# Assessment of outcome of cochlear implant in auditory neuropathy spectrum disorder using speech in noise test

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ORIGINAL STUDY

# Assessment of outcome of cochlear implant in auditory neuropathy spectrum disorder using speech in noise test

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**Abstract**

*Introduction*: **Children with auditory neuropathy spectrum disorder (ANSD) generally struggle with auditory and language treatment, and cochlear implant (CI) outcomes can be unpredictable environmental noise. The speech discrimination ability is the main complaint of those patients especially in the background.**

*Objective*: **Assessment of the ability of young children diagnosed with ANSD who underwent CI in one ear to discriminate speech by using 'speech in noise test' with 0 Signal to noise ratio (S/N) and comparing them with matched children with severe to profound sensori-neural hearing loss (SNHL).**

*Methods*: **The study was conducted on 27 children who received 'unilateral' CI of different companies divided into two groups: the control group 15 children with severe to profound sensori-neural hearing loss and ANSD group 12 children.**

**All children subjected to following evaluation 2 years postregular auditory language re habilitation: aided cortical auditory evoked potentials (CAEP) in which latency of P1 was analyzed, speech discrimination score (SD%), Speech perception in noise (SPIN) score test with 0 S/N ratio, categories of auditory performance (CAP), speech intelligibility rate (SIR).**

*Results*: **A total of 75 % of ANSD group showed response in aided CAEP and SD% and SPIN test with no significant difference between groups as regards to age, Latency of P1, CAP, and SIR, the mean ± SD of the SD% score in the control group was 89.9 ± 2.6 while it was 64 ± 4.9 in ANSD group the SPIN score with mean ± SD was 85.6± and 51.7 ± 7.4 in controls and ANSD groups, respectively, a significant negative correlation between P1 Latency associated with SPIN% in ANSD group was recorded. As regard CAP and SIR there was no significant difference between the study and the control group following 2 years of regular rehabilitation; indicating the beneficial effect of CI and 'auditory language rehabilitation' in children with ANSD.**

*Conclusion*: **CI in 'ANSD' children had a positive impact on language development, and the outcomes were on par with those children without 'ANSD'. Our study also highlighted that the SPIN score test is very important in determining how well CI works, especially for individual with 'ANSD'.**

*Recommendation*: **Long-term evaluation of children who had CI is necessary to determine the progressive and ongoing benefit of the treatment of those children with ANSD.**

***Keywords:*** Auditory neuropathy spectrum disorder, Auditory performance, Cochlear implant, Speech intelligibility rate, Speech perception in noise

## 1. Introduction

Auditory neuropathy spectrum disorder (ANSD) is a disorder in which pure tone audiometry results do not correspond with other measures of auditory function, such as ABR data and' speech discrimination scores'. One of the characteristics of 'ANSD' is the existence of 'oto-acoustic emissions or cochlear micro phonics (CM) which does not match with ABR threshold or pure tone audiometric threshold' [1,2].

According to the pathogenesis, ANSD is divided into presynaptic and postsynaptic forms.

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Presynaptic ANSD is characterized by dysfunction or loss of the cochlear inner hair cells, which reduces the amplitude or eliminates the receptor summating potential and reduces the auditory nerve activity. In the postsynaptic mechanisms multiple locations along the auditory nerve, such as the unmyelinated dendrites in the cochlea, myelinated dendrites and axons coursing centrally, and myelinated auditory ganglion cells, lead to auditory nerve dysfunction, a decrease in ganglion cells and nerve fibers may also be present [3].

Children with ANSD often face difficulties with auditory and language rehabilitation, and cochlear implants (CIs) results can vary [4].

The speech discrimination ability especially in noise is the main challenge for patients suffering from ANSD. CI for those patients has a variable outcome reported by previous literature findings, some of them found it effectively improves the ability of language development in children consequentially improving discrimination of speech [5], others, however, noticed little or no change. According to certain research, ANSD patients who had CI responded similarly to those who wore hearing aids [6,7].

Cortical auditory evoked potentials (CAEP), which represent the neuroelectric activity of primary and secondary auditory cortexes, offer important details on the processing of auditory information [8] have been used for a variety of purposes such as: an objective measure for estimating hearing thresholds for patients for whom behavioral hearing threshold is difficult to obtain, assessing cortical maturation, and verifying the effectiveness of hearing aids or CIs in pediatrics [6].

The waves of CAEP are P1, N1, and P2. When determining cortical maturity, the P1 component of CAEP is a helpful indicator. Due to constant auditory input, the P1 waves latency reduces with age starting at birth and peaks at 60 ms in middle-aged adults. The clinical significance of CAEP is it does not need child behavioral cooperation [9].

The objective of this study is to assess the auditory and language abilities of young children diagnosed with ANSD who underwent to unilateral CI to discriminate speech by using ‘speech in noise test’ with 0 ‘S/N ratio’ and comparing them with matched children with severe to profound sensorineural hearing loss (SNHL).

## 2. Procedures

The current investigation was carried out on patients who received a CI at the hearing and speech institution in Egypt as a case−control observational study. Participants' parents gave written, fully informed consent. The General Organization of Teaching Hospitals and Institutes' (GOTHI's) Institutional Review Board granted its consent. The patients were picked from a list of cochlear implant recipients that was kept at our facility.

### 2.1. Inclusion criteria

(1) Children Pre-lingual with severe to profound SNHL and ANSD.
(2) Age range at implantation from 3 to 5 years old.
(3) Satisfactory aided response by free field pure tone audiometry.
(4) IQ assessment greater than or equal to 90.
(5) Regularity in auditory and language rehabilitation sessions.

### 2.2. Exclusion criteria

(1) Patients who have imaging evidence of significant ‘inner ear malformations’.
(2) Post lingual severe to profound SNHL or ANSD.
(3) Children with IQ less than 90.
(4) Irregularity in therapy sessions.

The study was conducted on 27 children who received CI in one ear of different companies divided into two groups:

(1) Group (1) the control group composed of 15 children with severe to profound SNHL.
(2) Group (2) composed of 12 children with ANSD cases.

Both groups received CI and all of them received (auditory and language rehabilitation) therapy for 2 years duration (2 sessions/week each session 45 minutes) followed by instructions for parents to train their children through daily routines.

Both groups were subjected to: History taking of onset and duration of hearing loss and date of implantation and type of the processor.

Then all children underwent verification of the outcome of CI to ensure that there was satisfactory outcome by using: aided pure tone free field in sound-treated room (for the frequency range 500−4000 Hz), both study groups underwent to speech audiometry test by performing speech recognition threshold (SRT) using live Arabic bisyllabic phonetically balanced list for pediatrics then speech discrimination score using live ‘Arabic phonetically balanced monosyllabic list’ of pediatric (SD%) score using ‘Two Channel Audiometer’

(Inter-acoustics, model AC40) Denmark. The SPIN score test and aided CAEP also performed.

### 2.3. Speech perception in noise (SPIN) score test

In order to quantify speech discrimination scores, the SPIN test was utilized using Arabic (phonetically balanced) monosyllabic words with fixed 0 S/N ratio. Speech stimuli were placed at 40 dB above SRT. The speakers were used to transmit 'live-monitored speech stimuli' and 'speech noise'.

### 2.4. Aided cortical auditory evoked potentials test

The procedure was carried out with standardized tools in a sound-treated room using Evoked potential system (GSI Eclipse). Electrodes positioned over the head were used to record the electrical reactions. The test stimulus was (20 ms rise/fall plateau tones) at 2 khz, transduced by Loudspeaker at 45° azimuth, the electrode montage was single channel with non-inverting electrode (+ve) was on vertex, the inverting electrode on one mastoid and ground electrode on the other mastoid bone, with EEG filter High pass was 0.1 Hz and Low pass at 30 Hz. The repetition rate was one stimulus every 2 seconds. Number of repeats at least two. 65 dBnHL was the presentation level. After the second year of habilitation, (Aided CAEPs with CI) were documented for both groups. Latency and amplitude of P1 are among the other CAEP metrics that were recorded. For the analysis, the latency of the wave P1, was taken.

### 2.5. Phoniatrics assessment by applying

#### 2.5.1. Categories of auditory performance (CAP)

CAP are used to assess a child auditory skill following CI [10] the hierarchical scale of auditory perceptual ability which range from 0 to 7 are explained in the (Table 1).

A child with 'CI's speech intelligibility is evaluated using (SIR) by spontaneous speaking in natural settings [11]. SIR is broken down into 5 categories in (Table 2).

*Table 1. 'Categories of auditory performance score' (CAP) [10]*

| | |
|---|---|
| 0 | Not hearing sounds |
| 1 | Responding to background noises |
| 2 | 'React to speech sounds' |
| 3 | 'Recognises' environmental sounds |
| 4 | 'Discriminates' speech utterances |
| 5 | Without lip reading 'comprehend words' |
| 6 | Without lip reading 'Understands conversation' |
| 7 | Can use a telephone |

*Table 2. 'Speech intelligibility rate' (SIR)*

| | |
|---|---|
| Category 1 | 'Pre-recognizable words' in spoken language |
| Category 2 | Ambiguous uttered speech (unintelligible) |
| Category 3 | Connected speech is intelligible when the listener is concentrated |
| Category 4 | A listener who is unfamiliar with deaf speech can understand connected |
| Category 5 | The child is 'easily understood' in typical circumstances |

Baseline The scores for CAP and SIR acquired right away after implantation and those obtained two years later after therapy were compared. After two years of recovery, the CAP and SIR of the ANSD group were compared with the control group.

### 2.6. Statistical analysis

Presentation and analysis of statistics Using SPSS, a statistical software package for the social sciences, version 23, 2016 for Windows (SPSS Inc., USA).

The study of the statistical findings employed the mean and standard deviation. Chi-square analysis was performed to compare group differences, and T test and categorical results were given as percentages. All data were presented as mean standard deviation (SD).

## 3. Results

Our research was conducted on 27 children who underwent to CI. The children were divided into 2 groups. group (1) include 15 child patients with severe to profound SNHL with their mean age 3.8 ± 0.71 years, and compared with 12 children diagnosed by ANSD group (2) their mean age was 3.6 ± 0.8 the both groups are matched in age with *P*-value shows no statistically significant difference (Table 3).

Results of aided CEAP and SD% and SPIN score obtained from all children in control group while it could be obtained only from 9 (75 %) children (75 %) while 3 children (25 %) of ANSD group did not develop language and the no waves in aided CEAP can be recorded.

*Table 3. Age, sex, latency of P1, SRT %, and speech perception in noise score in controls and cases*

| Variable | Control $n = 15$ | Cases $n = 12$ | *P*-value |
|---|---|---|---|
| Age (y) mean ± SD | 3.8 ± 0.7 | 3.6.1 ± 0.8 | 0.638 |
| Sex no (%) | | | |
| Female | 6 (40 %) | 6 (50 %) | 0.5 |
| Male | 9 (60 %) | 6 (50 %) | |
| P1 latency means | 55.7 ± 2.02 | 42.1 ± 4.6 | 0.095 |
| SRT% mean ± SD | 89.9 ± 2.6 | 64 ± 4.9 | 0.042 |
| SPIN score mean ± SD | 85.6 ± 2.2 | 51.7 ± 7.4 | 0.003 |

*P*-value (<0.05 Significant- ≥0.05 non-significant).

The P1 latency in control group was 55.7 ± 2.02, and 42.1 ± 4.6 in group of cases with no statistically significant difference.

Sex, Age, and *P*-value less than 0.05 demographic data do not significantly differ between groups. SRT % and SPIN% for the cases are significantly different from the control, although P1 Latency is not significantly different between the cases and control.

Before initiating therapy and after two years of rehabilitation, the CAP and SIR scores were compared using the Wilcoxon signed-rank test.

## 4. Discussion

For children with severe to profound SNHL cochlear implantation is the sole option to restore hearing and promote for language development. Studies on the beneficial effect of children with ANSD related hearing and language development is still lacking.

The inner ear can detect sound in people with ANSD, but there is a problem transmitting the signal to the brain. Additionally, they have trouble interpreting language [12].

The objective of our study is to compare young children with ANSD and matched children with severe to profound SNHL who received unilateral CI to gauge their auditory and language skills.

Regarding the age of implantation, between the two groups, there was no statistically significant difference, additionally, both groups had language development evaluations after 2 years of implantation and rehabilitation therapy.

Comparing both groups regarding to SD% showed the mean ± SD of the SD% score in control group was 89.9 ± 2.6 while it was 64 ± 4.9 in ANSD group indicating benefit of CI which is also ensured by findings of the SPIN score with mean ± SD was 85.6±, and 51.7 ± 7.4in controls and ANSD groups, respectively, Sarankumar et al. [13] who evaluated the results of CI in a group of 10 children with ANSD who underwent CI also reported significant benefits in kids with ANSD who underwent CI as they recorded that SPIN scores in children with ANSD were 63 % at 0 dB SNR.

Fontenot et al. [14] performed open set speech perception test in case-control study between ANSD and non ANSD implanted children and they found there was no significant difference between the two groups, also Breneman et al. [15] stated that on the Multisyllabic Lexical Neighborhood (MLNT)-Easy and MLNT-Hard criteria, the ANSD and 'SNHL' groups achieved speech recognition scores of 80.5 % and 78.3 %, respectively. The groups' respective mean scores on the Lexical Neighborhood (LNT)-Hard and/or consonant-nucleus-consonant (CNC) criteria were 83.2 % and 77.0 %. There was little to distinguish them from one another.

The presence of statistically significant difference between the two groups in SD%, and SPIN score and low scores in our results may be attributed to that only 75 % of our cases had response to cochlear implantation.

According to Teagle et al. [16] children with ANSD who receive CIs are a diverse group with a wide range and deficits. Despite the fact that many of these children eventually benefit from CI, some of them will not, most likely as there is not electrical stimulation that causes neural synchronization.

Budenx and colleagues [17] recorded that children with ANSD who also have concurrent cognitive or developmental problems perform less better than those with ANSD alone, according to CI's study on 17 ANSD patients.

Aided CAEP was also performed to be an objective measure to evaluate the outcome of CI, comparison between both groups done by using the latency of wave P1 showed that the mean latency of PI in ANSD group was 42.1 ± 4.6, and 55.7 ± 2.02 in control group and there was no statistically significant difference (Table 1), and significant negative correlation between latency of P1 and SPIN score in ANSD group was found (Fig. 1 and Table 4) this was in agreement with Sarankumar et al. [13], as they also found significant negative correlation and they recorded in their study that the mid-frequency speech stimulus/g/was used in the aided CAEP, and the mean latency of the P1 wave at 12 months after habilitation was 60.1 ms.

According to Alvarenga et al. [18] latency of P1component was also found to be linked with the duration of hearing loss and to be a predictor of speech perception performance in children with CI. We would be able to assess the value of CI in ANSD with greater clarify if we performed a thorough examination of the P1 wave of the CAEP for speech stimuli covering all frequencies and correlating it with SPIN word and sentence tests. According to Dorman et al. [19] and Sharma [20] children's speech perception and language abilities increase along with a general decrease in the latency of P1.

There was a statistically significant difference in the data of CAP and SIR immediately after CI and 2 years of 'auditory and language therapy' (*P*-value for CAP = 0.002, SIR = 0.007). When comparing the two groups control and ANSD There was no statistically significant difference in the data of CAP and SIR two years of auditory and language rehabilitation (*P*-value for CAP = 0.150, SIR = 0.317). Alzahrani et al. [21] and Huang et al. [22] found no observable

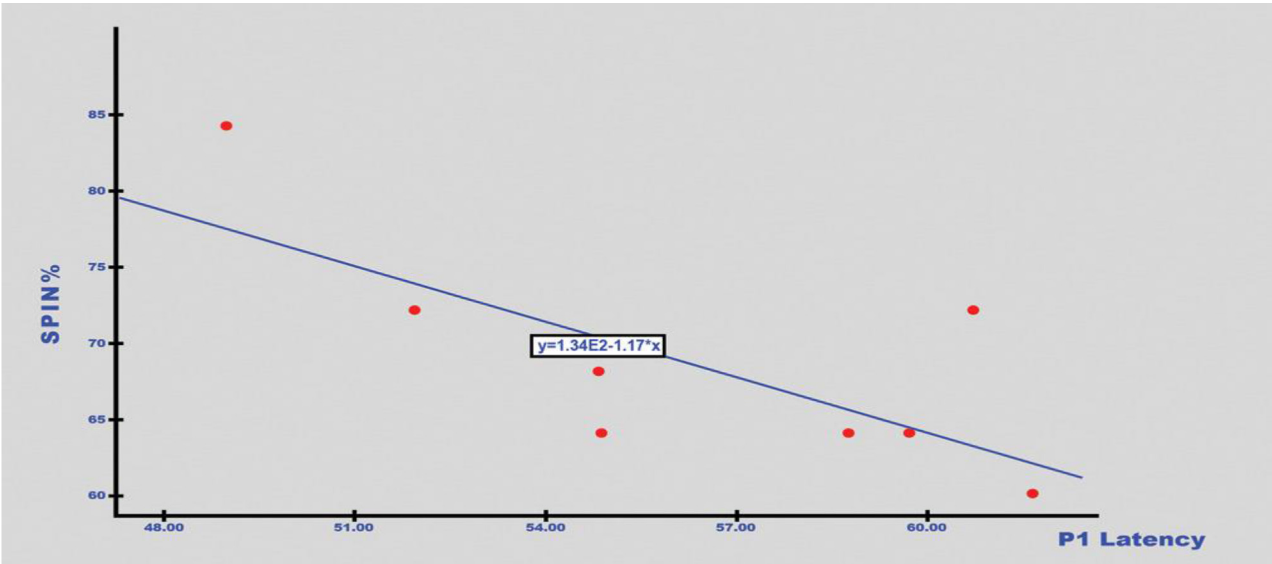


*Fig. 1. Correlation between Speech perception in noise% and P1 latency in cases.*

*Table 4. Correlation between Speech perception in noise score and latency of P1 in ANSD group (cases)*

| Variable | R | Sig |
|---|---|---|
| P1-lat SPIN% | −0.749 | 0.02 |

r (Correlation Coefficient).
It showed that there was a significant negative correlation between P1 Lat associated with Speech perception in noise% in ANSD group.
*Correlation is significant at the 0.05 level (2-tailed).

differences between patients with and without ANSD. According to these findings, children with auditory neuropathy who had early CI and language therapy experienced the same benefits as the control group. These findings suggest that children with ANSD should be encouraged to use early listening skills together with 'CI' and spoken language to help them communicate regularly with others.

### *4.1. Conclusion*

Despite having a mixed outcome, CI in 'ANSD' children had a positive impact on language development, and the outcomes were on par with those children without 'ANSD'. Our study also highlighted that the SPIN score test is very important in determining how well CI works, especially for individual with 'ANSD'.

### *4.2. Recommendation*

Long-term evaluation of children who had CI is necessary to determine the progressive and ongoing benefit of the treatment of those children with ANSD.

## Conflicts of interest

No conflict of interest.

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