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Consensus statement: Long-term results of ABI in children with complex inner ear malformations and decision making between CI and ABI

Levent Sennaroğlu¹, Vittorio Colletti², Thomas Lenarz³, Manuel Manrique⁴, Roland Laszig⁵, Helge Rask-Andersen⁶, Nebil Göksu⁷, Erwin Offeciers⁸, Shakeel Saeed⁹, Robert Behr¹⁰, Yıldırım Bayazıt¹¹, Jan Casselman^{12,13,14}, Simon Freeman¹⁵, Paul Kileny¹⁶, Daniel J. Lee¹⁷, Robert V. Shannon¹⁸, Mohan Kameswaran¹⁹, Abdulrahman Hagr²⁰, Andrzej Zarowski⁸, Mark S. Schwartz¹⁸, Burçak Bilginer²¹, Ameet Kishore²², Gonca Sennaroğlu¹, Esra Yücel¹, Sarp Saraç¹, Ahmet Atas¹, Lilian Colletti², Martin O'Driscoll¹⁵, In Seok Moon²³, Lutz Gärtner³, Alicia Huarte⁴, Gunnar Nyberg²⁴, Burçe Özgen Mocan²⁵, Gamze Atay¹, Münir Demir Bajin¹, Betül Çicek Çınar¹, Merve Özbal Batuk¹, Mehmet Yaralı¹, Fatma Esen Aydın¹, Filiz Aslan¹, Meltem Cigdem Kirazlı¹, Hilal Burcu Özkan¹, J. M. Hans²⁶, Julie Kosaner²⁷, Marek Polak²⁸

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Introduction

'Second Consensus Meeting on Management of Complex Inner Ear Malformations: Long Term Results of ABI in Children and Decision Making Between CI and ABI' took place on 5–6 April 2013 in Kyrenia, Northern Cyprus with the participation of 20 centers from 11 countries. These centers presented their auditory brainstem implantation (ABI) experience in children and infants and also provided

the selection criteria and cochlear implant (CI) results in patients with an abnormal cochlea and nerve. In addition, Cochlear and Med El companies provided presentations regarding solutions to problems during revision surgery and future projections of device technology.

According to the results from different centers, it was evident that an ABI is capable of providing hearing sensation in prelingually deafened children

with complex inner ear malformations and diseases. It is possible to obtain a pure tone average with an ABI between 30 and 60 dB HL in most of these patients. It has been observed that the majority of children obtain Categories of Auditory Performance (CAP) scores around 5, but occasionally certain ABI users obtained scores of up to CAP 8-II, NEAP-Nottingham Early Assessment Package. The Ear Foundation 2009). CAP scores tend to increase with earlier implantation, i.e. under 2 years of age. However, after 3 years of use, it usually reaches a plateau. Children with additional disorders cannot reach CAP scores of children without disorders. However, they obtain an improvement of cognitive functions but duration of ABI use and increasing chronological age also play a role in this improvement. It is also possible to develop closed and limited open set speech discrimination. In the current large series of 35 children with at least 1 year of follow up, 80% of children achieved scores above 50% with closed set pattern discrimination test, while approximately 30% reached maximum scores. Similarly, 30% of patients scored above 50% with open set sentence recognition test. It is commonly noted by clinicians that pediatric ABI users tend to only pay attention to sound in structured, quiet situations. They appear to have really to concentrate to hear and recognize sounds. Because of this, ABI users take longer than hearing aid or CI users to spontaneously respond to sound in their environments. Most ABI users cannot benefit from listening to environmental sounds which severely restricts their exposure to meaningful, spoken language.

When speech production outcome is taken into account, the children have low speech intelligibility scores. As hearing impaired, prelingually deafened children need to be exposed to an abundance of rich, spoken language to learn to talk, it seems unlikely that ABI users will develop spoken language easily. It is obvious that the ABI definitely supports the development of language, however, there is still a gap between the chronological age and language age. After long-term ABI use, expressive and receptive language development is behind peers with normal hearing and cochlear implantees; it is not possible to expect similar improvement in these children with complex inner ear malformations. They will need intensive auditory oral habilitation and an extremely supportive family. In addition, sign language and lipreading might be complementary for daily communication; therefore, it is advisable to consider use of both sign language and lipreading in pediatric ABI recipients. It should be noted that achieving a certain level of communication with visual enhancement can be considered as success in this particular patient group. Moreover, the

majority of the patients use their device daily (with less than 10% device nonuser ratio), which also suggests that these children achieve a certain level of benefit from this procedure, although progress is slower and their peak performance is inferior to most CI recipients. There may be some patients with no response. These points should be clearly mentioned in the informed consent to the family and it should be stressed that, generally speaking, the results are poorer than for CI recipients. However, it is possible to achieve sound detection, although language development is usually delayed and incomplete. Managing family expectancy and helping the family realize the level of commitment required is of foremost importance in patient counseling.

Different centers indicated that the type of the underlying medical condition, age at ABI activation, associated disabilities and number of active electrodes are among the significant prognostic factors. It must be pointed out that our comparison of outcome in children fitted with CIs and ABI has been performed without considering the severe associated disabilities that characterized most of the children fitted with ABI in this cohort. One centre in the present study reported that when the two groups of children (CIs and ABIs) are matched for age of implantation and presence or absence of disability, their results showed that children with ABI obtained similar outcomes as children using CI. The rest of the clinics reported that children with ABI could not reach the level of children using CI.

Timing of surgery is very important. As in children with CI, brain plasticity is very important in pediatric ABI. For this reason ideally ABI surgery should be done before the age of 2 and when possible between 1 and 1.5 years. It is highly unlikely that older children will be able to develop language skills. Implanting older children may result in unsuccessful outcomes running the risk of discrediting this method of treatment. Because of the increased neurosurgical risks only perfectly healthy children can undergo ABI around 1 year of age in experienced centers. This is also influenced by the experience of the center. Diagnosis, follow up and family counseling is likely to take up to a year so the child will be ideally ready for ABI surgery by the age of 1.

Contrarily, when the decision has been made to apply a CI first, than this should be performed around the age of 1 year in order to allow approximately 1 year for evaluation of the auditory development with CI (see also further in the section 'Postoperative evaluation') and in the case of insufficient benefit from CI, switching to ABI before the age of 24 months.

The participating centers reported minor complications. However, as in intracranial cerebellopontine angle (CPA) surgery, ABI in children carries more risks, including severe morbidity and even mortality, when compared to cochlear implantation and should be performed in centers which are experienced in CI surgery and neurosurgical procedures in the CPA together with audiological experience.

Success with an ABI decreases considerably in children with additional needs such as intellectual or cognitive deficits and visual impairment. On the other hand, these children are in great need of hearing to establish a new communication mode. The severity of the additional need is critical. If the pathology is so severe that it will interfere with the education of the child with ABI, decisions should be made with caution and certain cases might not be regarded as candidates. Therefore, counseling should be done accordingly. However, visual impairment should not be considered as a contraindication, as it is not such a major obstacle as impaired intellectual ability.

Audiological evaluation

As described below in the radiology section, ABI in children is indicated in two main groups of pathologies. Detailed audiological examination is necessary in all cases to check the potential benefit for a CI.

Preoperative evaluation

Behavioral testing

Behavioral audiological examination is very important in candidacy evaluation and depending on the age of child, behavioral observation audiometry, visual response audiometry, or play audiometry can be performed. Behavioral testing should be done by an experienced pediatric audiologist. Testing each ear with insert earphones, besides free field testing, is very important. Audiological responses can be helpful in determining the presence and function of an auditory nerve. If there is a response on one side, this can be predictive for the possible presence of a cochlear nerve and it is important that a CI trial is carried out particularly in patients with cochlear nerve deficiency.

Objective testing

A major objective test battery includes otoacoustic emission (OAE) testing, auditory brainstem response testing (ABR), auditory steady state response (ASSR), and acoustic reflexes. When possible, and appropriate, middle-latency, and cortical auditory evoked potentials should also be considered. OAE (transient-TEOAE or distortion-product-DPOAE) evaluates the function of outer hair cells in the cochlea. ABR testing provides information about the function of the auditory pathway from the distal

portion of the cochlear nerve to the nucleus of the lateral lemniscus. In patients with hypoplastic cochlear nerves, at times, cochlear microphonics (CM) may be the only electrophysiological responses identified during ABR testing. In addition to the CM, TEOAE, or DPOAE may also be present. Caution is advised in interpreting the presence of a CM or an OAE response as indicating the presence of normal outer hair cells, especially if these respective responses are only present at very high stimulus levels: the latter can also be the case in severe to profound sensorineural hearing loss due to inner hair cell dysfunction or perisynaptic audiopathy without a hypoplastic or absent cochlear nerve.

Electrically evoked ABR (eABR)

Ideally, transtympanic or through round window, eABR should be performed before CI or ABI surgery in patients with a suspicion of or confirmed hypoplastic cochlear nerves. Preoperatively, electrical stimulation can be useful as a prognostic indicator of outcomes in these cases. Although it is a great counseling and prognostic tool, complete absence of eABR should be further confirmed by imaging studies to ascertain an absent cochlear nerve. A positive response with eABR will be deterministic for the implant team to choose CI rather than ABI. But in the case of an absent response on preoperative, transtympanic promontory eABR, this needs to be correlated with temporal bone and cochlear nerve anatomy. It has been indicated that when eABR thresholds are low and the amplitude of wave V is high during preoperative testing, speech outcome is better. However, eABR results do not always correlate with specific outcomes of CI and ABI patients. It is of note that in addition to threshold ranges, and presence/absence of a response, response adaptation has been shown to correlate with a present, but hypoplastic or degenerated cochlear nerve. Adaptation is indicated by decreased eABR amplitudes, or a complete disappearance of the response on consecutive responses to the same stimulus level. The other signs of possible adaptation are the lack of improvement of the S/N ratio with increased number of the averages, or a decrease in the response amplitude at higher stimulation rates. Presence of neural adaptation may result in suboptimal, inconsistent benefit from a CI, and may or may not be an indication for an ABI, instead of a CI.

Improved quality of eABR registration can be achieved by using general anesthesia with muscle relaxants in order to decrease the influence of the myogenic potentials.

Educational assessment and follow up

Children who are being considered for ABI candidacy should be evaluated in terms of communication and learning behavior. Their participation in clinical and educational tasks is decisive for postoperative progress and building of a therapy program. Evaluation involves their communication attempts such as eye contact, speech reading, turn taking, initiating, and sustaining joint attention, following directions, etc. Although the main aim of ABI is to provide meaningful auditory input and help a child transpose these signals into receptive and expressive language, these nonverbal forms of communication reflect the child's ability to structure a particular communication method.

Children with cochlear nerve hypoplasia using CIs should be followed up in terms of development of behavioral and objective auditory performances. Those who have had at least 1 year of CI experience and participated in intensive auditory-verbal therapy sessions are expected to develop some open set identification of common phrases, such as 'all gone', 'here daddy comes', and 'do you want some milk?'. At the same time, eABRs or electrically evoked cortical responses (eCR) should be performed in order to confirm that the device is indeed stimulating the brainstem and the cortical areas. The end point for auditory perception skills is generally defined only by detection of speech and environmental sounds and closed set pattern and word perception. If the child is not performing successfully in auditory tasks (see also further in the section 'Postoperative evaluation') and learning by auditory processing within 1 year of CI use, the options for this child are an ABI in the contralateral ear or explanation of the CI and implantation of an ABI in the ipsilateral ear. In any case, for these children the therapeutic program should mainly be structured by auditory learning tasks where visual cues are used in a compensatory way.

Intraoperative evaluation

(1) Evoked compound action potentials (ECAP): neural response telemetry (NRT)/auditory response telemetry (ART)/neural response imaging (NRI) measurements should be performed during CI surgery. Electrically elicited stapedius reflex threshold measures (ESRT) performed intra-operatively will also provide information about whether the implant brings about hearing through stimulation of the hearing nerve. Typically, it is not possible to obtain ECAP's or ESRT's in cases of severe cochlear malformations like cochlear hypoplasia or incomplete partition type I. In cases of normal cochlear anatomy with hypoplastic cochlear nerve,

observing NRT/ART/NRI, ESRT is also rarely possible.

- (2) eABR: An intracochlear test electrode has been used in a limited number of patients during surgery to observe electrical responses with stimulation. This procedure necessitates a complete, facial recess approach CI surgery and using a test electrode before deciding to change the surgical approach to do an ABI. Such a test electrode may be useful in decision making between CI and ABI. If there is a positive eABR response, CI can be performed in the same setting.
- (3) Intraoperative monitoring, with eABR is to be used in all cases with ABI electrode stimulation as an indicator of appropriate placement. According to the findings slight adjustment to the final position of the ABI electrode is done. More than six active electrodes showing auditory stimulation are prognostic for a better outcome. Cranial nerves VII and IX can be monitored using EMG.

Postoperative evaluation

This is particularly important in patients with probable indications who already underwent CI surgery and are undergoing a trial period. It is important to decide whether the child is making sufficient progress with a CI. When results are negative, a decision to go ahead with an ABI should be done as early as possible. Delaying the ABI will decrease the benefit and therefore, timing is very important.

- (1) eCR: aided cortical assessment with a CI will provide information about whether or not the CI is assessing the user to adequate sound and allowing for auditory maturation. It is a valuable assessment tool to use when evaluating benefit of a CI before planning of an ABI. Low, mid, and high frequency regions can be screened using speech stimuli such as /t/, /m/ or /g/ at e.g. a dB SPL of 55 representing the level of quiet conversational speech. Presence of a P1 with decreasing latency at consecutive testing will indicate activation at cortical level is achieved by the CI.
- (2) eABR: postoperative eABR thresholds obtained with CI stimulation, typically correlate well with the supra-threshold so-called 'comfort levels' for each CI electrode. Response morphology can show neural maturation over time. eABR thresholds along with behavioral results, including speech and language development can be predictive of ABI need.
- (3) APE test: evaluation of the speech and language development with CI is crucial before taking the decision for potential switching to ABI. This must be performed before the age of 24 months, when most of the children show no language skills sufficient to perform the speech identification tests. In order to overcome this problem an auditory phoneme discrimination (APE) test was developed at the University ENT Department of AZ Sint Augustinus in Antwerp (currently the European

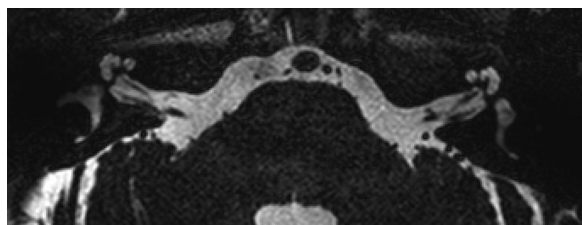


Figure 1 High-resolution image through the cochlear branch of the vestibulocochlear nerve, slices thickness = 0.25 mm.

Institute for ORL-HNS). This test evaluates detection and discrimination of the most important phonemic contrasts. The test is performed in the same manner as the child audiometry using the conditioned orientation reflex paradigm and is feasible already from the age of 7 to 8 months. The results of the APE test correlate well with the results of speech identification tests, while the APE test does not presume language skills and is generally independent of the mother language and the cognitive strength of a child.

- (4) Educational assessments: The major part of the follow up is educational assessment. Improvement in auditory skills and language must be the essential guide for the implant team.

If the team observes a continuous development with CI, ABI is not indicated. However certain cases show initial progress but then users come to a plateau and do not progress any more after this. This is most probably due to the capacity of the deficient cochlear nerve. ABI is also indicated in these patients to improve their auditory skills (1).

As can be seen, children with CN deficiency present a challenge for the decision-making process.

Useful measures to shorten the time interval include subjective assessments in the form of questionnaires allowing for collection of information about the child's functioning from family members and teachers, e.g. MAIS, Little EAR Auditory Questionnaire (LEAQ), and FAPCI. Scales and profiles such as CAP, SIR, and MSLDS should also be implemented. Use of such questionnaires and profiles both before and after ABI will help to define who is a candidate for ABI and standardize reporting of outcomes after ABI. Measures of sound field implant thresholds will also provide useful information about CI benefit along with results of objective tests such as changes in eABR latency and amplitude.

Radiological evaluation

The following congenital temporal bone anomalies identified on routine preoperative CI imaging raise a suspicion of a significantly hypoplastic, or absent cochlear branch of cranial nerve VIII, and should trigger additional imaging studies geared to specifically determine presence or absence of a cochlear nerve.

Radiological indications

In the previous consensus meeting, well-defined congenital indications were indicated and these are once again accepted as definite indications. These are:

- (1) Complete labyrinthine aplasia (Michel aplasia)
- (2) Cochlear aplasia
- (3) Cochlear nerve aplasia
- (4) Cochlear aperture aplasia

Probable indications:

- (1) Hypoplastic cochleas with cochlear aperture hypoplasia
- (2) Common cavity and incomplete partition type I cases with or without cochlear nerve
- (3) Patients with an unbranched cochleovestibular nerve
- (4) The hypoplastic cochlear nerve

Preoperative radiology and audiology protocols need to be discussed in great detail in these patients. It is necessary to show cochlear nerve presence by MRI and show its functional capability through audiological tests.

Imaging protocol for patients with VIIIth nerve aplasia/hypoplasia/dysplasia

The evaluation of the cochleovestibular nerve and especially its cochlear branch is of extreme importance prior to cochlear implantation. Differentiation between aplasia-hypoplasia and hypoplasia and a normal size of the cochlear branch can be very difficult and requires the highest possible resolution. The presence of a small internal auditory canal can make the diagnosis even more difficult. Visualization of the VIIIth nerve and its branches is performed on MR and when we are dealing with a CI-candidate or when the patient has congenital sensorineural hearing loss the following protocol is used.

MR – protocol

- (1) The patient is examined at the highest possible resolution.
- (2) The resolution of the heavily T2 weighted sequence in the axial plane is increased. The spatial resolution becomes: 0.29×0.29 mm in plane with a thickness of 0.25 mm (Fig. 1). The acquisition time is 10 min.
- (3) When an abnormality is seen or suspected on these axial images then *direct parasagittal images* with the same heavily T2 weighted sequence are made perpendicular on the nerves in the internal auditory canal and CPA. This is done on both sides so that bilateral pathology can be recognized or comparison with a normal side is possible (2×10 min again). These direct images have a better resolution and are sharper than reconstructed images which are acquired using the axial images. The angulation perpendicular on the nerves is set out on the original axial series and on coronal low resolution localizer images (Figs. 2–4).
- (4) In *young children* sufficient quality can only be acquired *under general anesthesia*.

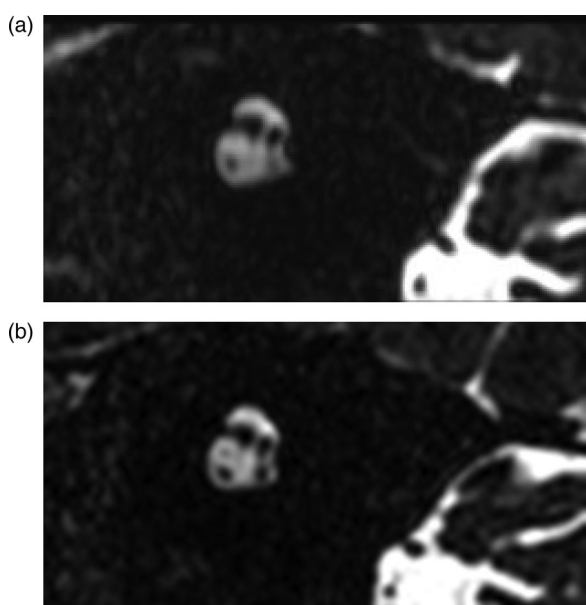


Figure 2 (A) Reformatted (right IAC). (B) Direct parasagittal (right IAC).

CT – protocol

High resolution is performed to confirm the presence of a canal for the cochlear nerve between the fundus of the internal auditory canal and the modiolus at the base of the cochlea. This is better seen at high resolution and can best be done with the lowest possible radiation burden as the patients are often very young. For both reasons *Cone Beam CT is preferred* over Multidetector conventional CT and boosts the resolution from 0.625/0.5 mm for MDCT to 0.15 mm for CBCT (Fig. 5). Again, in young children this is best performed *under anesthesia* (CT and MR should be performed during the same anesthesia). However, when good quality CBCT is not available (Newtom 5G, Accuitomo 170, CS 9300), then it is better to use MDCT as most of the other CBCT systems are not even able to provide the quality of MDCT.

In spite of these extensive imaging studies, there are patients with hypoplastic cochlear nerve who demonstrate sound detection with a CI at around 30–40 dB with very limited language development. These patients may benefit from an ABI. It is very important to shorten the time interval between CI and ABI in these children. Measures leading to decreasing the time interval are highlighted in the section on audiological postoperative evaluation.

This situation may be an indication for sequential CI and ABI surgery. Usually, at least a year is necessary to assess and decide whether the patient benefits from CI or not. In this type of situation, usually a contralateral ABI is decided on.

An alternative option is simultaneous CI and contralateral ABI. CI on the side with better cochlear

(surgically implantable) and cochlear nerve morphology and ABI on the contralateral side with worse anatomy. This may provide initially the best possible bilateral stimulation option for these patients. This approach however should be reserved for cases where there is clear anatomical and physiological evidence for the presence of a hypoplastic cochlear nerve on one side, and its absence on the contralateral side.

Another alternative approach might be use of ABCI where one electrode is in the cochlea and the other one is in the brainstem. This provides the option to stimulate both sites from the same audioprocessor. A disadvantage of this procedure may be the need to revise both electrode ends in case of the device failure and higher risk of meningitis if the child develops a mastoiditis due to the continuity of the ABI electrode with the CSF of the CPA. We have to mention that this has not been done in the pediatric population so far. A detachable electrode design may resolve this issue however, currently it is not available due to technological issues.

Contraindications

Children with special needs usually yield poorer performance with ABI. Severity of these additional needs is critical. In cases with minor disabilities, decision should be made in favor of surgery to improve the communication skills of the child. In situations where the handicaps are very complex, the probability that the child will benefit from ABI is very low. Intermediate cases should be evaluated together with other subspecialties such as pediatric psychiatry, neurology, speech-language pathology, genetics, pediatrics, and indications should be evaluated by the whole team.

Complications

This surgery should be performed in centers experienced in CI and neurotological surgeries. In these settings, the potential complications are rare in children. However, if a complication occurs additional medical and surgical interventions will be needed. ABI complications are usually related to NF-2 patient group which may be attributable to the underlying disease rather than ABI electrode placement.

Revisions: ABI revision was carried out in very limited number of children. In certain patients, the electrode was easily tracked, dissected, and removed completely. In other instances, it was fixed to the brainstem and could not be dissected out. One patient had a life threatening episode during revision surgery, and experienced neurological sequels involving lower cranial nerves following revision surgery.

The two major reasons for revision surgery are:

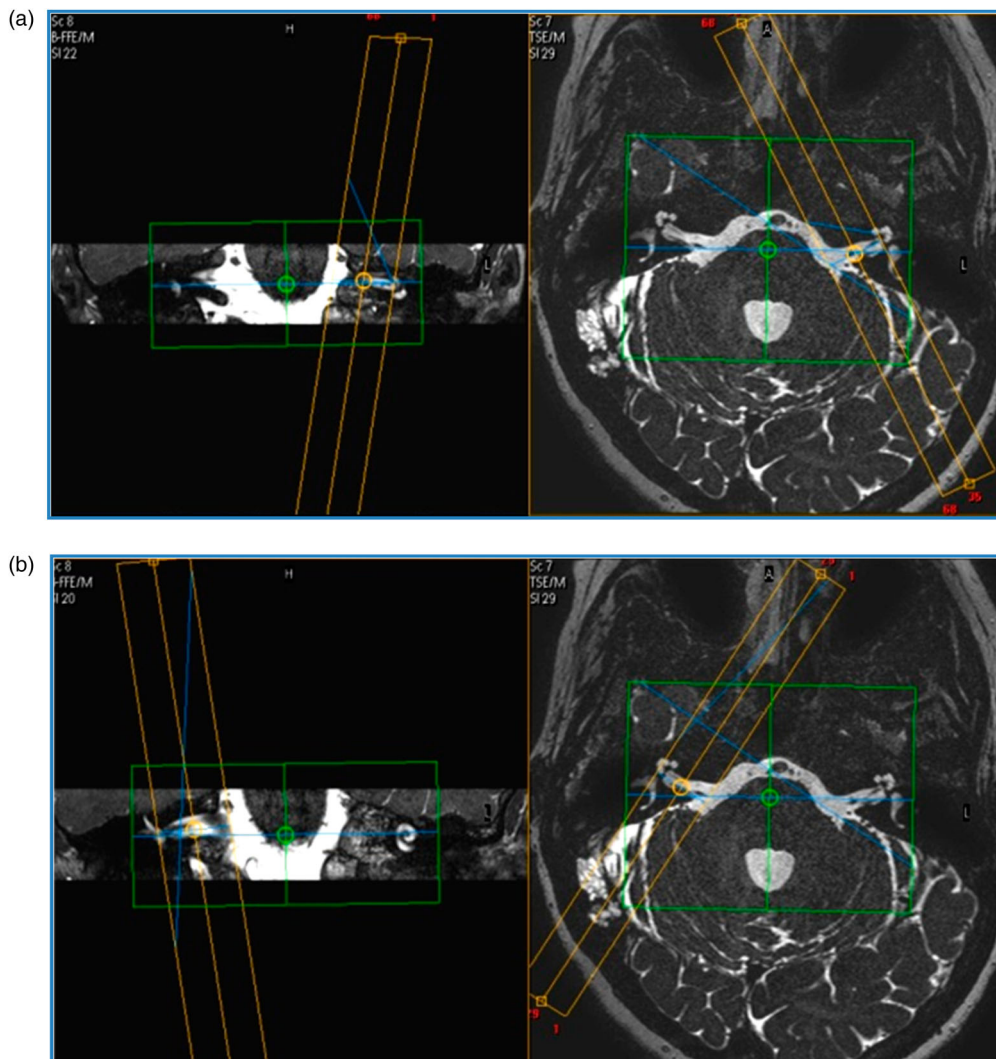


Figure 3 Parasagittal direct images set out on the axial images and coronal localizer images.

Device malfunction: The companies are urged to find technical alterations of the device itself enabling revision surgery without removing of the electrode from the brainstem. A detachable electrode where the implant body can be changed without removing the electrode from the lateral recess will be ideal for cases with device failure. However, the authorities expressed their opinion that a detachable electrode may bring other risks to the patient.

Electrode migration: Every effort should be made to avoid migration. It is advisable to place the electrode fully into the lateral recess. Trimming the mesh around the electrode is done by many surgeons and the electrode is supported by soft tissue at the back of the plate electrode in the lateral recess to avoid device migration and create good contact with the cochlear nucleus area in front. Usually, 1 mm of mesh around the electrode appears to be sufficient. Leaving the entire mesh intact around the electrode may create excessive fibrosis causing

integration of the plate electrode into the surrounding neural tissue. Then the electrode is positioned so that there will be excess lead between the bed and the lateral recess. In the case of a tense (stiff) electrode lead, the implant may be dislocated out of the recess when the child grows up. The torque on the electrode lead may cause device migration too.

In children it is more likely there will be electrode migration due to head growth, however, in patients with large foramen of Lushka or lateral recess (such as in adults) this is more likely to happen. In the majority of the patients, there are side effects due to the stimulation of nearby cranial nerves such as CN V, VII, IX, and X. In cases with severe side effects, electrode migration should be suspected (intraoperative eABR should be used for correct placement particularly in large recesses, however due to limited contact, eABR may be initially negative and later on when the cerebellum is repositioned to normal location adequate contact enables an eABR response). Intraoperative absence of eABR does not always have

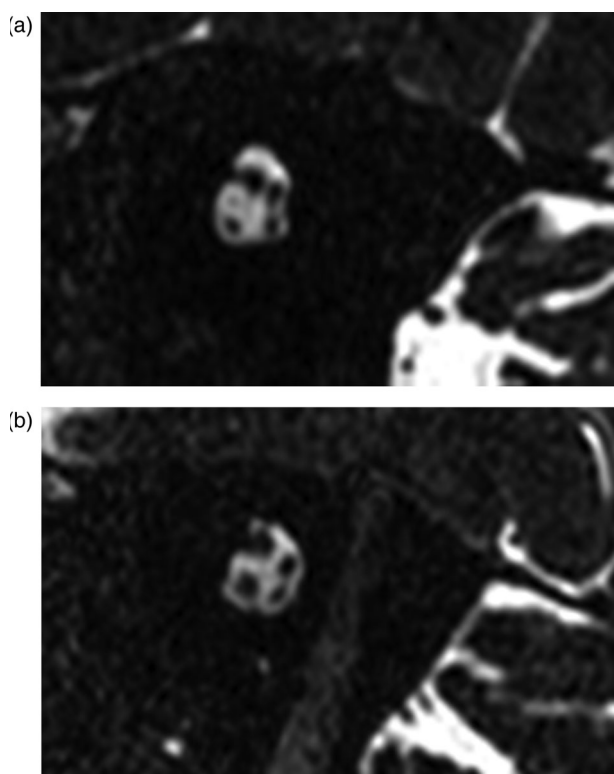


Figure 4 Direct parasagittal heavily T2-weighted images. (A) Right hypoplastic cochlear branch. (B) Left normal size cochlear branch.

to correlate with unfavorable outcome with postoperative ABI use.

For improving outcome and minimizing the possible nonauditory activity of the ABI, recently a new near field procedure of monitoring the electrical activity of the ABI on the ascending auditory system and on the surrounding cranial nerves has been introduced. This procedure, contrary to what is done with eABR which is a far field procedure allows a direct near field evaluation of the electrical compound action potentials directly generated contemporarily in the brainstem and on the nearby cranial nerves.

Close follow up

In order to monitor the speech and language development and the device function and to detect device migration as early as possible, a close follow up program is necessary by the implant center. The center is responsible for making proper arrangements in collaboration with local therapists of the child.

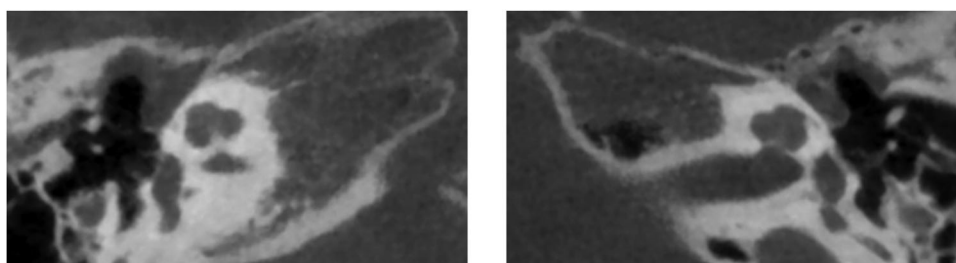
The follow up visits should be done under a standardized protocol at least every 3 months over the first 2 years, thereafter every 6 months and annually.

The standard protocol must include reports about child's behavior and reactions to auditory stimulation, use of the ABI, comments of the therapists, behavioral audiometry, check of device function, measurement of electrode impedances and NRT recording. In addition, if necessary recording of e-ABR and e-CR, CBCT imaging in cases of suspected device migration.

Performance should be monitored using a standardized age related set of tests (EEG subtest of the CRP). Results shall be reported in a standardized way using the CRP scale. They should be compared to normal listening and CI peers.

Conclusions

ABI has been the mode of auditory stimulation in children with severe inner ear malformations without any neural connection between the inner ear and the brainstem. More children have been implanted with ABI worldwide in recent years. Long-term audiological results indicate that children benefit from ABI in terms of hearing and language development, but generally speaking, the results of ABI in children did not reach the level obtained by children using CIs. If the children underwent a CI trial in a situation such as cochlear nerve deficiency and it is observed that they make very limited benefit from CI, the interval between CI and ABI should be minimized, in order to maximize the benefit from ABI. Performing CPA surgery in a child involves certain serious risks but, in experienced centers the complications are very rare. It should also be remembered that revision



CBCT 125 μm

Figure 5 (A) N. VIIIth aplasia on MR: no connection between fundus and base of the cochlea. (B) Normal VIIIth nerve on MR: fundus reaches the base of the cochlea.

surgery may sometimes be difficult and every effort should be taken to avoid revision in this population.

Disclaimer statement

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Conflict of interest All of the above authors state that they have no conflict of interest. This is a consensus report on ABIs; therefore, there is no abstract and reference number is very limited.

Ethics approval None.

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It is important for us to briefly explain the writing process of this article. This report was based on the data provided by all the participating groups during the meeting. During the afternoon of the second day, we discussed all the points presented and to be included in the consensus statement. The purpose

of this consensus report is to collect the pooled data of the participating centers to direct and guide the further investigations regarding management of complex inner ear anomalies. Therefore, this manuscript was based on only presented, discussed, or unpublished data. For that reason, it is natural that it does not contain any references as it is not a literature review rather a pooled data of experts in the field. This fact was included in the introduction part of the manuscript. It is also important for the reader to keep in mind that although there is a large authorship, there may be those who perform ABI who have not been included in the consensus paper.

This article includes the contribution of the whole participants and finalized with the consent of every single author. We would like to keep the manuscript in the present form and our main goal was the quickly publish and distribute this information, so that this unique group of patients can benefit from the opinion of the experts in this field. In our humble opinion, this will ultimately help healthcare providers to better manage these patients.