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The Therapeutic Dilemma of Cochlear Nerve Deficiency: Cochlear or Brainstem Implantation?

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Abstract

Objective. To compare the outcomes between 2 age-matched cohorts of children with cochlear nerve deficiency: those receiving auditory brainstem implants (group A) or cochlear implants (group B).

Study Design. Retrospective cohort study.

Setting. Tertiary referral center.

Subjects and Methods. Subjects were selected from a pool of 537 children fitted with cochlear implants (n = 443) or auditory brainstem implants (n = 94) over the past 14 years. Performance, examined with the Category of Auditory Performance scale, and complications were compared with a mean follow-up of 5 years.

Results. All children had bilateral profound sensorineural hearing loss and cochlear nerve deficiency. Magnetic resonance imaging documented an absent cochlear nerve (n = 12) and a small cochlear nerve (n = 8) in group A and an absent cochlear nerve (n = 11) and a small cochlear nerve (n = 9) in group B (P = 1.000). Children with cochlear implants had Category of Auditory Performance scores spanning from 0 to 3 levels of performance, and all required manual communication mode and visual supplementation. Children with auditory brainstem implants had Category of Auditory Performance scores spanning from 2 to 7, and most patients demonstrated behavioral responses irrespective of inner ear malformations and an absent cochlear nerve or small cochlear nerve ($P < .001$).

Conclusions. In children with cochlear nerve deficiency, patients fitted with cochlear implants did not develop speech understanding and production. Those fitted with auditory brainstem implants had the opportunity to develop open-set speech perception, acquiring verbal language competence using oral communication exclusively and participating in mainstream education. The overall complication rate of auditory brainstem implants was not greater than that of cochlear implants.

Keywords

cochlear implant, cochlear nerve deficiency, auditory brainstem implant

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Hearing restoration in children with cochlear nerve deficiency (CND) is a therapeutic challenge, with conflicting reports describing children who, despite cochlear nerve hypoplasia or aplasia on magnetic resonance imaging (MRI), show auditory responses to different procedures, including simple amplification,[1,2] cochlear implants (CIs),[3-6] and auditory brainstem implants (ABIs).[7-11] An evident caveat of most of these studies is the very small number of subjects in any given subgroup comparison.

Clearly, children with CND are a special population and generally perform more poorly than average pediatric CI recipients, but exceptions have been described. This raises medical and ethical matters of selecting the device and intervention that might prove most beneficial. However, the current literature at present indicates unequivocally that CIs and not ABIs are the first-line treatment for these children, even in the absence of any scientific evidence that CIs outperform ABIs in this cohort of children. So, in many centers, CIs continue to be offered to patients with CND, surmising that some cochlear nerve fibers are present but not visible due to MRI limitations or because they occur within the facial or vestibular nerve.[12,13]

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Supported by studies showing better outcomes in children with CND when fitted with ABIs compared with children with CIs,[14,15] ABI recently has been proposed as the first-line treatment in children with CND. This proposal has generated the therapeutic dilemma of selecting CI or ABI as the best treatment option to be offered to children with CND.

To clarify these issues, we reviewed our population of children fitted with ABIs (n = 94) and CIs (n = 443) over the past 14 years and extracted 2 age-matched groups of children diagnosed with CND and fitted with a CI or an ABI who were younger than 3 years and operated on by the same surgeon (V.C.). The aim of the investigation was to determine whether differences exist in the trajectories of auditory development of the 2 procedures to justify the option of ABI as a first-line treatment in children with CND.

Materials and Methods

The Verona University Ethics Board approved the study, and all families gave their informed consent.

From 1998 to 2013, we fitted 443 children with CIs and 94 with ABIs following the outcome of a personal preimplantation audiological assessment described in detail elsewhere.[16] The expected outcome, possible risks, and prevalence of the complications of CI and ABI surgery were discussed with the parents and their consent obtained. Consideration was given to the surgical indication of the referring doctor, but the final decision on the surgical procedure was adopted at the discretion of the family in agreement with the proposal of the surgeon. So far, 32 children have traveled internationally to have hearing restored with a bionic device, but the high or low socioeconomic status of the family has never interfered with the surgeon's selection of the procedure.

From the 2 groups of children fitted with CIs or ABIs, we were able to retrieve the clinical charts of 54 children who met the following criteria: bilateral profound hearing loss from congenital deafness with CND, absent or small cochlear nerves, cochlear and internal auditory canal (IAC) malformations, no prior hearing experience (including hearing aid use), no previous meningitis and no coexisting hindbrain anomalies, unilateral CI and ABI implantation,[3] and all operated on during the same period (2004-2009) before 3 years of age. From this pool of 54 children, 14 were excluded from the study (see **Figure 1** for details of exclusion criteria). Approximately 50% of these initial 54 children had other nonauditory disabilities.

So finally, from a total of 537 children fitted with CIs (n = 443) or ABIs (n = 94) over the past 14 years, only 2 groups of 20 children, matched for age and fitted with ABIs or CIs, fulfilled the selection criteria. Both groups were followed for up to 8 years to compare outcome measures.

The retrosigmoid and posterior tympanotomy approaches were used for the ABIs and CIs, respectively.[7,14-16] Electrically evoked auditory brainstem recordings (EABRs) were performed preoperatively, intraoperatively at the end of surgery, and during follow-up in all children. All children in each group had unilateral CIs (17 Cochlear devices, Sydney, Australia, and 3 Med-El devices, Innsbruck, Austria) or ABIs (18 Cochlear and 2 Med-El devices) fitted.

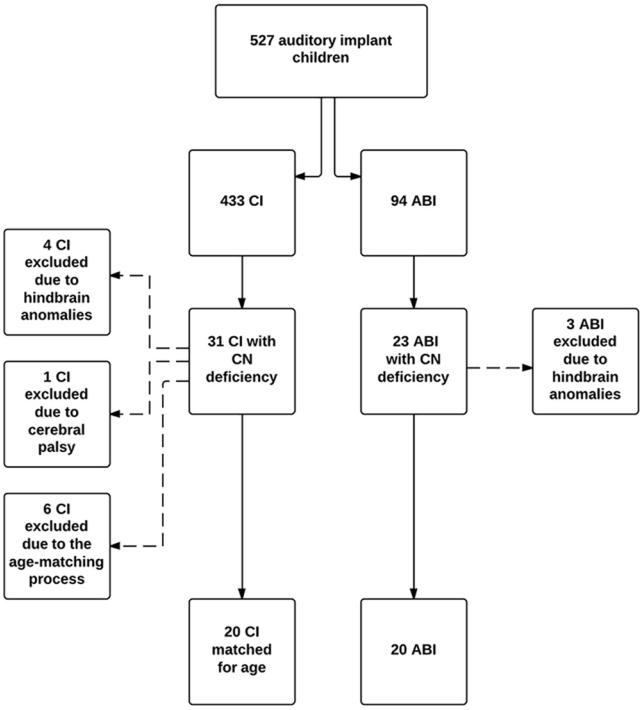

Figure 1. Flowchart for patient selection for inclusion in the auditory brainstem implant (ABI) and cochlear implant (CI) groups.

The algorithm for the rehabilitation of children fitted with CIs and ABIs included conditioned play audiometry, practiced at the beginning of every fitting session either with standardized instrumental sounds or with speech sounds (Six Ling's Sound Test) as a routine.

The evaluation of auditory perceptual ability was assessed with the Category of Auditory Performance (CAP) test[17,18] as previously illustrated.[15]

Statistical analysis included the *t* test, Wilcoxon Mann-Whitney test, Fisher exact test, and linear regression analysis, as appropriate.

Results

Demographic, clinical, and follow-up data are detailed in **Table 1**. All children completed the 24-month follow-up, while 16 subjects in each group were still enrolled in the study at 36 months.

Four children in group A (ABI) had associated cognitive deficits (among these subjects, 3 also had mild motor disabilities), 1 had behavioral impairment (attention-deficit hyperactivity disorder), 1 child was visually impaired, and 2 children were diagnosed with a polymalformative syndrome (Down and Moebius syndromes). Four children in group B (CI) also had associated cognitive deficits (1 also had mild motor disabilities), 1 child was visually impaired, and 3 children had other syndromes (Down, Shprintzen, and Moebius syndromes). There were 11 and 10 right ears and 9 and 10 left ears, respectively, in groups A and B ($P = 1.000$).

Table 1. Demographic Data for the 2 Study Populations.[a]

	Group A (ABI)	Group B (CI)	*P* Value
No. of patients	20	20	
Age at implantation, mean ± SD, y	1.4 ± 0.5	1.3 ± 0.4	.489[b]
Sex, male/female	13/7	11/9	.748[c]
Side, right/left	11/9	10/10	1.000[c]
Follow-up, median (interquartile range), y	6.9 (3.2-8)	4.7 (3.1-8)	.666[a]
Cochlear nerve deficiency, absent/small	12/8	11/9	1.000[b]
Auditory neuropathy spectrum disorders (normal cochleae)	5	4	1.000[b]
Associated cochlear malformations (subjects)	15	16	1.000[b]
Associated disabilities (subjects)	8	8	1.000[b]

Abbreviations: ABI, auditory brainstem implant; CI, cochlear implant.
[a]Values are presented as numbers unless otherwise indicated.
[b]*t* Test/Wilcoxon Mann-Whitney test as appropriate.
[c]Fisher exact test.

The EABR recordings performed intraoperatively demonstrated no auditory response in CI recipients and at least an auditory response on 8 to 11 (Cochlear) and 4 to 6 (Med-El) electrodes in children fitted with an ABI.

Imaging

Magnetic resonance imaging documented an absent cochlear nerve (ACN) and a small cochlear nerve (SCN) in 12 and 8 and in 11 and 9 children, respectively, in groups A and B ($P = 1.000$). Interestingly, among children with ACN, an open auditory nerve canal (ANC) was found in 5 and 4 children in groups A and B, respectively. The facial nerve (FN) had an aberrant course in 4 and 5 children in groups A and B, respectively.

Measurements of the IAC and ANC diameters were evaluated with high-resolution computed tomography (CT) scans for each child in both groups. The IAC was atretic in 4 and 3 children in groups A and B, respectively ($P = 1.000$). The diameter of the IAC was reduced (ie, less than 3 mm) in 12 and 13 children in groups A and B, respectively. The ANC diameter measurements showed abnormalities in children in both groups. A severe stenosis with an ANC diameter of less than 1.0 mm (0.31 ± 0.43 mm) was observed in 13 children in group A and 11 in group B. A moderate stenosis with a diameter of less than 1.8 mm was observed in 3 children in group A and 4 in group B (1.53 ± 0.25 mm). In the remaining children, the ANC was normal but empty on MRI. Because of the difficulty in obtaining clear auditory nerve (AN) diameter measurements, it was not possible to compute the correlation between the diameter of the AN and FN.

Cochlear abnormalities of different degrees were present in both groups: moderate in 6 and 5 children and severe in 9 and 11 children in groups A and B, respectively. Interestingly, cochlear morphology was normal on CT and MRI in 5 children in group A and 4 in group B, but the ANC was of abnormally reduced size in both groups. Severe vestibular malformations were associated with severe or extreme abnormalities of the cochlea in both groups. No child in the present 2 cohorts showed evidence of cochlear ossification.

Auditory Perceptual Abilities

The CAPs obtained before implantation scored 0 in all children in both groups. Both groups were tested with the CAP procedure at each visit after device activation, every 3 months for the first 24 months. After 24 months of device use, CAP scores showed significantly poorer outcomes in group B (0.7 ± 0.5) compared with group A (2.4 ± 1.3) ($P < .001$).

After the 24-month test, 5 children in group B were obtaining no benefit from the CI. After full discussion and informed consent from the parents, these children had the CI removed and an ipsilateral ABI fitted; these children dropped out of the present study. In the remaining children, CAP measurements were collected approximately every 6 months up to 8 years. At the 48-month follow-up, 1 child in group A could not be tested because the family went back to their original country and 4 more children in group B obtaining no benefit from the CI had the CI explanted and an ipsilateral ABI fitted. These children also dropped out of the study. At the 60- and 72-month follow-up, the number of ABI children remained the same, but the number of CI children dropped to 6 because 3 more children had the CI removed and had an ABI fitted ipsilaterally. **Figure 2** shows a scatterplot of the CAP scores of groups A and B as a function of ABI and CI experience. The CAP scores were higher in group A at all follow-ups of behavioral testing. After 2 years of device use, CAP scores continued to improve in group A, whereas group B reached a plateau at an approximate score of 2 within 4 years and did not improve significantly even after 8 years of CI experience (6.1 ± 1.0 vs 2 ± 0.8, $P < .0001$), with the exception of 2 patients, who were at least able to respond to speech sounds, without any identification skill, and to recognize very simple environmental sounds, such as continuous vs interrupted stimuli (**Figure 3**).

Nearly all ABI children demonstrated behavioral responses irrespective of inner ear and IAC morphology.

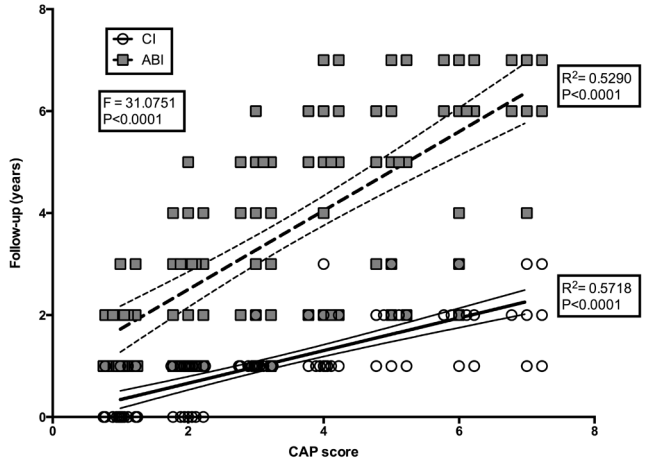

Figure 2. Category of Auditory Performance (CAP) developmental trajectory in children with cochlear nerve deficiency: auditory brainstem implant (ABI) vs cochlear implant (CI). The trend lines for the ABI and CI groups are represented by the dashed and solid lines, respectively.

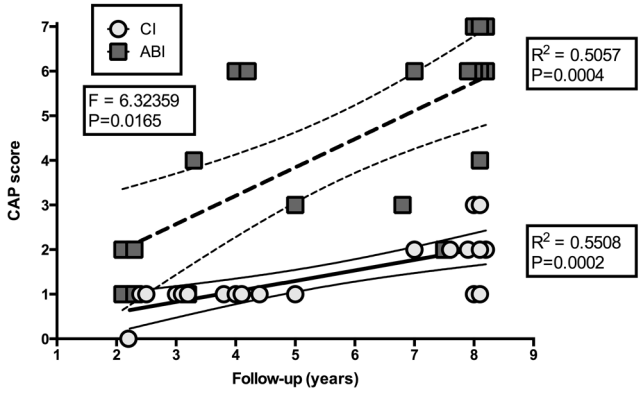

Figure 3. Category of Auditory Performance (CAP) scores and trend lines of 40 children with cochlear nerve deficiency fitted with an auditory brainstem implant (ABI) or a cochlear implant (CI) at the last follow-up.

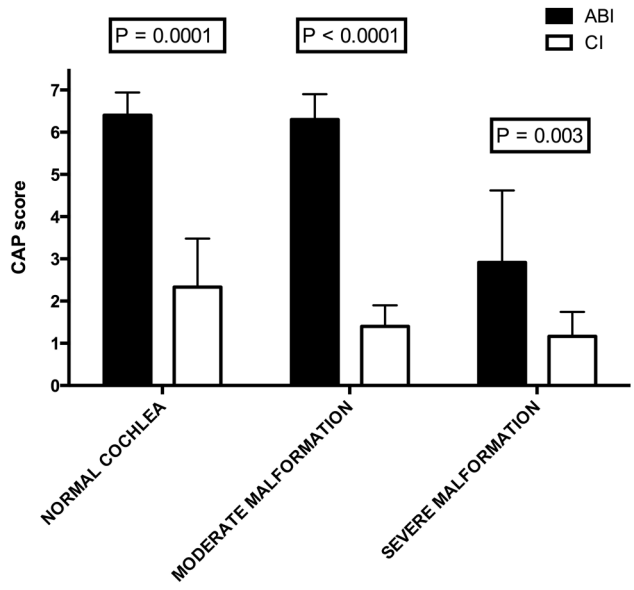

Figure 4. Last Category of Auditory Performance (CAP) scores of children with cochlear nerve deficiency fitted with an auditory brainstem implant (ABI) or a cochlear implant (CI) grouped by degree of cochlear malformation.

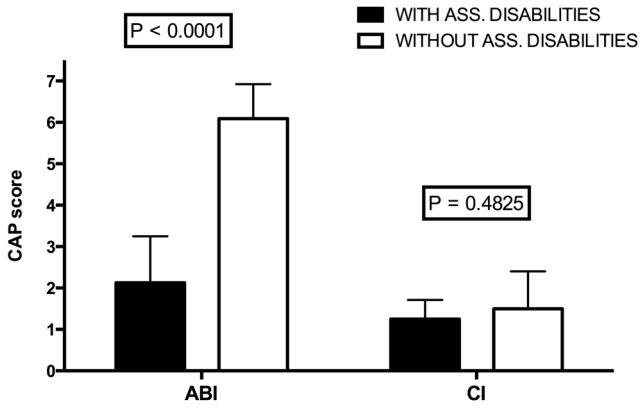

Figure 5. Last Category of Auditory Performance (CAP) scores in children with cochlear nerve deficiency fitted with an auditory brainstem implant (ABI) or a cochlear implant (CI) with or without associated disabilities.

The benefit from CI was limited to auditory awareness with behavioral responses induced at very high levels of charge units, often associated with nonauditory stimulation such as facial nerve stimulation and disequilibrium, so much so that in 5 patients, all electrodes had to be inactivated and the children explanted and fitted with ABIs.

The children with normal cochleae and either ACNs or SCNs fitted with ABIs demonstrated a significantly earlier and better perceptual outcome on the CAP test than did children with cochlear abnormalities; all children with normal cochleae had a CAP score of more than 5 at the last follow-up after ABI fitting (6.4 ± 0.5 vs 2.3 ± 1.2; $P <$ .0001) (**Figure 4**). No children with normal cochleae presented associated disabilities.

The ABI children without associated disabilities showed better auditory performance than children with associated disabilities at all follow-up intervals (6.1 ± 0.8 vs 2.1 ± 1.1; $P <$ .0001, at the last follow-up). Conversely, the CI children without associated disabilities demonstrated a small but not significant difference in performance at all follow-up intervals (1.5 ± 0.9 vs 1.4 ±0.4; P = .483, at the last follow-up) compared with children with disabilities (**Figure 5**).

Safety

No major anesthesiological or surgical complications such as cardiac arrest, facial palsy, or flap breakdown were observed in any child.

Among minor anesthesiological complications, 2 children aged 13 and 24 months in the ABI group experienced

transitory bronchospasm and hypotension, both of which resolved with medical treatment. Blood pressure range during surgery was not statistically significantly different in the 2 study groups ($P = .552$). No perioperative surgical complications were encountered in any children. Blood loss was recorded as less than 30 mL in all patients. There were 3 minor postoperative complications: 2 cases of wound seroma (1 in each group) and 1 case of wound infection in group B; all were treated conservatively. Children in groups A and B were discharged, respectively, after an average of 6.3 ± 2.1 and 2.6 ± 1.8 days ($P < .001$). Delayed wound healing (10 days after surgery) was observed in 1 child in group A and in 2 subjects in group B. Within 2 years of implantation, postoperative otitis media was observed in the same ear as the CI in 3 children. All were treated medically with no further complications. No complications related to ABI or CI activation or long-term use were evident in any subject, apart from those children who experienced facial nerve stimulation and had some CI electrodes deactivated.

Discussion

Earlier studies involving behavioral outcome measures in children with CND fitted with CIs have reported very poor results, leading to decisions not to provide a CI to these children.[19-22] However, more recent studies indicate that limited speech detection and discrimination and, very occasionally, higher levels of auditory performance may be observed in these children.[23-27] The recent innovative proposal of offering ABIs as first-line treatment in children with CND, corroborated by significantly better outcome compared with children fitted with CIs,[3,7-11,14-16] complicated the decision with regard to the best treatment option for children with CND and generated a pivotal therapeutic dilemma.

Clearly, if some reasonably good outcomes are achieved with CIs, it is difficult to decide in favor of an ABI as the initial treatment in these patients, considering the potentially serious risks of this intracranial procedure. These reservations, supported by the inability of preoperative MRI and EABRs to provide unambiguous information with regard to the status of the cochlear nerve, have suggested cautiously that children with CND should first undergo a trial with CIs to verify the benefit of the procedure and, only after confirming the inefficacy of the CI, could ABI possibly be considered.

A recent study[15] described a cohort of 21 children with a clinical diagnosis of CND fitted with CIs. Among these children 13 presented ACNs and 8 SCNs, respectively. As a result of failure of progression of auditory ability in all these children, the CIs were explanted and ABIs fitted ipsilaterally. At surgery, the so-called SCN was demonstrated in all cases to be the nervus intermedius. This very important observation confirmed that the determination of the individual nerves in ears with stenotic IAC is limited by the degree of spatial separation of the nerves.[12,13] In this cohort of children, the opportunity to develop open-set speech perception and acquire speech was obtained only after fitting an ABI.

The time course for the development of auditory perception in profoundly deaf children with CND following CI or ABI may extend over many years, and long-term investigations are needed to determine whether the 2 devices differ significantly in the trajectories of auditory development to justify the option of the ABI as a first-line treatment in these children. To provide a contribution to this theme and unravel the dilemma of the best treatment for children with CND, the present retrospective study was performed. To our knowledge, no such studies exist in the literature.

The outcome of the present investigation indicates that CAP scores were significantly poorer in the CI group compared with the ABI group: most children in the ABI group experienced a gradual increase in performance over time, whereas children in the CI group achieved some initial improvement in behavioral test scores without any further improvement even after long-term implant experience. Within the first year of activation, the entire ABI group obtained awareness of environmental sounds, and 45% responded to speech sounds. At the second year of follow-up, 50% of these young patients were able to recognize environmental sounds and 20% discriminated speech sounds, while in the third year of ABI use, 31.3% of group A were in open-set speech perception. Eight of 11 subjects who reached the fifth year of ABI fitting were able to understand simple commands with no lip reading, and 3 were capable of sustaining a telephone conversation with a familiar speaker. After 8 years of follow-up, 12 children from the CI cohort in the present study were explanted and fitted with ABIs, obtaining a partial recovery.

A comparison of the complications associated with ABI and CI surgery confirms that, even though the potential complications of a retrosigmoid craniotomy are clearly greater than those of the transmastoid approach of CI surgery, in practice, both major and minor complication rates are comparable in the hands of well-trained surgical teams.[27]

Further consideration should be given to the cost-benefit ratio and psychological involvement of the family of a child diagnosed with profound hearing loss and CND at the age of 3 to 4 months who is fitted first with a hearing aid for 6 to 12 months and then with a CI for a further 1 to 3 years and finally, only after all these inconveniences, receives the suggestion to have their child fitted with an ABI.

As a result of this study, we advocate EABR preoperative evaluation in CI and ABI candidates and intraoperative evaluation and programming with threshold determination in children with CND fitted with CIs and ABIs. Similarly, periodic EABRs should be performed to objectively assess CI or ABI device ''efficacy'' in these children and stratify candidates into those expected or not expected to achieve open-set speech perception.

The CI children who achieve poor speech perception results after 2 years of CI use and who have an abnormal EABR may receive limited benefit from their CI, and such candidates may profit from the ABI. The long-term outcome

study of the present article shows that children with CND and ABI do outperform those treated with CI.

We have learned that fitting a CI in a subject with CND, cochlear and IAC malformations, and no RW-EABRs may be a waste of time and expense. At the same time, a child fitted with CI showing no postoperative EABRs and no auditory progress for more than 2 years should not wait any further and should be fitted with a contralateral ABI.

Cochlear nerve deficiency is a relatively common cause of profound sensorineural hearing loss that challenges the decision-making process with regard to whether to proceed with a CI or an ABI.

In the present cohort of children with CND, those fitted with CIs did not develop speech understanding and production. Those fitted with ABIs frequently developed open-set speech perception, with some acquiring verbal language competence using oral communication and participating in mainstream education. Furthermore, since the overall complication rate of ABIs was not greater than that of CIs, consideration should be given to the use of ABI technology as the first surgical prosthesis of choice in this patient population.

Author Contributions

Liliana Colletti, conception and design of the study, acquisition of data, analysis and interpretation of data, drafting and revising the article, final approval; **Giacomo Colletti**, conception and design of the study, analysis and interpretation of data, drafting and revising the article, final approval; **Marco Mandalà**, conception and design of the study, acquisition of data, analysis and interpretation of data, drafting and revising the article, final approval; **Vittorio Colletti**, conception and design of the study, acquisition of data, analysis and interpretation of data, drafting and revising the article, final approval.

Disclosures

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References

1. Bamiou DE, Worth S, Phelps P, et al. Eighth nerve aplasia and hypoplasia in cochlear implant candidates: the clinical perspective. *Otol Neurotol*. 2001;22:492-496.
2. Bradley J, Beale T, Graham J. Variable long-term outcomes from cochlear implantation in children with hypoplastic auditory nerves. *Cochlear Implants Int*. 2008;9:34-60.
3. Buchman CA, Teagle HF, Roush PA, et al. Cochlear implantation in children with labyrinthine anomalies and cochlear nerve deficiency: implications for auditory brainstem implantation. *Laryngoscope*. 2011;121:1979-1988.
4. Teagle HF, Roush PA, Woodard JS, et al. Cochlear implantation in children with auditory neuropathy spectrum disorder. *Ear Hear*. 2010;31:325-335.
5. Young NM, Kim FM, Ryan ME, et al. Pediatric cochlear implantation of children with eighth nerve deficiency. *Int J Pediatr Otorhinolaryngol*. 2012;76:1442-1448.
6. Zanetti D, Guida M, Barezzani MG, et al. Favorable outcome of cochlear implant in VIIIth nerve deficiency. *Otol Neurotol*. 2006;27:815-823.
7. Colletti V, Carner M, Miorelli V, et al. Cochlear implant failure: is an auditory brainstem implant the answer? *Acta Otolaryngol*. 2004;124:353-357.
8. Choi JY, Song MH, Jeon JH, et al. Early surgical results of auditory brainstem implantation in nontumor patients. *Laryngoscope*. 2011;121:2610-2618.
9. Sennaroglu L, Ziyal I, Atas A, et al. Preliminary results of auditory brainstem implantation in prelingually deaf children with inner ear malformations including severe stenosis of the cochlear aperture and aplasia of the cochlear nerve. *Otol Neurotol*. 2009;30:708-715.
10. Grayeli AB, Bouccara D, Kalamarides M, et al. Auditory brainstem implant in bilateral and completely ossified cochleae. *Otol Neurotol*. 2003;24:79-82.
11. Cervera-Paz FJ, Manrique MJ. Auditory brainstem implants: past, present and future prospects. *Acta Neurochir Suppl*. 2007;97:437-442.
12. Thai-Van H, Fraysse B, Berry I, et al. Functional magnetic resonance imaging may avoid misdiagnosis of cochleovestibular nerve aplasia in congenital deafness. *Am J Otol*. 2000;21:663-670.
13. Ozdogmus O, Sezen O, Kubilay U, et al. Connections between the facial, vestibular and cochlear nerve bundles within the internal auditory canal. *J Anat*. 2004;205:65-75.
14. Colletti L, Zoccante L. Nonverbal cognitive abilities and auditory performance in children fitted with auditory brainstem implants: preliminary report. *Laryngoscope*. 2008;118:1443-1448.
15. Colletti L, Wilkinson EP, Colletti V. Auditory brainstem implantation after unsuccessful cochlear implantation of children with clinical diagnosis of cochlear nerve deficiency. *Ann Otol Rhinol Laryngol*. 2013;122:605-612.
16. Colletti L, Mandalà M, Colletti V. Cochlear implants in children younger than 6 months. *Otolaryngol Head Neck Surg*. 2012;147:139-146.
17. Archbold S, Lutman ME, Marshall DH. Categories of auditory performance. *Ann Otol Rhinol Laryngol*. 1995;104 (suppl 166):312-314.
18. Archbold S, Lutman ME, Nikolopoulos T. Categories of auditory performance: inter-user reliability. *Br J Audiol*. 1998;32:7-12.
19. Shelton C, Luxford WM, Tonokawa LL, et al. The narrow internal auditory canal in children: a contraindication to cochlear implants. *Otolaryngol Head Neck Surg*. 1989;100:227-231.
20. Gray RF, Ray J, Baguley DM, et al. Cochlear implant failure due to unexpected absence of the eighth nerve: a cautionary tale. *J Laryngol Otol*. 1998;112:646-649.
21. Maxwell AP, Mason SM, O'Donoghue GM. Cochlear nerve aplasia: its importance in cochlear implantation. *Am J Otol*. 1999;20:335-337.
22. Bradley J, Beale T, Graham J, et al. Variable long-term outcomes from cochlear implantation in children with hypoplastic auditory nerves. *Cochlear Implants Int*. 2008;9:34-60.

23. Acker T, Mathur NN, Savy L, Graham JM. Is there a functioning vestibulocochlear nerve? Cochlear implantation in a child with symmetrical auditory findings but asymmetric imaging. *Int J Pediatr Otorhinolaryngol*. 2001;57:171-176.
24. Song MH, Bae MR, Kim HN, et al. Value of intracochlear electrically evoked auditory brainstem response after cochlear implantation in patients with narrow internal auditory canal. *Laryngoscope*. 2010;120:1625-1631.
25. Ito J, Sakota T, Kato H, et al. Surgical considerations regarding cochlear implantation in the congenitally malformed cochlea. *Otolaryngol Head Neck Surg*. 1999;121:495-498.
26. Chadha NK, James AL, Gordon KA, et al. Bilateral cochlear implantation in children with anomalous cochleovestibular anatomy. *Arch Otolaryngol Head Neck Surg*. 2009;135:903-909.
27. Colletti V, Shannon RV, Carner M, et al. Complications in auditory brainstem implant surgery in adults and children. *Otol Neurotol*. 2010;31:558-564.