Audiological Performance in Children with Inner Ear Malformations Before and After Cochlear Implantation

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Abstract

Objective: To evaluate the auditory perception outcomes of cochlear implant (CI) in children with different types of inner ear malformations (IEMs) and to compare them with CI users with the normal cochlea. Design: Retrospective and prospective data collection. Settings: Tertiary referral hospital. Participants: There were 274 CI users with and without IEMs as two groups. Both groups' chronological age at implantation and duration of cochlear implant usage was matched (±8 months). Main outcome measures: All subjects were evaluated preoperatively and postoperatively with Ling's sound test and auditory perception test battery, which includes the Meaningful Auditory Integration Scale (MAIS), close-set Pattern Perception Test (PPT), and open-set Sentence Recognition Test (SRT). Also, children with IEMs were assessed for language development. Results: The incidence of IEMs were incomplete partition-II, 40 (29.19%), incomplete partition-I, 36 (26.2%), cochlear hypoplasia, 26 (18.9%), enlarged vestibular aqueduct, 14 (%10.2), incomplete partition-III, 10 (%7.2), common cavity, 8 (5.8%) and dilatation of vestibule, 3 (2.1%) patients. The significant difference was seen in Ling's sound test and auditory perception test battery scores of children with incomplete partition-I, cochlear hypoplasia, and common cavity (p-value $<$ .005). Conclusion: IEMs group showed different progress according to the type of ear anomaly. Although CI users with enlarged vestibular aqueduct (EVA) had the highest scores, users with common cavity had the lowest scores. Taking these results, caused by anatomical differences, in to account is very critical in follow-ups and rehabilitation programs. Each cochlear implant user should be evaluated according to his/her individual needs.

Keypoints

- Radiological results play a crucial role in detecting different types of IEMs.
- It is critical to evaluate the functional hearing and auditory perception skills of children with different types of IEMs in the first year after surgery.
- Depending on our clinical experience, children with IP-I, common cavity, and cochlear hypoplasia are developing slowly, and especially this group should be followed intensively in terms of auditory perception and language skills.
- Children with IEMs who did not develop as a result of follow-up, instead of the "wait and see" strategy, should be concluded quickly.

1 INTRODUCTION

Cochlear implantation (CI) has become a life-transforming device for children with severe and profound hearing loss (HL). However, in the beginning, CI was a contraindication for inner ear malformations (IEMs), which are, constitute approximately 20%–35% congenital sensorineural hearing loss cases.[1-4] Preoperative audiologic and radiologic evaluations are very important for cochlear implant candidates. With advanced imaging techniques such as CT and MRI, cochlear structures could be appropriately evaluated. With these advanced techniques, cochleovestibular malformations were classified, and it was reported that the majority of children with IEMs anomalies have bilateral severe to profound HL and are candidates for either CI or Auditory Brainstem Implant (ABI).[5-7] It was known that CI is an effective treatment modality for individuals with IEMs. Sennaroglu and Bajin[5] were classified IEMs eight groups: Complete Labyrinthine Aplasia (Michel Deformity), Rudimentary Otocyst, Cochlear Aplasia, Common Cavity, Cochlear Hypoplasia, Incomplete Partition of the Cochlea I-II-III, Enlarged Vestibular Aqueduct, Cochlear Aperture Abnormalities. The incidence of IEMs was reported at 11.5% in our clinic.[3]

Cochlear implant outcomes depend on different parameters such as onset and duration of HL, the age at implantation, presence of neurodevelopmental disorders, the anatomy of the inner ear and physiology of the auditory nerve, family support, socioeconomic level, access to rehabilitation services, and regular CI programming. Important information about cochleovestibular anomalies has allowed in the preoperative period with advanced technology in imaging methods.[6,7] Because of comorbidity of IEMs with different types, especially children, could be suffering from auditory perception. Many studies have compared the speech perception and language skills of children with and without IEMs. A study by Farhood et al,[8] showed the benefits of CI in the speech perception of patients with IEMs. Eisenman et al,[9]reported that the progression was slower in children with IEMs for auditory perception.

In the current study, the principal objective was to compare the preoperative and postoperative auditory performance of children with and without IEMs. The specific objective of this study was to elaborate auditory perception outcomes of IEMs types.

2 MATERIALS AND METHODS

2.1 Ethical considerations

[Blinded for review]

2.2 Participants

One hundred and thirty-seven children with various IEMs [Common Cavity, Enlarged Vestibular Aqueduct (EVA), Incomplete Partition of the CochleaI-II-III(IP-I, IP-II, IP-III), Cochlear Hypoplasia, Dilatation of Vestibule] were comprised the patient group. The normal cochlea group consisted of 137 children. Two hundred and seventy-four children with CI with or without IEMs were included in the study. Also, all children use unilateral cochlear implants.

All participants matched chronological age, age of implantation, and duration of CI use (±8 months). All of them use their CI regularly in daily life. The inner ear abnormalities' demographic distribution was summarized in Table 1. The preoperative data presented in this study were collected betweenJuly 2013 and March 2019.

2.3 Cochlear implant device programming

Approximately two-three weeks after the implantation, the initialactivation was done by a pediatric audiologist. The audiologists verify the children's hearing thresholds with the CI at each programming session using audiometric sound field-testing. Sound detection thresholds are measured at 0.25, 0.5, 1, 2, and 4 kHz using warble-tone or narrow-band stimuli, with speech presented by a monitored live voice. The children's thresholds currently range from 30 to 40 dB HL, with speech awareness thresholds at about 25-35 dB HL.

2.4 Assessment procedures

All participants were evaluated pre- and post CI with Ling's sound test, the Meaningful Auditory Integration Scale (MAIS), and Children's Auditory Perception Skills Test (CIAT) subtests.[10]

MAIS is a survey reported by parents' inventory and is used to evaluate functional listening skills in children with HL. It consists of ten items grouped into three main areas: implant use, awareness of sound, and sound recognition. Each item was rated by parents and scored from 0 to 4 (0 = never, 1 = rarely, 2 = occasionally, 3 = frequently, and 4 = always), with a total score ranging between zero and 40. All testing was initiated at 1-6 months post-CI activation, and at 1-3 yearly follow up intervals.

Functional auditory perception skills in daily life were used to evaluate children's auditory perception skills. Besides, comprehensive test battery CIAT was applied that includes close-set Pattern Perception Test (PPT) and open-set Daily Turkish Sentence Recognition Test (SRT). In PPT, 12 words with different numbers of syllables (one, two, or three syllables and compound words) were randomly presented, twice for each word, in auditory-only conditions, asking the child to point to or name the appropriate picture matching the word. If the child scored 18/24 or above, the researchers proceeded to the open-set Daily Turkish SRT. This test contained ten sentences and attempted to assess children's ability to recognize and comprehend speech. The children were told they would hear a series of simple questions, statements, and commands, which they might have heard in their daily lives.

Language skill was assessed using the Test of Early Language Development-Third Edition (TELD-3). This test thoroughly evaluates the children's language development in the fields of semantics, syntax, and morphology.[11]

2.5 Statistical analysis

Statistical analyses were performed using the SPSS software (version 22, SPSS Inc. Chicago, USA).All data showed normal distribution.ANOVA test was used to compare MAIS scores, pattern perception scores, sentence recognition scores, and language performance scores between the groups. Continuous variables (i.e., chronological age, CI age, and duration of CI use) were reported as means, standard deviations, and ranges by post-hoc t-test. The differences among mean with p -value $< .005$ accepted as representing statistically significant differences.

3 RESULTS

3.1. Demographics

There were 274 (137 study group and 137 control group) CI users in the study. IP-II constituted the most common subjects who were 40 of 137 anomalies (29.19%) in this study. Other malformations included IP-I (n: 36, 26.2%), cochlear hypoplasia (n: 26, 18.9%), EVA (n: 14, %10.2), IP-III (n: 10, %7.2), common cavity (n: 8, 5.8%) and dilatation of vestibule (n: 3, 2.1%). The results of children showed varying data depending on the types of IEMs, and auditory performances were ranked to order from high to low.

3.2The auditory perception outcomes

An improvement was observed in comparing Ling's sound test scores with the first evaluation period post-operative scores (Table 2). There was a significant difference between preoperative and postoperative in all children with IP-I ($F_{1\text{-}71} = 16.056$, p -value $< .005$), cochlear hypoplasia ($F_{1\text{-}51} = 18.769$, p -value $< .005$) and common cavity($F_{1\text{-}15} = 76.164$, p -value $< .005$). There were no preoperative and postoperative statistical differences compared to children with other IEMs.

The comparison of the preoperative MAIS scores and the one-six months and one-three years' postoperative scores showed that auditory integration improved significantly (Table 3), and all children with IEMs benefitted from CI (Figure 1). Also, there were remarkable differences between the MAIS scores of the different malformation groups in three years after surgery. A comparison of the MAIS scores of children with common cavity showed statistical difference at all periods ($F_{1\text{-}15} =10.186$, p -value $< .005$), but their rate of development was so slow than children with IEMs and normal cochlea.Also, children with (IP-I $F_{1\text{-}71} = 16.056$, p -value $< .005$) and cochlear hypoplasia ($F_{1\text{-}51}= 18.769$, p -value $< .005$) showed a significant difference between preoperative and postoperative in MAIS scores.

Table 4 and Figure 2 presented results of closed-set PPT and open-set SRT. All children with normal cochlea completed the closed-set and open-set tests in second evaluation period with relative success. However, children with IP-I, cochlear hypoplasia, common cavityand dilatation of vestibulehad not yet reached the open set SRT test within one-three years.Children with different IEMs also achieved very low scores, except EVA ($F_{1\text{-}27}$ = .955, p -value = .337; $F_{1\text{-}27}$ = 1.050, p -value = .315), IP-II ($F_{1\text{-}79}$ = .071, p -value = .791; $F_{1\text{-}79}$ = .062, p -value = .804) and IP-III ($F_{1\text{-}19}$ = 1.991, p -value = .175; $F_{1\text{-}19}$ = .188, p -value = .670)patients. A comparison of the PPT and SRT scores of children with IP-I ($F_{1\text{-}71}$ = 25.262, p -value < .005; $F_{1\text{-}71}$ = 47.216, p -value < .005), cochlear hypoplasia ($F_{1\text{-}51}$ = 33.196, p -value < .005; $F_{1\text{-}51}$ = 31.908, p -value < .005)common cavity($F_{1\text{-}15}$=33.286, p -value < .005; $F_{1\text{-}15}$=30.621, p -value < .005) and dilatation of vestibule ($F_{1\text{-}5}$ = 11.172, p -value < .005; $F_{1\text{-}5}$ = 16.000, p -value < .005)showed statistical difference.

Enlarged vestibular aqueduct obtained the first group of children with the highest scores in terms of auditory perception performance among IEMs. The children with EVA achieved open set SRT approximately 80%. Unfortunately, children with common cavity constituted the lowest score group for auditory perception results. In the Ling's Sound Test and MAIS evaluation performed before the CI, in common cavity had the lowest scores compared within the normal cochlea, and statistically significant differences were found (p -value < .005). In the MAIS test, between second evaluation periods after CI, in common cavity had a score of 25.38 points while in normal cochlea had a score of 40 full points. Although they could only complete the closed-set PPT, they were unable to open-set SRT.

3.3 The language outcomes

TELD-3assesses receptive and expressive language in children between the ages of 24 months to 95 months. TELD-3 was applied only to groups with all IEMs.

Children with EVA completed all the items of the receptive and expressive language tests. The mean age of the receptive language was 83 months, and the expressive language age was determined as 83 months.Children with IP-II had receptive language age of 83 months and expressive language age of 78 months. All IP-II children had completed the receptive language test items of the TELD-3. Children with dilatation of vestibule the receptive language age were average 71.66 months, while the expressive language age was average 58 months. While the receptive language age of the IP-III children were 68.9 months, the expressive language ages were 54 months. Children with IP-I, receptive language age was average 67.61 months, while the expressive language age was average 50 months. The mean ages of the receptive language of children with cochlear hypoplasia were 58.34 months and expressive language age of 41.53 months. Children with common cavity the receptive language age were average 56.25 months, while the expressive language age was average 42.5 months.

4 DISCUSSION

Despite CI success in children with IEMs, the results were highly variable. In this study, we aimed to determine the success of auditory perception skills in these children over the years. The study had one of the largest series of 137 using CI children with IEMs and reported auditory perception outcomes. CT and MRI have played a critical role in the diagnosis of children with ear abnormalities; thus, surgical planning has been facilitated. However, after the surgery, it was crucial to show up the functional hearing levels of all using CI children and to know how long they have achieved their development. The auditory perception outcomes of different types of IEMs have been compared in studies.

Children with IEMs were found to develop identification and comprehension toward environmental sounds and speech sounds after an average one year. The results indicated a delay in auditory perception performance in children with inner ear anomaly; however, their performance changes with time. It is necessary to follow more closely and to study in this direction in terms of the functionality of auditory perception performances. Although the auditory performances of children with IEMs were better in the closed-set test, their vocabulary was weak, and it was observed that they had struggled in attention and memory skills during the evaluation. Open-set tests involve thinking and predicting words in sentences using clues in the context to maintain the conversation; therefore, the use of language-based visual clues would be complicated skill to develop. [10]

The ten children who common cavity, incomplete partition, EVA, and membranous anomalies, in Luntz et al,[12] the study demonstrated speech awareness at 25 dB HL or better. They reported that children with

IEMs had more than 30 months of CI experience, and 75% of those achieved some degree of open-set sentence recognition. Dowell et al,[13] showed that children implanted before the age of four years had mean scores of 68% for open-set phonemes after three to five years of experience with the cochlear implant. The present study results showed that after one-three years of CI experience, children with IEMs have succeeded in the closed-set test, but not in the open-set SRT test.

Overall IEMs, the EVA group had a higher score than normal cochlea in the closed-set test. The EVA was a congenital anomaly with progressive sensorineural hearing loss. In the beginning, their hearing may be normal, but the process is progressive. It was thought children with EVA might have been suffering from HL in the peri- or post-lingual period rather than the prelingual period. EVA was better than IP-II because of the better than the modiolar defect, which is the reason to make the outcome slightly worse. EVA, IP-II, IP-III had comparable results with normal cochlea; this is because a good-sized cochlear nerve always accompanies them. ABI surgery is never indicated in EVA, IP-II, and IP-III. Some studies in the literature have shown that children with EVA exhibited high performance in auditory perception testing.[14-16]

As higher-level language skills had increased, it had been determined that children with IEMs (especially common cavity and cochlear hypoplasia) have difficulty with comprehension skills. Results indicate that the duration of CI use is significant than the chronological age for both receptive and expressive language development. As the child's chronological age increases, the gap between language skills scores wider. In later years, it will be challenging to learn comprehensive language skills and catch these tasks.[17]Tucci et al,[18] reported the same finding in their study with five children and one adult. All patients except one demonstrated limited language skills.In another study, researchers presented speech perception and production after follow-up for 36 months in children with common cavity scores for the CAP, SIR, MAIS, and closed-set/open-set auditory speech perception tests were lower than in the control group. Both of these studies reported lower results than the present study.[19]

Parents should be informed before surgery about the possible effects of ear anomalies on auditory performance and language skills. However, a comprehensive preoperative radiological examination, successful surgery, and an individually tailored postoperative rehabilitation program should be included.

4.1 Strength and limitation

This is the largest study so far documenting to evaluate and compare the auditory perception performance of children with IEM, and normal cochlear anatomy is one of the strengths.Secondly, all subjects with different ear anomaly that provided maximum and minimum benefit from CI were considered and ranked.Thirdly, the present study adds to the growing body of research that indicates auditory perception performance of children with various IEMs to determine the expectation after cochlear implantation. This study was limited by the absence of language evaluation in the control group. In future studies, both groups can be compared in terms of language performance.

5 CONCLUSION

Extensive sample data showed that children with IEMs might receive considerable benefit from CI. Children of IEMs detected speech sounds at various frequencies. After one year of monitorized, most of them developed pattern perception skills in a close-set mode. A transdisciplinary approach is suggested in pre-and post-implantation evaluation and follow-up. Auditory perception and language skills expectations should be different from those for CI with the normal cochlea. At least three years after CI, children with all IEMs showed varying degrees of auditory benefit. As a result, the early age of implantation is an essential parameter for CI users in IEMs. Audiological evaluation and intensive rehabilitation program are necessary.

CONFLICT OF INTEREST

The authors declare that they do not have funding, financial relationships, or conflicts of interest to disclose.

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Appendix A

CI	Cochlear Implantation
HL	Hearing Loss
IEMs	Inner Ear Malformations
EVA	Enlarged Vestibular Aquaduct
IP I-II-III	Incomplete Partition of the Cochlea I-II-III
CIAT	Children's Auditory Perception Skills Test in Turkish

CI	Cochlear Implantation
PPT	Pattern Perception Test
SRT	Sentence Recognition Test
MAIS	Meaningful Auditory Integration Scale
TELD-3	Test of Early Language Development – Third Edition

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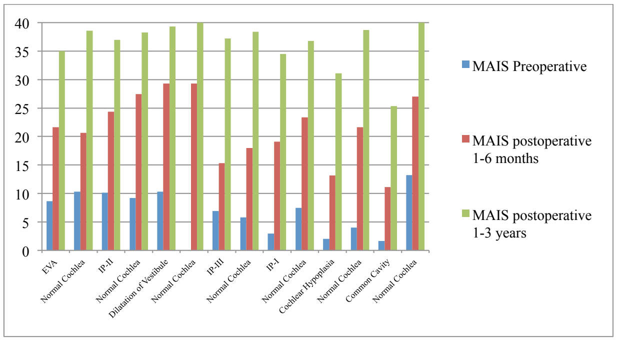
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EVA
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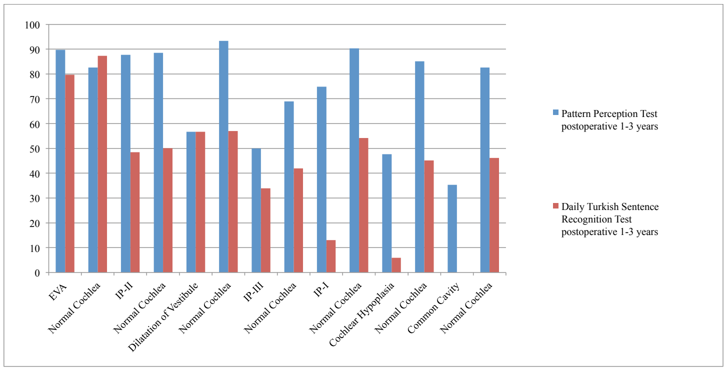
Dilatation of Vestibule
Normal Cochlea
IP-III
Normal Cochlea
IP-I
Normal Cochlea
Cochlear Hypoplasia
Normal Cochlea
Common Cavity
Normal Cochlea
MAIS Preoperative
MAIS postoperative 1-6 months
MAIS postoperative 1-3 years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Pattern Perception Test postoperative 1-3 years
Daily Turkish Sentence Recognition Test postoperative 1-3 years