
AUDITORY STEADY STATE RESPONSE IN CHILDREN WITH AUDITORY NEUROPATHY

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ABSTRACT:

Auditory neuropathy is a disorder characterized by preservation of outer hair cells function, with normal otoacoustic emission and/or cochlear microphonics, but an absent or distorted auditory brain stem response. this study aimed to study ASSR in children with auditory neuropathy and to compare these results with the outcome of study ASSR in children with normal hearing and with sensorineural hearing loss. thirty children with auditory neuropathy, 30 with normal hearing and 30 with sensorineural hearing loss all of them were examined. Audiological evaluation includes basic audiological tests, otoacoustic emission, auditory brain stem response and auditory steady state response. ASSR threshold was not well correlated with hearing threshold in normal hearing; however it is well correlated with sensorineural hearing loss. Only auditory neuropathy, ASSR percentage of response and ASSR threshold was more in high frequency than low frequency, the amplitude ASSR was more in sensorineural group than normal or auditory neuropathy children, however there is no significant difference between normal and auditory neuropathy.

KEY WORDS:

Auditory neuropathy

Auditory brain stem response

Auditory steady state response

Otoacoustic emission

INTRODUCTION:

Auditory Steady State Response (ASSR) is an evoked potential elicited in response to a periodically varying stimulus such as sinusoidal amplitude and/or frequency modulated tone (John et al., 2002). It is an objective recent method for detection of wide range of frequency specific hearing threshold. It gives hearing threshold within 5 dB from hearing threshold detected by auditory brainstem response (ABR). It detects the response by automatic detection algorithm and does not depend on visual detection of response as in the case of ABR (Tiffany et al., 2005). It is generally thought that there are multiple generators of ASSR as primary auditory

cortex, association area, auditory brain stem, cochlear nucleus, thalamus and inferior colliculus; depending on the modulation frequency. It must be emphasized that the presence of ASSR (at any modulation frequency) is determined in large part by the integrity of cochlea and eighth nerve (Ruggero, 1992).

One of the most challenging hearing disorders is auditory neuropathy (AN). AN is a hearing disorder that is characterized by absent ABR with normal outer hair cell function as indicated by intact otoacoustic emission and/or cochlear microphonic. Auditory neuropathy can affect any age from

neonates to old subjects; but its main targets are neonates, infant and young children (Starr et al., 2000). It has been shown that AN in infants and children are more common than it was initially thought. The prevalence of AN among infants and children with permanent hearing loss was found to be 11.01% (Sininger, 2002). Dys-synchrony or asynchrony within the auditory system is believed to be the underlying pathomechanism in AN (Starr et al., 1996). ABR, which is an onset response and depends on the synchronization within the auditory nerve and auditory brain stem, is essentially absent in cases of AN. Therefore, the actual threshold in infant and young children with AN can not be estimated using ABR. On the other hand, ASSR does not depend on the neuronal synchrony as much as ABR, Therefore; it might have a role in estimating hearing sensitivity in the young population with AN. Published studies about ASSR in AN are scarce.

This research is designed to study ASSR in children with AN and to compare ASSR results in children with auditory neuropathy to those in children with normal hearing sensitivity and sensorineural hearing loss (SNHL). Another aim of the current study is to correlate findings of ASSR in children with AN with other audiological findings such as otoacoustic emission, cochlear microphonics, speech discrimination and behavioral audiometry. Information gathered from the study results may help in the search for the pathology underlying auditory neuropathy and its site of lesion and may help in management of such challenging hearing disorder.

METHODOLOGY:

Material:

[A] **Patients:** A total number of 90 children participated in this study and divided into three equal groups. Group (I) included children with normal hearing sensitivity, group (II) included children with sensorineural hearing loss of different degrees, and group (III) included children with auditory neuropathy (as shown by intact otoacoustic emission and/or cochlear microphonic with absent ABR). No specific audiometric configuration was selected in this study. All subjects were selected from the outpatient clinic of Audiology unit, El-Minia university hospitals.

[B] **Equipment:** Two-channel audiometer, model amplified 309 calibrated according to ANSI (1969), Acoustic Immittance meter Madsen, model Zodiac 901 and Auditory evoked potential Intelligent Hearing System (IHS) with smart evoked potential software.

Method:

All participants in this study were subjected to the following:- Full history taking including prenatal, perinatal and postnatal history. Otological examination., Estimation of hearing sensitivity through one or more of The following procedure according to the age of child, Free field audiometry.,

Conditioned play audiometry and ABR:- using the smart intelligent hearing system (HIS) system. The stimuli were 100 μ s alternating click delivered through headphones at intensity level of 90 dB nHL. Repetition rate of the stimuli was 31.1p/s. Electrode

montage was high forehead to ipsilateral mastoid. The common electrode was placed on the contralateral mastoid. The response was filtered between 100 and 3000 Hz, amplified 10,000 times, recorded over 10.24 ms time window, and 2000 sweeps were averaged for each run. ABR testing for each child was carried under chloral hydrate sedation, Pure tone audiometry. If applicable, speech reception threshold (SRT) using Arabic Spondee Word (Soliman, 1975) and word discrimination score using Arabic Phonetically balanced words (Soliman et al., 1985). Immitancemetry consisting of tympanometry at varying pressure from +200 to -400 mm H₂O₂ and acoustic reflex threshold testing using pure tone of 500, 1000, 2000 and 4000 Hz. Distortion product otoacoustic emission.

Cochlear microphonic: it was recorded through surface electrodes using the same as ABR testing protocol except that the stimuli were delivered through insert ear phone in order to temporally separate the electromagnetic artifacts from the cochlear potentials. Separate recording was done for the three clicks polarities, namely, alternating, rarefaction, and condensation. The CM was considered present if the response reversed its polarity with change of the stimulus polarity and disappeared with alternating polarity and by clamping the tube of insert ear phone. Auditory steady state response (ASSR) measurement.

Electrode montage: testing was done using three electrodes, the active electrode was placed on high forehead, the reference electrode was placed on the mastoid process of examined ear, and the ground electrode was placed on the other mastoid.

RESULTS:

A- Descriptive data:-

- The present study comprised 90 children; 53 males and 37 females. The age range was between 7months and 14 years. The study group was further subdivided into 3 equal subgroups; group (A) had children with normal hearing; group (B) had children with sensorineural hearing loss and group (C) had children with auditory neuropathy.

- **Group (A)** consisted of 30 children, age ranged between 11months and 14 years with a mean value of 4.8 years and SD of 2.76. Twenty children were diagnosed with play-audiometry and the rest were diagnosed with ABR.

- **Group (B)** consisted of 30 children, 2 children had mild degree; 4 children had moderate degree; 15 children with moderately severe degree most of them had flat configuration, the remaining 9 children with severe to profound degree. Age of this group ranged between one year and 12 years with mean value of 6.4 years and SD of 2.89 years. Twenty two children were diagnosed with play-audiometry and the rest were diagnosed with ABR.

- **Group (C)** consisted of 30 children, age of this group ranged between 7 months and 17 years with mean value of 3.74 years and SD of 4.2 years. The diagnosis was based on absent ABR and intact OAE and/or CM. twenty one cases had pass OAE and 9 cases had partial pass OAE. CM was intact in all cases.

Table (1) shows the mean and SD of age in addition to the gender distribution in the three study groups. Table (2) and figure (14) show the mean and SD of

pure tone threshold in children with normal hearing and children with SNHL.

Possible etiology in sensorineural hearing loss was heredofamilial in 80%,

post-traumatic in 10% and idiopathic in 10%.

Possible etiology of auditory neuropathy was heredofamilial in 74%, neonatal jaundice in 23% and post-febrile in 3%.

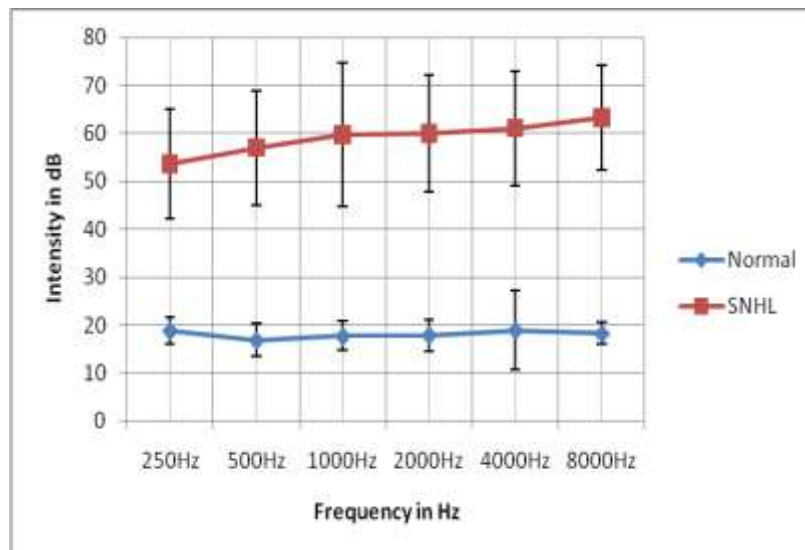


Figure (14): The mean and SD of pure tone threshold of group (1) and group (2)

B- ASSR in different study groups.

In children with normal hearing sensitivity (30 children; 60 ears) ASSR was present in 100% of the ears at the 4 different frequencies (500, 1000, 2000 and 4000Hz).

In children with sensorineural hearing loss, ASSR was present in 100% of ears in children who had pure tone audiometric threshold (21 children). This was valid at the 4 different frequencies. Nine children were diagnosed as having severe to profound SNHL based on the absence of ABR at maximum intensity used. 3 ears of them had intact ASSR at 500Hz and 1000Hz (16.6%), 2 ears

(11.1%) had intact ASSR at 2000Hz and no response at 4 KHz.

In children with AN, although ABR was absent at all ears (60 ears), ASSR was present in all cases, 12 ears at 500Hz (20%), 31 ears at 1000Hz (51.7%), 38 ears at 2000Hz (63.3%) and 40 ears at 4000Hz (66.7%). Table (3) & figure (15) described such results. Results showed that as the carrier frequency of the stimulus increased, the number of ears with intact ASSR increased. Such frequency effect was statistically significant as shown from the Chi-square test (table 4).

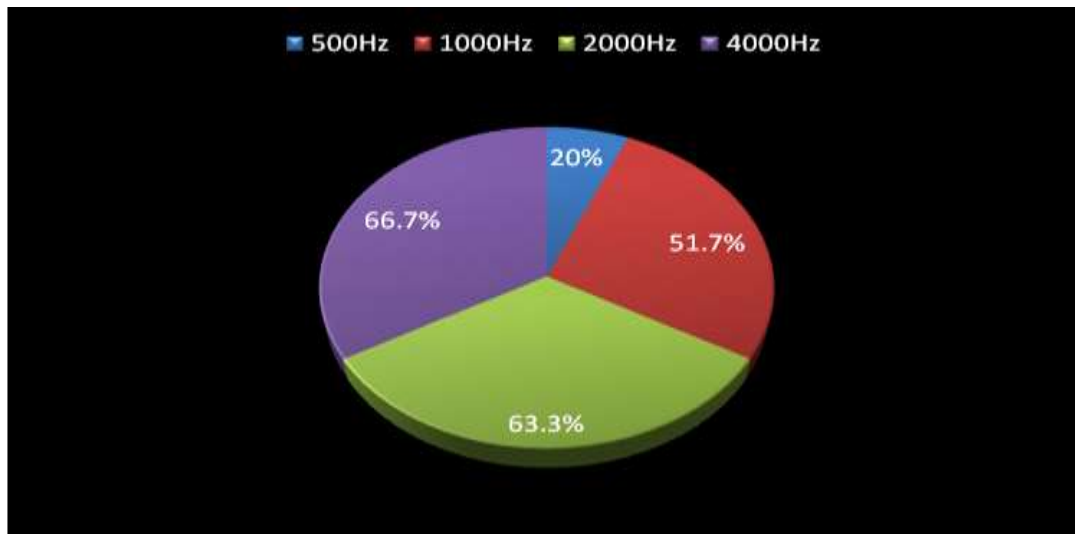


Figure (15): Percentage of ears with intact ASSR at different frequencies in children with AN (total number of the ears tested was 60 ears)

C- ASSR threshold.

Group (A): Table (6) and figure (16) show mean and SD of PTA threshold and ASSR threshold in children with normal hearing sensitivity. The smallest mean difference between PT threshold and ASSR was at 4 KHz (1.5 dBHL) and the largest was at 500Hz (5.2 dBHL). Although the mean difference was small, the SD was larger than the mean at each frequency and the range was so large (between 25 dBHL at 4 KHz and 60 dBHL at 0.5 KHz). Such results reflect

marked variability in ASSR threshold in children with normal hearing sensitivity.

Consistent with this finding, no significant correlation between PTA threshold and ASSR threshold in children with normal hearing sensitivity was found except at 4KHz. Table (5) describes these results. In addition, no statistically significant difference was found in ASSR threshold among frequencies as shown from paired sample T-test table (7).

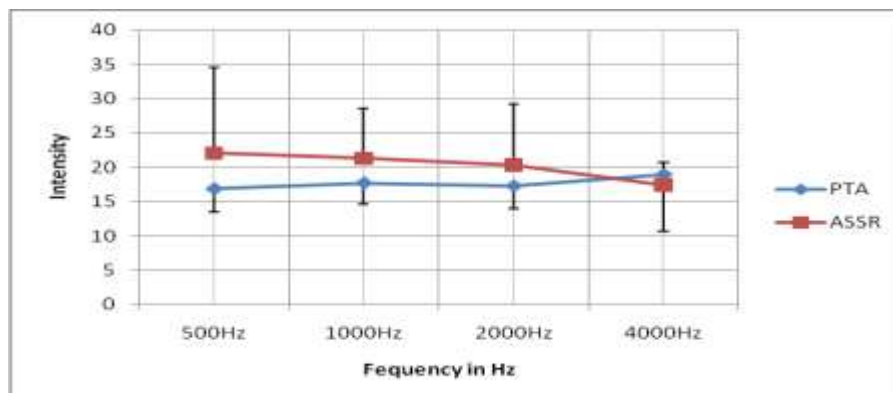


Figure (16): Mean and SD of PTA threshold and ASSR threshold in children with normal hearing sensitivity.

PTA threshold in dBHL, ASSR threshold in dBnHL.

- **Group (B):** Table (8) and figure (17) show the mean and SD of PT threshold and ASSR threshold in children with sensorineural hearing loss. Mean, SD, minimum, maximum and range of the

difference between both thresholds are also shown in table (8). Different than children with normal hearing sensitivity, there was statistically significant correlation between pure tone threshold and ASSR threshold in children with sensorineural hearing loss at all frequencies (table 9).

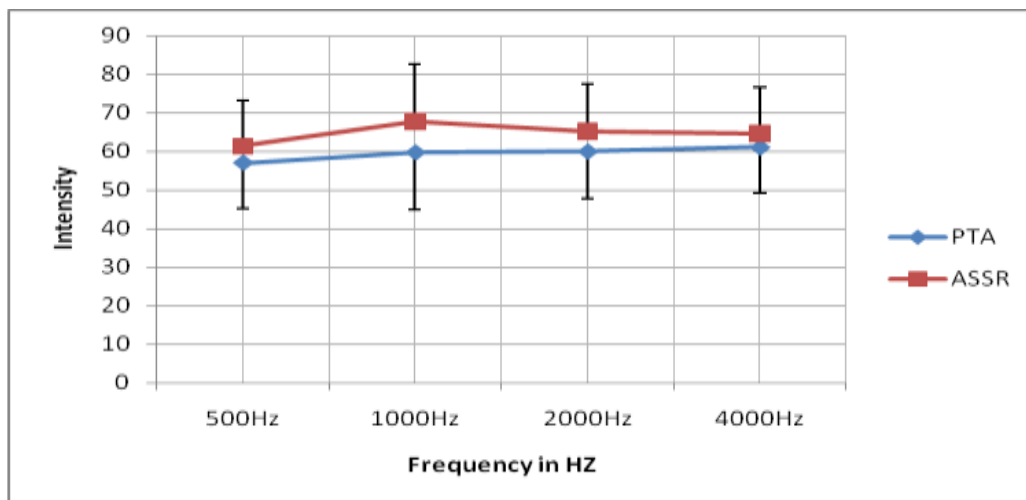


Figure (17): Mean and SD of PTA threshold and ASSR threshold in children with SNHL.

Group (C):- ASSR threshold in children with AN

- Table (10) & figure (18) show the mean, SD, minimum, maximum and the range of ASSR threshold in children with AN. Except for threshold at 500&1000Hz, ASSR Threshold was better at higher stimulus frequencies.

Paired sample t-test (table11) revealed statistically significant better ASSR threshold at 4 KHz than that at 500, 1000 and 2000Hz. No statistically significant difference in ASSR threshold was found among thresholds at 500, 1000 and 2000Hz.

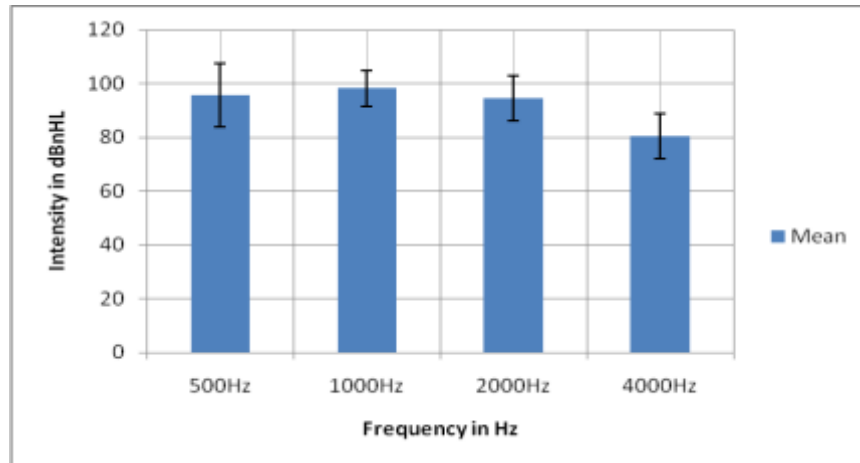


Figure (18): ASSR threshold in children with AN.

D- The description of ASSR amplitude in normal, SNHL and AN.

- Table (12) & figure (19) shows mean and SD of ASSR amplitude at the 4 test frequencies in the different study group. ANOVA test table (13) revealed statistically significant difference in the ASSR amplitude among the study groups in each of the test frequencies. - Interestingly the ASSR amplitude in SNHL group was larger than that of normal group and of AN group. Tukey

post-Hoc test (table 14) revealed that difference between the amplitude in SNHL group and each of normal and AN group was statistically significant. This was valid in the 4 test frequencies. Although the ASSR amplitude in the normal group was larger than that of AN group, the difference was not statistically significant as show from tuket test in table (14). Such results were the same at the 4 test frequencies.

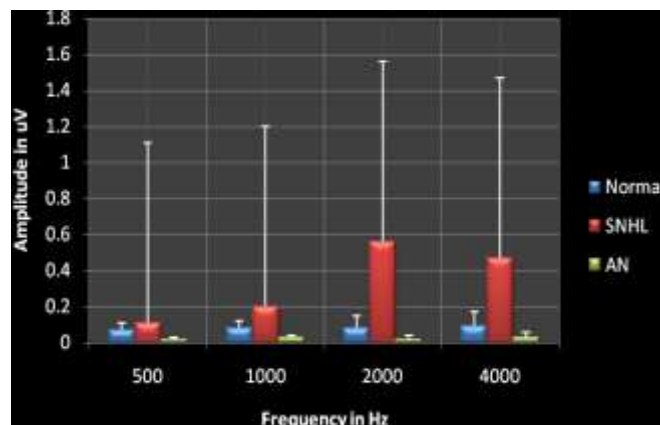


Figure (19): amplitude of ASSR in normal, SNHL and AN.

E- Relation between the amplitude of ASSR and degree of hearing loss.

- Figure (20) shows the amplitude of ASSR in different degrees of SNHL. Children with moderately severe degree have the largest ASSR amplitude. However, statistical comparison between the different degrees of SNHL

could not be done due to the small sample size in the categories of mild and severe to profound degrees. Overall no statistically significant correlation was found between the degree of SNHL and the amplitude of ASSR at different frequencies .

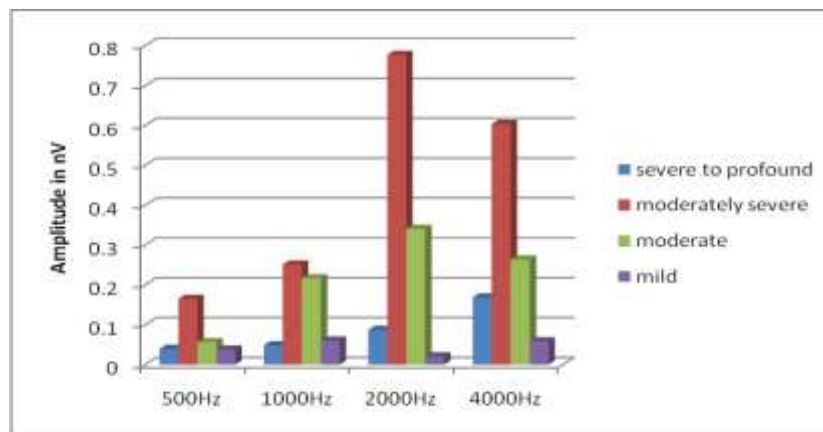


Figure (20): Amplitude of ASSR in different degrees of SNHL.

F- ASSR amplitude in different frequencies in the different groups.

- Paired sample t-test to compare ASSR amplitude among different frequencies (table 16 & 17) revealed such frequency effect was significant between 500 & 1000Hz and between 2000 & 4000Hz in children with normal hearing. In children with SNHL frequency effect was significant among the 4 different frequencies except between 2000 & 4000Hz.

figure (18) show the mean and SD of ASSR threshold at different frequencies in children with AN. Although there was statistically

significant effect of frequency on ASSR threshold, no statistically significant effect of frequency was found on ASSR amplitude as shown from paired sample t-test (table 18).

G- the relation between the amplitude of ASSR response and the age of the patient.

- Correlation between the children age and ASSR amplitude in different study group is shown in table (19), statistically significant correlation between children age and ASSR was found only in the AN group. The older children had the larger ASSR amplitude.