing impairment. Similar to these findings, Ngo et al. (2006) reported the prevalence of ANSD as 17.3% in all the hearing-impaired neonates whoever was screened. There was a large sample systematic study of ANSD was carried out by (Rance and his colleagues (1999) on 5199 Australian neonates who were at risk for hearing impairment. A total of 109 children out of 5199 neonates were diagnosed to have hearing loss, and 12 had ANSD, which showed that approximately 1 in 9 hearing impaired neonates are reported cases of ANSD. The prevalence rate of hearing loss in children is 0.23%, out of whom 11.01% were at risk of developing ANSD. The screening program conducted by Rodríguez Domínguez and his colleagues (2007) in the year, 2007 reported that 6(5.26%) out of 114 neonates diagnosed with unilateral or bilateral sensorineural hearing impairment had ANSD.

Duman et al. (2008) conducted a study on 75 school-going deaf children aged 6-17 years and concluded that three had ANSD (4%). Out of those three children, two did not have any risk factors for developing ANSD, but one got hearing impairment after vaccination. Another school-based study done by Lee and his co-workers (2001) suggested the prevalence of ANSD as 3% in 67 children aged 6-12 years. The epidemiological data displayed a decrease in the prevalence of ANSD with advancing age, i.e.; younger children are more prone to get ANSD compared to older children. However, this can be due to an ongoing process happening in ANSD, i.e., some outer hair cells can be spared initially, but with the progress of the condition, further damage can occur, or this can happen because of high gain amplification (Sanyelbhaa Talaat et al., 2009).

Studies carried out with adults showed a lower prevalence. Lofti & Mehrkian (2007) reported rates of 1.6%; Lee et al. (2001) found a prevalence of 2.5%, and Duman et al. (2008) reported a 4% prevalence rate. But in a study done by Penido & Isaac (2013), 23 adults in a total of 27 participants of their study were diagnosed as having ANSD. Four (14.8%) of the participants were between the ages of 0 and 20 y; nine (33.4%) were between the ages of 21 and 40 y; 12 (44.4%) were between the ages of 41 and 60 y, and two (7.4%) were above the age of 60 y.

In the Indian scenario, only limited studies are done in different cities of India. There was a register-based retrospective study conducted in Mysore by Kumar & Jayaram (2009) for the data of three years, considering all age groups individuals, and considering 21236 patients (11,712 males & 9524 females) to recognize the prevalence rate of ANSD. Results of their study suggested that 11205 were diagnosed with cases of permanent sensorineural hearing loss; among them, 61 were cases of ANSD, i.e., 1 in 183 individuals suffering from ANSD (0.54%). Among the studied population, there was a drastic rise in ANSD cases in the age range of 13 - 18 years.

2.3 Effects of ANSD

The effects of ANSD can be both auditory as well as non-auditory.

2.3.1 Auditory effects of ANSD

The patient's audiometric thresholds differ from hearing sensitivity being normal to profound degree of hearing loss. Some individuals might report reduced hearing or fluctuating hearing loss and poor speech recognition, especially in noisy environments but not in quiet conditions (Rance et al., 2007). Most of the time, the adult with ANSD might have a complaint of difficulty in discriminating speech sounds or difficulty understanding speech, possibly due to altered temporal functioning (Starr et al., 1996).

The correlation speech perception has with audibility does not hold well in people with ANSD. Instead, the limiting element is frequently the degree of temporal processing disturbance(Rance et al., 2004, 2010, 2012; Zeng et al., 2005). As a result, listeners who suffer from auditory neuropathy frequently describe their main complaint as they can "hear" speech but "can't understand what is said to them". Pre- and post-synaptic abnormalities similarly affect people. Individuals with Auditory neuropathy experience signal distortion even in quiet listening environments, making background noise extremely challenging. There are three potential causes for this phenomenon: impaired gap listening, disrupted spatial streaming and rapid loudness adaptation.

In addition, there is higher inter-subject variability in the usage of temporal cues in speech perception for individuals with ANSD (Berlin et al., 2010). Literature has shown that this affects children's capability to process quickly varying acoustic signals (auditory temporal processing); which results in normal-to-severely affected speech detection as well as pure tone thresholds(Kraus, Bradlow, Cheatham, Cunningham, King, Koch, Nicol, McGee, et al., 2000; Rance et al., 2002)

The ability to integrate timing discrepancies of around 50 microseconds is necessary for the localization of low-frequency sound sources and is severely compromised in this population (Zeng et al., 2005). Auditory neuropathy patients frequently struggle to hear changes of up to 90º in sound direction, but healthy listeners may detect changes of even 3 degrees(Rance & Starr, 2015). Further, children also reported impaired auditory processing abilities; in those situations, even amplification devices such as hearing aids may not be beneficial. In addition to this, the otological complaints might also be related to body temperature and climate changes (Sininger et al., 1998; Starr & Rance, 2015)

Tinnitus is another primary auditory concern in individuals with ANSD. According to Chandan et al. (2013), 63.75 percent of people with ANSD had tinnitus (95 out of 149). Their study found that 25 out of 30 participants (83.3 percent) reported tinnitus. Additionally, according to (Kumar & Jayaram, 2009), 50 percent of those with ANSD had bilateral tinnitus. Prabhu & Sneha (2014) gave thirty people with ANSD the self-report tinnitus handicap questionnaire (SR-THQ, Shanbal, 2002). They found that 13/30 participants with ANSD had moderate tinnitus impairment.

Prabhu, (2019) to ascertain whether tinnitus is a significant issue for individuals with ANSD, all study participants received the Tinnitus Handicap Questionnaires, Tinnitus functional index (TFI, Meikle et al., 2012) and tinnitus severity index (TSI, Meikle, 1995). The findings showed that those with ANSD experienced moderate functional impairment due to tinnitus.

2.3.2 Non-Auditory effects of ANSD

Non-auditory effects of ANSD can be any adverse effects of the condition on the individual's health and well-being, other than effects on the hearing organ. Many studies have reported the non-auditory effects of ANSD, such as vestibular symptoms, abnormal voice quality, and different psychosocial aspects. Auditory and vestibular symptoms may result from ANSD, a retro-cochlear condition that affects the vestibulocochlear nerve. Along with hearing issues, ANSD patients are said to also suffer from vestibular neuropathy. The literature is replete with reports of the vestibular branch's involvement in individuals with ANSD. The term "vestibuloacoustic neuropathy" was proposed by (K. Kumar et al., 2013) for people who have both vestibular and auditory dysfunction. According to (Sazgar et al., 2010), "audio-vestibular neuropathy" involving the two branches of the eighth cranial nerve is the most prevalent pathology. The condition where only either of the branches is affected, for instance, auditory neuropathy or vestibular neuropathy, is uncommon. According to Sinha et al. (2013), ANSD patients have atypical cervical and ocular evoked myogenic potentials (VEMP). In individuals with ANSD, Sinha et al. (2013) demonstrated that VEMP was absent, and the caloric test revealed hypofunctional responses. This shows that individuals with ANSD may have inferior and superior vestibular nerve involvement. In the study done by Prabhu & Jamuar (2017) to assess the prevalence of vestibular symptoms in ANSD individuals, the findings revealed that every one in five ANSD individuals is reported to have at least one vestibular symptom, and the prevalence of vestibular symptoms are more in females than in males.

A study done by Prabhu (2019) to examine the functional impairments due to tinnitus showed that individuals with ANSD face difficulties in terms of sleep annoyances, emotional distress, aggravating auditory challenges, and noticeable deterioration in the quality of life secondary to their prevailing tinnitus. This also makes them difficult to focus on their jobs, duties, and tasks or in social gatherings, which can cause them to be depressed, irritable, frustrated and stressed. Another study was conducted to determine the hearing handicap in ANSD individuals using Hearing Handicap Inventory in adults (HHIA,Ventry & Weinstein, 1983), and the Hearing handicap questionnaire (HHQ,Gatehouse & Noble,2004) was found to have a significant hearing handicap in these individuals. The handicap was majorly noticed in the domains of social and emotional of the HHIA and HHQ in most of the individuals. Also, it correlated well with their poor Speech identification Scores (SIS), whereas the poorer the SIS higher their hearing handicap. With their poor speech identification and thus a poorer potential for communicating in society, their quality of life is negatively affected by isolating themselves from their peers and family (Prabhu, 2017).

The voice characteristics of individuals with long-standing ANSD were analyzed and rated using Consensus Auditory perceptual evaluation of voice (CAPE-V). It suggested that the ANSD individuals characterized deviant voice characteristics such as rough voice, strain, reduced loudness, breathiness, and high-pitched voice. This was concluded to be due to their reduced auditory feedback. Also, a positive correlation was found between the duration of hearing loss and deviant voice Maruthy et al., (2019).

Administration of Quality of life scale questionnaire in ANSD individuals revealed a significant effect on the psychosocial domains such as occupational role, material well-being, Health, relationship with spouse, relatives and significant other, civic activities, creativity and personal expression, socialization, active and passive recreation, etc. (Sebastian et al., 2015). Another study where World Health Organization Quality-of-Life Scale (WHO-QOL,"Study Protocol for the World Health Organization Project to Develop a Quality of Life Assessment Instrument," 1993) were administrated revealed social isolation, emotional distress, and reduced social activities because of diminished self-esteem in their social skills, impacting their quality of life significantly (Parthasarathy & Shetty, 2021).

2.4 International Classification of Functioning, Disability, and Health (ICF)

The International Classification of Functioning, Disability, and Health (ICF) has emerged during the past four decades as a classification of health aspects pertaining to a person's experience. The World Health Organization (WHO) acknowledged the weak points of the International Classification of Diseases (ICD) in characterizing the consequences of non-acute diseases in the early 1970s. The International Classification of Impairment, Disability, and Handicaps (ICIDH) was developed to explain and classify disabilities (World Health Organisation, 2001). The ICIDH was identified as an advancement in rehabilitative environments as it focused on the repercussions of diseases. Unluckily, the ICIDH also failed to include the experiences of disability groups (Schneidert et al., 2009). In the mid-1980s, the ICIDH was revised, which proceeded to form an enhanced model of the ICIDH, and ultimately designed the International Classification of Functioning, Disability, and Health (ICF) in 2001 (WHO, 2001). The broader perspective of a person's health can be assessed from information on diagnosis plus information on functioning.

When addressing aspects of health, including human functioning or impairment, the International Categorization of Functioning (ICF) is both a classification and a conceptual model. It shows the health of people with health issues rather than classifying people, i.e., The ICF may be used to define the health and health-related states linked to all medical problems (WHO, 2001). The ICF is based on a bio-psycho-social approach to functioning and health. The Conceptualization concerns the prospect of the body, the individual, and society, and also it represents the inverse designations of functioning and disability. The classification delineates human functioning with a positive view of body functions, body structures, activities, and participation. Disability, in the ICF, is defined with negative terms such as impairments (i.e., problems in body structures or functions), activity limitations, and participation restrictions. Accordingly, disability is described in respect of functioning, i.e., when the level of functioning is 'below a determined threshold along a continuum for a specific health domain (Bickenbach, 2012), functioning becomes a disability.

Moreover, ICF explains external factors (i.e., contextual factors) to better understand human functioning and disability. These are described as environmental factors and personal factors. Hence, the ICF is multi-faceted, mentioning the significance of internal and external influences on human functioning and disability.

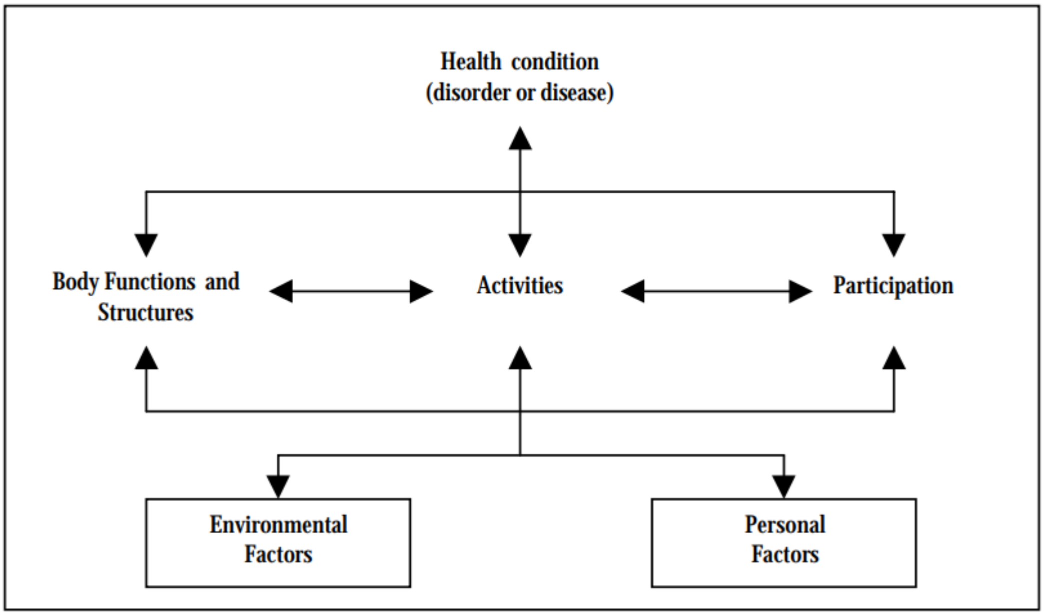
ICF is helpful with its wide range of applications, including social security, managed health care evaluation, and community surveys at the local, national, and worldwide levels. It provides a conceptual framework for knowledge relevant to personal health care, such as prevention, health promotion, and the enhancement of participation through removing or mitigating societal barriers and promoting the provision of social supports and facilitators. Additionally, it aids in the design of policies and the evaluation of health care systems.

2.5 ICF – A Conceptual Model

The ICF is an interactive model in which each factor can influence others in complex interactions. A fundamental standpoint in the model is the non-causal relationship between different components (Cieza et al., (2008); Cummins, (2009). Figure 2.1 shows the multifaceted method for categorizing functionality and impairment as a dynamic and evolving process. These interactions differ and are often not predicted to have one-to-one connections. Both directions of the relationship are active, and the existence of a handicap may even change the health status itself. It can frequently appear acceptable to infer a restriction in performance from one or more restrictions or a limitation on capacity from one or more impairments. However, it is crucial to gather information on each of these notions separately before examining relationships and causal connections between them. All elements are essential if the entire health experience is to be portrayed. Despite the fact that functioning is linked to a health condition, it is not necessarily seen as an explicit effect. Instead, it is a consequence of an intricate interaction between the health condition and contextual factors (Bickenbach, 2012).

Figure 2.1

Interactions between the Components of ICF (World Health Organization, 2001)



2.5.1 Activities & Participation

Activity is defined as "the execution of an activity or task by a person and should be evaluated regarding the nine life domains outlined in the ICF" (e.g., communication, interpersonal interactions and relationships, major life areas). Activities and participation are divided throughout these life areas; thus, either of these concepts may be present in any given area. An activity limitation is a challenge a person encounters when doing a task or action. (World Health Organisation WHO., 2001). 'Participation' is closely connected to the activity, defined as 'involvement in a life situation.' Participation restrictions are challenges a person could have when participating in life circumstances. Participation in a life situation means 'taking part,' 'being included', or 'being engaged' in an area of life (World Health Organisation WHO., 2001).

2.5.2 Body functions & Body structures

Body functions and body structures are the domains that can be found to the left in the model. Body functions deal with the physiological processes of body systems (including psychological functions), whereas body structures are related to the anatomical constituents of the body (organs, limbs). The negative aspect of these two concepts is denoted as the shared notion of 'impairment.'

2.5.3 Contextual factors

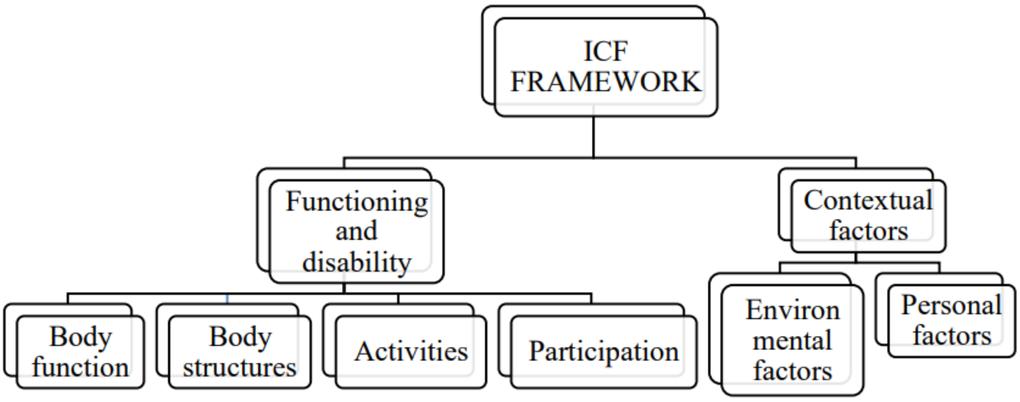
In the ICF, contextual factors have two attributes that might influence the health of an individual with a health condition: environmental and personal factors. The main difference between these two factors is the actual localization in relation to the individual, with environmental factors being viewed as external factors to the individual. In contrast, personal factors are internal. Personal factors are aspects of an individual's life and experiences that are unique to them and do not relate to their health or state of health. These variables might include things like gender, race, age, other health issues, fitness, way of life, habits, upbringing, and coping styles. (World Health Organisation WHO., 2001). Environmental factors in the ICF are viewed as human-related, i.e., social and attitudinal, or physical (World Health Organisation WHO., 2001). Environmental factors are always considered to have a positive or negative impact on the functioning of an individual in a particular circumstance and are therefore addressed as either facilitators or hindrances/barriers. The individual environment, often known as the natural or "face-to-face" environment, is how environmental factors are first and foremost defined. On the contrary, the societal environment, described as the informal and formal structural systems, is added to the individuals' living circumstances (World Health Organisation WHO., 2001).

2.6 ICF –Classification

As a whole, ICF classifies health conditions into two parts: (i) functioning and disability; and (ii) contextual factors. Figure 2.2 explains the classification of ICF under these two parts, which are further ramified into the following components: body functions, body structures, activities and participation, environmental factors, and personal factors (Schneidert et al., 2009). The physiological functioning of body systems is included in body function, such as sound detection. In contrast, anatomical parts of the body are included in the body structures (e.g. Ear and heart). An individual's execution of a task or action and participation comes under the activities. Environmental factors include the physical and social environment where people live (e.g. family support and relationships). Furthermore, the inherent aspect of the person not associated with the health condition is related to personal factors (e.g. age and gender).

Figure 2.2

ICF framework and Its Domains



2.7 ICF Core Sets

The WHO launched the ICF core sets projects to make it easier to use ICF in clinical settings or research.(Bickenbach, 2012; Stucki & Grimby, 2004). The ICF model has approximately 1500 categories, which makes it difficult to employ in clinical practice and research. ICF Core Sets have been created to solve this issue. ICF Core Sets contain the ICF categories that are most pertinent for defining how people with certain medical conditions function. A core set is defined as a collection of ICF categories of peculiar significance specific to a diagnosis or a health condition. There are core sets available for numerous conditions. There are two core sets: comprehensive core sets, comprising all ICF categories pertinent to the specific area, and brief core sets, which are a more condensed form of the comprehensive core sets (Stucki et al., 2014). Until 2017, 35 core sets for different health conditions, circumstances, situations, and generic core sets have been developed (Selb et al., 2017). The ICF core sets project targeting adults with HL was initiated in the year 2008 (Danermark et al., 2010).

In other core set projects, several other types of outcome measures have been identified when targeting the researcher's perspective, such as standard provider-reported or third-party-reported measures (e.g., clinician assessments), nonstandard measures (e.g., single questions) (Escorpizo et al., 2011), clinical measures (e.g., joint pain or joint swelling), and technical measures (e.g., X-ray) (Zochling et al., 2006). To conclude, ICF's benefits are more extensive than just measuring population health. With ICF, it is possible to identify those environmental factors that impact areas of participation, such as education, transportation, or housing, which may be determinants of health (Cerniauskaite et al., 2010). Including ICF in assessing noise-induced hearing loss will give a better outcome than the conventional audiological test battery.

2.8 ICF in other disorders

A number of studies have been reported in the field of speech and hearing where ICF classification is applied. For example, a study done to assess the application of ICF classification in individuals with voice disorders showed how their quality of life is affected in various dimensions though it is not a life-threatening condition most of the time (Ma et al., 2007). Various other hearing and balanced disorders have also been studied using ICF classification, such as hearing loss (Alfakir, 2021; Granberg et al., 2010), tinnitus (Manchaiah et al., 2022, van der Wal et al., 2021), single-sided deafness (Durisala et al., 2017), Meniere's Disease (Levo et al., 2010). In the year 2009, a group of professionals came together to develop a core set for hearing loss (Granberg et al., 2010). This will guide the professionals and other stakeholders from various dimensions associated with hearing loss to have a multidimensional view on the condition, which will assist in a better diagnosis and rehabilitation and a better perspective about the hearing impaired individual's condition. Not only have the particular disorders been studied but also the attitudes of people who are associated with individuals with various medical conditions have also been studied thoroughly using ICF classification (Pyykko et al., 2015).

ICF's value extends beyond population health measurement. With ICF, it is achievable to uncover environmental factors that impact involvement in sectors like education, transportation, or housing, activity limitations, and any participation restrictions or possibly even aspects of body functions that affect health (Cerniauskaite et al., 2010). ICF will be a better tool to provide a clearer perspective than just the standard battery of audiological tests used to evaluate ANSD because its effect on the individual is multi-dimensional.

Hence, the present study attempts to evaluate the use of ICF classification in determining the problems experienced by individuals with ANSD. It will also be attempted to correlate these findings with various audiological features of individuals with ANSD.

Chapter 3

METHODS

3.1 Research Design and Participants

The study was conducted as a combination of retrospective and prospective study designs, wherein data was collected through a qualitative interview method followed by correlating their audiological test results. Individuals with Auditory Neuropathy Spectrum Disorder (ANSD) who reported to the institute and underwent audiological testing at the department of Audiology were included in the study.

Thirty Individuals diagnosed with ANSD in the age range of 12-45 years with a mean age of 26.6 (SD=8.2) years participated in the study. Out of 30 individuals, 10 were males, and 20 were females. The degree of hearing loss ranged from Normal hearing sensitivity to Profound hearing loss. Individuals with both congenital and late-onset ANSD were included in the study. Only individuals who passed at least primary school and were proficient in English or Kannada languages were included in the study. Individuals with other co-morbidities, diagnosed with any other sensorineural or neurological conditions, and with neuropathy of other sense organs were excluded from the study.

3.2 Procedure

All the testing procedures in this study were carried out on humans using non-invasive techniques, adhering to the guidelines of the institutional research advisory board. All the procedures were explained to the participants, and informed consent was taken from all the study participants.

The individuals were diagnosed as having ANSD based on various audiological tests done at the institute, followed by correlating with the neurological reports. The different audiological tests done at the institute aiding in the diagnosis of ANSD includes pure tone audiometry, Speech audiometry, Immittance measurements, Oto-acoustic Emissions (OAE), and electrophysiological tests such as auditory brainstem response (ABR) and long latency responses (LLR). All the tests were conducted in an acoustically and electrically shielded room where the ambient noise levels were within the permissible limits per ANSI standards.

A calibrated two-channel diagnostic audiometer with TDH 39 supra-aural headphones, Etymotic insert earphones, and Radioear B-71bone vibrator were used for pure tone audiometry and speech audiometry. Through Puretone audiometry, air conduction thresholds and bone conduction thresholds were obtained for frequencies 250 Hz, 500 Hz, 1000 Hz, 2000 Hz, 4000 Hz, and 8000 Hz. Pure tone averages for air conduction thresholds were calculated for four frequencies, 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz. In speech audiometry, the speech recognition threshold was identified using a spondee word list and speech identification scores using a phonetically balanced list of 25 words (Yathiraj, 2005). The behavioral audiological evaluation also included speech identification in noise tests and uncomfortable loudness levels.

Objective tests included tympanometry for a 226 Hz probe tone and acoustic reflex test for both ipsilateral and contralateral testing at frequencies of 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz, which were performed using a calibrated middle ear analyzer (GSI Audiostar Pro, Grason-Stadler, Eden Prairie, Minnesota). Transient evoked otoacoustic emissions were recorded in a sound proof room for click stimulus at 80 dB peSPL. A response was measured to be normal whenever the reproducibility was more than 75%, and the overall signal-to-noise ratio (SNR) was greater than 6 dB SNR at least for three consecutive frequencies. If TEOAEs were not present, DPOAEs were also checked for two pure tones in the ratio of 1.2 with loudness levels of 65 and 55 dBSPLs. ILO-V6 otoacoustic emission equipment (Otodynamics Ltd, UK) was the instrument used for recording the OAEs.

Click evoked auditory brainstem response (ABR) testing was performed to check the integrity of the auditory system. Biologic Navigator Pro (Natud Medical, USA)instrument was used to record ABR. Absolute latency (Wave I, III & V), morphology, and amplitude of responses were noted down. Whenever there was a Cochlear Microphonic (CM) presence while recording ABR, the polarity was altered (condensation & rarefaction) to confirm its presence. In a few required cases, presence of LLR was also carried out for a better diagnosis. Based on the audiological evaluation, from the results obtained from the test battery, if the client was suspected of having ANSD, they were referred for neurological evaluation to confirm the diagnosis. Once the client was confirmed to have ANSD, consent was taken from the client to include as a participant in the study.

Case files were also collected from the institute's medical record section of those diagnosed with ANSD. The participants' demographic details (including age, gender, general health status, the onset of hearing loss, family history, and prior investigation) were noted.

The study was conducted in the following phases;

Phase I: Obtaining responses to open-ended questions

Two open-ended questions were adopted from Granberg et al., (2014). These questions were related to hearing-related problems faced due to ANSD and its life effects. These questions are a problem question (PQ) and a Life Effect Question (LEQ).

- PQ: Make a list of difficulties that you have as a result of your hearing problem. Write down as many as you can think of.
- LEQ: Make a list of the effects your hearing problem has on your life. Write down as many as you can think of.

The participants were asked to respond in their preferred mode, in writing, voice recording, or typed text. The participants were contacted through mobile phones and were informed about this study; only those willing to participate were included. They were given two options either answer these questions with the google form link or answer through the mobile phone. The mobile conversations were recorded and later transcribed verbatim with the help of native Kannada speakers. In cases where the participants could not understand the conversation through the phone, help from the available caregiver was sought.

Answers to these open-ended questions were coded based on different ICF components: body structure and functions, activities & participation, environmental factors, and personal factors that will cover participants' perspectives on aspects of the disability and the effects of ANSD on daily life. The questions were given to the three native Kannada speakers and were translated into Kannada. A translation that 2/3 of native speakers agreed upon was considered for use in the study. These questions were reverse translated into

English to see for translation errors (if any). The responses with translation errors were given to another blinded referee whose coding was used to accept or reject a particular code for the question. The finalized questions were loaded onto a google form containing three sections. Section 1 explained the purpose of the study, and informed consent was obtained. Section 2 targeted demographic details and the two open-ended questions (both in English and Kannada languages). Section 3 comprised auditory test results.

Phase II: International Classification of Functioning, Disability, and Health (ICF) coding

Using an analytical technique known as the "seven steps linking procedure," all of the data were linked to the ICF framework (Hsieh & Shannon, 2005). The seven steps are (1) meaningful unit identification, (2) defining the significant concept(s), (3) underlying meaning interpretation, (4) determining the linking unit(s), (5) appropriate ICF category derivation, (6) documenting the linking rule applied, and (7) verifying the representativeness of the ICF categories chosen. The transcribed responses were given to 3 coders to improve the reliability of the coding process. They were taught these coding processes and were asked to code for all the responses individually. The number of individual responses for PQ and LEQ and total responses (PQ + LEQ) were determined. If there was any disagreement in the coding, it was discussed between the coders, and a final consensus was obtained. Reliability analysis for these three coders' coded responses was carried out using IBM Statistical Package Social Sciences (SPSS) version 25.0 (IBM Corp. Released 2017. IBM SPSS Statistics for Windows, Version 25.0. Armonk, NY: IBM Corp.). The frequency counts for each code and overall domains were obtained using SPSS, while the graphic representation was conducted using Graph Pad Prism version 9.0 software (Graph Pad, California).

3.3 Quantitative Data Analyses

The statistical analysis was carried out using IBM SPSS version 25.0 and Graph Pad Prism 9 software. Descriptive statistics for means and standard deviation (SD) were obtained. The total number of responses for the PQ and LEQ questions was obtained. An inter-rater reliability check was carried out using Bland-Altman agreement analysis. This analysis was carried out for combined scores (PQ + LEQ) of the following domains: body function (BF), activity limitation and participation restriction (AL), and environmental factors (EF).

In addition, Bland–Altman agreement analysis was also done on the composite score (BF, AL, and EF). The difference in the ratings of the coders against the average ratings in all the above domains of ICF was compared. The limits of agreement are shown in a 95% confidence interval where variations between two raters should fall with ± 1.96 standard deviations (SD) of the difference scores (Giavarina, 2015) to determine any significant differences between the coder ratings. After determining the agreement between the coders, the normality check was performed using Shapiro Wilk's test. Based on the results of the normality test, Wilcoxon signed-rank test was conducted to check for the significant difference between the number of responses. Whenever significant, the effect size was calculated by using the formula $r = (Z/\sqrt{N})$. The test of Spearman's rho correlation coefficient was performed to determine the relationship between the problems mentioned in the PQ and LEQ questions with the audiological variables. The two-tailed significance level of $p < 0.05$ was considered statistically significant for all the analyses.

Chapter 4

Results

The present study aimed to profile the problems and life effects experienced by individuals with Auditory Neuropathy Spectrum Disorder using ICF classification. A total of 30 participants with ANSD were given two open-ended questions, and their responses were collected through phone interviews, google forms, and other similar modes convenient for the participants. These responses were transcribed verbatim, and three trained coders gave respective ICF codes for the keywords in their responses. The audiological data of all the participants were also collected.

Descriptive and inferential statistics were performed using SPSS (version 25.0) software. The data for this study were found to be non-normally distributed ($p < 0.05$) on Shapiro Wilk test of normality, following which . non-parametric inferential statistics were used. The study's findings are described in the sections below:

4.1 Inter-Coder reliability estimation

4.2 Quantification of problem and life effects of Auditory neuropathy spectrum disorder

4.3 ICF domain-specific Quantification of problem and life effects of Auditory neuropathy spectrum disorder

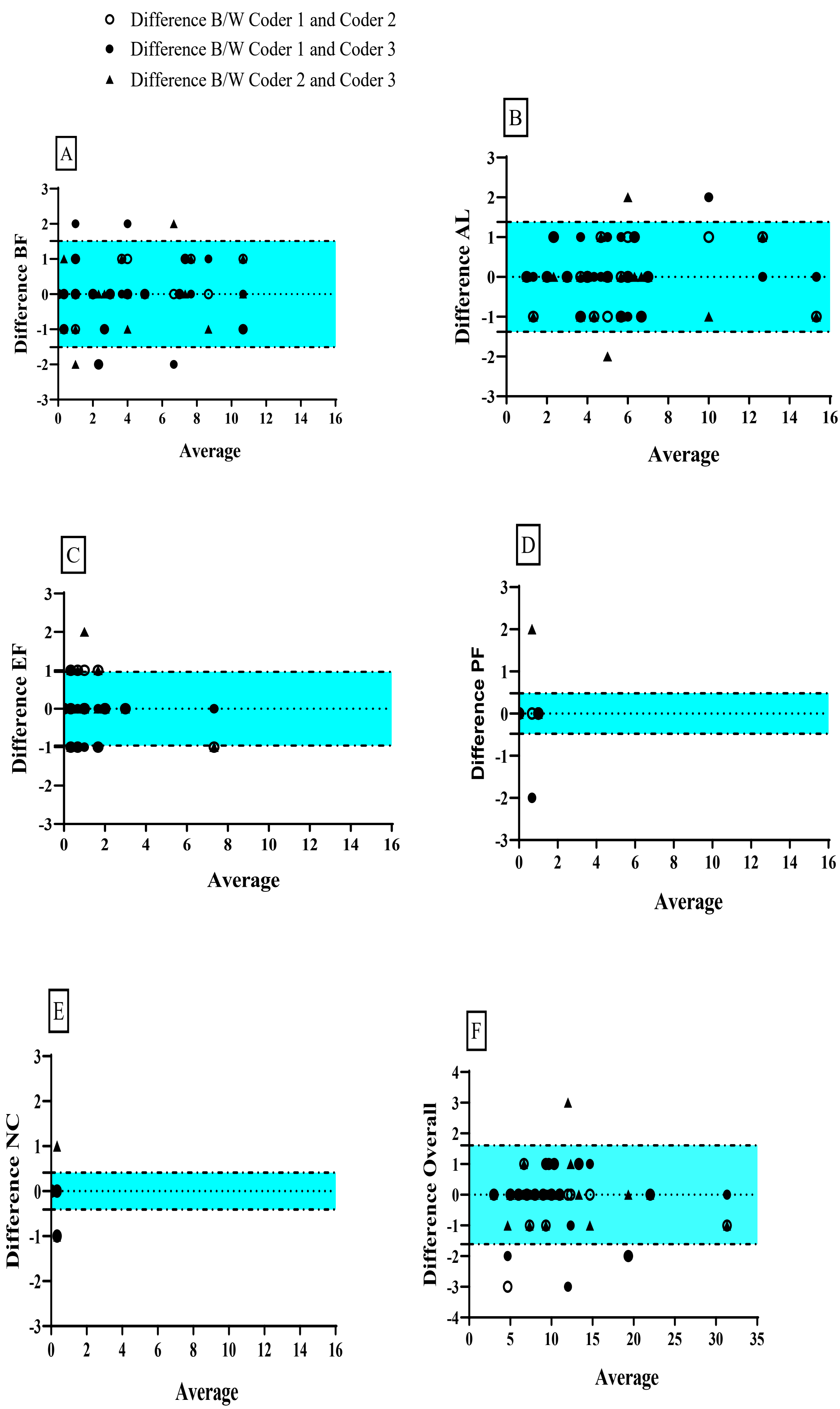
4.1 Inter-Coder reliability estimation

A Bland Altman plot analysis was carried out to estimate the inter-coder reliability among the three coders using Graph Pad Prism Version 9.4.0 (673).

The results of the Bland-Altman plot depicting the average of the total codes on the x-axis and the difference in total frequency of coder ratings (1 vs. 2; 1 vs. 3; 2 vs. 3) on the y-axis is shown in Figure 4.1 (A-F). It is evident through visual inspection itself in the Bland Altman plot that most of the composite frequency of the coders (84 out of 90) fell within the limits of variance (±1.96 SD, blue shaded area in Figure 4.1 F). The analyses of the outliers in Figure 4.1 (F) showed that 6 out of 90 observations did not correlate well, accounting for an error of 6.67%. The percentage of inter-rater reliability in composite scores was approximately 93.33%, indicating high reliability in the rater coding for overall responses. Similarly, Bland-Altman inter-rater agreement analyses for domain-wise coding indicated a biasing error of 7.78%, 3.33%, 21.11%, 2.22%, 4.44% for combined scores (PQ and LEQ) of body function (BF), activity limitation and participation restriction (AL), environmental factors (EF), Personal factors(PF), and Non-codable factors (NC). The corresponding inter-rater reliability was 92.22%, 96.67%, 78.89%, 97.78% and 95.55%. The highest intercoder reliability was observed for the domain of Personal factors, whereas the least was for Environmental factors.

Figure 4.1

Bland-Altman Plot Depicting Inter-Rater Variability for Each ICF Domain and Overall Codes


Difference B/W Coder 1 and Coder 2
Difference B/W Coder 1 and Coder 3
Difference B/W Coder 2 and Coder 3
A
Difference BF
Average
B
Difference AL
Average
C
Difference EF
Average
D
Difference PF
Average
E
Difference NC
Average
F
Difference Overall
Average

Note. (A) Body function, (B) Activity limitation and participation restrictions, (C) Environmental factors, (D) Personal factor (E), Non-codable, and (F) Overall. The blue shaded area represents the limits of agreement (± 1.96 SD) for each sub-domain and overall responses.

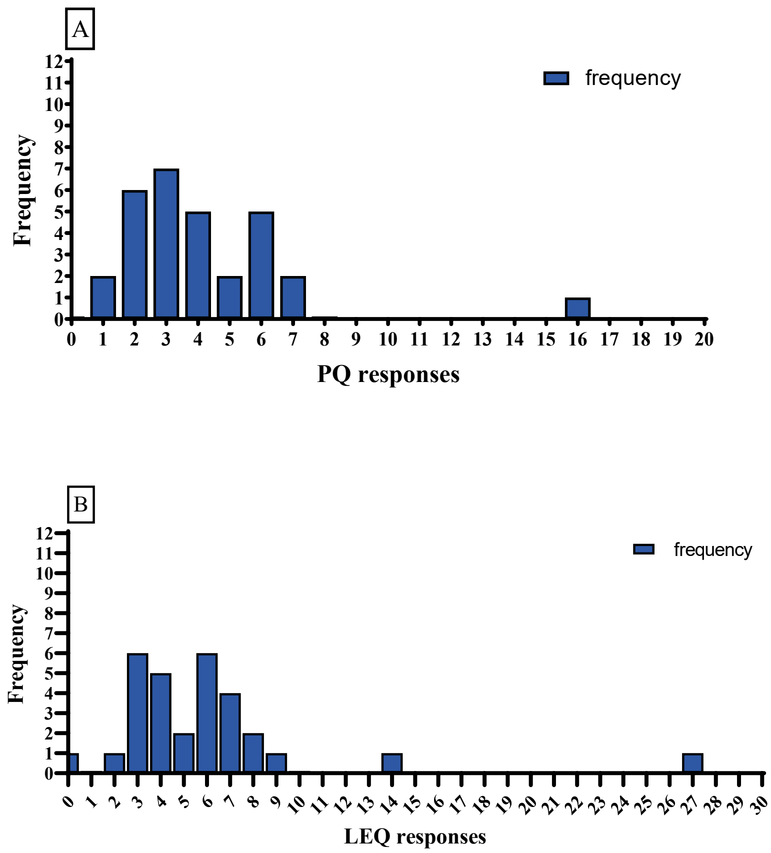
4.2 Quantification of problem and life effects of ANSD

A total of 305 responses (PQ: 125 and LEQ: 180) were obtained using the two open-ended questions from 30 adults with ANSD.

The number of meaningful responses ranged from 0 to 27, and most participants provided 2-3 meaningful responses for PQ with a maximum of 16 responses and 3-6 for LEQ with a maximum of 27 responses, as shown in Figure 4.2.

Figure 4.2

The bar graph depicting the number of problems listed in (A) PQ and (B)LEQ



For PQ and LEQ, the overall mean number of responses per participant was 4.17 (±0.52) and 6.00 (±0.87), respectively. There was a significant difference in the overall number of responses between the two open-ended questions, PQ and LEQ, as shown in Table 4.1.

Table 4.1

Number of Responses in Each of the ICF Domains Listed in the Two Questions

	PQ Mean (±one SD)	LEQ Mean (±one SD)	Wilcoxon (Z)	Sig. (*p*)	Effect Size r=(Z/√N)
Overall Responses	4.17(±0.5)	6.00(±0.87)	-2.28	<0.05	0.42

Note. SD= Standard Deviation

The Spearman's rho correlation between the number of responses to PQ and LEQ and the audiological variables such as PTA and SIS were carried out.

It showed that there was no significant relationship ($p > 0.05$) between the number of responses in PQ and LEQ questions and any of the indicated audiological variables (Table 4.2).

Table 4.2

Relationship Between the Number of the Responses to Problem and Life Effects Question and the Audiological Variables

Audiological Parameter	Spearman's rho correlation		Significance (*p*)	
	RE	LE	RE	LE
Correlation between overall PQ and audiological parameters				
PTA	0.05	-0.06	0.78	0.77
SIS	-0.02	-0.03	0.92	0.89
Correlation between overall LEQ and audiological parameters				
PTA	0.08	0.19	0.66	0.32
SIS	-0.09	-0.11	0.60	0.55

Note. RE= Right Ear, LE= Left Ear, PTA= Pure Tone Average, SIS= Speech Identification Score

4.3 ICF domain-specific Quantification of problem and life effects of Auditory neuropathy spectrum disorder

The greatest number of responses reported were the life effects of ANSD than the problem effects. Among the life effects, the most frequently listed responses were from the Activity limitations and Participation restrictions domain. There were a total of 161 responses from this domain. 102 of the total responses from the Activity limitation and participation restriction were from the LEQ, and the remaining 59 were from the PQ. The most occurred category was

“Communicating with - receiving - spoken messages” (d310), with a frequency of 54 responses. The second most frequently occurring category was “Emotional functions” (b152). Also, the domain with the second most responses was the Body function domain, with a total of 112 responses. Other frequently occurring categories were “Using telecommunication devices” (d3600), “Socializing” (d9205), “Hearing functions” (b230), “Speech discrimination” (b2301), and “Education, other specified and unspecified” (d839).

There was a total of 27 responses from Environmental factors, with a maximum number of responses from this domain as “Attitudes, other specified” (e498). There were also responses from the domain of Personal factors, majorly LEQ responses. Apart from these domains, there was also a non-codable factor.

The frequency of each code in domains such as Body function, Activity limitation, and Participation Restriction, and Environmental Factors are shown in Table 4.3 to Table 4.5.

Table 4.3

Frequency Counts of All the Responses Under the Domain of Activity Limitation and Participation Restrictions

Function	Codes	PQ n=59	LEQ n=102	Total n = 161
Communicating with - receiving - spoken messages	d310	31	23	54
Socializing	d9205	4	12	16
Using telecommunication devices	d3600	5	11	16
Education, other specified and unspecified	d839	1	11	12
Conversation	d350	3	6	9
Ceremonies	d9102	0	8	8
Communicating with - receiving - nonverbal messages	d315	3	2	5
Listening	d115	2	3	5
Higher education	d830	0	3	3
Using communication techniques	d3602	3	0	3
Parent-child relationships	d7600	0	3	3

Remunerative employment	d850	0	3	3
Relating with strangers	d730	0	3	3
Carrying out daily routine	d230	1	1	2
Informal relationships with friends	d7500	1	1	2
Undertaking a single task	d210	0	2	2
Shopping	d6200	0	2	2
Moving around in different locations	d460	1	0	1
Reading	d166	1	0	1
Recreation and leisure	d920	1	0	1
Using communication devices and technique	d360	1	0	1
Discussion	d355	1	0	1
Eating	d550	0	1	1
Handling stress and other psychological demands	d240	0	1	1
Acquiring skills	d155	0	1	1
Interpersonal interactions and relationships, other specified	d798	0	1	1

Maintaining a job	d8451	0	1	1
Acquiring, keeping and terminating a job	d845	0	1	1
Intimate relationships	d770	0	1	1
Seeking employment	d8450	0	1	1

Table 4.4

Frequency Counts of All the Responses Under the Domain of Impairments of Body Functions

Definition	**Codes**	**PQ n=63**	**LEQ n=49**	**Total n=112**
Emotional functions	b152	5	15	20
Hearing Functions	b230	9	5	14
Speech discrimination	b2304	7	6	13
Tinnitus	b2400	10	0	10
Sound detection	b2300	5	2	7
Range of emotion	b1522	0	6	6
Dizziness	b2401	5	1	6
Pain in head and neck	b28010	3	1	4

Hearing Functions, other specified	b2308	3	0	3
Pain in body part	b2801	2	1	3
Pain in body part,other specified	b28018	3	0	3
Voice functions	b310	2	1	3
Extraversion	b1260	0	2	2
Sound discrimination	b2301	0	2	2
Psychomotor control	b1480	1	1	2
Energy level	b1300	1	1	2
Attention functions	b140	1	0	1
Memory functions	b144	0	1	1
Temperament and personality functions	b126	1	0	1
Psychic stability	b1263	1	0	1
Optimism	b1265	0	1	1
Energy and drive function	b130	0	1	1
Sensation of falling	b2402	1	0	1
Irritation in the ear	b2404	1	0	1
Sensation of Pain	b280	1	0	1

Voice and speech functions, unspecified	b399	0	1	1

Table 4.5

Frequency Counts of All the Responses Under the Domain of Environmental Factors

Definition	Codes	PQ n=3	LEQ n=24	Total n=27
Attitudes, other specified	e498	1	4	5
General products and technology for communication	e1250	0	4	4
Individual attitudes of immediate family members	e410	0	4	4
Individual attitudes of acquaintances, peers colleagues, neighbours and community members	e425	0	3	3
Support and relationships, Immediate family	e310	0	2	2
Individual attitudes of friends	e420	0	2	2
Assistive products and technology for communication	e1251	0	2	2
General products and technology for personal use in daily living	e1150	1	0	1
Sound quality	e2501	1	0	1

Societal attitudes	e460	0	1	1
Individual attitudes of people in positions of authority	e430	0	1	1
Education and training services	e5850	0	1	1

The most commonly reported problems were from the domain of Activity limitations and Participation restrictions, where there were 59 PQ responses and 102 LEQ responses in total. The maximum number of responses was "Communicating with - receiving - spoken messages" (d310), totaling 54 responses.

There was no statistically significant correlation ($p>0.05$) between the PQ and LEQ responses and audiological parameters such as Pure Tone Average and Speech Identification Scores.

To summerise, the results of the study highlighted the following findings

1. There is a significant difference between the responses obtained for PQ and LEQ.
2. Activity Limitation Participation Restriction domain has the greatest number of responses reported.
3. There is no correlation between PQ, LEQ responses and audiological parameters (PTA, SIS).

Chapter 5

DISCUSSION

The study probed into the problems and life effects faced by 30 individuals with ANSD using an approach of open-ended questioning. A total of 305 responses were coded using ICF. Most of the responses ranged from 2-3 and 3-6 meaningful responses for PQ and LEQ, respectively. This indicates the extent of impact ANSD has on various domains of an individual's life. It also shows that incorporating an open-ended strategy is quite valuable when trying to learn about how ANSD affects individuals. The results showed a significant difference between the total number of responses obtained for the two open-ended questions. There was no evident correlation between the audiological variables and the responses obtained, which indicates there can be other determinants associated with ANSD.

The audiological parameters considered for the correlation analysis were pure tone average and speech identification scores in both ears and the PQ and LEQ responses. The pure tone averages among the participants varied from Normal hearing to Profound Hearing loss. This variability in the PTA of individuals with ANSD had been reported in previous studies (Norrix & Velenovsky, 2014; Starr et al., 1996, 2000). 11 out of 30 participants had an SIS of 0% in both the ears and another 11 participants had an SIS of less than 50% in either one or both of their ears. These findings are on par with the previous studies (Rance et al., 2004, 2007, 2010, 2012; Zeng et al., 2005). The results from the Spearman correlation analysis indicate no statistically significant correlation between the audiological parameters and the responses to open-ended questions, which points to other confounding variables that can have a toll on the lives of individuals with ANSD.

When comparing the responses obtained for the open-ended questions, LEQ, the life effect question, marks the highest number of responses, and the difference was found to be statistically significant. This clearly suggests the impact on the day-to-day life of individuals with ANSD. 180 out of 305 responses, which is nearly 60% of the overall responses, were accounted under the Life effect question. Although Hearing-related problems were reported in individuals with ANSD, the consequences of hearing problems affect those individuals to a major extent. Sonawane & Nandurkar, (2017) reported in their study that many covert aspects take a toll on the day-to-day functioning of ANSD individuals in the dimensions of social, emotional, psychological, and economic aspects.

Now, considering responses in each domain of ICF classification, the most frequent responses were from the domain of Activity limitation and Participation restriction, which comes under part 1 of ICF, Functioning, and Disability. More than half of the overall responses were accounted from this domain. This domain mainly deals with the action or task to be carried out by an individual and the restrictions they face in participating in many life situations. Thus, it shows the intensity of challenges the individuals with ANSD face in their day-to-day life. The most frequently reported response under this domain was “Communicating with - receiving - spoken messages” (d310).

Similarly, “Socializing” (d9205) and “Using telecommunication devices” (d3600) were the other most frequently reported areas that were affected.

"Education, other specified and unspecified" (d839), carrying out conversation" (d350), and attending different "ceremonies" (d9102) were also reported by a significant number of participants. "Seeking employment, Problems related to "remunerative employment" (d850) are also affected in these individuals. Many individuals with ANSD reported having normal hearing, although they face difficulty in understanding speech, thus affecting their communication with others. This, in turn, affects their educational and employment areas of life, where interaction with people becomes inevitable. This affects their quality of life as a whole, wherein they are bound to limited interactions with people and unable to progress in life due to a lack of education and seeking employment.

The second most affected domain was impairments of body functions, where a total of 112 responses were obtained. The most reported response in the body function domain was "Emotional functions" (b152), where many individuals reported a range of difficulties they face in regulating various emotions such as sadness, fear, happiness, anger, anxiety, tension, etc. "Hearing functions" (b230), "Speech discrimination" (b2301), "Tinnitus" (b2400), and "Dizziness" (b2401) are the other major functions affected in individuals with ANSD as reported in this study. These findings were on par with the literature exploring the incidence of tinnitus and vestibular issues in ANSD (Chandan et al., 2013; Kumar & Jayaram, 2009; Prabhu, 2019; Prabhu & Jamuar, 2017).

Under the domain of Environmental Factors, "Attitudes, other specified" (e498) was reported to have a significant effect. The participants reported negative attitudes of people around them, including their immediate family members, friends, neighbors, etc. These views have a greater potential to affect the individual's behavior and social life on all scales, from close friendships and neighborhood associations to political, economic, and judicial systems. Especially, there were a substantial number of responses from "Individual attitudes of immediate family members" (e425) and "Individual attitudes of acquaintances, peers, colleagues, neighbors and community members" (e425) specifically. Based on these findings assessing these domains and including family members and others during the treatment will be a better option to overcome these hurdles. Grenness et al., 2016, with the help of a case study, explained how audiologists could implement patient and family-centered care when working with older adults and their significant others.

Other responses noted from this domain included complaints related to "General products and technology for communication" (e1250). This was majorly reported by those participants who had already made an effort to use hearing aids and other amplification systems, which became inefficacious and started to have a deleterious effect on them. Different studies have explored the limitations posed by conventional hearing aids in this population (Giraudet & Avan, 2012; Narne & Vanaja, 2008; Raveh et al., 2007).

Other than these major domains, four personal factors were reported in response to LEQ. All these four responses were regarding their future. Apart from these, there was one non-codable response. Altogether, the problems and life effects listed by individuals with ANSD were diverse.

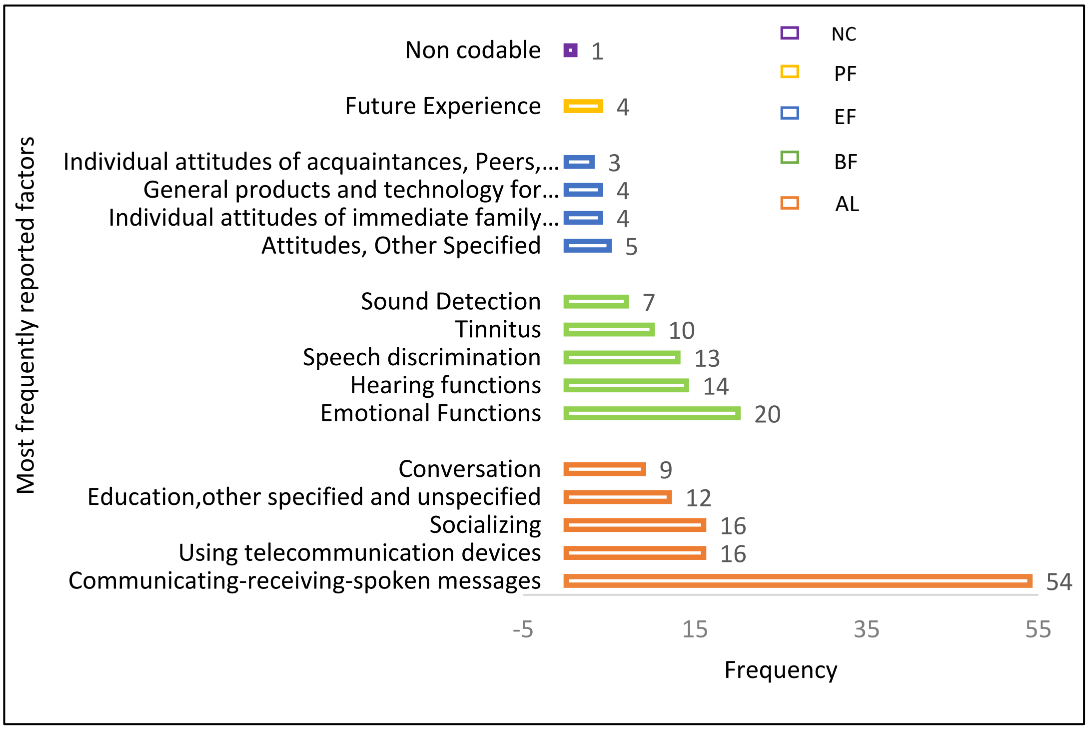
Chapter 6

SUMMARY AND CONCLUSIONS

The current study explored the problems and life effects experienced by people with ANSD. There was a significant difference in the total number of responses for PQ and LEQ. The most affected domain is the Activity limitation and Participation restriction, where "Communicating with - receiving - spoken messages" (d310) were the most occurring response. This was followed by the domain of Impairments to Body functions, where “Emotional functions” (b152) posed the greater trouble. “Hearing functions” (b230) and “Speech discrimination” (b2301) were also reported to be affected. The most frequently occurring responses related to environmental factors were “Attitudes, other specified” (e498) and "Individual attitudes of immediate family members" (e410). Furthermore, there were responses related to personal factors and a non-codable factor. The most frequently occurring responses are given in Figure. 6.1.

Figure 6.1

The Bar Graph Depicting the Most Frequently Reported Factors Impacted Due to ANSD in All Domains.



Note. NC-Non Codable factors, PF- Personal Factor, EF- Environmental Factor, BF- Body Function, AL- Activity Limitation Participation Restriction

The PQ and LEQ were also found to be not correlated with the audiological parameters such as Pure tone average and Speech Identification Scores. This indicates the need to recognize the additional perplexing factors linked to ANSD. It is apparent from the findings that the open-ended questions were instrumental in obtaining the necessary information with a diverse set of concerns. Based on these results, future audiological evaluation methods for Auditory neuropathy spectrum disorder might focus on these critical areas in addition to the audiological test battery to get better results.

6.1 Implications of the study

Understanding the difficulties faced by individuals with Auditory neuropathy spectrum disorder using a biopsychosocial approach will be an eye opener, especially to the clinicians, family members of these individuals, etc. From the clinician's point of view, the assessment procedures should focus on each and every aspect of a person affected by a particular problem or a condition. Based on the finding from the present study, it is clear that the people with Auditory neuropathy spectrum disorder are not experiencing the issues in one domain but in all the domains (e.g., Activity limitation and participation restriction - Communicating with - receiving - spoken messages, Communication devices Body function – emotional functions, Hearing functions; and Environmental factors - Individual attitudes of immediate family members", "extended family members" and "friends"). Therefore, the ICF framework can and should be used to guide audiology services for adults with hearing loss (Grenness et al., 2016), including those with ANSD. Considering all the domains specific to the problems reported by individuals with ANSD as a response to these open-ended questionnaires, a more elaborate questionnaire can be developed and validated which can alleviate the limitations of the existing questionnaires.

Rehabilitating ANSD individuals is still unsuccessful in majority of the cases (Giraudet & Avan, 2012; Narne & Vanaja, 2008; Raveh et al., 2007) since it is not only the audibility that is affected in these individuals. Moreover, they face difficulties in speech understanding and other psychosocial aspects that vary at different levels, which is evident from the study. Thus, there is a need for an individualized holistic treatment plan, including counseling sessions that can address their problems specific to different domains.

ANSD is a multifaceted condition with many variations, especially in terms of the degree of hearing loss. However, more or less, the speech understanding issues they face is a common symptom. And from this present study as well as from previous studies, it is evident that they face a lot more issues other than in the hearing domain (Parthasarathy & Shetty, 2021; Prabhu, 2017b, 2019; Sonawane & Nandurkar, 2017) which can limit their participation in the society. In spite of that, these individuals are not eligible for government beneficiaries or Disability compensation if they don't have the stipulated degree of hearing loss. Addressing the eligibility of individuals with ANSD for such provisions with respect to their difficulties in terms of speech identification scores would be beneficial, especially to those who are not able to continue their education and finding difficulty in seeking and maintaining jobs.

6.2 Strengths and limitations of the Study

Incorporating open-ended questions itself is the major strength of this study which have the benefit of covering more topics than structured ones. Especially from the results, it is evident that the responses were arising from a variety of domains, which will be restricted in a conventional interview-based questionnaire. Also, the subjects were given multiple options to submit their responses, which helped bring the responses in a much fine way. Considering the aim of this study, this approach was very relevant and could yield a high number of responses. Moreover, ICF coding provided high reliability since it is an internationally recognized classification system. Including three coders and analyzing their intercoder reliability makes this study more valid.

The method of data collection also included online mode as well as telephonic interviews. In certain cases, the data were collected with the help of the immediate family members or friends of the individuals who faced difficulty understanding speech over the phone. This could have affected the responses from these individuals to a certain extent.

Although this study is a preliminary attempt to understand the psychosocial impact of ANSD using the standard ICF guidelines, the low sample size of the study limits its generalization. Hence future studies might address this shortcoming by including more participants.

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