AUDIOLOGICAL AND LANGUAGE OUTCOMES IN CHILDREN USING HEARING AIDS: A RETROSPECTIVE COMPARATIVE STUDY OF ANSD AND COCHLEAR HEARING LOSS

Apoorva prathibha K S

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CERTIFICATE

This is to certify that this dissertation entitled **'Audiological and language outcomes in Children using hearing aids: A retrospective comparative study of ANSD and Cochlear hearing loss '** is a bonafide work submitted in part fulfilment for degree of Master of Science (Audiology) of the student Registration Number: **19AUD009**. This has been carried out under the guidance of faculty of the institute and has not been submitted earlier to any other University for the award of any other Diploma or Degree.

Dr. M. Pushpavathi

Director

All India Institute of Speech and Hearing

Manasagangothri, Mysuru-570006

Mysuru

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Dr. K V Nisha

Guide

Scientist B, Department of Audiology

All India Institute of Speech and Hearing

Manasagangothri, Mysuru-570006

Dr Ajith Kumar U

Co guide

Professor in Audiology, Department of Audiology

All India Institute of Speech and Hearing

Manasagangothri, Mysuru-570006

DECLARATION

This is to certify that this dissertation entitled **'Audiological and language outcomes in Children using hearing aids: A retrospective comparative study of ANSD and Cochlear hearing loss'** is the result of my own study under the guidance of Dr. K V Nisha, Scientist B, Department of Audiology, All India Institute of Speech and Hearing, Mysuru, and co-guidance of Dr Ajith Kumar U, Professor in Audiology, Department of Audiology, All India Institute of Speech and Hearing, Mysuru and has not been submitted earlier to any other University for the award of any other Diploma or Degree.

Mysuru **Registration No: 19AUD009**

September, 2021

DEDICATED

TO LORD GANESHA

AND

TO MY FAMILY

who deserve endless gratitude,

I'm grateful to my parents whose constant love and support keeps me motivated and confident.

Deepest thanks to my sibling, Sudeepthi who keeps me grounded, reminds me of what is important in life, and is always supportive of my adventures.

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ABSTRACT

The current study aimed to compare the hearing aid (HA) outcomes and speech and language outcomes in children with Auditory neuropathy spectrum disorder (ANSD) and Sensorineural hearing loss (SNHL). The retrospective data collected from medical reports of 24 participants, aged 1-4.9y was categorized into two groups based on the patho-physiology of the deficit: ANSD (n = 12, mean age:2.43±1.21 SD) and SNHL (n = 12, mean age:2.43±1.21y SD). The group equivalency in HA benefit (aided hearing aid scores within speech banana) and audiological data (PTA, degree of HL, Symmetry, SRT, ABR, OAEs, CM, aided and unaided free-field thresholds, and language scores) at 3 evaluations (baseline, follow up-1, & follow up-2) was considered preliminary criteria for inclusion of both groups. The hearing aid benefit (functional gain) was quantified as difference between free-field unaided and aided thresholds obtained at three evaluations for 4 frequencies (500, 1000, 2000 and 4000 Hz), while speech and language outcome was quantified as difference of chronological and language age on the three measures [receptive (RLA) and expressive (ELA) language age, combined (C-LA) language age]. For hearing aid benefit, the results of two-way repeated measure ANOVA revealed significant main effect of groups, frequencies and significant interaction effects between them. Follow up test t-test revealed functional gain of hearing aid in SNHL group was significantly greater than ANSD group at all frequencies in the baseline, while the similar group differences confined to only 500 Hz at follow up 1. No group differences in functional gain were seen in follow up 2. This disparity in hearing aid benefit (despite group equivalency in age, degree and configuration) in baselines but not follow-up can be traced to the acclimatization differences between groups, with ANSD group requiring greater acclimatization period for the realization of hearing aid benefits.

With respect to speech and language outcomes, repeated measure revealed significant main effect of timeline and language score and significant interaction effects. On post-Hoc independent t-tests, significantly higher language scores on all measures (RLA, ELA & C-LA) were observed for SNHL compared to ANSD at baseline. In contrast, at follow up 1 and follow up 2, SNHL had significantly better language scores over ANSD only on C-LA as the test SECS considers not only verbal reception and expression, but also non-verbal part of it. Although both the groups have equivalent functional hearing aid benefit, the differences seen in C-LA at the follow up 2 implies non-verbal language perception advantage of SNHL children, despite similar hearing aid acclimatization impact on verbal language reception (RLA) and expression (ELA).

Keywords: Auditory neuropathy spectrum disorder, cochlear hearing loss, hearing aids, functional gain, speech and language development.

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Chapter 1

INTRODUCTION

Auditory neuropathy spectrum disorder (ANSD) is a disorder that alerts how auditory inputs are processed in the brain (Sharma & Cardon, 2015). Patients with this condition can respond adequately to sounds, but their capacity to understand speech is impaired. ANSD is a relatively new condition. Clinical researchers began to characterise groups of individuals with normal or slightly increased audiogram pure tone thresholds but no or significantly aberrant auditory brainstem responses in the late 1970s. These patients were confirmed to have normal cochlear function when the OAEs were discovered in the mid-1980s. ANSD was first defined in 1996 as the presence of normal cochlear function but impaired brainstem responses.

The incidence and prevalence of ANSD is difficult to determine because it depends on the breadth of the inclusion criteria. Approximately 1-3 infants per 10,000 births have ANSD (Dolphin, 2004) and shows no gender preferences also (Starr et al., 2000). Kumar & Jayaram (2006) estimated the prevalence of auditory dys-synchrony in mysore, Karnataka. It was found that around 1 in 183 (0.54%) in individuals with sensori-neural hearing loss (SNHL), around 60% of the individuals had no measurable speech identification scores. The female to male ration of auditory dys-synchrony was 2:1. Sininger (2002) estimated that ANSD occurs in about 1 in 10 children with permanent hearing loss(Ching et al., 2013). Berlin, et al.(2010) reported onset of ANSD before 4 years of age in more than 50% of all cases of ANSD and bilateral ANSD in 92% of the affected.

Literature has not only highlighted the vast prevalence of this disorder, but also points out at the diagnostic hallmarks of the disorder. The diagnostic hallmark of ANSD is the combination of normal activity of the outer hair cell and abnormal afferent and efferent auditory neural functions probably at the level of the eighth cranial nerve and brainstem. In terms of the audiological correlates, outer hair cell activation is assessed indirectly by the acoustic energy emitted by the inner ear (otoacoustic emissions, OAEs) and the electrical response from the cochlea (cochlear microphonics, CM). The type of VIII nerve abnormality present in these patients is not known. The disorder could be at the level of the inner hair cells, the synapse between inner hair cells and VIII nerve fibres, the ganglion neurons, the nerve fibres or any combination of the above, which are inferred audiologically using auditory brainstem responses (ABR). The outer hair cells in the cochlea are presumed to be normal, based on the finding of normal OAEs. The status of the inner hair cells cannot be assessed as there are no procedures currently available for this purpose. Prieve et al. (1991) have suggested the possibility of a specific disturbance of inner hair cells in a patient with a severe hearing loss and preserved otoacoustic emissions.

Given such variability in the pathophysiology of ANSD, amplification and auditory rehabilitation for these patients is challenging. Although the prevalence rates of ANSD in children have been succinctly reported in literature, Audiologists are still seeking effective treatments to rehabilitate this population. There are three current forms of hearing technology that are recommended as intervention for children with ANSD: hearing aids, cochlear implants (CI), and use of frequency modulation (FM) systems. Considerable heterogeneity is reported with respect to benefit derived from these hearing rehabilitation options is available for individuals with ANSD (Roush et al., 2011).

Hearing aids (HA) with or without the FM system should be first stage of management. A trial of HA amplification should be performed depending on the audiological information. Of those studies that explored HA benefit in children, poor or negligible effect was seen on outcome measures (Berlin et al., 1999; Berlin et al., 2003; Raveh et al., 2007). This leads us to the argument that HAs merely amplify sound, which cannot suffice the distortion occurring at the neural level in ANSD children. These findings have led to general uncertainty as to whether or not children with ANSD should utilize HAs, but there are some limitations to applying this line of research for children who are have hearing impairment secondary to cochlear lesion. No tests are currently available, however, to determine whether an individual with ANSD might benefit from a HA or not. The use of FM system is also recommended in both ANSD patients with normal hearing and HA and/or CI because of the improved speech understanding in noisy conditions. In addition, the cochlear implantation candidacy is also determined in ANSD (if need be) based on the audiological findings, HA benefit, status of speech language development and overall development skills.

Out of the several research strides in the rehabilitating ANSD deficits, Yuvaraj and Jayaram (2016) analyzed the benefit from HAs for adults (aged 16-30 years) with ANSD. The authors checked the association between benefit from HAs and auditory profile. The two groups classified in the study based on HA benefit (who benefitted and those who did not), were also found to be significantly different from each other in their mean pure tone thresholds (PTA) too. Those who benefitted from hearing aid had significantly poorer thresholds ($p < 0.05$) than those who did not benefit. This reflects the relationship between the PTA threshold and HA benefits in augmenting sound audibility for adults with ANSD. In persons with poorer hearing thresholds, augmentation of the signal through HAs may help in the detection of the signal. However, this study was done on a fewer sample size i.e, 38 individuals and also did not try to correlate other audiological measures to predict the HA outcomes in the ANSD group. On other hand, Yuvaraj and Jayaram (2016) reported that mean speech identification scores of those who benefitted from HAs did not vary from those who did not, implying that benefit from HA and speech identification scores are not associated, as expected in ANSD population. In contrast, Starr et al. (Starr et al., 1996) reported that HAs were found to be less beneficial in both children and adults with ANSD. Use of CIs in children with ANSD, though found to be largely beneficial, has limitations such as the lack of electrical induced neural synchronization, the detrimental effects of other associated conditions, or combination of both (Teagle et al., 2010). Hence, the authors instead of recommending CI for all children with electrophysiological evidence of ANSD, the stepwise procedure including comprehensive profiling would allow for the identification of children who benefit from amplification before CI is fitted.

The lack of consensus on utility of hearing aids and CI as rehabilitative options in ANSD can be attributed to the inherent nature of the disorder. Contrasting this with rehabilitation options for children with cochlear sensorineural hearing loss (SNHL), it can be hypothesized that this population would relatively enjoy higher benefits from HAs as it compensates for the defective functioning of outer hair cells by amplification. Plethora of research strides document the effectiveness of HAs as rehabilitative options in children with SNHL (Berlin et al., 2010; Roush et al., 2011). However, we currently lack evidence-based protocols for using behavioral observations to fit HAs in a clinically valid manner for children with ANSD. In summary, we know little about children with ANSD who have disabling hearing loss, have been early identified and have been fit using best practice recommendations, in comparison to children with similar degrees of SNHL.

Need for the Study

Given the overwhelming clinical variability of ANSD individuals on audiological tests and variability in the conventional HA outcomes in this population, there is a strong need to profile the audiological measures and rehabilitative outcomes in children with ANSD. The current study addressed this need of profiling the audiological and speech and language outcomes in ANSD and compared them with the children having cochlear-SNHL. Further, the study also addressed the need of monitoring the established outcomes at regular follow-up intervals.

1.2. Aim of the study.

The study aimed to compare the audiological and speech and language outcomes measures in ANSD and compared them with the cochlear-SNHL children using hearing aids at different timelines (baseline and two follow-up sessions). The study also aimed to identify and classify audiological profiles of ANSD and cochlear-SNHL to best determine the factors that can predict HA outcomes in the two groups.

1.3. Objectives.

The specific objectives of the study are-

1. To retrospectively compare the HA benefit outcomes (functional gain) in children with ANSD and cochlear-SNHL, fitted with HA across evaluations (baseline, follow up 1, follow up 2).
2. To retrospectively compare the speech and language outcomes (receptive language age: RLA, expressive language age: ELA & combined language age: C-LA) in children with ANSD and SNHL fitted with HAs, across evaluations (baseline, follow up 1, follow up 2).
3. To correlate the HA benefit outcomes with speech and language outcomes in each group separately across evaluations (baseline, follow up 1, follow up 2).
4. To determine the audiological measures which best predict the group differences (ANSD and SNHL) in HA and language outcomes across evaluations (baseline, follow up 1, follow up 2).

Chapter 2

REVIEW OF LITERATURE

2.1. Definitions:

Auditory neuropathy spectrum disorder (ANSD) is characterized by absent or severely abnormal auditory brainstem response (ABR) with intact outer hair cell function, in the presence of evoked otoacoustic emissions and/or cochlear microphonics. Though the terminology implies dysfunction of the auditory nerve, possible causes include damage to the inner hair cells, to the synapse between the inner hair cell and auditory nerve, or in the auditory nerve itself (Roush et al., 2011). ANSD is a disorder of hearing or a condition in which a patient's otoacoustic emissions or cochlear microphonics are present with abnormal or absence of auditory brain-stem responses (Berlin et al., 2010).

'Cochlear- Sensorineural hearing loss (SNHL)' occurs when there is a dysfunction in conversion of sound waves to electrical signals by the inner ear of nerve impulses to the brain' (Mary, 2012)

2.2. Prevalence

2.2.1 ANSD:

Researchers have conducted a number of research to determine the prevalence of ANSD in diverse populations, as the prevalence rate varies depending on the community analysed. The prevalence of ANSD has been found to reach 40% among newborns at high risk for hearing loss, particularly in babies kept in neonatal intensive care units (Rea & Gibson, 2003). According to studies that looked at newborns with profound hearing loss, the prevalence of ANSD was as high as 5.26 % (Talaat et al., 2009), 13.4 % (Domínguez et al., 2007), and 19 % (Maris et al., 2011). The prevalence rate of ANSD in babies is extremely high when compared to school children. Within the age range of 6–12 years, the prevalence rate of ANSD in deaf school students is 2.46 % (Lee et al., 2001).

2.2.2 SNHL:

In a study conducted by Naeem & Newton (1996), the prevalence of hearing loss in Asian children was found to be between 5.09 and 9.61 per 1000, compared to 1.4 to 3.51 per 1000 in non-Asian children, and the relative risk measure revealed that the Asian group was 2.42–3.61 times more likely to have a hearing loss. However these findings are for both cochlear and neural types.

2.3. Etiology:

2.3.1 ANSD

ANSD can be caused due to both genetic and acquired factors. Genetic can be both syndromic (group of disorders) and non-syndromic (isolated). Faulty coding of genes DFNB9, DFNB59 and AUNA1 leads to mutation causing ANSD due to non-syndromic cause & examples for syndromic types are Charcot-Marie-Tooth/ Friedreich's ataxia.

Acquired causes mainly occur during perinatal period disturbances but can also occur in later stages of life. The early acquired ANSD are caused due to hypoxia, hyperbilirubinemia and prematurity, whereas the late acquired would be in the childhood or in adolescence which basically is due to genetics. (Norrix & Velenovsky, 2014)

2.3.2 Cochlear- SNHL:

Cochlear- SNHL is caused by any injury to the end organ of hearing i..e cochlea. Degenerative processes linked with age, which decrease the functioning of the cochlea, long-term exposure to loud sounds, therapeutic drug consumption with ototoxic side effects, and chronic diseases are the main causes of in Cochlear- SNHL in adults. However, in children, genetic abnormalities can be the primary cause of SNHL, which can include both syndromic and non-syndromic hearing loss, as well as congenital deformities, inner ear infections, and head trauma. Hereditary hearing loss is the most frequent among infants, affecting around 1 in 1000 live births.

2.4. Audiological test battery:

The audiological characteristics of ANSD and cochlear-SNHL groups, mentioned in the literature (Madden et al., 2002) are outlined in Table 2.1.

Table 2.1.

Comparison of the audiological characteristics between the ANSD and Cochlear-SNHI groups

	Tests/ Particulars	ANSD	Cochlear- SNHL
1	Audiometry		
	Degree	Normal to profound	Mild to profound
	Configuration	Rising pattern (LFHL)	Falling (HFHL)
2	Correlation with PTA	No correlation	Good correlation
3	SIS Scores	Irrespective of degree, poor scores	Greater the loss, poorer the performance
4	SPIN scores	Very poor (< 60 %)	Greater the loss, poorer the performance
5	Acoustic reflexes	Always absent	Mild (Present); > moderate (Absent)
6	OAEs	Present (robust)	Mild (Present), Moderate (present in DP)

			> moderate (Absent)
7	CM	Present	Absent
8	ABR	Absent	Absent response for severe and profound degree of HL
9	LLR	May or may not be present	Usually Present

2.5. Rehabilitation options:

Currently, three forms of hearing technologies that are recommended— hearing aids (HAs), FM devices and cochlear implantation (CI)—for children with ANSD. Not surprisingly, there has been considerable debate among clinicians and investigators regarding the choice of and expected benefit from these technologies.

2.5.1 Hearing aids and Auditory Neuropathy Spectrum Disorder

Walker et al., (2016) compared the speech production, speech perception, and language outcomes of children with ANSD with hearing loss and children with similar degrees of mild to moderately severe SNHL who are fitted with binaural HAs. They reported no significant differences between the groups on language measures. In quiet, children with ANSD demonstrated functional speech perception skills. There was also a trend that the ANSD group fared worse in background noise with hearing aids than the SNHL group.

2.5.2. Cochlear implantation in Auditory neuropathy spectrum disorder

Opponents of traditional amplification point to the alleged limited benefits of HA use for people with normal outer hair cell function, claiming that acoustic amplification just provides louder, more distorted signals (Trautwein et al., 2000). Researchers are concerned about the possible effects of acoustic damage on existing outer hair cells (Roush et al., 2011). Concerns have also been raised about the suitability of CIs for children with ANSD due to reports of auditory nerve involvement (Miyamoto et al., 1999).

Others have expressed reservations about cochlear implantation, citing reports of spontaneous improvements in hearing thresholds in a subset of children, particularly those with hyper bilirubinaemia (Madden et al., 2002), which is a common cause for ANSD in children. Electrical stimulation of the auditory nerve by a CI may increase neuronal activity synchronisation.

Improved audiological performance, good implants evoked brainstem responses, and satisfactory neural response telemetry (NRT) after implantation in ANSD children has been documented in literature (Katada et al., 2004). The improvement in audiological performance in ANSD is shown to be directly related to the intervention age. A recent study Cardon and Sharma (2013) using long latency response (LLR) found a significant difference in auditory cortical maturation between ANSD patients who received CI before and after the age of two years. In over 70% of cases, the former resulted in a normal P1 component, whereas the latter resulted in delayed P1 latencies in the same proportion (Cardon & Sharma, 2013). The presence of P1 component, can be utilised to track cortical maturation, predict speech perception ability and rehabilitative benefits in ANSD children (Sharma et al., 2002). In contrary, Rance and Barker (2009) reported no significant differences in the speech and language outcomes in 12 ANSD children using CI compared to their age matched SNHL group.

2.5.3.Hearing aids in sensorineural hearing impairment

For children with cochlear-SNHL, HA fitting is a standard of care. The improved audibility provided by HA use would give children most opportunities to successfully perceive speech and thus learn their community's speech and language patterns. A study by Tomblin et al.(2014) found that providing HAs to children with mild to severe HL at a young age is likely to result in better speech and language development, especially if the child receives good audibility from the HAs and has had more time to wear the HA. The advantages of HA manifested also applied to children with mild HL (Tomblin et al., 2015; Walker et al., 2016). In addition, Tomblin (2014) also cautioned that quality of the HA will determine the degree of audibility, and the benefits derived henceforth in SNHL children.

2.5.4. Cochlear implantation in sensori neural hearing impairment

For children with profound deafness to hear and acquire speech understanding, paediatric cochlear implantation is clearly essential. Up to ten years following cochlear implantation, children with cochlear implants have demonstrated hearing performance and language acquisition comparable to normally hearing children, with word recognition scores of 85 % in the best implant users (Peixoto et al., 2013). 75% of implanted children are in mainstream education by the time they reach high school, with only 5% requiring full-time educational support and customised educational settings. When it came to speech results in children with substantial SNHL, those who had implants outperformed those who had regular hearing aids (Sharma et al., 2020).

2.6. Understanding Variability and Prognosis with rehabilitative options in ANSD.

One way of understanding the variability in rehabilitative outcomes can be attributed to the very nature of ANSD disorder, with this population exhibiting a wide range of clinical manifestations, prognoses, and underlying etiologies associated with the disorder (Feiern et al., 2013). Acceptance of amplification devices among this population is very low because of its minimal efficacy. But there are many studies which have shown cochlear implantation to be beneficial but in contrast recent researchers' state that the benefit from the cochlear implantation or other amplification devices will solely depend on the site of lesion.

The current management of individuals presenting with ANSD varies according to the severity of the impairment. However, management remains challenging and is frequently tailored case-by-case. It is based on bottom-up (auditory skills restoration by hearing aids) and top-down procedures (hearing and speech training). Generally, a multidisciplinary approach is favoured.

Chapter 3

METHODS

3.1. Study design.

Retrospective standard group comparison, involving both within and across group comparisons (Schiavetti & Metz, 2006).

3.2. Participants.

Retrospective data of 24 participants, aged 1- 4.9 y with mild to profound degree of hearing loss based on the 4-frequency pure-tone average (Mazzoli et al., 2003) were considered for the study. The data collection for profiling was done retrospectively by studying the case reports from the medical records section, department of clinical services (DCS), AIISH, Mysore. The data was gathered in a retrospective longitudinal design with the entries of minimum of two follow ups in a span of 2 years.

The participants were divided into two groups based on the pathophysiology of the deficit: Auditory neuropathy spectrum disorder, ANSD (n = 12, mean age:2.43±1.21 SD, 7 males, 5 females) and cochlear- Sensorineural hearing loss, SNHL (n = 12, mean age:2.43±1.21y SD, 9 males, 3 females). The criteria adopted to diagnose ANSD are according to the recommendations of Starr et al (2000), including preserved cochlear amplification, reflected by the presence of transient evoked oto-acoustic emissions and/or the presence of cochlear microphonics, altered auditory nerve responses as indicated by absent or severely abnormal auditory brainstem responses, and normal otological and tympanometric findings with absent acoustic reflexes. On other hand, age and hearing degree matched children were considered in cochlear-SNHL group.

All the participants considered in the study were previously fitted with the digital hearing aid (HA), by a Rehabilitation Council of India (RCI) certified audiologist. The case files of the selected participants had similar features of the HA such as number of channels (8 channels), prescription formula (NAL-NL1), noise cancellation, feedback suppression, directionality, and volume control (deactivated). The audibility of sounds from the HA was ensured retrospectively by analysing aided thresholds, which fell in the speech banana range. Further, all the participants had atleast two visits to AIISH and reported of no other comorbid conditions such as secondary neuropathies, cognitive impairments or other developmental disabilities (as ruled out during the case file scrutiny process).

3.3. Ethical guidelines.

The study adhered to the 'Ethical guidelines for bio-behavioral research involving human subjects' and ethical committee approval was obtained (Venkatesan & Basavaraj, 2009). The demographic details and medical history of all the participants in two groups are shown in Table 3.1.

Table 3.1.

Demographic details and basic audiological characteristics of the participants in two groups. Mean values are mentioned along with one standard deviation in parentheses.

Demographic details	ANSD	SNHL
Age in years	2.43±1.21	2.43±1.21
Gender		
No. of males	7	9
No. of females	5	3
Onset of the problem	Early	Early
Risk factors /Significant medical history (if any)	Nil	Nil

3.4 Procedure.

The retrospective data stored in case files were segregated under three evaluations/phases: baseline, follow-up 1 and follow up 2 evaluations. The auditory (HA benefit) and speech and language outcomes (language scores) were enumerated in each phase, along with the participants audiological profiles. Medical records of all the children diagnosed with ANSD in the age range of 1 to 5y were scrutinised for the year 2010 to 2018. Out of the 65 files which matched the age and diagnosis criteria, only 12 fulfilled the inclusion and exclusion criteria (see section 3.2), considered in the study. The medical records for SNHL participants, who were age- and degree-matched to the participants in the ANSD group were also scrutinised, and 12 files which satisfied the matched-criteria were included. Table 3.3. shows the age of the participant at different phases of data collection.

Table 3.2.

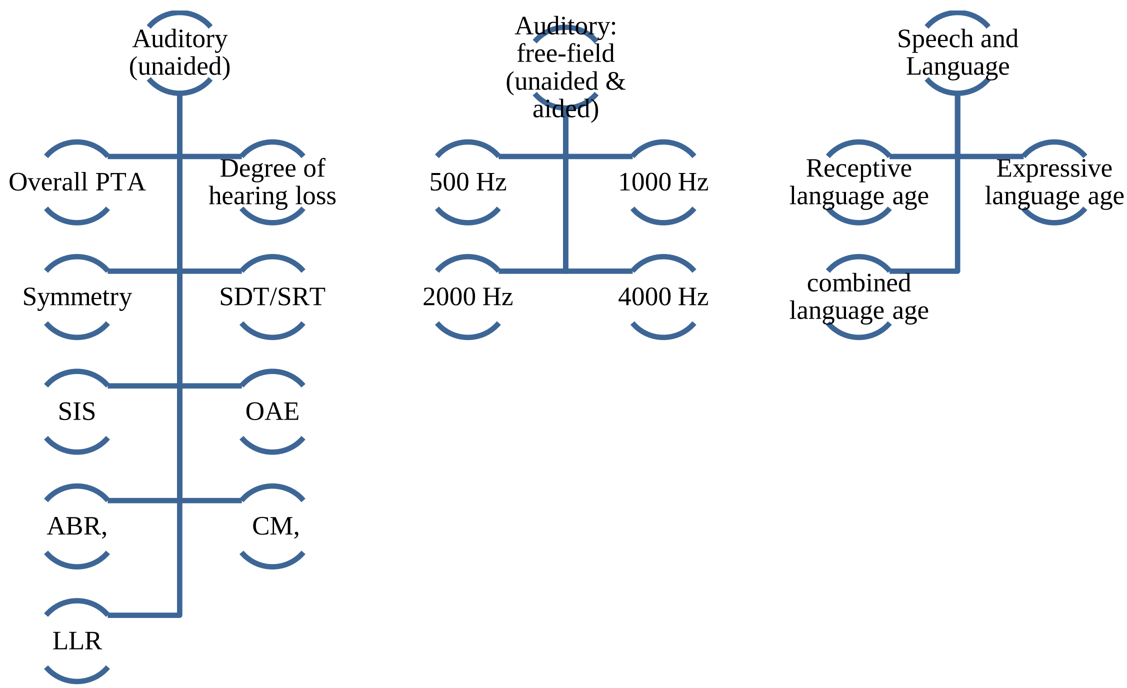
The age at time of evaluation of children with ANSD and cochlear-SNHL at baseline, follow-ups 1 and follow-up 2.

Participants	Age at the time of data extraction/evaluation (in y)					
	Baseline		Follow up 1		Follow up 2	
	ANSD	SNHL	ANSD	SNHL	ANSD	SNHL
1.	4	3.6	4.8	3.6	5.2	4.6
2.	1.6	1.6	2	2.1	2.8	3
3.	3.6	2.0	4.4	3.8	5	4.8
4.	1.6	1.5	2.2	2.2	3.2	3
5.	4.9	1.6	5.8	5	6.2	5.8
6.	1.5	1.9	2.2	2.3	3	2.11
7.	1.6	4	5.1	5.2	5.7	5.9
8.	1	1.11	1.8	2.1	2.7	3
9.	4.6	3.2	4.9	3.10	5.3	4.9
10.	1.4	1,5	2	2.6	2.11	3.2
11.	1.6	2.1	2.2	2.7	3	3.4
12.	4	3	4.2	3.8	5	4.9

3.4.1. Phase I: *Baseline assessment.* Phase 1 involved elucidation of the data obtained in first visit (baseline), under the different parameters as shown in Figure 3.1.

Figure 3.1.

Schematic diagram of different parameters profiled in the three evaluations of the study.



The audiological parameters (Figure 3.1.) documented in the case file were obtained. The standard test protocol followed at Jayachamendra block in DCS, AIISH for conducting these audiological test battery is as follows:

1. Using a modified Hughson and Westlake technique (Carhart & Jerger, 1959) pure tone air conduction thresholds and bone conduction thresholds obtained from a calibrated Inventis Piano audiometer (Inventis SRL Inc.), were noted.
2. Speech identification assessed using a 40 dB SL monitored live voice presentation of phonemically balanced Kannada words or monosyllables (Rajashekhar, 1976) were noted.

3. The status of the middle ear assessed using a calibrated immitance metre, the GSI-Tympstar V (Grason-Stadler, Inc.) were noted.
4. The status of outer hair cell function inferred from otoacoustic emissions (OAE) test measured using a calibrated ILO V6 DP Echoport were noted.
5. The auditory brainstem response (ABR), long latency response (LLR), and cochlear microphonics (CM) recorded using Biological Navigator Pro (Natus Medical Inc.) or SmartEP (Intelligent Hearing Systems) coupled with an ER-3A insert receiver were also noted.

In addition to these measures, free-field aided and unaided scores across 4 frequencies (500 Hz, 1000 Hz , 2000 Hz and 4000 Hz), and the speech and language measures [RLA, ELA from REELS(Receptive expressive emergent language scale) and C-LA(Combined language age) which is the combination of CRLA and CELA from SECS was obtained]. Hearing aid benefit was measured as functional gain, which is derived from the difference of aided and unaided thresholds, while the speech and language outcomes (RLA, ELA, C-LA) were derived as difference of language age and the chronological age.

3.4.2. *Phase II: Comparison with the first follow up visit.*

The details collected at first follow visit (within 1 year from the baseline assessment, Table 3.2) of the participants were retrospectively segregated in the phase II, of the study. The details of the audiological parameters described above in Phase 1 (Figure 3.1) was collected. The aided and unaided scores across 4 frequencies (500 Hz, 1000 Hz , 2000 Hz and 4000 Hz), and the speech and language measures (receptive language age: RLA, expressive language age: ELA evaluated on receptive expressive emergent language test:

REELS and combined language age: C-LA from scales for early communication skills for hearing impaired children: SECS) was obtained. The HA benefit was measured as functional gain, which is derived from the difference of aided and unaided thresholds, while the speech and language outcomes (RLA, ELA, C-LA) were derived as difference of language age and the chronological age.

3.4.3 *Phase III*: *Comparison with second follow up visit*

The data collected in phase III was similar to Phase II, except that the reports were obtained from second follow up (conducted within 2 y from the baseline assessment) as shown in Table 3.2. The hearing aid and speech and language outcomes were also quantified, as discussed in section.

3.5 Data coding for Analyses

The data in the baseline evaluation were coded in IBM Statistical Package Social Sciences, version 25.0 (SPSS Inc, Chicago). for further analyses. Table 3.3. shows the codes used for data obtained from the three phases.

Table 3.3:

Codes and labelling of Audiological parameters profiled on SPSS for analysis

		Label	Code
1	AC thresholds	Baseline: AC R/L, FU1: AC1 R/L, FU1: AC2 R/L	numerical values
2	BC thresholds	Baseline: BC R/L FU1: BC 1 R/L FU2: BC 2 R/L	numerical values
3	Pure tone average	Baseline: PTA R/L, FU1: PTA 1 R/L,	numerical values

		FU2: PTA 2: R/L	
4	Degree	Baseline: R/L Degree, FU1: R/L1Degree, FU2: R/L2Degree	1-normal, 2-mild, 3-moderate, 4-moderately severe, 5-severe, 6-profound
5	Symmetry		1-Symmetrical, 2-Asymmetrical
6	Configuration	Baseline: Configuration RT /LT, FU1: Configuration RT 1/LT 1 FU2: Configuration RT 2/LT 2	1-flat, 2-sloping, 3-rising
7	SRT	Baseline: SRT RT /LT, FU1: SRT RT 1/LT 1 FU2: SRT RT 2/LT 2	numerical values
8	Reflex	Baseline: Reflex RT /LT, FU1: Reflex 1R/1L, FU2: Reflex 2R/2L	1-present, 2-absent
9	OAEs	Baseline: OAEs R/L FU1: OAEs1R/1L FU2: OAEs2R/2L	1-present, 2-absent
10	ABR	Baseline: ABR R/ L FU1: ABR1R/ 1L, FU2: ABR 2R/2L	1-present, 2-absent
11	CM	Baseline: CM R/ L FU1: CM1R/1L, FU2: CM2R/2L	1-present, 1-absent
12	Aided scores	AR, A1R, A2R AL, A1L, A2L	numerical values
13	Speech	RLA, RLA1, RLA2 (Baseline, F/U 1, F/U2) ELA, ELA1, ELA2 (Baseline, F/U 1, F/U2) C-LA, C-LA1, C-LA2 (Baseline, F/U 1, F/U2)	1: 0-6months(m), 2: 7-12m, 3:13-18m, 4:19-24m, 5:25-30m, 6:31-36m,7:37-42m, 8:43-48m, 9:49-54m, 10:55-60m

3.5 Statistical analyses.

The raw scores of the retrospectively collected data were subjected to statistical analysis using IBM Statistical Package Social Sciences, version 25.0 (SPSS Inc, Chicago). In order to profile the auditory charateristics of ANSD and SNHI children using HAs, descriptive statistics (mean, median, standard deviation, and interquartile) were done for all the measures (HA functional gain, RLA, ELA and C-LA outcomes). Following this, the Shapiro-Wilk test of normality was administered. The functional gain of HA were across frequencies and timelines were compared using three way- repeated measure ANOVA (4 frequencies × 3 timelines × 2 groups), while similar analyses was also done for speech and language outcomes (2 language outcomes × 3 timelines × 2 groups). This was followed up by independent t-test, wherever significant main effects were seen. Whenever significant differences were noticed, the corresponding effect size were reported.

Furthermore, Fisher's linear discriminant function analysis (FLDA) was carried out for group classification based on the auditory data of the better ear recorded in the baseline, follow-up 1 and follow-up 2 measurements from all the participants in the study. The rational for the use of better ear in audiological data, is based on the assumption that HA and speech and language outcomes will be correlated to the better ear's audiological characteristics. FLDA is a multivariate analysis technique that attempts to categorize groups based on measures obtained from the same set of variables (Bennett & Bowers, 1976). The FLDA generates a mathematical operation ($Di = a + b_1x_1 + b_2x_2 +\ldots+ b_nx_n$; Di = predicted discriminant score; a = constant, x = predictor; and b = discriminant coefficient, Lachin & Schachter (1974) based on weights generated for each test variable, that maximizes the differences in performance for each group. Based on the weights obtained in the FLDA, the best tests for group segregation were determined. In addition, the error rate for accurate group-prediction by FLDA was calculated by performing a casewise tally of predicted membership (discriminant function) from the original (pre-determined) membership. The FLDA derived group assignment was compared with the otherwise original membership of the individual to determine the overall error rate for group segregation.

Chapter 4

RESULTS

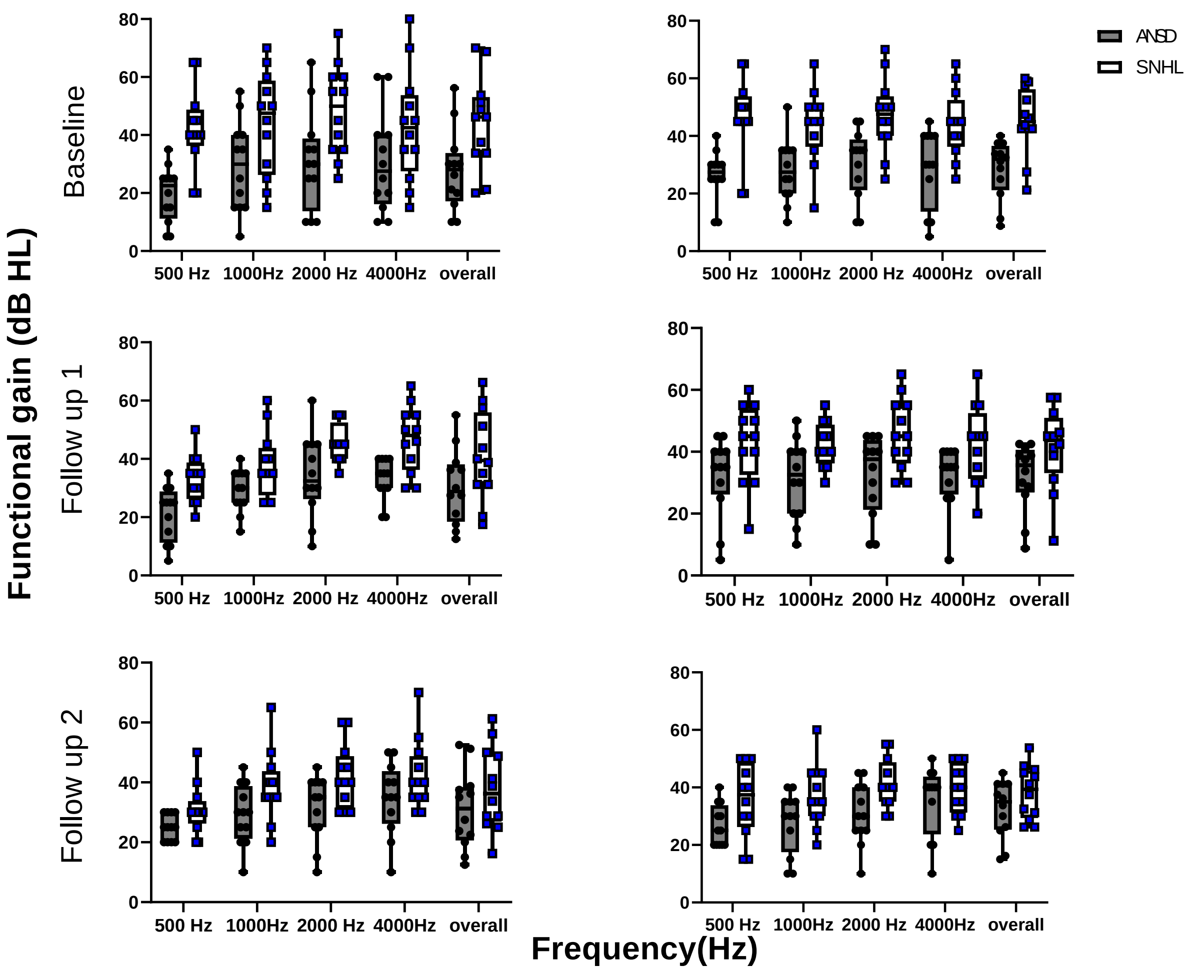
The study aimed to compare the hearing aid (HA) outcomes in children with ANSD and SNHL on functional gain and speech and language outcomes, in a retrospective time-series design. In addition the audiological predictors of hearing aid benefit were also explored in these children at three timelines (baseline, follow up 1 and follow up 2). The following sections discuss the results of the such comparisons.

4.1. Comparison of hearing aid benefit on audiological outcomes between ANSD and SNHI children.

Hearing aid benefit (unaided threshold -aided threshold) was assessed at 4 frequencies (500 Hz, 1000 Hz, 2000 Hz, & 4000 Hz) in all the phases of evaluation (baseline, follow up 1 and follow up 2) Figure 4.1. depicts the functional gain of HA at the three phases as a function of frequency. The mean functional gain in ANSD was comparatively lesser than that the mean gain of SNHL, at all the three phases indicative of poorer hearing aid benefit in ANSD children. This was seen for both right and the left ear. Although this difference was strikingly evident in the baseline, the group differences in mean was reduced at follow up I and became negligible at follow up II.

Figure 4.1

Comparison of hearing aid benefit on functional gain across frequencies between ANSD and SNHI children. The box plot in the figure indicates the spread of data, the error bar indicates one standard deviation and the individual data points in black and white corresponds to ANSD and Cochlear-SNHL.



Repeated measures ANOVA with phases, frequency, and ear as within subject factors and group as across subject factor revealed main effect of group [F(1,22) = 6.11, p = 0.02, partial eta square - η_p^2= 0.22] as shown in Table 4.1. The other main effects and the interaction effects are shown in the Table 4.1. Wherever significant effects were found, partial eta square (η_p^2) is given as a measure of effect size.

Table 4.1

Main effects and interaction effects with respect to functional HA gain.

Main and interaction Effects	F ratio	*p*	Partial eta square ($\boldsymbol{\eta_p^2}$)
Group	F(1,22)= 6.11	0.02*	0.22
Phase	F(2,44)= 0.91	0.41	
Frequency	F(3,66)= 1.57	0.21	
Ear	F(1,22)=0.01	0.92	
Group*phase	F(2,44)=7.30	0.00**	0.25
Group*frequency	F(3,66)= 0.27	0.85	
Group*Ear	F(1,22)= 2.87	0.11	
Phase*frequency	F(6,132)= 2.34	0.03*	0.10
Phase*ear	F(2,44)= 1.82	0.17	
Frequency*Ear	F(3,66)= 4.60	0.00**	0.17
Group*phase*frequency	F(6,132)= 0.44	0.85	
Phase*frequency*Ear	F(6,132)=3.06	0.01**	0.12
Ear*group* phase	F(2,44)= 3.07	0.05*	0.12
Frequency*ear*group	F(3,66)= 0.47	0.71	
Frequency*ear*group* phase	F(6,132)= 1.68	0.13	

Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

Due to the significant interaction effects between the variables observed, the follow-up independent t tests were performed, with Bonferroni correction for functional gain comparisons at each frequency (500 Hz, 1000 Hz, 2000 Hz, & 4000 Hz), for right and left ear separately. The results of the t test showed significantly greater benefit from all the frequencies for SNHL group compare to ANSD at baseline, as shown in Table 4.2. However at follow up 1 SNHL had significantly better functional gain scores only at 500Hz compared 500Hz ANSD. In contrast, no statistical differences were observed for any of the frequencies at follow up 2.

Table 4.2

Comparison of group differences in functional HA gain at baseline, follow up 1 and follow up 2 across frequencies

Frequency (Hz)	Baseline					
	Rt ear			Lt ear		
	t(22) =	*P*	Cohen's d	*t*(22) =	*p*	Cohen's d
500	-4.57	0.000***	1.86	-4.07	0.001**	1.66
1000	-2.20	0.038*	0.89	-3.25	0.004**	1.32
2000	-2.61	0.016*	1.06	-3.28	0.003**	1.34
4000	-1.67	0.10	0.64	-2.97	0.007**	1.21
	Follow – up 1					
500	-2.95	0.007**	1.20	-2.62	0.015*	1.07
1000	-1.22	0.23	0.50	-1.75	0.09	0.71
2000	-1.82	0.81	0.74	-1.86	0.07	0.76
4000	-1.23	0.23	0.50	-1.89	0.07	0.77
	Follow – up 2					
500	-1.36	0.188	0.55	-0.75	0.45	0.31

1000	-1.36	0.19	0.55	-1.60	0.12	0.65
2000	-1.26	0.22	0.51	-2.15	0.04	0.87
4000	-0.68	0.50	0.27	-0.96	0.34	0.39

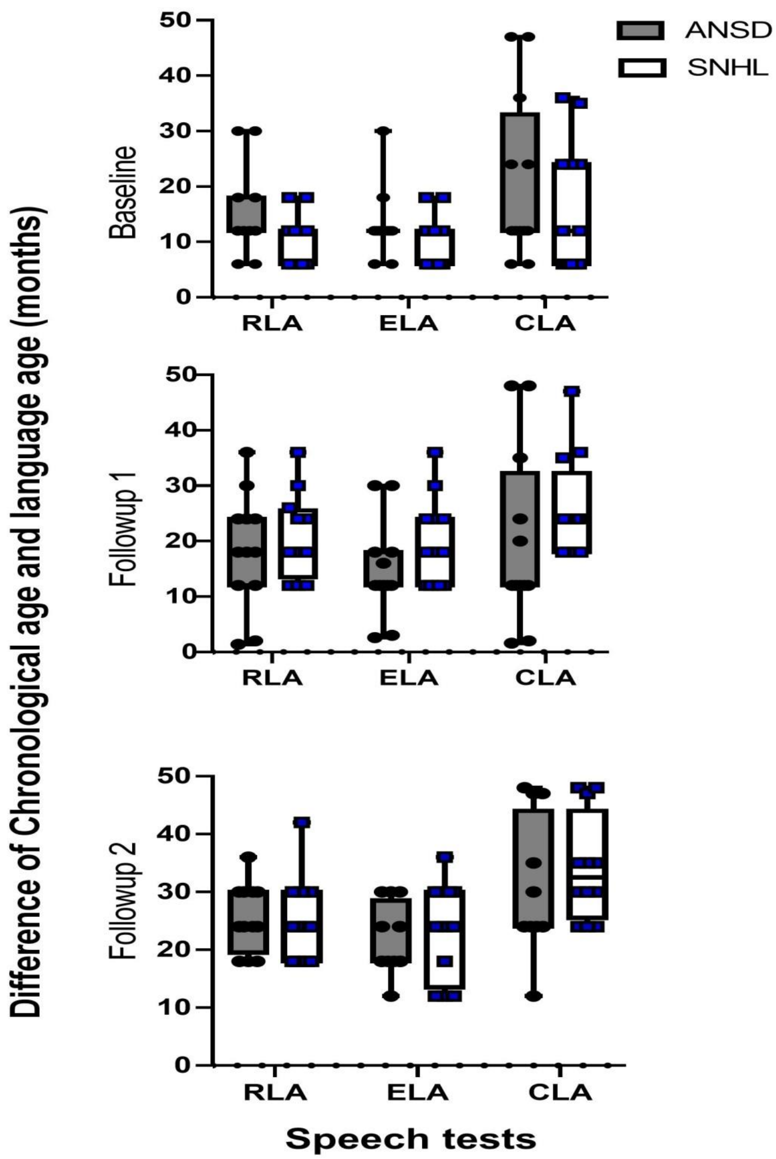
Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

4.2. Comparison of hearing aid benefit on speech and language outcomes between ANSD and SNHI children.

The graph depicting the change in language age with amplification as a function of speech test scores, is shown in Figure 4.2. Language level prognosis (unaided threshold -aided threshold) was assessed with 2 speech tests (REELS and SECS) and 3 different outcome measures (RLA, ELA, CLA) in all the phases of evaluation. Both the groups showed improvement in language scores over time. The group SNHL derived greater improvements in language compared to the group ANSD, at all the three phases. At baseline the language-based difference observed between the groups were greater compared to follow up 1. At follow up 2 the difference was very minimal.

Figure 4.2.

Comparison of hearing aid benefit on speech and language outcomes between ANSD and SNHI children. The box plot in the figure indicates the spread of data, the error bar indicates one standard deviation and the individual data points in black and white corresponds to ANSD and Cochlear-SNHL.



Repeated measures ANOVA with phase and language score as within subject factors and group as across subject factor and revealed main effect of phase [F(2,44)=16.69, p < 0.001, η_p^2= 0.27] and language score [F(2,44)= 13.35, p < 0.001, η_p^2= 0.39], as shown in Table 4.3. The other main and interaction effects, are also tabulated in the Table 4.3.

Table 4.3

Main effect and Interaction effect with respect to speech and language outcomes

Main and interaction Effects	F	*p*	Partial eta square (η_p^2)
Group	F(1,22)=0.36	0.55	
Phase	F(2,44)= 16.70	<0.001***	0.26
Language score	F(2,44)= 13.35	<0.001***	0.40
Group*Phase	F(2,44)=11.32	<0.001***	0.14
Group*language score	F(2,44)= 5.51	0.01**	
Phase*language score	F(4,88)= 1.01	0.40	
Group*Phase*language score	F(4,88)= 0.75	0.56	

Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

Due to the significant interaction independent t test was performed for each of the language outcome separately. The results of the t test are shown in the Table 4.4, which revealed significantly higher RLA, ELA, CLA for SNHL compared to ANSD at baseline. However no statistical differences were observed for RLA and ELA between the groups at follow up 1 and follow up 2. In contrast, at follow up 1 and follow up 2 SNHL had significantly better CLA scores compared to ANSD.

Table 4.4.

Comparison of language outcomes between groups at baseline, follow up 1 and follow up 2

	Baseline		
	t(22)	*p*	Cohen's d
RLA	-2.75	0.012*	0.11
ELA	-2.71	0.013*	0.02
CLA	-3.30	0.003**	0.30
	Follow up 1		
RLA	0.28	0.77	0.24
ELA	0.39	0.69	0.39
CLA	2.42	0.02*	0.63
	Follow up 2		
RLA	-0.28	0.77	0.05
ELA	-0.83	0.68	0.21
CLA	2.09	0.05*	0.61

Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

4.3 Correlation between functional gain and language outcomes

Pearson's correlation (r) analysis was done to assess the correlation between the hearing aid benefit and language scores (in months). No significant correlation between the overall functional gain of HA and language scores (in months) were seen in the ANSD group, across all the phases. In contrast, for SNHL group, a significant correlation in the between the overall functional gain of HA and C-LA was seen in baseline and follow up – 2 across phases as shown in Table 4.6.

Table 4.5

Correlation between functional gain (dB HL) and language scores (in months) across phases . Degree of correlation (Pearson co-efficient (r)) and corresponding significant values for each parameter for comparisons are shown.

		RLA		ELA		CLA	
		(r)	*p*	(r)	*p*	(r)	*p*
Baseline	ANSD	-0.15	0.48	-0.16	0.46	-0.21	0.32
	SNHL	-0.04	0.95	-0.01	0.87	-0.69	<0.001***
Follow up I	ANSD	-0.38	0.06	-0.35	0.09	-0.12	0.56
	SNHL	0.02	0.94	0.09	0.67	0.07	0.75
Follow up II	ANSD	-0.28	0.18	-0.32	0.12	-0.24	0.25
	SNHL	-0.65	0.06	-0.47	0.06	-0.10	0.63

Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

4.4. Discriminant Analysis identifying the best audiological metric of group differences

Fisher discriminant analysis (FDA) was done to identify a selected set of the measures (PTA, degree of HL, Symmetry, SRT, ABR, OAEs, CM) that most effectively distinguish the pathology based group differences. In the present study, the data obtained from two groups of participants (ANSD and SNHL) in each phase of evaluation (baseline, follow up 1 and follow up 2) was subjected to FDA. The discriminant function (DF) accounted for total of 100 % of variability among the groups in all the phases of evaluation as shown in Table 4.6. Eigen value of the given function, defined as degree of the observed variance for each factor is also shown in the table 4.6. Larger the Eigen value, greater the variance in the dependent variable that is explained.

The statistical significance of these functions in segregating the groups was obtained using Wilk's lambda (λ), which did not show DF to significantly classify the group differences. Smaller values of Wilk's lambda (λ) indicate greater discriminatory ability of the function.

Table 4.6.

Eigen values for audiological characteristics at baseline, follow up 1 and follow up 2

Evaluation/ phase	*Eigen value*	*% of variance*	*Wilk's lambda*	*Chi-square*	*Df*	*Significance*
Baseline	0.75	100	0.57	10.40	7	0.03*
Follow up 1	18.12	100	0.52	35.41	20	0.01*
Follow up 2	3.61	100	0.22	19.86	18	0.34

Note: * indicates effect is significant at 0.05 level (2-tailed), ** indicates effect is significant at 0.01 level (2-tailed), *** indicates effect is significant at 0.000 level (2-tailed).

Further the results of FDA also yielded standardized discriminant function coefficient values which depict the relative contribution of various audiological measures (PTA, degree of HL, Symmetry, SRT, ABR, OAEs, CM) in discriminating the groups, which is shown in Table 4.7. Coefficients with large absolute values correspond to variables with greater discriminating ability. In the current study, out of the audiological measures PTA and symmetry emerged as best predictors of group differences between ANSD and SNHL. Among these two metrics, PTA which had the largest discriminability in the baseline, became decreasingly important for group segregation in the follow –up 2. In contrast the symmetry of hearing which was 2nd largest matric in discriminating groups at baseline became increasing important in follow-up one and replaced PTA as the most important measure of group differences. This trend became even more evident at follow-up 2 where symmetry became the major predictor for group differences.

Table 4.7.

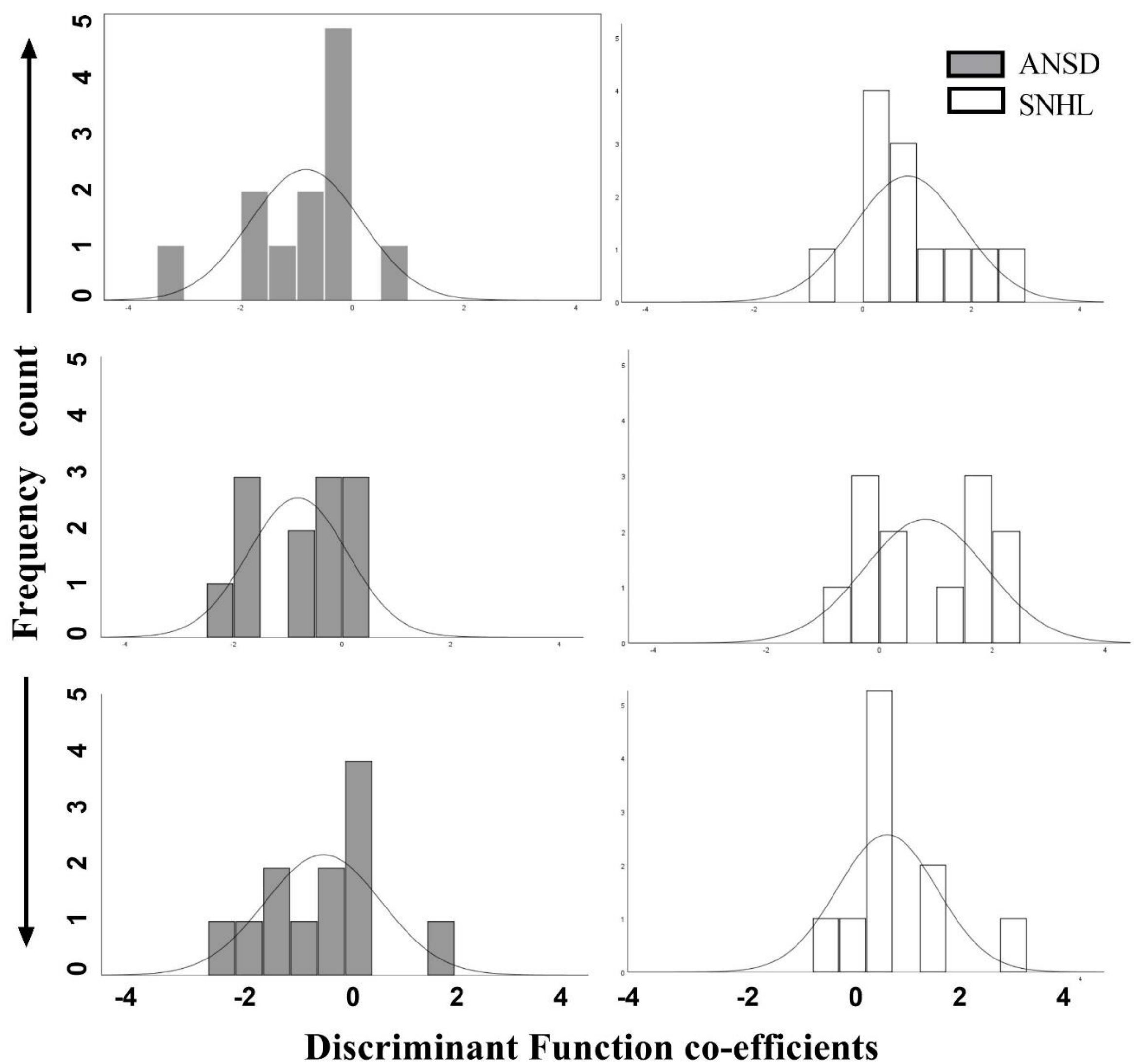
Standardized Canonical Discriminant Function Coefficients for audiological measures at baseline, follow up 1 and follow up 2. Grey shaded boxes represent higher group discrimination ability of the highlighted measure.

Evaluation phase	*Baseline*		*Follow up 1*		*Follow up 2*	
Measures	*Weights/ coefficients*	*Structure matrix*	*Weights/ coefficients*	*Structure matrix*	*Weights / coefficients*	*Structure matrix*
PTA	0.61	0.53	0.42	-0.04	-0.12	-0.52
Degree	-0.39	-0.37	0.35	0.25	-0.50	-0.31
Symmetry	0.29	-0.36	0.55	0.25	0.87	0.52
SRT	-0.28	-0.08	-0.49	-0.27	-0.95	-0.68
OAE	0.29	-0.37	-0.69	-0.58	-0.52	-0.49
ABR	-0.41	-0.57	0.51	0.47	-0.58	0.00
CM	0.52	0.57	-0.29	0.000	-0.18	0.000

Each participant's score on the discriminant function was calculated by multiplying the standardized canonical DF coefficient by the test score of each individual on the measures and summing these products. Thus, calculated frequency (y-axis) for each discriminant score (x-axis) is shown in Figure 4.3. It is clear from the figure that the DF separates the ANSD group from the SNHL group.

Figure 4.3:

Bar graphs representing the Discriminant Function scores for the segregation of both the groups.



Furthermore, the classification results of discriminant function analysis (Table 4.8) also revealed that differentiating the groups based on an increasing trend in the total number of confusions in group membership prediction across timelines of evaluations. While the 91.7 % of original grouped participants (both ANSD and SNHL) were correctly classified in baseline, this overall classification accuracy reduced to 70.8 % in follow up 1 and follow up 2 phases. While the classification accuracy of 75.0 % in ANSD and 66.7% in SNHL was observed in the first follow up phase, a further reduction in classification accuracy of ANSD (58.3 %) was seen in the follow up 2, as shown in Table 4.8. Although the classification accuracy of SNHL marginally improved at follow up 2 (83.3%).

Table 4.8.

Classification results for groups in baseline, follow up I and follow up II phases.

	Phase	*Groups*	*Original count* *ANSD*	*SNHL*	*Percentage correctly classified* *ANSD*	*SNHL*
Predicted group membership	Baseline (91.7 % of original grouped cases correctly classified)	ANSD	11	1	91.7	8.3
		SNHL	1	11	8.3	91.7
	Follow up I (70.8 % of original grouped cases correctly classified)	ANSD	9	4	75	33.7
		SNHL	3	8	25	66.7
	Follow up II (70.8 % of original grouped cases correctly classified)	ANSD	7	2	58.3	16.7
		SNHL	5	10	41.7	83.3

Chapter 5

DISCUSSION

The purpose of the study was to compare the functional gain and speech and language outcomes in children with auditory neuropathy spectrum disorder (ANSD) and cochlear-sensorineural hearing loss (SNHL) using hearing aids (HAs). The study used retrospective data from medical records of 24 children participants, divided into the above two groups of 12 each.

The findings of the study is discussed under the headings.

- 5.1 Comparison of hearing aid benefit on audiological outcomes between ANSD and cochlear-SNHL children.
- 5.2 Comparison of hearing aid benefit on speech and language outcomes between ANSD and cochlear-SNHL children.
- 5.3 Correlational analysis between hearing aid benefit and language score across evaluations
- 5.4 Discriminant analysis identifying the best metric of group differences across evaluations.

5.1 Comparison of hearing aid benefit on audiological outcomes between ANSD and cochlear-SNHL children.

At all three phases, the mean functional gain in ANSD children was lower than the mean functional gain in SNHL children, indicating a lower benefit from HAs in ANSD children in all three evaluations (Figure 4.1.). These differences in the mean functional gain can be attributed to physiological and anatomical differences between ANSD and SNHL groups. Rance and Starr (2015) discussed various structural changes such as hair cell loss, presynapse problems (ribbon synapse), and post synaptic disorders (demyelination) in ANSD group. In cases of cochlear origin SNHL children, the lesion will be at the level of hair cells, with the higher auditory pathway remaining intact. These distinctions in the anatomical aspects can be mirrored with audiological outcomes of both aided thresholds, which translates into functional gain differences in HA.

Results of ANOVA also revealed main effect of group on functional gain of HA (Table 4.1). However, on the post-hoc independent t-test the differences were found only for baseline, with ANSD performing poorer than children with cochlear-SNHL. This difference was confined to 500Hz at follow up 1 and no significant group differences were observed at follow up 2 (Table 4.2). This finding is very important because although unaided performance was comparable between the two groups (due to controlled and matched the degree of hearing loss, age and HA characteristics at the start of study), there was a difference in aided performance between the two groups, which got reflected as higher functional gain in SNHL compared to ANSD group. This could be related to disrupted neural synchrony in ANSD group (Santarelli et al., 2015 ; Raveh et al., 2007). In the ANSD group, the use of HAs would benefit only audibility and not neural synchrony. Although the audibility problem is solved here (aided thresholds verified using speech banana), the synchrony problem could not be replaced, resulting in this poorer functional gain in ANSD children. On converse, once the audibility issue in SNHL children was resolved using HAs, the physiologically intact neural synchrony in them (Nikolopoulos, 2014) would have reflected as higher functional benefit from the HA at the baseline.

However, both the groups get acclimatized with HA functionalities and features with time. Therefore, these group differences reduced gradually with time. The differences only confined to 500 Hz in follow up 1, and no significant group differences were observed in follow up 2. This finding could be attributed to 2 reasons:

1. The children with ANSD require higher time for auditory acclimatization with HA, than children with SNHL, perhaps as long as 2 y. The HA benefits subsequent to acclimatization happens sooner in children with SNHL and the benefits are realized earlier, while more auditory experiences are needed in ANSD children before realization of HA benefits.
2. In follow up 1, the retrospective analysis of the data accumulated in our study showed 50% (6/12) of OAE recorded in baseline for ANSD children reduced to 8.33% (1/12) in follow up 1, which further more reduced to 0 % (0/12) at follow up 2. The reduction in OAEs attributed to loss of OHCs cells, can atleast in theory remove the lesion confining to hair cells, with only probability of other two lesions (pre-synaptic and demyelination) in ANSD (Rance & Starr, 2015). This along with acclimatization in ANSD would have facilitated equalization of HA function gain in them at follow ups, although this might take as long as 2 y as reported in the present study.

The above finding should draw the attention of Audiologists for a longer follow-up in ANSD for monitoring HA benefits. Further, this finding suggests that ANSD affected children should not automatically be considered cochlear implant candidates, as realisation of auditory benefits will take longer time. In addition, this finding also indicates the need for the greater enhanced auditory stimulation including auditory and language training in ANSD group, to facilitate faster acclimatization and language learning outcomes.

5.2 Comparison of hearing aid benefit on speech and language outcomes between ANSD and SNHI children.

Improvement in the language scores (receptive language age: RLA, expressive language age: ELA & combined language age: C-LA), were assessed for both groups using two tests (receptive expressive emergent language test: REELS and scales for early communication skills for hearing impaired children: SECS). The results showed that language scores improved over time. At all three phases, the SNHL group outperformed the ANSD group in terms of language improvement (Figure 4.2). The language-based differences observed between the groups were greater at baseline than at follow-up 1, with significantly higher benefits in SNHL children compared to ANSD. The difference at follow-up 2 was negligible, except for C-LA, where SNHL children had significantly higher language scores than matched SNHL counterparts (Table 4.4). Similar findings were found by Rance and Barker (2009), on language outcomes (Peabody picture vocabulary test-PPVT) in children with ANSD who were treated with cochlear implants (CIs) or HAs. Rance and Barker (2009) found that 9/10 ANSD children implanted with CI, had comparable performance on PPVT test was not only similar to matched cohort of CI users with cochlear-SNHL using CI (n=10), but also ANSD children who were long-term (4 y) HA users (n = 10). This finding implies that candidates with ANSD using HAs should be given long acclimatization period to match language outcomes to a HA user with cochlear- SNHL, in terms of spoken language measures (PPVT). Similarly, though SNHL children enjoyed significant advantage over SNHL children in baseline on both verbal (RLA, ELA) and non-verbal language measures (C-LA) in our present study, the group differences became negligible in follow up-2 indicating the HA acclimatization related changes in ANSD on verbal language (RLA, ELA) outcomes. The equal scores on RLA and ELA of ANSD in follow up 2 compared to SNHL, despite compromised neural synchrony in them can be understood from the nature of language development: in order to comprehend an acoustic signal, a child may not require a very precise acoustic cue. Even a degraded signal is sufficient to understand a message, and repeated exposure to the same stimuli would have formed a deviant auditory image, facilitating the child's association of the image with the respective symbols. Similar observations were also noted by Praveena, Prakash, and Rukmangathan (2014), who reported RLA and ELA improved with HA and auditory training in their case report of 3 children with ANSD.

In contrary, non verbal language outcomes (CLA) in SNHL continued to enjoy an advantage over ANSD, in follow up 2 also, suggestive of sensitivity of C-LA in identifying group differences in long-term language outcomes. This finding could also stem from the differences type of amplification used between the two studies. While the ANSD children in Rance and Baker (2009) were CI users, the current study had HA users. In summary from the findings on language outcomes, we suggest the combined use of verbal language measures (RLA, ELA) and non-verbal language (C-LA) measures to effectively categorize the group differences in language outcomes based on the timeline of assessment.

5.3 Correlational analysis between hearing aid benefit and language score across timelines

Correlational analysis was performed to infer the relationship between overall functional gain of HA and the three language scores (RLA, ELA, C-LA) at three phases: baseline, follow up 1, and follow up 2 (Table 4.5). Results revealed no correlation between the measures (overall functional gain of HA and language scores) for the ANSD group. The reason for this finding can be attributed to the a large variability in neural synchrony among subjects with ANSD disorder (De Siati et al., 2020), which in turn gets reflected as large variability in HA and language outcomes.

In contrast, a significant correlation between overall functional HA gain and language scores were seen in SNHL at baseline, while with follow up even children with SNHL tend to have variable outcomes on the speech and language measures (Table 4.5). This finding showed the variability in language scores with acclimatization in SNHL children.

5.4 Best audiologic metric for identifying group differences across evaluations

With baseline, the group differences were more apparent on PTA followed by symmetry, while with follow ups, the best metric changed to symmetry (Table 4.7). This finding should alert Audiologists to focus more on the PTA and symmetry measures while looking for group differences across evaluations. Even though the degree of HL was balanced between the groups in the baseline, the differences were still visible to PTA because thresholds assessed in ANSD might have been obtained on BOA, where there is a high scope for examiner-based bias. However, with time the variability across measures seen in ANSD children would have unequivocal shifts in the thresholds (PTA) between ears, which might have affected the symmetry as seen in follow up 2. The asymmetry seen with time in ANSD, could be a marked factor to distinguish progression of pathology in ANSD children using HA.

With respect to the classification results, the ANSD error is increasing over time. This could be attributed to the absence of OAEs that were present at the start. Though the ANSD group was very different from the Cochlear-SNHL group at the baseline, with the follow up or progression, certain parameters such as OAEs, CMs known to disappear with time in them (owing to the amplification given) could have resulted in the disappearance of these group differences, leading to increased errors in ANSD group-prediction at follow ups.

Chapter 6

SUMMARY AND CONCLUSIONS

The is a scanty literature in terms of hearing aid (HA) benefit and language outcome in children with Auditory neuropathy spectrum disorder (ANSD), is the rationale for the current investigation. The purpose of the study was to compare the HA outcomes and speech and language outcomes in children with ANSD and cochlear-Sensorineural hearing loss (SNHL). The retrospective data obtained by scrutinising the medical reports of 24 participants, aged 1-4.9y was categorized as two groups based on the diagnosis of the participant. Data belonging to ANSD subjects was segregated as Group I (n = 12, mean age:2.43±1.21 SD), while matched cohort with diagnosis of SNHL (n = 12, mean age:2.43±1.21y SD) were categorised under Group II. Audiological parameters (PTA, degree of HL, Symmetry, SRT, ABR, OAEs, CM, aided and unaided free-field thresholds) and language scores (receptive (RLA) and expressive (ELA) language age, combined (C-LA) language age) at 3 evaluations (baseline, follow up-1, & follow up-2) was considered preliminary criteria for inclusion of both groups. The hearing aid benefit (functional gain) was quantified as difference between unaided and aided thresholds obtained at three evaluations for 4 frequencies (500, 1000, 2000 and 4000 Hz), while speech and language outcome was quantified as difference of chronological and language age on the three measures [RLA, ELA, CLA].

For hearing aid (HA) benefit, the results of two-way repeated measure ANOVA revealed significant main effect of groups, frequencies and significant interaction effects between them. Follow up test t-test revealed functional gain of HA in SNHL group was significantly greater than ANSD group at all frequencies in the baseline, while the similar group differences confined to only 500 Hz at follow up 1. No group differences in functional gain were seen in follow up 2. This finding shows that children with ANSD benefited less (lesser functional gain) at first, but their benefit improved over time and became comparable to that of children with cochlear-SNHL. This finding is significant because children with ANSD do not readily improve their prescribed HA gain at first fit. More auditory experience is required before the benefits of HAs can be realised. The materialization of functional HA gain benefits in ANSD with time is also reflected on their verbal language (RLA, ELA) improvement, while they lacked in non-verbal language (C-LA) compared to SNHL in follow up 2, showing despite acclimatization with HAs, children with ANSD do lag behind the SNHL peers in language development.

Lack of correlation between overall HA functional gain and language scores in ANSD can be attributed to the variability among ANSD subjects. The discriminant analysis cues audiologists to focus more on PTA, and Symmetry at Baseline whereas for the symmetry of hearing should be the focus in follow up 1 and follow up 2, for understanding the group differences with time. To conclude, while evaluating the hearing aid benefit using functional gain for PT threshold in children with ANSD it should be kept in mind that they should require more auditory experience, hence they should be given enough time to adjust to the HAs before deciding the cochlear implant candidacy.

Limitations and Future directions.

To understand the impact of HA users with ANSD on language acquisition, it is crucial to be aware of the factors involved. Many factors such as the type and degree of hearing impairment, the age of onset and detection, the methods used to treat the impairment, communication strategies, language models available in the home and community, and the child's intellectual and remaining sensory abilities, can be play an important role. Though all of these aspects could not be controlled in the study, effort was taken to minimize the variability by matching ANSD and SNHL groups at the start of study. Also, the study was done on a small population (12 each group), owing to which the readers are advised to exercise caution before generalizing the study findings.

Chapter 7

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