1. INTRODUCTION

"To form a fist our five fingers have to come together in congruence. In the same way, human beings are blessed by five senses vision, hearing, touch, taste, smell. None of our senses functions in absolute isolation from others. All the senses contribute to provide a meaning for experience in life, but hearing & vision, the distance senses are most crucial."

Bess & MC Conell (1981)

1.1 Introduction of Hearing:

Hearing is act of perceiving sound. Hearing is an important distance sense. At a basic level, it is important for us in order to remain in touch with our auditory environment which contains even basic warning signals and at a higher level, it is considered the primary channel for verbal communication.

1.2 Definition of Hearing Impairement:

"Hearing impairment is used to mean a deviation or change for the worse in either auditory structure or auditory function, usually outside the range of normal." **(ASHA, 1981)**.

According to **WHO (1980)** "Impairment is any loss or abnormality of physiological or anatomical structure or function".

Hearing loss is thus characterize by decreased hearing sensitivity to sound in comparison to normal. The presence of a hearing loss affects functioning at different levels. The effects depend on the type and degree of hearing loss. Hearing can be deteriorated by multiple factors. How ever, know a days auditory neuropathy (AN) is one of the prominent cause of hearing loss.

1.3 Introduction to Auditory Neuropathy:

The term 'Auditory Neuropathy' was originally described by **(Starr et al., 1996)**. Other workers have preferred terms such as 'Auditory Dys-synchrony',**(Berlin et al., 2002)** 'Auditory Dys-synchrony' or 'Auditory mismatch', feeling that these terms are better attempt to describe what is happening in the auditory system without implying a particular locus of pathology **(Rapin et al.,2003).** To encompass these different opinions, the term 'Auditory Neuropathy/Dys-synchrony (AN/AD)' came into use, and was used in previous versions of the NHSP guidelines. At the **International Guidelines Development Conference at Como, Italy, in 2008,** a consensus reached to adopt the term 'Auditory Neuropathy Spectrum Disorder' (ANSD). This term includes both true auditory neuropathy (i.e. a true neural abnormality) and other possible underlying mechanisms resulting in neural dys-synchrony, as well as delayed maturation of the lower level auditory pathway. The term ANSD was also considered helpful as it expresses the wide range of presentations, prognosis and underlying etiologies associated with the disorder.

1.3.1 Underlying Physiology:

This suggests relatively normal activity in the outer hair cells, but disruption of transmission at some point from the inner hair cells along the neural pathway to the brainstem. In some cases, the underlying reason for this initial pattern of test results will become evident, whereas in others the underlying reason may not be found. In some cases, neural firing may be occurring but with a lack of synchrony, so that no clear ABR is recordable. In some cases, dys-synchrony may also arise due to delayed maturation or myelination of the auditory pathway. ANSD may affect neural processing of auditory stimuli, which may reduce a child's ability to understand

speech and may affect ability to detect sound to various degrees. These patients often complain they can hear sounds but cannot understand speech.

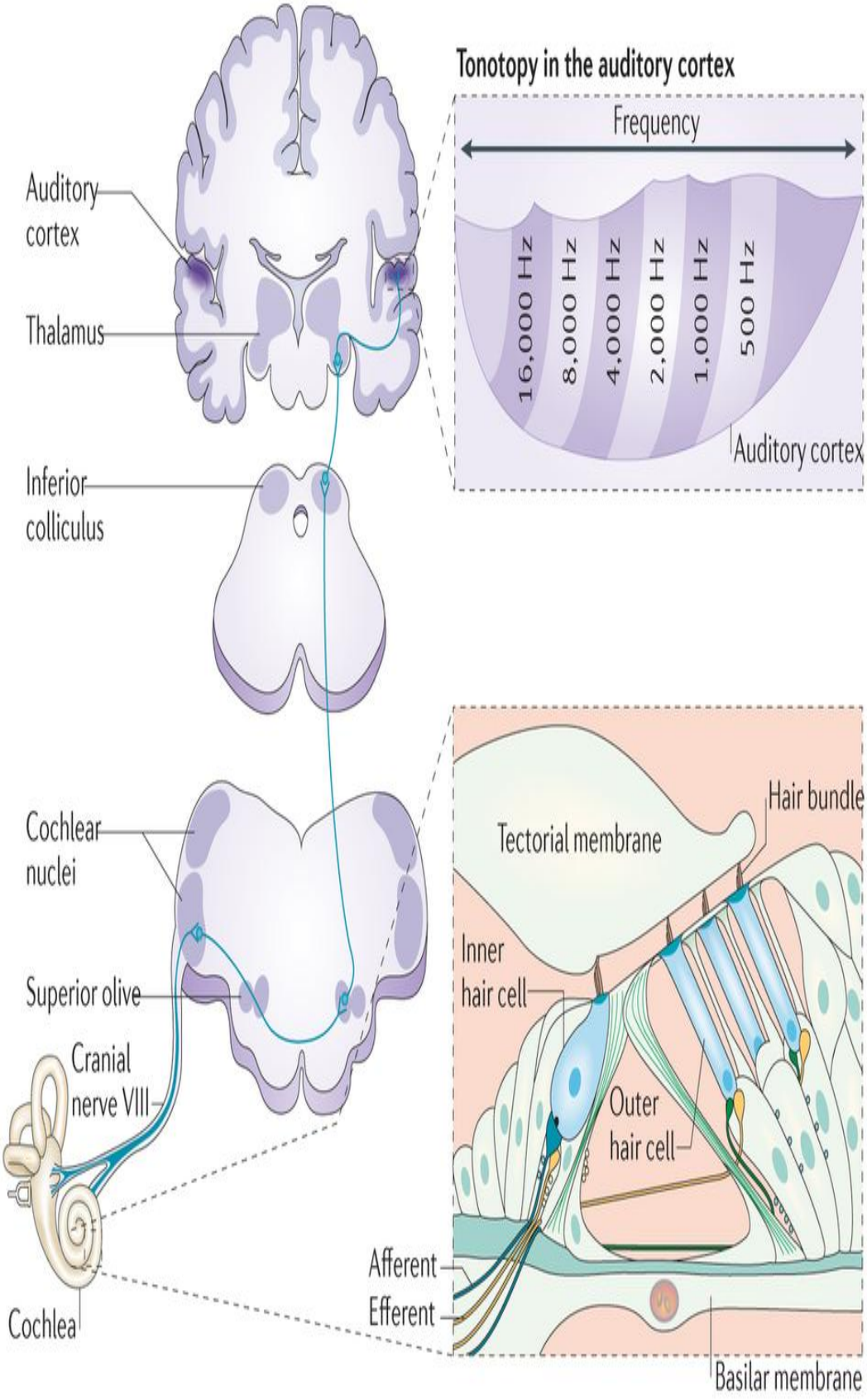

Fig 1.1: Showing Anatomical Structure of Inner Ear & Auditory Pathway

1.4 Prevalence Rate:

The prevalencerate of ANSD is presently unknown with estimates varying from 0.5-15%. Several researchers have given varies prevalence rate as per their studies and number of subjects.**Sininger et. al(2002)** estimates that ANSD occurs in about 1 in 10 children with permanent hearing loss, though prevalence estimates vary between studies. The true prevalence of ANSD in the paediatric population with hearing loss has not been determined in large, prospective multi-centre investigations and is therefore uncertain. Initial prevalence figures from the **English NHSP** are in line with the **Sininger etal (2002)** estimate. These children, because of absent ABR, might at first sight be thought to have severe/profound sensorineural (cochlear) hearing loss until tests of cochlear function are carried out.

Although the majority of ANSD cases occur among special care / neonatal intensive care babies, some studies have indicated that a significant number may occur in the well-baby population **(Sininger et. al, 2002) .**Many newborn hearing screening programs, including the **NHSP** protocol, currently only screen for evidence of ANSD in infants admitted to NICU, and do not offer ABR screening to all well babies. Cases of ANSD occurring in the well-baby population may therefore remain undetected. Cases of ANSD may be referred at a later stage and will need to be investigated, identified and managed following diagnosis.

1.5 Related Etiology:

The aetiology of ANSD seems to be multifactorial **(Varga et. al, 2003).**ANSD may arise from a diverse range of aetiologies. Infants with ANSD therefore require assessment, investigation and monitoring of neurodevelopmental progress by a physician with appropriate skills and an understanding of the condition. Diagnosis of

the underlying aetiology may determine the most appropriate further management, including specific intervention if indicated.

1.5.1Risk factors for ANSD from the neonatal history include:

1) Extreme prematurity <28 weeks gestation **(Sininger et. al, 2002).**
2) Low birth weight / intrauterine growth restriction **(Berg et. al, 2005).**
3) Hyperbilirubinemia reaching exchange transfusion levels **(Madden et. al, 2002).**
4) Hypoxic ischemic encephalopathy / intraventricular hemorrhage (as is likely to occur in infants with prolonged assisted ventilation / severe sepsis; (**Rance et. al, 1999).**
5) Genetic conditions that may give rise to this pattern of test results include, among others:
    - Otoferlin mutations (DFNB9 – autosomal recessive; **Varga et. al, 2003).**
    - Pejvakin mutations (DFNB59 – autosomal recessive; **Delmaghani et. al, 2006)**
    - Familial delayed auditory maturation **(Aldosari et. al, 2004)**
    - Neurodegenerative conditioned Charcot Marie Tooth, Friedreich's Ataxia **(Starr et. al, 1996).**
    - Metabolic conditioned e.g. Maple syrup urine disease**. (Spankovich et. al, 2007).**
    - Mitochondrial disorders **(Corley et. al, 1999).**

1.5.2 Associated Anatomical Anomalies

Some anatomical anomalies may also give rise to ANSD such as

- Hydrocephalies **(Starr, et.al, 1996, Berg et. al, 2005).**
- Brainstem anomalies **(Huang et. al, 2010).**

- Auditory nerve hypoplasia or aplasia **(Huang et. al, 2010).**

1.5.3 Other Anatomical Brain Anomalies:

- microcephaly,
- Space-occupying lesions such as cerebellar tumors etc.

Auditory Neuropathy Spectrum Disorder is one of the disorders, which affect cochlea and subsequent areas resulting in sensori neural hearing loss. Comparatively, its effects are even more worsening in younger children than adults. According to pathophysiology of auditory neuropathy spectrum disorder, it defuncts the sensory system, which in turn dys-synchronises the neural firing of sensory cells. This abrupt process in neural firing causes mild to severe sensori neural hearing loss.

1.6 Need for the Study:

Over the time, the number of cases with auditory neuropathy spectrum disorder has been changing depending upon its etiology and lack of facility and knowledge in areas of early identification and intervention. As a result, its prevalence rate has also been changing from time to time and from region to region depending upon the size of demography and availability of health care facilities. Auditory neuropathy spectrum disorder in recent times, demands a high- check procedure to identify its sign and symptoms as early as possible. Moreover, clinical presentation and audiological findings of pediatric auditory neuropathy shows it usually associated with various syndromes and other pathological sign and symptoms. Therefore, it demands a perennial approach of study to derive a reliable contemporary data on various aspects of this disorder viz. prevelance rate, auditory profile and emerging trend of associated problem if either. However, stringent initiatives should be taken in the direction of promoting Universal Hearing Screening Programme (UHSP) with

conscious participation of multi-disciplinary approach, that we can have a clear check on new cases with auditory neuropathy spectrum disorder.

Clinically it has become mendatory to rule out:

- Audiological findings of ANSD.
- Prevalence rate of ANSD.
- Associated problem with ANSD.
- Common risk factor or etiological factor, which can lead to cause ANSD among pediatric population.
- To compare children with ANSD to children having simple SNHL

1.7 Objectives of the study:

1. To determine prevalence rate of auditory neuropathy.
2. To determine the related etiologies.
3. To determine the audiological profile of children with auditory neuropathy.

1.8 Hypotheses: The list of hypothesis are as given below

- Prevalence of auditory neuropathy spectrum disorder:

  There will be no significant prevalence rate of auditory neuropathy spectrum disorder.

- Related etiologies of children with ANSD.

  There will be no much etiologies associated with auditory neuropathy.

- For comparison of audiological findings of children with ANSD:

  3.1. There will be no significant difference in between the audiological profile of children with ANSD and children with SNHL.

  3.2. There will be no significant sex difference in between the audiological profile of children with ANSD and children with SNHL.

3.3. There will be no significant difference in between the audiological profile of male children with ANSD and SNHL.

3.4. There will be no significant difference in between the audiological profile of female children with ANSD and SNHL.