Original Article

Dysmorphic Features and Cochlear Implantation Outcome in Children with Sensorineural Hearing Loss; the Headmost Study

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Abstract

Introduction: Recently, treatment of children with severe-to-profound sensorineural hearing loss (SNHL) has been influenced by diagnostic improvements and technological treatment advances, specifically new cochlear implant prospects. Multiple handicaps children and children with syndromes and conditions resulting disabilities, such as dual sensory loss, cerebral palsy, somatic abnormalities, and autistic spectrum disorder, are now not routinely precluded from receiving a cochlear implant. The primary aim of this study was to determine the effects of dysmorphic feature on cochlear implant outcome. **Materials and Methods:** In this cohort study, we evaluated 336 cochlear implanted children from 2007 to 2009. The case group consists of 53 patients (15/7%) with dysmorphic features and control group consisted at 53 patients with normal features and without behavioral and developmental disorders. All patients received auditory and speech rehabilitation and we evaluated their speech and auditory outcome. One year after cochlear implantation, the patient was assessed by categories of auditory perception (CAP) and speech intelligibility rating (SIR) tests. **Results:** We included 106 out of 336 cochlear implanted children, with the mean age of 30.42 ± 12.16 (maximum 48 months), 52 cases (49.1%) were girls and 54 (50.9%) were boys. There was a significant difference in SIR between case and control groups (3.26 ± 0.98) versus (4.06 ± 0.94) ($P < 0.001$), and a significant difference in CAP (4.09 ± 1.26) versus (5.43 ± 1.23) as well ($P < 0.001$). **Conclusion:** In this study, the prevalence of dysmorphic feature in children with severe-to-profound SNHL is 15.7%. One year after cochlear implant SIR and categories of auditory perception in these patients are significantly lower than children without dysmorphic feature, but cochlear implant will help these children.

Keywords: Categories of auditory perception scales, cochlear implantation, dysmorphic feature, sensorineural hearing loss, speech intelligibility rating

Introduction

About 3% of infants are born with congenital abnormalities including congenital hearing loss which is one of the most common permanent disabilities.[1] These problems are one of the factors for spreading diseases, mortality and disability among children. Fifty percent of defects are unknowable but some studies approved that 18% have chromosome origin, <10% are environmental, and chemical problems and 25%–30% are genetically and acquired problems.[2-4] Some of these abnormalities are hydrocephaly, microcephaly, macrocephaly, auricle, cleft lips, heterocromy, and strabismus. All of these disabilities and congenital abnormalities impose expensive cost to society.[5-8] In other hand, some of severe congenital abnormalities cause abortion or intrauterine fetal death. Therefore, it is logical to identifying and prevention rather than treatment or rehabilitation in congenital abnormality. Apparent and physical defects such as cleft lips or external ear with hearing loss could have embryonic origin.[9] Hearing loss is the most common cause of scenes defect in children which is divided into hearing impaired before and after speech development. Cochlear implantation (CI) is performed on bilateral hearing loss <90 db and postlingual deafness children caused by infection.[3,10] This method is more than 80% curable and improves considerably speech and hearing

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ability of children. Children who have multiple syndromes such as bilateral hearing loss, cerebral palsy, delayed global development, and autism spectrum disorders with hearing defects do not ban for routinely CI.[5,11,12] In the beginning, candidates for CI were children between 1 and 4 years old with bilateral sever hearing loss and did not have any physical and mental problem, but during the development of CI, this method has been performing on children with cerebral palsy, behavioral disorders, and optical problems.[12,13] The past studies about CI have reported this method as an effective and cost benefit for curing children with bilateral severe hearing loss. We conducted this study to determining speech an auditory sequence in deaf children with physical dimorphism pre- and post-CI operation.

Materials and Methods

Study design and participants

This cohort study was performed on children with profound SNHL that underwent CI at Baqyiatallah CI Center between 2007 and 2009. The children who were selected for control group included these criteria: (1) Permanent congenital SNHL, (2) onset of hearing loss before 6 months of age, (3) the use of amplification and/or intervention program emphasizing spoken language, (4) Persian as the language of communication, (5) the maximum age was 4 years old, and (6) without any evidence of dysmorphic features, anatomical, behavioral, and developmental disorders.[5] We enrolled all children who had undergone CI and then divided them into two groups based on physical abnormalities. Every patient was assessed by a clinical psychologist, an audiologist, a pediatrician, and a speech/language pathologist.

Intervention

Each children who have more than 15 db hearing falling considers as hearing impaired and if someone's hearing ability falls more than 70 db considers as deaf person. CI is performed on children who have lower 90 db hearing ability. Speech ability means able to produce sounds and understand the speaking which is assessed by speech intelligibility rating (SIR) [Table 1]. Hearing skill means able to understand voices which are assessed by CAP [Table 1].

All patients (children with PMD) had profound levels of sensorineural hearing impairment based on the average of preimplantation unaided pure-tone thresholds over 90 dB hearing loss. The profound SNHL was confirmed by pure speech audiometry, auditory brainstem response with click and tone burst methods, transient-evoked otoacoustic emissions, and distortion-product otoacoustic emissions. None of children had the experience of speech perception from properly fit power hearing aids.

All patients received audiological, speech perception, language skills, neurological, and psychological assessment immediately before CI. Imaging study consists of magnetic resonance imaging was performed for finding the central nervous system abnormality and overruling other cause of SNHL. The nucleus 22 channel device and a speech processors device were used routinely although other options have been considered in special subjects.

Table 1: Speech intelligibility ratings

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

Simple mastoidectomy was the surgery approach on children under general anesthesia. By posterior tympanotomy, the middle ear space was accessible. Then, the bone surrounding the round window was cut and its' membrane was perforated to drain the endolymph. Finally, electrodes using the "advance off-stylet insertion technique," the less endamage to the mediolus, were inserted into the cochlea completely. If there was no possibility to insert a cochlear electrode completely, because of severe labyrinthitis ossificans, the partial insertion was used. After insertion, correct placement of electrodes was examined using of the fluoroscopy.

Outcome assessments

Speech skills are ability of producing voice and concept of speech based on SIR criteria and Auditory skills defined as the ability of sound understanding which is assayed on CAP criteria. All children were examined by speech and auditory tests which were CAP (categories of auditory performance) and SIR.

We included children based on CAP and SIR as case group, but some of them with behavioral problem and evolutional delayed problem categorize in this group too. Children, with the absence of: (1) behavioral problem and (2) evolutional delayed problem, were chosen as control group. All children between 1 and 4 years old with hearing loss or deaf were observed by personnel of Baqiyatallah CI and were referred to pediatrics neurology clinic for advanced examination. Children were categorized by existing of physical abnormalities and adequate information such as deaf problem, and demographics data were received from parents. Physical abnormalities included defects in the eyes, ears, mouth, etc., which observed by a pediatric neurologist [Table 2]. Control group was children without any physical abnormalities that candidate for CI. During 12 months, after CI, all children participated in auditory

performance and speech intelligibility classes. Professional trainer saved scores in all exams such as invention, identifying, distinction, and perception.

Scientific and ethic considerations

Medical records requested with permission from outside hospitals and from Baqiyatallah records. All procedures were approved by children's parents. This investigation was approved by the Ethical Review Board at Baqiyatallah University of Medical Sciences, Tehran, IR Iran.

Statistical analysis

All quantitative variables have normal distribution examined by using Kolmogorov–Smirnov test. The frequency and prevalence, Paired and independent *t*-test and ANOVA test in addition to repeated measures ANOVA were used for analyzing CAP and SIR score between baseline and 3 or 6 months after implantation.

Results

Three hundred and thirty-six children were undergone CI during 2 years. Out of these children, 53 (15.7%) had abnormal physical problem. A total of 53 children did not have other concurrent problem. Totally 106 children were enrolled in the study, and 52 (49.1%) were male and 54 (59.1%) female. Among children with abnormal physical problem, 30 (56.5%) were male and 23 (43.4%) were female, and in 53 children who did not have abnormal physical problems, 24 (45.3%) were male and 29 (54.7%) were female. The prevalence of anatomical abnormality in children according to gender is showed in Table 3.

The mean age of all participated children was 30.48 ± 12.16 month with age range of 12–48 months. The mean age of case group was 31.7 ± 11.12 and mean age of control group was 29.26 ± 13.11. CAP and SIR were performed for all groups 1 year after CI operation, and in all of them, an average CAP score was 4.76 ± 1.41 and SIR was 3.66 ± 1.41.

The outcomes of auditory speech perception tests were compared before surgery and 24 months after the device was switched on for all cases and then were compared between our two groups. The scores of CAP and SIR are shown in children with or without malformation after CI operation [Tables 4 and 5]. SIR and CAP could not be assessed before CI operation so average score be considered zero. Thus, difference and development in CAP and SIR scores was considered 12 months later. The sequence of SIR and CAP is considered as the change of SIR and CAP score. CAP score of children with physical malformation was assessed 4.09 ± 1.26 after CI operation. CAP score of children without physical malformation was assessed 5.43 ± 1.23 after CI operation. There is significant difference in CAP score of both the groups ($P < 0.001$). SIR score of children with physical malformation was assessed 3.26 ± 0.98 after CI operation. SIR score of children without physical malformation was assessed 4.06 ± 0.94 after CI operation. There is difference in comparison of SIR score of both the groups ($P < 0.001$). The most common physical abnormalities were microcephaly (28.3%), lop ear (17%), epicanthic fold (17%), and low set air (15.1%). Other abnormalities are showed in Table 6.

Table 2: Categories of auditory perception scale

Categories	Description
Category 0	No awareness of environmental sounds
Category 1	Awareness of environmental sounds
Category 2	Response to speech sounds (e.g., "go")
Category 3	Identification of environmental sounds
Category 4	Discrimination of some speech sounds without lipreading
Category 5	5 understanding of common phrases without lipreading
Category 6	Understanding of conversation without lipreading
Category 7	Use of telephone with known listener books

Table 3: The prevalence of anatomical abnormality in children according to gender

	Abnormal anatomical problem (%)	Without abnormal anatomical problem (%)
Male	30 children (56.5)	24 children (45.3)
Female	23 children (43.4)	29 children (54.7)

Discussion

This study was performed to evaluate the effect of physical abnormalities on hearing scores before and after CI in children with congenital SNHL. This study showed that the prevalence of physical abnormalities on children with congenital SNHL has been about 15%; on the other hand, other studies have shown that 2%–3% of infants are born with one congenital abnormality. Hence, there are many accompaniments with congenital SNHL and physical abnormalities, and physician should pay attention to this point.

Auditory perception and speech intelligibility tests had not assessed before CI, so these tests were performed 12 months later. The result showed that these children made a profit with CI and this could improve their CAP and SIR scores significantly.

In comparison with physical abnormalities patients and normal group, there was a significant meaning between CAP and SIR. In this way, normal group made a profit and improved after CI, but more important point that children with physical abnormalities also made a profit with CI less than normal group. Training and education could similar these children with normal group.

We could categorize children with disability as physical abnormality group. It has been some studies about children with disability or other defects in addition to hearing loss.[14-16] Waltzman *et al*. studied on deaf children who suffered

Table 4: CAP criteria scores after cochlear implantation in children with physical abnormalities and normal children

CAP criteria after CI	1	2	3	4	5	6	7
Physical abnormalities (%)	0	2 (3.8)	18 (34)	17 (32.1)	9 (17)	3 (5.7)	4 (7.5)
Normal (%)	0	0	3 (5.7)	8 (15.1)	21 (39.6)	5 (9.4)	16 (30.2)

CI: Cochlear implantation

Table 5: Speech intelligibility ratings criteria scores after cochlear implantation in children with physical abnormalities and normal children

SIR criteria after CI	1	2	3	4	5
Physical abnormalities (%)	1 (1/91)	11 (20/8)	25 (37/7)	15 (28/3)	6 (11/3)
Normal (%)	0	3 (5.7)	13 (24.5)	15 (28.3)	22 (41.5)

SIR: Speech intelligibility ratings, CI: Cochlear implantation

Table 6: The prevalence of Physical abnormalities

Physical abnormalities	Number of person (%)
Heterochromia iridis	5 (9.4)
Pseudostrabismus	5 (9.4
Ophthalmic problem (asymmetry of eye-strabismus)	2 (3.8)
Epicanthic fold	9 (17)
Wide nasal root	1 (1.9)
External ear atresia	1 (1.9)
Lips hyperplasia	1 (1.9)
Lips anomaly	1 (1.9)
Frontal bossing	3 (5.7)
Lop ear	9 (17)
Low set ear	8 (15.1)
Simian Line	2 (3.8)
Cleft palate	2 (3.8)
Cleft lips	2 (3.8)
White forelock	4 (7.5)
Cubic form head	1 (1.9)
Microcephaly	15 (28.3)
Macrocephaly	2 (3.8)
Hydrocephaly	1 (1.9)

from disability, and after CI, they made a profit with this operation.[14] Filipo *et al*. agreed this subject in their own article. We approved in our study previous researches and suggest that there is no contraindication between physical abnormality and CI.[17]

Conclusion

Although there were not any other studies about these subjects and studies only were performed on appearance disorders patients, it seems this study is one of the first researches about effects of physical abnormalities and results of CI. In our center, the prevalence of dysmorphic feature in children with severe-to-profound SNHL is 15.7%. One year after cochlear implant SIR and categories of auditory perception in these patients are significantly lower than children without dysmorphic feature, but cochlear implant will help these children.

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Conflicts of interest

There are no conflicts of interest.

References

1. Amirsalari S, Saburi A, Radfar S, Yousefi J, Nouhi S, Hassanalifard M, *et al*. Hearing loss prevalence and risk factors among Iranian deaf children: The first report from Iran. J Clin Neonatol 2014;3:191-4.
2. Available from: https://www.cdc.gov/ncbddd/hearingloss/genetics.html. [Last reviewed on 2018 Apr 11].
3. Paludetti G, Conti G, DI Nardo W, DE Corso E, Rolesi R, Picciotti PM, *et al.* Infant hearing loss: From diagnosis to therapy official report of XXI conference of Italian society of pediatric otorhinolaryngology. Acta Otorhinolaryngol Ital 2012;32:347-70.
4. Matsunaga T. Value of genetic testing in the otological approach for sensorineural hearing loss. Keio J Med 2009;58:216-22.
5. Amirsalari S, Yousefi J, Radfar S, Saburi A, Tavallaie SA, Hosseini MJ, *et al.* Cochlear implant outcomes in children with motor developmental delay. Int J Pediatr Otorhinolaryngol 2012;76:100-3.
6. Najafiyan B, Saburi A, Amirsalari S. Report of cochlear implantation outcome in a case of waardenburg syndrome. Int J Pediatr Otorhinolaryngol 2012;76:761.
7. Amirsalari S, Ajallouyean M, Saburi A, Haddadi Fard A, Abed M, Ghazavi Y, *et al.* Cochlear implantation outcomes in children with Waardenburg syndrome. Eur Arch Otorhinolaryngol 2012;269:2179-83.
8. Tarhan E, Oğuz H, Safak MA, Samim E. The carpenter syndrome phenotype. Int J Pediatr Otorhinolaryngol 2004;68:353-7.
9. Ma X, Li YW, Ma L, McPherson B. Chinese children with nonsyndromic cleft lip/palate: Factors associated with hearing disorder. Int J Pediatr Otorhinolaryngol 2016;88:117-23.
10. Hasanalifard M, Ajalloueyan M, Amirsalari S, Saburi A. Outcome of cochlear implantation in post-meningitis deaf children. Iran Red Crescent Med J 2013;15:15-7.
11. Steven RA, Green KM, Broomfield SJ, Henderson LA, Ramsden RT, Bruce IA, *et al.* Cochlear implantation in children with cerebral palsy. Int J Pediatr Otorhinolaryngol 2011;75:1427-30.
12. Birman CS, Elliott EJ, Gibson WP. Pediatric cochlear implants: Additional disabilities prevalence, risk factors, and effect on language outcomes. Otol Neurotol 2012;33:1347-52.
13. Peters JP, Ramakers GG, Smit AL, Grolman W. Cochlear implantation in children with unilateral hearing loss: A systematic review. Laryngoscope 2016;126:713-21.

14. Waltzman SB, Scalchunes V, Cohen NL. Performance of multiply handicapped children using cochlear implants. Am J Otol 2000;21:329-35.
15. Schwartz SR, Chen BS. The role of preoperative imaging for cochlear implantation in postlingually deafened adults. Otol Neurotol 2014;35:1536-40.
16. Khosravi A, Amirsalari S, Ajalloueyan M, Honaramooz A, Tafreshi R, Saburi A. The frequency of congenital long QT syndrome based on new formula in children with sensori-neural hearing loss. Indian J Otol 2015;21:114-8.
17. Filipo R, Bosco E, Mancini P, Ballantyne D. Cochlear implants in special cases: Deafness in the presence of disabilities and/or associated problems. Acta Otolaryngol Suppl 2004:552:74-80.