Otology & Neurotology
24:887–891 © 2003, Otology & Neurotology, Inc.

Reprinted from OTOLOGY & NEUROTOLOGY
Vol. 24, No. 6 November 2003

Cochlear Implants in Aplasia and Hypoplasia of the Cochleovestibular Nerve

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Objective: To report on the outcome of four patients with aplasia or hypoplasia of the cochleovestibular nerve who have received a cochlear implant.
Study Design: Retrospective case review.
Setting: Tertiary referral center.
Patients: Four patients with: 1) type I aplasia; 2) type IIa aplasia; 3) type IIa hypoplasia; and 4) type IIb aplasia received a cochlear implant. All patients had corner audiograms even with hearing aids.
Intervention: Three patients received a LAURA multichannel implant, and one patient received a Nucleus 24 implant.
Main Outcome Measure: Auditory performance, educational setting.
Results: The patients with type I and type IIb aplasia did not have auditive perception with their implant and became nonusers. Both are now in a total communication educational setting. The patients with type IIa aplasia and hypoplasia had moderate audiological results with the implant with audiometrical thresholds of approximately 40–60 dB HL (pure tone average), moderate phoneme discrimination, and poor word discrimination. One child is in a total communication educational setting and the other in an oral educational setting, but the preferred mode of communication remains total communication for both. Both appear to benefit from the implant nonetheless.
Conclusion: Patients with aplasia/hypoplasia of the cochleovestibular nerve should be counseled with caution with respect to cochlear implantation, but particular circumstances may justify the intervention. At present these circumstances seem to be a type IIa aplasia or hypoplasia in which the end organ (cochlea or common cavity) still connects to a neural structure on MRI. **Key Words:** Aplasia—Hypoplasia—Cochlear implant—Sensorineural hearing loss.
Otol Neurotol **24**:887–891, 2003.

Congenital bilateral sensorineural hearing loss (>30 dB HL) occurs in approximately 1.2 to 3.2 per 1000 live births (1–3). One of the causes is an aplasia or hypoplasia of the cochleovestibular nerve (CVN). This radiologic entity was first described in 1997 (4). It was shown that MR images (axial [0.7 mm] three-dimensional Fourier transformation-constructive interference in steady state [3DFT-CISS] images) with parasagittal reconstruction images perpendicular on the course of the VCN allowed identification of the facial nerve and of the VCN with its cochlear, inferior, and superior vestibular branches. A classification was suggested based on the affected branch of the nerve and the related labyrinthine dysplasia (Table 1, Fig. 1). It was anticipated that the cochlear branch would always be involved in this anomaly, and to date, no isolated hypoplasia or aplasia of the vestibular nerve, in the presence of a normal cochlear nerve, has already been described. Previous reports had already mentioned the existence of a narrow internal auditory meatus on CT-scan, from which a hypoplastic cochleovestibular nerve was inferred and the results of cochlear implantation in these cases were poor (5–6). In consequence, one would expect that aplasia and hypoplasia of the cochlear nerve would be a contraindication for cochlear implantation. Other reports however have claimed good results after implantation. In most of these cases, the diagnosis of aplasia or hypoplasia of the CVN was made after the implantation. These reports justify a more detailed analysis of the results of cochlear implantation in the different types of aplasia/hypoplasia of the CVN. In our series of 17 cases of aplasia/hypoplasia of the CVN, four received a cochlear implant, and they represent four different types of aplasia/hypoplasia. Three patients had an aplasia (type I, IIa, and IIb) and one a hypoplasia (type IIb) of the CVN. The results will be discussed.

PATIENTS AND METHODS

A retrospective case study was carried out on four patients that are known in our department with aplasia/hypoplasia of the

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TABLE 1. *Classification of hypoplasia and aplasia of the CVN*

Type	Affected nerve on imaging	Remarks
I	Cochleovestibular nerve	The labyrinth may be normal or dysplastic, the internal auditory canal is stenotic
IIa	Cochlear branch with labyrinth dysplasia	Labyrinth dysplasia ranges from a minor dysplasia, like in case 3, to a common cavity
IIb	Cochlear branch with normal labyrinth	
III ?	Vestibular branch	Was anticipated not to exist as an isolated aplasia and has not been reported so far

cochleovestibular nerve and that have been implanted with a cochlear implant. No other patients of the seventeen patients with aplasia/hypoplasia of the CVN that have been diagnosed to date by the authors have been implanted.

The diagnosis of the deafness was made with routine techniques of clinical audiology, such as pure tone audiometry with and without hearing aids, auditory brainstem responses (ABR), and transient otoacoustic emissions. In addition, before surgery, all children underwent an electrical trial stimulation, either with subjective responses or with brainstem evoked responses. For this, an active electrode was placed in the outer ear canal, which was filled with physiological fluid, or at the round window. The reference electrode was placed on the skin of the mastoid. Biphasic pulses (150 seconds per phase) of varying intensities (200–1400 A) and repetition rates (30–200 Hz) were given as stimulus and the responses were registered.

The diagnosis of aplasia/hypoplasia of the CVN was made on MR imaging with axial (0.7 mm) three-dimensional Fourier transformation-constructive interference in steady state (3DFT-CISS) images and parasagittal reconstruction images perpendicular on the course of the VCN (4). Only for the first case, no parasagittal reconstructions were made, because this was a later development.

A LAURA multichannel cochlear implant (Antwerp Bionic Systems, Antwerp, Belgium) was given to the first three cases, and a Nucleus 24M multichannel cochlear implant (Cochlear Ltd., Sydney, Australia) was given to the last case and for reimplantation.

Postoperative audiograms are given, as well as the results on the phoneme discrimination test, which is part of the A§E (Auditory Speech Sound Evaluation; PJ Govaerts, Antwerp-Deurne, Belgium). This test is an essential part of the routine test battery that is used by our department for the selection and evaluation of cochlear implantees.

RESULTS

Case 1: Aplasia type IIa

A male patient was born after an uncomplicated pregnancy. At the age of 13 months he was referred for hearing evaluation because of parental concern. Otoscopy was normal. Conditioned orientation audiometry did not show any hearing thresholds. Transient evoked otoacoustic emissions were bilaterally absent. Auditory Brainstem Responses (ABR) were absent at 120 dB click stimulation. No mutations were found in the connexin-26 gene. Medical imaging (CT scan and axial 3DFT-CISS images) revealed a cochleovestibular dysplasia with a common cochleovestibular cavity with dysplastic semicircular canals. At that moment, no anomalies were diagnosed at the level of the CVN. Only later, after thorough re-evaluation of the images, the CVN that was present in the inner ear canal did not appear to divide into separate cochlear and vestibular branches. The common nerve made contact with the common cavity. At the age of 15 months the boy received hearing aids and was enrolled in an early rehabilitation program. The auditory rehabilitation was unsuccessful. Aided thresholds were high (70–120 dB) at the low frequencies (125, 250, and 500 Hz) and absent at the frequencies above 500 Hz. A trial with a multichannel vibrotactile stimulator (Tactaid; Audiological Engineering Corp., Sommersville, MA, U.S.A.) was not tolerated by the patient. Electrical stimulation with an outer ear canal electrode could elicit responses at both ears. After thorough counseling the de-

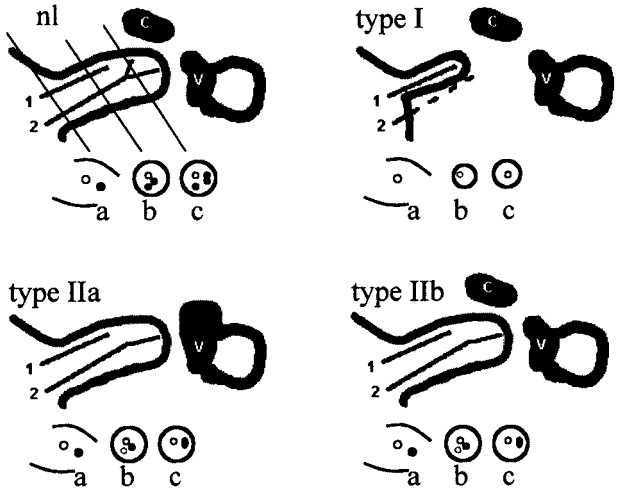

FIG. 1. Schematic representation of the MR images of the normal situation (nl), and the different types of hypoplasia/aplasia of the cochleovestibular nerve (type I, IIa and IIb). Each figures contains two schemes, the upper representing the axial image through the middle part of the left internal auditory canal; and the lower representing the parasagittal reconstructions perpendicular to the course of the nerves at three levels: (a) cerebellopontine angle, (b) middle third and (c) lateral third of the internal auditory canal. In the normal situation, the facial nerve (1 on the axial image, circle on the parasagittal image) and the CVN (2 on the axial image, dot on the parasagittal image a) are present in the inner auditory canal. The CVN contains a cochlear branch and a common vestibular branch. This can be seen at level (b) where in clockwise direction, the facial (circle), the common vestibular nerve and the cochlear branch (dots) are seen. At level (c), the common vestibular nerve has split into a superior and inferior branch. Type I is recognized by a stenotic internal auditory canal in which no CVN can be seen. The facial nerve runs in a separate canal and is seen at level (a) and (c). At level (b) the canal can be too narrow to allow visualization of the facial nerve. The labyrinth may be normal or dysplastic.

Type IIa is recognized by the absence of the cochlear branch at level (c) and sometimes at level (b) in the presence of a dysplastic labyrinth. Type IIb is similar to type IIa, but with a normal labyrinth.

cision was made to implant the left ear. At the age of 3 years, a LAURA multichannel cochlear implant (Antwerp Bionic Systems) was placed in the left ear. Only four electrodes could be inserted in the common cavity. Because of the cochleovestibular dysplasia, a monopolar configuration was selected. Postoperatively, electrical stimulation with the implant yielded good subjective perception with a flat audiogram at 65–70 dB HL 3 months after the operation. During the following years, the audiometric thresholds improved to 40 dB HL and the boy made good progress in terms of social behavior and speech development, although his main mode of communication remained sign and body language in a total communication educational setting. Four years after implantation he was able to discriminate the majority of a set of phoneme pairs that is routinely used by our department (Table 2). After 5 years, the implant had to be removed because of technical failure (electrical leakage) and was replaced with a Nucleus 24M multichannel implant (Cochlear Ltd.), of which 12 electrodes could be inserted. After 1 year, the audiogram showed thresholds of 45 dB HL and all phoneme pairs of Table 2 could be discriminated except /u/-/o/, /u/-/y/, and /y/-/I/. The word score on a closed set identification test (Erber 12) was 50 percent. The patient has been moved to an oral educational setting.

Case 2: Aplasia type I

A female patient was born from an insulin-dependent diabetic mother who had multiple episodes of vaginal bleeding during gestational weeks 8 to 12. Infancy was uneventful, and the patient entered the kindergarten at 2.5 years where she received speech therapy for delayed speech development. At the age of 4.5 years, a profound sensorineural hearing loss was diagnosed and the child was referred to a school for the deaf, where she received hearing aids. Oral education failed to improve the communication and at the age of 6 years the patient was moved to a total communication class. She was considered mentally retarded with autistic behavior. With total communication the behavior changed dramatically and the girl became a very good student with an open and communicative character. At the age of 10 years, she was referred for cochlear implantation. Otoscopic examination was negative at the left side and revealed a cholesteatoma the right side (which was surgically removed). Audiometry showed a corner audiogram. Aided thresholds showed no responses at 1000 Hz or higher frequencies, with elevated thresholds (80–110 dB HL) at the low frequencies (125, 250 and 500 Hz). Click evoked otoacoustic emissions were present at the left ear and absent at the right ear. Medical imaging (CT scan and axial 3DFT-CISS images) showed a type I aplasia of the CVN. Both internal auditory canals were stenotic and a CVN could not be visualized. Both labyrinths were normal and the facial and trigeminal nerves were present. Electrical stimulation with an outer ear canal electrode could elicit responses at the left ear. After thorough counseling, and in the absence of precedents in the literature, the decision was made to implant the left ear. At the age of 11 years, a LAURA multichannel cochlear implant (Antwerp Bionic Systems) was placed in the left ear. Postoperatively, electrical stimulation with the implant failed to elicit subjective perception. Electrically evoked brainstem responses could not be elicited. The patient discontinued wearing the speech processor. A vibrotactile aid (Tactaid; Audiological Engineering Corp.) was tried, but not tolerated, by the girl.

TABLE 2. *Case 1: results on the phoneme discrimination test**

Able to discriminate	Unable to discriminate
a-r	æ-a
u-ˢ	E-A
u-l	æ-E
I-a	v-z
u-a	
o-a	
u-o	
æ-u	
æ-o	
I-E	
æ-ie	
y-l	
u-y	
z-s	
m-f	
m-z	
m-r	
s-ˢ	

*The phoneme discrimination test is part of the A§E (Auditory speech sound evaluation). The first phoneme of a pair is presented as the background phoneme and the second as the odd phoneme. The phonemes are presented at 70 dB HL (re 1kHz narrow band noise).

Case 3: Hypoplasia type IIa

A male patient was born at 31 weeks gestational age due to a premature dehiscence of the placenta. At birth it was noted that the auricles were small and displaced inferiorly with supernumerary ear tags at the right side. No other deformities were withheld. The child did not pass the neonatal screening with transient otoacoustic emissions and was referred at the age of 2 months. ABR with air-conducted clicks of 120 dB HL did not show responses, but with bone-conducted clicks, responses were recorded at 50 dB HL. Medical imaging (CT scan and axial 3DFT-CISS images) showed a stenotic outer ear canal at the right side with major ossicular malformations at both ears. Both cochleas had a normal aspect, the vestibula were slightly dysplastic, and the semicircular canals were normal. At both sides the CVN could be visualized and the vestibular branch appeared to be normal but the cochlear branch was very hypoplastic. No connexin-26 gene mutations were found. The boy received hearing aids at the age of 5 months and was enrolled in an early rehabilitation program. The auditory rehabilitation was unsuccessful, and based on the working hypothesis of a mixed hearing loss, a bone anchored hearing aid (BAHA) was given at the age of 3 years, after a positive 3-month trial with a conventional bone con-

TABLE 3. *Case 3: results on the phoneme discrimination test**

Able to discriminate	Unable to discriminate
a-r	z-s
u-l	m-z
l-a	
u-a	
s-^{s}	

Only the minimal set of the A§E is used for this patient.

ducting hearing aid. Soon after the placement of the BAHA, the bone-conduction thresholds were reported to go down and the boy did not seem to benefit form the device. Electrical stimulation with an outer ear canal electrode could elicit responses at both ears, and based on these results, a cochlear implant was given at the age of almost 4 years. A LAURA multichannel cochlear implant (Antwerp Bionic Systems) was placed in the left ear. Postoperatively, electrical stimulation with the implant yielded good subjective perception with thresholds of 65–70 dB HL at 500–4000 Hz 3 months after the operation. Electrically evoked brainstem responses could be elicited at the 5 most basal channels. The boy made moderate progress in terms of audiological performance and speech development, and his main mode of communication remained sign and body language in a total communication educational setting. Three years after the implantation, the LAURA device (Antwerp Bionic Systems) had to be removed because of technical failure (electrical leakage) and was replaced with a Nucleus 24 multichannel cochlear implant (Cochlear Ltd.). Six months later, audiometry showed thresholds of 65 dB HL at the low frequencies and 45 dB at the frequencies 1000–4000 Hz, with discrimination of some phonemes (Table 3).

Case 4: Aplasia type IIb

A female patient was born with the Goldenhar syndrome (oculoauriculovertebral dysplasia) after an uncomplicated pregnancy. ABR did not show any responses to air conducted clicks of 120 dB HL. Medical imaging (CT scan and axial 3DFT-CISS images) revealed a normal labyrinth (cochlea, vestibulum, and semicircular canals) with a narrow internal ear canal in which a cochleovestibular nerve could be visualized with a vestibular branch but no cochlear branch. No mutations in the connexin-26 gene were found. The patient received hearing aids at the age of 9 months, but the aided thresholds were never better than 80–100 dB HL at 250 and 500 Hz without responses above 500 Hz. She was enrolled in an early intervention program with total communication. She was referred for cochlear implantation at the age of 19 months because of the poor audiological and speech developmental progress. Electrical promontory stimulation with ABR could elicit a reproducible peak at 3.8 ms, which was interpreted as an auditory brainstem response. Based on these results and after careful counseling, the girl received a Nucleus 24M multichannel cochlear implant (Cochlear Ltd.) at the age of 2 years, with full insertion of the electrode array. During the fitting sessions, no responses could be elicited and the girl did not appear to benefit from her implant. One year later, she discontinued wearing her implant and received a vibrotactile aid (Tactaid; Audiological Engineering Corp.) that appears to contribute to her level of communication and comfort of life.

DISCUSSION

After the first report on the MR diagnosis of aplasia and hypoplasia of the cochleovestibular nerve (4), many other authors have confirmed this diagnosis and have stressed the importance of systematic use of MR imaging in the workup of congenital profound sensorineural hearing loss (7–17). Recently, aplasia of the CVN was also histopathologically confirmed in two cases (probably type I and IIb) (18). In the first publication, a classification was proposed based on the affected branch of the nerve and the related labyrinthine dysplasia (Table 1). Since then, 17 cases with aplasia/hypoplasia of the CVN have been diagnosed in our department and their distribution is shown in Table 4.

All reported cases had profound sensorineural hearing loss and would meet the commonly used criteria for cochlear implantation. As could be expected, several papers report bad results after implantation (5–7), and some authors claim that aplasia/hypoplasia of the CVN is a formal contraindication for cochlear implantation (8,10). It has been suggested that these patients could benefit from a brainstem implant (17). Yet, other papers report good results with cochlear implantation (19,13,15).

In our series of 17 cases, 4 received a cochlear implant. Two patients appear to benefit from it and the other two are non-users. In the first case, the aplasia was overlooked and was only diagnosed after implantation. The next three cases represent different types of aplasia/hypoplasia and this justified the implantations. Obviously, the parents were carefully counseled and were aware of the potential lack of effect. An encouraging argument was the positive electrical stimulation that was performed in all four candidates before surgery. One test was even done under anesthesia with evoked brainstem responses and the result was interpreted positively. This was probably a misinterpretation of a myogenic response. Electrical stimulation has been abandoned in our department because of its poor prognostic value in general (20). A common feature of all four children was the corner audiogram, with little audiometrical improvement with hearing aids. This is obviously in line with the

TABLE 4. *Distribution of different types of aplasia and hypoplasia of the cochleovestibular nerve*

	Type I (%)	Type IIa (%)	Type IIb (%)
Bilateral	18	35	6
Unilateral	12	18	12

little benefit that was achieved with hearing aids. It is noteworthy that transient otoacoustic emissions were present in one patient (case 2). This was the case in only one ear, but the contralateral ear suffered from middle ear problems. The inner ears were normal on imaging.

Despite early detection and rehabilitation in three children, none developed speech or oral communication. The cochlear implant had no affect in two patients, one with a type I aplasia (case 2) and one with a type IIb aplasia (case 4). Both have discontinued wearing their implant. The second child appears to benefit from her vibrotactile device. A brainstem implant is being considered, especially for the younger of both (case 4). The other two patients do benefit from their implants, be it with moderate auditory outcome. The reason for this is probably that in both cases, the cochlear implant electrode is situated in the vicinity of an afferent nerve containing auditory fibers. In case 1 (type IIa aplasia), this is because the labyrinth is so dysplastic, that only a common cavity remains, from where the common cochleo-vestibular nerve runs to the brainstem. In case 3, this is because the cochlear nerve is hypoplastic, which means that even on MR imaging, neural tissue can be recognized at the place of the cochlear branch. Probably the remaining neural fibers suffice to provide reasonable detection and frequency discrimination of sound.

In conclusion, these data have led us to believe that aplasia/hypoplasia of the CVN is not a formal contraindication for cochlear implantation. Medical imaging may be of important prognostic value. If the local anatomy is such that the electrode cannot be located in the close vicinity of the nerve, it may not be justified to proceed with implantation. This is the case in type I and type IIb aplasia. If, in contrast, the electrode can be positioned in the close vicinity of the nerve, it may be worthwhile implanting the child. This is the case in type IIa hypoplasia or in a type IIa aplasia with major labyrinthine malformation, such as a common cavity. The authors recommend the routine use of this classification in case of hypoplasia or aplasia of the cochleovestibular nerve.

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