# The Outcome of Cochlear Implantations in Deaf-Blind Patients: A Multicenter Observational Study

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**Objective:** This multicenter study aimed to evaluate the auditory and speech outcomes of cochlear implantation (CI) in deaf-blind patients compared with deaf-only patients.
**Study Design:** Retrospective cohort study.
**Setting:** Multiple cochlear implant centers.
**Patients:** The current study was conducted on 17 prelingual deaf-blind children and 12 postlingual deaf-blind adults who underwent CI surgery. As a control group, 17 prelingual deaf children and 12 postlingual deaf adults were selected.
**Intervention:** Cochlear implantation.
**Main Outcome Measure(s):** Auditory and linguistic performances in children were assessed using the categories of auditory performance (CAP) and Speech Intelligibility Rating (SIR) scales, respectively. The word recognition score (WRS) was also used to measure speech perception ability in adults. The mean CAP, SIR, and WRS cores were compared between the deaf-only and deaf-blind groups before CI surgery and at “12 months” and “24 months” after device activation. Cohen's d was used for effect size estimation.
**Results:** We found no significant differences in the mean CAP and SIR scores between the deaf-blind and deaf-only children before the CI surgery. For both groups, SIR and CAP scores improved with increasing time after the device activation. The mean CAP scores in the deaf-only children were either equivalent or slightly higher than those of the deaf-blind children at “12 months post-CI” (3.94 ± 0.74 vs 3.24 ± 1.25; mean difference score, 0.706) and “24 months post-CI” (6.01 ± 0.79 vs 5.47 ± 1.06; mean difference score, 0.529) time intervals, but these differences were not statistically significant. The SIR scores in deaf-only implanted children were, on average, 0.870 scores greater than the deaf-blind children at “12 months post-CI” (2.94 ± 0.55 vs 2.07 ± 1.4; $p = 0.01$, d = 0.97) and, on average, 1.067 scores greater than deaf-blind children at “24 months post-CI” (4.35 ± 0.49 vs 3.29 ± 1.20; $p = 0.002$; d = 1.15) time intervals. We also found an improvement in WRS scores from the “pre-implantation” to the “12-month post-CI” and “24-month post-CI” time intervals in both groups. Pairwise comparisons indicated that the mean WRS in the deaf-only adults was, on average, 10.61% better than deaf-blind implanted adults at “12 months post-CI” (62.33 ± 9.09% vs 51.71 ± 10.73%, $p = 0.034$, d = 1.06) and, on average, 15.81% better than deaf-blind adults at “24-months post-CI” (72.67 ± 8.66% vs 56.8 ± 9.78%, $p = 0.002$, d = 1.61) follow-ups.
**Conclusion:** Cochlear implantation is a beneficial method for the rehabilitation of deaf-blind patients. Both deaf-blind and deaf-only implanted children revealed similar auditory performances. However, speech perception ability in deaf-blind patients was slightly lower than the deaf-only patients in both children and adults.
**Key Words:** Auditory performance—Blind—Cochlear implant—Deaf—Speech intelligibility.

*Otol Neurotol* **43:**908–914, 2022.

## INTRODUCTION

Cochlear implantation (CI) has become a well-established surgical method for restoring the audition of patients suffering from severe to profound sensorineural hearing loss (SNHL) when conventional hearing aids have minimal or no beneficiary

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The authors have no relevant financial conflicts or other conflicts of interest to disclose.

**DOI: 10.1097/MAO.0000000000003611**

outcome (1–3). These patients can use the increased access to acoustic stimuli provided by CI devices to promote speech production, speech perception, and language reading abilities over time (4–6). The development of lexical skills, as well as expressive and receptive language skills in cochlear implanted individuals, permits verbal communication as the main route of communication; moreover, these skills may develop better with earlier implantation (7–9).

It has been shown that visible facial articulators (e.g., jaw or lips) will help humans to transmit important aspects of their speech, such as information regarding the place of articulation (e.g., nonbilabial vs. bilabial sounds) and roundedness of vowels and consonants (10). Certainly, adding visual cues to auditory information will assist normal-hearing subjects to perceive speech sounds more precisely, particularly when there is a competing signal in the environment and acoustic quality is poor (10,11). In such cases, visual cues provided by visible articulatory organs operate as functional cues that supplement the auditory information transmitted by the speech signal.

Dual-sensory impairment (DSI) or the presence of both SNHL and vision loss is a complicated condition and its prevalence increases with age. Patients with DSI have limited choices for communication and rely primarily on tactile stimulation (12,13). It has been shown that individuals with DSIs have poorer social and physical functioning and higher rates of psychological symptoms than patients with a single-sensory impairment (i.e., auditory or vision loss alone) (14,15). CI can substantially benefit the rehabilitation of deaf-blind subjects, and such patients may receive the greatest relative improvement from the hearing provided. Several studies have demonstrated that early implantation in deaf-blind patients has a positive effect on their communication abilities and improves their quality of life (QoL) (16–20).

Usher syndrome is the most common genetic cause for the dual handicap of blindness and deafness. Hartel et al. (18) reported the beneficial effect of CI in 16 postlingual adults (mean age, 59 years at CI surgery) with Usher syndrome. Their findings indicated that cochlear implantation increases speech perception and improves the QoL in patients with Usher syndrome.

According to the perceptual compensation hypothesis, sensory deprivation within one sensory system will stimulate compensatory perceptual alternations in another sensory system. In the case of early visual deprivation, there is some debate over whether blindness leads to an auditory system dysfunction or an enhancement in auditory performance (10). It seems that early-blind patients may show superior frequency discrimination and sound localization skills (21,22).

Although several studies have indicated higher speech perception abilities in blind individuals, much less is known regarding the influence of visual impairment on auditory and speech performances in CI recipients. The current multicenter study was designed to determine the outcomes of CI in deaf-blind individuals compared with their deaf-only peers in both children and adult populations during a 2-year follow-up period.

## METHODS

### Study Population

This retrospective cohort study was conducted on 17 children and 12 adults with blindness and severe to profound hearing impairment identified as CI candidates. All patients were recruited from the seven cochlear implant centers of public hospitals (five centers in Iran and two centers in the United States) between 2004 and 2017.

Inclusion criteria of CI for both children and adults were as follows: 1) bilateral prelingual severe to profound SNHL, 2) complete insertion of CI electrodes into the cochlea, and 3) regular attendance at preoperative and postoperative aural rehabilitation sessions. Children/adults with cochlear malformations or auditory nerve agenesis were excluded from the study. Furthermore, individuals with a history of ear infection, meningitis, neurologic problems, and cognitive or developmental disorders were excluded.

All deaf-blind children had congenital, complete visual loss, classified as Classes 5, 4, or 3 in the International Classification of Diseases of the World Health Organization (see Table 1). As the control group, CI candidates had perfect (10/10) visual acuity (deaf-only group) (see Table 2).

**TABLE 1.** *Demographic and clinical characteristics of deaf-blind children*

| Case No. | Sex | Implanted Ear | Age at First CI, yr | Duration of Sev.-Prof. SNHL, yr | Duration of Blindness, yr | Cause of Deafness | Cause of Visual Impairment |
|---|---|---|---|---|---|---|---|
| 1 | Female | Right | 10 | 6 | 10 | Usher syndrome | Retinitis pigmentosa |
| 2 | Female | Right | 4 | 4 | 4 | Usher syndrome | Retinitis pigmentosa |
| 3 | Female | Right | 5 | 2 | 5 | Unknown | Congenital |
| 4 | Male | Left | 2 | 2 | 2 | Unknown | Optic atrophy |
| 5 | Male | Right | 5s | 3 | 5 | Unknown | Anterior segment dysgenesis |
| 6 | Male | Right | 4 | 3 | 4 | Unknown | Choroideremia |
| 7 | Male | Right | 7 | 4 | 7 | Usher syndrome | Retinitis pigmentosa |
| 8 | Male | Left | 4 | 4 | 4 | Usher syndrome | Retinitis pigmentosa |
| 9 | Male | Left | 2 | 2 | 2 | Usher syndrome | Retinitis pigmentosa |
| 10 | Male | Right | 3 | 3 | 3 | Usher syndrome | Retinitis pigmentosa |
| 11 | Male | Right | 10 | 7 | 10 | Usher syndrome | Retinitis pigmentosa |
| 12 | Male | Left | 4 | 2 | 4 | Usher syndrome | Retinitis pigmentosa |
| 13 | Male | Right | 2 | 2 | 2 | Usher syndrome | Retinitis pigmentosa |
| 14 | Male | Left | 3 | 3 | 3 | Usher syndrome | Retinitis pigmentosa |
| 15 | Female | Bilateral | 5 | 3 | 5 | Usher syndrome | Retinitis pigmentosa |
| 16 | Female | Right | 2 | 2 | 2 | Vestibulocochlear dysplasia | Anterior segment dysgenesis |
| 17 | Male | Right | 5 | 3 | 5 | Usher syndrome | Choroideremia |

CI indicates cochlear implantation; Sev.-Prof., severe to profound; SNHL, ensorineural hearing loss.

**TABLE 2.** *Demographic and clinical characteristics of deaf-only children (control group)*

| Case No. | Sex | Implanted Ear | Age at First CI, yr | Duration of Sev.-Prof. SNHL, yr | Cause of Deafness |
|---|---|---|---|---|---|
| 1 | Female | Right | 2 | 2 | Congenital SNHL |
| 2 | Female | Left | 4 | 4 | Congenital SNHL |
| 3 | Male | Right | 5 | 5 | Congenital SNHL |
| 4 | Male | Left | 4 | 4 | Congenital SNHL |
| 5 | Female | Bilateral | 5 | 4 | Idiopathic |
| 6 | Male | Right | 5 | 3 | Congenital SNHL |
| 7 | Male | Right | 3 | 3 | Congenital SNHL |
| 8 | Female | Right | 5 | 4 | Congenital SNHL |
| 9 | Male | Right | 3 | 3 | Congenital SNHL |
| 10 | Female | Right | 4 | 4 | Congenital SNHL |
| 11 | Male | Right | 5 | 3 | Idiopathic |
| 12 | Male | Left | 3 | 3 | Congenital SNHL |
| 13 | Male | Right | 4 | 4 | Congenital SNHL |
| 14 | Male | Right | 4 | 2 | Idiopathic |
| 15 | Male | Right | 3 | 3 | Congenital SNHL |
| 16 | Female | Right | 2 | 2 | Congenital SNHL |
| 17 | Female | Right | 4 | 4 | Congenital SNHL |

CI indicates cochlear implantation; Sev.-Prof., severe to profound; SNHL, ensorineural hearing loss.

Our study population also consisted of 12 postlingual adults (seven male patients) with congenital blindness (deaf-blind group) who underwent CI surgery (see Table 3). Diagnosis of visual problems was conducted according to International Classification of Diseases criteria of the World Health Organization (Classes 5, 4, or 3). The control group consisted of 10 postlingual implanted adults (seven male patients) who perfect visual acuity (10/10) or vision corrected by lenses, resulting in near-perfect vision (deaf-only group) (see Table 4).

The study was approved by the local ethics committee at each participating institution and was in complete accordance with the ethical principles of human studies set by the Helsinki declaration (2014).

## Procedures

### *Categories of Auditory Performance*

The categories of auditory performance (CAP) scale was used to assess the auditory receptive abilities of the implanted children (23). The CAP comprises a hierarchical scale (see Table 5) based on the Likert scale ranging from 0 (“no awareness of environmental sound”) to 7 (“use of telephone with a known listener”). It has been shown that CAP scale has a satisfactory test-retest reliability (0.82), concurrent validity (0.64), and construct validity (0.58–0.74) in cochlear implanted children (24).

### *Speech Intelligibility Rating*

The Speech Intelligibility Rating (SIR) scale was used to measure the verbal intelligibility of the cochlear implanted children in real-life situations (25). The SIR is a five-point hierarchical scale ranging from “prerecognizable words in the spoken language” to “connected speech is intelligible to all listeners” (see Table 6). The SIR scale has an acceptable construct validity (0.66–0.69), concurrent validity (0.69), and test-retest reliability ($r$ = 0.99) in cochlear implanted children (24).

### *Word Recognition Score)*

Speech recognition in adults was evaluated via a standardized lists of open-set monosyllabic words in a free-field environment. The test was performed without lip-reading, at 65 dB SPL intensity in a quiet environment, and with a signal to noise ratio of 10 dB SPL noise below the signal intensity. The speech test was conducted in sound-proofed audiometric rooms with a calibrated loudspeaker at 0 azimuth, 100 cm from the patient.

**TABLE 3.** *Demographic and clinical characteristics of deaf-blind adults*

| Case No. | Sex | Implanted Ear | Age at First CI, yr | Duration of Sev.-Prof. SNHL, yr | Duration of Blindness, yr | Cause of Deafness | Cause of Visual Impairment |
|---|---|---|---|---|---|---|---|
| 2 | Female | Right | 48 | 6 | 6 | Progressive SNHL | Traumatic injury |
| 3 | Female | Right | 21 | 4 | 4 | Usher syndrome | Retinitis pigmentosa |
| 4 | Male | Left | 43 | n/a | n/a | Usher syndrome | Retinitis pigmentosa |
| 5 | Female | Right | 34 | 4 | 2 | Usher syndrome | Retinitis pigmentosa |
| 6 | Male | Right | 40 | 16 | yr | Idiopathic | Cataract |
| 7 | Male | Right | 39 | n/a | 3 | Norrie syndrome | Leukocoria, cataract |
| 8 | Female | Bilateral | 50 | n/a | yr | Idiopathic | Optic atrophy |
| 9 | Male | Right | 70 | 8 | 10 | Progressive SNHL | Macular degeneration |
| 1 | Male | Right | 20 | 5 | n/a | Usher syndrome | Retinitis pigmentosa |
| 10 | Male | Bilateral | 48 | 6.5 | yr | Cogan syndrome | Recurrent cornea inflammation |
| 11 | Female | Bilateral | 70 | 16 | n/a | Idiopathic | Glaucoma, traumatic injury |
| 12 | Male | Left | 35 | 5 | 6 | Progressive SNHL | Optic atrophy |

CI indicates cochlear implantation; n/a, not available; SNHL, ensorineural hearing loss.

**TABLE 4.** *Demographic and clinical characteristics of deaf-only adults (control group)*

| Case No. | Sex | Implanted Ear | Age at First CI, yr | Duration of Sev.-Prof. SNHL, yr | Cause of Deafness |
|---|---|---|---|---|---|
| 1 | Male | Right | 28 | 5 | Progressive SNHL |
| 2 | Male | Bilateral | 51 | 7 | Progressive SNHL |
| 3 | Male | Right | 33 | 2.5 | Idiopathic Sudden SNHL |
| 4 | Male | Left | 48 | 6 | Progressive SNHL |
| 5 | Female | Bilateral | 34 | 9 | Progressive SNHL |
| 6 | Male | Right | 32 | 2.5 | Idiopathic sudden SNHL |
| 7 | Male | Right | 45 | 3 | Progressive SNHL |
| 8 | Female | Bilateral | 42 | 4 | Idiopathic Sudden SNHL + NIHL |
| 9 | Male | Right | 65 | 5.5 | Progressive SNHL |
| 10 | Female | Right | 54 | 3 | Sudden SNHL |
| 11 | Female | Right | 68 | 8.5 | Progressive SNHL |
| 12 | Female | Left | 42 | 6 | Idiopathic Sudden SNHL + NIHL |

CI indicates cochlear implantation; NIHL, noise-induced hearing loss; SNHL, sensorineural hearing loss.

Patients with CIs had their devices activated approximately 3 weeks after the surgery, and their auditory and speech abilities were measured before surgery and at 12 and 24 months after CI activation.

### Data Analysis

The $\chi^2$ test was used to investigate associations between two categorical variables, such as sex and study group (deaf-blind vs. deaf-only). The Kolmogorov-Smirnov and Levene's tests were also performed to evaluate the normality of the distribution of numeric variables and equality of variances, respectively. The effects of cochlear implant on the auditory and speech performance in deaf-only and deaf-blind patients ("group" factor) across different timepoints ("test interval" factor) were assessed using a repeated-measures analysis of variance (ANOVA) test. Data were analyzed using SPSS version 24.0 (SPSS Inc., Chicago, IL). Statistical significance level was set as $p$ value less than 0.05.

## RESULTS

### Demographic and Clinical Characteristics

The demographic and clinical characteristics of deaf-blind (DSI) and deaf-only children are presented in Tables 1 and 2, respectively. In this study, 17 deaf-blind (12 male patients) and 17 deaf-only (10 male patients) children participated. No significant differences were noted in terms of sex ($\chi^2$ test; $p = 0.721$) and age of implantation (deaf-blind, 4.52 ± 2.47 years; deaf-only, 3.76 ± 1.1 years; independent sample $t$ test; $p = 0.14$) between both groups of children. A total of deaf-blind (five male patients) and 12 deaf-only (five male patients) adults participated in this study. According to our data, no significant differences were observed between the mean age of implantation in the deaf-blind and deaf-only adults (deaf-blind: 43.17 ± 15.78 years, deaf-only: 45.16 ± 12.77 years, Mann-Whitney $U$ test; $p = 0.554$).

### CAP Measurement in Children

Table 7 shows the mean CAP scores for both groups of children (deaf-blind and deaf-only) at preimplantation and during the follow-up period. The repeated-measures ANOVA showed the significant effect of "test interval" ($F$ (2,64) = 316.56; $p < 0.001$). However, the "group" ($F$ (1,32)=3.72; $p = 0.63$), and "group"-"test interval" interaction ($F$ (2,64) = 2.81; $p = 0.68$) effects were not statistically significant. We found no significant difference in CAP scores between the deaf-blind and deaf-only children before the CI surgery (deaf-blind, 0.88 ± 0.71; deaf-only, 0.71 ± 0.47; $p = 0.29$). Pairwise comparisons demonstrated that there was a significant improvement in mean CAP scores from "preimplantation" condition to "12-month post-CI" and "24-month post-CI" conditions in deaf-only children (preimplantation, 0.71 ± 0.47; 12-month post-CI, 3.94 ± 0.74; 24-month post-CI, 6.0 ± 0.79; $p < 0.001$). Pairwise comparisons also indicated that there was a significant improvement in mean CAP scores from "preimplantation" condition to "12-month post-CI" and "24-month post-CI" conditions in deaf-blind children (preimplantation, 0.88 ± 0.71; 12-month post-CI, 3.24 ± 1.25; 24-month post-CI, 5.47 ± 1.06; $p < 0.001$). Further analysis revealed that the mean CAP scores in the deaf-only children were either equivalent or slightly higher than those of the deaf-blind children during follow-up intervals, but these differences were not statistically significant ("12-month post-CI" phase: mean difference, 0.706; $p = 0.054$; and "24-month post-CI" phase: mean difference, 0.529; $p = 0.111$).

### SIR Measurement in Children

The mean SIR scores for both deaf-blind and deaf-only children at preimplantation and follow-up periods are presented in Table 7. Our findings revealed that speech intelligibility in both deaf-blind and deaf-only children improved with increasing time after cochlear implantation. The repeated-measure ANOVA showed a significant main effect of "group" ($F$ (1,29)=7.8; $p = 0.01$), "test interval"

**TABLE 5.** *The CAP criteria*

| Category | Criteria |
|---|---|
| 0 | No awareness of environmental sounds |
| 1 | Awareness of environmental sounds |
| 2 | Response to speech sounds |
| 3 | Identification of environmental sounds |
| 4 | Discrimination of some speech sounds without lip-reading |
| 5 | Understanding of common phrases without lip reading |
| 6 | Understanding of conversation with a familiar talker without lip reading |
| 7 | Use of telephone with a familiar talker |

CAP indicates categories of auditory performance.

**TABLE 6.** *The SIR criteria*

| Category | Criteria |
|---|---|
| 1 | Connected speech is unintelligible. Prerecognizable words in spoken language, primary mode of communication may be manual |
| 2 | Connected speech is unintelligible. Intelligible speech is developing in single words when context and lip-reading cues are available |
| 3 | Connected speech is intelligible to a listener who concentrates and lip-reads within a known context |
| 4 | Connected speech is intelligible to a listener who has little experience with a deaf person's speech |
| 5 | Connected speech is intelligible to all listeners. Child is understood easily in everyday contexts |

SIR indicates Speech Intelligibility Rating.

($F$ (2,58) = 193.8; $p < 0.001$), and "group"-"test interval" interaction effect ($F$ (2,58) = 8.9; $p = 0.001$). The mean SIR scores before CI surgery in the deaf-blind children were not significantly different from those of the deaf-only children (deaf-blind, 1.21 ± 0.58; deaf-only, 1.24 ± 0.44; $p = 0.90$). Pairwise comparisons on the significant main effect of "test interval" indicated that SIR scores in deaf-only children was significantly higher than deaf-blind children at "12-month post-CI" (mean difference, 0.870; $p = 0.01$) and "24-month post-CI" (mean difference, 1.067; $p = 0.002$) time intervals.

### WRS Measurement in Adults

The mean preoperative WRS values for the deaf-blind, and deaf-only adults was 44.86% (range, 32%–64%) and 54.0% (range, 38%–82%), respectively. The repeated-measures ANOVA revealed a significant main effect of "test interval" ($F$ (2,34)=211.3; $p < 0.001$), group ($F$ (2,34)=11.0; $p < 0.001$), and "group"-"test interval" interaction effect (F (1,17)=6.89; $p = 0.02$). There was no statistically significant difference in WRS values between either group before the CI surgery (deaf-blind: 44.86% ± 11.65%, deaf-only: 54.0% ± 9.26%; $p = 0.07$). Pairwise comparisons on the significant main effect of "test interval" indicated that there was a significant improvement in WRS values from the "preimplantation" to "post-CI" time-points in deaf-only adults (preoperative phase: 54.0% ± 9.26%, 12-month post-CI phase: 62.3% ± 9.1%, 24-month post-CI phase: 72.7% ± 8.7%; $p < 0.001$). Pairwise comparisons also demonstrated that there was a significant improvement in WRS values from the "preimplantation" to "post-CI" time-points in deaf-blind adults (preoperative phase: 54.0% ± 9.26%, 12-month post-CI phase: 51.71% ± 10.7%, 24-month post-CI phase: 56.86% ± 9.8%; $p < 0.001$). Furthermore, the mean WRS in the deaf-only adults was significantly higher than those in the deaf-blind adults at "12-month post-CI" (mean difference: 10.6%, $p = 0.03$) and "24-months post-CI" (mean difference: 15.8%, $p = 0.002$) follow-ups.

## DISCUSSION

In recent years, there has been great interest in the potential advantage of cochlear implantations in deaf-blind patients (26,27). Patients with dual sensory deficits, including hearing loss and blindness, experience a challenging situation that may affect their physical, psychological, and social performances. The visual system primarily influences the physical and spatial environment, whereas the auditory system influences social functioning in daily interactions (12,13,28). Capella-McDonnall (29) found that patients with dual sensory loss are more likely to experience symptoms of depression compared with those with no-sensory and single-sensory impairments.

Our results indicate that CI improves auditory performance and speech intelligibility skills in patients with concurrent visual and hearing deprivations. El-Kashlan et al. (16) reported that deprivation of one sensory input (e.g., vision) promoted an increase in cortical function serving another sensory input, such as hearing. Our results revealed that cochlear implantation has promoted auditory performance (i.e., CAP score) in deaf-blind and deaf-only children. In the present study, approximately 70% of children (deaf-only group, 12 of 17; deaf-blind group, 11 of 17) had a chance of reaching a CAP score of 6 or 7 within 2 years after cochlear implantation. These results correspond to "understanding of conversation with a familiar talker without lip-reading" (Level 6) or "use of telephone with a familiar talker" (Level 7). According to our results, it can be inferred that CI can lead to relatively high auditory function levels in deaf-blind and deaf-only children.

Although blind individuals may display higher speech perception abilities in several tasks, such as speech discrimination and voice processing (30,31), much less is known about the impact of blindness on speech production in CI recipients. Burlingham (32) reported that blind babies indicate longer babbling phases, as well as delays in the production of their first words. It seems that blindness result in delayed

**TABLE 7.** *Mean (± SD) of CAP, SIR, and WRS scores across different time intervals*

| Parameters | Group | Time of Assessment | | |
|---|---|---|---|---|
| | | Preimplantation | 12-mo Post-CI | 24-mo Post-CI |
| CAP | Deaf-blind children (n = 17) | 0.88 ± 0.71 | 3.24 ± 1.25 | 5.47 ± 1.06 |
| | Deaf-only children (n = 17) | 0.71 ± 0.47 | 3.94 ± 0.74 | 6.01 ± 0.79 |
| SIR | Deaf-blind children (n = 17) | 1.21 ± 0.58 | 2.07 ± 1.41 | 3.29 ± 1.20 |
| | Deaf-only children (n = 17) | 1.24 ± 0.44 | 2.94 ± 0.55 | 4.35 ± 0.49 |
| WRS (%) | Deaf-blind adults (n = 12) | 44.86 ± 11.65 | 51.71 ± 10.73 | 56.86 ± 9.78 |
| | Deaf-only adults (n = 12) | 54.01 ± 9.264 | 62.33 ± 9.09 | 72.67 ± 8.66 |

CAP indicates categories of auditory performance; SD, standard deviation; SIR, Speech Intelligibility Rating; WRS, word recognition score.

morphologic, pragmatic, and lexical development compared with sighted children (10). The current study indicated that speech intelligibility (i.e., SIR score) in deaf-blind and deaf-only children improved as the duration of cochlear implant use increased. We also found that speech performance in deaf-only CI children improved slightly higher than in deaf-blind CI children. Moreover, 2 years after cochlear implantation, the SIR scores reached Categories 4 or 5 in about 70% (14/19) and 80% (15/19) of deaf-only and deaf-blind children, respectively.

In a similar study, Nair et al. (19) compared the mean CAP, SIR and hearing-related QoL scores between the children with Usher's syndrome and nonsyndromic children (control group) who underwent CI. The authors reported that CI resulted in improved auditory performance, speech recognition, and hearing-related QoL from "3-month" to "12-month" post-CI intervals in both disease groups. However, the mean CAP and SIR scores in children with Usher's syndrome was significantly lower than the nonsyndromic implanted children.

Several studies suggested that improved speech recognition abilities in implanted individuals are directly correlated with earlier time of CI (before the age of 4 years), as well as with an emphasis on regular attendance at pre-CI and post-CI auditory training sessions. Thus, patients with long-term prelingual deafness obtain reduced benefits in objective auditory-only speech perception. The speech perception values in our study were higher than Hoshino et al. (33) results; they reported a 56% WRS score for patients with Usher syndrome. The mean age at implantation was 18.9 years (range, 5–49 yr), and they had received their devices at a later age. They showed that detection of sounds could be achieved with late CI, but speech recognition was only improved in subjects with previous acoustic stimulation (e.g., hearing aid fitting). Henricson et al. (7) indicated that phonologic and lexical abilities and working memory capacity in children with Usher's syndrome (seven children, aged 7 to 16 yr) after cochlear implantation were similar to hearing-impaired children using hearing aids.

Wiley et al. (34) studied the effect of CI surgery on language outcomes of deaf-blind children (age range, 8 mo to 8 yr). In terms of receptive language skills, 32% of children reached a level of sound detection, and 21% could identify words. In terms of expressive language skills, only 49% of children had sound production skills, and 18% could communicate with some words. Their findings showed great heterogeneity in CI language outcomes for children with DSIs. Takano et al. (12) also reviewed eight older adults with severe SNHL and visual impairment. They compared speech perception rates for words and sentences between subjects with visual (72.3% for word recognition, and 86.0% for sentence recognition) and without visual (62.1% for word recognition, and 78.5% for sentence recognition) deficits who had undergone implantation. However, they found no significant differences in the average speech perception scores between patients with and without visual impairment.

The effectiveness of CI on speech perception and oral communication skills has been documented by Jatana et al. (35). They showed that 92% of children (n = 26; age range, 13–30 mo) diagnosed with Usher syndrome developed significant open-set speech perception and oral communication abilities, with 69.2% using primarily oral communication by the time of the last follow-up.

Our findings indicated that multichannel CI is a beneficial procedure for rehabilitation of deaf-blind patients in both children and adults' populations, and can adequately restore auditory and speech abilities with stable results over time. According to our results, the deaf-blind implanted children had reached to similar auditory performance level of the deaf-only children. However, it takes longer time to reach the speech production levels similar to the deaf-only control.

Most studies that have been reported the effect of CI in deaf-blind patients are case reports or have been designed cross-sectionally and carried out in a single center. Then, one of the strengths of the current multicenter study was to assess the outcomes of CI in deaf-blind individuals during a 2-year time course. Furthermore, our study will provide an opportunity for professionals and clinicians to see the outcomes of CI in both children and adults' populations with varying etiologies.

The current study has its own limitations. Because of the complexities of rehabilitation process for the deaf-blind population, we may require additional time and expertise for optimal outcomes. Moreover, diverse population in different countries with different languages and rehabilitation methods, and different device used may affect the outcomes.

## CONCLUSION

Our findings indicated that auditory performance and speech production in deaf-blind individuals who were CI users improved over time. However, speech perception improvement in deaf-blind CI adults was significantly lower than that of their deaf-only counterparts. Despite some difficulties of CI in patients with dual sensory loss, this method can significantly enhance the ability to perceive and articulate speech, particularly when patients are implanted in appropriate time. Deaf-blind patients need unique and comprehensive care before CI, as well as further aural rehabilitation after CI, to obtain beneficial results.

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