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Cochlear implantation in children with cochlear nerve deficiency: A report of nine cases

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ABSTRACT

Objective: Cochlear implantation for children with cochlear nerve deficiency remains controversial, as the presence of the cochlear nerve has been central to the success of cochlear implantation. This study sought to investigate whether there is any benefit from cochlear implantation for children with cochlear nerve deficiency.
Methods: Nine children with cochlear nerve deficiency and bilateral prelingual profound sensorineural hearing loss were included in this study. Inner ear and internal auditory canal structures were evaluated using magnetic resonance imaging and temporal bone computed tomography scans. Meaningful auditory integration scales, categories of auditory performance scores, speech intelligibility ratings and pure tone average threshold with cochlear implantation were measured for evaluation of hearing and speech performance.
Results: Only four (44.4%) children had a significant improvement in pure tone average threshold with the cochlear implant device (77.5 dBHL, 45 dBHL, 51.3 dBHL and 68.8 dBHL). No child achieved sufficient speech intelligibility or perception ability during a follow-up of at least one year after surgery.
Conclusions: The decision to perform cochlear implantation in children with cochlear nerve deficiency must be undertaken with caution as it has limited effectiveness and uncertain cost-benefit.

1. Introduction

Cochlear implantation (CI) has been proven to be the most promising and effective method for restoring the auditory stimulus in children with bilateral prelingual severe or profound sensorineural hearing loss (SNHL). Furthermore, the worldwide use of CI, as well as technological improvements, has expanded the indications for CI, which means more and more children can benefit. Previously viewed as relative contraindications, factors such as age (<12 months) [1], single-sided deafness [2], tinnitus [3], inner ear malformations [4], and auditory neuropathy [5], are now becoming legal indications under certain circumstances [6].

Nonetheless, CI for some children with severe inner ear malformations, especially with cochlear nerve deficiency (CND), remains controversial, as the presence of the cochlear nerve (CN) has been central to the success of CI. Choosing between the relative indications for CI and the family's desperate need for aid is a difficult clinical issue for many physicians. Until now, in developing countries such as China, CI is the only possible chance for improving hearing and speech ability in these children, due to the absence of, and lack of experience with, auditory brainstem implantation (ABI). To our knowledge, only a few of authors have reported their experience of CI in children with CND [7–11].

As is generally known, the functional outcome of pediatric CI depends on several major prognostic factors [12], including the functional presence of CN, age at implantation, genetic mutations (e.g., connexin 26), inner ear malformations, and meningitis. Therefore, it is essential to identify all potential adverse factors at the outset [10]. This study sought to investigate whether there is any benefit from CI for children with CND. We focused on postoperative functional outcome in order to analyze the possible effectiveness of CI in these children.

2. Materials and methods

This retrospective study has been approved by the Ethical Committees of Xinhua hospital, Shanghai Jiaotong University School of Medicine. Children with bilateral prelingual profound

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SNHL, who were referred to our center, were considered for this study. All children with radiographically documented CND who had undergone CI between 2005 and 2010, were included in our analysis.

All children had completed a series of detailed assessments, including medical, audiological, speech and language assessment, and neuroimaging. Standard audiological assessment included otoacoustic emissions (transient-evoked otoacoustic emission and distortion product otoacoustic emission), auditory brainstem evoked responses (ABR), auditory steady-state responses (ASSR), and acoustic emittance. Aided hearing threshold was evaluated using visual reinforcement audiometry testing. The tests were routinely performed two or three times during the observation period before surgery.

One week before surgery, an electrically evoked auditory brain stem response (EABR) was performed under general anesthesia (propofol) in the operating room, as previously described [13]. The stimuli were generated by a custom-made stimulator triggered by a 5V-TTL pulse from a Bio-logic Systems Navigator evoked potential system (Bio-logic Systems Corp, Mundelein, IL). The custom-made stimulator was controlled by programming software "SCLIN 2000" (Advanced Bionics Co). Stimuli consisted of balanced biphasic constant current pulses (75–150 μs per phase) ranging in level from 0 to 2040 CU at a rate of 23 per second.

All children commenced daily training and daily sessions with the speech therapist after switching on at 1 month. Children either had a cochlear implant device alone, or in combination with a contralateral traditional power hearing aid (HA). After surgery, the meaningful auditory integration scale (MAIS) [14], the categories of auditory performance (CAP) score [15] and the speech intelligibility rating (SIR) [16,17] were used to measure speech perception ability. The MAIS is a parent report scale designed to assess a child's spontaneous response to environmental sound. The CAP score is a scale designed to evaluate outcomes of auditory perception in everyday life following CI. The SIR test uses subjective estimations of the proportion of words understood to quantify the intelligibility of continuous speech. Pure tone average (PTA) threshold with cochlear implant device in the free field was used to assess auditory levels after surgery. All auditory tests were performed by the same experienced pediatric audiologist (Dr. Yun Li). The postoperative audiological results were recorded and compared at 1 month, 6 months and 1 year after the implant was switched on.

Neuroimaging of temporal bone, including temporal bone computed tomography (TBCT) scans and magnetic resonance imaging (MRI), was routinely performed to evaluate CND and other inner ear structural abnormalities. A sedative (chloral hydrate) was used for children who could not fall asleep during auditory tests and neuroimaging procedures. Direct axial and coronal TBCT scans were performed with a dual source CT scanner (Siemens Somatom Definition) using a standard temporal bone protocol. CT examinations were performed with 120 kV and 230 mA, with a section thickness of 1.25 mm and a pitch of 0.562:1. The Fov was 16 cm using a 512 × 512 matrix. The window width was 4000 HU and the window level was 600 HU. A 2D multi-planar reconstruction(MPR) of the coronal image was obtained. MRI was performed using a 3.0-T (GE, HDx) or 1.5-T system (GE Twin speed plus) with an eight-channel sensitivity encoding head coil. A high resolution 3D FIESTA sequence was chosen with a TR of 7.9 ms, and a TE of 4.2 ms. The Fov was 16–20 cm using a 320 × 256 matrix, with 0.8 mm-thick sections. The total scan time was approximately 4 min. The vestibulocochlear nerves (VCN) were constructed with 0.4 mm-thick sections and a 3.2 cm Fov, which was perpendicular to the internal auditory canal (IAC). A 3-D maximum intensity projection (MIP) reconstruction was chosen for depicting the inner structures. The nerves at the porus and in the IAC were identified in the reconstructed parasagittal oblique plane perpendicular to the IAC.

The CND was defined as the diameter in the midportion of the IAC less than that of the adjacent facial nerve (FN), or absent on the reconstructed parasagittal oblique plan, either congenital or acquired [18]. It were classified using Govaerts' system, which is based on the affected branch of the nerve and the related labyrinthine dysplasia on MRI findings [7]: Type I, total absence of the cochleovestibular nerve (CVN); Type IIa, CN branch absent or hypoplastic, VN present, dysplasia of the cochleovestibular labyrinth; and Type IIb, CN branch absent or hypoplastic, VN present, normal morphology of the cochleovestibular labyrinth. Facial nerve function was assessed using the House–Brackmann (H–B) grading system.

3. Results

3.1. Children and clinical features

Between 2005 and 2010, nine children, seven girls and two boys, with bilateral prelingual profound hearing loss and CND, were included in this study. Their clinical features can be seen in Table 1. The mean age at diagnosis was 7.8 ± 7.5 months (birth to 24 months). With the exception of one patient (No. 1) who had cerebral palsy, the remaining eight children were diagnosed with non-syndromic hearing loss. The children were mostly from rural areas, which explains why only three cases were detected in the universal newborn hearing screening (UNHS) program, and problems in the other six cases were detected by their own families. All children had normal preoperative FN function, H-B grade I.

Five children (Nos. 4, 5, 7, 8 and 9) accepted a *GJB2* gene test, among whom one patient (No. 8) was found to be homozygous for p.V27I and p.E114G. One patient (No. 9) was heterozygous for p.V27I. No polymorphisms or mutations in *GJB2* were detected in the other three children (Nos. 4, 5 and 7).

Six Medel C40+, one Medel Pulsar CI100, one HiRes 90K and one Nucleus N24R were implanted in the nine children. One implantation (No. 1) was performed in 2005, two (Nos. 2 and 3)

Table 1
Clinical features of children.

No.	Children	Gender	Age at diagnosis (months)	Detection	Speech training	Growth and cognitive development level	Age at surgery (months)	Cochlear implant device	Implanted ear
1	LXC	F	10	Family	RC	Delayed with CP	48	Medel C40+	Right
2	CJQ	F	5	Family	Family	Delayed	21	Medel C40+	Right
3	CSY	F	1	UNHS	Family	Normal	24	Medel C40+	Right
4	WSM	F	14	Family	RC	Normal	49	Medel C40+	Right
5	ZLX	F	1	UNHS	Family	Normal	18	HiRes 90K	Right
6	WKY	F	24	Family	Family	Normal	25	Medel C40+	Right
7	LXZ	F	8	Family	Family	Normal	14	Medel C40+	Right
8	TJB	M	6	Family	Family	Normal	25	Nucleus N24R	Left
9	WXY	M	1	UNHS	Family	Normal	14	Medel Pulsar 100	Right

RC: rehabilitation center; CP: cerebral palsy; UNHS: universal newborn hearing screening.