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Cochlear implantation in children with cochlear nerve deficiency

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ABSTRACT

Objective: The aim of this study was to report on auditory performance after cochlear implantation in children with cochlear nerve deficiency.
Methods: A retrospective case review was performed. Five patients with pre-lingual profound sensorineural hearing loss implanted in an ear with cochlear nerve deficiency participated in the study. Postoperative auditory and speech performance was assessed using warble tone average threshold with cochlear implant, speech perception categories, and speech intelligibility ratings. All patients underwent high resolution computed tomography and magnetic resonance imaging.
Results: According to Govaerts classification, three children had a type IIb and two a type IIa cochlear nerve deficiency. Preoperatively, four patients were placed into speech perception category 1 and one into category 2. All patients had an improvement in hearing threshold with the cochlear implant. Despite this, at the last follow-up (range 18–81 months, average 45 months), only one girl benefited from cochlear implantation; she moved from speech perception category 2 to 6 and developed spoken language. Another child developed closed set speech perception and had connected speech that was unintelligible. The other 3 children showed little benefit from the cochlear implant and obtained only an improved access to environmental sounds and improved lipreading skills. None of these 4 children developed a spoken language, but they were all full-time users of their implants.
Conclusions: The outcomes of cochlear implantation in these five children with cochlear nerve deficiency are extremely variable, ranging from sporadic cases in which open set speech perception and acquisition of a spoken language are achieved, to most cases in which only an improved access to environmental sound develops. Regardless of these limited outcomes, all patients in our series use their device on a daily basis and derive benefits in everyday life. In our opinion, cochlear implantation can be a viable option in children with cochlear nerve deficiency, but careful counseling to the family on possible restricted benefit is needed.

1. Introduction

Cochlear implantation is widely considered a highly effective method for auditory rehabilitation in children with severe and profound sensorineural hearing loss (SNHL). As experience in the field of cochlear implantation has increased, the candidacy criteria have been expanded and children with inner ear malformations or additional disabilities are now considered as candidates [1–4]. Since cochlear implants work by stimulating the cochlear nerve, an implicit cornerstone to achieve satisfactory outcomes is the presence of peripheral neural population sufficient for the development of a synchronized neural response on electric stimulation. Accordingly, the absence of the cochlear nerve has been considered an absolute contraindication to cochlear implantation. Recent advances in magnetic resonance imaging (MRI) have allowed individual evaluation of the four nerves within the internal auditory canal (IAC) and the assessment of presence and size of the cochleovestibular nerve (CNV). It has been demonstrated that cochlear nerve dysplasia can encompass different situations ranging from a truly aplasia (cochlear nerve could not be identified on the axial, coronal, and oblique plane sagittal images) to hypoplasia (when it appears decreased in size compared with the other nerves of IAC); furthermore, an extremely small nerve that would appear absent owing to the limited spatial

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Table 1
Govaerts classification of hypoplasia and aplasia of the vestibulocochlear nerve (7).

Type	Cochleovestibular nerve features	Labyrinthine remarks
I	Total absence of the cochleovestibular nerve	Normal or dysplastic labyrinth
IIa	Cochlear branch absent or hypoplastic; vestibular nerve present	Labyrinth dysplasia ranging from minor dysplasia to a common cavity
IIb	Cochlear branch absent or hypoplastic; vestibular nerve present	Normal labyrinth

resolution of MRI has been defined as "imperceptible cochlear nerve" [5]. Currently, cochlear nerve deficiency (CND) is the most widely used term in the literature to describe both an absent and a small cochlear nerve. CND is observed in up to 18% of children with SNHL [6]; recently, a classification of eighth nerve dysplasia into three categories – based on the affected branch of the nerve and the related labyrinthine dysplasia – has been proposed [7].

Several authors state that patients with CND have no chance to benefit from traditional cochlear implants and suggested direct stimulation of the cochlear nuclei by means of auditory brainstem implantation (ABI) in order to improve auditory-verbal skills in such patients [8–10]. Yet, some authors claimed satisfactory performances after cochlear implantation in CND and suggested that the absence of an identifiable cochlear nerve on imaging does not preclude auditory innervation of the cochlea [11,12]. In this study we report our experience in cochlear implantation of children with CND focusing on radiologic findings and postoperative rehabilitative outcomes.

2. Materials and methods

The Parma Cochlear Implant Program database was used to search for children with CND who underwent cochlear implantation. Patient charts were reviewed for medical history, audiometric results, radiologic findings, and pre- and post-implantation performance. The study protocol was approved by the institutional review board; an informed written consent was obtained from all patients. Preoperatively, all children underwent high resolution computed tomography (HRCT) using a standard temporal bone protocol (Somatom Sensation 64 slice, Forchheim, Germany; collimation 0.6 mm; 120 kV; variable mAs with current modulation; care dose). A brain MRI study was subsequently performed on a 1.5 Tesla system (Philips Achieva, Best the Netherlands) including in the scan protocol a 3D T2-weighted DRIVen Equilibrium (DRIVE) sequence (slice thickness 0.8 mm) and parasagittal reconstruction to evaluate the course of the eighth cranial nerve within the IAC. CND was classified using Govaerts system (Table 1) [7]. The cochlear nerve was defined as hypoplastic when the diameter of the midportion of the IAC was less than that of the facial nerve or was defined as absent when it could not be identified on the reconstructed parasagittal oblique plan.

Audiological assessment included transient-evoked otoacoustic emissions, auditory brainstem responses (ABR) and acoustic imittance. Aided hearing thresholds were evaluated using visual reinforcement audiometry testing. Speech and language assessment was performed by an experienced pediatric speech therapist. After cochlear implantation, children were evaluated using age-appropriate speech perception tests. An Italian equivalent of the early speech perception (ESP) test [13] and of the Glendonald Auditory Speech Perception (GASP) test [14] for words and sentences was used to measure pre- and post-operative speech perception. The speech materials were presented in a hearing-only condition using a monitored live-voice through the sound field at an intensity level of 70 dB sound pressure level in quiet. On the basis of the speech perception test results, children were assigned to one of the six classes of performance as described by Geers and Moog [15]. Speech perception results were evaluated preoperatively (with hearing aids), at 1, 3, 6, 9 and 12 months after implantation, and then at yearly intervals. The Speech Intelligibility Rating (SIR) scale [16] was used to assess the intelligibility of the speech produced by these children. Warble tone average threshold with cochlear implant in the sound field was used to evaluate hearing levels after surgery. For the present study, results were collected at 6-, 12-months and at the last follow-up post-implantation interval. The follow-up period ranged between 18 and 81 months (average, 45 months).

3. Results

Five children, 3 boys and 2 girls, who had been diagnosed with CND before surgery and underwent cochlear implantation in the Cochlear Implant Center of the University of Parma, were identified. Etiology of SNHL was variable. Syndromes diagnosed included Noonan, Dandy-Walker and Rubinstein-Taibi; the remaining two children had no known etiology of their SNHL. The relevant preoperative patient data are given in Table 2.

According to Govaerts classification [7], three children had a type IIb and two a type IIa malformation in the implanted ear (Table 3). The IAC had a normal diameter on both sides in two children (cases 1 and 3), and was bilaterally stenotic (diameter <2 mm) in other two patients (cases 4 and 5); in the remaining child (case 2) the IAC was normal in one ear and stenotic on the contralateral side. Cochlear nerve was judged hypolastic on both sides in two children (cases 1 and 4), bilaterally absent in other two patients (cases 2 and 5), hypolastic in one ear and absent on the controlateral side in the remaining child (case 3). The cochlea was considered normal on both sides in two patients (cases 1 and 3), while in 2 other cases (4 and 5) there were bilateral abnormalities; in the remaining case, the cochlea was normal on the implanted side and malformed on the controlateral ear. MRI findings for all 5 patients are displayed in Figs. 1–5. Despite unambiguous responses to sounds with hearing aids, all children failed to make adequate progresses in terms of auditory performance and speech development during a trial period of amplification with intensive auditory training. The results of preoperative audiological assessment are reported in Table 4. Preoperatively, four patients were

Table 2
Clinical features of the patients.

Patient	Sex	Etiology	Neurocognitive development	Age at amplification	Age at surgery	Implanted ear	Cochlear implant device
1	F	Noonan syndrome	Normal	6	24	Right	Nucleus 24 RE
2	M	Dandy-Walker syndrome	Normal	22	36	Left	MXM Digisonic
3	M	Unknown	Normal	18	23	Left	Nucleus 24 RE
4	F	Rubinstein-Taibi syndrome	Delayed	20	30	Left	MXM Digisonic
5	M	Unknown	Normal	6	18	Right	Nucleus 24 RE