

# Auditory Brainstem Implantation After Unsuccessful Cochlear Implantation of Children With Clinical Diagnosis of Cochlear Nerve Deficiency

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**Objectives:** We compared the perceptual auditory abilities of 21 children with suspected cochlear nerve deficiency (CND) and a surgically verified absent cochlear nerve (CN) who first underwent cochlear implantation (CI) and subsequently underwent auditory brainstem implantation (ABI).

**Methods:** In this retrospective cohort study, from 2000 to 2011, 21 children initially underwent CI at an outside institution and failed to progress in their perceptual auditory abilities. Before CI, all of the children had severe to profound sensorineural hearing loss and a diagnosis of CND. Magnetic resonance imaging (MRI) documented an absent CN in 13 children and a small CN in 8 children. We performed explantation of the cochlear implant and simultaneous ABI on the same side. We performed MRI if no previous MRI results were available. All surgical videos were reviewed to determine the presence or absence of the CN. Measures of the patients' perceptual auditory abilities obtained after CI and after ABI were converted to the Category of Auditory Performance (CAP) scale.

**Results:** At surgery, all patients demonstrated an absent CN. After CI, all patients had a CAP score of 2 or less (mean,  $0.52 \pm 0.68$ ). After ABI, all patients had a CAP score of 2 or more (mean,  $4.33 \pm 1.68$ ); the improvement was statistically significant ( $p < 0.001$ ). The complication rates were similar for CI and ABI.

**Conclusions:** In this cohort of patients who had poor performance after CI, ABI achieved significantly improved performance as measured by the CAP and was shown to successfully rehabilitate hearing. Cases of a small CN may in reality represent an absent CN. Although this cohort was selected from patients with failed CI, the results have implications for the selection of device for patients with CND, in that ABI is a potential alternative to CI in select cases. In patients who fail to progress with intensive rehabilitation with CI or who have no progression in evoked auditory brainstem response, ABI must be considered early.

**Key Words:** auditory brainstem implant, cochlear implant, cochlear nerve absence, cochlear nerve deficiency, inner ear malformation.

## INTRODUCTION

In patients with cochlear nerve deficiency (CND), the cochlear nerve (CN) is either absent or small (small cochlear nerve, SCN). Typically it is identified on high-resolution imaging during evaluation of sensorineural hearing loss, in association with inner ear malformations and narrow internal auditory canals (IACs). On occasion, CND may be also observed in children with normal inner ear morphology. In these children, electrophysiological investigations may suggest the diagnosis of auditory neuropathy spectrum disorder, and cochlear implantation (CI) may be indicated for hearing restoration. With CND, electrical stimulation of the cochlea by the implant may not be able to reach the cochlear nucleus complex and effectively activate the ascending auditory system. Most reports of CI in children

with CND have shown poor results. A few reports have documented limited benefit in some patients.<sup>1,2</sup> Those children with CND who achieve limited auditory benefit from CI often do so with high charge density stimulation and with wide pulse widths.<sup>2</sup> This uncertainty is compounded by the inability of the routine preoperative radiologic and electrophysiological evaluations to provide definitive information regarding the presence or absence of a functioning CN.

In a previous report,<sup>3</sup> we described the performance achieved with auditory brainstem implantation (ABI) in 5 patients who had been previously treated unsuccessfully with CI. This group of patients included 3 adults with complete cochlear ossification, 1 child with bilateral absent CN, and 1 child with auditory neuropathy spectrum disorder.

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TABLE 1. DEMOGRAPHIC INFORMATION, ASSOCIATED DISORDERS, AND CAP SCORES WITH CI AND ABI IN PATIENTS WITH PRIMARY DIAGNOSIS OF COCHLEAR NERVE DEFICIENCY

Pt	Degree of Cochlear Malformation	Sex	Ear	Other Disorders	Age at CI (y)	Interval From CI to ABI (y)	Age at ABI (y)	CAP Score Before ABI	ABI CAP Score at Last Follow-Up	ABI Follow-Up (y)	CAP Score by Year of ABI Follow-Up								
											1 y	2 y	3 y	4 y	5 y	6 y	7 y	8 y	
11	None	F	R		0.99	2.01	3	1	5	2	2	5							
12	None	M	L		1.6	1.4	3	1	6	2	4	6							
1	Severe	M	R		0.91	0.79	1.7	0	4	3	1	3	4						
2	None	F	R		1	0.91	1.91	0	7	3	3	5	7						
17	Moderate	M	R		2.1	1.9	4	1	5	3	1	3	5						
7	None	F	R		1.58	1.22	2.8	1	6	4	3	4	6	6					
15	Severe	M	L	Moebius syndrome	1.5	1.8	3.3	0	4	4	2	2	3	4					
19	Severe	M	R	Cognitive	2.3	2.2	4.5	0	3	4	2	3	3	3					
3	Severe	M	R	Cognitive + SLI (DVD)	1.42	0.68	2.1	0	3	5	1	2	3	3	3				
20	Severe	F	R	Down syndrome	1.6	3.4	5	0	3	5	2	3	3	3	3				
5	None	M	L		1.08	1.32	2.4	2	7	6	3	4	7	7	7	7			
6	Moderate	M	L	Cognitive + motor	1.16	1.54	2.7	0	4	6	2	2	2	2	4	4			
8	None	F	L		1.75	1.15	2.9	1	6	6	4	5	6	6	6	6			
13	Severe	M	R		1.08	1.92	3	1	4	6	2	3	3	4	4	4			
18	Severe	M	R	Cognitive	1.66	2.44	4.1	0	2	6	0	0	0	1	2	2			
21	Severe	F	L	Cognitive + visual	2.33	7.67	10	0	2	6	1	2	2	2	2	2			
9	Severe	F	L	Cognitive + motor	1.91	1.09	3	0	3	7	1	1	2	3	3	3	3		
4	None	M	L		1	1.2	2.2	2	7	8	3	5	7	7	7	7	7		
10	Severe	F	L	Shprintzen syndrome	1.2	1.8	3	0	2	8	0	1	1	2	2	2	2	2	
14	Moderate	F	R		2	1	3	1	5	8	2	3	4	5	5	5	5		
16	Severe	M	L	Cognitive	1.75	2.25	4	0	3	8	2	2	2	3	3	3	3	3	

CAP — Categories of Auditory Performance; CI — cochlear implantation; ABI — auditory brainstem implantation; SLI — specific language impairment; DVD — developmental verbal dyspraxia.

Other centers have also reported on successful ABI after CI.<sup>4</sup> Auditory brainstem implantation has been proposed as first-line therapy in this category of deaf patients currently treated with CI.<sup>5-7</sup>

The present investigation describes the outcomes of a large group of children with CND who sequentially underwent CI and, after explantation, ABI. We describe the differences in eighth nerve anatomy between the radiologic studies and the findings at surgery, and we review the outcomes after both CI and ABI.

#### METHODS

The Ethical Institutional Review Board of the University of Verona Hospital and Medical School approved this study. A consecutive group of 21 children with a clinical diagnosis of CND initially underwent CI at various outside institutions. After the children failed to progress in perceptual auditory abilities, their cochlear implants were explanted and

they underwent ABI on the same side at our institution or elsewhere by the senior author (V.C.). All children had severe to profound hearing loss, and all had their cochlear implants removed after 0.68 months to 7.67 years (mean,  $1.98 \pm 1.55$  years), either at their own institution or at our department. A retrosigmoid surgical approach was used in all children, all were fitted with either a Cochlear 24 ABI device (Cochlear Ltd, Sydney, Australia) or a Med-El ABI device (Med-El, Innsbruck, Austria), and all followed the same rehabilitation program.

There were 12 male and 9 female patients. The mean ages at surgery were  $1.52 \pm 0.44$  years for CI and  $3.41 \pm 1.73$  years for ABI. The mean follow-ups were  $1.89 \pm 1.47$  years for CI and  $5.24 \pm 1.97$  years for ABI. Demographic information for the patients is detailed in Table 1.

*Evaluation of Auditory Perceptual Abilities.* Auditory perception was assessed with the Categories of Auditory Performance (CAP) test<sup>8,9</sup> in children

TABLE 2. CATEGORIES OF AUDITORY PERFORMANCE<sup>8,9</sup>

7	Use of telephone — known speaker
6	Understand conversation, no lip-reading
5	Understand common phrases, no lip-reading
4	Discrimination of speech sounds
3	Identification of environmental sounds
2	Response to speech sounds (e.g. "go")
1	Awareness of environmental sounds
0	No awareness of environmental sounds

followed at our institution (Table 2). The CAP was used because of the different languages of the patients undergoing surgery, and because of its proven interobserver reliability. The Early Speech Perception Test (ESP) and the Glendonald Auditory Screening Procedure (GASP) were used to evaluate the auditory performance of the 9 children followed in other institutions, outside Italy. The outcomes of these children were pooled together with the outcomes of the other 12 children by converting the ESP and GASP scores into equivalent CAP scores according to performance level. For example, discrimination of words on the ESP was assumed to be equivalent to category 4 on the CAP.

*Preoperative Imaging.* Inner ear and CN abnormalities were diagnosed before CI on the basis of the results of high-resolution magnetic resonance imaging (MRI) and computed tomography (CT).

The definition of an absent CN or a small CN was obtained with MRI by evaluating the CN in the IAC in the oblique sagittal plane. An absent CN was defined as a nondetectable nerve, and a small CN was defined as a nerve diameter smaller than that of the adjacent facial nerve. The CN was considered normal when its diameter was twice that of the facial nerve.<sup>10-14</sup>

Cochlear abnormalities were categorized as moderate (Mondini malformation), severe (presence of only basal turn or common cavity), or absent (Michel or cochlear aplasia). The patency of the bony CN canal was measured by either CT or MRI and was classified as normal if the canal was at least 1.4 mm in diameter and as reduced if it was less than 1.4 mm in diameter.<sup>11,12</sup> The IAC was measured on the axial view of the temporal bone CT scan and was defined as normal if the diameter was at least 3 mm and as reduced if the diameter was less than 3 mm. Abnormalities of the vestibule were characterized as mild (involving only the lateral semicircular canal and vestibule), moderate (involving 2 semicircular canals), severe (involving all of the canals), or an absence of the vestibule.

## RESULTS

*Preoperative Imaging.* In 14 children, the MRI

findings made before CI were available for review. In the remaining 7 children, MRI was performed after cochlear implant removal, just before ABI. The MRI documented bilateral absent CNs in 13 children and small CNs in 8 children. Both the seventh and eighth cranial nerves were absent on the side contralateral to the operated ear in 2 children. The seventh cranial nerve had an aberrant course in 5 cases (2 bilateral and 3 unilateral).

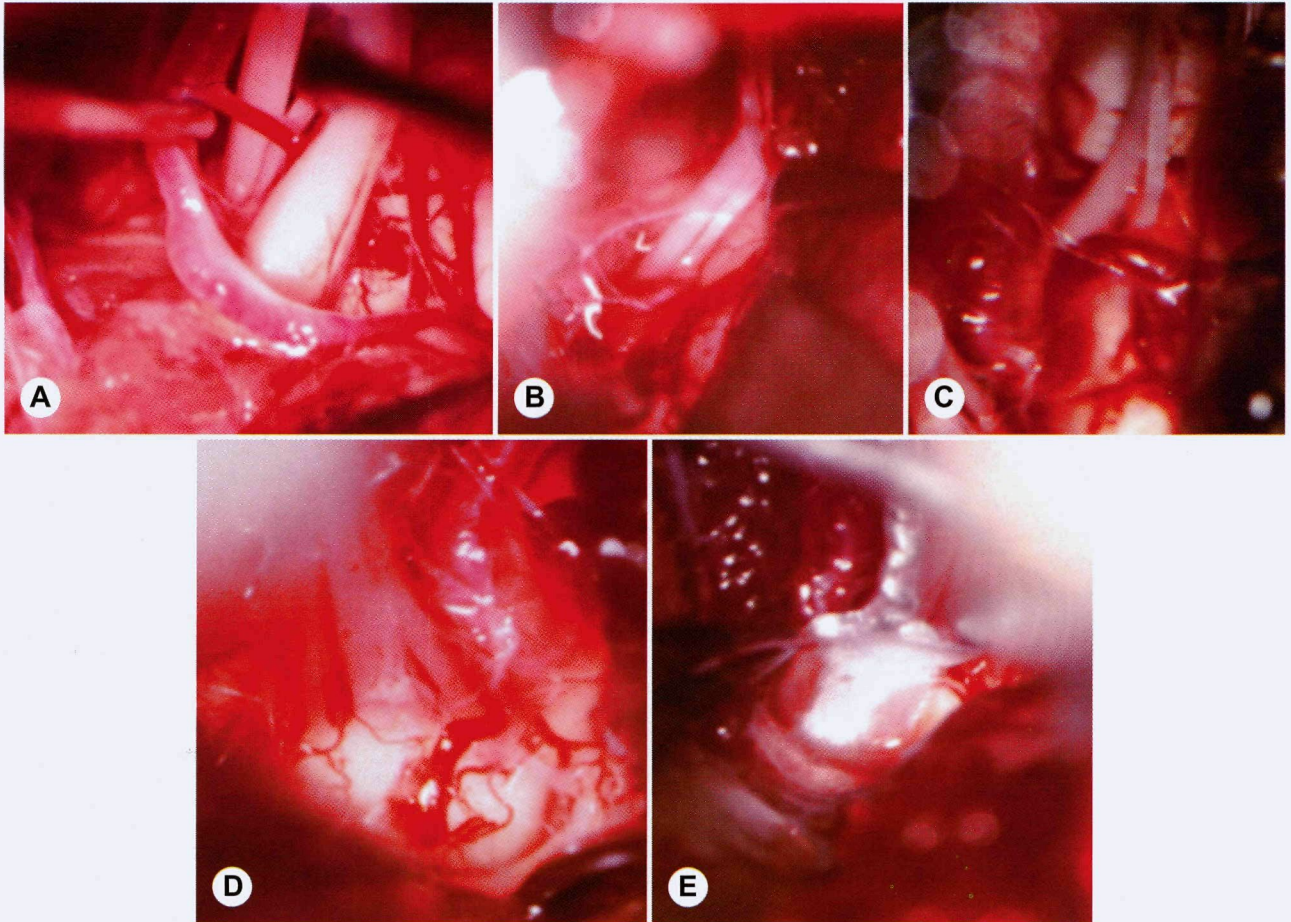
In the cisternal portion of the cerebellopontine angle (CPA), 13 patients had only 1 single identifiable nerve (seventh cranial nerve) and 8 patients had 2 identifiable nerves defined as the seventh nerve and a small CN. In the IAC, the seventh nerves were clearly identified in all but the 5 patients with IAC atresia who presented an aberrant course of the nerves. The so-called small CNs were difficult to follow in the IAC because of a significantly reduced diameter of the IAC in the remaining patients. Interestingly, among the 13 children with absent CNs, 5 were found to have a normal-size IAC, and there was an open bony CN canal in 4 of the 5.

Fourteen children had cochlear abnormalities of different degrees: moderate in 3 and severe in 11. Six had normal cochlear morphology on both CT and MRI. The IAC was normal in 6, was atretic in 5, and had a reduced diameter of 1.5 to 2.3 mm in the remaining 10 children. Eight children had vestibular abnormalities: mild in 2 and severe in 6. Severe vestibular malformations were associated with moderate or severe abnormalities of the cochlea. No child in the present series had evidence of cochlear ossification. The bony CN canal was normal in 5 patients, small in 3 patients, and absent in 13 patients.

*Identification of CN in CPA.* To confirm whether the CN was present or absent in the cisternal part of the CPA, and to define its size and point of origin from the brainstem, we looked for the proximal portion of the CN, which normally entered the acoustic recess and coalesced with the surface of the pons.

In the present series of children with CND, the CN was radiologically defined as absent in 13 children and small in 8 children. However, at surgery, no eighth nerve could be visualized emerging from or close to the cochlear nucleus complex (Fig 1B-D). At high magnification, the very thin nerves observed at surgery close to the seventh nerve, described in the MRI reports as a small CN, were indeed seen to be the nervus intermedius. Normally (Fig 1A), both the seventh nerve and the nervus intermedius emerge from the brainstem slightly above and anterior to the eighth nerve. An example of an inserted auditory brainstem implant is shown in Fig 1E.

*Aberrant Course of Facial Nerve in CPA and*



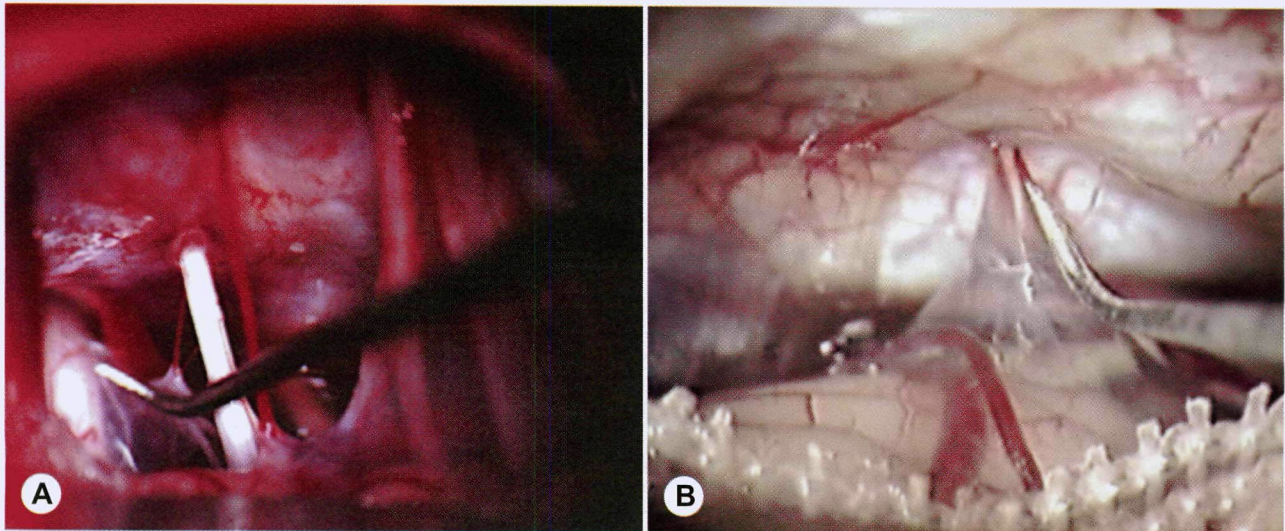
**Fig 1.** Intraoperative photographs. **A)** Cerebellopontine angle (CPA) view of normal seventh and eighth cranial nerve complex with anterior inferior cerebellar artery (AICA) and communicating artery running from AICA to brainstem, between nervus intermedius and eighth nerve. **B,C)** CPA views of patients with eighth nerve aplasia. Seventh nerve and nervus intermedius may be seen at their entry zone. AICAs of both patients (bottom of figures) have been retracted to facilitate view of entry zone of these nerves and of foramen of Luschka. On left side of both images, lower cranial nerves, including ninth nerve, may be seen. **D,E)** Following direction of ninth nerve, foramen of Luschka is identified (**D**) and auditory brainstem implant is inserted (**E**).

IAC. In the present series of children, the seventh nerve was found entering an IAC of a normal size in 6 children and entering an IAC of a reduced diameter of 1.5 to 2.3 mm in 10 children. An aberrant course of the intracranial seventh nerve was identified in 5 cases: 2 bilateral and 3 unilateral. Two of the unilaterally affected patients had seventh nerve palsy. All 5 had complete atresia of the IAC (Fig 2), and the seventh nerve was dislocated in its cisternal segment anteriorly and inferiorly. In the 2 ears with seventh nerve palsy, the cisternal pathway of the seventh nerve could not be identified on radiology. The facial nerve fibers at the level of the Gasserian ganglion took a sharp lateral turn and entered a minute separate canal to reach the geniculate ganglion (Fig 2A).

*Perceptual Auditory Abilities.* The CAP scores were obtained with the cochlear implant immediately before explantation, and the CAP score was 0 in 12 children. In 4 patients, most of the electrodes

were deactivated progressively because of facial nerve stimulation. The CAP data are summarized by patient in Table 1 and are plotted in Fig 3 and grouped by final CAP score in Fig 4. After CI, the children had a CAP level of 2 or less ( $0.52 \pm 0.68$ ), whereas after ABI all children had a CAP level of 2 or greater ( $4.33 \pm 1.68$ ); the difference between the two sets of scores was statistically significant ( $p < 0.001$ ). The development of speech perception did not occur in any of the 21 children after CI, but did occur in 13 children after ABI. According to parental reports, after ABI all children gained improved access to environmental and speech sounds. Open-set speech recognition was observed in 41% of the children after ABI.

The children with normal cochleas and either an absent CN or a small CN demonstrated a significantly earlier and better perceptual outcome on the CAP test than did children with cochlear abnormalities; all children with normal cochleas had a CAP



**Fig 2.** CPA views in patients with eighth nerve aplasia, internal auditory canal (IAC) atresia, and severely stenotic IAC. **A)** Fifth nerve (designated by instrument) and seventh nerve may be seen. Seventh nerve is anteriorly dislocated and enters minute bony canal between fifth nerve and atretic IAC. IAC is identified by exit of small blood vessel running posteriorly from atretic meatus parallel to seventh nerve. Eighth nerve is absent. At extreme right, lower cranial nerves are visualized. **B)** Close-up view of severely stenotic porus and IAC. Hypoplastic seventh nerve and small vessel are entering porus. Eighth nerve and nervus intermedius are absent.

score of more than 5 at 4 to 6 years after ABI.

Figure 5 shows the best CAP scores measured over time according to whether the children underwent ABI before or after 3 years of chronological age. Both age groups had CI CAP levels of either 2 or below. At 3 years after ABI, the gap in CAP scores between the two groups is one of more than 3 full categories.

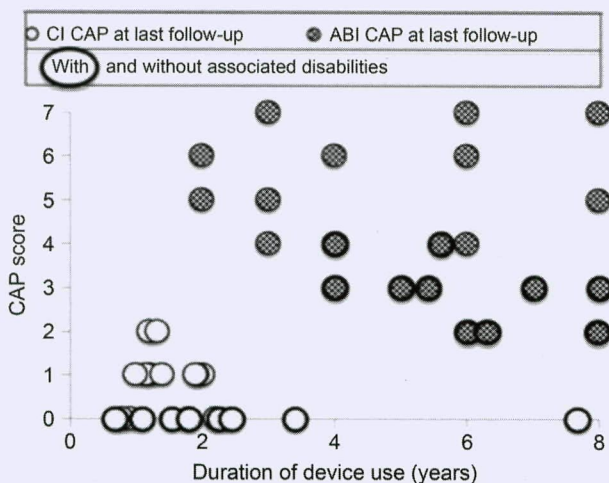
Figure 6 shows the ABI CAP scores over time in the 21 children according to whether they had CNDe only or CNDe with associated disabilities. Children without associated disabilities showed better audi-

tory performance after ABI than did children with associated disabilities at all follow-up intervals; this finding suggests that associated disabilities limit the potential benefit of ABI. Although children with special needs showed lower levels of auditory performance, they still showed improved awareness of their environment and benefits in cognitive development.<sup>15</sup> There were no differences in complication rates between CI and ABI surgeries.

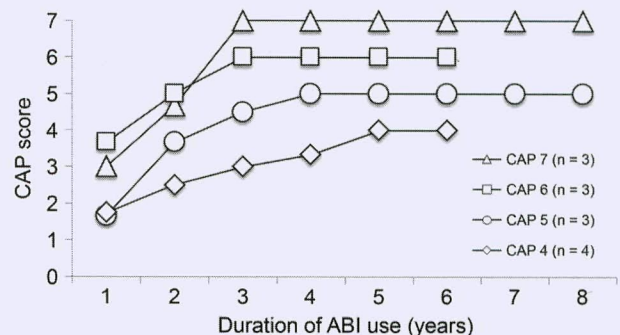
DISCUSSION

Appropriate management of children with clinically diagnosed CNDe has not been fully standardized. Previous studies regarding the outcomes of CI for children with auditory neuropathy spectrum disorder and CNDe have provided conflicting results.<sup>16-19</sup> However, since the time of those studies, auditory neuropathy spectrum disorder has become an acceptable indication for CI.

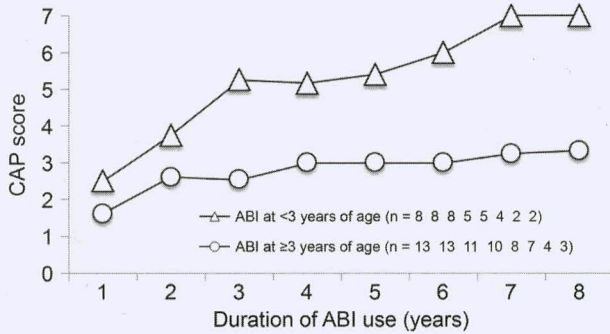
Cochlear nerve deficiency may be seen in up to



**Fig 3.** Categories of Auditory Performance (CAP) scores over time in 21 children with cochlear nerve deficiency with and without associated disabilities who underwent cochlear implantation (CI) and auditory brainstem implantation (ABI).



**Fig 4.** Evolution to final CAP score over time in 21 patients, grouped by final CAP score.



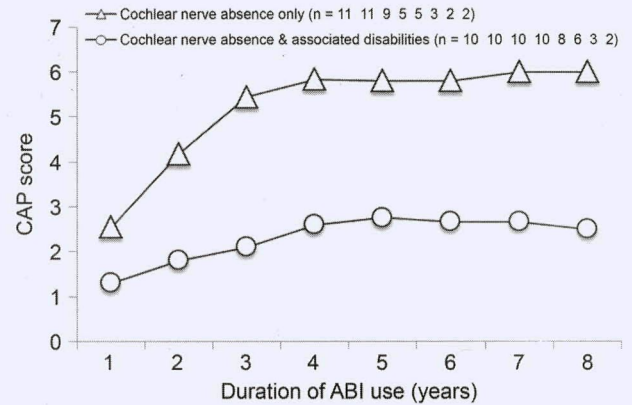
**Fig 5.** CAP score progress in patients who underwent ABI at less than and more than 3 years of age.

18% of patients with sensorineural hearing loss. In CND, the electrical activity elicited within the cochlea may fail to reach the cochlear nuclei and effectively activate the ascending auditory system, resulting in poor CI outcomes in many patients.<sup>17,18</sup> Nevertheless, at many centers CI continues to be offered as first-line treatment for patients with CND. These centers hold that an absence of the CN observed on imaging studies does not preclude the possibility that some CN fibers may indeed be present but may not be identifiable because of current imaging resolution or the presence of CN fibers traveling with the facial or vestibular nerves and not as a distinct bundle. Considering the plasticity of the auditory cortex in young children, it is believed that even activation of a limited number of CN fibers has the potential to provide benefit to these children.<sup>19-23</sup>

**Results of CI.** Although our study has a selection bias in that the present cohort had failed CI, important points can be learned from the CI experience of the children in this cohort. It is somewhat concerning that even after verification that the benefit from CI was limited to auditory awareness and behavioral responses induced at very high levels of charge units (often with associated nonauditory stimulation), ABI was delayed, sometimes for an extended period.

Several children with CI in this series had CAP 1 or CAP 2 levels of performance. It is often questioned how patients with a small or absent CN receive any auditory percept if the CN fiber population is small. It has been proposed that vestibular or somatosensory perception may be playing a role in these patients' sound detection.<sup>2</sup>

Regrettably, CI in this group of children may have had a part in delaying the correct rehabilitation procedure and losing the opportunity and advantages of a critical period in auditory development. The type of CND (small or absent CN as seen on high-resolution imaging) has not been shown to cause a substantial difference in the development of significant levels of speech perception abilities, and all of the



**Fig 6.** CAP scores over time in patients with cochlear nerve deficiency, grouped by presence or absence of associated disabilities.

present children with CND with cochlear implants needed communication-mode and visual supplementation.

In the present series of children with CI, a year passed on average before the parents realized that the device was not providing the child with the desired acoustic stimulation and sought alternative modes of hearing rehabilitation. Additional time was spent in organizing the procedure of explanting the cochlear implant and implanting and activating the auditory brainstem implant. Considering that today most CI teams are fitting patients with congenital sensorineural hearing loss at earlier and earlier ages, a delay of several years is excessive. Thus, rapid assessment of auditory stimulation with a cochlear implant is a very important goal.<sup>13,21,22</sup> In 4 children, because of suspicion of device failure, the device was explanted and replaced with a new cochlear implant — in 1 child twice during the same session, and in the other 3 children after 8, 9, and 12 months. Subsequently, when it was recognized that there was not a device problem, ABI was considered.

We believe that if a child who has CND and no associated disabilities has been fitted with a cochlear implant, has followed rehabilitation sessions for more than a year, and has not developed any significant auditory ability, then ABI should be strongly considered without any further delay. Additionally, an evoked auditory brainstem response that is absent or inconsistent over time suggests that prolonged cochlear implant stimulation is not reaching the cochlear nuclei and is not inducing changes in the pattern of auditory brainstem activation.

**Results of ABI.** Despite the fact that ABI was performed in ears with previous CI, the ABI procedure was demonstrated to be relatively safe. In part, surgeries were performed on the ipsilateral ear in order to determine the status of the CN in relation to

the cochlear implant function previously obtained, as well as to perform MRI without the cochlear implant in place. Others<sup>4</sup> have performed ABI on the contralateral ear in cases in which ABI is used as salvage. The choice of whether to perform ABI on the ear ipsilateral to or contralateral to the cochlear implant's former location should be individualized and based on anatomic concerns, including the CN anatomy and the sigmoid sinus anatomy, and on the level of function of the cochlear implant. We anticipate that as further patients undergo ABI, more bilateral ABI/CI and ABI/ABI conditions will allow for further study of combined-modality treatment in children with CND.

Furthermore, despite the delay in ABI in our cohort, ranging from 0.68 to 7.67 years, the outcomes were statistically significantly better at the last follow-up than were the outcomes after CI. Among the children of the present cohort, 85.7% had clear responses to speech sounds and were able to identify environmental sounds, 61.9% demonstrated ability to discriminate some speech sounds without lipreading, 42.8% showed the ability to comprehend simple phrases without lipreading, 28.6% were able to converse with family members and classmates in the auditory mode, and 14.3% were able to use a telephone with known listeners.

In younger children without additional disabilities, progressive improvement of auditory performance was possible after ABI. Several children demonstrated improvement rates comparable to those reported for CI patients without CND.<sup>23</sup> Not surprisingly, concomitant disabilities were associated with limited development of auditory function after ABI, potentially correlated to delays in cognitive function rather than lack of auditory stimulation itself. These results are consistent with reports by several authors.<sup>4-7,24,25</sup>

Considering the older ages of the children of the present cohort at the time of ABI, compared to the very young age at which CI is currently performed,<sup>26-32</sup> we believe that ABI holds great promise for patients with narrow IACs and CND if it is performed early.

*Roles of Imaging and Electrophysiology in Determining Intervention.* The preoperative evaluation of a normal CN or an absent or small CN is currently dependent on imaging and electrophysiological studies. These are important, but are not diagnostic for the absence or presence of a CN. In fact, in 5 children in the study with an absent CN the IAC was of a normal size, and in another 4 children with an absent CN the bony CN canal was open. High-

resolution temporal MRI with a parasagittal view on the IAC<sup>5,10,13</sup> may identify the number of nerves and clarify the CN's integrity. However, if the IAC is small, the MRI resolution may prevent precise differentiation between an absent CN and a small CN.

In the present study, MRI of the cisternal portion of the CPA clearly showed that 13 children had only 1 single identifiable nerve: the seventh nerve. However, in the 8 children who had 2 identifiable nerves, 1 of the nerves was clearly identified as the seventh cranial nerve but the other, described as a small CN in the radiologic report, was on intraoperative observation clarified to be the nervus intermedius. This finding confirms that the precise determination of the individual nerves in ears with a CND and a small IAC is limited by the resolution of the MRI system and by the degree of spatial separation of the nerves.

Evoked auditory brainstem response testing has been proposed as a prognostic means of defining the status of the CN and predicting the results of CI in children with ambiguous results on imaging studies.<sup>33-37</sup> However, it is unclear whether the CN fibers in children with no radiographic evidence of a nerve can be confirmed by preoperative evoked auditory brainstem response testing. This procedure has been used in many centers with large variations in technique and outcome. Indeed, some patients with CND and no clear response on promontory evoked auditory brainstem response testing have been reported to achieve limited auditory responses after CI.

*Complications of ABI and CI.* The potential complications of retrosigmoid craniotomy are greater than those of the transmastoid approach of CI. However, in practice, the major and minor complication rates of the procedures are comparable in the hands of well-trained surgical teams.<sup>38</sup>

## CONCLUSIONS

Cochlear nerve deficiency can complicate the decision regarding whether to proceed with CI or ABI. Because the study patients had already undergone failed CI, it is difficult to extrapolate our findings regarding CI to all patients with CND. However, in this cohort of patients who had a clinical diagnosis of CND and imaging of an absent or small CN who underwent ABI, the outcomes of ABI were superior to those of CI. In such children who fail to progress with CI, the best opportunity for developing open-set speech perception and acquiring speech may be ABI. Auditory brainstem implantation should be considered early if cochlear implant performance is suboptimal.

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