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Speech perception and production in children with inner ear malformations after cochlear implantation

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ABSTRACT

Objective: The aim of this study was to assess the speech perception and speech intelligibility outcome after cochlear implantation in children with malformed inner ear and to compare them with a group of congenitally deaf children implantees without inner ear malformation.
Methods: Six deaf children (five boys and one girl) with inner ear malformations who were implanted and followed in our clinic were included. These children were matched with six implanted children with normal cochlea for age at implantation and duration of cochlear implant use. All subjects were tested with the internationally used battery tests of listening progress profile (LiP), capacity of auditory performance (CAP), and speech intelligibility rating (SIR). A closed and open set word perception test adapted to the Modern Greek language was also used. In the dysplastic group, two children suffered from CHARGE syndrome, another two from mental retardation, and two children grew up in bilingual homes.
Results: At least two years after switch-on, the dysplastic group scored mean LiP 62%, CAP 3.8, SIR 2.1, closed-set 61%, and open-set 49%. The children without inner ear dysplasia achieved significantly better scores, except for CAP which this difference was marginally statistically significant ($p = 0.009$ for LiP, $p = 0.080$ for CAP, $p = 0.041$ for SIR, $p = 0.011$ for closed-set, and $p = 0.006$ for open-set tests).
Conclusion: All of the implanted children with malformed inner ear showed benefit of auditory perception and speech production. However, the children with inner ear malformation performed less well compared with the children without inner ear dysplasia. This was possibly due to the high proportion of disabilities detected in the dysplastic group, such as CHARGE syndrome and mental retardation. Bilingualism could also be considered as a factor which possibly affects the outcome of implanted children. Therefore, children with malformed inner ear should be preoperatively evaluated for cognitive and developmental delay. In this case, counseling for the parents is mandatory in order to explain the possible impact of the diagnosed disabilities on performance and habilitation.

1. Introduction

According to studies of the last decade [1–6], the incidence of inner ear malformations was reported to range from 6.9 to 35% of the congenitally deaf children who underwent cochlear implantation. Malformation of the inner ear was initially considered to be a contraindication to cochlear implantation. Earlier studies showed that in malformed cochleas the spiral ganglion cell populations were substantially diminished [7,8]. Schmidt [8] found 7677 cochlear neurons in a case of Mondini dysplasia. Paparella and el-Fiky [9] reported a case of Mondini dysplasia that had 10% of the normal number of the spiral ganglion cells (normal hearing subjects have approximately 35,000 neurons) in the basal turn of cochlea and 40% of the normal number in the apex. Since a certain amount of proximal neural elements should be present for an encoded signal to be effectively transmitted towards the auditory cortex, the question of how beneficial the cochlear implantation could be in such cases had been raised. It was then demonstrated in cochlear implant users that the minimal number of ganglion cells required for good speech perception was quite low, about 5000 cells [10]. Moreover, it was supported that children with inner ear malformation who underwent cochlear implantation demonstrated significant improvements in their speech recognition skills in comparison with their preoperative performance with hearing aids [1–6,11–23]; in addition, their audiological results were even comparable to that of children with implants who had radiographically normal cochlea [4,12,13].

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Table 1
Clinical data and surgical aspects of patients with malformed cochlea.

Child	Sex	Type of deafness	Age at CI (years)	Duration of CI use	Malformation	Systemic handicaps	Type of CI	Surgical complications
1. L.P.	M	C	5	27	Incomplete partition	CHARGE syndrome	Medel Sonata	Gusher
2. T.A.	F	C	4.5	23	Incomplete partition	Marginal mental retardation	Freedom straight	Gusher, meningitis
3. P.P.	M	C	5	28	Incomplete partition	No	Medel Pulsar	No
4. P.E.	M	C	3	28	Incomplete partition	No	Cochlear Contour	No
5. Pr.G.	M	P	15	25	Hypoplasia of vestibule and aplasia of semicircular canals	CHARGE syndrome	Cochlear Contour	No
6. Pz.G.	M	C	7	120	Incomplete partition	Mild mental retardation, epilepsy	Nucleus 24 M	No

CI: cochlear implant, M: male, F: female, C: congenital, P: progressive.

The purpose of this study was to present our experience about the outcome of speech perception and speech intelligibility after cochlear implantation in children with malformed inner ear and to compare them with a group of congenitally deaf children implantees without inner ear malformation. A review of the literature was also attempted regarding the speech outcome of children with inner ear malformation in order to better determine the performance of this category of deaf children after cochlear implantation.

2. Material and methods

2.1. Subjects

Six deaf children (five boys and one girl) with inner ear malformations who were implanted and followed in our clinic were included. The demographic and clinical data of all children are demonstrated in Table 1. Using the radiological classification of Papsin [1], five children were presented with incomplete partition and one with isolated posterior labyrinthine dysplasia (hypoplasia of vestibule and aplasia of semicircular canals) (Table 1).

Perioperatively, in child 1, a slight gusher occurred which was stopped after electrode insertion, with no other particular measures.

Child 2 was first implanted with Freedom cochlear implant (Cochlear Contour) on the left ear, but gusher occurred at the moment of cochleostomy; although the gusher was stopped after sealing the cochleostomy with temporalis fascia and tissue col, cerebrospinal fluid (CSF) otorrhea has reappeared postoperatively. In the following days, the child developed meningitis and the cochlear implant was removed. Two years later, this girl underwent cochlear implantation successfully on the right ear (Freedom with straight electrode).

In child 5, initial attempt to implant was on the left side at the age of 12 years, but the creation of a cochleostomy was not possible due to an unexpected medial rotation of the cochlea; two years later, a total insertion of Freedom implant (Cochlear Contour) was performed on the right ear using Navigator system.

In the other children (cases 3, 4, 6) the electrode insertion was complete and the postoperative course was uneventful.

2.2. Methods

Informed consent was obtained from the parents of all participating children. The study was approved by the Bioethics Committee of the Medical School of the Aristotle University of Thessaloniki.

The following internationally used and reliable instruments were administered in our children to measure auditory-speech perception and speech intelligibility: the listening progress profile (LiP), which was devised to assess the early stage of auditory-speech perception after implantation (Table 2) [24]; the capacity of auditory performance (CAP), which comprised a nonlinear hierarchical scale of auditory receptive abilities, ranging from 0 to 7 (Table 3) [25]; the speech intelligibility rating (SIR), which is a 5-point rating scale that quantifies the speech production abilities (Table 4) [26]. All these tests battery were appropriately adapted to the Greek language.

Our sample were also subjected to the Greek Spondee word test, which included five closed sets of four spondee pictured words familiar to the child and were spoken with equal emphasis on each syllable at an average presentation level of 70 dB SPL. Each child was required to match the picture with the correct word (the total score of the test was 100).

Our open-set battery included two subtests: The first consisted of list of 50 words, 40 bisyllabic words and 10 mono-, trisyllabic words; this was because the Modern Greek language consists mostly of bisyllabic words and there are a limited number of monosyllabic words. The words were administered in the auditory-only mode and the children were instructed to repeat the words that they heard. The second subtest consisted in "Mr Potato Head and his bucket of parts" [27], in which the child was asked to recognize his body parts; this test included two lists of 10 questions, and the performance was scored on the basis of words and sentences correctly identified. Although technically the above test was a closed-set test, it was progressively modified to an open-set test as the number of alternatives became larger. The median score of both subtests was obtained in terms of the percentage of words and phonemes correctly understood.

The above tests were performed during a follow-up generally after an average implant use of two years, except for one case which was evaluated 10 years after cochlear implantation (Table 1). During this assessment, the aided thresholds of all these children for frequencies of 0.5–4 kHz achieved with their cochlear implant were within the region of 30–40 dB (A). No measurable closed/open set speech perception and production was found during the preoperative evaluation period. In the same period, all

Table 2
The listening progress profile (LiP).

Response to environmental sounds
Response to drum (elicited)
Response to musical instrument (elicited)
Response to voice-elicited
Response to voice-spontaneous
Discrimination between 2 different instruments
Discrimination between loud and quit drum
Discrimination between single and repeated drum
Identification of environmental sounds
Response to [oo, ah, ee, ss, mm][a]
Discrimination between long and short speech sounds[a]
Single/repeated sounds[a]
Loud/quiet speech sounds[a]
2 of ling five sounds[a]
All of Ling sounds[a]
2 family names of different syllable length[a]
Identification of own name in quiet

The score (never = 0, sometimes = 1, always = 2) was modified for %.
[a] Adapted for Greek language.