Effect of Acoustical Enhancement of Speech on Audio-visual Perception in Individuals with Auditory Neuropathy Spectrum Disorders

DOCTORAL THESIS

Submitted to the University of Mysore,

for the award of

Doctor of Philosophy (Ph.D.) in Audiology

By

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DECLARATION

I declare that this thesis entitled **'Effect of Acoustical Enhancement of Speech on Audio-visual Perception in Individuals with Auditory Neuropathy Spectrum Disorders'** which is submitted for the award of the degree of Doctor of Philosophy in Audiology to the University of Mysore, is the result of work carried out by me at the All India Institute of Speech and Hearing, Mysuru, under the guidance of Dr. Sandeep M., Reader in Audiology, All India Institute of Speech and Hearing, Mysuru. I further declare that the results of this work have not been previously submitted for any other degree.

Place: Mysuru Jithin Raj B.

Date:

CERTIFICATE

This is to certify that the thesis entitled **'Effect of Acoustical Enhancement of Speech on Audio-visual Perception in Individuals with Auditory Neuropathy Spectrum Disorders'** submitted by Mr. Jithin Raj B., for the degree of Doctor of Philosophy in Audiology to the University of Mysore was carried out at the All India Institute of Speech and Hearing, Mysuru, under my guidance. I further declare that the results of this work have not been previously submitted for any other degree.

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CERTIFICATE

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Date:

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DEDICATED TO

DADDY, SANJAY SIR,

PRIYANKA

&

AURA

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ABSTRACT

The study aimed to assess the effect of acoustical enhancement of speech on audio-visual perception in individuals with Auditory Neuropathy Spectrum Disorders (ANSD).Two groups of individuals in the age range of 16 to 35 years participated in the study. The clinical group included 40 participants diagnosed as having ANSD, while the control group had 40 age and gender matched individuals with typical auditory abilities. Syllable identification was assessed in auditory, visual, and auditory-visual (AV) modalities. There were three types of stimuli (primary, companded, & envelope enhanced), presented in quiet and 0 dB SNR conditions. The syllable identification scores were compared across modalities, stimuli and conditions to derive the relative benefits of visual cues and acoustic enhancement on speech perception of individuals with ANSD. Sequential Information analysis (SINFA) was also done to derive the feature-wise information transmitted in different test conditions in each group. The results showed maximum syllable identification score in the AV modality followed by auditory and least in visual modality. This was true in both the groups. Syllable identification scores along with the results of SINFA showed that both auditory and visual modalities play an important role, particularly in challenging listening conditions. The individuals with ANSD were able to make better use of visual cues than the control group, as evident in the visual gain score. However, acoustic enhancement of speech did not significantly enhance speech perception. Based on the results it can be concluded that there is definite benefit of auditory as well as visual cues to individuals with ANSD, suggesting the need to facilitate both the modalities as part of the audiological rehabilitation.

Chapter 1

INTRODUCTION

Auditory neuropathy refers to a unique hearing disorder in which the neural conduction is impaired with normal cochlear amplification. The clinical diagnosis of the condition will be based on the presence of otoacoustic emissions/cochlear microphones, and the absence/abnormality of auditory brainstem responses (Starr, Picton, Sininger, Hood, & Berlin, 1996). At first, the disorder was termed as auditory neuropathy as the majority of the affected individuals were reported to have associated peripheral neuropathy. Later, in view of the lesion restricting to inner hair cells in some of the cases (Miyamoto, Kirk, Renshaw, & Hussian, 1999), the term auditory dys-synchrony was suggested (Berlin et al., 2002). Sininger and Hayes (2008) suggested the term auditory neuropathy spectrum disorder (ANSD) considering that the damage is not confined to a particular site in most of these persons, rather there are different affected loci. Henceforth in this study the condition will be uniformly referred to as ANSD.

Research has revealed an alarming incidence and prevalence of ANSD among individuals with hearing impairment. The incidence of ANSD in patients with profound hearing loss is estimated to be 10% with a prevalence of 0.23% among high-risk babies (Kraus, Ozdamar, Stein, & Reed, 1984; Rance et al., 1999). Rance et al. (1999) from a hospital-based statistics assessed 5199 ‘at-risk’ children for ANSD. The prevalence of ANSD among children at-risk was 1 in 433 (0.23%) and in children with hearing impairment was 1 in 9 (11.01%). It was estimated that 2% to 15% of infants with hearing loss may exhibit ANSD (Rance et al., 1999; Sininger, 2002).

Davis and Hirsh (1979) reported that 1 in 200 children with hearing impairment exhibit the clinical trait of ANSD. Tang, McPherson, Yuen, Wong and Lee (2004) investigated the frequency of occurrence of ANSD in school-aged children with hearing-impairment and reported a prevalence of 2.44%. The prevalence of ANSD in India has been reported to be 0.54% among individuals with sensorineural hearing loss (Kumar & Jayaram, 2006).

Starr, Sininger and Praat (2000) reported the occurrence of peripheral neuropathy in 80% of individuals with ANSD in whom age of onset of the disorder was above 15 years. It was also reported that in 96% of individuals with ANSD, the occurrence is bilateral in nature and no gender difference was noted. On the contrary, Narne, Prabhu, Chandan and Deepthi (2016) reported a female to male ratio of 1.25:1 in Indian population.

The hearing thresholds in individuals with ANSD could vary from normal hearing to severe degree of hearing loss (Rance et al., 1999). The majority of them are reported to have mild to moderate sensorineural hearing loss (Starr et al., 1996) while the configuration of the audiograms is generally variable. The audiograms could be of rising configuration (Rance et al., 1999), flat and unusual configuration, and rising configuration with a peak at 2 kHz (Kumar & Jayaram, 2005). It was also noted that the individuals with ANSD with peaked audiograms showed better speech discrimination skills compared to individuals with audiograms of other configurations (Kumar & Jayaram, 2005; Jijo & Yathiraj, 2012).

The cardinal feature of individuals with ANSD is the poor speech perception ability (Starr, Picton, Sininger, Hood, & Berlin, 1996; Zeng, Kong, Michalewski, & Starr, 2005), more so in adverse listening conditions (Shallop, 2002). The speech

perception abilities are disproportionate to their hearing thresholds and have been reported to correlate with the disrupted temporal processing in them (Kumar & Jayaram, 2005; Narne & Vanaja, 2009; Zeng et al., 2005). Temporal resolution deficits are reported to be the cause of impaired perception of short and dynamic auditory signals (Kraus et al., 2000). The impaired temporal resolution is likely to affect the perception of acoustic cues such as voice onset time, burst duration and formant transitions, thereby resulting in poor perception of consonants, mainly the stops/plosives (Kumar & Jayaram, 2011).

Management of speech perception difficulties of persons with ANSD is always a challenge to the Audiologists. Conventional amplification devices such as hearing aids do not address their temporal processing deficits and are therefore known to yield limited benefit (Miyamoto, Kirk, Renshaw, & Hussain, 1999; Shallop, Peterson, Facer, Fabry, & Driscoll, 2001; Sininger, Hood, Starr, Berlin, & Picton, 1995) in improving the speech perception in them. Frequency modulated (FM) devices are reported to provide better benefit compared to conventional hearing aids by improving the SNR of the target speech (Rance, Corben, Du Bourg, King, & Delatycki, 2010). However, the utility of FM devices is limited to a few listening situations. Cochlear implants (CI) are known to benefit individuals with ANSD only if the lesion is presynaptic or synaptic (Berlin, Hood, Morlet, Wilensky, Li, & Mattingly, 2010; Miyamoto, Kirk, Renshaw, & Hussain, 1999).

In view of the poor spectral and temporal processing abilities of persons with ANSD, attempts have been made to enhance the input speech signal in various ways to facilitate speech perception in them. Companding is one such method of spectral enhancement wherein the peak to valley difference in the spectrum is increased

(Bhattacharya & Zeng, 2007). Such spectral contrast enhancement was expected to compensate for the poor frequency resolution in individuals with hearing impairment (Tyler, Fernandes, & Wood, 1980). As a supporting finding, companding was found to improve speech perception using simulated CI processing among individuals with normal hearing (Oxenham, Simonson, Turicchia, & Sarpeshkar, 2007) and also among the CI users (Bhattacharya & Zeng, 2007). Narne, Barman, Deepthi and Shachi (2014) found companding to improve speech perception in persons with ANSD. But the improvement was limited at 0 dB SNR condition compared to quiet condition.

Speech perception deficits seen in persons with ANSD are also thought to be due to their failure in utilizing the low-frequency envelope cue of the signal (Narne, 2013). These individuals are known to have abnormal perception of temporal modulation and therefore require more modulation depth in order to perceive the modulation in the signal (Kumar & Jayaram, 2005). Studies in cochlear hearing loss have shown that envelope enhancement can improve speech perception even in the presence of background noise (Baer, Moore, & Gatehouse, 1993). Bhattacharya, Vandali and Zeng (2011) studied the combined effect of spectral expansion and temporal enhancement spectral maxima (TESM) in CI users. They found an improvement in the participants' vowel and consonant recognition even in the presence of noise. In view of these findings, Narne and Vanaja (2008) enhanced the envelope of the speech signal by a magnitude of 15 dB and found an improvement in the speech perception of persons with ANSD. They reported that envelope bandwidth enhancement in the 3 to 30 Hz region resulted in best possible improvement.

The other signal enhancement strategies used to facilitate speech perception in persons with ANSD include stretching of formant transition duration (Kraus et al., 2000) and time-scale modification of short acoustical cues in speech (Kumar & Jayaram, 2011). Although these strategies have been found to improve speech perception (Kumar & Jayaram, 2011; Kraus et al., 2000; Narne et al., 2014), current technology does not support implementation of these strategies in the online modification of signal, as in hearing aids and cochlear implantation. Jijo (2015) studied the effect of stretching and spectro-temporal modification on speech perception in ANSD. It was reported that stretching the speech by 25% had improved speech perception, whereas spectro-temporal modifications did not result in significant improvement.

In instances where auditory cues are compromised, as in the presence of competing-noise, visual cues are typically used to facilitate speech perception (MacLeod & Summerfield, 1987; Munhall, Kroos, Jozan, & Vatikiotis-Bateson, 2004; Tye-Murray, Sommers, & Spehar, 2007). In challenging listening environments, integration of auditory and visual cues is found to enhance speech perception (Anderson, 2006; Ross, Saint-Amour, Leavitt, Javitt, & Foxe, 2006). Evidences from both behavioral and neurophysiological experiments support the existence of auditory-visual (AV) integration (Grant, Walden, & Seitz, 1998) and among individuals with hearing impairment, speech perception in AV modality is reported to be better compared to auditory alone or visual alone conditions. This is true in degraded acoustic conditions as well as in silent discourse (Grant et al., 1998).

Ramirez and Mann (2005) studied speech perception in ANSD in different modalities and found that persons with ANSD primarily relay on visual cues to

understand speech in quiet and in noise. They did not find a significant difference between the scores of visual and AV modalities, suggesting that persons with ANSD ignore auditory modality. However, Maruthy and Geetha (2011) reported that speech perception in AV modality is driven by cues of both auditory and visual modalities.

1.1 Justification for the Study

ANSD as a challenging clinical condition has drawn the interest of clinicians and researchers alike. This has resulted in in-depth understanding of the condition in terms of its pathophysiology, audiological characteristics and the underlying mechanisms of perceptual deficits. In spite of this understanding, successful management of the speech perception deficits in these individuals has been an unattainable task for audiologists. Persons with ANSD have negligible benefit from the conventional amplification devices. Although assistive listening devices like FM systems have proved to be beneficial compared to conventional hearing aids (Rance, Corben, Du Bourg, King, & Delatycki, 2010), they do not address the primary psychoacoustical difficulties (temporal processing deficits) encountered by these individuals and hence their utility is expected to be limited to a few listening conditions only. Cochlear implantation is known to benefit individuals with ANSD only if the lesion is presynaptic or synaptic (Berlin et al., 2010; Miyamoto et al., 1999; Sininger & Trautwein, 2002).

Acoustic enhancement strategies were found to improve speech perception in persons with ANSD. Independent studies have shown that both companding and envelope enhancement strategies are beneficial (Narne, Barman, Deepthi, & Shachi, 2014; Narne & Vanaja, 2009; Mathai & Yathiraj, 2013; Kumar & Jayaram, 2011). However, which of these strategies is more beneficial is not known. This warrants a

study that compares the different strategies in the same group of persons with ANSD. The findings from such a study would be helpful in making evidence-based recommendations for the implementation of acoustic enhancement strategies in amplification devices used for persons with ANSD.

The presence of background noise is known to severely reduce speech perception in persons with ANSD. Considering that background noise is invariably present in most of the listening environments, it is important to validate the utility of the above mentioned acoustic enhancement strategies in the presence of noise. The strategy that improves speech in noise perception should be recommended for use in persons with ANSD.

Although the acoustic enhancement strategies have been found to show improvements, none of these techniques led to complete or near complete speech perception. The benefit derived is further reduced in the presence of background noise (Narne, Barman, Deepthi, & Shachi, 2014; Narne & Vanaja, 2009). This means that the persons with ANSD are less likely to be satisfied with the management that involves only the acoustic enhancement strategies. In such situations, it is advisable to recommend AV modality for better speech perception.

The use of visual cues has always been advised during the management of persons with ANSD. However, a clear documentation of the benefit obtained from the visual cues for speech perception has not been done till date. Ramirez and Mann (2005) assessed speech perception in 4 persons with ANSD and found that they completely rely on visual modality while perceiving speech in the AV modality and ignore auditory modality. However, considering that ANSD is a heterogeneous condition, it warrants evidence from a larger group. Contrary to Ramirez and Mann

(2005), Maruthy and Geetha (2011) have shown evidence for persons with ANSD utilizing cues from both auditory and visual modalities. The equivocal results in terms of relative contribution of visual and auditory cues for the AV speech perception warranted further studies in this direction.

Furthermore, considering that the auditory input is distorted in ANSD, during the process of AV integration in these individuals, there could be mismatch in the input from the auditory and visual modalities, possibly leading to inherent incongruence between the two modalities. This in turn could negatively influence speech perception. Therefore, dynamics of AV speech perception in persons with ANSD needs to be evidenced before advising it as a rehabilitative option. The role of visual cues in AV speech perception also has to be established in quiet as well as in the presence of noise, to determine its realistic benefit to persons with ANSD.

The benefits of visual cues and acoustic enhancement have been documented in independent studies. However, these reports do not provide a clear picture about which of these yields better speech perception in individuals with ANSD. Therefore, it is important to compare their relative contribution in improving speech perception of persons with ANSD. This will guide the clinical audiologist in choosing the right strategy for the best possible management of ANSD. Furthermore, it is important to understand whether the combination of acoustic enhancement and visual cues supplementation results in better benefit compared to a single strategy. The interaction between the facilitation provided by the two strategies warranted a systematic investigation. It is also important to highlight the individual differences in the benefit derived from the acoustic enhancement and visual cues supplementation, which needs definite consideration in deriving the clinical benefit of these strategies.

It is well understood that individuals with hearing impairment who depend on speech reading pay less attention to auditory information. In such a case, one cannot simply advise to combine speech reading with signal enhancements. Also, if the cues derived from speech reading and signal enhancements are the same, additional visual cues may not enhance speech perception. Therefore, dynamics of AV speech perception in persons with ANSD needs to be investigated before advising it as a rehabilitative option. Hence, the present study was taken up.

1.2 Aim of the Study

To investigate the effect of acoustical enhancement of speech on audio-visual perception in individuals with auditory neuropathy spectrum disorders.

1.3 Objectives of the Study

The specific objectives of the present study were

1) To compare normal hearing individuals and persons with ANSD for their syllable identification, auditory gain and visual gain scores.
2) To compare normal hearing individuals and persons with ANSD for their feature transmission index derived from SINFA.
3) To investigate the effect of stimulus, modality, and condition on syllable identification scores of persons with ANSD.
4) To investigate the effect of stimulus, modality and condition on feature transmission index derived from SINFA, in persons with ANSD.
5) To determine the predictors of benefit from AV modality in persons with ANSD.
6) To correlate the duration of hearing loss, pure tone average, and speech identification scores of persons with ANSD with their respective auditory and visual gain scores.

1.4 Hypotheses of the Study

The present study tested the following null hypotheses

1) There is no significant difference between normal hearing individuals and persons with ANSD in their syllable identification, auditory gain and visual gain scores.
2) There is no significant difference between normal hearing individuals and persons with ANSD in their feature transmission index derived from SINFA.
3) There is no significant effect of stimulus, modality, and condition on syllable identification scores of persons with ANSD.
4) There is no significant effect of stimulus, modality, and condition on feature transmission index derived from SINFA, in persons with ANSD.
5) Syllable identification scores in the auditory and visual modality are not significant predictors of benefit from AV modality.
6) There is no significant correlation of duration of hearing loss, puretone average, and speech identification scores of persons with ANSD with their respective auditory and visual gain scores.

Chapter 2

REVIEW OF LITERATURE

In this chapter, the available literature in the major areas of concern in ANSD such as, speech perception in ANSD, different signal enhancement strategies used for the management of ANSD and audio-visual speech perception in individuals with ANSD is reported in detail. The review of literature is presented under the following major sections

1. Audiological Profile in ANSD
2. Age of onset of ANSD
3. Aetiology and pathophysiology of ANSD
4. Psychoacoustic abilities in ANSD
5. Speech Perception in ANSD
6. Management of ANSD
7. Auditory-visual perception of speech

2.1 Audiological Profile in ANSD

2.1.1 Hearing sensitivity

The hearing thresholds in individuals ANSD could vary from normal hearing to severe degree of hearing loss (Zeng et al., 2005; Rance et al., 1999). Configuration of hearing loss could be either typical rising (Sininger & Starr, 1997; Hood, 1998; Rance et al., 1999), rising with peak at 2 kHz (Kumar & Jayaram, 2005) or flat in nature. Persons with ANSD having peaked audiogram are reported to have better speech discrimination abilities compared to other configurations (Kumar & Jayaram, 2005; Jijo & Yathiraj, 2012).

2.1.2 Middle ear muscle reflexes (MEMRs)

MEMRs are known to be present in only a few of the persons with ANSD. Starr et al. (2000) found the presence of MEMRs in only 7% of persons with ANSD tested. Similar findings have been obtained in Sininger and Oba (2001) and Cheng et al. (2005). Kumar and Jayaram (2006) reported absence of MEMRs in all of their subjects. The absence of MEMRs has been attributed to the inability of afferent pathway in generating sufficient synchronized neural discharge that trigger stapedius muscle contraction (Starr et al., 1998). The presence of non-acoustic middle-ear muscle reflexes in ANSD (Gorga, Stelmachowicz, Barlow, & Brookhouser, 1995; Starr et al., 1998) suggests normal functioning of the efferent part of the MEMR arc.

2.1.3 Otoacoustic emissions

Persons with ANSD are found to have higher mean amplitude of TEOAEs compared to their normal hearing controls (Hood, Berlin, Bordelon, & Rose, 2003; Kumar & Jayaram, 2005). Higher amplitude is attributed to the lack of efferent suppression in ANSD. However, the lack of efferent suppression and acoustic reflexes which are thought to protect the cochlea from loud sounds may result in permanent OHC damage over time (Berlin et al., 1993; Sininger et al., 1995; Starr et al., 1996). Reduced OAE amplitude and deterioration of OAEs has been found in persons with longstanding ANSD (Deltenre et al., 1999). This has been reported to be due to the use of hearing aids or may be due to the effect of OTOF mutation in OHCs (Rodriguez- Ballestros et al., 2003). Researchers have reported that the presence or absence of OAE however does not relate to the speech perception in persons with ANSD (Rance et al., 1999; Starr, Sininger, & Pratt, 2000).

2.1.4 Auditory evoked potentials

Auditory brainstem responses (ABRs) are known to be absent or abnormal in persons with ANSD. While most show absent ABRs, a few of them show present but abnormal ABRs. Starr et al. (2000) reported that 73% of the patients tested had absent ABR, whereas 21% had fifth peak present with reduced amplitude and 6% of them had the third and fifth peak present.

Electrocochleography (EcochG) is recommended in ANSD to confirm the peripheral functions (Kraus, Ozdamar, Stein, & Reed, 1984; Arslen, Turrini, Lupi, Genovese, & Orzan, 1997). The presence of summating potential in EcochG indicates normal functioning of inner hair cells (Durrant, Wang, Ding, & Salvi, 1998). Shi Kempfle and Edge (2012) reported that the input-output (I/O) function of cochlear microphonics helps in differentiating the site of lesion in persons with ANSD. If the I/O function of cochlear microphonics shows good nonlinearity, it indicates that the site of lesion could be either inner hair cells, synapse between IHCs and eighth nerve, or at the eighth nerve itself. On the contrary, reduced nonlinearity in the I/O function of cochlear microphonics indicates that the site of lesion could be at the synapse between IHCs and eighth nerve or at the eighth nerve itself.

Satya-Murti, Wolpaw, Cacace and Schaffe (1983) observed cortical auditory evoked potentials (CAEPs) for the first time in 6 individuals in whom the ABR was absent. Starr et al. (1996) could detect N1 and P2 component of CAEPs in three out of five individuals with ANSD. Rance, Cone-Wesson, Wunderlich and Dowel (2002) reported the presence of CAEP in 50% (9 out of 18) of individuals with ANSD. Since the CAEPs do not depend on the neural synchrony as much as the earlier potentials, temporal disruption has minimal effect on the cortical potentials (Hood, 1998; Rapin

& Gravel, 2003). Kumar and Jayaram (2005) reported the presence of P1 and N1 in 10 out of 14 individuals with ANSD being tested whereas P2 and N2 components were present in all the 14 individuals. In their study, mismatch negativity was also recordable in 9 out of 14 participants. Furthermore, there was no significant difference in the mismatch negativity between the normal and ANSD even though persons with ANSD were not able to discriminate the stimulus contrast behaviorally. On the contrary, delay in the late latency responses has been reported for tonal (Kraus et al, 1993; Starr et al., 2004), speech (Narne & Vanaja, 2008) as well as gaps in noise (Michalewski, Starr, Nguyen, Kong, & Zeng, 2005) stimuli. Compared to controls, a delay of up to 60 ms has been reported in individuals with ANSD.

In normal hearing individuals, cortical response to unvoiced stimulus has two peaks; one corresponds to the burst/aspiration (usually labelled as p1' in recording) and the second corresponds to the onset of voicing (Sharma & Dorman, 1999). The early peaks were not detected in ANSD (Kraus et al., 2000). The absence of P1' suggests a poor representation of transient cues which may be related to stimulus burst.

The relationship between the CAEPs and the speech perception abilities in ANSD has also been investigated. Kumar and Jayaram (2005) reported that there is no correlation of speech perception with the latency or amplitude of CAEPs. Whereas, Narne and Vanaja (2009a) grouped the participants ANSD as good (SIS>50%) and poor performers (SIS<50%) based on their speech identification scores. The comparison of late latency responses between these two groups showed that the amplitude of N1-P2 complex was lower for poor performers compared to good performers.

Rance, Cone-Wesson, Wunderlich and Dowell (2002) correlated the aided speech perception scores of individuals with ANSD with their late latency responses. They found that ANSD children with measurable speech recognition scores had the presence of good late latency responses. late latency responses positively correlated with the aided performance in ANSD. Those individuals who showed presence of CAEPs had an average speech perception score of 60%, while those without CAEPs had an an average perception score of only 6%. Based on their findings it was concluded that recording late latency responses can predict the speech perception score in ANSD. The authors hypothesized that preserved synchrony at the cortical level may be the contributing factor for better speech perception.

Alvarenga, Amorim, Agostinho-Pesse, Costa, Nascimento and Bevilacqua (2012) studied the correlation of late latency responses with the speech perception abilities in children with ANSD. In their study, P1 was recordable in 12 of 14 (85%) children using cochlear implants. Authors concluded that the P1 component can be an indicator of central auditory cortical development and a predictor of speech perception in implanted children with ANSD.

2.2 Age of Onset of ANSD

Berlin et al. (2010) studied the occurrence of ANSD in 260 patients and reported that 85.76% of their participants had an onset below the age of 12 years. A very few of them had an onset during puberty and adulthood. On the contrary, the other studies indicate the onset to be in the second decade of life. The onset of ANSD in Indian scenario is reported to be between 10 and 20 years (Jijo & Yathiraj, 2012), more frequently between 10 and 14 years of age (Kumar & Jayaram, 2006). Similar findings were reported by Prabhu et al. (2012) and Shivashankar, Satishchandra,

Shashikala, and Gore (2003). Wang, Gu, Han, and Yang (2003) reported late onset ANSD in their study. Rance (2005) found that nearly in 80% of individuals with ANSD, symptoms started after 15 years of age.

2.3 Aetiology and Pathophysiology of ANSD

The etiological factors of ANSD include genetic, infectious, toxic-metabolic (hypoxia, hyperbilirubinemia) and immunological disorders (drug reaction, demyelination). In most cases, the origin of ANSD is reported to be idiopathic in nature (Starr, Zeng, Michalewski, & Moser, 2008; Berlin et al., 2010; Starr, Sininger, & Praat, 2000). Conditions such as hyperbilirubinemia, ototoxic drug regimen, low birth weight, low APGAR scores, exposure to aminoglycosides, hyponatremia, anoxia and family history of deafness are also found to be the causative factors (Berlin, Hood, Morlt, Rose, & Brashears, 2002). Leonardis et al. (2000) reported a gypsy family with hereditary motor and sensory neuropathy (Lom HMSN-L) associated with ANSD. Studies have reported X-linked recessive inheritance and autosomal recessive inheritance in individuals with ANSD (Wang et al., 2003).

The conditions usually associated with ANSD include Charcot Marie Tooth disease, Friedrich Ataxia, Rufson syndrome and Gullian Barre syndrome (Starr et al., 1996) and multiple sclerosis (Cevette, Robinette, Carter, & Knops, 1995). Friedrich's ataxia (FRDA) is a neurodegenerative condition that is believed to be restricted to the brainstem and cerebellar parenchyma (Rance, 2005). FRDA is due to mutations in the FXN gene (Durr et al., 1996). Histological evidence shows spared cochlear structure and damage to the cochlear nerve, hence showing the features of ANSD (Spoendlin, 1974).

ANSD is also reported to be associated with other syndromes such as Harding disease, multiple sclerosis-like conditions which is caused by mutation of 11778mtDNA, (Berlin, Hood, & Morlet, 2003). The isolated case of ANSD is associated with rare genetic disorders such as Ehlers-Danlos syndrome (Sininger & Oba, 2001) and Stevens-Johnson syndrome (Doyle, Sininger, & Starr, 1998). ANSD was also reported to be associated with syndromes affecting mitochondrial enzymes (Deltenre, Mansbach, Bozet, Clercx, & Hecox, 1997; Corley & Crabbe, 1999).

Hyperbilirubinemia is known to be one of the most prevalent causative factors of ANSD (Kraus et al., 1984; Rance et al., 1999). The excessive amount of bilirubin usually causes damage to the CNS and peripheral nervous system, especially the cochlear nucleus (Chisin Perlman, & Sohmer, 1979; Kraus et al., 1984; Vohr et al., 1989). Sustained hypoxia is reported to be the other etiology of ANSD (Delterne et al., 1979; Harrison, 1988; Rance et al., 1999). In prolonged hypoxia, inner hair cells are more prone to damage than the outer hair cells (Shirane & Harrison 1987a; Billet et al., 1989). Apart from these more prevalent causative associations, ANSD has been reported to be secondary to mitochondrial disorders (Delterne et al., 1997; Corley & Crabbe, 1999), childhood measles/mumps (Prieve et al., 1991), and acute led poisoning (Starr et al., 2000). Prabhu et al. (2012) reported that among non-syndromic late onset ANSD, the causative factors are reported to be the hormonal, genetic and idiopathic conditions.

The possible site of lesion of ANSD includes inner hair cell (IHC), synapse between IHC and the VIII nerve, and the VIII nerve itself (Starr et al., 1996; Berlin, Hood, & Rose, 2001). Other possible location of dysfunction in ANSD include generation of receptor potential by IHC, transmitter release from IHC, nerve impulse

generation in VIII nerve dendrites, and the VIII nerve ganglion cell dysfunction (Starr, Sininger, Derebery, Oba, & Michalewski, 1998). ANSD is reported to be mainly of two types. Type I ANSD, which is postsynaptic, may have an associated peripheral neuropathy, which can be hereditary or inflammatory in origin (Starr et al., 20001a; Starr et al., 1996; Butinar et al., 1999). Whereas in type II ANSD, hearing loss is not confined to eighth nerve but lesion sites may also involve IHCs and also synapse of IHC with auditory nerve (Starr et al., 2001b).

Starr et al. (2003) conducted a histopathological investigation of the cochlea and auditory nerve in an individual with ANSD. It revealed normal organ of corti in the basal turn with nearly 30% loss of outer hair cells at the apex of the cochlea. The inner hair cells were reported to be normal throughout the length of the cochlea, but there was a significant loss of ganglion cells. The proximal part of the eighth nerve showed a marked reduction in the number of auditory fibers. Furthermore, the myelin sheath on the surviving auditory nerve fibers was thin indicating an incomplete myelination. MacDonald (1980) reported that in demyelinating neuropathy, the conduction velocity through the nerve slows down once the neural impulses pass through a demyelinated segment of the axon and then regain normal speed when that segment is passed. Thus, demyelination of varying degrees in different nerve fibers carry neural impulses at different velocities and results in neuronal de-synchrony. Demyelination is reported to result in an increase in membrane capacitance and decrease in membrane resistance, leading to a delayed excitation, reduction in the velocity of action potential propagation, and an increase in conduction vulnerability (McDonald & Sears, 1970; Rasminsky & Sears, 1972; Pender & Sears, 1984). The dys-synchronous firing of auditory neurons disrupts the ABR waveform along with

auditory perception which depends on temporal cues (Kraus et al., 2000; Starr et al., 1991; Zeng et al., 1999, 2005).

Barman (2008) assessed the temporal processing in ANSD by means of psychophysical methods and reported temporal processing deficits in individuals with ANSD. Studies have also reported normal or near normal temporal integration in ANSD (Zeng et al., 1999). They inferred that the perceptual deficits in ANSD are mostly caused by the demyelination or axonal loss of auditory nerve. McMahon, Pattuzi, Gibson and Sanli (2008), based on their findings of EcochG, and the eABR after cochlear implantation, reported the existence of pre and postsynaptic ANSD. Out of the fourteen subjects they tested, seven showed EcochG with delayed summating potential (with or without CAP) and superior eABR consistent with a presynaptic lesion. Whereas six subjects with normal summating and dendritic potential showed poor morphology of eABR or absent eABR consistent with a postsynaptic lesion.

A presynaptic form of ANSD may be the result of mutation of OTOF (Otoferlin) gene, which is an important for membrane trafficking, which affect the release of neurotransmitter (Rodríguez-Ballesteros et al., 2003; Roux et al., 2006; Varga, Kelley, Keats, Starr, & Leal, 2003). The OTOF is hence known to play an important role in synaptic vesicle trafficking and/or fusion to the plasma membrane (Yasunaga et al., 2000). Wang et al. (2010) reported OTOF mutation in 4 out of 73 ANSD subjects (5.5%) in Chinese population. The OTOF mutation in p1515t has also been found in temperature-sensitive ANSD (Varga et al., 2006). In case of demyelinating neuropathy, locus of the gene is reported to be on chromosome number 8 (8q24). Due to MPZ gene mutation, ANSD can have peripheral as well as the vestibular neuropathy (Starr et al., 2003). Further, mutation of ANUAI gene is

reported to be responsible for an autosomal dominant form of ANSD (Kim et al., 2004) and the ANSD is also found to result from a genetic disorder affecting peripheral myelin protein 22 (PMP-22) on chromosome 7p11.2 (Kovach et al., 1999).

Impaired perception of high-frequency information in ANSD is reported to be due to the limitation of the neural refractory period (Rance, 2005). Whereas, the impaired low-frequency hearing may be due to the poor timing accuracy in representing the low-frequency information. Kumar and Jayaram (2006) opined that the longest auditory nerve fibers which innervate the apical region are more prone to get disrupted due to the longer course. Shortest fibers are those which exit from the second half of the cochlea, mediate mid frequency and the basal part has the length in between the former two fibers. Hence, mid frequencies are less affected compared to low and high frequencies (Starr et al., 2001).

Temperature-dependent disorder of auditory function is also reported in ANSD. It is reported to be caused due to conduction block rather than disruption of timing (Marsh, 2002). This kind of pathology is consistent with demyelinating neuropathies (Starr et al., 1998). Starr et al recorded nerve conduction velocity on sural, peroneal and median nerve on both sides at normal body temperature and also at 39º C. The results showed a normal velocity at increased temperature, indicating the absence of other neuropathic conditions. Authors opined that maintenance of nerve conduction in the paranoidal region of demyelinated axons is temperature dependent. With slight increment in temperature the voltage-gated Na+ channels become inactivated more rapidly compared to normal temperature, resulting in failure of impulse transmission. Moreover, authors suspect both conduction block and deafness with elevated body temperature in individuals with ANSD.

In persons with ANSD, ABR in the affected ear is either absent or abnormal because of the paucity of neural element or disruption of temporal integrity. In the former case, as in the case antineoplastic drugs (carboplatin), which cause selective damage of IHCs, volume conducted neural activity is too low to detect by scalp electrode (Rance, 2005). In the latter case, ABR is absent or grossly abnormal due to compromised neural synchrony (Berlin et al., 2001). The ABR peaks represent the synchronous spike discharge at the neural tracts whereas the cortical potentials correspond to the summation of excitatory postsynaptic potentials. The unit contribution of ABR is biphasic and of shorter duration, and hence it tends to cancel when the response occurs at a difference of fraction of milliseconds in individuals with ANSD (Kraus et al., 2000).

2.4 Psychoacoustic Abilities in ANSD

The subjects with ANSD are reported to show marked deficits in their ability to resolve rapid stimulus changes (Michalewski, Starr, Nguyen, Kong, & Zeng, 2005; Starr et al., 1991; Zeng et al., 1999, 2005). The studies that measured gap detection thresholds have shown that normal hearing individuals could perceive silent periods of less than 5 ms within a continuous signal, whereas individuals with ANSD required a gap of 20 ms or more. This inability to perceive small gaps in speech signal was reported to affect the perception of brief vowel feature such as 3rd formant onset frequency. Similarly, discrimination of manner of articulation of consonants which is based on the small difference in voice onset time is reported to be affected secondary to reduced GDT in ANSD.

Kumar and Jayaram (2005) estimated the temporal modulation transfer function in normal hearing individuals and individuals with ANSD. They reported

that individuals with ANSD required significantly higher modulation depth to detect the modulations compared to normals. Further, they found that at higher modulation frequencies, individuals with ANSD were unable to detect the modulation even with 100% modulation depth. Similarly, studies have reported that individuals with ANSD experience difficulty to follow faster and even slow (<10 Hz) amplitude envelope changes over time (Rance, McKay, & Grayden, 2004; Zeng et al., 1999; Zeng et al., 2005). It has been reported that ANSD perform poorly for the task involving timing cues and they found a correlation between the temporal processing abnormalities and the speech perception abilities. The impaired temporal processing is reported to affect the ability to cope up with the dynamic nature of speech signal causing speech perception deficits in ANSD.

Psychophysical evidence has shown that subjects with ANSD have more problems with simultaneous and non-simultaneous masking compared to normal listeners (Kraus et al., 2000; Vinay & Moore, 2007; Zeng et al., 2005). Kraus et al. (2000) and Zeng et al. (2005) studied temporal processing in individuals with ANSD using forward and backward masking experiments. Results showed that the perception of short duration signals was affected even with masker to signal delays of 100 ms whereas normal hearing subjects showed limited masking effects beyond 10 to 20 ms of the masker. When tested on masking level difference, individuals with ANSD had little or no masking release (Berlin, Hood, Cecola, Jackson, & Szabo, 1993; Starr et al., 1996). This was inferred as the inability to combine the neural code from the two ears in ANSD. Poor backward masking thresholds was seen in ANSD, indicating that they are poorer than normal at separating noise sounds in time. Kraus et al. (2000) found that persons with ANSD had poorer ability to separate a brief tone from a noise which is remote from the frequency of the tone, making them a poor

listener in the noisy environment. They are also found to show abnormal temporal measures such as GDT, TMTF (Rance et al., 2004), wider temporal window in forward-backward masking (Kraus et al., 2000: Zeng et al., 2005) and abnormal binaural processing (Zeng, Oba, Garde, Sininger, & Starr, 1999; Starr et al., 2012). The authors also opined that, in ANSD location-based binaural timing cues was poorly perceived, but the perception of inter-aural intensity difference required for the judgment of lateralization was preserved.

Kumar and Jayaram (2011) examined the effect of lengthened transition duration on speech perception and Just Noticeable Difference (JND) in transition duration of stop consonants in individuals with ANSD. Results revealed a significant difference is JND between normal and ANSD groups. Improvement in the perception of place of articulation of phonemes was noted with lengthened transition duration of the stimuli. The results of Sequential information analysis (SINFA) showed that lengthening the transition duration resulted in better transmission of the place information compared to voicing information. It was also noted that JND of individuals with ANSD was almost 3 to 4 times longer than that of normals indicating impaired temporal processing in ANSD. The authors hypothesized that lengthening the transition duration would have reduced the modulation frequency without affecting modulation depth or overall spectrogram of the signal. Moreover, individuals with ANSD have difficulty following faster modulation. Hence the reduction in modulation frequency (by lengthening the transition duration) was reported to augment their speech perception as the modulation detection is better at the lower frequency compared to higher modulation frequencies. Other studies also reported JND of individuals with ANSD to be approximately 4.5 times higher than the normals (Starr et al., 1991; Zeng, Oba, & Starr, 2001).

For the steady-state puretone of 4 kHz or higher, frequency discrimination is primarily cued by the place of excitation on the basilar membrane (Moore, 1973; 2008). On the contrary, frequencies less than 4 kHz are discriminated based on the temporal cues. Zeng et al. (2001) found abnormal frequency discrimination at low frequencies while the discrimination was normal at higher frequencies. Rance et al. (2004) found a strong direct relationship between difference limen of frequency and speech perception in ANSD. Abdala, Sininger and Starr (2000) generated DPOAE suppression tuning curves in individuals with ANSD and their controls, by systematically changing the level and frequency of the ipsilateral noise. The suppression tuning curve thus obtained in ANSD was similar to normal, suggesting normal cochlear level frequency selectivity in individuals with ANSD. Hence it can be inferred that individuals with ANSD exhibit normal frequency resolution, intensity discrimination, but impaired temporal resolution. On contrary, individuals with cochlear hearing loss demonstrate normal temporal resolution and impaired frequency resolution (Hassan, 2011).

2.5 Speech Perception in ANSD

The cardinal feature of ANSD is the poor speech perception that does not relate to their degree of hearing loss (Starr et al., 1996; Starr et al., 2000; Zeng et al., 2001). The poor speech perception in ANSD is known to be due to the impaired ability to process the dynamic cues of speech. Earlier studies have shown that the disrupted neural synchrony in individuals with ANSD impairs their ability to use envelope cues as well as spectral cues of speech (Rance, 2005; Zeng et al., 1999).

The speech perception in ANSD is reported to further deteriorate in the adverse listening conditions such as in the presence of background noise (Kraus et al.,

2000; Shallop, 2002; Star, Sininger, Winter, Derby, & Michalewski, 1998). The drastic reduction in the speech perception ability in the presence of noise is known to be due to the "line busy effect" in which the noise activates the auditory nerves and reduces the response to the other signals (Derbyshire & Davis, 1935; Powers, Salvi, Wang, Spongr, & Qiu, 1995; Spreng (2000). The auditory perceptual deficits in subjects with ANSD are reported to be mainly due to the disruption of temporal cues (Kraus et al., 2000; Starr et al., 1991). The perceptual deficits in ANSD are found to correlate to their abnormal temporal and masking functions (Vinay & Moore, 2007; Zeng et al., 1999).

In individuals with ANSD, fricatives are perceived better compared to the other consonant groups due to the preserved high-frequency discrimination (Hassan, 2011). The perception of nasal consonants is known to be affected in them which are attributed to their impaired ability to use low-frequency spectral cues (Narne & Vanaja, 2008). Authors also reported place errors for stops as a major concern in ANSD. This was suggested to be due to the impairment in utilizing the burst amplitude and formant transition that contribute mainly to the perception of stop consonants. Kumar and Jayaram (2011; 2013) also reported impaired perception of voice onset time, burst and formant transitions, resulting in the poor perception of stops. They attributed it to the impaired temporal processing in individuals with ANSD. Zeng et al. (1999) reported that individuals with ANSD have impaired perception of fast modulation of speech. This results in the poor perception of burst duration and transition duration which are crucial in the perception of stops.

In individuals with ANSD, synchrony at the level of eight nerves and brainstem level which play a major role in speech perception is affected. Along with

that, they are not able to make use of the neural mechanism that represents the temporal fine structure of the stimulus, which is important for speech perception in noise (Kraus et al., 2000). Difficulty understanding speech in background noise has been attributed to the impaired ability to process the envelope of the signal (Houtgast & Steeneken, 1985). The perception of auditory signals during simultaneous masking is found to be more affected in ANSD compared to normal (Kraus, 2000; Zeng et al., 2005). Excessive masking effect which is 10-20 dB higher than normal has been reported in ANSD (Kraus et al., 2000). The findings also suggested that in ears with normal OAEs, some form of central masking mechanism exists in ANSD. Overall, the forward and backward masking experiments showed that a short signal with the proximity of 100 ms of the masker is difficult to perceive in individuals with ANSD. This is likely to deleteriously affect perception of the running speech.

Typically in ANSD, speech perception is poorer than that seen in cochlear hearing loss. But not all individuals show unusually poor speech identification scores in quiet. This may be due to the fact that in some individuals with ANSD, the disease process may be less severe (Rance, 2005). Some of the factors contributing to poor speech perception in ANSD include reduced ability to follow fast and slow temporal modulation as evidenced by TMTF, reduced gap detection and affected frequency discrimination at low frequency (Rance et al., 2004; Starr et al., 1996). Rance at al. (2004) also reported a strong correlation between speech perception and temporal modulation in ANSD. Shanon, Zeng, Kamath, Wygonski and Ekelid (1995) reported that the reduced ability of individuals with ANSD to perceive cues contained in the temporal envelope results in poor speech in noise perception. They also found that the peak sensitivity for modulation detection in ANSD was -3.4 dB for individuals with SIS less than 30%, and -14.3 dB for individuals with SIS of more than 30%.

Drullman, Festen and Plomp (1994) studied the speech perception in normal by reducing the modulation depth and degrading the amplitude modulation and flattening the spectral change in the auditory stimulus. It was found that the individuals with normal hearing experience difficult to extract the salient cues for consonant-vowel distinction and spectral contrast. This was comparable to perceptual deficits seen in ANSD. Narne and Vanaja (2008) reported that in individuals with ANSD voicing cues are poorly perceived compared to place or manner of articulation. Gnanatheja and Barman (2011) studied the perception of place, manner, and voicing in cochlear hearing loss and ANSD. They reported that all the three cues are poorly perceived in ANSD compared to the cochlear hearing loss. They also reported that in individuals with ANSD, manner cues were perceived better compared to place and voicing. Rance and Barker (2008) compared the perception of vowels, diphthongs and semivowels in ANSD and cochlear hearing loss. Their results revealed that perception of vowels was similar in both the groups, whereas the perception of diphthongs and semivowels were poorer in persons with ANSD compared to cochlear loss.

Prabhu, Avilala and Barman (2011) found no difference in the perception of unfiltered and low pass filtered speech with a cutoff frequency of 1700Hz. It may be attributed to the low-frequency hearing loss in ANSD, caused by poor phase locking of low-frequency information by Type I fibers. The authors opined that greater loss at low frequency leads to increased temporal asynchrony and poor perception of low-pass filtered speech in ANSD. They concluded that individuals with ANSD may not make use of phase locking cues due to neural dys-synchrony but make use of high-frequency information for understanding speech.

2.6 Management of ANSD

2.6.1 Amplification devices

As far as the management is concerned, majority of the studies show that conventional amplification yields little or no benefit for ANSD (Berlin, Hood, Hurely, & Wen, 1996; Hood, Wilensky, Li, & Berlin, 2004) although some studies have reported just adequate aided benefit among a few clients (Cone-Wesson, Rance, & Sininger, 2001; Rance et al., 1999). Berlin (1999) advised not to use hearing aid until OAEs diminish. But there are studies that report of normal OAEs even after long-term hearing aid usage (Katona, Büki, Farkas, Pytel, Simon-Nagy, & Hirschberg, 1993; Doyle et al., 1998; Rance et al., 1999; Berlin et al., 2000; Starr et al., 2000; Lee, McPherson, Yuen, & Wong, 2001; Sininger & Oba, 2001).

Studies have reported improvement in speech perception with hearing aid even in individuals with late-onset ANSD (Mathai & Yathiraj, 2013; Vanaja & Manjula, 2004). However, Plomp (1988) found that conventional hearing aids with nonlinear amplitude compression reduces the amplitude fluctuation and in fact may deteriorate the speech perception in ANSD. Cone-Wesson et al. (2001) reported that the unaided performance-intensity function (PIPB) in ANSD provides insight into the condition and also helps in better management. A flat PIPB and rollover indicates limited benefit with amplification devices while a rising PIPB function indicates improvement with amplification devices (Cone-Wesson et al., 2001; Mathai & Yathiraj, 2013). The use of frequency transposition in hearing aid is one of the proposed suggestions to minimize the frequency discrimination deficits in ANSD (Zeng et al., 2001). As the discrimination of low-frequency sounds is affected to a greater degree compared to the higher frequency, the authors suggested that filtering

the low-frequency sounds or transposing the acoustic signal to high frequency may improve speech perception in ANSD.

Prabhu and Barman (2017) studied the effect of low-cut modified amplification and channel free hearing aid on the speech perception of ANSD. This was done in comparison to conventional multichannel compression hearing aids. Results showed improved performance in low-cut modified amplification compared to multichannel compression hearing aids. They also found improved performance in channel free hearing aids compared to multichannel compression hearing aids. Channel free hearing aids were reported to be more beneficial in individuals with ANSD who had good speech discrimination compared to individuals with poor discrimination.

Vanaja and Manjula (2004) also reported that individuals with ANSD who had a higher amplitude cortical response show better speech perception and benefitted with hearing aids compared to those with lesser amplitude cortical potential. On the contrary, Hood (1999) had reported a client with Charcot Marie tooth disease who had good CAEP, robust OAE and poor ABR with moderate to severe hearing loss did not find a benefit with hearing aids. Therefore, one can infer that a clear cortical response may not always be predictive of good prognosis with hearing aids.

In individuals with ANSD who do not benefit with conventional hearing aids, Frequency Modulated (FM) systems is an alternative, particularly in noisy environments. Rance et al. (2010) has shown better benefit with FM systems in individuals with ANSD compared to conventional hearing aids.

Irrespective of the degree of hearing loss, ANSD who have not shown improvement in learning language through hearing aids and auditory verbal therapy,

cochlear implants should be considered as an alternative management strategy (Berlin et al., 2003). Cochlear implant has been reported to be a successful option in ANSD although the outcome varies significantly across individuals (Miyamoto et al., 1999). It was noted that electrical stimulation can actually restore neural synchrony in case of ANSD (Zhou, Abbas, & Assouline, 1995), promote neural endurance (Araki, Kawano, Seldon, Shepard, Funasaka, & Clark, 1998) and restore temporal coding (Shannon, 1993).

Santarelli, Scimemi, Monte, Genovese and Arslan (2006) reported a case of ANSD caused by systemic sclerosis in which the lesion was assumed to be involving the distal portion of auditory nerve fibers and/or synapses with inner hair cells. Authors recorded transtympanic electrocochleography (ECochG) with clicks stimuli, one month prior to cochlear implantation. Electrically evoked neural response was also obtained through cochlear implant stimulation after the implantation. The ECochG recordings showed the presence of the cochlear microphonics with normal amplitude and compound action potential. Compound action potential was obtained only at high stimulation intensity, while the electrically evoked neural response was clearly identifiable at all the recording sites during neural response telemetry. They concluded that the synchronous neural discharge in ANSD could be achieved by electrical stimulation by means of the cochlear implant.

Studies have shown that one-third of individuals with ANSD meet the candidacy criterion for cochlear implantation (Trautwein, 2002). More recently, children and adults with ANSD are being implanted, provided they show good electrical ABRs during the candidacy assessment. Shallop et al. (2001) studied the outcome of multichannel cochlear implantation in 5 individuals with ANSD. Their results indicated that neural synchrony was restored in these patients as evidenced by

the presence of electrical-ABR before implantation. It was found that most of the children with ANSD showed improvement in listening and communication skills after implantation. More importantly, the outcomes in these cases are reported to be similar to that of cochlear hearing loss (Trautwein et al., 2001).

Before prescribing cochlear implants for the individuals with ANSD it is mandatory to make sure that the lesion is pre-synaptic for the successful outcome (Buchman, Roush, Teagle, Brown, Zdanski, & Grose, 2006). Studies have reported that if the lesion is presynaptic or endocochlear, as seen in OTOF mutation, good prognosis can be expected from cochlear implants in individuals with ANSD (Varga, Kelley, Keats, Starr, & Leal, 2003). However if the pathology is at the postsynaptic site, i.e. at the auditory nerve itself, the electrical signal may not be able to propagate through the nerve, depending on the severity of the problem (Jeong, Kim, kim, Bae, & Kim, 2007). Currently there are limited noninvasive methods to distinguish between pre and post synaptic form of ANSD.

Madden, Rutter, Hilbert, Greinwald Jr and Choo (2002) suggested that in case of young children with ANSD caused due to hyperbilirubinemia, chances of spontaneous recovery by around 18 months are more and the improvement in hearing is attributed to the neural maturation. Hence cochlear implantation should be suggested only if no further improvement is observed on repeated tests during the first years of life. However, the findings of the earlier studies reveal that cochlear implant usually benefits the ANSD although a few cases have been reported to show limited benefit.

Current trends in cochlear implants incorporate higher rate of stimulation to provide cues for encoding temporal cues (Zeng, 2004). Among individuals with

ANSD caused due to demyelination or axonal loss, a higher rate of stimulation may have an adverse effect such as neural fatigue or conduction block (Stephanova, Daskalova, & Alexandrov, 2004). The decline in speech perception at higher presentation rate can be attributed to forward and backward masking by the neighboring speech elements (Nagarajan et al., 1998). Individuals with ANSD who were mapped with ACE (stimulation rate of 900 Hz/channel) performed poorly initially and improved subsequently when the stimulation rate was reduced to 720 Hz /channel. Hence a lesser rate of stimulation may be the choice for implanted individuals with ANSD.

2.6.2 Signal enhancement strategies

Based on the psychoacoustical abnormalities of individuals with ANSD, attempts have been made to advent different signal enhancement strategies to improve speech perception (Hassan, 2011; Mathai & Yathiraj, 2013; Kumar & Jayaram, 2011; Narne & Vanaja, 2008). The successful strategies are suggested to be incorporated as digital algorithms in the hearing aids.

Kumar and Jayaram (2006) lengthened the burst, transition and the VOT of various stop consonants according to the just noticeable differences and found that the perception improved with lengthening of these cues. But the extent to which the cues need to be lengthened varied across individuals, across cues and also across speech sounds. Therefore, such temporal enhancements, although beneficial, are not practical with hearing aids in the present day where real-time processing is necessary.

Narne and Vanaja (2008) studied the amount of information transmitted in the envelope enhanced condition. It was reported that when the signal bandwidth was 3-30 Hz, it transmitted the maximum information compared to other bandwidth.

Moreover, it was reported that in the envelope enhanced condition, manner and place cues were better transmitted compared to voicing. They found an improvement of up to 36% and reported that algorithm with real-time enhancement of the envelope is practical to be included in the hearing aids. The improvement was significant for a moderate degree of impairments, but improvement was negligible for the profound degree of hearing impairment.

Vasistha and Barman (2012) compared the effect of companding on speech perception in normal hearing individuals and individuals with ANSD. Speech perception was assessed both in quiet, and noise at different SNR. The results revealed improvement in speech perception. The companding is expected to enhance the spectral and temporal contrasts and therefore facilitate individuals with ANSD in their speech perception. The improvement was noticed both in quiet and noisy conditions.

Mathai and Yathiraj (2013) investigated consonant perception in ANSD in different vowel contexts (/a/, /i/ & /u/) using VCV syllables. The consonant perception was assessed in the unprocessed and three temporally stretched conditions (25%, 35% & 50%). The results showed that the perception of consonant was better in the context of /a/ and /u/ as compared to /i/. Moreover, it was also reported that place and voicing cues were better perceived with /a/ and /u/ contexts than that with /i/. Among the stretched condition, 25% of signal stretching yielded maximum scores in speech identification. Studies have shown that speech intelligibility in ANSD and cochlear loss can be improved using clear speech, which includes reduced speaking rate, enhanced temporal modulation, expanded voice pitch range and vowel space (Krause & Braida, 2002). Researchers also reported that clear speech enhances speech

perception in ANSD due to the enhanced envelope of speech, enhanced consonant energy and also due to improved spectral contrast (Zeng & Liu, 2006).

2.6.2 Training based management of ANSD

Structured auditory training was also found to be beneficial in individuals with ANSD. Yadav and Yathiraj (2010) studied the effect of fine-grained speech identification training on temporal cues and speech perception. Training was shown to improve discrimination of voiced-voiceless contrast, mid versus high-frequency vowels and speech identification of words. This, in turn was shown to improve speech perception. Tallal et al. (1996) suggested intensive training with the acoustically modified speech to improve speech perception in ANSD. However this is a grey area and warrants more studies to validate the training related improvements in individuals with ANSD.

2.7 Auditory-visual Perception of Speech

Rosenblum (2005) has reported that human speech is a multimodal function, usually comprehended by hearing as well as lip reading. Reduction in either external redundancy (noisy situation or reverberation) or internal redundancy (due to hearing impairment) impairs the perception of speech through the auditory mode (Walden, Busacco, & Montgomery, 1993; Anderson, 2006). In such instances where the auditory cues are distorted, visual input improves speech intelligibility by providing the missing cues (Tye-Murray, Sommers, & Spehar, 2007; Munhall, Kroos, Jozan, & Vatikiotis-Bateson, 2004; MacLeod & Summerfield, 1987). Researchers have reported that bimodal presentation of a speech signal yields more benefit when the auditory stimuli are degraded (Sumby & Pollack, 1954; Neely, 1956; Erber, 1969; Grant & Seitz, 2000; Rudmann, Mc Carley, & Kramer, 2003; Bernstein, Auer, &

Takayanagi, 2004; Ross, Saint-Amour, Leavitt, Javitt, & Foxe, 2007). Sumby and Pollack (1954) reported that the addition of visual cues improves the perception of speech in noise by an amount equivalent to a 5 to 18 dB increase in the signal-to-noise ratio (SNR) which is equivalent to 60% increase in word recognition. Erber (1969) also reported an improvement of 60% in word recognition scores from auditory-only condition to AV condition at -10 dB SNR for young adults.

The term 'auditory-visual integration' (Massaro, 1998) and 'auditory-visual (AV) benefit' (Grant, Walden, & Seitz, 1998) have been used to denote this process employed by individuals to combine the information extracted from auditory and visual sources. Anderson (2006) examined the amount of redundancy necessary for optimal AV integration by reducing the redundancy of the auditory signal. The performance of participants was explored under four conditions; degraded auditory only, visual only, degraded auditory + visual, and non-degraded auditory + visual. Listeners achieved higher performance in the auditory + visual mode in a degraded condition rather than the auditory alone or visual alone condition. Author has reported that the auditory modality gives information about voicing, place, and manner of articulation, while the visual modality gives information only about the place of articulation in speech. Studies have reported that older adults are less successful in integrating information than younger adults due to their impaired ability to combine information across two or more sensory modalities (Shop & Binnie, 1979; Plude & Doussard-Roosevelt, 1989). Moreover older adults with hearing loss relay more on the visual cues compared to those with normal hearing (Tye-Murray, Sommer, & Sephar, 2007). Literature review reveals that geriatrics are less successful in integrating auditory and visual cues during AV perception due to either degeneration in the visual modality or central auditory processing mechanism (Shoop & Binnie,

1979; Middelweerd & Plomp, 1987; CHABA, 1988; Walden et al.,1993; Cienkowski, 1999; Cienkowski & Carney, 2002; 2004). In instances where information in auditory modality is compromised, individuals tend to depend on the visual modality.

2.7.1 Auditory visual speech perception in individuals with hearing impairment

Studies have reported that the degree of hearing loss affects the fusion of auditory and visual cues (Grant, Walden, & Seitz, 1998). They assessed integration abilities across hearing-impaired listeners using a variety of AV integration measures. Congruent and discrepant nonsense syllables were degraded using a band-pass filter bank with four non-overlapping filter bands between 300 and 6000 Hz. Congruent stimuli are described as having the auditory signal "match", or be in synchrony with, the visual articulators. Discrepant stimuli, on the other hand, are described as having the auditory signal and visual cue "out of sync". These stimuli can either be misaligned or have another auditory signal, dubbed on to a different visual cue. These degraded syllables were then presented to listeners in the auditory alone , visual alone and AV conditions. Results showed that even with an extremely reduced auditory signal, AV benefit was still significantly higher. It was reported that AV integration is independent of a person's ability to extract auditory and visual information from speech (Grant, 1998). Studies have shown little association between integration measures derived from nonsense syllable tests and those derived from sentence tests (Grant & Seitz, 1998).

AV speech perception was reported to be better and more precise compared to A-alone or V-alone modalities among hearing impaired especially in profoundly hearing-impaired individuals (Erber, 1972: Grant, Walden, & Seitz, 1998). The improvement in AV perception was seen not only during degraded acoustic

environments (caused by environmental noise, or reverberation) but also in intact speech signal. It was noted that the children with hearing impairment are able to use and combine both visual and auditory stimuli to process information (Lachs, Pisoni, & Kirk, 2001). They investigated the ability of prelingually deaf children with cochlear implants to combine perceptual information from two sensory modalities (audition and vision). It was seen that the children's performance was better with the audiovisual condition than the auditory alone and visual alone conditions.

2.8.2 Auditory-visual speech perception in individuals with ANSD

Ramirez and Mann (2005) studied the speech perception of 4 individuals with ANSD in auditory, visual and AV modalities. They reported that individuals with ANSD compensate for their perceptual deficits in quiet as well as degraded conditions by focusing on visual cues. With the visual cues, the participants showed definite improvement in identifying the place of articulation compared to their performance with auditory cues alone. Speech perception in the auditory and AV modalities was assessed in quiet, low-noise, moderate-noise, and high-noise conditions. The identification scores of syllables in the visual modality was the baseline and the identification of syllables in the other conditions was measured and compared with the baseline. Results showed that the combination of auditory cues and visual articulatory cues did not boost the speech perception of their participants above the visual baseline except at the high-noise condition. Addition of visual cues improved the perception of speech in noise in ANSD in terms of place of articulation, only for stop consonants. The lack of difference in perception between the visual and AV modalities led to the conclusion that individuals with ANSD depend exclusively on the visual articulatory cues.

Maruthy and Geetha (2011) studied speech perception of individuals with ANSD across auditory, visual and AV modalities. Stimuli used were /ba/, /da/ and /ga/ syllables. Stimuli were presented in both congruent and incongruent conditions in the AV modality. Results showed poor perception in individuals with ANSD compared to normals in all the three modalities. An impaired visual processing was also reported in individuals with ANSD compared to normals. AV modality in ANSD yielded a better perception of place of articulation compared to auditory and visual modalities.

Chapter 3

METHODS

The present study incorporated a repeated measure standard group comparison design (Jones & Kenward, 2014) to test the overall null hypothesis that there is no significant difference in the benefit yielded by acoustic enhancements and visual cues in the speech perception of persons with ANSD. The study was executed in two phases.

Phase I: Generation of test stimuli

Phase II: Assessment of syllable identification score in different test conditions

3.1 Phase I: Generation of the Test Stimuli

In the study, the participants were tested for their speech perception in Auditory only (A), Visual only (V) and Auditory-Visual (AV) modalities. Six consonant-vowel (CV) syllables which were non-meaningful in Kannada language were the test stimuli used for speech perception. The consonants in the syllables were plosives, velar /k/, retroflex /ṭ/ and bilabial /p/, and their voiced counterparts, /b/, /ḍ/ and /g/. The vowel used was /a/ in all the syllables. The choice of these syllables was based on the previous studies wherein persons with ANSD have been found to have more difficulty in processing short duration dynamic sounds due to their impaired temporal processing (Kraus et al., 2000; Zeng et al., 2005). Perception of stop consonants was reported to be more challenging compared to fricatives, affricates and nasals in these individuals (Narne & Vanaja, 2008; Ramirez & Mann, 2005; Kumar & Jayaram, 2011). Classification of the above-mentioned consonants based on their phonetic features is depicted in Table 3.1.

Table 3.1: *Classification of consonants used in the present study based on their phonetic features*

Feature	**/p/**	**/ʈ/**	**/k/**	**/b/**	**/ɖ/**	**/g/**
Manner	Plosive	plosive	plosive	plosive	plosive	plosive
Place	Bilabial	retroflex	velar	bilabial	retroflex	velar
Voicing	Unvoiced	unvoiced	unvoiced	voiced	voiced	voiced

The auditory stimuli were enhanced using two methods; envelope enhancement and companding. Visual stimuli included the video of an adult male speaker articulating the target syllables.

3.1.1 Instrumentation and software

The following equipment and software were utilized for the generation of test stimuli in the study:

a) A note book (Sony Vaio – 64-bit) with windows 7 OS, installed windows movie maker and Video pad video editor version 4.22 (NCH Software, Canberra, Australia) was used for video editing.
b) Recording, editing audio stimuli was done using Adobe Audition software version 3 (Adobe Systems Incorporated, San Jose, CA, USA).
c) Unidirectional microphone (AHUJA AUD-101 XLR) was used for audio recording.
d) Sennheiser headphone (HDA-200) was used for presentation of auditory stimulus for clarity rating.
e) MATLAB-7 (The Math Works, Natick, USA) was used for generation of acoustically enhanced syllables.

f) Sony HXR-MC2500 professional camera [Recording frame rate: X (24Mbps) 1920 x 1080/50i, 25p, 16:9, 1280 x 720/50p, 16:9] was used for video recording of the visual part of the stimulus.

3.1.2. Recording of primary auditory stimuli

The test stimuli were audio recorded using a unidirectional microphone (AHUJA AUD-101 XLR), placed approximately 6 cm away from speaker's mouth. The microphone was connected to a computer with adobe audition (version 3) software, where the recording was saved. The syllables were digitized at a sampling frequency of 44,100 Hz and 16 bit digitization.

The six syllables were spoken by five adult males, who were native speakers of Kannada. It was ensured that all the speakers had normal speech. The speakers were instructed to produce the syllables clearly at a normal conversational level, avoiding exaggeration in articulation. Each syllable was recorded three times and out of the three samples, the best audio sample in terms of the perceptual quality was selected. The duration of auditory stimuli ranged from 300 to 470 ms. The recorded syllables were normalized (root mean square normalization) in order to minimize the differences in the energy across the syllables, using adobe audition (version 3) software.

The recorded syllables were then played to 10 experienced audiologists, through Sennheiser headphones (HDA-200). The listeners were instructed to rate the clarity of the recorded syllables on a 3-point scale (1: unclear, 2: clear, & 3: very clear). Based on their ratings, only the syllables which were rated 'very clear' by all the listeners were short listed. It was found that all the six syllables spoken by one of the five speakers were rated as 'very clear' by all the listeners. Therefore, the audio

samples of that speaker were used for speech perception testing. These six recorded syllables are operationally termed as '*primary syllables'* in this study. The recording and editing of the auditory stimuli were done using Adobe Audition software version 3.

3.1.3 Generation of envelope enhanced auditory stimuli

The primary syllables were temporally enhanced using the procedure recommended by Apoux, Tribut, Debruille and Lorenzi (2004). MATLAB-7 (The Math Works, Natick, USA) was used for the purpose. The syllables were divided into four bands using band pass filters (3^{rd} order Butterworth) of 150-550, 550-1550, 1550-3550, and 3550-8000 Hz. The temporal envelopes E (t) was extracted from each band by full-wave rectification and low pass filtering (3rd order Butterworth) with a cut-off frequency of 32 Hz. This cut-off frequency was selected based on the results of the investigation done by Narne and Vanaja (2008), wherein they found that mean consonant identification score was best in this cut-off frequency. The extracted envelope was either left intact or raised to the power K, with value of K ranging from 4 to 0.3 as a function of the instantaneous envelope amplitude value (Ei). The exponent K was set in such a way that maximum expansion (K max = 4) was applied to the lowest envelope amplitude value (E min), and the maximum compression (K min = 0.3) was applied to the highest envelope amplitude value. The expression for K is given in Equation 1.

$$Ki(b) = e^{\frac{(Ei - E\,min)}{t}(K\,max - K\,min) + K\,min} \quad \text{.....Eq.(1)}$$

(Wherein, b represents a specific band, t is a constant (0.5 for each word) within the band).

The minimum envelope amplitude value (E min) was computed over the whole signal duration within the band. A correction factor was then obtained by computing the ratio of the expanded and original envelopes for each sample. The obtained correction factor was then multiplied with the original band-pass signal at each corresponding point in time, and finally the resulting bands were added to get the enhanced signal and low pass filtered (3rd order Butterworth filter) with a cut-off frequency of 8000 Hz. All the syllables were processed using this scheme and the resultant syllables are called '*Envelope Enhanced syllables'*. The RMS amplitude of the expanded signals was then equated to that of the primary signals. Block diagram showing the different stages of signal processing to generate envelope enhanced syllables is shown in Figure 3.1. The waveform of six primary and the corresponding envelope enhanced syllables used in the study are shown in Figure 3.2.

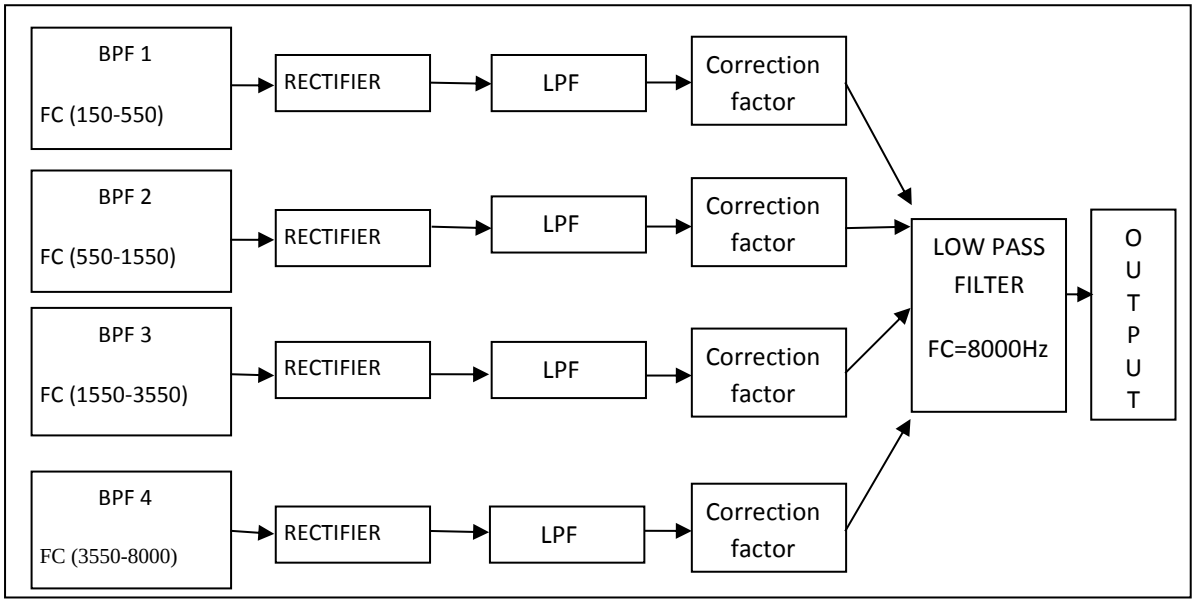

Figure 3.1: Block diagram showing the different stages of signal processing to generate envelope enhanced syllables.

Note. BPF: band-pass filter, FC: cutoff frequency, LPF: low-pass filter

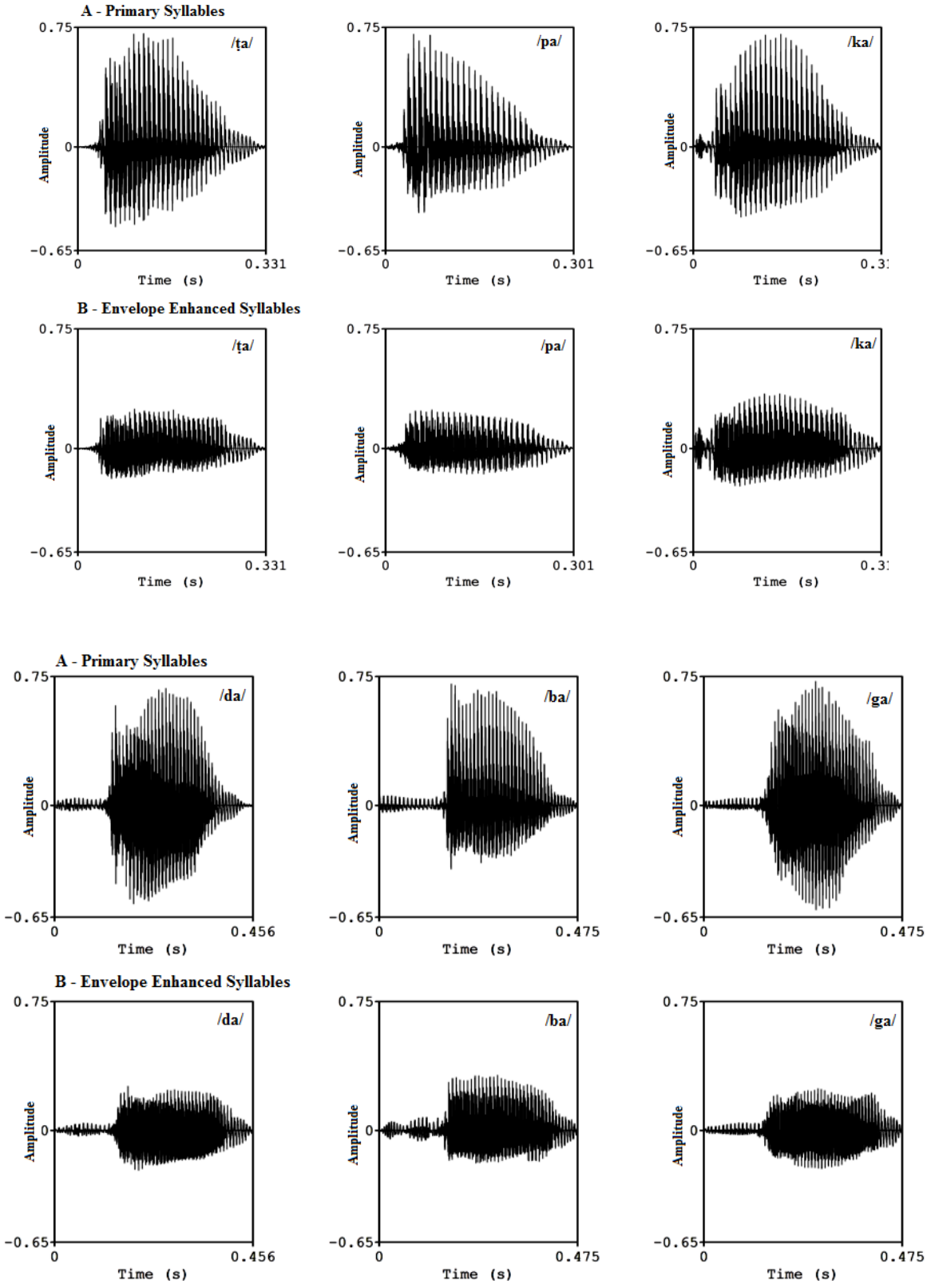

Figure 3.2: Waveforms of six primary (A) and the corresponding envelope enhanced syllables (B) used in the study.

3.1.4 Generation of companded auditory stimuli

The primary syllables were spectrally enhanced through companding as described by Turicchia and Sarpeshkar (2005). MATLAB-7 software was used for the purpose. The companding strategy incorporates a non-coupled filter bank and compression-expansion block. The channels in the companding strategy had a relatively broad pre-filter, a compression block, a relatively narrow post-filter and an expansion block. The center frequency that was logarithmically spaced to cover the desired spectral range was kept the same for the pre and post filter in each channel.

In the process of companding, initially the input syllable was divided into several frequency channels by a bank of relatively broad band-pass filters. The signal within each channel was subjected to amplitude compression. The amount of compression required was determined by the output of envelop detector (ED) and the compression index (n1). The value of n1 was 0.3. This provides third root compression on the input signal in the compression block. Then the output was passed through a relatively narrow band-pass filter G, before the signal was expanded. The n2 is the expansion index having a maximum value of 1. Whenever n2 is equal to 1, the expansion block cancels the effect of the compression block and the channel becomes linear on the time-scale of the envelope-detector dynamics. When the value of n2 is less than 1 and greater than zero then the channel does syllabic compression with an overall compression index of n2. The expansion happens in the expansion block only if n2 is more than n1. The gain of the expansion block depended on the corresponding ED output and the ratio n2/n1. The RMS amplitude of the enhanced signals was then equated to that of the original signals. The output of all the channels was summarized to obtain the processed signal. The resultant syllables are called '*companded syllables*' in this study. Block diagram showing the different stages of

signal processing to generate companded syllables is shown in Figure 3.3. The comparison of spectra of the six companded syllables against that of the respective primary syllables is shown in Figure 3.4.

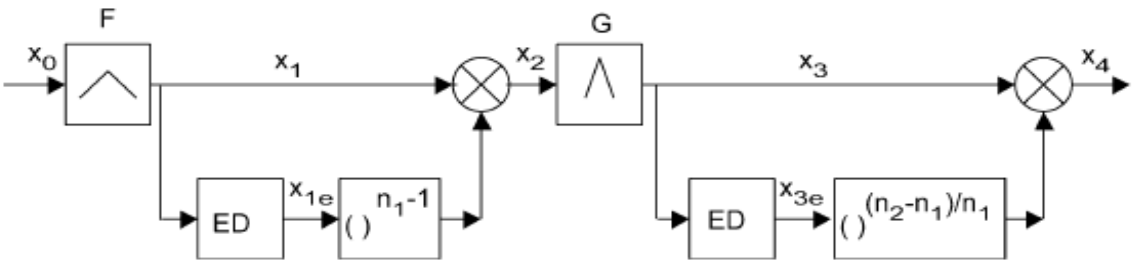

Figure 3.3: Block diagram showing the different stages of signal processing used to generate companded syllables (Turicchia & Sarpeshkar, 2007).

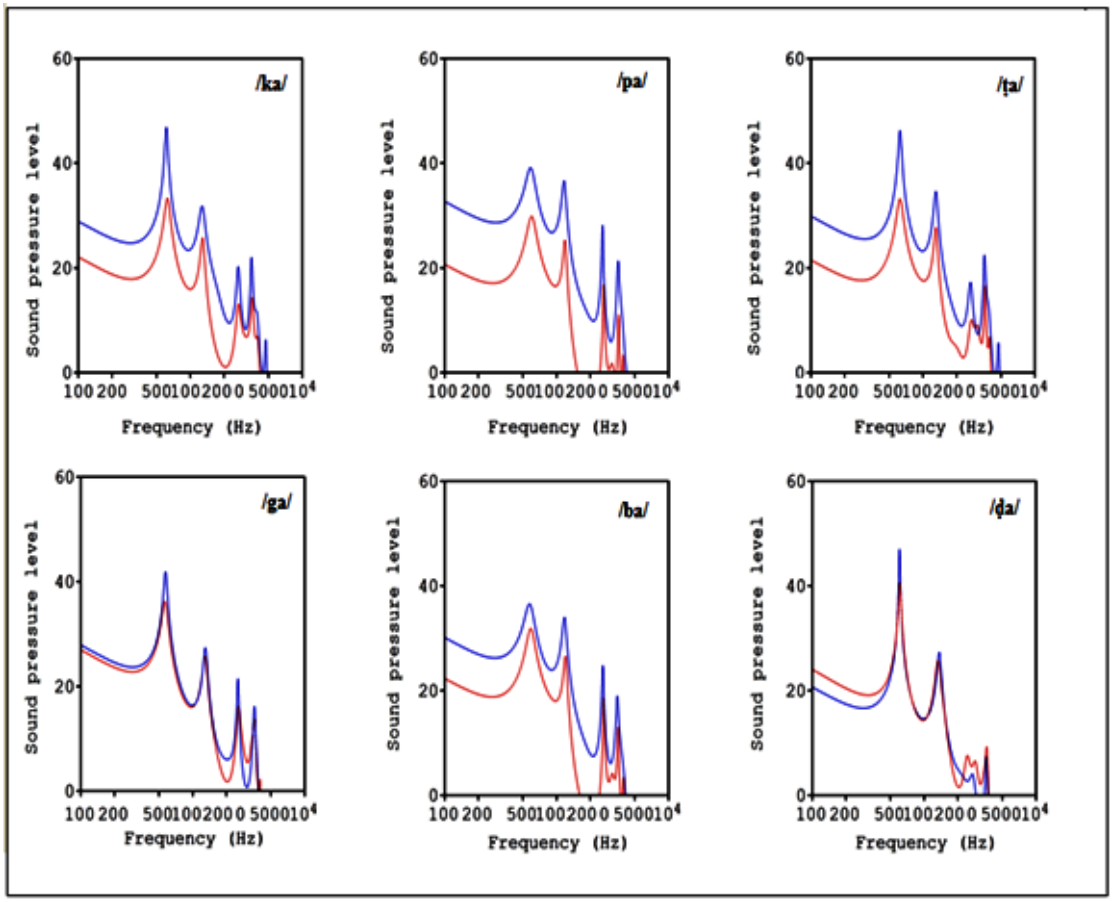

Figure 3.4: Spectra of the six companded syllables (blue color) against that of the respective primary syllable (red color).

3.1.5 Recording of visual stimuli

The close-up video of an adult male uttering the six syllables served as the visual stimuli. Video recording was done in the same individual whose audio samples got the highest clarity rating. The video was recorded by a professional videographer using high definition Sony HXR-MC2500 professional camera [Recording frame rate: X (24Mbps) 1920 x 1080/50i, 25p, 16:9, 1280 x 720/50p, 16:9]. The recording was done with appropriate lighting and a white screen was used as a background. For video recording, the video camera was kept on a tripod stand at a distance of 3 feet from the speaker. The speaker was instructed to produce the syllables clearly without exaggerating the articulation. He was also informed to minimize eye blinks and avoid head movements, during the recording. The syllables which were articulated unclearly were recorded twice. The recording was edited to improve the picture clarity and to keep the duration of each visual stimulus to 4 seconds. The initial one second was a steady video (without articulation) and the articulation began at the end of first second. After the end of articulation of the syllable, the steady video was continued till the end of the 4^{th} second. The set up used for video recording is depicted in Figure 3.5. The picture sequence showing the production of /pa/ is shown in Figure 3.6. A note book (Sony Vaio-64-bit with windows-7 OS), installed with windows movie maker and Video pad video editor version 4.22 was used for video editing. The video of each syllable was saved separately.

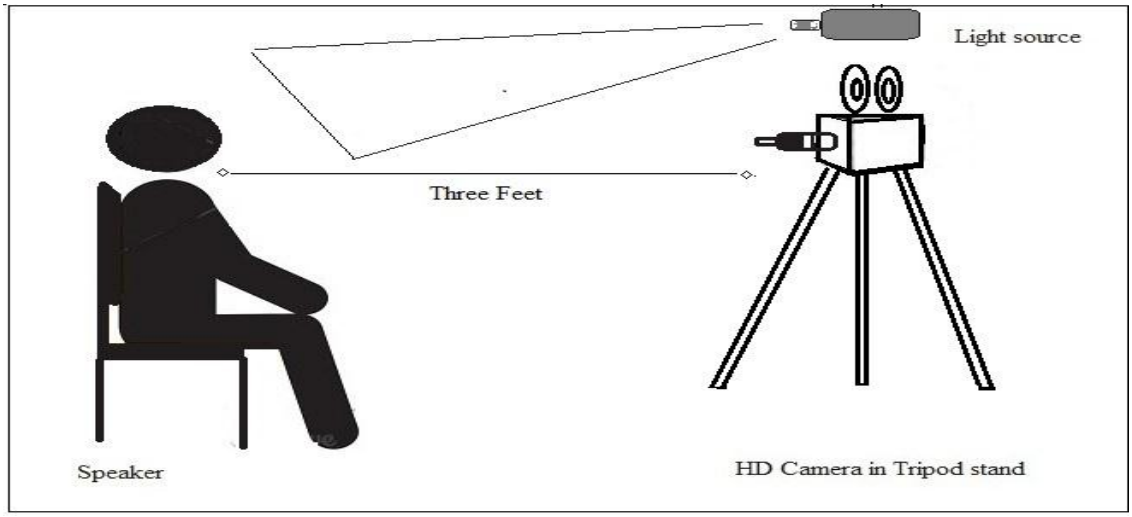

Figure 3.5: Depiction of setup used for video recording the test stimulus.

Figure 3.6: Picture sequence showing the production of /ba/ and the instantaneous timeframe of each picture in the video.

3.1.6 Generation of auditory-visual Stimuli

In order to generate the AV stimuli, each auditory stimulus was dubbed onto the corresponding visual stimulus. While dubbing, the two stimuli were time aligned so that the articulation of the speaker starts and ends with the auditory stimulus. The

release of the articulator was aligned to burst of the plosive. This was done using video pad editor software (version 4.2.2). For the enhanced AV stimulus conditions, acoustically enhanced (companded and temporal enhanced) stimuli were time aligned to the visual stimuli. The AV stimuli thus prepared were played to 5 experienced audiologists to judge the synchrony between auditory and visual components of the stimuli. All five audiologists confirmed good synchrony in all the six syllables.

3.2 Phase II: Assessment of Syllable Identification Score in Different Test Conditions

3.2.1. Test environment

All the tests (behavioural and the electrophysiological) were administered in a sound treated and electrically shielded room. The noise level in the room was as per the recommendation of ANSI S3.1 (1999).

3.2.2. Participants

The two groups of participants, a clinical group and a control group, were included in the study. ***The clinical group*** had 40 participants (19 male & 21 female) with confirmed diagnosis of Auditory Neuropathy Spectrum Disorder (ANSD). The age range of participants in the clinical group was 16 to 35 years (mean age = 24.19 years with SD being 7.4). ANSD was diagnosed by qualified audiologists based on the criteria recommended by Starr, Sininger, and Praat (2000).The demographic details and audiological profile of participants in ANSD group are given in Appendix 1.

Most of the participants with ANSD (n = 37) had sensorineural hearing loss. The severity of hearing loss ranged upto moderate degree with the puretone average ranging upto 55 dB HL (right ear mean = 33.29 dB HL, SD = 11.96 and left ear mean

= 31.04 dB HL, SD = 11.88). Among persons with ANSD three of them had normal hearing sensitivity. The speech identification scores in quiet ranged from 0% to 100% in the two ears (right ear Mean = 55.12%, SD = 31.06 and left ear Mean = 53.84%, SD = 31.74). The minimum duration of hearing loss in these participants was 1 year and the maximum duration of hearing loss was upto 20 years and all of them had acquired ANSD postlingually.

The presence of external or middle ear pathology was ruled out by an experienced Otologist. The normal middle ear function was further confirmed with Immittance evaluation. They had normal outer hair cell function revealed by the presence of transient Otoacoustic emissions (amplitude>6 dB SPL) or cochlear microphonics. They had absent ABR indicative of neuronal dys-synchrony.

All the participants had also undergone neurological examination by neurologist to rule out the presence of space occupying lesion. Neurological evaluation included clinical examination, CT scan and/or MRI as recommended by the neurologist.

The ***control group*** consisted of 40 normal hearing participants (25 male & 15 female). They were in the age range of 16 to 35 years (mean = 22.71 years & SD = 4.84). Based on a structured interview, it was ascertained that none of them had any history of speech, hearing and neurological disorders. None of them reported of difficulty in understanding speech in daily listening conditions. Their speech identification scores in quiet were 100%. The presence of normal middle ear function in both the ears was confirmed by immittance evaluation which revealed type 'A' tympanogram with the presence of both ipsilateral and contralateral reflexes. Participants in this group had normal speech, language and hearing abilities. Their

hearing thresholds were within 15 dB HL at octave frequencies between 250 Hz and 8 kHz (ANSI, 1996). Auditory brainstem responses and transient otoacoustic emissions revealed normal findings in all the participants of this group.

The participants of both the groups were native speakers of Kannada and all of them belong to Karnataka, India. They all had normal or corrected visual acuity (6/6). It was also made sure that all participants in both the groups were literate and had passed secondary school examinations. Informed consent was taken from each of the participants before carrying out the test. The method was approved by 'AIISH ethical committee for bio-behavioral research project involving human subjects' (Venkatesan, 2009) on 16.05.2013.

3.2.2 Equipment and software used

The following equipment and software were used in the phase II of the study

a) A calibrated 2 channel GSI Audiostar pro diagnostic audiometer with TDH-50 headphone was used to find out the air-conduction thresholds and also for speech audiometry. Bone conduction thresholds were estimated using Radio ear B-71 bone vibrator attached to the same Audiometer. The speakers (Seismic audio speakers SA-15 T, designed in USA) attached to the audiometer had an intensity range of -10 dB to 80 dB HL, with a flat frequency response from 125 Hz to 8000 Hz. It was ensured that the audiometer was calibrated once in three months as per the recommendations of ANSI, S3.6 (2004), during the period of the data collection
b) Calibrated GSI Tympstar (version 2) was used to carry out tympanometry and acoustic reflex evaluation. The instrument was calibrated as per the recommendations of ANSI, S3.39 (R1996).

c) Biologic AEP system (version 7.2.1) was used to record and analyze auditory brainstem response and late latency response.
d) ILO V6 Echoport (version 6.40.0.0) was used to record transient evoked oto-acoustic emissions.
e) Paradigm software (version 2.5.0.68, Perception Research Systems Incorporated, Lawrence, KS 66046, USA) was used for stimulus presentation in auditory, auditory-visual and visual modalities.
f) Samsung 21" LCD was used to display the test stimuli visually.
g) Sony Vaio laptop (Intel core 2 duo processor, 4 GB RAM) was used for the presentation of stimuli through paradigm software.
h) Feature Information Xfer (FIX) (developed by University College of London, Department of Linguistics) software was used for carrying out sequential information analysis (SINFA).

3.2.3. Test Procedure

Each participant was subjected to two types of evaluations; (1) preliminary evaluations and (2) experimental test procedure.

Preliminary evaluations

Initially, the participants were evaluated to ensure that they meet all the inclusion criteria of the study. Preliminary evaluation included case history/structured interview, pure-tone audiometry, speech audiometry, Immittance evaluation, Otoacoustic emissions (OAEs), Auditory Brainstem Responses (ABR), Auditory Late Latency Response (ALLR) and neurological evaluation.

Pure-tone thresholds were estimated using modified version of Hughson and Westlake procedure. Pure-tone thresholds were estimated at octave frequencies

between 250 Hz and 8000 Hz in air conduction, and between 250 Hz and 4000 Hz in bone conduction mode. Speech recognition thresholds were obtained monaurally in the two ears using paired-words in Kannada developed by department of Audiology, AIISH, Mysore. Speech identification score was obtained monaurally at MCL for phonetically balanced words developed by Yathiraj and Vijayalakshmi (2005). A calibrated 2-channel GSI Audiostar pro diagnostic audiometer with standard accessories was used for pure tone audiometry and also for speech audiometry. Bone conduction thresholds were estimated using Radio ear B-71 bone vibrator. It was ensured that the audiometer was calibrated once in three months as per the recommendations of ANSI, S3.6 (2004), during the period of the data collection.

Tympanogram and acoustic reflex thresholds were measured using 226 Hz probe tone. A calibrated GSI-Tympstar, version-2 middle ear analyzer was used for the purpose. Ipsilateral and contralateral acoustic reflex thresholds were measured at 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz in the two ears.

ABR and ALLR were recorded using Biologic Navigator evoked potential system (version 7.2.1). Each recording was repeated to ensure reproducibility of the responses. The stimulus and acquisition parameters used to record ABR and LLR are given in Table 3.2.

TEOAEs were measured for clicks at 80 dB +/-5 dB pe SPL using ILO V6 Echoport (version 6.40.0.0) equipment. TEOAEs were considered to be present if the waveform reproducibility was more than 75% and the overall amplitude is more than 6 dB in at least 3 consecutive frequencies of measurement.

Table 3.2: *Stimulus and acquisition parameters used to record click evoked ABR and ALLR*

Stimulus Parameters			**Acquisition Parameters**		
	ABR	**LLR**		**ABR**	**LLR**
Stimulus	Clicks	500 Hz TB	Filter	100- 3000 Hz	0.1- 100Hz
Polarity	Rarefaction	Alternating	Window	10.6 ms	533 ms
Level	90 dB nHL	80 dB nHL	Montage	Cz-M1 and Cz- M2, Nasion-ground	
Duration	100 μs	60 ms			
Number of sweeps	2000	500			
Rate	11.1/s and 90.1/s	1.1/s			
Artifact rejection	+/- 22 μV	+/- 45μV			
Transducer	ER 3A Inserts ear phones				

Experimental Test Procedure

All the participants who met the inclusion criteria were subjected to experimental test procedures. In the experimental test procedure, each participant was individually tested for their syllable identification in different stimulus and test conditions. The procedure included assessment of closed set identification of syllables in auditory alone (A), visual alone (V) and auditory-visual (AV) modalities. In the ANSD group, perception of the primary, companded and envelope enhanced syllables were assessed and AV modalities, in quiet and at 0 dB SNR conditions. (The approximate SNR in quiet condition was 30 dB) On the other hand, the control group was tested in an additional -5 dB SNR condition. This added condition was tested to avoid ceiling effects and also to compare the pattern of AV interaction in the two

groups of participants. In the V modality, perception was tested for primary syllables only in quiet in both the groups. The different modalities, stimuli and test conditions used in the present study are shown in Figure 3.7. The procedure is in line with the earlier studies wherein speech perception in the V modality was tested only in quiet whereas the A and AV speech perception were tested in different SNRs (Anderson, 2006; Ramirez & Mann, 2005; Ross, Saint-Amour, Leavitt, Javitt, & Foxe, 2006).

Stimuli were presented to the participants through Paradigm software (version 2.5.0.68) installed in a Sony Vaio laptop (Intel core 2 Duo processor, 4 GB RAM). The laptop computer was connected to the GSI Audiostar pro audiometer via 3.5 mm auxiliary cable to route audio signal to free field speaker. The speakers were kept at 45^0 Azimuth at a distance of one meter from the participants. The laptop was also connected to a Samsung 21" LCD via VGA cable to display the video part of the test stimuli. The LCD screen was kept at 0^0 Azimuth, at a distance of one meter from the participants. The wireless mouse was connected to laptop and the responses were saved to the Sony-Vaio laptop in which the stimulus was being presented. The block diagram of the experimental setup is shown in Figure 3.8. The screenshot of the syllables displayed on LCD screen is shown in Figure 3.9.

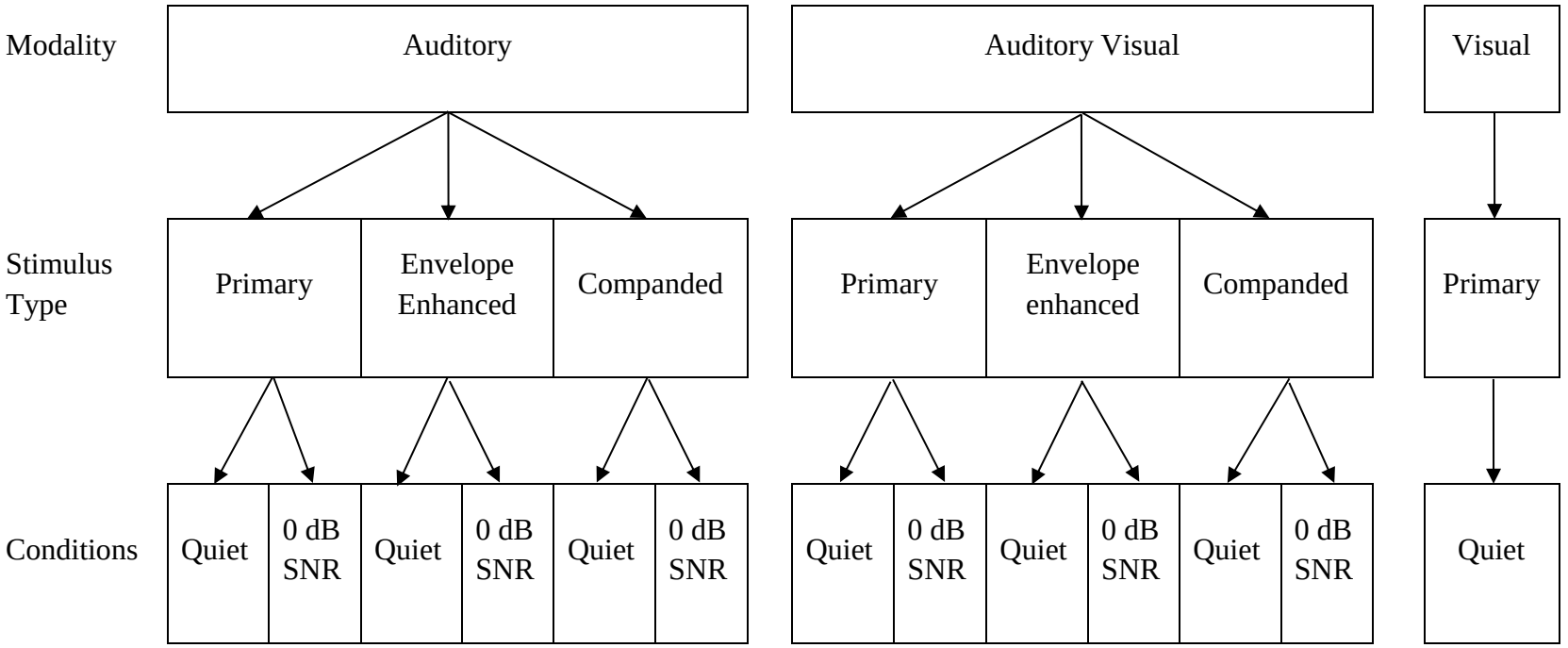

Figure 3.7: Block diagram depicting different modalities, stimuli and test conditions used in the present study. In control group, there was an additional condition of -5 dB SNR in the A and AV modalities for all the 3 stimulus types.

Note. SNR: signal to noise ratio

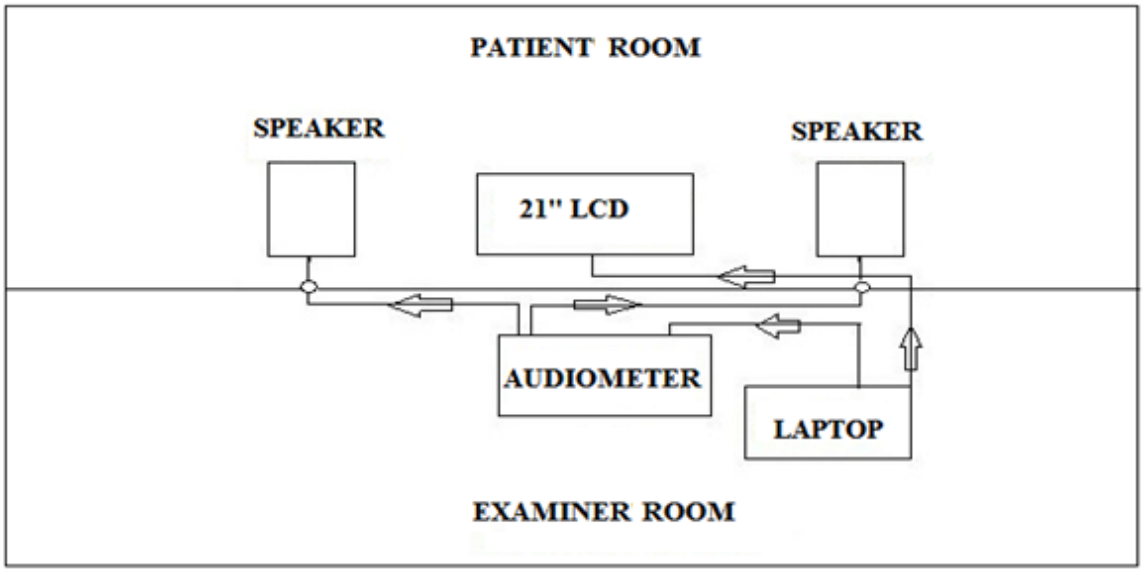

Figure 3.8: Block diagram of the experimental set up.

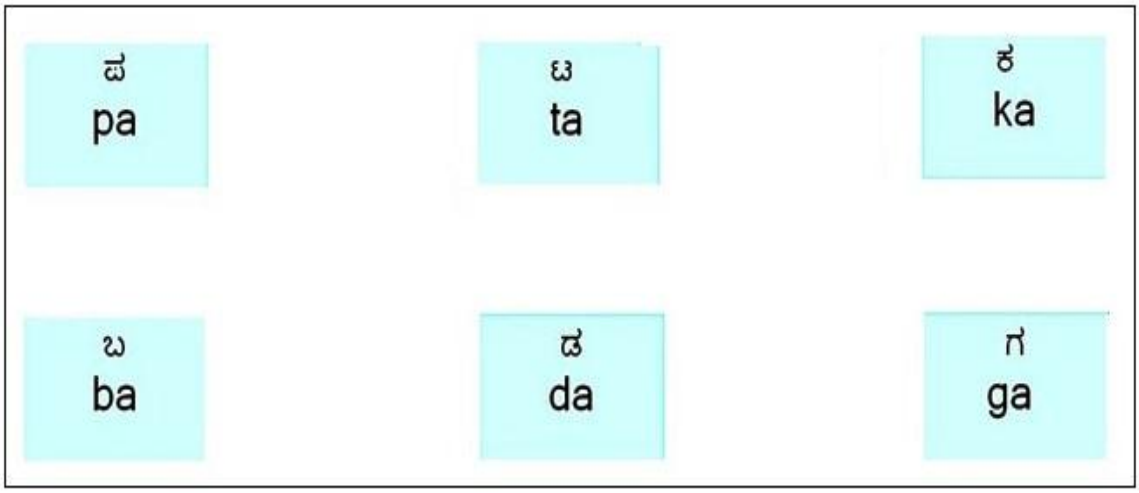

Figure 3.9: Screenshot of the syllable display on LCD screen.

The stimulus modality was randomly chosen. Just after presentation of each test stimuli all the six syllables were displayed on the LCD screen. The participants were instructed either to point or click on the syllable heard using the left mouse button. Practice trials were given to each participant to ensure that they all understood the task to be done during the test. Each syllable was presented 10 times in a random sequence. Hence normal hearing individuals heard a total presentation of 1140 and persons with ANSD heard a total of 780 presentations. Participants were given two

minute break after finishing each condition (each condition includes presentation of 6 syllables for 10 times) and the whole test conditions were undergone in the same day of testing. The stimulus conditions and the total number of stimuli presented for clinical group is represented in Table 3.3.

Table 3.3: *Total number of stimulus presented in each modality and SNR*

Group	Modality	Stimulus	SNR	Syllables	Repetition	Subtotal	Total
Control	A	3	3	6	10	540	1140
	AV	3	3	6	10	540	
	V	1	1	6	10	60	
ANSD	A	3	2	6	10	360	780
	AV	3	2	6	10	360	
	V	1	1	6	10	60	

3.2.4 Scoring of the responses

A response was considered correct if a participant selected the syllable correctly matching with the presented stimuli in each mode. Each correct response was given a score of one, and incorrect response a score of zero. The total number of correct responses of each participant was noted as the raw score.

The absolute difference between the AV modality and A modality [VG = (AV - A)] was obtained to derive the visual gain (VG). Similarly, absolute difference between the AV mode and V mode was calculated [AG = (AV - V)] to estimate the auditory gain (AG). The method used for calculation was based on the original method provide by Sumby and Pollack (1954). The maximum possible score in each condition was 60. The mean and standard deviation of AG and VG were analyzed between the two groups to compare the type of enhancement seen.

In order to determine the percentage of information transmitted for each phonetic feature in each condition, Sequential Information Analysis (SINFA) was carried out using the software, Feature Information Xfer (FIX) (developed by University College of London, Department of Linguistics) which follows the procedure described by Wang and Bilger (1973). For the purpose of analysis using SINFA, identification scores of each of these conditions were added across participants and a summed single confusion matrix was created. This was done for all conditions and then analyzed. The six CVs being tested were classified based on the place, manner and voicing features as listed in Table 3.1. This analysis was meant to derive the transmission index of each of the phonetic features in the different stimulus conditions used in the study. As the 'manner of articulation' was common across all the syllables, it was excluded from the SINFA.

3.2.5 Statistical analyses

The group data was statistically analyzed using Statistical Package for Social Sciences (SPSS, version 21). The data of the two groups was compared using nonparametric tests. The effects of stimulus, modality and condition on the syllable identification scores were tested using parametric tests. Pearson product moment correlation and linear regression were also used to test the objectives of the study.

Chapter 4

RESULTS

The study aimed to assess the relative benefits of visual cue supplementation and acoustic enhancements in improving speech perception of individuals with ANSD. Syllable identification score and its derivatives were the dependent variables. The group data were statistically analyzed to derive the effect of group (control & ANSD), stimulus (primary, companded, envelope enhanced), modality (auditory, auditory-visual & visual), and the condition (quiet & 0 dB SNR) on syllable identification scores and its derivatives.

Initially, the data of both the groups were tested for its distribution using Shapiro-wilk test of normality. It was found that all the data in the ANSD group were normally distributed, whereas the data of the control group in some of the conditions showed skewed distribution. Therefore, for between-group comparisons (in most cases, where distribution was skewed in the control group) non-parametric tests were used, while for within-group (ANSD group) comparisons, parametric tests were used.

The syllable identification scores of ANSD group were compared with that of the control group using Mann-Whitney U test. Auditory gain and visual gain between the two groups were compared using independent sample t-test and Mann-Whitney U test.

The difference in mean identification score obtained in the two modalities (A & AV), two conditions (quiet & 0 dB SNR) and three stimuli (primary, companded & envelope enhanced) in the ANSD group was tested using three-way repeated measures ANOVA. Subsequently, the effects stimulus, modality and condition were tested using one-way repeated measure ANOVA, paired t-test and Bonferroni pair-

wise comparisons. The correlation among the variables was tested using Pearson product moment correlation while the predictive variables were tested using linear regression model. The test-retest reliability was derived based on Cronbach's Alpha. There were interesting results from these analyses. The results are reported under the following major headings:

1) Comparison of syllable identification Scores and derivatives between the two groups of participants
2) Effect of stimulus, modality and condition on syllable identification scores of ANSD group
3) Effect of Stimulus, Modality and Condition on Feature Transmission Derived from SINFA, in ANSD group
4) Predictors of benefit from AV modality in ANSD group
5) Relationship of duration of hearing loss, puretone average and speech identification scores with auditory gain (AG) and visual gain (VG) scores in ANSD group

4.1 Comparison of Syllable Identification Scores and its Derivatives between Control and ANSD group

In this section, the syllable identification scores of ANSD group were compared with that of control group using Mann-Whitney U test. This section addresses the first two objectives of the study.

4.1.1 Comparison of syllable identification scores between control and ANSD groups

The mean, standard deviation, median and range of syllable identification scores obtained for the different stimulus types, modalities and conditions in the two groups is given in Table 4.1.

The mean identification scores in the ANSD group were lower than that in the control group. This was true for all the three stimuli, in all the three modalities and in both the conditions. Figure 4.1 and 4.2 shows individual identification scores of the two groups of participants for the primary stimulus in the three modalities (A, AV & V) in quiet and 0 dB SNR conditions respectively. The scores of ANSD have been depicted with reference to their scores for the primary stimulus in AV modality in ascending order. The two figures show that most of the participants of ANSD group scored lower than all the participants of control group in A and AV modalities. However in V modality even though, many participants of ANSD group performed poorer than that of the control group, the vice versa was also seen in some of the cases.

Table 4.1: *Mean, standard deviation (SD), median and range (minimum-maximum) of syllable identification scores obtained in the two groups of participants in the A and AV modalities for the three stimulus type, in quiet and 0 dB SNR*

Modality	Stimulus	Condition	Group	Mean	SD	Median	Min-Max
Auditory	Primary	Quiet	Control	59.50	0.96	60.00	58-60
			ANSD	34.15	12.41	35.00	10-60
		0 dB SNR	Control	51.55	05.95	52.00	39-60
			ANSD	18.02	07.34	16.50	06-36
		-5 dB SNR	Control	44.67	07.88	42.00	19-39
	Companded	Quiet	Control	58.35	02.07	59.00	54-60
			ANSD	32.80	12.14	37.00	13-53
		0 dB SNR	Control	46.42	07.94	45.50	32-60
			ANSD	17.32	07.23	16.00	04-36
		-5 dB SNR	Control	37.92	10.71	36.00	20-60
	Envelope enhanced	Quiet	Control	59.45	01.18	60.00	55-60
			ANSD	33.80	12.77	34.00	11-58
		0 dB SNR	Control	51.30	06.72	52.00	37-60
			ANSD	19.85	09.45	18.00	06-39
		-5 dB SNR	Control	41.05	11.70	41.00	20-59
Auditory-Visual	Primary	Quiet	Control	59.52	0.78	60.00	57-60
			ANSD	41.58	10.08	42.00	25-58
		0 dB SNR	Control	55.43	03.18	55.50	50-60
			ANSD	27.83	5.14	28.00	15-39
		-5 dB SNR	Control	51.27	05.39	52.00	14-60
	Companded	Quiet	Control	59.47	0.68	60.00	58-60
			ANSD	42.50	09.45	44.50	27-58
		0 dB SNR	Control	51.53	04.79	51.50	39-60
			ANSD	28.15	04.61	28.00	20-37
		-5 dB SNR	Control	47.03	06.58	47.00	35-60
	Envelope enhanced	Quiet	Control	59.28	0.91	60.00	57-60
			ANSD	40.33	09.12	40.00	29-60
		0 dB SNR	Control	54.98	04.69	56.00	48-60
			ANSD	30.03	05.25	30.50	24-39
		-5 dB SNR	Control	49.48	06.71	51.00	32-59
Visual	Primary	Quiet	Control	28.35	04.32	28.00	19-39
			ANSD	23.30	05.55	23.00	13-36

Note. Maximum possible score was 60. Min: minimum, Max: maximum

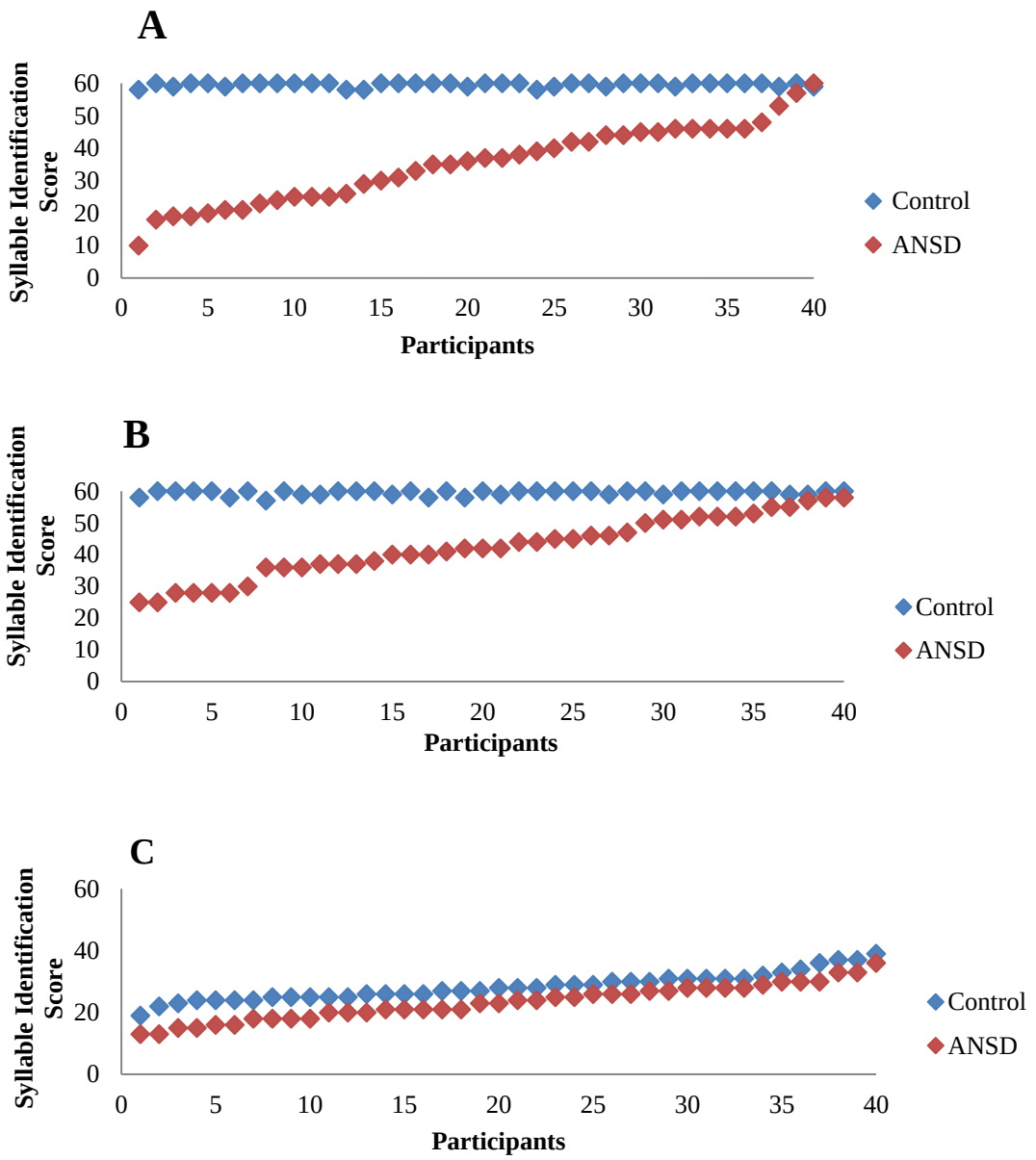

Figure 4.1: Individual syllable identification scores of the two groups of participants for the primary stimulus in the auditory (A), auditory-visual (B) and visual (C) modalities in the quiet condition. The scores are represented with reference to the scores of ANSD group in the ascending order.

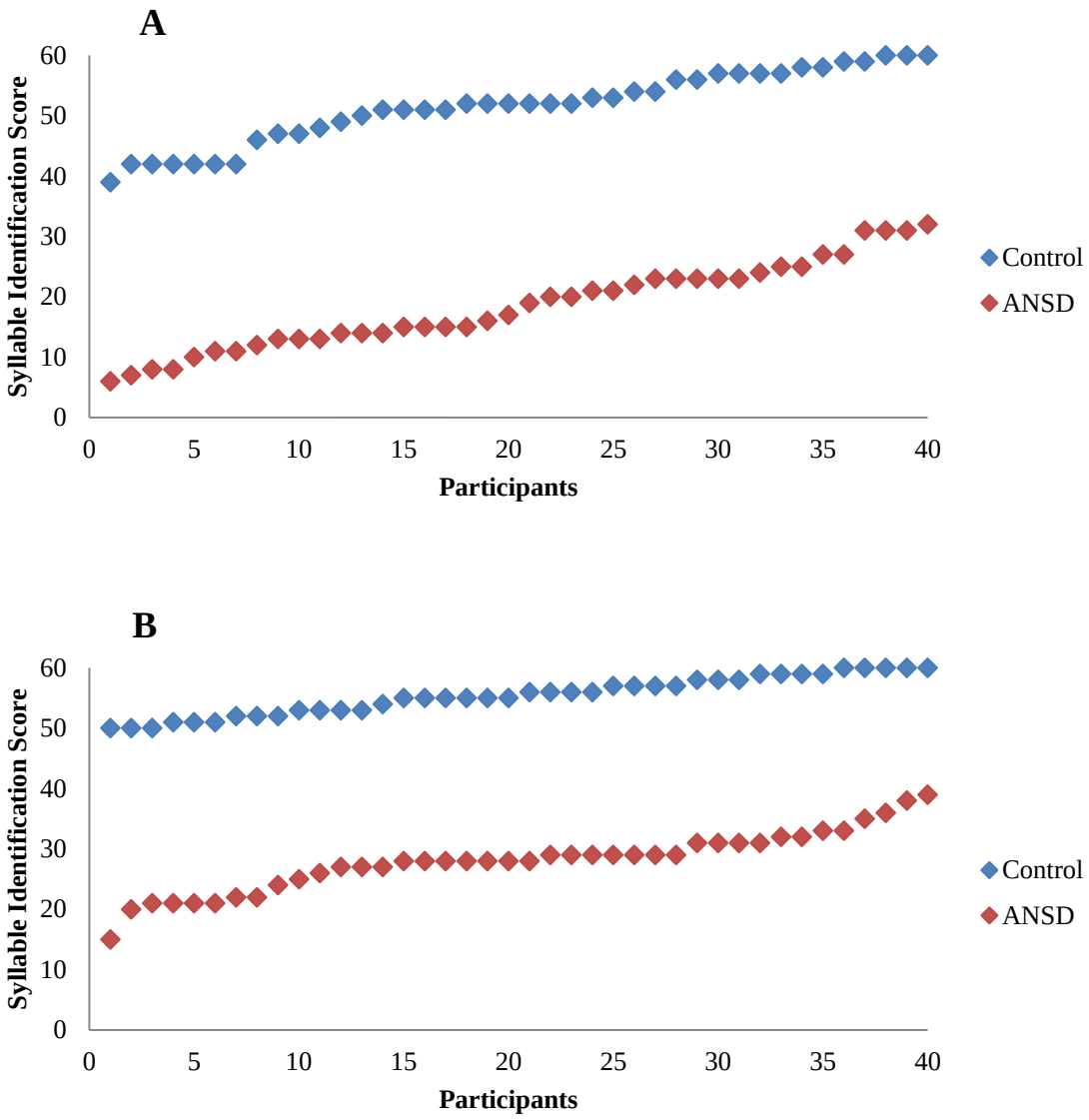

Figure 4.2: Individual syllable identification scores of the two groups of participants for the primary stimulus in the A modality (A) and AV modality (B) in the 0 dB SNR condition. The scores are represented with reference to the scores of ANSD group in the ascending order.

Figure 4.3 and Figure 4.4 show individual identification scores of the two groups of participants for the companded and envelope enhanced stimuli in A-modality in the quiet and 0 dB SNR conditions respectively. The scores of ANSD

have been depicted with reference to their scores for the companded stimulus in A-modality in ascending order.

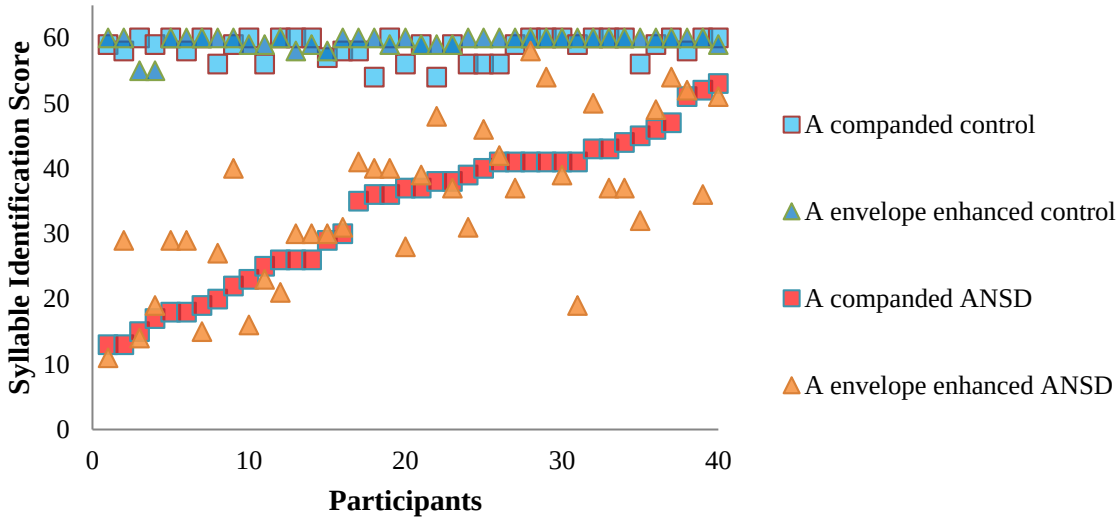

Figure 4.3: Individual syllable identification scores of the two groups of participants for the companded and envelope enhanced stimuli in quiet condition in auditory modality.

Note. A: Auditory modality, ANSD: auditory neuropathy spectrum disorder

The two figures (Figure 4.3 & 4.4) show that most of the participants of ANSD group scored lower than all the participants of control group for the enhanced stimuli (companded & envelope enhanced) in the two conditions (quiet & 0 dB SNR). The difference in score between the two groups was more pronounced in the 0 dB SNR condition compared to the quiet condition. Individual scores in each group showed minor difference between companded and envelope enhanced stimuli in quiet and 0 dB SNR conditions. In the quiet condition, a few participants scored better for the envelope enhanced stimuli compared to the companded stimuli, whereas in the 0 dB SNR condition, majority of the participants showed better scores with envelope enhanced stimuli compared to companded stimuli.

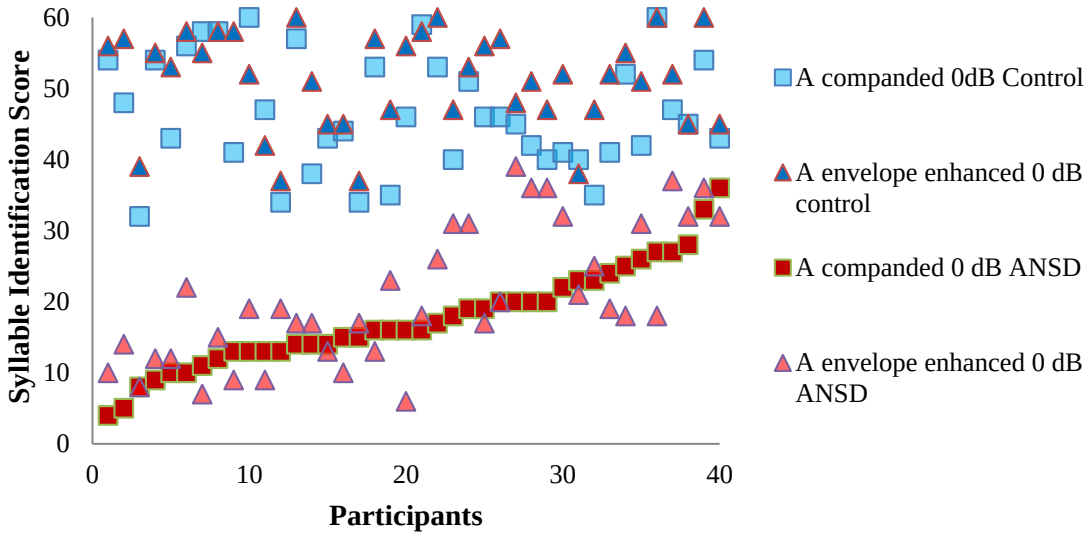

Figure 4.4: Individual syllable identification scores of the participants of the two groups for the companded and envelope enhanced stimuli in 0 dB SNR condition in auditory modality.

Note. A: auditory modality, ANSD: auditory neuropathy spectrum disorder, SNR: signal to noise ratio.

Figure 4.5 and Figure 4.6 show the individual syllable identification scores of the two participant groups for the companded and envelope enhanced stimuli in AV modality in the quiet and 0 dB SNR conditions respectively. The scores of ANSD have been depicted with reference to their scores for the companded stimuli in AV modality in the ascending order.

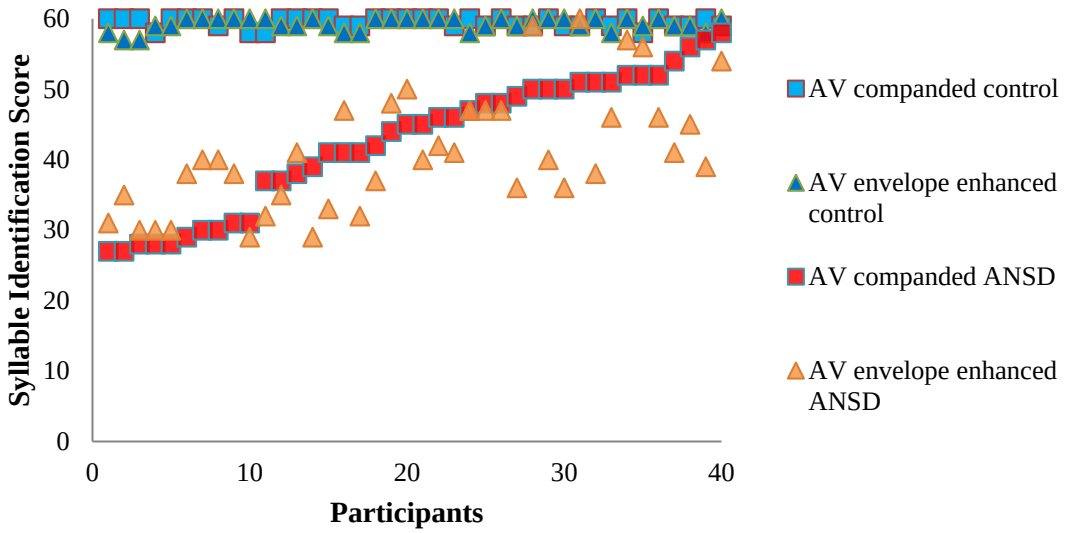

Figure 4.5: Individual syllable identification scores of the two participant groups for the companded and envelope enhanced stimuli in quiet condition in AV modality.

Note. AV: auditory-visual modality, ANSD: auditory neuropathy spectrum disorder

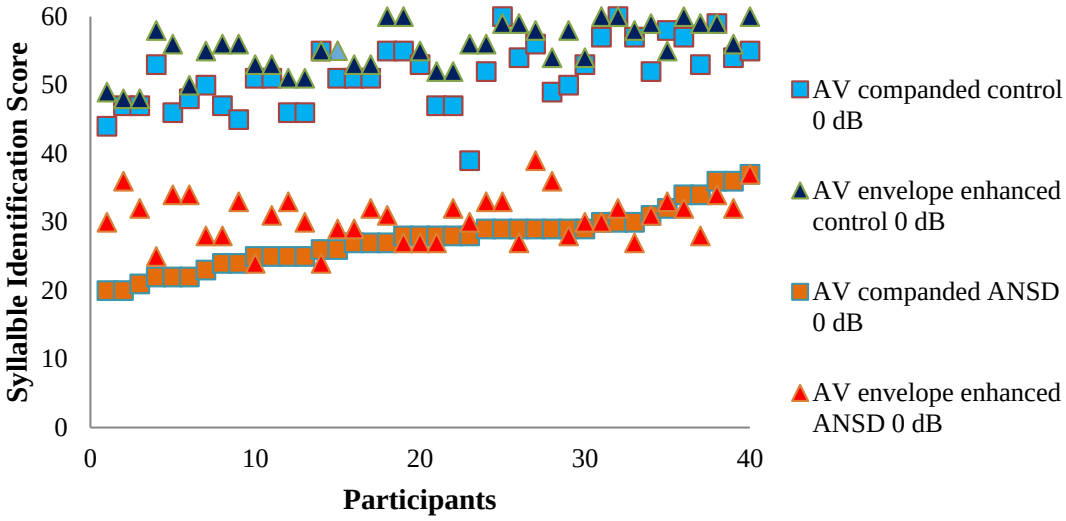

Figure 4.6: Individual syllable identification scores of the two participant groups for the companded and envelope enhanced stimuli in 0 dB SNR condition in AV modality.

Note. AV: auditory-visual modality, ANSD: auditory neuropathy spectrum disorder

The two figures (Figure 4.5 & 4.6) show that most of the participants of ANSD group scored lower than all the participants of control group for the enhanced stimuli (companded & envelope enhanced). This was true in both conditions (quiet & 0 dB SNR). The difference in score between the two groups was more pronounced in the 0 dB SNR condition compared to quiet condition.

The differences in the mean scores were tested for statistical significance using Mann-Whitney U test. Results (Table 4.2) showed a significant difference between the groups in all the three modalities, for all the three stimuli and in both the test conditions.

From Table 4.1, it can also be observed that the mean scores of control group obtained at -5 dB SNR were better than the scores obtained by ANSD group with at 0 dB SNR condition as well as quiet conditions. This was true for all the three stimuli in both A and AV modalities.

4.1.2 Comparison of auditory gain scores between the two groups

From the raw syllable identification scores obtained in A, AV and V modalities, two derivatives were obtained; Auditory gain (AG) and Visual gain (VG). The AG was derived by subtracting individual V score from the respective AV score at each SNR condition. The AG score of 0 dB SNR condition was derived by subtracting V score of quiet from AV score of 0 dB SNR. This was done for the three stimuli. Table 4.3 shows mean and standard deviation of AG in the two groups of participants for the three stimuli, in different SNR conditions.

Table 4.2: *Results of Mann-Whitney U test comparing control and ANSD groups for their syllable identification scores, in three stimuli, in three modalities and two conditions*

Stimuli	Modality	Condition	Z	*p*	*r*
Primary	A	Quiet	-7.64	<0.01	0.74
	AV		-7.79	<0.01	0.77
	V		-3.94	<0.01	0.18
	A	0 dB SNR	-7.71	<0.01	0.75
	AV		-7.71	<0.01	0.75
Companded	A	Quiet	-7.73	<0.01	0.76
	AV		-7.79	<0.01	0.77
	A	0 dB SNR	-7.64	<0.01	0.74
	AV		-7.71	<0.01	0.75
Envelope enhanced	A	Quiet	-7.84	<0.01	0.78
	AV		-7.37	<0.01	0.69
	A	0 dB SNR	-7.64	<0.01	0.74
	AV		-7.69	<0.01	0.75

Note. *r*: effect size

The mean AG score was higher in the control group compared to ANSD group with, irrespective of the conditions. In the control group, mean AG reduced in enhanced stimuli in quiet, 0 dB as well as -5 dB SNR. The AG score obtained for the three stimuli in quiet and 0 dB SNR conditions were compared between the two groups using independent sample t-test (as the data were normally distributed). Results (Table 4.4) showed a significant difference in AG between the two groups for all the three stimuli, in both the conditions.

Table 4.3: *Mean and standard deviation of AG scores in the two groups of participants for the three stimuli, in different SNR conditions*

Stimuli	Conditions	Control group		ANSD Group	
		Mean	SD	Mean	SD
Primary	Quiet	31.82	5.35	16.27	13.03
	0 dB SNR	27.70	6.07	5.25	7.07
	-5 dB SNR	22.92	6.28		
Companded	Quiet	31.15	4.90	21.63	10.07
	0 dB SNR	23.90	5.14	7.70	9.88
	-5 dB SNR	19.32	6.46		
Envelope enhanced	Quiet	30.83	4.63	19.25	8.85
	0 dB SNR	27.55	5.18	9.62	9.08
	-5 dB SNR	21.93	7.12		

Note. Maximum score is 60.

Table 4.4: *Result of independent sample t-test comparing the AG scores between the two groups*

Stimuli	Modality	*t*	*df*	*p*	*r*
Primary	Quiet	6.80	78	<0.01	1.89
	0 dB SNR	16.27	78	<0.01	3.69
Companded	Quiet	5.43	78	<0.01	1.44
	0 dB SNR	8.63	78	<0.01	2.27
Envelope enhanced	Quiet	7.31	78	<0.01	1.90
	0 dB SNR	10.74	78	<0.01	2.75

Note. *r*: effect size

4.1.3 Comparison of visual gain scores between the two groups

The individual VG scores were derived by subtracting the individual A score from the respective AV score. The VG score of 0 dB SNR condition was derived by subtracting V score of quiet from AV scores of 0 dB SNR. Mean, standard deviation (SD) and median of VG scores in the two groups of participants for the three stimuli, in different SNR conditions are shown in Table 4.5.

Table 4.5: *Mean, SD and median of VG scores in the two groups of participants for the three stimuli, in different SNR conditions*

Stimuli	Conditions	Control group			ANSD group		
		Mean	SD	Median	Mean	SD	Median
Primary	Quiet	0.05	1.24	0.00	7.30	8.50	7.00
	0 dB SNR	3.88	4.50	3.50	10.53	6.10	11.00
	-5 dB SNR	6.60	5.62	6.00			
Companded	Quiet	0.00	1.04	0.00	9.03	7.80	10.00
	0 dB SNR	4.63	6.11	4.00	10.00	7.97	13.00
	-5 dB SNR	6.60	5.62	6.00			
Envelope enhanced	Quiet	-0.20	1.68	0.00	4.90	8.48	4.00
	0 dB SNR	3.63	5.26	3.00	8.38	7.65	8.50
	-5 dB SNR	7.58	8.32	6.00			

Note. Maximum possible score is 60.

Contrary to the AG score, the mean VG score was higher in ANSD group in comparison to control group, for all the three stimuli both in quiet and 0 dB SNR conditions. The difference in VG score between the two groups was tested using Mann Whitney U test (as the data were not normally distributed). The results (Table 4.6) showed that VG scores were significantly different between the two groups for the all three stimuli, in both the conditions.

Table 4.6: *Result of Mann Whitney U test comparing VG scores between the two groups*

Stimuli	Modality	Z	*P*	*r*
Primary	Quiet	-6.11	<0.01	0.54
	0 dB SNR	-4.93	<0.01	0.30
companded	Quiet	-4.93	<0.01	0.35
	0 dB SNR	-3.23	<0.01	0.10
Envelope enhanced	Quiet	-4.25	<0.01	0.22
	0 dB SNR	-3.04	<0.01	0.12

Note. *r*: effect size

4.1.4 Comparison of feature information transmitted between the two groups for the primary stimulus

Figure 4.7 shows the total information transmitted in the two groups in the three modalities, in the two conditions analyzed by SINFA. This was only for the primary syllable. Total information transmitted is much lesser in ANSD group compared to that in control group. This was true in all the three modalities and in both quiet and 0 dB SNR. The difference in the total information transmitted between the two groups was more in A-modality compared to AV modality in both quiet and 0 dB SNR. The difference was least in V modality. The total information transmitted in the control group at -5 dB SNR was more than that of ANSD group at better SNRs (quiet & 0 dB SNR) in any of the modality.

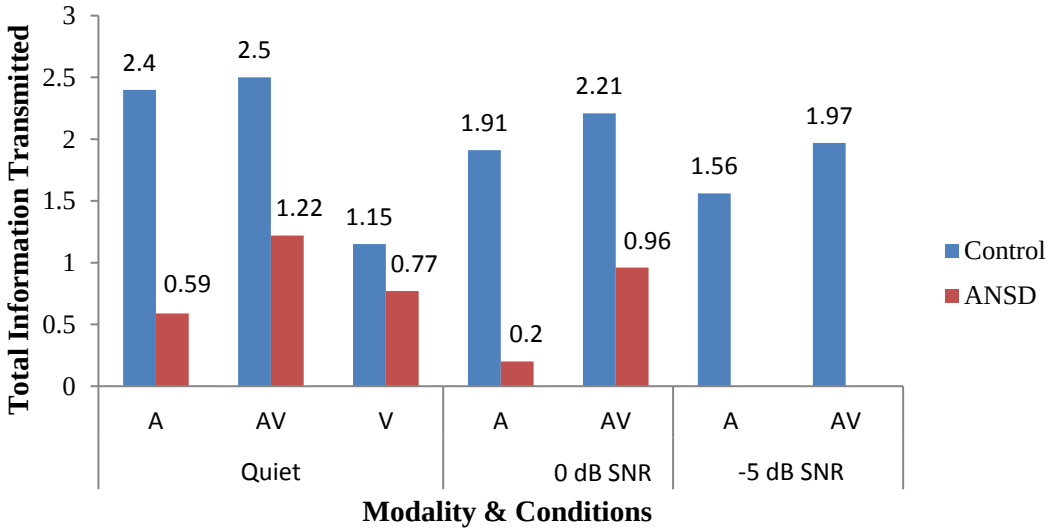

Figure 4.7 Comparison of total information transmitted in the two groups of participants in the three modalities in different conditions. The data were derived by SINFA on the scores of primary syllable.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder

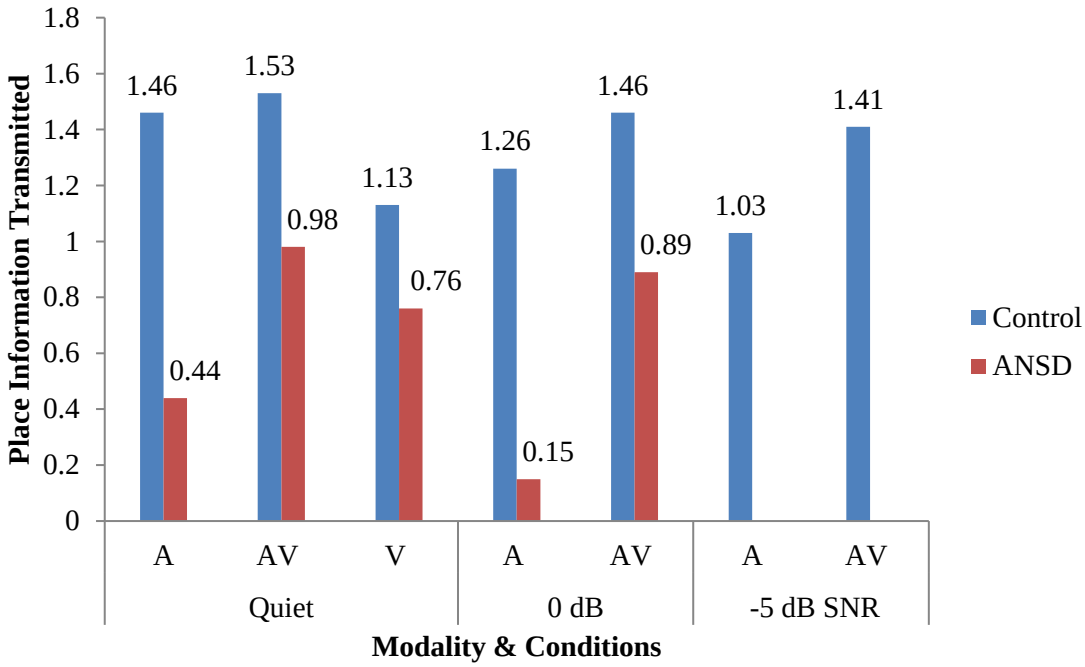

Figure 4.8: Comparison of place feature transmitted in the two groups of participants in the three modalities in different conditions for the primary stimuli.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder

The feature-wise information transmission for the primary syllable derived for place feature (Figure 4.8) and voicing feature (Figure 4.9) showed lower information transmission in the ANSD group compared to control group for both the feature.

Similar to the total information transmitted, the difference in the place feature transmission between the two groups was highest in A-modality followed by AV modality and least in V modality. The pattern was similar in quiet and 0 dB SNR conditions. The place feature transmission in the control group at -5 dB SNR was more than that of the ANSD group at better SNRs in any of the modalities.

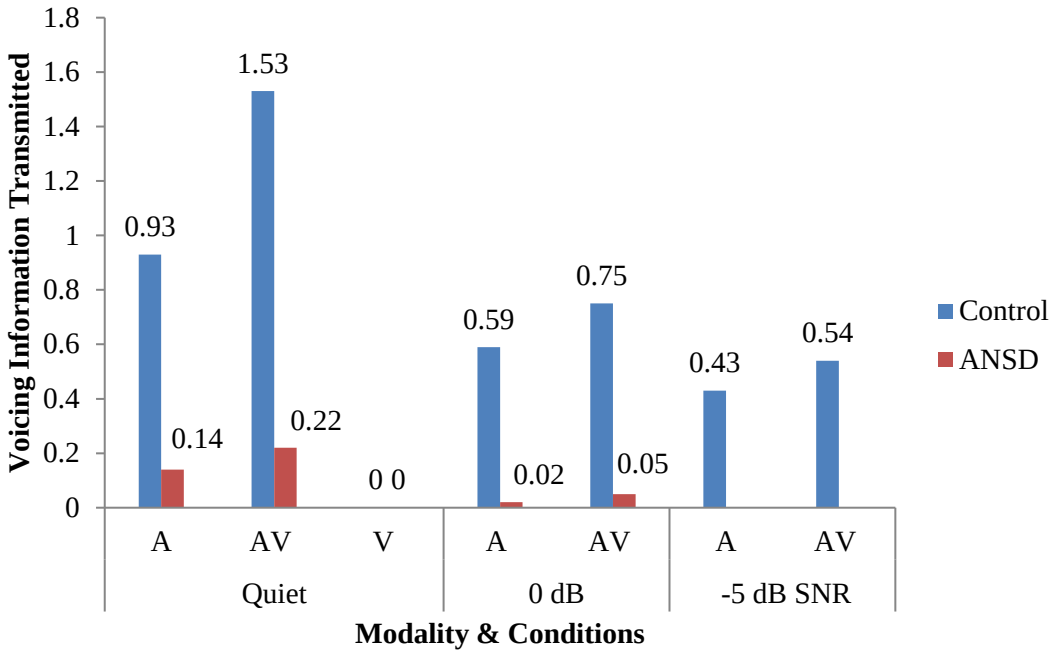

Figure 4.9: Comparison of voicing feature transmitted in the two groups of participants in the three modalities in different conditions for the primary stimuli. *Note.* A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder

On the contrary, voicing feature was not transmitted in V modality in either of the groups. In A and AV modalities, the voicing feature transmitted in ANSD group was lesser compared to control group. The voicing feature transmitted in the control group at -5 dB SNR was higher than that of ANSD group at better SNRs in any of the modalities.

4.1.5 Comparison of feature information transmitted between the two groups for the acoustically enhanced stimuli

Figure 4.10 shows the total information transmitted for the companded and envelope enhanced stimuli between the two groups in the two modalities and in the two conditions. Total information transmitted is much lesser in ANSD group

compared to that of the control group. This was true in A and AV modalities, both quiet and 0 dB SNR conditions and for the two stimuli. The difference in the total information transmitted between the two groups was more in A-modality compared to AV modality in both quiet and 0 dB SNR conditions. The total information transmitted in the control group at -5 dB SNR in each modality was more than that of ANSD group at better SNRs (quiet & 0 dB SNR) in the same modality.

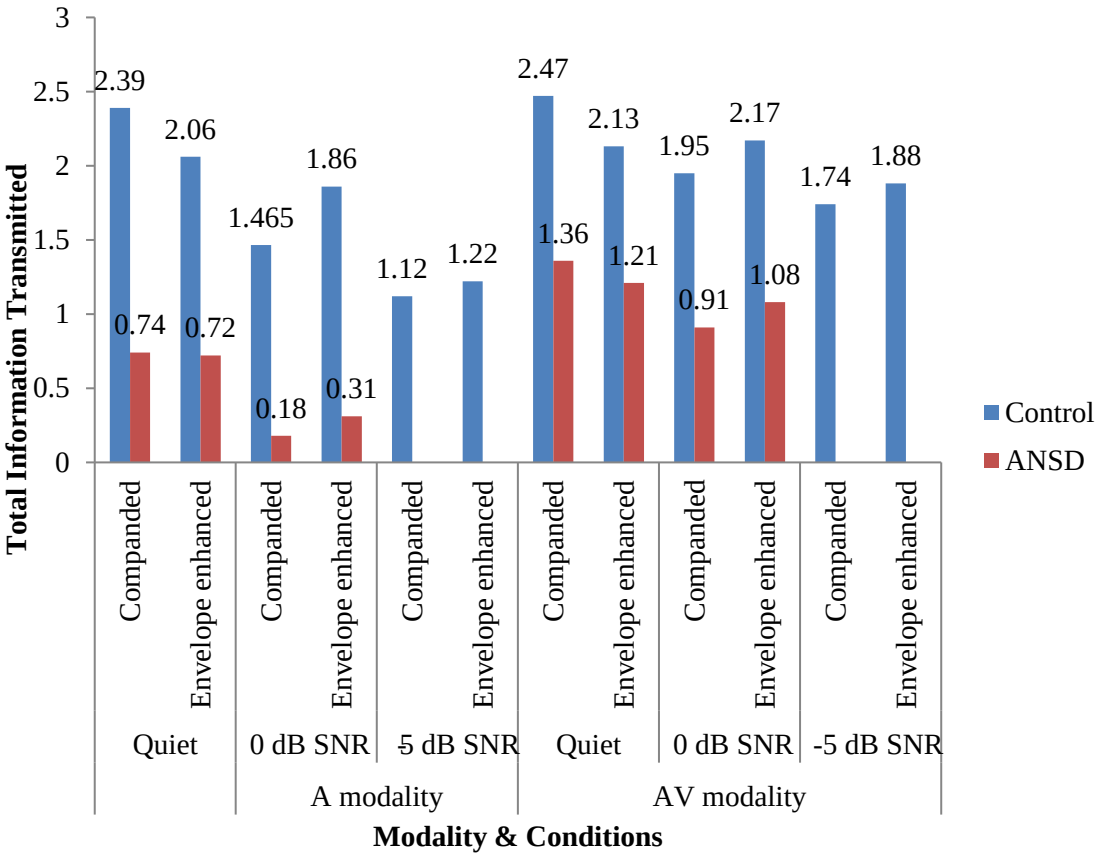

Figure 4.10: Comparison of total information transmitted in the two groups of participants for the companded and envelope enhanced stimuli in the two conditions. The data were derived using SINFA on the scores of acoustically enhanced syllable.

Note. A: auditory modality, AV: auditory-visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder

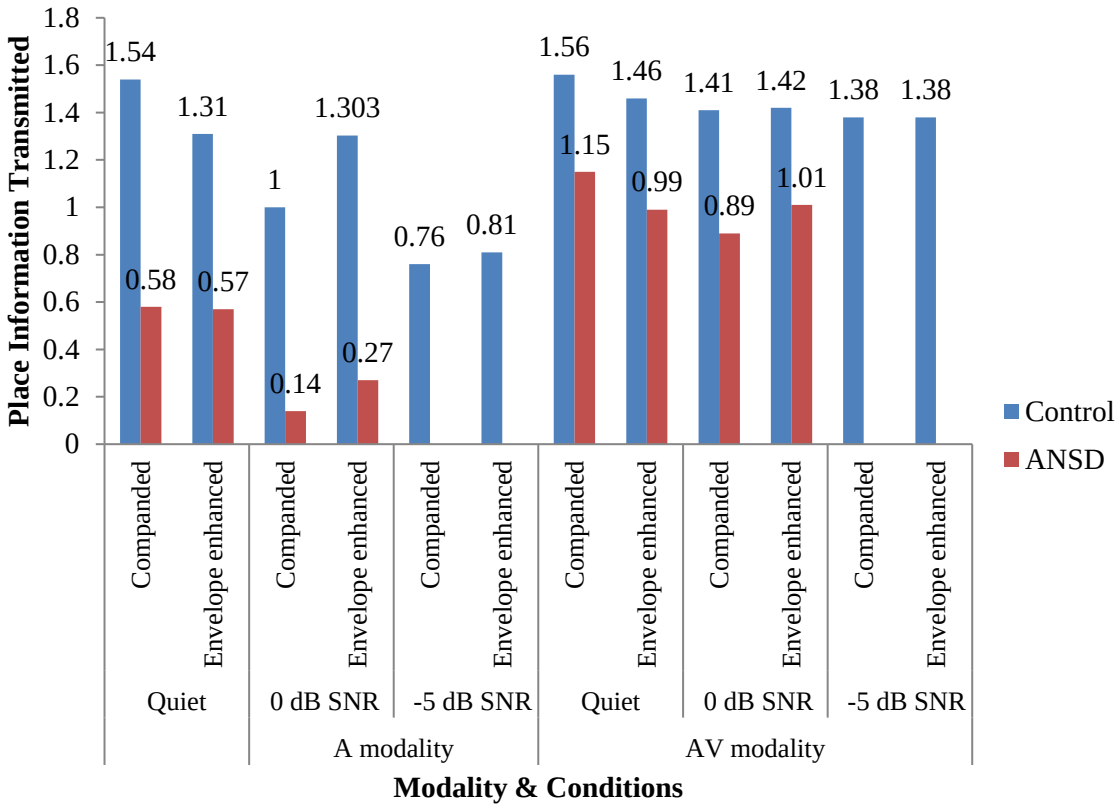

Figure 4.11: Comparison of place information transmitted in the two groups of participants for the companded and envelope enhanced stimuli in the two conditions. The data were derived by SINFA on the scores of acoustically enhanced syllables.

Note. A: auditory modality, AV: auditory-visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder.

Similar to the total information transmitted, the difference in the place feature transmission between the two groups was highest in A-modality followed by AV modality. The pattern was similar in quiet and 0 dB SNR conditions. Among the two conditions, the difference was more at 0 dB SNR compared to the quiet condition. The place feature transmission in the control group at -5 dB SNR was more than that of the ANSD group at better SNRs in any of the modalities.

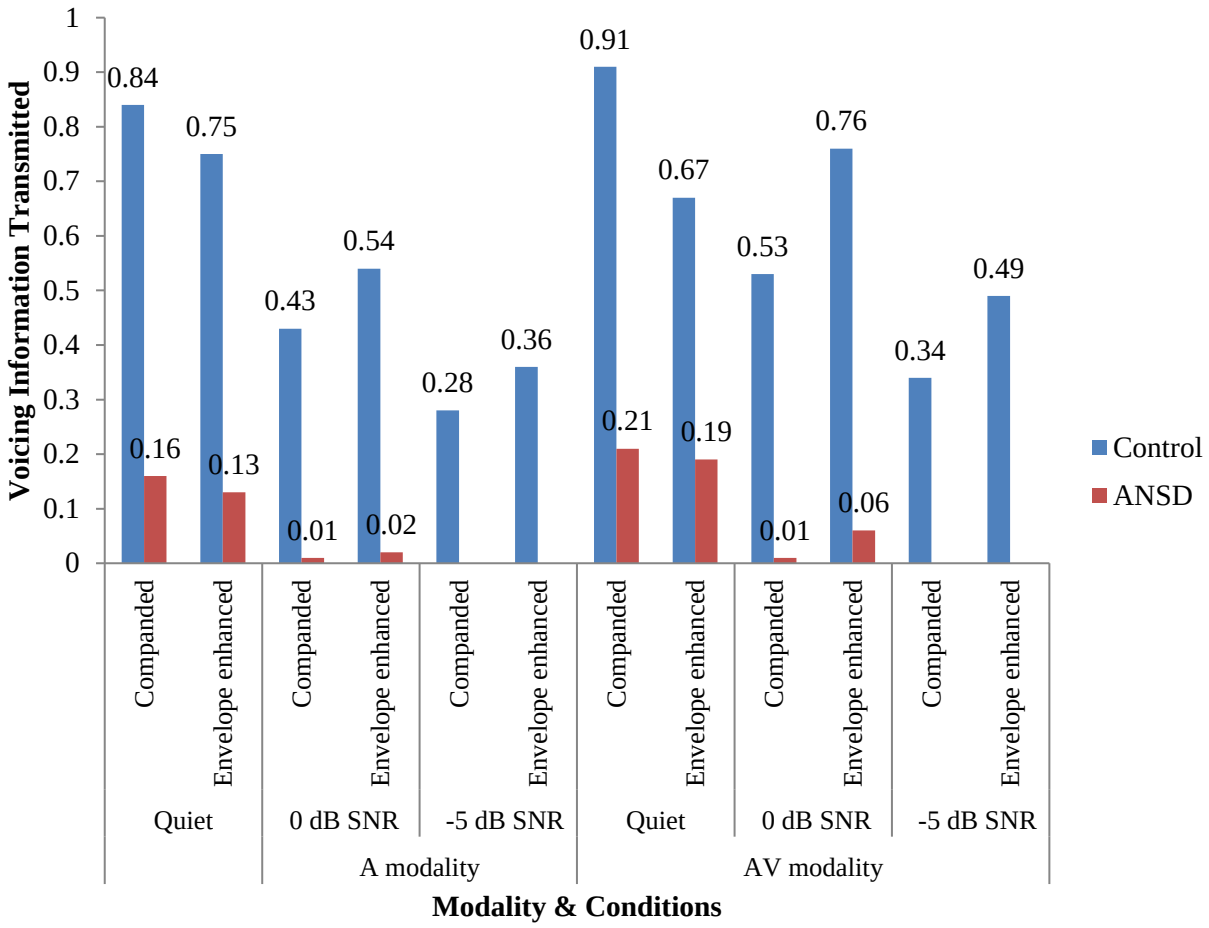

Figure 4.12: Comparison of voicing information transmitted in the two groups of participants for the companded and envelope enhanced stimulus in the two conditions. The data were derived by SINFA on the scores of acoustically enhanced syllables *Note.* A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio, ANSD: auditory neuropathy spectrum disorder

The voicing feature transmitted in ANSD group was also lesser compared to control in the companded and envelope enhanced stimuli. The difference between the two groups was lesser in AV modality compared to A-modality. Among the two conditions, 0 dB SNR transmitted lesser voicing information compared to quiet condition. This was more evident in ANSD group compared to the control group. The voicing feature transmission in the control group at -5 dB SNR was more than that of the ANSD group at better SNRs in any of the two modalities.

4.2 Effect of Modality, Condition and Stimulus on Syllable identification Scores of ANSD Group

This section addresses the third objectives of the study. The mean and standard deviation of syllable identification score of ANSD group obtained for the three types of stimuli (primary, companded & envelope enhanced syllables) in the three modalities (Auditory, Auditory-Visual & Visual), in quiet and 0 dB SNR conditions are shown in Table 4.7.

Table 4.7: *Mean and standard deviation (SD) of syllable identification score obtained for the three types of stimuli (primary, companded & envelope enhanced syllables) in Auditory, Auditory-Visual and Visual modalities, in quiet and 0 dB SNR conditions*

Condition	Stimulus	Modality		
		Auditory	Auditory-Visual	Visual
		Mean (SD)	Mean (SD)	Mean (SD)
Quiet	Primary	35.15 (11.87)	42.43 (9.50)	23.35 (5.75)
	Companded	33.6 5 (11.46)	42.68 (9.54)	
	Envelope enhanced	34.76 (12.07)	41.05 (8.51)	
0 dB SNR	Primary	18.45 (7.14)	28.00 (5.04)	
	Companded	17.60 (7.13)	27.60 (4.26)	
	Envelope enhanced	20.68 (9.55)	30.70 (3.44)	

Note. Maximum possible score was 60 (6 CV*10 times).

As seen in the table, the mean identification scores were higher in the AV modality compared to A-modality for primary, companded and envelope enhanced stimuli. Within A modality, in the quiet condition, the mean score was higher for primary syllables followed by envelope enhanced and was the least for companded stimuli. On the contrary in 0 dB SNR condition, the mean score was highest for envelope enhanced stimuli followed by primary and companded stimuli.

In AV modality, mean score of quiet condition showed comparable scores for primary and companded syllables, while the mean score for envelope enhanced stimulus was lower than the other two stimuli. On the contrary, in 0 dB SNR condition, envelope enhanced stimuli showed higher mean scores compared to the other two stimuli.

The mean score in the visual modality was lower than that in the A and AV modalities in quiet condition. In the 0 dB SNR condition, mean score in visual modality (in quiet) was higher than that in the A modality. The pattern of results was same for all the three stimuli.

The difference in mean identification score obtained in the two modalities (A and AV), two conditions (quiet and 0 dB SNR) and three stimuli (primary, companded & envelope enhanced) was tested using three-way repeated measures ANOVA (2*2*3). The results showed a significant main effect of condition [F (1, 39) = 159.48, $p<0.01$] and modality [F (1, 39) = 105.58, $p<0.01$]. There was no significant main effect of stimulus [F (2, 78) = 2.48, $p>0.05$]. Three-way interaction was not significant. But there was a significant two-way interaction between stimulus and condition [F (2, 78) = 6.24, $p<0.01$] and, modality and condition [F (1, 39) = 5.01, $p<0.05$]. There was no significant two-way interaction between stimulus and modality [F (2, 78) = 0.97, $p>0.05$]. Because there were significant two-way interactions, the effect of stimulus, modality and the condition were separately tested.

4.2.1. Effect of stimulus on syllable identification scores of individuals with ANSD

Subsequent to two-way interaction between stimulus and condition, the effect of stimulus was separately tested in each condition using one-way repeated measure

ANOVA. Results showed no significant main effect of stimulus in quiet condition [F (2, 78) = 63.57, $p>0.05$], while there was a significant main effect of stimulus in the 0 dB SNR condition [F (2, 78) = 4.44, $p<0.05$]. Bonferroni pair-wise comparison showed that identification score of envelope enhanced stimuli was significantly different from that of companded and primary stimuli ($p<0.05$). Figure 4.13 shows individual identification scores for each stimulus type (primary, companded & envelope enhanced), in the quiet (A) and 0 dB SNR (B) conditions in the auditory modality.

The individual score showed that some of the participants had higher scores for the envelope enhanced and companded stimuli compared to the primary stimuli. But the vice versa was also seen. The number of individuals who showed higher score with envelope enhancement is more than those showing better scores with the companded stimuli. Compared to quiet, in the 0 dB SNR condition, the improvement with envelope enhanced stimulus was larger. Figure 4.14 shows the individual identification scores in each stimulus type (primary, companded & envelop enhanced) in the quiet (A) and 0 dB SNR (B) conditions in AV modality.

The individual scores showed that some of the participants had higher scores for the envelope enhanced and companded stimuli compared to the primary stimuli. The vice versa was also seen. The number of individuals who showed higher score with companded stimuli is more than those who showed higher scores with the envelope enhancement stimuli in the quiet condition. But the vice versa was seen in the 0 dB SNR condition.

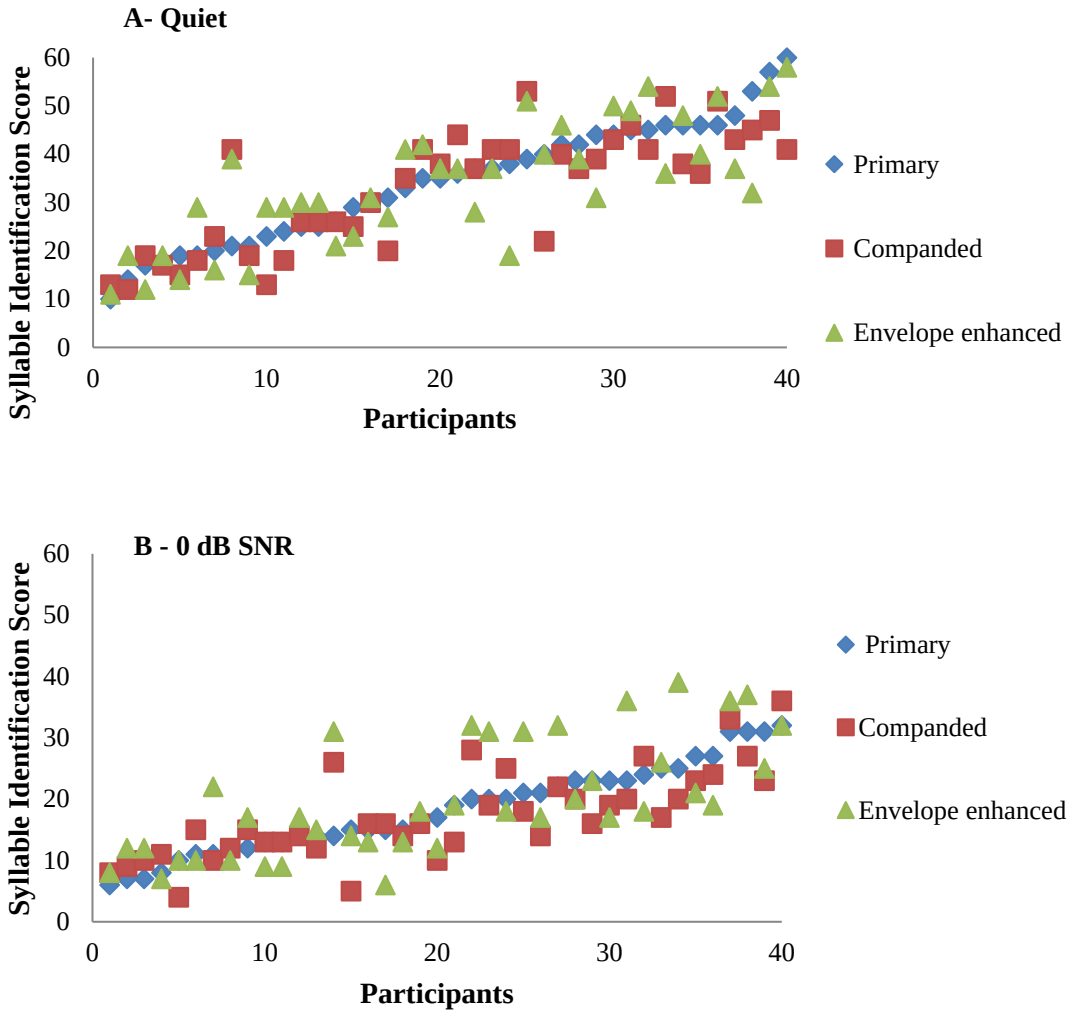

Figure 4.13: Individual identification scores of each stimulus type in quiet (A) and 0 dB SNR conditions (B) in the auditory modality. The scores are depicted with reference to the scores for the primary stimuli arranged in ascending order.

Note. Maximum possible score is 60.

Comparison of the individual scores in the A modality shows that there is no particular trend in the way scores varied across the three stimuli. However in the AV modality, most of the participants scored higher for envelope enhanced stimulus compared to the primary and companded stimuli. Therefore the effect of stimulus was assessed separately for A and AV modalities within the 0 dB SNR condition. Results in the A modality showed a significant main effect of stimulus [$F(2, 78) = 4.44$, $p<0.05$]. Bonferroni multiple pair-wise comparisons showed mean score of envelope

enhanced stimuli to be significantly higher than that of companded stimulus. The other two comparisons were not significantly different (p>0.05).

The results in AV modality also showed a significant main effect of stimulus type [F (2, 78) = 8.90, p<0.05]. Bonferroni multiple pair-wise comparison showed mean scores of envelope enhanced stimuli were significantly higher than that of primary and companded stimuli (p<0.05). There was no significant difference between the scores of primary and companded stimulus (p>0.05).

Additionally, in the Figure 4.14 (B), it was observed that the difference between the envelope enhanced and primary stimuli was larger when the identification scores of the primary syllable were poorer. The difference narrowed as the scores in the primary stimuli increased. Therefore it was of interest to study whether the improvements with envelope enhanced stimulus differ between good and poor performers. To do this the participants were categorized into two groups based on the scores obtained for primary stimulus in the AV modality in 0 dB SNR condition. The 95% confidence interval was considered for grouping. Accordingly, Good performers had scores of more than 29 (18 individuals) and Poor performers had scores of less than 26 (12 individuals). The absolute difference in identification scores of envelope enhanced and primary syllable in the AV modality in 0 dB SNR condition was calculated for the two groups (Good & Poor performers) for comparison. The mean, standard deviation and median of the difference in identification scores for the envelope enhanced and primary stimuli in 0 dB SNR condition in the two groups is given in Table 4.8.

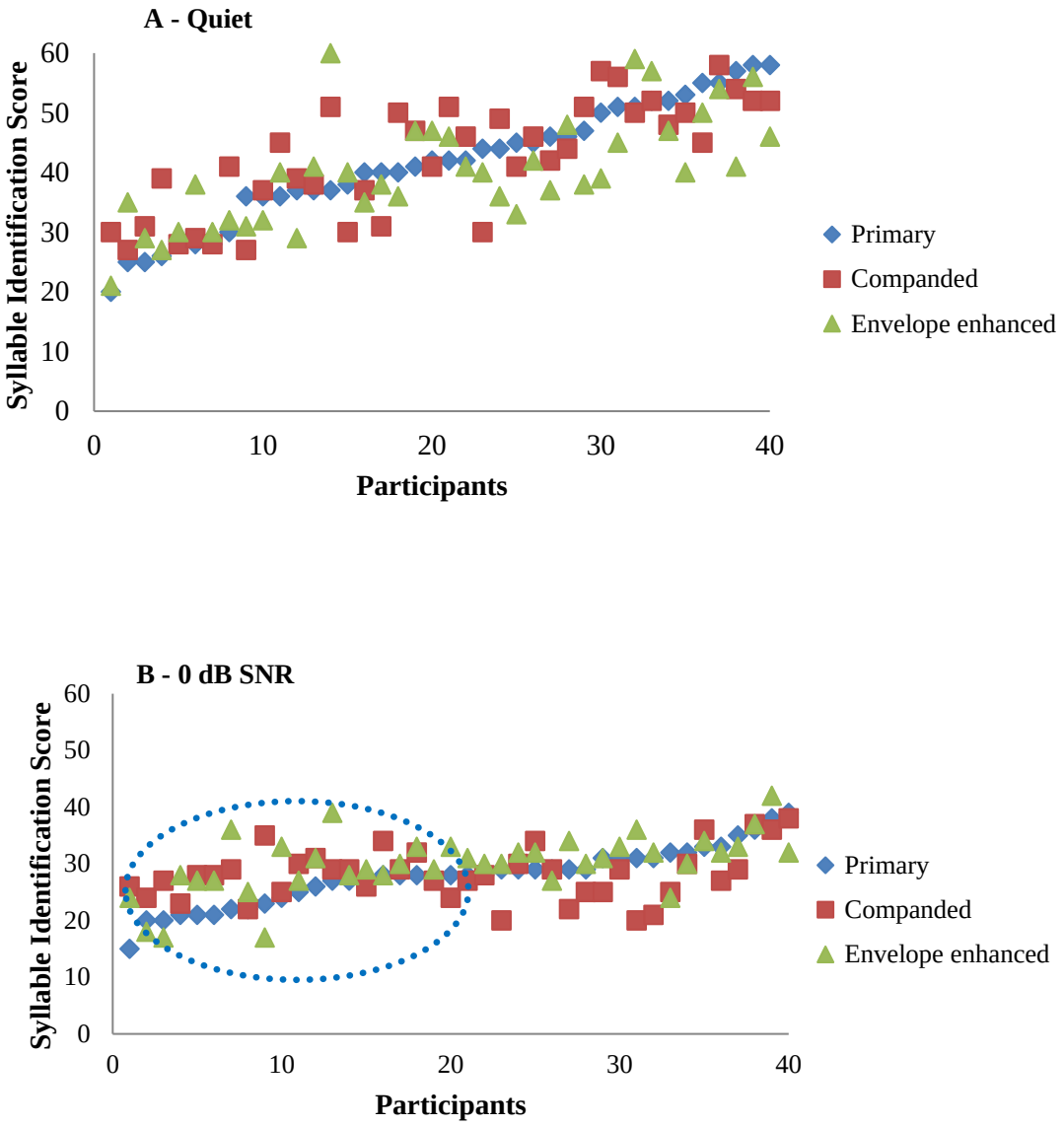

Figure 4.14: Individual identification scores in each stimulus type in quiet (A) and 0 dB SNR conditions (B) in the AV modality. The scores are depicted with reference to the scores in the primary stimuli arranged in ascending order. The area marked with circle denotes higher scores for the envelope enhanced stimuli in those individuals who scored lesser for the primary stimulus.

Note. Maximum possible score is 60.

Table 4.8: *The mean, standard deviation (SD) median and range of the difference in identification scores of envelope enhanced and primary syllable in 0 dB SNR condition in the two groups (Good & Poor performers)*

Group	Mean	SD	Median	Min - max
Poor performer	4.17	5.70	5.50	-06 - +14
Good performer	0.28	3.58	1.00	-08 - +05

The mean score was higher for poor performers compared to good performers indicating higher benefit with envelope enhancement in poor performers. The difference in mean score between the two groups was tested using Mann Whitney U test. Results showed significant difference between the two groups (Z = -2.17, $p<0.05$).

There was no interaction between stimulus and modality. Therefore, effect of stimulus was not tested separately in each modality.

4.2.2. Effect of condition on syllable identification scores of individuals with ANSD

There was a significant interaction between stimulus and condition. In view of this, the two conditions (quiet & 0 dB SNR) were compared separately in each stimulus type using paired t-test. Results showed that the scores in the quiet condition were significantly higher than that in the 0 dB SNR in primary (t = 11.82, df = 39, $p<0.05$), companded (t = 10.80, df = 39, $p<0.05$) and envelope enhanced stimuli (t = 8.02, df = 39, $p<0.05$). Similarly, because there was an interaction between modality and condition, the two conditions (quiet & 0 dB SNR) were compared separately in A and AV modalities using paired t-test. Results of both A (t = 11.82, df = 39, $p<0.05$)

and AV modality (t = 10.49, df = 39, p<0.05) showed significant difference between the two conditions.

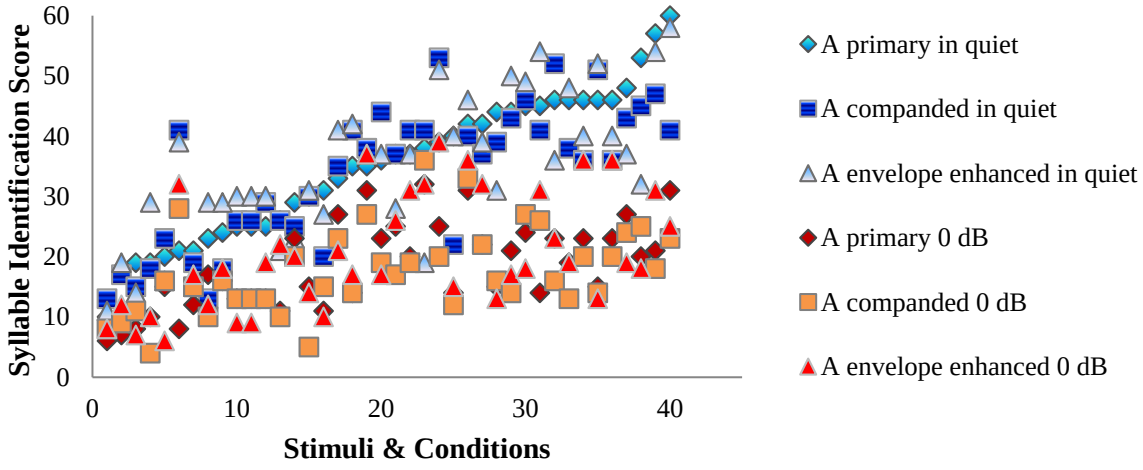

Figure 4.15: The individual identification scores in quiet and 0 dB SNR condition for the three stimuli in the A modality. The scores are depicted with reference to the scores obtained for primary stimulus in A-modality in quiet.

Note. A: auditory modality, Maximum possible score is 60.

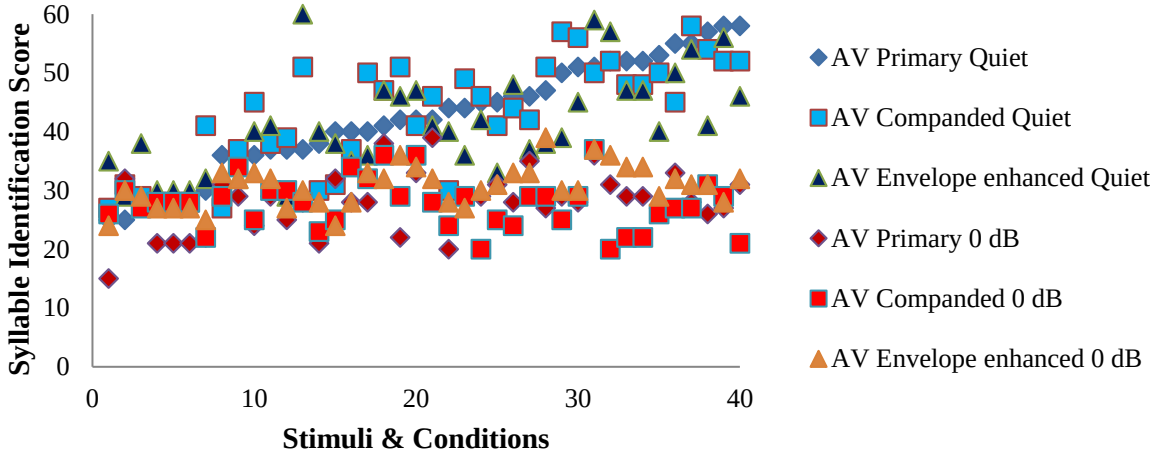

Figure 4.16: The individual identification scores in quiet and 0 dB SNR condition for the three stimuli in the AV modality. The scores are depicted with reference to the scores obtained for primary stimulus in AV modality in quiet.

Note. AV: auditory-visual modality, Maximum score is 60.

The individual identification scores in quiet and 0 dB SNR condition for the three stimuli in the A modality is shown in Figure 4.15. Similarly, the individual identification scores in quiet and 0 dB SNR condition for the three stimuli in the AV modality is shown in Figure 4.16. From the figures (4.15 & 4.16), it can be observed that the identification scores in quiet condition were better than 0 dB SNR condition. This was true for all the three stimuli in both the modalities.

4.2.3 Effect of modality on syllable identification scores of individuals with ANSD

In the identification of primary stimuli, the mean score in quiet was best in AV followed by A and least in V modality. Whereas in 0 dB SNR, the mean score was higher in AV than in A-modality, but the mean score in A-modality was lesser than that in V modality of quiet condition. In Figure 4.17, it can be observed that, in most of the participants, score obtained in the AV modality was better than that in A and V modalities. This was true both in quiet and 0 dB SNR conditions. In the quiet condition, among the A and V modalities, most of them obtained higher identification scores in A compared to V modality. However, exceptional cases did exist wherein, A was better than AV, V was better than AV and, V was better than A. On contrary, at 0 dB SNR, the individual scores in the A modality was poorer than that in the V modality (obtained in quiet) in many instances.

The identification scores across the three modalities were compared separately in each condition (quiet & 0 dB SNR). In the quiet condition, the scores were compared using one-way repeated measures ANOVA and the results showed a significant main effect of modality [$F(2, 78) = 63.71$, $p<0.05$]. In the Bonferroni pair-wise comparison, significant differences were found across all the three modalities. In

the 0 dB SNR, the identification scores in A and AV modalities were compared using paired t-test. The results showed a significant difference between the two modalities (t = -9.60, df = 39, p<0.05). There was no interaction between stimulus and modality in the 3-way ANOVA. Therefore, the effect of modality was not further analyzed in each stimulus. Figure 4.17 shows the individual scores in each modality, in the quiet (A) and 0 dB SNR (B) conditions for the primary stimulus. The scores are depicted with reference to the scores obtained for the primary stimulus in A-modality.

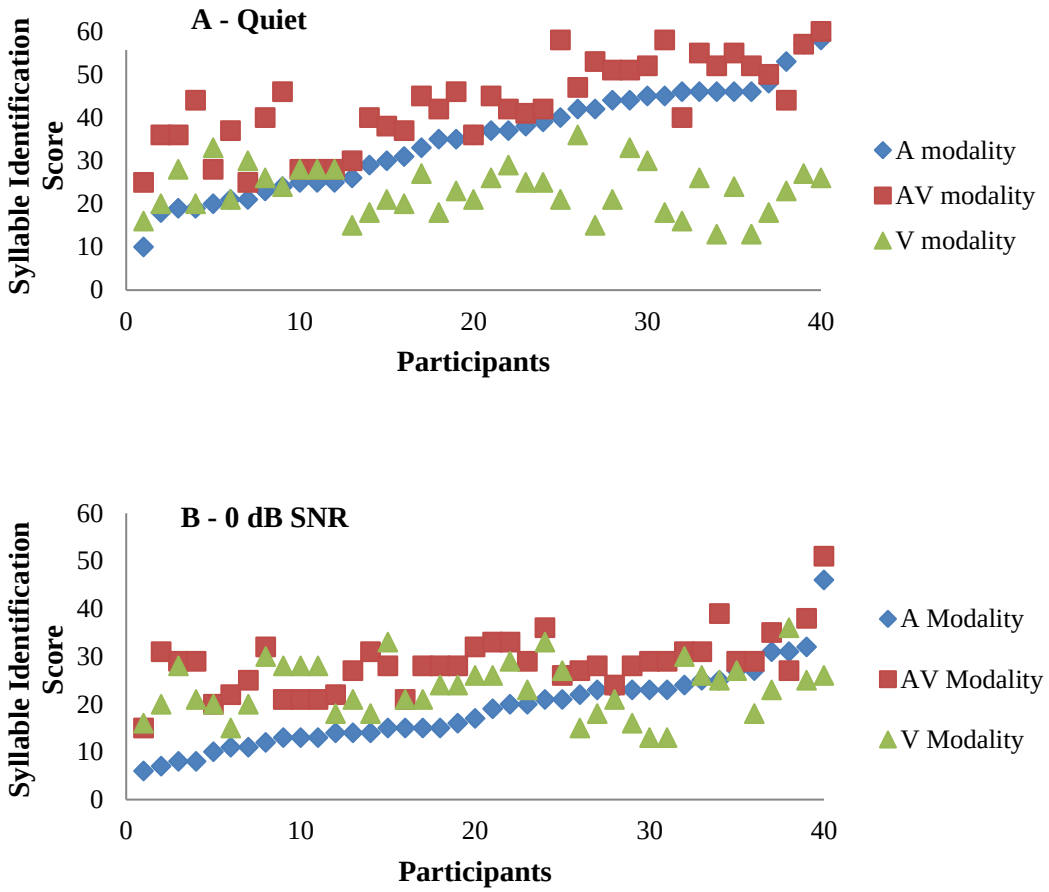

Figure 4.17: The individual identification scores in three modalities in the quiet (A) and 0 dB SNR condition (B) for the primary stimulus. The scores are depicted with reference to the scores obtained for the primary stimulus in A-modality.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality

It can be observed from the figure that the scores were better in the quiet compared to 0 dB SNR condition. This was true for all the three modalities. Scores in AV modality was higher in both conditions compared to A and V modalities. V modality scores were the least in quiet condition, whereas A modality scores were the least in 0 dB SNR condition.

4.3 Effect of Stimulus, Modality and Condition on Feature Transmission Derived from SINFA, in ANSD group

This section addresses the fourth objective of the study**.** The total information transmitted in ANSD for the primary companded and envelope enhanced stimuli in the A and AV modalities in quiet and 0 dB SNR conditions is shown in Figure 4.18. Total information transmitted in the AV modality was higher than the A modality. This was true for all the three stimuli and both the conditions. The difference in total information transmitted between the A and AV modality was more in 0 dB SNR compared to quiet condition. The total information transmitted for the primary stimuli in A-modality was lesser compared to companded and envelope enhanced stimuli. In the AV modality, the total information transmitted for the primary and envelope enhanced stimuli was same, which was lesser compared to companded stimuli. Total information transmitted in the V modality in quiet was more than that in A-modality for all the three stimuli. But it was lesser than that in the AV modality.

The total information transmitted for the companded and envelope enhanced stimuli is comparable in A modality in quiet. Whereas in 0 dB SNR envelope enhanced stimuli showed total information transmitted higher compared to companded stimuli. On contrary in the AV modality, the information transmitted for

the companded stimuli was higher compared to the envelope enhanced stimuli in the quiet condition. But the vice versa was seen in the 0 dB SNR condition.

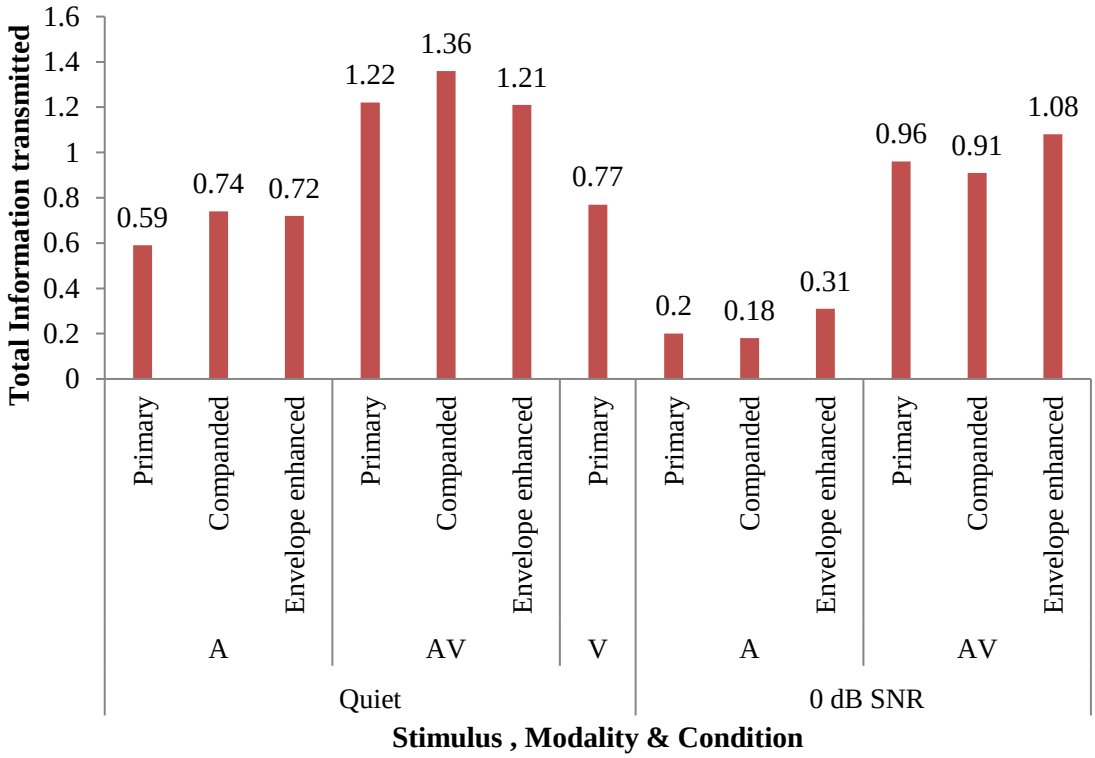

Figure 4.18: Total information transmitted for the three stimuli in different modalities in the two conditions.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio

Place and voicing feature transmitted for the two enhanced stimuli is shown in Figure 4.19 and Figure 4.20 respectively. The place feature transmitted in A-modality for quiet condition was equal for both companded and envelope enhanced stimuli and these two were higher than that for the primary stimuli. In the AV modality in quiet condition, companded stimuli transmitted higher place information compared to envelope enhanced and primary stimuli. Visual modality transmitted higher place information compared to A-modality in quiet condition. In the 0 dB SNR condition,

place information transmitted by envelope enhanced stimuli was higher than the primary and companded stimuli. This was true in both A and AV modalities.

The place feature transmitted in the ANSD group was better in the quiet condition compared to 0 dB SNR condition. This was true for both A and AV modalities. Place information transmitted in the AV modality was higher compared to A-modality and V modality. This was true for all the three stimuli in both the conditions.

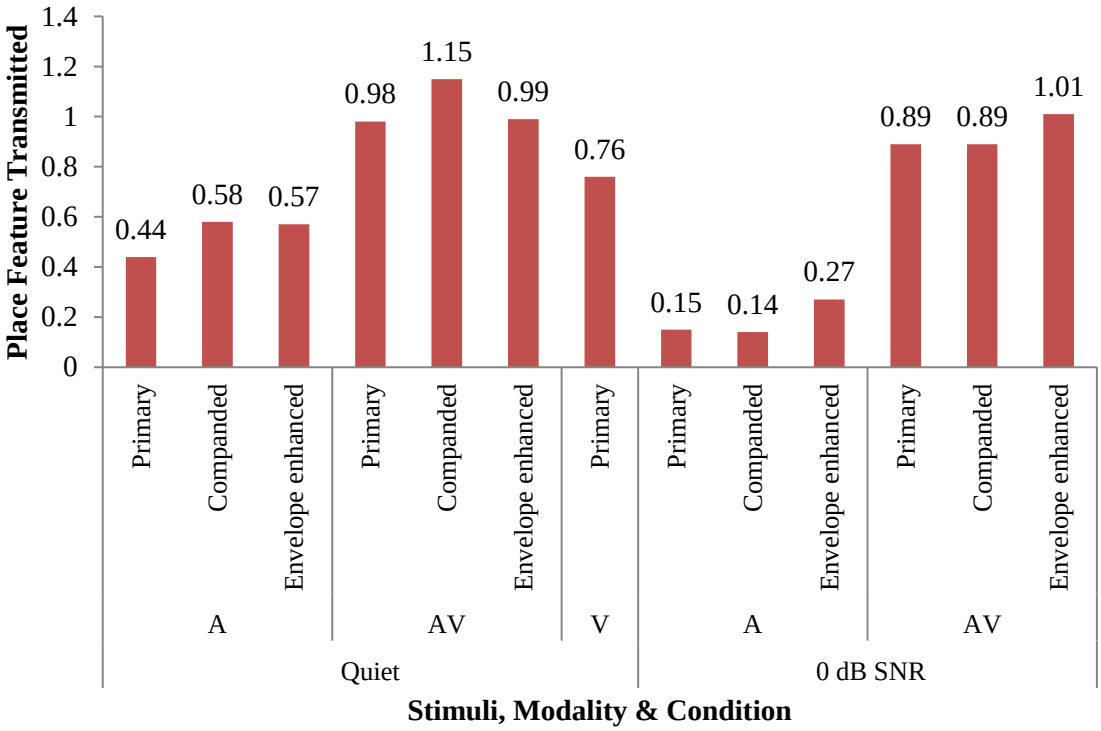

Figure 4.19: Place feature transmitted for the companded and envelope enhanced stimuli in the auditory and auditory-visual modalities in quiet and 0 dB SNR conditions.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio

The voicing feature transmitted was higher for the companded stimuli compared to envelope enhanced stimuli and the primary stimuli. This was true both in A and AV modalities in the quiet condition. On the contrary, the voicing information transmitted for the envelope enhanced stimulus was higher compared to companded stimulus in the A and AV modalities in 0 dB SNR condition. Voicing feature transmitted was higher in AV modality compared to A-modality in the quiet condition. This was true for both the stimulus in quiet and for the envelope enhanced stimuli in 0 dB SNR condition. Whereas in 0 dB SNR condition, voicing feature transmitted was equal for the companded stimuli in both modalities. The voicing information transmitted in ANSD was better in the quiet condition compared to 0 dB SNR condition. This was true for both A and AV modalities.

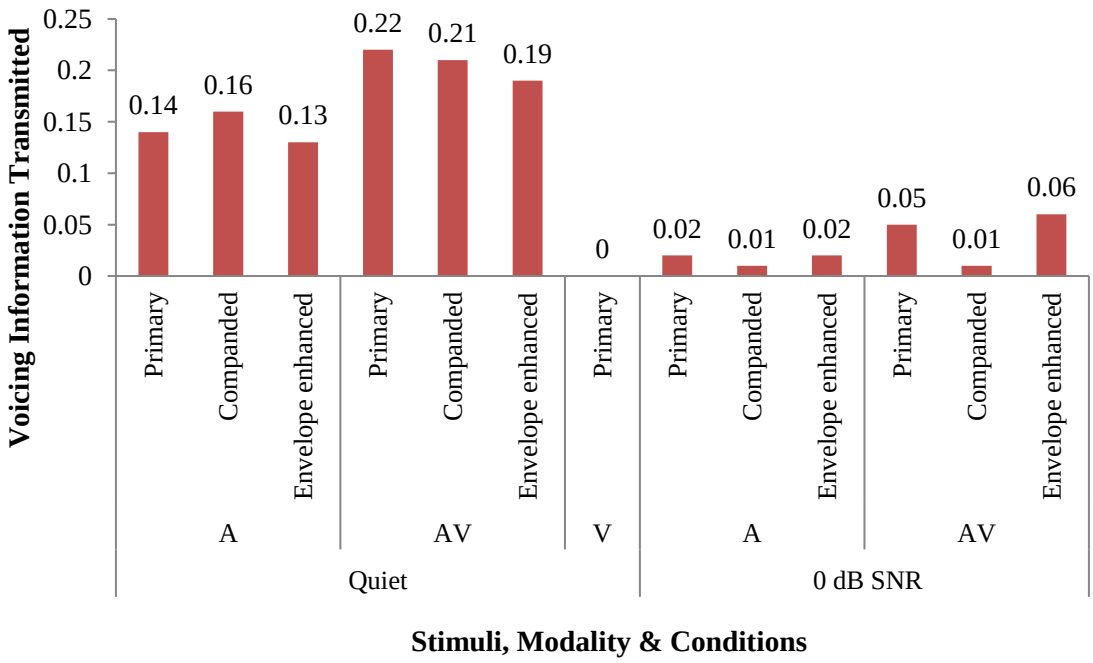

Figure 4.20: Voicing feature transmitted for the primary companded and envelope enhanced stimuli in the A and AV modalities in quiet and 0 dB SNR conditions.

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality, SNR: signal to noise ratio

It can be seen from the figures that place feature was transmitted better than voicing feature in all the three stimuli, in the three modalities and in both the conditions.

4.4 Predictors of Benefit from AV Modality

This section addresses the fifth objective of the study. It was observed that AV modality provided maximum identification scores compared to other two modalities and this was true in all the conditions. Hence attempt was made to identify the predictive variables, if any, for scores of AV modality. The scores obtained in the auditory and visual modality was tested as predictive variables in regression model. Additionally the score obtained in quiet condition was tested for its role in the scores of 0 dB SNR. Results of regression (Table 4.9) showed that scores of auditory modality in both quiet and 0 dB SNR conditions are significant predictors of scores of AV modality in ANSD group. The scores of AV modality in quiet condition and the scores of visual modality scores were significant predictors of AV modality in 0 dB SNR. However, scores of visual modality were not significant predictors of scores of AV modality in quiet condition. Furthermore, score of auditory modality in quiet condition was a significant predictor of that in 0 dB SNR. But the score of auditory modality was not a significant predictor of score in visual modality.

4.5. Relationship of Duration of Hearing Loss, Puretone Average and SIS with AG and VG in ANSD group.

This section addresses the sixth objective of the study. The study also determined the relationship of AG and VG scores with the degree of hearing loss, reported duration of ANSD and the syllable identification scores. Pearson's correlation was used for the purpose. This was done only for AG and VG derived

from scores of primary stimuli. The results (Table 4.10) showed that there is no significant correlation of AG and VG score with any of the three parameters tested (degree of hearing loss, duration of hearing loss & syllable identification scores).

Table 4.9: *Results of regression analysis indicating the predictive variables of performance in AV modality in ANSD group*

Predictor	Constant	Regression	Residual	F	*p*	R^2
A Quiet	AV Quiet	1	38	64.67	<0.01	0.63
V Quiet	AV Quiet	1	38	1.04	>0.05	0.03
A Quiet	AV 0 dB SNR	1	38	5.65	<0.05	0.13
V Quiet	AV 0 dB SNR	2	37	07.62	<0.01	0.29
A 0 dB SNR	AV 0 dB SNR	1	38	13.42	<0.01	0.26
AV Quiet	AV 0 dB SNR	1	38	7.21	<0.01	0.16
A Quiet	V Quiet	1	38	0.43	>0.05	0.11
A Quiet	A0 dB SNR	1	38	29.43	<0.01	0.43

Note. A: auditory modality, AV: auditory-visual modality, V: visual modality

Table 4.10: *Results of Pearson's correlations of AG and VG for the primary and enhanced stimuli with PTA, duration of hearing loss and SIS in ANSD group*

Variable	Condition	Correlation	PTA	Duration of loss	SIS
AG primary	Quiet	r	0.75	-0.11	-1.22
		p	0.65	0.49	0.45
	0 dB SNR	r	-0.05	0.21	0.24
		p	0.74	0.20	0.13
VG primary	Quiet	r	-0.50	0.17	0.04
		p	0.77	0.29	0.78
	0 dB SNR	r	0.26	0.03	0.67
		p	0.09	0.86	0.68

PTA and SIS mentioned is the ear having better PTA and SIS and the test was done in the sound field.

Note. N = 40, PTA: puretone average, SIS: speech identification score, AG: auditory gain, VG: visual gain.

4.6 Test Re-test Reliability of the Syllable Identification Score Obtained in Individuals with ANSD

Test re-test reliability was determined for 10% of data (4 participants) for the three stimuli (primary, companded & envelope enhanced), in the three modalities (A, AV & V) and in two conditions (quiet & 0 dB SNR). The participants were tested within of one month of initial evaluation. Cronbach's Alpha test was used to assess the test re-test reliability. The results are shown in Table in 4.11.

The result showed acceptable to excellent test re-tests reliability based on the classification of internal consistency using Cronbach's Alpha (Dunn, Baguley, & Brundsen, 2014). This suggests the reliability of the test on multiple evaluations.

Table 4.11: *Results of test re-test reliability of syllable identification scores for the three stimuli, three modalities and two conditions*

Stimuli	Modality	Condition	Cronbach's Alpha
Primary	A	Quiet	0.81
	AV		0.94
	V		0.88
	A	0 dB SNR	0.95
	AV		0.95
Companded	A	Quiet	0.97
	AV		0.96
	A	0 dB SNR	0.94
	AV		0.91
Envelope enhanced	A	Quiet	0.75
	AV		0.88
	A	0 dB SNR	0.97
	AV		0.88

Chapter 5

DISCUSSION

The management of Auditory Neuropathy Spectrum Disorder (ANSD) is always a challenge to audiologists. In spite of several attempts in the past, none of the strategies available have been able to enhance speech perception to the satisfaction of persons with ANSD. The resultant ineffective communication has obviously been leading to significant negative effects on the psychological and social aspects of their life. In view of this, as part of the continuing efforts towards effective audiological management of persons with ANSD, the present study was taken up. The study aimed to compare the benefits of visual cues, acoustic enhancements, and the combination of the two in improving speech perception in persons with ANSD. The findings of the study are discussed under the following headings;

1. Speech perception in persons with ANSD
2. Relative benefits of auditory and visual cues in the speech perception of ANSD
3. Benefits of acoustic enhancement in the speech perception of ANSD
4. Benefits of combination of visual cues and acoustic enhancements
5. Predictors of benefit with AV speech perception
6. Influence of hearing sensitivity, speech identification scores and duration of the condition on the benefits of AV speech perception

5.1 Speech Perception in Persons with ANSD

Typical of ANSD, the study found significantly poorer speech identification in participants with ANSD compared to control group. The results were same irrespective of the stimulus type (primary, companded & envelope enhanced) and in

auditory as well as AV modalities. The poor speech identification is the cardinal feature of ANSD attributable to the impaired temporal processing, secondary to dys-synchronous neural firing (Starr et al., 1991; Kraus et al., 2000; Rance et al., 2004; Kumar & Jayaram, 2005). Speech perception further reduced in the presence of noise and the negative effects of noise was more in persons with ANSD, similar to that reported in the earlier studies (Mattys, Davis, Bradlow, & Scott, 2012; Starr et al., 1996; Kraus et al., 2000; Shallop, 2002).

The speech identification of individuals with ANSD in quiet was poorer than the speech identification of control group at -5 dB SNR. In the present study -5 dB was the minimum SNR used and the finding suggests that the dys-synchrony of the nerve fibers in the persons with ANSD results in deterioration of the SNR and the resultant inherent SNR is lower than -5 dB. This is an indirect inference about the SNR loss in ANSD. It is recommended that future studies can measure SNR-50 to determine the SNR loss in persons with ANSD.

The dynamics of AV speech perception was seen to be different in ANSD compared to individuals with normal hearing. The performance of persons with ANSD was poorer in visual modality compared to control individuals. This is in agreement with earlier study by Maruthy and Geetha (2011) and hints at deficit visual processing in persons with ANSD. In spite of poorer speech identification in the visual modality compared to control group, individuals with ANSD showed higher visual gain scores than the control group. This means that the addition of visual cues benefitted persons with ANSD more than the individuals with normal hearing. This supports the use of AV modality in persons with ANSD. The visual gain was higher both in quiet and 0 dB SNR in ANSD group compared to control group. Within the

two listening conditions, visual gain was higher in the presence of noise. These findings suggest that persons with ANSD are able to make better use of visual cues compared to individuals with normal hearing, more so in the presence of noise. The precise reason for this is not known. However, one can speculate that lower scores in the auditory modality could be one of the important factors influencing it. Studies in the literature (Tye-Murray et al., 2007; Munhall et al., 2004; MacLeod & Summerfield, 1987) have shown that the importance of visual cues increases as the listening environment becomes more challenging. Considering that perception through auditory modality is compromised in ANSD, the role of visual cues seems to be crucial. Further, reported duration of onset of ANSD in the present study was up to 20 years. Therefore, it is likely that they would have trained themselves to better utilize the available visual cues, as a compensatory mechanism, in view of the compromised auditory input. On the other hand, control group was only dependent on the auditory modality and perceiving speech in the AV modality was not a regular scenario for them. This would have been the probable reason for their poorer visual gain. However, caveat for such an inference is that individuals with ANSD had poorer mean identification scores in the visual modality compared to individuals with normal hearing.

On the contrary, persons with ANSD showed lesser auditory gain compared to control group. This again indicates that, while perceiving speech in the AV modality, persons with ANSD depend less on the auditory modality unlike control individuals. The lesser dependency of individuals with ANSD on the auditory modality was seen in both quiet and 0 dB SNR.

The results of the auditory and visual gain in control group suggest that they were able to make efficient use of the cues provided by the auditory modality and their dependency was less on the visual cues. The dependency of control group on auditory modality was also supported by the lesser visual gain even at 0 dB SNR. The lower visual gain in the control group however can be contributed to the ceiling effect in the auditory modality.

In the control group, the visual gain increased with decrease in SNR of speech. The control group had maximum visual gain at -5 dB SNR. This suggests that even the control group use visual cues in instances where the auditory cues are compromised. Earlier studies have also shown that the contribution of visual cues increases as the listening environment becomes more challenging (Tye-Murray et al., 2007; Munhall et al., 2004; MacLeod & Summerfield, 1987).

The results of SINFA also indicated significant difference between the control and ANSD groups in terms of information transmitted. This was true in all the three modalities, in all the stimulus types and in both the conditions. The difference between the two groups was more in the auditory modality compared to AV modality, indicating that persons with ANSD are able to utilize the visual cues to compensate for the deficit input through the auditory modality. Feature transmission index indicated that the both place and voicing features are poorly transmitted in individuals with ANSD attributable to the dys-synchronous firing of auditory neurons.

Taken together, the findings of speech identification, auditory gain, visual gain and the information transmission support that the speech perception in individuals with ANSD is significantly poorer than the control group. Therefore, the first two hypotheses that ‘there is no significant difference between normal hearing

individuals and persons with ANSD in their syllable identification, auditory gain and visual gain scores' and 'there is no significant difference between normal hearing individuals and persons with ANSD in their feature transmission index derived from SINFA' are rejected.

5.2 Relative Benefits of Auditory and Visual Cues in the Speech Perception of ANSD

The results showed a significant difference across auditory, AV and visual modalities in the speech identification of ANSD. In the quiet condition, they performed the best in AV modality followed by auditory modality and least in visual modality. This indicates that they utilize cue from both auditory and visual modalities while identifying speech in the AV modality. The finding is in partial agreement with the previous study (Ramirez & Mann, 2005). In their study, the scores in the AV modality did not differ from the visual-alone modality. Based on this, it was inferred that persons with ANSD primarily depend on visual cues, with insignificant role of auditory cues. However in the present study, we found that scores in the AV modality were significantly higher than that in visual modality. This suggests that persons with ANSD make use of auditory as well as visual cues for their speech perception. The individual scores showed that speech identification in the AV modality was higher than auditory as well as visual modality scores in most of the participants. This indicates that audiologists can recommend visual cue supplementation as a strategy to facilitate speech perception in persons with ANSD.

The present finding that both auditory and visual cues play a role in the speech perception of ANSD suggests that both these modalities need to be facilitated to the best possible extent in these individuals. The auditory modality may be enhanced

through signal enhancement strategies such as FM devices (Rance et al., 1999), companding (Narne, Barman, Deepthi, & Shachi, 2014) and envelope enhancement (Narne & Vanaja, 2009b). The visual modality on the other hand can be enhanced either through training in speech reading using standardized methods (Kinzie & Kinzie, 1931; Bruhn & Mueller-Walle, 1949) or though anticipatory compensatory strategies.

The current study used non-meaningful monosyllables with stop consonants to assess the speech identification. Monosyllables have least redundancy and stop consonants in particular are most challenging for individuals with ANSD in terms of perception (Hassan, 2011; Narne, 2013). Therefore one can expect greater benefits with visual cues when words or sentences are used. This however needs to be investigated. It is also important to note that none of the participants of the current study were systematically trained for speech reading. If trained they may be able to derive greater benefits from the AV modality for speech perception.

The speech identification was poorer in the presence of noise compared to quiet condition both in auditory and AV modalities. The reason for such reduction is primarily due to their inability to extract the envelope and fine structure cues from speech (Buss, Hall, & Grose, 2004) in the auditory modality. The reduction in speech perception was seen in all the participants.

Despite reduction in speech perception in the presence of noise, the benefit derived from the visual cues was retained. In fact the mean difference showed that the benefit derived from visual cues was more in the presence of noise. This is in agreement with the earlier studies in individuals with hearing impairment (Ross et al., 2006; Bernstein et al., 1969; Grant & Seitz, 2000). Participant's speech perception in

the AV modality was significantly better than that in the visual modality even in the presence of noise. This indicates that individuals with ANSD use auditory cues even in degraded listening environments. This is in contradiction with the reports of Ramirez and Mann (2005), although the exact reason for differences in the findings of these two studies is not known. The finding of the present study is derived from data of 40 individuals with ANSD, while Ramirez and Mann (2005) had reported their finding from 4 individuals with ANSD. The difference in the range of speech identification scores across participants and the difference in the scoring pattern would have contributed for the difference in the results of the two studies.

It was also observed that the identification scores of most the participants in auditory modality at 0 dB SNR condition were poorer compared to that in visual modality at quiet condition. In the present study, speech identification in the visual modality was tested only in quiet, in line with the earlier studies (Ross et al.,2006; Sumby & Pollack, 1954). These studies had shown similar speech identification across different signal to noise ratios in the visual alone modality. In view of this, it can be inferred that the visual processing shall be more useful in the degraded listening environment. Also, the speech identification in the AV modality must be primarily contributed by the visual cues.

Persons with ANSD are known to have erroneous auditory perception. In the present study we had hypothesized that the erroneous auditory perception when combined with visual cues may result in McGurk like effect in their perception. The support for the notion can be drawn from the consonant confusion matrix in Kumar (2006). During the closed set identification of the consonants, there was consonant substitution and the substituted consonant differed from the target, in place of

articulation and voicing. Therefore, if an ANSD individual focuses on auditory as well as visual cues during the AV mode of presentation, the information from the two modalities may be perceived as incongruent, in turn distorting the resultant perception. Results of the present study showed enhancement in speech perception in the AV mode. This suggests that McGurk like effect proposed in the present study is less likely and even if present, it is not influencing speech perception to a large extent.

The results of SINFA in ANSD showed that total transmitted information was lesser compared to typical participants in all the listening conditions. The total information transmitted was lower in the 0 dB SNR condition compared to that in quiet, and in quiet it was lesser in auditory modality compared to that in visual modality. The information transmitted was highest in AV modality indicating that the addition of visual modality supplemented cues for the correct identification, which was true for both place and voicing of the consonant. The benefit derived from the visual cues was more for the correct identification of place of articulation compared to that for voicing. This advantage was seen more in the presence of noise. The information is of high relevance to the audiologists and is crucial while counselling the individuals with ANSD about the benefit derived from the AV modality.

Overall, the addition of visual cues showed significant enhancement in speech perception both in quiet and 0 dB SNR conditions. This is an empirical evidence to support recommendation of AV mode for speech perception in ANSD, as a management strategy.

The results of the present suggested that audiological characteristics such as degree of hearing loss, unaided speech identification and the duration of the condition do not relate to auditory and visual gain. The findings were similar in quiet and 0 dB

SNR. This may be due to the heterogeneity in the individual audiological profile which is typical of ANSD. The finding suggests that an individual with ANSD will benefit from the AV mode without significant influence of their degree of hearing loss, speech identification scores and duration of the condition. In other words, all persons with ANSD will benefit from the AV modality to a similar extent supporting its use uniformly across the group.

5.3 Benefits of Acoustic Enhancement in the Speech Perception of ANSD

In the present study, two types of acoustic enhancements were used; companding and envelope enhancement. Both these enhancements have earlier been shown to benefit person with ANSD in their speech perception (Narne, Barman, Deepthi, & Shachi, 2014; Narne & Vanaja, 2009b). While companding, compensate for their poor spectral resolution, envelope enhancement is meant to address their deficit in temporal processing. In contrary to the previous studies, neither of the two acoustic enhancements showed significant benefits in speech perception in the present study.

Narne and Vanaja (2009a) had shown benefits of envelope enhancement only in individuals with good speech identification scores at 0 dB SNR. However, the present study showed that the benefits were negligible both in good as well as poor speech identification groups. The procedures used in the present study for companding and envelope enhancement were exactly same as that of the earlier studies (Bhattacharya & Zeng, 2007; Narne et al., 2014; Hassan, 2011). Narne et al. (2014) had found benefits of companding only in quiet and not at 0 dB SNR in individuals with ANSD. But in the present study, benefits of companding were absent both in quiet and 0 dB SNR. The absence of benefits of acoustic enhancement may be

primarily due to the test stimuli used in the present study. The present study used only stop consonants whereas the previous studies had included other classes of consonants also. Considering that the person with ANSD have more difficulty with perception of transient sounds, the perception of stop consonants would be a challenge and this would have led to the absence of appreciable benefits with acoustic enhancements. However, this is only a speculation and needs to be systematically investigated. Furthermore in the study by Narne and Vanaja (2009a), words were used as test stimuli, which possess greater redundancy. The use of monosyllables in the present study would have hindered the benefits derived with acoustic enhancements. It is proposed that future studies may be undertaken to investigate the effect of different type of stimuli on the benefit derived from acoustic enhancement in ANSD.

The role of acoustic enhancements for the perception of consonants should not be totally ruled out based on the present findings. The participants were not exposed to companded and envelope enhanced speech prior to the testing in this study. Listening to the acoustically enhanced speech was a naïve experience to them. Therefore it is suggested that future studies be undertaken to train the individuals with ANSD for listening to the acoustically enhanced speech and then conclude on the benefits derived from it.

Hence the null hypotheses as ‘there is no significant effect of stimulus, modality, and condition on syllable identification scores of persons with ANSD’ and ‘there is no significant effect of stimulus, modality, and condition on feature transmission index derived from SINFA, in persons with ANSD’ are partly rejected.

5.4 Benefits of Combination of Visual Cues and Acoustic Enhancements

In the present study, we were also interested to investigate the combined effect of visual cue supplementation and acoustic enhancement on speech perception of individuals with ANSD. It was found that there is no benefit of combining the two strategies and the benefit derived from the combined input was only due to visual cues. That is, there was no integration benefit when both the strategies were delivered together to the persons with ANSD.

However, on detailed inspection of the individual data, it was seen that in the AV modality, in 0 dB SNR condition, envelope enhanced syllables resulted in better scores compared to primary syllables. This benefit was observed only for individuals who scored poor in the identification of primary syllables. This indicates that envelope enhancement is helpful for persons with ANSD in adverse listening conditions in the AV modality. Temporal envelope cues are known to be beneficial in speech in noise perception. The results suggest that the enhancement of the temporal envelope has facilitated speech in noise perception in persons with ANSD. However, why such an improvement was observed only AV modality is not clear from the current findings. The findings suggest that combining AV modality with envelope enhancement is helpful to persons with ANSD, but in only few listening conditions.

5.5 Predictors of Benefit with AV Speech Perception

The results suggested that performance of persons with ANSD in the AV modality can be predicted from their performance in auditory modality. Performance in the auditory modality served as predictor of performance in AV modality, both in quiet and noise conditions. However, their speech identification in the visual modality could predict the performance in the AV modality only in the with-noise condition.

Further, their performance in the auditory modality in the with-noise condition could be predicted from their performance in the quiet condition. These findings can be used to counsel the persons with ANSD about the probable benefit they get from using the AV modality. Hence the null hypothesis that 'syllables identification scores in the auditory and visual modality are not significant predictors of benefit from AV modality are partly rejected.

5.6 Influence of Hearing Sensitivity, Speech Identification Scores and Duration of ANSD on the Benefits of AV Speech Perception

The results of the study suggested that audiological characteristics such as degree of hearing loss, unaided speech identification scores and the duration of ANSD do not relate to auditory and visual gains. The findings were similar in quiet and 0 dB SNR. This may be due to the heterogeneity in the individual audiological profile which is typical of ANSD. The finding suggests that an individual with ANSD will benefit from the AV modality without significant influence of their degree of hearing loss, speech identification scores and duration of the condition. In other words, all individuals with ANSD will benefit from the AV modality to a similar extent supporting its use uniformly across the group. Hence, the null hypothesis that 'there is no significant correlation of duration of hearing loss, puretone average, and speech identification scores of persons with ANSD with their respective auditory and visual gain scores' is accepted.

Chapter 6

SUMMARY AND CONCLUSIONS

The study aimed to investigate the relative benefits of visual cue supplementation and acoustic enhancements on the speech perception of persons with ANSD. The purpose was to identify the strategy that is more beneficial and accordingly make recommendation for the audiological management of ANSD.

The study used a repeated measure standard group comparison research design. There were two groups of participants in the age range of 16 to 35 years. The clinical group included 40 participants diagnosed to have ANSD, while the control group had 40 age and gender-matched individuals with normal auditory abilities. They were assessed for their syllable identification of six monosyllables in auditory, visual and auditory-visual (AV) modalities. There were three types of stimuli; primary, companded and envelope enhanced. The identification was assessed in closed set task in quiet as well as 0 dB SNR conditions. In the visual modality, identification of only the primary stimuli was assessed, and only in the quiet condition. An additional -5 dB SNR was assessed used in the control group in the A and AV modalities.

The identification scores were compared across modalities, stimuli and conditions to derive the relative benefits of visual cues and acoustic enhancement on speech perception of individuals with ANSD. Further, from the syllable identification scores, auditory gain and visual gain scores were derived, and the two groups were compared for these scores. The group data were subjected to SINFA to derive the feature-wise information transmitted in different test conditions, in the two groups.

The results showed maximum syllable identification score in AV modality followed by auditory and least in visual modality. This was true in both the groups. Identification scores along with the results of SINFA showed that both auditory and visual modalities play an important role, particularly in challenging listening conditions. The individuals with ANSD were able to make better use of visual cues than the control group, as evident in the visual gain scores. However, acoustic enhancement of speech did not significantly enhance speech perception. When acoustic enhancement and visual cues were simultaneously provided, speech perception was determined mainly by visual cues. The evidence from individual data showed that most of the individuals benefit from AV modality. The scores in AV modality could be predicted from the scores of auditory modality and visual modality. There was no correlation of pure tone average, speech identification scores and duration of ANSD with the auditory or visual gain scores.

The findings indicate that dynamics of speech perception in the AV mode is different between ANSD and control individuals. There is definite benefit of auditory as well as visual cues to individuals with ANSD, suggesting the need to facilitate both the modalities as part of the audiological rehabilitation. The benefits derived are independent of degree of hearing loss, duration of ANSD and speech identification scores. The benefit of the AV modality is present even in the presence of noise. Future studies can focus on independently facilitating the two modalities and testing the benefits in the AV modality of speech perception in individuals with ANSD. The results have important implications in clinical Audiology. The findings indicate that both auditory and visual modality needs to be facilitated in ANSD to enhance speech perception. The acoustic enhancements in the current form have negligible influence. However the inference shall be restricted to the perception of stop consonants.

REFERENCES

Abdala, C., Sininger, Y. S., & Starr, A. (2000). Distortion product otoacoustic emission suppression in subjects with auditory neuropathy. *Ear and hearing*, *21*(6), 542-553.

Alvarenga, K. F., Amorim, R. B., Agostinho-Pesse, R. S., Costa, O. A., Nascimento, L. T., & Bevilacqua, M. C. (2012). Speech perception and cortical auditory evoked potentials in cochlear implant users with auditory neuropathy spectrum disorders. *International journal of pediatric otorhinolaryngology*, *76*(9), 1332-1338.

American National Standards Institute. (1999). *American National Standard Maximum Permissible Ambient Noise Levels for Audiometric Test Rooms*. Standards Secretariat, Acoustical Society of America.

Anderson, E. (2006). *Audiovisual Speech Perception with Degraded Auditory Cues*. Undergraduate honors thesis, The Ohio State University.

ANSI, A. (2004). S3. 6–2004, Specification for audiometers. *American National Standards Institute*.

ANSI. Specifications for Immittance, American National Standards Institute: New York, NY, S3.39-1987 (R1996).

Apoux, F., Tribut, N., Debruille, X., & Lorenzi, C. (2004).Identification of envelope-expanded sentences in normal-hearing and hearing-impaired listeners. *Hearing research*, *189*(1-2), 13-24.

Araki, S., Kawano, A., Seldon, L., Shepard, R.K., Funasaka, S., & Clark, G.M. (1998). Effects of chronic electrical stimulation on spiral ganglion neuron survival and size in deafened kittens. *The Laryngoscope, 108*, 687-95.

Arslan, E., Turrini, M., Lupi, G., Genovese, E., & Orzan, E. (1997). Hearing threshold assessment with auditory brainstem response (ABR) and ElectroCochleoGraphy (ECochG) in uncooperative children. *Scandinavian audiology. Supplementum*, *46*, 32-37.

Baer, T., Moore, B. C., & Gatehouse, S. (1993). Spectral contrast enhancement of speech in noise for listeners with sensorineural hearing impairment: effects on intelligibility, quality, and response times. *Journal of Rehabilitation Research and Development, 30* (1), 49–72.

Barman, A. (2008). *Psycho acoustic profile in Normals and Individuals with Auditory Dys-Synchrony* (Unpublished doctoral thesis). University of Mysore, Mysore.

Berlin, C. I., Hood, L. J., & Rose, K. (2001). On renaming auditory neuropathy as auditory dys-synchrony. *Audiology Today, 13,* 15–17. 32

Berlin, C. I., Hood, L. J., Hurely, A., & Wen, H. (1996). Hearing aids: Only for hearing impaired patients with abnormal otoacoustic emissions. In C.I. Berlin (Ed.), *Hair cells and hearing aids* (pp. 99-111). San Diego: Singular publishing group.

Berlin, C. I., Li, L., Hood, L. J., Morlet, T., Rose, K., & Brashears, S. (2002). Auditory neuropathy/dys-synchrony: after the diagnosis, then what? In *Seminars in hearing* (Vol. 23, No. 03, pp. 209-214).Copyright© 2002 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, USA

Berlin, C. I., Morlet, T., & Hood, L. J. (2003). Auditory neuropathy/dyssynchrony: its diagnosis and management. *Pediatric Clinics*, *50*(2), 331-340.

Berlin, C.I. (1999). Auditory neuropathy. *Seminar in Hearing, 20*, 307-314.

Berlin, C.I., Hood, L.J., Cecola, R.P., Jackson, D.F., & Szabo, P. (1993). Does Type I afferent neuron dysfunction reveal itself through lack of efferent suppression? *Hearing Research, 65*, 40–50.

Berlin, C.I., Hood, L.J., Morlet, T., Wilensky, D., Li, L., & Mattingly, K.R. (2010). Multi-site diagnosis and management of 260 patients with auditory neuropathy/dys-synchrony (auditory neuropathy spectrum disorder). *International Journal of Audiology, 49*, 30 – 43.

Bernstein, L.E., Auer, E.T., Jr., & Takayanagi, S. (2004). Auditory speech detection in noise enhanced by lipreading. *Speech Communication, 44(1),* 5-18.

Bhattacharya, A., & Zeng, F. G. (2007). Companding to improve cochlear-implant speech recognition in speech-shaped noise. *The Journal of the Acoustical Society of America*, *122*(2), 1079-1089.

Bhattacharya, A., Vandali, A., & Zeng, F. G. (2011). Combined spectral and temporal enhancement to improve cochlear-implant speech perception. *The Journal of the Acoustical Society of America*, *130*(5), 2951-2960.

Billett, T. E., Thorne, P. R., & Gavin, J. B. (1989). The nature and progression of injury in the organ of Corti during ischemia. *Hearing research*, *41*(2-3), 189-197.

Bruhn, M. E., & Müller-Walle, J. (1949). *The Mueller-Walle method of lipreading for the hard of hearing*. Volta Bureau.

Buchman, C.A., Roush, P.A., Teagle, H.F., Brown, C.J., Zdanski, C.J., & Grose, J.H. (2006). Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear and Hearing, 27(4)*, 399–408

Buss, E., Hall, J. W., & Grose, J. H. (2004). Temporal Fine-Structure Cues to Speech and Pure Tone Modulation in Observers with Sensorineural Hearing Loss. *Ear and Hearing*, *25* (3), 242–250. http://doi.org/10.1097/01.AUD.0000130796.73809.09

Butinar, D., Zidar, J., Leonardis, L., Popovic, M., Kalaydjieva, L., Angelicheva, D., ... & Starr, A. (1999). Hereditary auditory, vestibular, motor, and sensory neuropathy in a Slovenian Roma (Gypsy) kindred. *Annals of neurology*, *46*(1), 36-44.

Cevette, M. J., Robinette, M. S., Carter, J., & Knops, J. L. (1995). Otoacoustic emissions in sudden unilateral hearing loss associated with multiple sclerosis. *Journal of American academy of audiology*, *6*, 197-197.

CHABA, Committee on Hearing and Bioacoustics, Working Group on Speech Understanding and Aging. (1988). Speech understanding and aging. *Journal of the Acoustical Society of America, 83*, 859–895.

Cheng, X., Li, L., Brashears, S., Morlet, T., Ng, S. S., Berlin, C., & Keats, B. (2005). Connexin 26 variants and auditory neuropathy/dys-synchrony among children in schools for the deaf. *American Journal of Medical Genetics Part A*, *139*(1), 13-18.

Chisin, R., Perlman, M., & Sohmer, H. (1979). Cochlear and brain stem responses in hearing loss following neonatal hyperbilirubinemia. *Annals of Otology, Rhinology & Laryngology, 88*(3), 352-357.

Cienkowski, K. M. (1999). Auditory-visual speech perception across the lifespan [Doctoral Dissertation, University of Minnesota, 1999]. *Dissertation Abstracts International*, *60(1)*, 116.

Cienkowski, K. M., & Carney, A. E. (2002). Auditory-visual speech perception and aging. *Ear and Hearing*, *23*, 439–449.

Cienkowski, K. M., & Carney, A. E. (2004). The Integration of Auditory-Visual Information for Speech in Older Adults. *Journal of Speech-Language Pathology and Audiology*, *28(4)*, 169-172.

Cone-Wesson, B., Rance, G & Sininger, Y. (2001). Amplification and Rehabilitation Strategies for Patients with Auditory Neuropathy. *Auditory Neuropathy: A New Perspectives on Hearing Disorders.* Singular Publishing.

Corley, V. M., & Crabbe, L. S. (1999). Auditory neuropathy and a mitochondrial disorder in a child: case study. *Journal of American academy of audiology*, *10*, 484-488.

Davis, H., & Hirsh, S.K. (1979). A slow brainstem response for low frequency audiometry. *Audiology, 18*, 445-465.

Deltenre, P., Mansbach, A. L., Bozet, C., Christiaens, F., Barthelemy, P., Paulissen, D., & Renglet, T. (1999). Auditory neuropathy with preserved cochlear microphonics and secondary loss of otoacoustic emissions. *Audiology*, *38*(4), 187-195.

Deltenre, P., Mansbach, A. L., Bozet, C., Clercx, A., & Hecox, K. E. (1997). Auditory neuropathy: a report on three cases with early onsets and major neonatal illnesses. *Electroencephalography and Clinical Neurophysiology/Evoked Potentials Section*, *104*(1), 17-22.

Derbyshire, A. J., & Davis, H. (1935). The action potentials of the auditory nerve. *American Journal of Physiology-Legacy Content*, *113*(2), 476-504. DOI: 10.1007/s10162-004-5014-5

Doyle, K. J., Sininger, Y., & Starr, A. (1998). Auditory neuropathy in childhood. *The laryngoscope*, *108*(9), 1374-1377. https://doi.org/10.1097/00005537-199809000-00022

Drullman, R., Festen, J. M., & Plomp, R. (1994). Effect of reducing slow temporal modulations on speech reception. *The Journal of the Acoustical Society of America*, *95*(5), 2670-2680.

Dunn, T. J., Baguley, T., & Brunsden, V. (2014). From alpha to omega: A practical solution to the pervasive problem of internal consistency estimation. *British Journal of Psychology*, *105*(3), 399-412.

Dürr, A., Cossee, M., Agid, Y., Campuzano, V., Mignard, C., Penet, C., & Koenig, M. (1996). Clinical and genetic abnormalities in patients with Friedreich's ataxia. *New England Journal of Medicine*, *335*(16), 1169-1175. DOI: 10.1056/NEJM199610173351601

Durrant, J. D., Wang, J., Ding, D. L., & Salvi, R. J. (1998). Are inner or outer hair cells the source of summating potentials recorded from the round window?. *The Journal of the Acoustical Society of America*, *104*(1), 370-377.

Erber, N. (1969). Interaction of audition and vision in the recognition of oral speech stimuli. *Journal of Speech and Hearing Research, 12*, 423–425. 34

Erber, N.P. (1972). Auditory, visual, and auditory-visual recognition of consonants by children with normal and impaired hearing. *Journal of Speech and Hearing Research. ,15*, 413–422.

Gnanateja, G. N., & Barman,A. (2011). *Relation between consonant perception and psychoacoustic measures in individuals with auditory dys-synchrony*. (Unpublished dissertation). University of Mysore, Mysore.

Gorga, M. P., Stelmachowicz, P. G., Barlow, S. M., & Brookhouser, P. E. (1995). Case of recurrent, reversible, sudden sensorineural hearing loss in a child. *Journal of American academy of audiology*, *6*, 163-163.

Grant, K. W., & Seitz, P. F. (2000). The recognition of isolated words and words in sentences: Individual variability in the use of sentence context. *Journal of the Acoustical Society of America*, *107*, 1000–1011.

Grant, K. W., Walden, B. E., & Seitz, P. F. (1998). Auditory-visual speech recognition by hearing-impaired subjects: Consonant recognition, sentence recognition, and auditory-visual integration. *Journal of the Acoustical Society of America*, *103*, 2677–2690.

Hassan, D. M. (2011). Perception of temporally modified speech in auditory neuropathy. *International Journal of Audiology*, *50* (1), 41–49. http://doi.org/10.3109/14992027.2010.520035

Hood, L. J. (1998). Auditory neuropathy: what is it and what can we do about it?. *The Hearing Journal*, *51*(8), 10-12.

Hood, L. J., Berlin, C. I., Bordelon, J., & Rose, K. (2003). Patients with auditory neuropathy/dys-synchrony lack efferent suppression of transient evoked otoacoustic emissions. *Journal of the American Academy of Audiology*, *14*(6), 302-313.

Hood, L.J., Wilensky, D., Li, L., & Berlin, C.I. (2004). The role of FM technology in the management of patients with auditory neuropathy/dys-synchrony. In: Fabry DA, De Conde Johnson C, eds. ACCESS: Achieving, Clear Communication Employing Sound Solutions–2003. Proceedings of the 1st International FM Conference: Chicago. IL. 35

Houtgast, T., & Steeneken, H. J. (1985). A review of the MTF concept in room acoustics and its use for estimating speech intelligibility in auditoria. *The Journal of the Acoustical Society of America*, *77*(3), 1069-1077.

Jeong, S.W., Kim, L.S., Kim, B.Y., Bae, Y.W., & Kim, J.R. (2007). Cochlear implantation in children with auditory neuropathy: Outcomes and rationale. *Acta Oto-Laryngologica*, *558,* 36-43.

Jijo, P. M., & Yathiraj, A. (2012).Audiological characteristics and duration of the disorder in individuals with auditory neuropathy spectrum disorder (ANSD)–a retrospective study. *Journal Indian Speech and Hearing Association*, *26*(1), 17-26.

Jijo, P.M. (2015). *The Effect of Intensity and Temporal Enhancement on Speech Perception in Individuals with Auditory Neuropathy Spectrum Disorder (ANSD*). (Unpublished doctoral dissertation).University of Mysore, Mysuru.

Jones, B., & Kenward, M. G. (2014). *Design and analysis of cross-over trials*. Chapman and Hall/CRC. New York.

Katona, G., Büki, B., Farkas, Z., Pytel, J., Simon-Nagy, E., & Hirschberg, J. (1993). Transitory evoked otoacoustic emission (TEOAE) in a child with profound hearing loss. *International journal of pediatric otorhinolaryngology*, *26*(3), 263-267.

Kinzie, C. E., & Kinzie, R. (1931). *Lip-reading for the deafened adult: with a foreword by His Grace the Duke of Montrose*. The John C. Winston company.

Kovach, M. J., Lin, J. P., Boyadjiev, S., Campbell, K., Mazzeo, L., Herman, K., & Gelber, D. (1999). A unique point mutation in the PMP22 gene is associated with Charcot-Marie-Tooth disease and deafness. *The American Journal of Human Genetics*, *64*(6), 1580-1593.

Kraus, N., Bradlow, A.R., Cheatham, J., Cunningham, C.D., King, D.B., Koch, T.G., Nicol, T.J., McGee, L.K., Stein, L.K., & Wright, B.A. (2000). Consequences of neural asynchrony : A case of auditory neuropathy. *Journal of the Association for Research in Otolaryngology, 1 (1),* 33–45.

Kraus, N., Özdamar, Ö., Stein, L., & Reed, N. (1984). Absent auditory brain stem response: peripheral hearing loss or brain stem dysfunction?. *The Laryngoscope*, *94*(3), 400-406.

Krause, J. C., & Braida, L. D. (2002). Investigating alternative forms of clear speech: The effects of speaking rate and speaking mode on intelligibility. *The Journal of the Acoustical Society of America*, *112*(5), 2165-2172.

Kumar, U. A., & Jayaram, M. (2011). Speech perception in individuals with auditory dys-synchrony, *The Journal of Laryngology and Otology, 125*, 236–245.

Kumar, U. A., & Jayaram, M. (2013). Speech perception in individuals with auditory dys-synchrony: effect of lengthening of voice onset time and burst duration of speech segments. *The Journal of Laryngology & Otology*, *127*(7), 656-665.

Kumar, U.A. (2006). *Perception of Some Temporal Parameters of Speech in Individuals with Auditory Dys-synchrony*(Unpublished doctoral dissertation).University of Mysore, Mysore.

Kumar, U.A., & Jayaram, M. (2005). Auditory processing in individuals with auditory neuropathy, *Behavioral and Brain Functions*, 1-21.

Kumar, U.A., & Jayaram, M. (2006). Prevalence and audiological characteristics in individuals with auditory neuropathy/auditory dys-synchrony. *International Journal of Audiology, 45(6),* 360-366.

Lachs, L., Pisoni, D.B., & Iler Kirk, K. (2001). Use of audiovisual information in speech perception by prelingually deaf children with cochlear implants: A first report. *Ear and Hearing*, 22, 236-251. 36

Lee, J. S., McPherson, B., Yuen, K. C., & Wong, L. L. (2001). Screening for auditory neuropathy in a school for hearing impaired children. *International journal of pediatric otorhinolaryngology*, *61*(1), 39-46.

Leonardis, L., Zidar, J., Popovič, M., Timmerman, V., Löfgren, A., Van Broeckhoven, C., & Butinar, D. (2000). Hereditary motor and sensory neuropathy associated with auditory neuropathy in a Gypsy family. *Pflügers Archiv*, *439*(1), r208-r210.

MacDonald, W.I. (1980). Physiological consequences of demyelination. In: Sumaer AJ, ed. The physiology of peripheral nerve disease. Philadelphia, W.B. Saunders, 265-86.

MacLeod, A., & Summerfield, Q. (1987). Quantifying the contribution of vision to speech perception in noise. *British Journal of Audiology*, *21*, 131-141.

Madden, C., Rutter, M., Hilbert, L., Greinwald Jr, J. H., & Choo, D. I. (2002). Clinical and audiological features in auditory neuropathy. *Archives of otolaryngology–head & neck surgery*, *128*(9), 1026-1030.

Marsh, R. R. (2002). Is it auditory dys-synchrony?. *Audiology Today*, *14*(3), 36-37.

Maruthy. S., & Geetha, C. (2011). Audiovisual perception and processing in individuals with auditory dys-synchrony. Unpublished project funded by AIISH research fund submitted to AIISH, Mysore.

Massaro, D. W. (1998). Perceiving talking faces: From speech perception to a behavioral principle. Cambridge, Massachusetts: MIT Press.

Mathai, J. P., & Yathiraj, A. (2013). Audiological findings and aided performance in individuals with auditory neuropathy spectrum disorder (ANSD)–a retrospective study. *Journal of Hearing Science*, *3*(1), 18-26.

Mattys, S. L., Davis, M. H., Bradlow, A. R., & Scott, S. K. (2012). Speech recognition in adverse conditions: A review. *Language and Cognitive Processes*, *27*(7-8), 953-978.

McDonald WI, & Sears TA. (1970). The effects of experimental demyelination on conduction in the central nervous system. *Brain, 91*, 583-598.

McMahon., Pattuzi, R., Gibson, W., & Sanli, H. (2008). Frequency-Specific Electrocochleography Indicates that Presynaptic and Postsynaptic Mechanisms of Auditory Neuropathy Exist. *Ear and Hearing, 29*, 314–325. 37

Michalewski, H.J., Starr, A., Nguyen, T.T., Kong, Y-Y., & Zeng, F.G. (2005). Auditory temporal processes in normal- hearing individuals and in patients with auditory neuropathy. *Clinical Neurophysiology, 116*, 669–680.

Middelweerd, M., & Plomp, R. (1984). The effect of speech reading on the speech reception threshold of sentences in noise. *Journal of the Acoustical Society of America, 82*, 2145–2147.

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

Moore, B. C. (1973). Frequency difference limens for short-duration tones. *The Journal of the Acoustical Society of America, 54*(3), 610-619.

Moore, B. C. (2008). The role of temporal fine structure processing in pitch perception, masking, and speech perception for normal-hearing and hearing-impaired people. *Journal of the Association for Research in Otolaryngology*, *9*(4), 399-406.

Munhall, K.G., Kroos, C., Jozan, C., & Vatikiotis-Bateson, E. (2004). Spatial frequency requirements for audiovisual speech perception. *Perceptions and Psychophysics, 66 (4),* 574 – 583.

Nagarajan, S. S., Wang, X., Merzenich, M. M., Schreiner, C. E., Johnston, P., Jenkins, W. M., & Tallal, P. (1998). Speech modifications algorithms used for training

language learning-impaired children. *IEEE Transactions on Rehabilitation Engineering*, *6*(3), 257-268.

Narne, V. K. (2013). Temporal processing and speech perception in noise by listeners with auditory neuropathy. *PloS one*, *8*(2), e55995.

Narne, V. K., & Vanaja, C. S. (2008). Effect of envelope enhancement on speech perception in individuals with auditory neuropathy. *Ear and hearing*, *29*(1), 45-53.

Narne, V. K., & Vanaja, C. S. (2009a). Perception of envelope-enhanced speech in the presence of noise by individuals with auditory neuropathy. *Ear and hearing*, *30*(1), 136-142.

Narne, V. K., & Vanaja, C. S. (2009b). Perception of speech with envelope enhancement in individuals with auditory neuropathy and simulated loss of temporal modulation processing. *International journal of audiology*, *48*(10), 700-707.

Narne, V. K., Barman, A., Deepthi, M., & Shachi. (2014). Effect of companding on speech recognition in quiet and noise for listeners with ANSD. *International journal of audiology*, *53*(2), 94-100.

Narne, V. K., Prabhu, P., Chandan, H. S., & Deepthi, M. (2016). Gender Differences in Audiological Findings and Hearing Aid Benefit in 255 Individuals with Auditory Neuropathy Spectrum Disorder: A Retrospective Study. *Journal of the American Academy of Audiology*, *27*(10), 839-845.

Neely, K. K. (1956). Effects of visual factors on intelligibility of speech. *Journal of the Acoustical Society of America, 28*, 1276-1277.

Oxenham, A. J., Simonson, A. M., Turicchia, L., & Sarpeshkar, R. (2007). Evaluation of companding-based spectral enhancement using simulated cochlear-implant processing. *The Journal of the Acoustical Society of America*, *121*(3), 1709-1716.

Pender, M.P., & Sears, T.A. (1984). The pathophysiology of acute experimental allergic encephalomyelitis in the rabbit. *Brain, 101,* 699-726. 38

Plomp, R. (1988). The negative effect of amplitude compression in multichannel hearing aids in the light of the modulation-transfer function. *The Journal of the Acoustical Society of America*, *83*(6), 2322-2327.

Plude, D. J., & Doussard-Roosevelt, J. A. (1989). Aging, selective attention, and feature integration. *Psychology and Aging*, *4*, 98115.

Powers, N. L., Salvi, R. J., Wang, J., Spongr, V., & Qiu, C. X. (1995). Elevation of auditory thresholds by spontaneous cochlear oscillations. *Nature*, *375*(6532), 585.

Prabhu, P., & Barman, A. (2017). Effectiveness of Low Cut Modified Amplification using Receiver in the Canal Hearing Aid in Individuals with Auditory Neuropathy Spectrum Disorder. *International archives of otorhinolaryngology*, *21*(3), 243-249.

Prabhu, P., Avilala, V. K. Y., & Manjula, P. P. (2012). Predisposing factors in individuals with late-onset auditory dys-synchrony. *Asia Pacific Journal of Speech, Language and Hearing*, *15*(1), 41-50.

Prabhu, P., Avilala, V., & Barman, A. (2011). Speech perception abilities for spectrally modified signals in individuals with auditory dys-synchrony. *International journal of audiology*, *50*(5), 349-352.

Prieve, B. A., Gorga, M. P., & Neely, S. T. (1991). Otoacoustic emissions in an adult with severe hearing loss. *Journal of Speech, Language, and Hearing Research*, *34*(2), 379-385.

Ramirez, J., & Mann, V. (2005).Using auditory-visual speech to probe the basis of noise-impaired consonant–vowel perception in dyslexia and auditory neuropathy. *The Journal of the Acoustical Society of America*, *118*(2), 1122-1133.

Rance, G. (2005). Auditory neuropathy/dys-synchrony and its perceptual consequences. *Trends in Amplification*, *9*(1), 1-43.

Rance, G., & Barker, E. J. (2008). Speech perception in children with auditory neuropathy/dyssynchrony managed with either hearing aids or cochlear implants. *Otology & Neurotology*, *29*(2), 179-182.

Rance, G., Barker, E., Mok, M., Dowell, R., Rincon, A., & Garratt, R. (2007). Speech perception in noise for children with auditory neuropathy/dys-synchrony type hearing loss. *Ear and Hearing*, *28*, 351–360.

Rance, G., Beer, D.E., Cone-Wesson, B., Shepard, R., Dowell, R.C., King, A.M., Rickards, F.W., & Clark ,G.M. (1999). Clinical findings for a group of infants and young children with auditory neuropathy. *Ear and Hearing*, 20, 238-252.

Rance, G., Cone-Wesson, B., Wunderlich, J., & Dowell, R.C. (2002). Speech perception and cortical event related potentials in children with auditory neuropathy. *Ear and Hearing*, *23*, 239-253.

Rance, G., Corben, L. A., Du Bourg, E., King, A., & Delatycki, M. B. (2010).Successful treatment of auditory perceptual disorder in individuals with Friedreich ataxia. *Neuroscience*, *171*(2), 552-555.

Rance, G., McKay, C., & Grayden, D. (2004). Perceptual characterization of children with auditory neuropathy. *Ear and Hearing* 25: 34–46

Rance, G., Ryan, M. M., Carew, P., Corben, L. A., Yiu, E., Tan, J., & Delatycki, M. B. (2012). Binaural speech processing in individuals with auditory neuropathy. *Neuroscience*, *226*, 227-235.

Rapin, I., & Gravel, J. (2003). "Auditory neuropathy": physiologic and pathologic evidence calls for more diagnostic specificity. *International journal of pediatric otorhinolaryngology*, *67*(7), 707-728. https://doi.org/10.1016/S0165-5876(03)00103-4

Rasminsky, M., & Sears, T.A. (1972). Internodal conduction in undissected demyelinated nerve fibers. *Journal of Physiology*, *221*, 323-350. 39

Rodríguez-Ballesteros, M., del Castillo, F. J., Martín, Y., Moreno-Pelayo, M. A., Morera, C., Prieto, F., & Navas, C. (2003). Auditory neuropathy in patients carrying mutations in the otoferlin gene (OTOF). *Human mutation*, *22*(6), 451-456.

Rosenblum, L. D. (2005). Primacy of multimodal speech perception. In D. B. Pisoni, & E. R. Remez (Eds.), The handbook of speech perception (pp. 51-78). Blackwell Publishing.

Ross, L.A., Saint-Amour, D., Leavitt, V., Javitt, D.C. & Foxe J.J. (2006). Do you see what I'm saying? Optimal Visual gain of Speech Comprehension in noisy environments. *Cerebral Cortex, 17(5),* 1147-53.

Roux, I., Safieddine, S., Nouvian, R., Grati, M. H., Simmler, M. C., Bahloul, A, & Triller, A. (2006). Otoferlin, defective in a human deafness form, is essential for exocytosis at the auditory ribbon synapse. *Cell, 127*(2), 277-289.

Rudmann, D.S., McCarley, J.S., & Kramer, A.F. (2003). Bimodal display augmentation for improved speech comprehension. *Human Factors, 45,* 329-336.

Santarelli, R., Scimemi, P., Monte, E.D., Genovese, E., & Arslan, E. (2006). Auditory neuropathy in systemic sclerosis: a speech perception and evoked potential study before and after cochlear implantation. *European archives of Oto-rhino-laryngology, 263*, 809-815.

Satya-Murti, S., Wolpaw, J. R., Cacace, A. T., & Schaffer, C. A. (1983). Late auditory evoked potentials can occur without brain stem potentials. *Electroencephalography and clinical neurophysiology*, *56*(4), 304-308.

Shallop, J., Peterson, A., Facer, G., Fabry, L., & Discoll, C. (2001).Cochlear implants in five cases of auditory neuropathy: Postoperative findings and progress. *Laryngoscope, 111*, 555-562. 40

Shallop, J.K. (2002). Auditory neuropathy/dys-synchrony in adults and children. *Seminars in Hearing*, 23 (3), 215–223.

Shannon, R.V. (1993) Quantitative comparison of electrically and acoustically evoked auditory perception: Implications for location of perceptual mechanisms. *Progress for Brain Research*, *97*, 261-269.

Shannon, R.V., Zeng, F.G., Kamath, V., Wygonski, J., & Ekelid, M. (1995). Speech recognition with primarily temporal cues. *Science*, *270*, 303-304.

Sharma, A., & Dorman, M. F. (1999). Cortical auditory evoked potential correlates of categorical perception of voice-onset time. *The Journal of the Acoustical Society of America*, *106*(2), 1078-1083. https://doi.org/10.1121/1.428048

Shi, F., Kempfle, J. S., & Edge, A. S. (2012). Wnt-responsive Lgr5-expressing stem cells are hair cell progenitors in the cochlea. *Journal of Neuroscience*, *32*(28), 9639-9648.

Shirane, M., & Harrison, R. V. (1987). The effects of hypoxia on sensory cells of the cochlea in chinchilla. *Scanning microscopy*, *1*(3), 1175-1183.

Shoop, C., & Binnie, C. A. (1979). The effects of age upon the visual perception of speech. *Scandinavian Audiology*, *8*, 3–8.

Sininger, Y. (2002). Identification of auditory neuropathy in infants and children. *Seminar in Hearing*, *23(3)*, 193-200.

Sininger, Y. S., & Starr, A. (1997). Auditory neuropathy in children. *SIG 9 Perspectives on Hearing and Hearing Disorders in Childhood*, *7*(1), 6-11.

Sininger, Y. S., & Trautwein, P. (2002).Electrical stimulation of the auditory nerve via cochlear implants in patients with auditory neuropathy. *Annals of Otology, Rhinology & Laryngology, 111*(5_suppl), 29-31.

Sininger, Y. S., Hood, L. J., Starr, A., Berlin, C. I., & Picton, T. W. (1995). Hearing loss due to auditory neuropathy. *Audiology Today*, *7*(2), 10-13.

Sininger, Y., & Hayes, D (2008, June). At the Consensus Conference on Auditory Neuropathy/Dys-synchrony, Como, Italy.

Sininger, Y., & Oba, S. (2001). Patients with auditory neuropathy: who are they and what can they hear. *Auditory neuropathy: A new perspective on hearing disorders*, 15-35.

Spoendlin, H. (1974). Optic and cochleovestibular degenerations in hereditary ataxias: II. Temporal bone pathology in two cases of Friedreich's ataxia with vestibulo-cochlear disorders. *Brain*, *97*(1), 41-48.

Spreng, M. (2000). Central nervous system activation by noise. *Noise and health*, *2*(7), 49.

Star, A., Sininger, Y., Winter, M., Derby, M.J., & Michalewski, H.J. (1998). Transient deafness due to temperature sensitive Auditory neuropathy. *Ear and Hearing*, *19*, 169-79.

Starr, A., Isaacson, B., Michalewski, H. J., Zeng, F. G., Kong, Y. Y., Beale, P., ... & Lesperance, M. M. (2004). A dominantly inherited progressive deafness affecting distal auditory nerve and hair cells. *Journal of the Association for Research in Otolaryngology*, *5*(4), 411-426.

Starr, A., Kim, C.S., Kim, D., Butinar, D., & Linthicum, F., (2001b, July). Neuropathology of type I auditory neuropathy. Abstracts of the 17th Biennal Symposium of the International Evoked Response Audiometry Study Group (IERASG), Vancouver, BC.

Starr, A., McPherson, D., Patterson, J., Don, M., Luxford, W., Shannon, R, Sininger, Y., Tonakawa, L., et al. (1991). Absence of both auditory evoked potential and auditory percepts dependent on timing cues. *Brain*, *114*, 1157-1180.

Starr, A., Michalewski, H.J., Zeng, F.G., Brooks, S.F., Linthicum, F. Kim, C.S., Winnier, D., & Keats, B. (2003). Pathology and physiology of auditory neuropathy with a novel mutation in the MPZ gene**.** *Brain*, *126***,** 1604-1619.

Starr, A., Picton, T. W., Sininger, Y., Hood, L. J., & Berlin, C. I. (1996). Auditory neuropathy. *Brain*, *119*(3), 741-753.

Starr, A., Picton, T.W., & Kim, R. (2001a). Pathophysiology of auditory neuropathy. In: Y. Sininger, & A. Starr (Eds.), *Auditory neuropathy: A new perspective on hearing disorder* (67-82). Canada: Singular publishing group.

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

Starr, A., Sininger, Y., Nguyen, T., Michalewski, H.J., Oba, S., & Abdala, C. (2001b). Cochlear receptor (microphonic and summating potentials, otoacoustic emissions) and auditory pathway (auditory brainstem potentials) activity in auditory neuropathy. *Ear and Hearing, 22*, 91-99. 42

Starr, A., Sininger, Y., Winter, M., Derebery, M. J., Oba, S., & Michalewski, H. J. (1998). Transient deafness due to temperature-sensitive auditory neuropathy. *Ear and hearing*, *19*(3), 169-179.

Starr, A., Sininger, Y.S., & Pratt, H. (2000). The varieties of auditory neuropathy. *Journal of Basic Clinical Physiology and Pharmacology, 11(3)*,215–230

Starr, A., Zeng, F., Michalewski, H., & Moser, T. (2008). *Perspectives on Auditory Neuropathy: Disorders of Inner Hair Cell, Auditory Nerve, and Their Synapse. In: The Senses: A Comprehensive Reference*. New York: Academic Press.

Stephanova, D. I., Daskalova, M., & Alexandrov, A. S. (2005). Differences in potentials and excitability properties in simulated cases of demyelinating neuropathies. Part I. *Clinical neurophysiology*, *116*(5), 1153-1158.

Sumby, W., & Pollack, I. (1954).Visual contribution to speech intelligibility in noise. *Journal of Acoustic Society of America, (26)*, 212–215.

Tallal, P., Miller, S. L., Bedi, G., Byma, G., Wang, X., Nagarajan, S. S., & Merzenich, M. M. (1996). Language comprehension in language-learning impaired children improved with acoustically modified speech. *Science*, *271*(5245), 81-84.

Tang, T. P., McPherson, B., Yuen, K. C., Wong, L. L., & Lee, J. S. (2004). Auditory neuropathy/auditory dys-synchrony in school children with hearing loss: Frequency of occurrence. *International Journal of Pediatric Otolaryngology, 168*, 175–183.

Trautwein, P. (2002, November). *Auditory Neuropathy: Diagnosis and Case Management.* Paper presented at 4th ACFOS international conference on The Impact of Scientific Advances on the Education of Deaf Children, Paris, France.

Trautwein, P.G., Shallop, J., Fabry, L. & Friedman, R. (2001). Cochlear Implantation of Auditory Neuropathy. *Auditory Neuropathy, A New Perspectives on Hearing Disorders.* Singular Publishing.

Turicchia, L., & Sarpeshkar, R. (2005). A bio-inspired companding strategy for spectral enhancement. *IEEE transactions on speech and audio processing*, *13*(2), 243-253.

Tye-Murray, N., Sommers, M. S., & Spehar, B. (2007). Audiovisual integration and lip-reading abilities of older adults with normal and impaired hearing. *Ear and Hearing*, *28(5)*, 656-668.

Tyler, R. S., Fernandes, M., & Wood, E. J. (1980). Masking, temporal integration and speech intelligibility in individuals with noise-induced hearing loss. In *Disorders of auditory function* (pp. 211-236).

Vanaja, C. S., & Manjula, P. (2004). LLR as a measure of benefit derived from hearing devices with auditory dys-synchrony. In *first conference on Auditory Neuropathy* (pp. 136-146). Department of Speech pathology and Audiology, National Institute of Mental Health and Neuro sciences.

Varga, R., Kelley , P.M., Keats, B.J., Starr, A., & Leal , S.M. (2003). Nonsyndromic recessive auditory neuropathy is the result of mutations in the otoferlin (OTOF) gene. *Journal of Medical Genetetics, 40,* 45– 50.

Vasistha, S., & Barman, A. (2012). *Perception of spectrally enhanced speech through companding in individuals with auditory neuropathy*. (Unpublished dissertation). University of Mysore, Mysore.

Venkatesan, S. (2009). Ethical guidelines for bio behavioral research involving human subject. All India Institute of Speech and Hearing. Mysuru. doi:10.1017/CBO9781107415324.004

Vinay, C., & Moore, B.C.J. (2007). Ten (HL)-test results and psychophysical tuning curves for subjects with auditory neuropathy. *International Journal of Audiology*, *46(1)*, 39–46. 44

Vohr, B. R., Barry, L., Rapisardi, G., O'Dea, C., Brown, L., Peucker, M., & Oh, W. (1989). Abnormal brain-stem function (brain-stem auditory evoked response) correlates with acoustic cry features in term infants with hyperbilirubinemia. *The Journal of pediatrics*, *115*(2), 303-308.

Walden, B. E., Busacco, D. A., & Montgomery, A. A. (1993). Benefit from visual cues in auditory-visual speech recognition by middle-aged and elderly persons. *Journal of Speech and Hearing Research*, *36*, 431–436.

Wang, M. D., & Bilger, R. C. (1973). Consonant confusions in noise: A study of perceptual features. *The Journal of the Acoustical Society of America*, *54*(5), 1248-1266.

Wang, Q., Gu, R., Han, D., & Yang, W. (2003). Familial auditory neuropathy. *The Laryngoscope*, *113*(9), 1623-1629.

Yadav, A.V., & Yathiraj, A. (2010). *Effect of listening training in perception of voicing of stops in individuals with Auditory Dys synchrony* (Unpublished dissertation). Mysore University, Mysore.

Yasunaga, S. I., Grati, M. H., Chardenoux, S., Smith, T. N., Friedman, T. B., Lalwani, A. K., & Petit, C. (2000). OTOF encodes multiple long and short isoforms: genetic evidence that the long ones underlie recessive deafness DFNB9. *The American Journal of Human Genetics*, *67*(3), 591-600.

Yathiraj, A., & Vijayalakshmi, C. S. (2005). Phonemically Balanced Word List in Kannada: Developed in Department of Audiology. *Mysore: AIISH*.

Zeng, F. G. (2004). Trends in cochlear implants. *Trends in amplification*, *8*(1), 1-34.

Zeng, F. G., & Liu, S. (2006). Speech Perception in Individuals with Auditory Neuropathy. *Journal of Speech, Language, and Hearing Research, (49)*, 367–380.

Zeng, F. G., Oba, S., & Starr, A. (2001). Supra threshold processing deficits due to desynchronous neural activitiesin auditory neuropathy. In D.J. Breebaart, A.J.M. Houstma, A. Kohlrausch, V.F. Prijs and R. Schoonhoven (eds.), *Physiological and Psychophysical Bases of Auditory Function* (pp. 365–372). Maastricht, Netherlands: Shaker Publishing BV.

Zeng, F.G., Kong, Y.Y., Michalewski, H.J., & Starr, A. (2005). Perceptual consequences of disrupted auditory nerve activity. *Journal of Neurophysiology 93*, 3050–3063

Zeng, F.G., Oba, S., Garde, S., Sininger, Y., & Starr, A. (1999). Temporal and speech processing deficits in auditory neuropathy. *NeuroReport, 10,* 3429-3435.

Zhou, R., Abbas, P.J. & Assouline, J.S. (1995). Electrically evoked auditory brainstem responses in myelin-deficient mice. *Hearing Research*, 88, 98-106. 45

APPENDIX 1

Demographic details and audiological findings of 40 participants with ANSD.

No	Age (years)/ Gender	Duration of loss (years)	PTA (dB HL)		SIS (%)		Tymp	Reflex		OAE /CM	ABR	LLR
			R	L	R	L	R/L	Ipsi	contra	R/L	R/L	R/L
1	22/F	04	37.50	50.00	36	32	A/A	NR	NR	P	NR	NR
2	16/ F	02	42.40	32.50	00	00	Ad/A	NR	NR	P	NR	NR
3	16/M	03	48.75	50.00	00	00	A/A	NR	NR	P	NR	P
4	23/M	04	22.50	26.25	24	16	A/A	NR	NR	P	NR	P
5	26/F	06	28.75	22.50	48	80	As/As	NR	NR	P	NR	P
6	35/M	01	30.00	22.50	40	44	A/A	NR	NR	P	NR	P
7	21/M	02	31.25	35.00	68	44	A/A	NR	NR	P	NR	NR
8	28/M	01	10.00	12.50	76	88	A/A	NR	NR	P	NR	P
9	16/F	01	27.50	27.50	64	88	A/A	NR	NR	P	NR	NR
10	35/F	03	45.00	43.75	20	24	As/As	NR	NR	P	NR	P
11	36/M	12	46.75	47.50	84	80	A/As	NR	NR	P	NR	p
12	35/M	12	55.00	55.00	44	52	As/A	NR	NR	NR/P	NR	P
13	30/F	01	21.25	27.50	00	00	As/AS	NR	NR	P	NR	P/NR
14	20/F	05	17.50	15.00	96	96	As/As	NR	NR	P	NR	P
15	16/M	03	37.50	28.75	64	68	A/A	NR	NR	P	NR	NR
16	19/M	04	25.00	20.00	96	56	A/A	P	P	P	NR	NR
17	18/F	04	36.25	23.75	76	84	Ad/A	NR	NR	P	NR	P
18	22/M	03	37.50	33.50	48	60	Ad/Ad	NR	NR	P	NR	P
19	36/F	12	50.00	28.75	84	88	A/A	NR	NR	P/P	NR	P
20	22/F	07	33.75	21.25	46	52	A/A	NR	NR	P	NR	P
21	20/F	01	26.25	18.75	00	00	As/As	NR	NR	NR/P	NR	P
22	21/F	02	13.75	17.50	96	100	A/A	P	P	P/P	NR	P
23	21/F	10	38.75	43.70	60	20	As/A	NR	NR	P	NR	P
24	35/F	20	3.75	7.50	72	44	A/A	NR	NR	P	NR	P
25	21/F	06	50.00	41.25	00	00	A/A	NR	NR	P	NR	NR
26	18/M	03	28.75	25.00	92	96	A/A	NR	NR	P	NR	P
27	16/F	01	37.50	28.75	72	80	As/As	NR	NR	P	NR	P
28	17/M	04	32.50	41.25	36	16	As/A	NR	NR	P/P	NR	P
29	35/M	01	22.50	18.75	64	80	A/A	NR	NR	P	NR	P
30	35/M	20	31.25	30.00	76	76	A/A	NR	NR	P	NR	P
31	25/M	13	28.30	46.60	28	12	Ad/Ad	NR	NR	P	NR	NR
32	19/F	03	41.25	38.75	84	84	A/A	NR	NR	P	NR	NR
33	21/M	01	18.75	25.00	100	56	As/A	NR	NR	P	NR	P
34	26/M	06	36.25	47.50	32	28	A/A	NR	NR	P	NR	p
35	18/F	01	48.75	52.50	60	40	A/A	NR	NR	P	NR	NR
36	16/F	01	38.75	20.00	36	56	A/A	P	P	P	NR	P

37	24/M	10	43.75	30.00	88	84	Ad/A	NR	NR	P	NR	P
38	17/F	07	27.50	33.75	98	88	As/As	NR	NR	P	NR	NR
39	36/F	20	55.00	36.67	80	84	A/A	NR	NR	P	NR	P
40	23/F	05	35.00	26.25	32	60	A/A	NR	NR	P	NR	NR

Note. PTA: puretone average, SIS: speech identification score, Tym: tympanometry, Reflex; acoustic reflex, OAE: otoacoustic emissions, CM: cochlear microphonics, ABR: auditory brainstem response, LLR: late latency response, NR: no response, P: present, R: right ear. L: left ear, ipsi: ipsilateral, contra: contralateral.