Eur Arch Otorhinolaryngol
DOI 10.1007/s00405-013-2378-3

REVIEW ARTICLE

Indications and contraindications of auditory brainstem implants: systematic review and illustrative cases

Paul Merkus · Fillipo Di Lella · Giuseppe Di Trapani · Enrico Pasanisi · Milo A. Beltrame · Diego Zanetti · Maurizio Negri · Mario Sanna

Received: 21 August 2012 / Accepted: 22 January 2013

Abstract The number of non-neurofibromatosis type 2 (NF2) indications for auditory brainstem implant (ABI) in the literature is increasing. The objective of this study was to analyze and discuss the indications for ABI. Retrospective chart review and systematic review were conducted at Quaternary referral skull base center and referring centers. Analysis of ABI cases with non-NF2 indications and systematic review presenting non-NF2 ABI cases were performed. Fourteen referred cases with ABI were identified. All cases had unsatisfactory results of ABI and all could have been rehabilitated with a cochlear implant (CI). Of these 14 cases, 9 improved with a cochlear implant, and 2 with a hearing aid, two are still planned for CI, one received bilateral CI, no ABI. In literature, we found 31 articles presenting 144 non-NF2 ABI cases with at least 7 different indications other than NF2. ABI should be restricted to those patients who have no other rehabilitation options. Patency of the cochlea and evidence of an intact cochlear nerve should be examined with imaging and electrophysiologic testing. Sometimes a CI trial should be planned prior to proceeding with ABI. We have shown that in many cases a CI is still possible and CI provided better results than ABI. In vestibular schwannoma in the only hearing ear, cochlear otosclerosis, temporal bone fractures, (presumed) bilateral traumatic cochlear nerve disruption, auto-immune inner ear disease and auditory neuropathy primarily CI are indicated. Traumatic bilateral cochlear nerve disruption is exceptionally unlikely. In cochlear nerve aplasia, testing should be performed prior to meeting indications for ABI. In malformations, ABI is indicated only in severe cochlear hypoplasia or cochlear aplasia.

Keywords Auditory brainstem implant · Deafness · Treatment · Meningitis · Otosclerosis · Temporal bone fracture · Cochlear nerve · Vestibular schwannoma · Auditory neuropathy · Cochlear implant · Labyrinth malformation

Introduction

The indication for an auditory brainstem implant (ABI) in neurofibromatosis type 2 (NF2) patients is well known as it

Electronic supplementary material The online version of this article (doi:10.1007/s00405-013-2378-3) contains supplementary material, which is available to authorized users.

P. Merkus (✉)
Department of Otorhinolaryngology and Head and Neck Surgery, VU University Medical Center, KNO 1d-114, PO Box 7057, 1007 MB Amsterdam, The Netherlands
e-mail: p.merkus@vumc.nl

F. D. Lella · G. D. Trapani · M. Sanna
Department of Otology and Skull Base Surgery, Gruppo Otologico, Piacenza-Rome, Italy

E. Pasanisi
Section of Middle Ear Microsurgery and Otoneurosurgery, Department of Otolaryngology-Head and Neck Surgery, University of Parma, Parma, Italy

M. A. Beltrame
Department of Otolaryngology, S. Maria del Carmine Hospital, Rovereto, Italy

D. Zanetti
Department of Otolaryngology, University of Brescia and S. Gerardo Hospital, Monza, Italy

M. Negri
Department of Otolaryngology, Ramazzini Hospital, Carpi, Italy

has been used for years [1]. Next to NF2, several other indications have been reported over the years and seem plausible in the cases where no cochlear implant (CI) or other means of rehabilitation can be used. Nevertheless, some surgeons choose ABI over a more traditional cochlear implant surgery, even when CI placement was still possible. We have seen those cases in our own clinic and in literature. Proper diagnostic evaluation needs to be done before a decision to implant an ABI can be made. Cochlear implantation results are much more predictable and have better hearing rehabilitation results compared with an ABI [2]. The primary goal should be the evaluation for a CI instead of an ABI as the means of rehabilitation. We conducted this study to distinguish the patients who received an ABI, but who perform better with a CI, and to comprehend the steps needed before an ABI indication exists.

We present 14 cases of ABI placement in other clinics, in which ABI was chosen as a means of rehabilitation instead of an alternative. The diagnosis and indications for ABI in these cases, as well as the indications mentioned in the literature, are summarized and discussed herein. We believe that ABI should be a 'last resort' means of rehabilitation. As in many cases, a stepwise strategy has to be completed before an ABI decision is applicable. In order to discuss all the known indications and contraindications of ABI, we conducted a systematic review of the literature concerning ABI cases, with the focus being on non-NF2 patients. Furthermore, we present several illustrative cases of patients, who have received an ABI, and discuss their indication as well as the possible strategy and rehabilitation alternatives.

Methods

ABI cases

All non-NF2 patients who were already implanted with an ABI were analyzed. Some were referred to our clinic but others were seen and treated in our referral clinics. All non-NF2 patients implanted with an ABI in our clinic were also included in this analysis.

Systematic review of the literature

A search was performed in PubMed and Embase in January 2012. We assessed the articles for inclusion/exclusion criteria by evaluating title, abstract, full article and checking for related articles in the references. The filters and inclusion/exclusion criteria were Limits: Human, English, German, Italian, Dutch; Inclusion/exclusion criteria: concerning patients, case series or case reports, no review, no phantom models, no cadaver study. Reporting (also) on ABI, not only CI. Reporting (also) on non-NF2 patients; Search syntax and results are shown in Online resource 1.

Results

ABI cases

From October 1986 through September 2011, 24 NF2 patients received 25 ABIs and 5 NF2 patients received a CI. Because of NF2 these patients were excluded from this study and analyzed separately (Sanna et al. [2]). In the non-NF2 group, 8 CI's were placed in the contralateral ear in cases of solitary vestibular schwannomas (VS) in the only hearing ear. Three ABIs were placed in cases of bilateral fully ossified cochlea after drill-out procedure and CI attempt in a single procedure. Bilateral fully ossified cochleae have been the only non-tumor ABI indication in our clinic. We have been able to rehabilitate all other patients, with presumed ABI indications, by means other than ABI. In our outpatient clinic, we have seen 13 patients who had an ABI in another clinic and one who was referred for auditory brainstem implantation but received bilateral CI implantation. These 14 patients have been analyzed. Of the patients with ABI, 12 of them were non-tumor patients; one had VS in the only hearing ear. All had a dissatisfactory result of their ABI. These cases are shown in Table 1, and their full description is accessible as digital supplement (Online resource 2). In all 14 cases, a cochlear implantation was or is possible, because of a patent cochlea and no absence of the cochlear nerve on imaging. The speech outcomes of the cases with CI after ABI are presented as a line plot in Fig. 1 and show an improvement after CI. In addition, two patients who did not have satisfactory results with their ABI were refitted with a hearing aid contralaterally. They had an improvement of their hearing, as shown in Fig. 1. Maybe these two patients will also receive a CI, depending on personal motivation and audiological criteria, as there is no contraindication on imaging.

In short, fourteen referred cases with questionable indications for ABI were identified. Of these 14 cases, 11 of them improved their hearing with rehabilitation via different means: nine of them were reoperated after ABI placement and received a CI and two were refitted with a hearing aid. Two are still planned for CI, one received bilateral CI and no ABI. The indications of all 14 patients will be discussed below, next to all indications seen in literature.

Systematic literature review

The search resulted in 1,115 and 1,021 articles in PubMed and Embase, respectively. After eliminating duplicates, 1,122 articles remained. A filter was used (human studies, language English, German, Italian, or Dutch): 587 papers

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

Table 1 Of the 14 presented cases, 13 of them (A–H, J–N) showed unsatisfactory results after ABI placement

Etiology	Pt	Sex	Age (years)	Right ear	Left ear	Figure	First treatment & results	2nd treatment	Results of 2nd treatment (see also Fig. 1)
VS only hearing ear	A	M	41	Anacusis due to sudden SNHL, 20 years before	SNHL: 35 db PTA; 85 dB SDS Extrameatal VS (1.5 cm)		ABI left side with tumor removal (SO) (Other Dpt 2003) *benefit declined over time* Facial nerve HB grade III	CI right side (Parma University 2010) pre-operative CT scan: evidence of bilateral cochlear patency	After 18 months from activation WR: 75 %; SR: 90 %; daily use of CI; no use of ABI
Bilateral cochlear otosclerosis	B	M	37	Profound mixed hearing loss: PTA BC: 50 dB PTA AC: 95 dB SDS: 65 % Hearing aid	Anacusis	2	ABI left side (Other Dpt: 2006) *ABI used for lip reading*	CI left side (2009) CI, standard transmastoid facial recess approach	After 36 months from activation WR: 71 %; SR: 50 %; daily use of CI; no use of ABI
	C	M	30	Mixed hearing loss: PTA 60 db with 10 dB ABG SDS: 100 % at 100 dB (2007)	Mixed hearing loss: PTA 60 db with 10 dB ABG SDS: 90 % at 100 dB		ABI left side (Other Dpt: 2007) *no benefit from ABI* ABI fitting problems and lower CN stimulation PTA 90 dB, no measurable ABG	HA right ear, (Brescia University) after CT scan evidence of cochlear patency next to cochlear remodelling (scan resembles Fig. 2)	Aided right side WR: 50 % max SDS: 60 % at 90 dB (2010) no use of the ABI CI still possible
	D	F	60	Profound SNHL Hearing aid	Profound mixed hearing loss Hearing aid		ABI right side (Other Dpt: 2003) *no benefit of ABI* and could not use HA	HA fitting left side (2010)	Aided WR: 60 % Aided SR: 100 % Aided speech comprehension: 100 % no use of the ABI
	E	M	68	Profound SNHL	Severe to profound hearing loss Hearing aid		ABI right side (Other Dpt: 2005) *no benefit of ABI*	CI right and left side (Rovereto Hospital, 2009& 2010) sequential bilateral CI after evidence of bilateral cochlear patency on CT scan	After 24 months after first CI, WR and SR with both CI almost 100 % with telephone use no use of the ABI
Head trauma/ temporal bone fracture	F	M	55	Anacusis Post-meningitis deafness on R side (at 12 years of age)	Anacusis Temporal bone fracture on L side (2006)		ABI left side (Other Dpt: 2006) free-field PTA 55 dB with WR 35 % with visual and auditory stimulation. 6 active electrodes	CI right side (Parma University 2009) CI after CT (cochlear patency on the right and partial obliteration on the left)	After 24 months from CI, WR 90 %, SR 90 % with telephone use no use of the ABI

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

Table 1 continued

Etiology	Pt	Sex	Age (years)	Right ear	Left ear	Figure	First treatment & results	2nd treatment	Results of 2nd treatment (see also Fig. 1)
	G	F	40	Anacusis Head trauma (2001) Total bilateral deafness	Anacusis; head trauma (2001) Total bilateral deafness and left facial nerve palsy		ABI right side (Other Dpt: 2002) *no hearing results*	CI right side (2006) CI after CT and MRI evidence of bilateral cochlear patency and intact cochlear nerve	After 6 months from activation WR: 75 %; SR: 60 %; daily use of CI; no use of ABI
	H	M	61	Anacusis; head trauma (2001) Total bilateral deafness	Anacusis	3	ABI right side (Other Dpt: 2002) early satisfactory audiological results that declined over time. *In 2010 WR: 0 %.*	CI right side (2010) CI after CT and MRI evidence of cochlear patency and intact cochlear nerve	After 18 months from CI activation, WR 95 %, SR 90 % with daily use of the implant no use of the ABI
	I	M	55	Anacusis; head trauma (2010) Total bilateral deafness	Anacusis		Bilateral simultaneous CI (2010) CT/MRI: evidence of bilateral cochlear patency and bilateral intact cochlear nerves	n.a.	After 6 months for CIs, SR 100 % with both CI
Hereditary/ progressive profound bilateral SNHL	J	F	31	Anacusis; Cogan syndrome	Anacusis Cogan syndrome	ESM_4	ABI right side (Other Dpt: 2003) WR 30 %, SR 50 % and comprehension 60 % (auditory and visual stimulation)	CI left side (2008) CI after evidence of cochlear patency on CT and MRI	After 36 months from CI, WR 100 %, SR 100 %, comprehension 100 %, telephone use No use of the ABI
	K	F	16	Hereditary Profound hearing loss	Hereditary Profound hearing loss	ESM_5	ABI left side (Other Dpt: 2005) *no WR scores*. Epilepsy and mioclonia during fitting of the implant	CI left side (2010) CI after evidence of cochlear patency on CT and MRI.	After 18 months from CI, WR 75 %, SR 40 % no use of the ABI
	L	F	69	Profound SNHL Progressive SNHL on right side	Profound SNHL Sudden SNHL on left side (2002)		ABI left side (Other Dpt: 2003), complicated by cerebellar edema, palpebral ptosis and dyplopia; WR 45 %.	CI right side (Ramazzini Hospital, Carpi, 2010) CI after evidence of cochlear patency on CT and MRI	After 24 months from CI, WR 80 %, sentence comprehension 80 %, daily use of CI, no use of ABI
	M	M	60	Severe SNHL HA: Progressive bilateral SNHL of unknown etiology	Severe SNHL HA: Progressive bilateral SNHL of unknown etiology		ABI right side (Other Dpt: 2003) anacusis, 5/21 active electrodes with a WR score of 40 %	Scheduled for CI, but afraid of new operation evidence of cochlear patency on CT and MRI	n.a.

Journal : Large 405 Dispatch : 1-2-2013 Pages : 11
Article No. : 2378 ☐ LE ☐ TYPESET
MS Code : EAORL-D-12-00785 ☑ CP ☑ DISK

Table 1 continued

Etiology	Pt	Sex	Age (years)	Right ear	Left ear	Figure	First treatment & results	2nd treatment	Results of 2nd treatment (see also Fig. 1)
Cochlear malformation	N	M	45	Severe SNHL	Severe SNHL Common cavity		ABI right side (Other Dpt: 2006) *No functional results*	CI right side is scheduled, but patient has postphoned it CT and MRI: evidence of a normal developed labyrinth R, cochlear patency R and intact cochlear nerve both sides	n.a.

In all 14, it was or is technically possible to insert a CI. In many cases, a cochlear implant or hearing aid proved to be a better method of rehabilitation (see Fig. 1). The ABI indications of these patients are discussed within the text. The second treatment was performed at the Gruppo Otologico in Piacenza, Italy, if no other location is mentioned. Full description of the cases is visible as supplementary digital content on the website (Online resource 1)

NF2 Neurofibromatosis type 2, *FN* facial nerve, *SDS* speech discrimination score, *Dpt* department, *SO* suboccipital approach, *AMI* auditory midbrain implant, *HA* hearing aid, *VS* vestibular schwannoma, *SNHL* sensorineural hearing loss, *HB* House and Brackmann grading, *WR* word recognition score, *SR* sentences recognition score, *ABG* air-bone gap, *lower CN* lower cranial nerves, *n.a.* not applicable, *GG* geniculate ganglion, *ESM* Electronically Supplemental Material (online resource)

remained. Subsequently, we assessed these articles for inclusion/exclusion criteria, on title (121 papers remained), in the abstract (49 papers remained). The full text of 49 papers was read. The articles that published on only NF2 patients were excluded. Articles with the clinical data of one or more patients who were implanted with an ABI for other reasons than NF2 were included. This resulted in 29 valuable articles about indications for ABI in non-NF2 cases. In the references, we discovered 2 additional papers not identified in our original search (Online resource 1). An overview of the 31 included articles is given in Table 2 and Online resource 3a & 3b. In these articles, a total of 144 non-NF2 ABI cases with at least 7 different indications other than NF2 were reported. Literature concerning ABI indications in NF2 cases is discussed in another paper [2]. The following non-NF2 indications are mentioned in literature (Table 2 and Online resource 3a & 3b):

- Vestibular schwannoma in the only hearing ear
- Post-meningitis ossification of both cochleae
- Otosclerosis
- Cochlear trauma/cochlear nerve disruption
- von Hippel-Lindau disease
- Bourneville-Pringle disease (tuberous sclerosis of the brain)
- Auditory neuropathy, idiopathic and due to bony entrapment (hyperostosis)
- Cochlear nerve aplasia
- Cochlear malformation

In the discussion, we present several patients and their outcomes to critically review the above-mentioned indications.

Discussion

In spite of some good ABI performers, the overall ABI speech perception results do not match the good results seen in modern cochlear implantation. Nonetheless, auditory sensations provided by ABI can be very helpful in facilitating oral communication and supporting lip-reading. We still believe that ABI is only indicated in patients with profound hearing loss or total bilateral deafness in which other means of rehabilitation, like a hearing aid or cochlear implant, are impossible. At least for many indications, a step-wise work-up should be performed prior to ABI placement. Although the empirical evidence for ABI indications is weak, we hope to explain this with the presented cases and literature.

VS in the only hearing ear

In patients with a unilateral vestibular schwannoma in the only hearing ear, there seems to be no indication for an ABI. The deaf ear is almost always suitable for CI, as

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

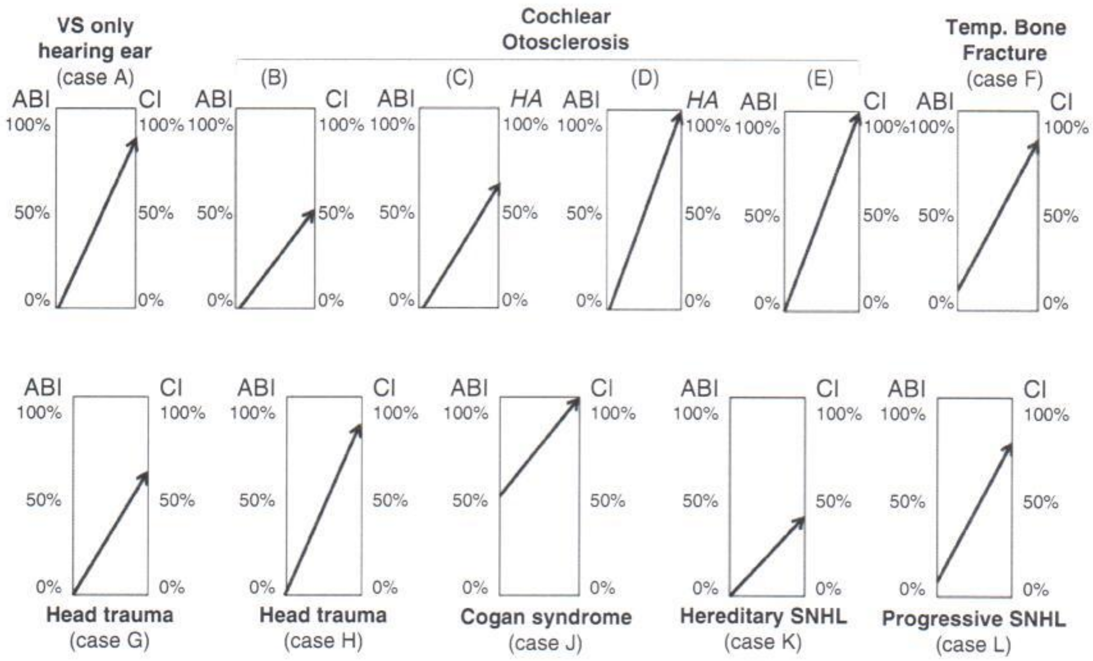

Fig. 1 Speech perception line plots per patient. In 11 patients ABI gave unsatisfactory results, which improved with other rehabilitation modalities. Nine patients received a CI, two a contralateral hearing aid (case *C* and *D*). The results of the hearing prior counseling (ABI) and post counseling and rehabilitation [CI or hearing aid (HA)] per patient are shown here, in percentage correct score. *Left-side*, start of arrow, ABI speech results and *right-side*, end of arrow, the CI/HA results

shown here in case A. In a recent analysis of our group, this topic has been addressed, showing that many other options are possible before ABI is considered [3]. The options *before* surgery in these VS cases include: a hearing aid in the affected ear if surgery can wait or a cochlear implant in the contralateral side [4]. The result of CI prior tumor removal can sometimes be poor, advocating ABI placement at the time of tumor removal [5]. However, a primary indication to first place an ABI before trying a contralateral CI seems incorrect. Also, several options during removal of the VS are possible: hearing preservation during the removal of a small VS [6], or preservation of the cochlear nerve and placement of a CI in the ipsilateral ear during the same surgery [7]. The abovementioned options leave almost no room for ABI indication in patients with a VS in the only hearing ear. Case A illustrates this strategy.

Post meningitis ossification of the cochlea

Since 2003, several papers have discussed the indication for ABI in cases of postmeningitic ossified cochleae [8–11]. They are all case reports or very small series, making definite conclusion difficult. It is pointed out that an ABI is indicated in cases of bilateral fully ossified cochleae, confirmed by scanning *and* preferably after CI attempt. A CI and ABI comparison is hard to investigate, as the presented hearing outcomes are often incomparable. An international accepted standardized way of presenting CI and ABI data could help to clarify this discussion.

Recently, a postmeningitis follow-up protocol was published [12] to secure cochlear implantation in the deafened patients before total obliteration of the cochlea has occurred. In cases presented months to years after the meningitis, MRI is essential to investigate the patency of the cochlea. Surgery to perform CI should be scheduled and different strategies should be anticipated: scala vestibuli insertion [13, 14], partial insertion [15], basal turn drill-out or double array insertion [16]. In our opinion, with clear cochlear ossification the operation should be done by ear canal wall down procedure to oversee the whole area including carotid artery and to provide access to the complete basal turn of the cochlea. If no cochlear lumen is present, one could convert to a translabyrinthine approach for auditory brainstem implantation or refer to an ABI center. Good results with CI are well known and also in some single cases ABI performance can be comparable [11]. Both CI and ABI seem to have a place in the rehabilitation of patients with ossified cochleae. To decide directly for ABI or first try a cochlear drill-out procedure seems a matter of personal preference since clear evidence is lacking.

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

Table 2 Results of a systematic review: 31 papers presenting patients who received an ABI because of another indication than NF2

First author	Publ year	Clinic	Ref	Period	Age	NF2	VS	Non-tumor	Patency coch	Nerve aplasia	Malf coch	Head trauma	Aud neurop	Specification
15 papers of Verona covering the same series (ref 24, 38–50)														
14× Colletti	2001–2010	Verona		1997–2008	11 months–70 years	31	5	78	33*	24	8	8	5	*22 otosclerosis, 7 meningitis, 1 Cogan
1× Eisenberg														
4 papers of Paris covering the same series (ref 8, 9, 53 & 54)														
3× Grayeli	2003–2008	Paris		1994–2006	17 years–71 years	23	3	5	4		1			3 meningitis, 1 otosclerosis
1× Coez														
1× Sollmann	2000	Freiburg	10, 56	1992–2001	17 years–58 years	49	6	3	2				1	1 BP disease, 1 HL tumor, 1 meningitis
1× Marangos														
2× Choi	2011	Seoul	52, 53					11	3	11				
Sanna	2006	Piacenza	11	2006	12 years	0	0	1	1					Meningitis
Sennaroglu	2009	Ankara	33	2006–2008	30–56 months	0	0	11		9	11			
Waterval	2011	Maastricht	31		44 years			1						Hyperostosis
Pallares	2011	Bns.Aires	57					4		4				
Morera	2011	Valencia	58		2 months–12 years			10						
Trabalzini	2011	Siena	59					2	2*					2 meningitis
Manrique	2011	Pamplona	60					4		4				
Kishore	2011	New Delhi	61					1		1	1			
Total of presented non-NF2 tumor ABI cases=							14	130	=Total of presented non-tumor ABI cases					
Total of presented non-NF2 ABI cases in world literature=							144							

A total of 144 non-NF2 ABI cases were distinguished. Papers sorted on clinic where the auditory brainstem implantation took place (four sets of rows: Verona, Paris, Freiburg, Seoul, and other centers). Total number of the presented series resembles the sum of the NF2, Vestibular Schwannoma (VS), and non-tumor patients presented in that paper. VS patients and non-tumor patients per clinic were calculated for each clinic. In the right columns, the non-tumor patients are divided in 'cochlear patency' problems (which are subdivided in the last column), nerve aplasia, malformed cochlea, head trauma (including nerve disruption), and auditory neuropathy. m = months, y = years, NF2 = neurofibromatosis type 2, HL = Hippel Lindau, BP = Bournevill-Pringle. Bns.Aires = Buenos Aires. The complete Table 2 is presented as Online 3a and the references as Online 3b

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

Otosclerosis

In severe retrofenestral otosclerosis, the temporal bone has otospongeotic lesions encompassing the otic capsule [17]. There is often intracochlear calcification, which is most prominent in the scala tympani. These lesions can hamper cochlear implantation and several complications can occur. First, malplacement of the electrode into a false lumen created by the otospongeotic ring around the cochlea may occur. Second, calcification of the scala tympani has to be bypassed and a scala vestibuli insertion or drill-out of the basal turn must be performed. Third, a CSF leak can occur during surgery, as the otic capsule is completely spongeotic with direct contact of the scalae to the CSF. The electrode can also have an entry and exit of the cochlea with this defect [18]. Fourth, facial nerve stimulation can occur postoperatively requiring reprogramming the electrode activity [18, 19].

Despite these potential complications, cases B, D, and E (Table 1; Figs. 1, 2) provide the experience and facts that support cochlear implantation as means of rehabilitation in patients with otosclerosis and severe to profound hearing loss. Difficulties with CI placement can be expected in cases of severe retrofenestral otosclerosis/otospongeosis. Therefore, these cases should be accurately evaluated and planned preoperatively. Recently, these severe retrofenestral otosclerosis patients have been proposed to be CI candidates, even before they fit standard CI criteria, because of the expected difficulties if surgery is postponed [20]. An indication for ABI before an attempt to place a CI seems incorrect.

Cochlear trauma and cochlear nerve disruption

After head trauma, two possible ways are mentioned in literature that could lead to total bilateral deafness and auditory brainstem implantation: a bilateral post-traumatic fracture of the cochlea or a cochlear nerve disruption. Several cases of post-traumatic deafness who received ABI (cases F to I) showed at least partial cochlear patency and have undergone a successful CI placement, as shown in Table 1 and Figs. 1, 3, and 4.

Cochlear trauma

When the otic capsule or the internal auditory canal is involved in the fracture, total deafness can occur. In very rare cases, both otic capsules could be fractured resulting in total bilateral deafness. As soon as the patient has recovered from this trauma, ossification of the cochlea should be assessed, similar to postmeningitis patients. For example, cases G and H had a partially obliterated labyrinth, but still enough lumen for a cochlear implant. In our opinion, there

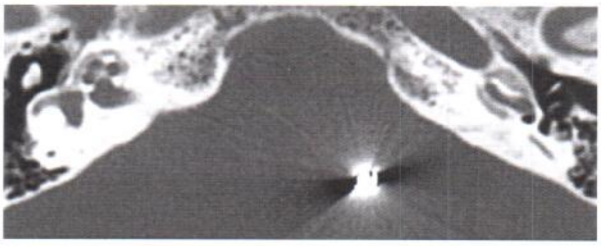

Fig. 2 Case *B*. Bilateral otospongeotic otic capsule but with a minimal obliterated cochlea. The left-side ABI is well seen, gave unsatisfactory results, and CI was still possible in this case. In otosclerosis CI seems, although sometimes difficult, always possible

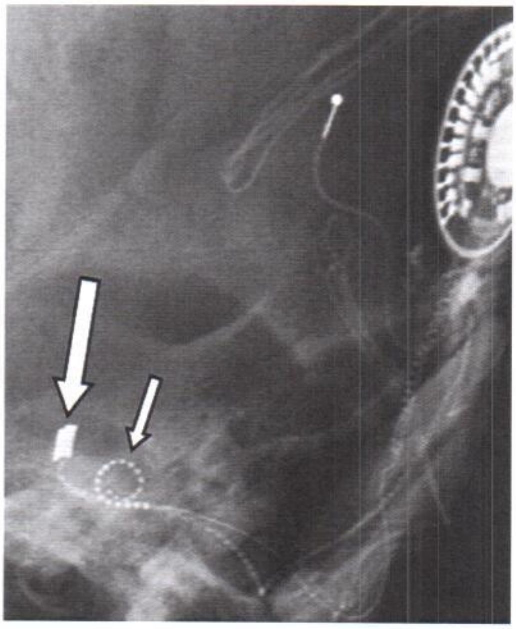

Fig. 3 Case *G*: X ray of the skull after placement of cochlear implant (*small arrow*). The patient received an ABI previously for the same indication (*big arrow*). Traumatic bilateral disruption of the cochlear nerve seems not a viable indication for ABI

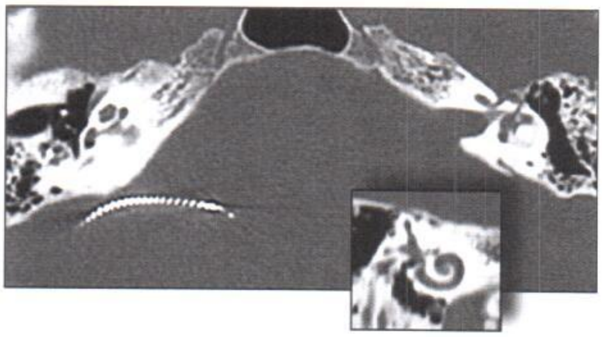

Fig. 4 Case *H*. CT scan of a patient after head trauma. The ossification of the lateral canal on the left side is visible. Both cochleae were patent on this scan (*bottom figure* and *insert*) even though an ABI is in place. Even though head trauma can lead to fractures and ossification, it seems that after MRI imaging (evaluating cochlear fibrosis) a CI attempt often is successful

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | LE | TYPESET
MS Code : EAORL-D-12-00785 | CP | DISK

is only an indication for ABI in cases with complete obliteration of both cochleae and after an unsuccessful cochlear drill-out attempt. There has not been a single case in the literature with bilateral complete ossification of the cochleae due to head trauma. Therefore, this indication for an ABI remains theoretical. Cochlear implantation seems to be the means of rehabilitation in cases of cochlear trauma, as proven in literature [21–23] and cases F–I.

Cochlear nerve disruption

The other theoretically valid indication for an ABI in literature due to trauma is bilateral disruption of the cochlear nerve [24]. This indication has also been mentioned in a consensus paper on the indications of ABI [25]. Surprisingly, in a literature review of all neuroradiological papers or books concerning this topic, there has only been one single case recently reported presenting a unilateral nerve rupture [26]. Cochlear nerve disruption *in both ears* has never been reported and seems practically impossible without fatal damage to the head and brain. Furthermore, cochlear nerve disruption is coupled with a facial nerve disruption [26] and could be clearly visible on MRI. Not a single case showed a nerve disruption on MRI or bilateral facial nerve disruption in the presented cochlear trauma cases (F–I). In literature, we could not find the existence of a complete rupture of the cochlear nerve in the internal auditory canal bilaterally among the survivors of serious head trauma. It seems, therefore, *very unlikely* to be an ABI indication, as MR images of a bilateral rupture have never been published and to our opinion it is not compatible with the survival of head trauma cases.

ABI indications with a patent cochlea?

In literature and in the presented cases, there are ABIs placed in patients with a patent cochlea, like in cases with Auto-immune inner ear disease (Cogan), von Hippel-Lindau disease, Bourneville-Pringle disease, Auditory neuropathy and idiopathic bony entrapment (hyperostosis). Each pathology will be discussed separately as they are all completely different and only have cochlear patency in common.

In autoimmune inner ear disease, we know that cochlear calcification rarely occurs, but bilateral fully obliterated cochleae in these cases have not been reported [27]. As shown in case J, a patient with Cogan syndrome and patent cochleae (Table 1; Fig. 1 and Online resource 4 and 5), CI will provide much better results than ABI.

In three more patients (K–M, Fig. 1), who were implanted with an ABI elsewhere due to different reasons of hearing loss (e.g., profound hereditary or idiopathic sudden SNHL), we have not seen a radiological, anatomical, or surgical reason why an attempt for a cochlear implant would not be feasible.

We believe as long as there is an intact nerve and an open cochlea at least on one side, there should be a CI attempted. Even if the result with a CI is not as good as predicted, reprogramming, replacing, or evaluating the other ear for CI should be done before even considering ABI, as CI gives us more predictable outcomes than ABI.

von Hippel-Lindau disease, Bourneville-Pringle disease, and auditory neuropathy are mentioned in literature to be ABI indications (Table 2, Online resource 3a & 3b). In all of these indications, it is not likely to have a bilateral obstructed cochlea or absent cochlear nerve. Therefore, an ABI indication is doubtful. The first two have not been described in detail, so it is hard to comment on the exact cases. In general, von Hippel-Lindau disease is associated with endolymphatic sac tumor, which can destroy the labyrinth. Still, successful bilateral CI placement in von Hippel Disease has been published [28]. A bilateral complete destruction of the labyrinth and cochlea seems very exceptional, but would be the only ABI indication in von Hippel-Lindau disease. It is not possible to comment on Bourneville-Pringle disease as an indication for ABI, as there were no hits in PubMed on 'Bourneville' AND 'hearing' or in other search strategies.

Auditory neuropathy (AN) is also mentioned as a primary ABI indication in the literature. This diagnosis is predominantly present with a normal cochlea and an intact nerve. As long as in general the results of an ABI seem less favorable and the literature shows average to good results with CI in AN patients, CI should be attempted first [29].

Furthermore, in AN cases with poor outcomes after CI, we believe that the poorer outcomes have to do with the development of the complex of cochlea, cochlear nerve, and cochlear nucleus. This is supported by the report of Jemec et al. [30] who showed that congenital facial nerve palsy is often due to a brainstem nucleus abnormality. This is felt to occur more often than previously believed and could be parallel to a congenital cochlear nucleus abnormality presenting as a peripheral deficiency.

Diseases in which the internal auditory canal is slowly narrowed by the progressive bone formation leading to entrapment of the cranial nerves, like hyperostosis, have been presented as an ABI indication [31]. The authors point out that, in their case and according to literature, decompression of the internal auditory canal followed by cochlear implantation should have been the first step before an ABI indication exists. Furthermore, the presented case [31] underlines the severe side effects of an intradural approach, like a retrosigmoid auditory brainstem implantation.

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK

Cochlear nerve aplasia and cochlear malformation

In the most common labyrinth malformations such as common cavity, incomplete partition type 2 and others, there is some form of cochlear lumen. It seems incorrect to proceed directly to an ABI in these cases, as performed in the case N with a common cavity on one side. Imaging showed a bilateral intact cochlear nerve and a patent cochlea contralateral (Table 1, Online resource 1, case N).

Sennaroglu has published the latest extensive description of all possible malformations of the labyrinth [32]. From this publication and our experience, it is clear that only in a Michel deformity there is no labyrinth available for cochlear implantation. There can be also an insufficient lumen for a CI when there is severe cochlear hypoplasia or aplasia. These cases can also be associated with a small internal auditory canal with hypoplasia or aplasia of the cochlear nerve on imaging [33]. Still, two recent publications point out that the absence of a visible cochlear nerve on imaging does not preclude auditory innervation of the cochlea [34, 35]. Cochlear implantation can be a valuable option for patients with apparent cochlear nerve aplasia as long as they have undergone appropriate testing [34]. Electrically evoked ABR is critical in the evaluation of this patient group [34, 36, 37]. If there is proof of a bilateral absent cochlear nerve fibers, bilateral complete cochlear aplasia, or bilateral Michel deformity, ABI is the only solution to secure any chances on hearing development [33]. It seems that in the majority of labyrinth malformations there is *no* indication for an ABI. In addition, if imaging is pointing out an absence of cochlear nerve, testing still has to be done to prove an absence of nerve fibers running along other nerves in the internal auditory meatus.

In summary, due to the fact that at this moment the ABI results are unpredictable and overall much worse than cochlear implantation results, we believe the following:

- CI seems indicated in case with normal cochlea and acoustic nerve present. ABI seems contraindicated.
- CI seems indicated in solitary vestibular schwannoma in the only hearing ear. ABI is not primarily indicated [3].
- In cases with fully ossified cochleae on scanning, the decision to go directly for ABI or first try a cochlear drill-out procedure seems a matter of personal preference.
- CI seems indicated in post-traumatic deafness. ABI seems contraindicated [23].
- Cochlear nerve disruption seems not to exist bilaterally in head trauma survivors [26].
- CI seems indicated in most of the malformations of the cochlea (e.g., common cavity) and ABI only indicated in the absence of the cochlear nerve proven on scanning and in testing [34, 35].
- Bilateral complete cochlear aplasia and inner ear aplasia (Michel deformity) are ABI indications.

Conclusion

Due to the better results with cochlear implantation, ABI rehabilitation should be restricted for those patients who have no other rehabilitation options. Patency of the cochlea as well as an intact and functional cochlear nerve needs to be examined and sometimes explored before an ABI indication exists. We have shown that in many cases a CI is still possible, and CI provided better results than ABI. In vestibular schwannoma in the only hearing ear, cochlear otosclerosis, temporal bone fractures, (presumed) bilateral traumatic cochlear nerve disruption, auto-immune inner ear disease and auditory neuropathy primarily CI are indicated. In cochlear nerve aplasia, testing should be done before an ABI indication exists. ABI indications only exist in cases of severe cochlear hypoplasia or aplasia. Cochlear nerve disruption bilaterally is very unlikely to exist.

Acknowledgments We like to thank Spencer Voth, ENT surgeon, USA, for his excellent help.

References

1. Schwartz MS, Otto SR, Brackmann DE, Hitselberger WE, Shannon RV (2003) Use of a multichannel auditory brainstem implant for neurofibromatosis type 2. Stereotact Funct Neurosurg 81(1–4):110–114
2. Sanna M, DiLella F, Guida M, Merkus P (2012) Auditory brainstem implants in NF2 patients. Results and review of the literature. Otol Neurotol 33(2):154–164
3. Di Lella F, Merkus P, Di Trapani, G, Taibah A, Guida M, Sanna M (2012) Vestibular schwannoma in the only hearing ear: the role of the cochlear implant. Ann Otol Rhinol Laryngol (in press 12-5422)
4. Thedinger BA, Cueva RA, Glasscock ME III (1993) Treatment of an acoustic neuroma in an only-hearing ear: case reports and considerations for the future. Laryngoscope 103(9):976–980
5. Ramsden R, Khwaja S, Green K, O'Driscoll M, Mawman D (2005) Vestibular schwannoma in the only hearing ear: cochlear implant or auditory brainstem implant? Otol Neurotol 26(2): 261–264
6. Yamakami I, Yoshinori H, Saeki N, Wada M, Oka N (2009) Hearing preservation and intraoperative auditory brainstem response and cochlear nerve compound action potential monitoring in the removal of small acoustic neurinoma via the retrosigmoid approach. J Neurol Neurosurg Psychiatry 80(2):218–227
7. Aristegui M, Denia A (2005) Simultaneous cochlear implantation and translabyrinthine removal of vestibular schwannoma in an only hearing ear: report of two cases (neurofibromatosis type 2 and unilateral vestibular schwannoma). Otol Neurotol 26:205–210
8. Grayeli AB, Bouccara D, Kalamarides M, Ambert-Dahan E, Coudert C, Cyna-Gorse F, Sollmann WP, Rey A, Sterkers O

(2003) Auditory brainstem implant in bilateral and completely ossified cochleae. Otol Neurotol 24(1):79–82

9. Grayeli AB, Kalamarides M, Bouccara D, Ambert-Dahan E, Sterkers O (2008) Auditory brainstem implant in neurofibromatosis type 2 and non-neurofibromatosis type 2 patients. Otol Neurotol 29(8):1140–1146
10. Sollmann WP, Laszig R, Marangos N (2000) Surgical experiences in 58 cases using the nucleus 22 multichannel auditory brainstem implant. J Laryngol Otol Suppl 27:23–26
11. Sanna M, Khrais T, Guida M, Falcioni M (2006) Auditory brainstem implant in a child with severely ossified cochlea. Laryngoscope 116(9):1700–1703
12. Merkus P, Free RH, Mylanus EAM, Stokroos R, Metselaar M, van Spronsen E, Grolman W, Frijns JHM (2010) Dutch Cochlear Implant Group (CION) consensus protocol on postmeningitis hearing evaluation and treatment. Otol Neurotol 31:1281–1286
13. Steenerson RL, Gary LB, Wynens MS (1990) Scala vestibuli cochlear implantation for labyrinthine ossification. Am J Otol 11:360–363
14. Bacciu S, Bacciu A, Pasanisi E, Vincenti V, Guida M, Barbot A, Berghenti T (2002) Nucleus multichannel cochlear implantation in partially ossified cochleas using the Steenerson procedure. Otol Neurotol 23(3):341–345
15. Kemink JL, Zimmerman-Phillips S, Kileny PR, Firszt JB, Novak MA (1992) Auditory performance of children with cochlear ossification and partial implant insertion. Laryngoscope 102(9): 1001–1005
16. Lenarz T, Battmer RD, Lesinski A, Parker J (1997) Nucleus double electrode array: a new approach for ossified cochleae. Am J Otol 18(6 Suppl):S39–S41
17. Rotteveel LJ, Proops DW, Ramsden RT, Saeed SR, van Olphen AF, Mylanus EA (2004) Cochlear implantation in 53 patients with otosclerosis: demographics, computed tomographic scanning, surgery, and complications. Otol Neurotol 25:943–952
18. Ramsden R, Bance M, Giles E, Mawman D (1997) Cochlear implantation in otosclerosis: a unique positioning and programming problem. J Laryngol Otol 111(3):262–265
19. Toung JS, Zwolan T, Spooner TR, Telian SA (2004) Late failure of cochlear implantation resulting from advanced cochlear otosclerosis: surgical and programming challenges. Otol Neurotol 25(5):723–726
20. Merkus P, van Loon MC, Smit CF, Smits C, de Cock AFC, Hensen EF (2011) Decision making in advanced otosclerosis: an evidence-based strategy. Laryngoscope 121:1935–1941
21. Camilleri AE, Toner JG, Howarth KL, Hampton S, Ramsden RT (1999) Cochlear implantation following temporal bone fracture. J Laryngol Otol 113(5):454–457
22. Serin GM, Derinsu U, Sari M, Gergin O, Ciprut A, Akdaş F, Batman C (2010) Cochlear implantation in patients with bilateral cochlear trauma. Am J Otolaryngol 31(5):350–355
23. Simons JP, Whitaker ME, Hirsch BE (2005) Cochlear implantation in a patient with bilateral temporal bone fractures. Otolaryngol Head Neck Surg 132(5):809–811
24. Colletti V, Carner M, Miorelli V, Colletti L, Guida M, Fiorino F (2004) Auditory brainstem implant in posttraumatic cochlear nerve avulsion. Audiol Neurootol 9(4):247–255
25. Sennaroglu L, Colletti V, Manrique M, Laszig R, Offeciers E, Saeed S, Ramsden R, Sarac S, Freeman S, Andersen HR, Zarowski A, Ziyal I, Sollmann WP, Kaminsky J, Bejarano B, Atas A, Sennaroglu G, Yucel E, Sevinc S, Colletti L, Huarte A, Henderson L, Wesarg T, Konradsson K (2011) Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. Otol Neurotol 32(2):187–191
26. Corrales CE, Monfared A, Jackler RK (2010) Facial and vestibulocochlear nerve avulsion at the fundus of the internal auditory canal in a child without a temporal bone fracture. Otol Neurotol 31(9):1508–1510
27. Aftab S, Semaan MT, Murray GS, Megerian CA (2010) Cochlear implantation outcomes in patients with autoimmune and immune-mediated inner ear disease. Otol Neurotol 31(8):1337–1342
28. Jagannathan J, Lonser RR, Stanger RA, Butman JA, Vortmeyer AO, Zalewski CK, Brewer C, Surowicz C, Kim HJ (2007) Cochlear implantation for hearing loss associated with bilateral endolymphatic sac tumors in von Hippel-Lindau disease. Otol Neurotol 28(7):927–930
29. Breneman AI, Gifford RH, Dejong MD (2012) Cochlear implantation in children with auditory neuropathy spectrum disorder: long-term outcomes. J Am Acad Audiol 23(1):5–17
30. Jemec B, Grobbelaar AO, Harrison DH (2000) The abnormal nucleus as a cause of congenital facial palsy. Arch Dis Child 83(3):256–258
31. Waterval JJ, Stokroos RJ, Dings J, Van Overbeeke JJ, Manni JJ (2011) Cerebral vasospasm after auditory brainstem implantation in a patient with hyperostosis cranialis interna. Clin Neurol Neurosurg 113(10):904–908
32. Sennaroglu L, Saatci I (2002) A new classification for cochleovestibular malformations. Laryngoscope 112(12):2230–2241
33. Sennaroglu L, Ziyal I, Atas A, Sennaroglu G, Yucel E, Sevinc S, Ekin MC, Sarac S, Atay G, Ozgen B, Ozcan OE, Belgin E, Colletti V, Turan E (2009) 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 30(6):708–715
34. Warren FM III, Wiggins RH III, Pitt C, Harnsberger HR, Shelton C (2010) Apparent cochlear nerve aplasia: to implant or not to implant? Otol Neurotol 31(7):1088–1094
35. Song MH, Kim SC, Kim J, Chang JW, Lee WS, Choi JY (2011) The cochleovestibular nerve identified during auditory brainstem implantation in patients with narrow internal auditory canals: can preoperative evaluation predict cochleovestibular nerve deficiency? Laryngoscope 121(8):1773–1779
36. Colletti V, Shannon RV, Carner M, Veronese S, Colletti L (2010) Complications in auditory brainstem implant surgery in adults and children. Otol Neurotol 31(4):558–564
37. Beltrame MA, Bonfioli F, Frau GN (2000) Cochlear implant in inner ear malformation: double posterior labyrinthotomy approach to common cavity. Adv Otorhinolaryngol 57:113–119

Journal : Large 405 | Dispatch : 1-2-2013 | Pages : 11
Article No. : 2378 | ☐ LE | ☐ TYPESET
MS Code : EAORL-D-12-00785 | ✔ CP | ✔ DISK