Current status of pediatric auditory brainstem implantation in inner ear malformations; Consensus statement of the third pediatric ABI meeting

Pediatric auditory brainstem implantation (ABI) is performed in severe inner ear malformations (IEM) and the indications are well defined [1]. Before 2000 it was not possible to habilitate hearing in these anomalies as the cochlear implantation was contraindicated.

The first ABI was performed in House Ear Institute (HEI) in Los Angeles, by the pioneering work of Drs. William House and William Hitselberger after removal of an acoustic neuroma in 1979. In 2000 Vittorio Colletti reported the first use of ABI in prelingually deafened children with severe IEMs. We believe that Vittorio Colletti opened a new pathway for the habilitation of children with severe IEMs.

Third Pediatric ABI Symposium was organized by Hacettepe Implant Team between 3-5 September 2020. 39 centers from 19 countries participated to this meeting. Because of COVID-19 pandemic the meeting was organized as an online zoom meeting. There were 12 sections in the meeting. This paper contains different aspects of pediatric ABI discussed in the meeting, including the latest outcome results of pediatric ABI reported by the participating centers.

1-Classification and Imaging Findings in Inner Ear Malformations

IEM's represent approximately 20% of congenital sensorineural hearing loss and contain a variety of anomalies with different surgical options. There are eight groups of anomalies according to Sennaroglu classification of IEM [2]. Some categories (enlarged vestibular aqueduct, incomplete partition (IP) type II and IP-III) always have a well-developed cochlear nerve and are not an indication for auditory brainstem implantation (ABI). Complete labyrinthine aplasia, rudimentary otocyst, cochlear aplasia, cochlear aperture aplasia and cochlear nerve aplasia are definite indications for ABI. Controversy still persists in cochlear hypoplasia, IP-I and common cavity with hypoplastic cochlear/cochleovestibular nerve. In this latter group, audiological and radiological findings are used to make the decision between cochlear implantation (CI) and ABI.

Histopathological findings reveal the difficulties in demonstrating the hypoplastic cochlear nerve. According to histopathological evaluation of temporal bone specimens with IEMs, a hypoplastic cochlear nerve may be present but if it is located adjacent to the wall of the internal auditory canal it may be difficult to demonstrate on magnetic resonance imaging (MRI) because of absence of fluid signal around it. MRI may be unable to demonstrate the hypoplastic cochlear nerve (CN) in a narrow internal auditory canal (IAC) because of the same reason. In addition, CN may be too thin to demonstrate. As a result, CN is reported as absent. In some specimens, CN may be absent in spite of a normal cochlea, cochlear aperture and internal auditory canal (IAC). Histopathological evaluation of cochlear specimens also demonstrated asymmetrical development between two sides; in clinical situation, it is very important to determine the more developed side for CI and less developed side for ABI. This is done by considering audiological as well as radiological findings.

Examination of the CN is extremely important because the choice between a CI and an ABI in the habilitation of a patient with IEMs depends mainly on the presence (and size) of a neural structure other than the facial nerve which is directly connecting with a normal or dysplastic cochlea. MR is the evaluation tool 'par excellence' and in difficult cases direct parasagittal Turbo Spin Echo T2 images or advanced Gradient Echo T2 images which can be reformatted in the parasagittal plane without quality loss should be used (e.g. balanced-Fast Field Echo XD images 0.3 to 0.5 mm^3). The advantage of the Gradient Echo T2 technique is that the nerves are grey and bone is black which may help to differentiate the presence of hypoplastic CN from bone in a narrow IAC. The diameter of the cochleovestibular nerve is larger than the facial nerve in the CPA. In the IAC the cochlear nerve is expected to be equal and larger than the facial nerve. If the CN is smaller than the facial nerve, CN hypoplasia/aplasia is suspected.

Warning signs of an absent nerve on cone beam computerized tomography (CBCT) or CT are an abnormal labyrinth, a narrow IAC and a narrow (< 1.7 mm) or absent cochlear nerve canal or cochlear aperture.

Hacettepe University reported the imaging findings in patients with positive audiological response who do not have a separate discernible CN visible on imaging. In an attempt to investigate if there are any imaging features that might predict the side with a positive auditory response in patients with bilateral aplastic CN, imaging studies of 15 patients with bilateral CN absence and unilateral auditory response were assessed. There was no significant correlation with the diameter of the vestibulocochlear nerve (VCN) compared to the facial nerve to the side with audiological response. In patients with IEMs except isolated cochlear nerve canal (CNC) stenosis, audiological response was always on the side with relatively more differentiated inner ear structures. In 6/7 patients with isolated CNC stenosis, audiological response was on the side with relatively wider internal auditory canal.

The Bruges St Jan – Antwerp St Augustinus radiology departments in Belgium reported that imaging of the brainstem and auditory pathways reveals important information in patients who are candidates for CI and ABI. Multi-echo sequences (e.g. m-FFE, MERGE, MEDIC) allow visualization of the cochlear nuclei and these and other sequences also allow visualization of the anatomy and pathology of other parts of the central auditory system in CI and ABI candidates. fMRI, PET-CT (due to its important radiation avoided or not used in young children) and Functional Near-Infrared Spectroscopy (fNIRS) can provide the following additional important functional information in difficult cases:

a) distinction between cochlear nerve aplasia or hypoplasia

b) decision on which side the CI or ABI should be installed

c) follow-up of cortical activity after CI or ABI placement (research setting – using fNIRS).

fMRI remains the golden standard functional technique although several technical challenges make the technique itself challenging in very young children, which is not the case in fNIRS but the latter technique is however suffering from very low spatial resolution.

2-Unusual Indications

In this section unusual indications for ABI are discussed.

Hacettepe University reported a case of bilateral common cavity who experienced facial nerve stimulation after CI surgery on the right side. This developed after surgery and decreased the benefit from CI considerably. It still persisted after repositioning of the electrode with a revision CI surgery. As there was a hypoplastic cochleovestibular nerve on the contralateral side, an ABI surgery was performed to the left side. CI-ABI use resulted in improvement in thresholds with the devices and auditory-language skills. The improvements were observed in MAIS, closed-set sentence recognition in auditory-verbal condition as well as CAP, SIR and aided thresholds. As a conclusion, severe facial nerve stimulation limits mapping and may prevent benefit from CI and contralateral ABI may be a solution in these complex situations if there is an ABI indication.

Results of CI in two cases of cochlear aplasia are reported by Hacettepe University. Normally cochlear aplasia is an indication for ABI. However, both of these cases, demonstrated listening behavior on one side and liked to wear hearing aids on that side. It was thought that cochlear nerve fibers were present within the vestibular nerve. Therefore, CI surgery was performed on the side with behavioral response and thresholds were obtained around 45 dB. To provide better auditory perception and improve language skills, these children were implanted with an ABI on the contralateral side. In general, only CI is not sufficient for language development alone in these cases, necessitating a contralateral ABI.

Burlo Garofolo Pediatric Institute, Ljubljana University, and Hacettepe University presented a case of unilateral deafness whose hearing was completely normal on the right side. Left side had an IP-I malformation with stapes footplate fistula which caused recurrent meningitis and further deafness on the contralateral ear. Although the fistula was repaired on the left side, meningitis resulted in severe sensorineural hearing loss on the right side. The child did not make benefit from CI possibly because of a post-meningitic cochlear nerve damage. As a result, ABI surgery was performed on the right side for partial recovery of previous hearing capacities. It is concluded that it is very important to diagnose and treat a stapes footplate fistula in a single sided deafness which may cause meningitis leading to an ABI procedure in the only hearing ear of a patient.

Universities of Groningen and Leiden presented the surgical difficulties of CHARGE syndrome for CI. These difficulties consist of sclerotic mastoid, abnormal venous structures, aberrance of facial nerve and round window stenosis; in around 40% of cases a trapped cochlea is present, making ABI the only viable solution. However, developmental, anatomical and other more life-threatening medical issues (feeding disorders, need for cardiac surgery) influence timing and outcome of CI as well as ABI care. Tailor-made individual evaluation and decision making is advised, considering that general outcomes are less favorable than in the general pediatric population. Age of implantation should be kept in mind, with timely transition to ABI when CI does not yield favorable results.

3-Audiological Findings

In this section audiological test methods and findings in case of hypoplastic/aplastic cochlear nerve were discussed.

Sheba Medical Center (SMC) presented the pivotal role of objective measures in the diagnosis of hearing loss and auditory function in young CI/ABI candidates with CN deficiency. Notable challenges in these young children include: obtaining unaided and aided behavioral audiograms with ear-specific data and appropriate masking in single sided deafness/Asymmetric HL cases; determining frequency-specific thresholds in the presence of an auditory neuropathy spectrum disorder profile; and difficulties in behavioral testing due to high prevalence of additional comorbidities (e.g. speech and language impairment). Accumulating clinical evidence from SMC shows that recording cortical auditory evoked potentials to speech tokens is feasible and provides valuable information regarding access to sound, functional integrity of the central auditory system, and cortical responsiveness to speech stimuli while using a hearing aid and/or a CI.

Hacettepe University reported the importance of behavioral testing to determine the best treatment modality between CI and ABI in cases where the presence of hypoplastic CN is doubtful. It necessitates two experienced pediatric audiologists to condition and observe the behavioral response of the child at the same time. Each ear is tested separately with insert ear phones. Behavioral evaluation is very critical to see ear specific response to sound and together with imaging findings appropriate intervention is decided.

Massachusetts Eye and Ear infirmary reported that behavioral audiometry allows for evaluation of frequency and ear specific detection levels that may not be otherwise obtainable due to the limitations of electrophysiologic testing such as stimulus intensity limits and unusual response morphologies. Additionally, the ability to complete operant conditioning tasks provides insight into the child's cognitive development and ensures that reliable responses can be obtained for ABI programing."

The use of intracochlear test electrode (ITE) to decide between CI and ABI was also reported by Hacettepe University. The electrode was developed by Med El company and has 3 intracochlear contacts. If wave V was obtained on eABR, a CI was implanted. This was the situation in two cases of normal

anatomy and IP-II. If the wave V was absent, operation continued as ABI as in two cases of absent CN. There were two cases that had clear responses on preoperative behavioral testing and hypoplastic CN was shown MRI. Even though we did not get a response with ITE, CI was applied. Intraoperatively no response was elicited even with actual CI electrode. Therefore, it is important to know that false negative result is possible; if the test is negative it does not completely rule out the presence of a hypoplastic CN. This is most probably due to severe anatomical abnormalities of the cochlea.

Hacettepe University reported the outcome of CI in 137 children with inner ear malformations (IEMs). The enlarged vestibular aqueduct group scored better on auditory perception tests than children with IP-II, possibly because of the modiolar defects in the latter group. Scores of children with IP-III were slightly lower but still acceptable with CI. As the need for high-level audiological outcomes increases, it is revealed that children with more complex IEMs, especially IP-I, cochlear hypoplasia and common cavity have challenges in identifying and comprehensive abilities. The most important factor for this result appears to be the size of the CN. According to this study some cases with IP-I, cochlear hypoplasia and common cavity become candidates for an ABI.

Manchester University reported the outcomes of CI in children with CND. Eleven of 25 children achieved limited audition (predominantly CAP 3 – 5) and speech intelligibility (SIR 2 – 3) and used sign as their main mode of communication. The 12-month CAP score predicted long term outcome. The CAP score did not improve beyond 24 months in most children. The size of the vestibulocochlear nerve in the cerebellopontine angle on MRI and preoperative transtympanic round window EABR were both significantly associated with the 12-month CAP score. They also reported late recovery of natural hearing in 3 children with CN deficiency in whom previous behavioral and electrophysiological investigations had demonstrated no audition. Late recovery of limited audition was observed at 5 to 6 years of age. They postulated that ABI may allow stimulation and development of central auditory pathways for later hearing aid or CI use. They suggested long term follow up to identify late improvements with hearing aids or CIs.

Importance of multidisciplinary team approach in the management of pediatric ABI is highlighted by University of Southern California. Children using ABIs benefit from a coordinated approach to intervention given the existence of intersecting challenges between programming and habilitation. Important considerations during patient candidacy and patient management post implant include individual and environmental factors. Recent experience confirms the importance of collaboration, communication, and consistency among caregivers. To gain optimal auditory benefit from an ABI and establish communicative competence, interdisciplinary collaboration is essential.

4-Decision Making Between CI and ABI

In this section clinics presented case reports on how to choose between CI and ABI in their practice. In general decision is not made solely according to imaging findings; but considering the audiological as well as radiological results. Centers reported cases where MRI failed to demonstrate a CN but there was a response during audiological evaluation particularly with behavioral testing. It is very clear that it is necessary to have better imaging modalities to demonstrate CN for decision making between CI and ABI. In situations where CN is reported as absent, if there is an audiological response during behavioral testing, CI is performed on the side with audiological response.

This requires appropriate counselling of the family. The reason is that the team is performing a CI in spite of a report indicating CN aplasia. Parents should also be informed that unilateral implantation may not provide sufficient hearing and language outcome in this situation. Children with hypoplastic CN or

cochleovestibular nerve are true indications for bilateral implantation. Therefore, proper counselling of the family is necessary and their expectations should be appropriate.

All centers preferred to perform a CI if there is hint of a CN and there is an implantable hypoplastic cochlea. The same is true for common cavity if there is a well-developed cochleovestibular nerve between the cavity and the brainstem. The reason is that it is possible to obtain some results (sometimes surprising good results) with a CI in this situation. The progress of the child is then followed closely, and CI or ABI is performed on the contralateral side. It is very important to perform initial CI surgery as early as possible; may be when the child is around 9-10 months old, to avoid the delay of the ABI procedure on the contralateral side. This makes it possible to evaluate the progress between 1-1.5 years of age until a decision for contralateral side is made. If the decision for ABI is delayed, then the chances for a better outcome decreases.

It is important to perform the initial CI surgery to the audiologically better side. If the CI is done to the better side but the child does not show sufficient progress, ABI is planned to the contralateral worse side. It is not advisable to remove CI and place ABI ipsilaterally. CI on the better side and ABI on the worse side will provide best bilateral stimulation option for the child. If the opposite has been done and the team is faced with a patient whose worse ear had been implanted with CI initially (instead of the better audiological site), this is a challenge to the team. Best management is to replace the CI with an ABI and implant a CI on the contralateral better side. Therefore, initial management is of paramount importance.

One other option is to make bilateral CI surgery in the management of CN hypoplasia and monitor the progress. If there is insufficient progress, the side with worse progress is converted to an ABI. It is very important to perform the initial surgery when the baby is around 9-10 months old so that conversion to ABI will not be done late. However, some cases were presented showing that conversion from CI to ABI can be challenging in certain situations. If bilateral CI is performed around the age of 8-9 months, then CI can be replaced with ABI on the contralateral worse side around the age of 1.5-2 years which can provide better outcome.

In this particular situation there may be reimbursement issues which need to be solved before going forward. The team does not know the exact outcome of this procedure at the start; it involves a kind of trial with CI in the situation of a hypoplastic CN. The regulatory board should be ready to reimburse the third device in this situation.

Intraoperative trial with intracochlear test electrode developed by Med El company can be used in decision making between CI and ABI. When evaluating the results, the implant team must be careful about the false negative results; the test can be negative in the presence of a hypoplastic cochlear nerve because of the anatomical abnormalities in the cochlea. A decision of performing intracochlear EABR using a custom-made electrode (Med El CME for EABR) should be taken before the child reaches the age of 12 months. The formal CI approach in the anatomically favorite side should be taken. A desirable EABR waveform is a reassurance for the future rehabilitation, but regardless the presence or absence of a good EABR response, a CI should be performed in the same setting. By doing so, a reliable stimulation to the hypoplastic nerve would be expected. If a child still does not make audiological/speech and language developmental progress with the CI in situ, a decision of offering ABI to the contralateral side of the CI should be made between age of 18 to 24 months. Ideally, the ABI surgery should be performed before the child reaches the age of 2 years.

In cases of definite ABI indications (Complete labyrinthine aplasia, rudimentary otocyst, cochlear aplasia, cochlear aperture aplasia and internal auditory canal aplasia) there is no need to wait for the outcome of a CI and direct ABI can be performed. Every effort should be done to look for a possibility of a CI on the contralateral side. If there is a definite ABI indication on one side, and a possible ABI indication on the

contralateral side, CI and ABI should be performed. Although majority of the participating centers performed this as a staged procedure, one center performed this simultaneously. In the latter situation CI surgery started first and ABI is done after CI surgery is finished. This procedure allows stimulation of both hearing centers as early as possible.

If there are definite ABI indications on both sides there is no place for CI in this situation and the team can proceed directly with an ABI. The age of surgery depends upon the experience of the team and majority of the centers performs the procedure when the child is at 1-1.5 years old. Bilateral ABI surgery is done by a limited number of centers. In case of a device failure in a child who uses a unilateral ABI only, it may be a catastrophe if the implant cannot be replaced. Bilateral ABI is definitely done as a staged procedure to minimize the surgical risk. Benefit will be evaluated better when the number of bilateral ABI users is increased.

Children with hypoplastic cochlear/cochleovestibular nerve need bilateral implantation more than any other group. Every effort should be done to provide bilateral stimulation: bilateral CI, CI and ABI or bilateral ABI.

5-Surgical Technique

ABI surgery is an otoneurosurgical intervention that requires the collaboration of otologist, neurosurgeon, anesthetist and audiometry team. In addition, the surgical team must have experience in CI, ABI in NF2 and surgeries in pediatric patients in order to avoid complications.

Most centers use a retrosigmoid approach for the placement of ABI in pediatric patients. A template to plan the skin incision so as to not disturb future plastic surgical reconstruction of pinna is useful. If there is unusual surface anatomy image guidance may be helpful to identify the position of the junction of the lateral and sigmoid sinuses in order to plan the craniotomy.

King's College Hospital of London recommended to place the internal receiver in a shallow well created in the skull before opening the dura. The bed should be positioned immediately above the craniotomy to allow sufficient length of the electrode to adjust to skull growth. Fixing the implant body is very important to avoid movement and hence, migration in case of seroma or CSF collection. It is important to prevent blood and bone dust from entering the subdural space to prevent aseptic meningitis. The cisterna magna should be accessed to drain cerebrospinal fluid in order to decompress the cerebellum and without this maneuver it is difficult to safely access the cerebello-pontine angle. The surgeon should not continue the procedure if the cerebellum cannot not be decompressed. Hyperventilation and mannitol are other resources that can help decompress the cerebellum.

Exposure of VII, IX, X and XI. cranial nerves are important. A 'V' shape identifies the landmark for the flocculus, then choroid plexus leading to the foramen of Luschka. The cochlear nucleus is located on the floor of brainstem which is seen as a pale area with veins on it. The vein of the cerebellomedullary fissure could well guide ABI electrode array insertion intraoperatively. Minimal manipulation and accurate insertion may reduce injury of the cochlear nucleus and local fibrotic changes. Some centers suggest using the test electrode to see the response on electrophysiological testing and then use definitive electrode insertion. Watertight closure of dura is very important to avoid seroma collection and CSF leakage. The Cochlear Implant center of Prof Diamante stressed the importance of tight closure of the dura, (if there is a defect suturing the dura with interposing muscle) and suturing the planes appropriately to avoid a potential cerebrospinal fluid leak.

Replacement of bone was controversial. Some centers replace the bone removed at the beginning of the surgery. Other centers do not close the bony craniotomy defect. No centers reported any complication resulting from not replacing the bone. Therefore, both methods provide healing without complications. In

addition, sealing of the exposed mastoid cells during the approach is important to avoid CSF leak into mastoid which may result in rhinorrhea.

University of Sao Paulo preferred to use the retrolabyrinthine approach for ABI in children exclusively. According to their experience this approach allowed good access, small chance of morbidity, and faster surgery (important in children). In adults, preservation of the labyrinthine block resulted in labyrinthine function and hearing similar to retrosigmoid approach without cerebellum retraction. In their series of 32 patients they did not have any complication.

The size of the flocculus of the cerebellum is variable and impacts the ease of surgical access to the lateral recess in the retrolabyrinthine approach for ABI placement in pediatric non-tumoral ABI. A simplified classification has been reported by the Madras ENT Research Foundation based on the variability of the floccular size ranging from Grade 1 to Grade 4 (grade 1 being a very small flocculus and Grade 4 being a large flocculus needing more extensive dissection). In general, the size of the flocculus has an inverse relationship to the size of the choroid plexus. However, the flocculus size does not impact the outcome and is only indicative of the degree of surgical difficulty in accessing the lateral recess. However, larger flocculus size-particularly Grade 4, has a higher risk of post-operative vestibular disturbance.

Imaging may have a potential role in preoperative planning for access in ABI patients. In some patients, foramen of Luschka may be closed. According to preliminary assessment of 25 patients at the Hacettepe University, imaging can identify closure of foramen of Luschka. An asymmetrically enlarged cerebellar flocculus may be a clue for a closed foramen. In these cases, the surgeon needs to elevate a flap to provide access to the lateral recess. Preoperative imaging is also important to evaluate the retrosigmoid incision area regarding venous sinuses. They may cause profuse bleeding which makes it impossible to continue in a retrosigmoid fashion. In such cases presigmoid retrolabyrinthine approach should be selected. Also position of the anterior inferior cerebellar artery (AICA) can be assessed prior to surgery on MRI. In such cases where AICA crosses the foramen of Luschka the vessel should be carefully dissected away.

The University of Navarra reported that in more than 75% of the cases, the active electrodes corresponded to those located in the central-basal portion of the ABI array region. These were placed closer to the ventral part of cochlear nucleus complex. It is estimated that this was caused by the cochlear nucleus complex (CNC) convexity. This finding advises changing the implant design by giving it a greater curvature. This design change will achieve greater proximity of the ABI paddle to the dorsal part of the CNC. This will also aid analyzing the effects of stimulating the auditory pathway at the cortex, by generating stimuli at different points: the cochlea, cochlear nuclei, inferior colliculus and the cortex. The possibility of analyzing the cortical response provoked on various points could be very useful to understand the physiological role of the auditory pathway centers and improve the function of new ABI designs.

The Hannover Medical School stressed the importance of the fixation of the implant electrode which can be a special challenge due to the growing head of the child and also the long cable leading to the implant electrode. Different modes of fixation can be tried but migration is still possible. There is a need for proper fixation technique which allows uneventful growing as well as ease for possible revision surgery. They reported about several cases where movement of the implant postoperatively has to be assumed. Proper analysis by electrophysiology and imaging is mandatory in order to have a proper indication for a revision surgery. Revision surgery is a big challenge for this patient group in terms of hearing and life-threatening complications.

6-Complications

Possible complications in ABI surgery are discussed in detail. In contrast to NF2, ABI in children is just for hearing restoration which is not guaranteed, therefore, complications should be avoided from the start.

University of Marburg highlighted the preoperative considerations such as looking for additional malformations which may interfere with surgical procedure, and discussion on possible postoperative problems with anesthesiologist especially in syndromic patients. Children are less compliant than adults and this makes them more prone to CSF pressure increase. The implant has to be fixed because in case of CSF collection implant body may start floating. During the surgery proper monitoring of cranial nerves along with eABR and monitoring of end-tidal CO2, body temperature, fluid balance is important.

In terms of surgical act, craniotomy has to be at the correct location and large enough. The dura is advised to be opened one cm at the base and CSF is to be drained before complete opening. One of the important points is to prevent blood and bone dust from entering the cerebrospinal space to avoid aseptic meningitis.

Cochlear Implant Center Prof Diamante reported that sometimes cerebellar swelling can be seen at the beginning of the surgery. In order to overcome this problem, opening and draining the cerebellopontine cistern, hyperventilation and mannitol are resources that can help. However, one caution during surgery is to discontinue the procedure if the cerebellum cannot be depressed sufficiently. In order to prevent complications, drilling the bony bed before opening the dura is advisable in order to avoid aseptic meningitis. In addition, meticulous hemostasis before closure to avoid any postoperative bleeding is important.

A subtle dissection of arachnoid membrane and wide opening of lateral recess of the 4th ventricle is advised. Then the electrode is pushed gently into the recess and laid over the cochlear nucleus. Electrode should be handled carefully.

In order to define the best place for stimulation some centers advised to perform sufficient EABR measurements with the test electrode and check the presence of auditory responses showing that auditory pathway is intact. If a test electrode is used, it is then replaced with the stimulating electrode which should be placed exactly at the same location. It is fixed with fibrin glue, and surgicell to avoid misplacement. The lead should not be fixed to the skull base as it may lead to tension and migration out of the recess. Lastly, a final stimulation via the implant has to be done before closing the dura.

Another important procedure is to perform a meticulous suturing of the dura. Sealing of the exposed mastoid cells during the approach, suturing the dura with interposing muscle and suturing by planes will avoid a potential cerebrospinal fluid leak. Some centers preferred to replace the bone flap to prevent scaring, which is important for revisions. However, there were centers who did not implant the bone flap at the end of the procedure without experiencing any side effects.

After the surgery is finished, patients should be kept one night at the ICU with slow recovery and a CT scan is to be done at day one after the surgery. The parents and nurses should be instructed to maintain an elevated head and body for the next days, even at sleep in order to prevent CSF leak.

One other postoperative problem is electrode migration in ABI patients. Electrode migration especially in children was mentioned to be one of the reasons of non-stimulation. Hanover Medical School reported that in a group of 28 patients electrode migration was observed in 4 cases resulting in non-stimulation, despite positive intraoperative EABR. In those cases, a revision has to be done in order to replace the electrode. Proper EABR measurements after surgery are mentioned to be mandatory combined with neural response telemetry to verify a stable position of the electrode carrier. Otherwise radiographic analysis is necessary. It is advisable to take one CT immediately after surgery. In case of stimulation problem, the position of the electrode can be compared with the initial CT to diagnose migration. Sometimes minor migration can be handled by reprogramming the electrodes. A case of a major migration necessitates a revision procedure. In order to avoid migration, one has to consider the skull

growth and leave sufficient extra electrode. Fixation of the electrode in the lateral recess is also very important to avoid migration.

7-Revision Surgery

Revision surgery for ABI is always a difficult task, and indication should be discussed with all disciplines involved. Causes for revisions can roughly be classified into intradural and extradural problems. Among intradural problems requiring revision are hematoma, infection, CSF leak, dislocation, hydrocephalus, and infarction. Postoperative CSF leakage may cause extradural swelling in the surgical field. General surgical complications like hematoma and infection are relatively seldom. In the series of University of Marburg consisting of 42 children, there was only one child that needed shunting due to hydrocephalus. Implant specific complications occurred three times in the same series: one electrode dislocation and 2 implant failures, one technical and one after fall on the implant. All these three patients underwent revision surgeries, resulting in 7.14 % of the 42 patients. The patient with electrode dislocation did not show improvement after revision surgery, whereas the other two showed good results.

According to the experience of University of Marburg, seven different scenarios occurring during workup in the indication process of revision surgery were defined:

1- The first scenario is gross dislocation, revealing good eABR intraoperatively but no or worse eABR at first fitting in anesthesia. In the CT, clear shift of electrode such as upside-down flip can be observed.
2-The second scenario involves the same eABR findings with first scenario, but in the CT there is an unclear shift of electrode.
3-The third scenario involves good eABR intraoperatively and at first fitting but no progress of hearing achievements. Late dislocation of electrode and change of electrode position in CT is observed.
4-Scenario IV involves the same eABR findings with the previous scenario, but deterioration of good hearing achievements is observed.
5-In scenario V, a good eABR intraoperative and at first fitting in anesthesia is observed but there is no hearing sensation, also electrode shift is not clear in this scenario observed in a syndromic patient.
6-Scenario VI involves implant failure after fall observed in a non-syndromic patient. There is good progress of hearing or achieved hearing level.
7-The last scenario is implant failure after fall for syndromic patient, moderate to worse progress/achievement of hearing was observed.

It was mentioned that in decision making, the risk of revision and the possible benefit in syndromic and non-syndromic patients should be balanced. They suggested that, scenario V and VII should probably not be revised. The others are possible candidates for revision if the patient makes benefit from ABI in hearing and language development as long as no other contraindication exist. In general, the revision provides an improvement of the hearing levels compared to pre-revision status. Complete changes of the system and just repositioning of the electrode are also possible.

In addition to the scenarios mentioned above, Medizinische Hochschule Hanover suggested another scenario in which implant failure is independent of impact to the implant and confirmed by implant test procedures. In this case also, revision should be considered.

Uppsala University Hospital reported outcomes of a pediatric revision surgery. This was a pediatric patient who had a technical device failure and needed a re-implantation. During surgery there was very tight fibrotic attachments between the implant and the surrounding tissue which made it difficult to remove. Intraoperative electrical auditory brainstem measurements (eABRs) gave unclear responses after re-implantation. Four years after re-implantation the patient has got CAP 4, the same as with the first implant, and is a full-time user.

Hacettepe University presented the outcomes of revision surgery of five pediatric ABI users which corresponded to 4 % of the pediatric ABI users. Initial and revision surgery video of the same patient was shown and it was indicated that revision surgery is much more difficult where fibrosis alters the anatomy making identification and preservation of landmarks and neural structures more difficult. Statistical comparisons of MAIS, expressive and receptive language scores before device failure and after revision surgery showed improvement, whereas aided thresholds and pattern perception scores did not change. Continued training between malfunction diagnosis and revision surgery is important to obtain similar or improved performance.

8- Intraoperative Monitoring & Postoperative Programming

Intraoperative monitoring is an important part of the ABI surgery. We can estimate the correct location of the stimulating electrodes of the ABI with the electrically evoked auditory brainstem response (eABR) measurements. Recorded intraoperatively, the eABRs guide the placement of the ABI electrode, and recorded weeks later, pre tune up, they assist with the device fitting by helping choosing the electrodes that provide better responses. Cochlear Implant Center Professor Diamente reported that they performed the eABRs intraoperatively and postoperatively in the twelve pediatric patients implanted with an ABI in their centre. The eABRs elicited by different electrodes were evident in all patients, and correlated with the behavioral measurements.

According to the experience of New York University, the eABR at initial stimulation (IS) is more closely aligned than EABR in the operation room (OR) to useful electrodes in programming. Neither EABR (OR) or EABR (IS) measures were predictive of speech perception performance. This is possibly due to comorbid factors. The predictive value of the eABR for speech perception outcomes remains unclear and requires more long-term data.

Verona University reported that near-field electrical compound action potential recording during ABI surgery can significantly improve potentials threshold definition and the number of auditory and extra-auditory waves generated (replacing electromyography). It provides important information for best coupling and fitting improving the overall open-set speech perception.

Massachusetts Eye and Ear Infirmary reported the benefits of repeating the eABR after ABI surgery and prior to the activation. A post-operative eABR prior to pediatric activation has three main purposes: 1) to assess changes in the ABI electrode array position, 2) to verify which electrodes are most likely to give an auditory or non-auditory percept when stimulated, and 3) to establish a safe range of monopolar stimulation for the pulse trains and live speech used in processor programming on each electrode. Purpose 1 and 2 are beneficial because eABRs change from the intraoperative to postoperative evaluations, despite using the same bipolar stimulation pairs, in 50% of cases and thus the prediction of auditory vs. non-auditory percepts from individual electrodes also changes. Purpose 3 provides a safer initial activation by having identified which if any electrodes elicit serious non-auditory responses such as cardiac or respiratory irregularities before the awake activation of a child with little to no language.

Cochlear Implant Center Professor Diamente reported that the complexity of audiological issues with ABI implantation is greater than with CI. Side effects during activation are not uncommon in ABI. To establish auditory or non-auditory stimulation is difficult in children. Associated handicaps, increase fitting challenges to set T/C levels and to decide what is auditory vs. non-auditory. Stimulation levels to reach audibility are generally higher. Hearing skills gradually improve with progressive variations even after 5 years of ABI use. Programming parameters should be changed in accordance with assessment. Counseling, intensive intervention and follow-up to parents, therapists and school are necessary during all the process. Evaluation in the controls according to the evolution is necessary: Hearing thresholds in free

field, Ling Six Sound test, vowels, consonants, Early Speech Perception (ESP) Test in Low Verbal or Standard version, closed set and open set speech perception tests, and Meaningful Auditory Integration Scale (MAIS) or Infant Toddler MAIS (IT-MAIS).

São Paulo University reported that despite the majority of the reports in the literature, their top performers, with open set speech recognition in quiet are NF2 adult patients. All of their pediatric patients were referred to total communication programs with sign language support. The eABR sometimes is present intraoperatively with higher currents that cannot be achieved postoperatively when non-auditory effects are evoked with lower currents. All of their children could demonstrate their non–auditory sensations or they were observed with a cartoon chart. Pitch order might not be a concern in young children with no previous auditory experience, that plasticity may induce a new tonotopic cortical reorganization, but it showed an impact in the outcomes of post lingual deaf adults with NF2.

Top performer NF2 patients are programmed in bipolar (BP) mode, with lower pulse widths and hence lower charges than poor outcomers. Whenever possible (available in the software), they choose BP mode to manage non-auditory stimulation, focusing the electric field, and allowing to maximize the number of active channels with different combinations of electrode pairs. They concluded that implanting at younger age is crucial. This is not sufficient if it is not supported with a good rehabilitation program with the participation of the family. Top performers need less charge than the others, probably due to the proximity to the cochlear nucleus and better positioning. If fibrosis or new tissue formation separates the electrode from the CN, it does not seem to interfere or be ´tangible´ in the impedances measures.

Verona University and Med-El developed and verified a fitting method in children implanted with ABI that is based on eABR and subjective responses. Evaluated eABR morphology in congenitally deaf children implanted with ABI; presence of the wave P2 is necessary in eABR. Evaluation of non-auditory stimulation with eABR and subjectively confirmed with the minimum follow up of 6 months after the initial ABI fitting with minimum of 3 fitting sessions for each patient

Outcomes

- The eABR seems to be reliable tool to judge ABI electrode placement (reconfirmation of the original findings by [3]
- The eABR based fitting helps children with ABI to achieve faster auditory perception and development; this time increases with the smaller age at the implantation and other handicaps of a child, it is depended on the ability to condition the child, it may be up to 12 months of child's auditory developmental period)
- In the tested children implanted with ABI, eABR based fitting helped to predict any possible non-auditory stimulation.
- If eABR is performed during the initial stimulation, no need to fit the child in the intensive care unit (ICU).

9-Outcomes of Pediatric ABI

All participating centers agreed that pediatric patients benefited from their ABIs. In some cases, the device failure can be observed, and re-implantation is possible for ABI users.

Sienna University reported the outcomes of auditory perception performances of their 127 pediatric ABI patients. This included the patients in Verona University where Vittorio Colletti started the first pediatric ABI surgery. Only 26% of them underwent ABI surgery before three years of age. The mean age of pediatric ABI patients was 3.63±2.83 years old. 46.25% had additional disabilities, which decreased their auditory perception outcomes. They reported that there was a statistical difference in the median CAP scores between etiology groups (such as cochlear nerve deficiency, cochlear malformation, bilateral

cochlear ossification) at 2, 3, and 4 years post-ABI. There was a broad variety on CAP-II scores of their pediatric patients. The children who had auditory input prior to ABI had the best outcomes, i.e., cochlear ossification, cochlear fracture, NF2. After at least 5 years of follow-up, six of them reached item-9 on CAP II, five participants on 8, and eight participants had 5 on this rating. Children who used ABI before the age of 3 had better CAP-II scores than children who were older at implantation. Additionally, the presence of additional needs was highly related to time to achieve CAP 5 score, regardless of etiology groups.

The outcomes of four children with ABI and their improvements on auditory perception were presented by the Uppsala University. Two of the four children had genetic syndromes (Goldenhar and CHARGE), one had post-ossification meningitis, and one had bilateral cochlear aplasia. Two children had used their ABI full-time, one of them had used it part-time and one was a non-user. They did not observe severe side effects.

The Chinese University of Hong Kong contributed auditory perception assessment especially on tonal-languages. They emphasized that suprasegmental perception on the pitch was one of the essential milestones for the tonal languages in the training process. They presented 11 failed CI patients who had ABI at the age of 1 to 3-year-old. Two out of 11 children had a global delay. Cantonese has lexical tones which is the most difficult aspect for children with ABI. Some children are able to develop tone perception as well as children using CI. There is indeed tonotopic arrangement in the brainstem with the contribution to tone perception. In terms of pediatric ABI patients, children with an auditory nerve and a cochlear malformation performed better than subjects with a cochlear nerve deficiency / auditory neuropathy spectrum disorder. All active users showed a variable degree of benefits from ABI. Five out of eight children achieved open-set word recognition. 50% of the children achieved satisfactory tone perception and production.

There were 20 patients using ABI (2 patients had bilateral ABI) in the University of Navarra. The sound recognition improved in one of the bilateral ABI users after the second ABI. Auditory sensation and quality of life improved after ABI in these patients.

Hacettepe University team reported that they implanted 137 primary pediatric ABI patients while 21 used their device for more than 10 years. They reported that 13 patients had their ABI at the age of one, and 33 patients were implanted under the age of 2. Children without additional needs had better auditory perception and language development outcomes if they had ABI surgery under the age of 3. Children with additional needs had poorer outcomes than without additional needs. The categories of auditory performance scores varied between 2 and 8. The mean score was 4 which was “discriminates some speech sounds without lip-reading”. The speech intelligibility rating scores of pediatric ABI patients ranged from 1 to 5. Additionally, 16.46% of children were able to use the telephone. Family support and intensive multidisciplinary therapy practices are crucial in children with ABI.

The speech comprehension development of 22 ABI children was reported from Hannover Medical School. High-performing child reached CAP = 8 after 10 years of ABI use. Many children remained at CAP 4 level. The transition from CAP 4 (discrimination of speech sounds) to CAP 5 (understanding of common phrases) seems to be a difficult step. Migration was found to be an issue in ABI and was solved in one case by re-programming the map. Migration and device failure were the main reasons for poor performance (CAP = 0). Auditory performance should correlate with the age of implantation, number or percentage of activated electrodes, and applied stimulation charge, but this hypothesis could not be confirmed due to the limited data set.

The Los Angeles Pediatric ABI clinical trial was designed to establish safety and efficacy in 10 eligible children (2-5 years of age) who did not demonstrate benefit from cochlear implants in House Ear Clinic. Six children underwent ABI surgery, followed by behavioral mapping and communication assessments over the course of 3 to 5 years. Speech perception results suggest that ABI progress is slower than that of children with cochlear implants based on clinical observation and published reports. The six children in this study have not progressed beyond closed-set speech perception tasks at 3-5 years post-op, except for one child who has progressed to open set. Children continue to show slow but steady progress. For all of the children, visual input remains essential for communication.

All but one of the 17 children who received an ABI at their institution under the guidelines of an FDA approved protocol obtained some degree of auditory benefit with the majority limited to sound/speech awareness in the New York University Grossman School of Medicine. The benefit is not equal to that obtained with a cochlear implant, nor do any of the children use oral language as a primary mode of communication with 12 out of 17 children using either American sign language or total communication. Some of the challenges to achieving improved performance with an ABI include 1) the current processing strategy which is designed to stimulate the eighth nerve, not the cochlear nucleus, 2) lack of programming guidelines and 3) school placement and therapeutic interventions which do not support oral/aural language development.

London Pediatric ABI service cohort's time post implantation is low; therefore, patients have not had time to reach their full potential/benefit. Parents need to support to stay motivated to provide a sound and spoken language rich environment alongside sign when it takes such a long time for any ABI benefit to be seen in their children. Most children have added developmental complexity, which can negatively impact their outcomes. In the St Thomas' Hearing Implant Center, most of children (8/9) like their ABI and want to wear it consistently, most (8/9) have improved in their speech intelligibility and also most (8/9) have improved in their awareness and/or understanding of sound. Most children (8/9) use sign as a predominate mode of communication, whilst 1/9 uses spoken English as their predominant mode of communication.

Experience from University Hospital Antwerp concerning bilateral ABI in children demonstrated a trend similar to the era when bilateral cochlear implantation was the understudy. With two children using bilateral ABI, the sample size is limited, and the children have had their second ABI with different intervals. Their auditory performance is characterized with the CAP score as 4 and 5, but the additive value could not be substantiated with this scoring system. This anecdotal data on better speech sound evaluation with the second ABI switched on with the so-called A§E phoneme discrimination test® test. Just the same, the fact is that neither of these children witnessed any adverse event when both ABI devices were switched on, and there is no evidence that bilateral ABI in one patient is not tolerated. Manchester University NHS Foundation Trust had carried out ABI placement in 12 patients and looked after 8 others from different centers. Of 20 ABI users, two of them had bilateral ABI. The mean pure tone average in the users is 35dBHL. CAP scores for 12 ABI patients ranged from 0 to 6 and the mean score of CAP was 4.16. Their SIR scores ranged from 1 to 4, and the mean SIR scores were 2. The CAP/SIR correlation coefficient value was 0.71. Eight of nine of those with CAP 4 or better had at least 50% of electrodes active. Age at implantation was not correlated with CAP scores. There were great individual variations in all outcomes in pediatric ABI users. The auditory development of children with ABI is prolonged for a long time.

10-Outcomes of CI & ABI patients

The NYU Grossman School of Medicine reported five patients with bimodal stimulation. They were not benefiting from their CIs and underwent ABI in the contralateral ear. Their age at ABI was between 2 and

6-year-old. CAP scores of those children ranged from 1 to 5. Two of them used total communication and the rest of the children preferred ASL. Following ABI, each subject developed further auditory and language skills in varying degrees. Those with more consistent bimodal use reported greater subjective benefit. Particular educational settings and primary modes of communication may contribute to using the device and performance.

The University of Navarra shared their experience in 4 out of 15 pediatric ABI users who had bimodal stimulation. One patient had bilateral ABI. Three of four bimodal users had additional needs, i.e., cognitive delay, CHARGE syndrome, and ADHD. Three children scored 1 in CAP, one child achieved 3, and one child had 5. Only one child used the spoken language, the rest of them preferred the sign language.

Cochlear Implants Center "Prof. Diamante" reported two children with bimodal stimulation. The first child achieved closed-set word identification and the second child reached open-set word recognition 45% disyllabic words in bimodal stimulation. Children use the devices in a permanent way and their speech performance with CI improved after ABI. Parents reported good compliance to both CI and ABI usage by their children who demonstrated better awareness, detection, and localization of environmental sounds. Intensive auditory training and permanent counseling with CI, ABI, CI+ABI with constant monitoring are necessary.

In Hacettepe University, 34 children with ABI were in bimodal stimulation group, two child had bilateral ABI, and the outcome of 23 children presented in the consensus meeting. 74% of all children reached above 20 in the MAIS scores. According to speech perception, 70% had more than half of the word discrimination score, and 48% of them had better than 50 out of 100 in the word identification. Nearly half of them (43.5%) achieved more than half of the total scores in open-set sentence recognition. While 70 percent preferred auditory-verbal communication and rest used total communication.

To sum up the presentations, we could imply that speech perception scores improved with bimodal stimulation. In a subset of these patients, continued improvement in CI performance over time was observed, even if no benefit was evident before the decision for ABI. This could suggest that ABI and CI have a synergistic effect, or it could simply be the adaptive ability of the developing brain to utilize the signals coming from these devices. There is preliminary evidence to support choosing the ear contralateral to the CI for an ABI in a pediatric patient with bilateral cochlear nerve deficiency. Most of the bimodal CI and ABI users often used their devices regularly. The auditory perception and language outcomes are heterogeneous; the associated handicaps have repercussions on results. The first communication mode could be sign language followed by and oral communication with lip reading.

Hacettepe University presented the result of simultaneous CI and contralateral ABI procedure in five pediatric cases. This procedure was performed in patients with a definite ABI indication on one side. It was reported that users could easily adapt to bimodal use in a relatively short time period when compared to sequential bimodal implanted group. Intensive auditory training and permanent counseling with CI, ABI, and bimodal stimulation with constant monitoring are necessary.

11-Rehabilitation

Cochlear Implant Center "Professor Diamante" reported that they taught children with ABI to listen and communicate orally through the development of detection, discrimination, identification, recognition and comprehension skills. They believed that this work is possible due to the combination of other areas that cannot be neglected, such as the constant improvement of oral language and cognitive levels in all their aspects. The language used at the beginning of their sessions was simple and composed of short phrases.

As for the development of auditory skills, they worked with Ling's detection and identification test, including whispering, distance and background noise. They had reached excellent outcomes following a distance of two meters. Identifying closed lists is part of their strategies, and they started rehabilitation stages with lists of four stimuli and they had come to work with lists of more than thirty stimuli with children who have ABI belonging to category five according to Moog and Geers.

Leiden University Medical Center reported on the importance of the additional needs. While comparing the children with CIs and ABIs, the influence of additional handicaps should be considered. On the basis of follow-up of 1-7 years (median almost 3 years) of a series of 10 children implanted in Leiden it is concluded that on average, the language skills with ABI matched those of children with CIs with additional disabilities. Six out of seven children with ABI could ultimately respond to speech or identify environmental sounds within one-year of ABI rehabilitation. Two children with ABI could even use spoken language. Language skills developed at a slower rate than in children with CIs, especially for speech production, but could reach the same competence level when additional disabilities were absent. Children with ABI need longer and more intense rehabilitation and support, with focus on auditory input, although sign language will continue to play an important role in communication for many children. It is very important to manage expectations with ABI, especially in the presence of additional handicaps.

Sheba Medical Center shared their experience with a post-meningitic child, with ossified cochleae, who had only partially inserted cochlear implants bilaterally through drill-out procedures. When extra-cochlear stimulation evolved on one side some years after surgery, and speech perception deteriorated, their dilemma was to choose between attempt at re-insertion of a cochlear implant or ABI. After consultations it is decided to perform an ABI on the right side. After ABI, initially the patient had thresholds around 45 dB with her ABI. But the performance showed variation and some deterioration. In addition, there were difficulties with programming of the ABI. They reported that they now face a 3rd dilemma, as her other ear is deteriorating as well.

According to the experience of Hacettepe University, in the cohort of pediatric ABI patients, %35 of children had additional needs. Most common difficulties were ADHD, eye-sight problems, syndromes (such as CHARGE), developmental disabilities, and neurodevelopmental disorders (such as autism). Out of 81 unilateral pediatric ABI users, 26.25% of them had additional needs. The auditory perception and language development outcomes of children with additional needs were lower than the children with only hearing loss in the ABI group. Especially, children with cognitive delay, neurodevelopmental delay, and global developmental delay showed limited development in CAP scores and language development scores. Their MAIS scores ranged between 10 and 35; closed-set speech perception scores varied 0 to 50 out of 100 points; and open-set sentence perception scores ranged 0 to 20 out of 100 points. In the rehabilitation, holistic approach is recommended to improve their quality of life. Not only auditory and language development areas, but all developmental areas should be assessed and supported in these children Multisensory educational materials should be used while teaching children with additional needs to enrich their auditory perception and language skills.

Twelve patients with cochlear nerve deficiency received an ABI in the NYU Grossman School of Medicine. Patients were evaluated with age-appropriate speech perception and production assessments, and health-related quality of life (HRQoL) surveys for parents of subjects and for subjects if age appropriate. 11 out of twelve received some auditory benefit from their ABI. Parental HRQoL ratings were positive for all domains with the exception of communication. Self-reported overall HRQoL metrics from two subjects were also positive. The presence of additional disabilities and health problems resulted in less positive HRQoL outcomes. Their results emphasize the need to assess outcomes in these patients beyond speech perception and communication.

Massachusetts Eye and Ear Infirmary (MEEI) presented a case review of their 10 pediatric ABI patients, which indicated that the level of sign language and cognitive skills influenced overall communication

outcomes. As a result, modifications were made to the MEEI pediatric candidacy process. Modifications included beginning the pediatric ABI candidacy evaluation process at a minimum of 18 months of age, development of age-appropriate nonverbal social-pragmatic skills and family involvement in and progression with sign language skills. An informational handout was also developed to support family education. The informational handout reinforced realistic expectations of limited auditory skills development with an ABI and indicated that sign language would be the most accessible language for communication and learning.

Cochlear Implant Center Prof Diamante shared the outcomes of 12 pediatric ABI patients. They reported that most aspects involved in children's quality of life –emotional and physical wellbeing, self-reliance, general functioning, social interactions, family and friends have shown a clear improvement after ABI implantation, showing that children can rely more on themselves, function with less assistance in their daily life, interact with others and manage better their emotions. ABI has proven positive results, which show that patients feel adapted, comfortable and obtain good profit of their implants, independently from audiological and speech perception outcomes.

12-Worldwide Demand for Pediatric ABI Centers

Parallel to the progress of implantation, almost all kinds of etiology are managed with different methods of implantation. Pediatric ABI is a difficult surgery which is increasingly being performed in different parts of the world in children with congenital bilateral profound SNHL who cannot receive a CI. These are children with complex IEMs without a cochlear nerve. As of now, the outcomes are still variable and are being collated from centers across the world. In general, outcome of pediatric ABI is not as high as CI in normal anatomy. But bimodal stimulation with CI and ABI provides much better stimulation and should be among the surgical options in centers dealing with implantation.

Paediatric ABI is a high cost, low volume complex healthcare intervention. The precise incidence of infants and children who might benefit for an ABI is unknown. However, it can be estimated around 1 in 100 children who would be considered for a CI. Therefore, we talk about a very important intervention which is not as frequent as CI. Both surgery, programming and rehabilitation are more challenging when compared to CI surgery.

The mission of the experienced centers must be to make this procedure available to different parts of the world. For the moment being only people with very good financial resources can afford to travel abroad and have the ABI surgery. Insurance usually may not reimburse the cost of the ABI implant and the procedure. If everything is successful it is commonly seen that family has difficulty to come back for programming. Therefore, not only the surgery, but audiology and rehabilitation need to be planned carefully.

There are, regions in the world where this intervention is not performed. One such area is Russia and post-soviet countries. Similarly, there is no center in the continent of Africa, performing this surgery.

Therefore, patients with complex IEMs, total ossification, bilateral IAC tumors have no chance to get hearing (re)habilitation or have to go abroad for expensive ABI surgery. Mapping and further sessions are also expensive and results of rehabilitation can be poor due to language barrier. Undervalued currencies and poor socio-economic factors make it unaffordable for people to travel abroad for expensive ABI surgery and further mapping and therapy sessions that may be necessary. The need for ABI Center and development of governmental program has become evident.

It is not possible to develop a pediatric ABI facility in every center that performs cochlear implantation. Centers with expertise in cochlear implantation and cerebellopontine angle surgery would be the logical

choice to perform this surgery. Also, habilitation in these children tends to be more challenging and protracted.

The minimal requirements for a startup ABI team are: experience with CI and skull base/neurotology surgery, team of CI audiologists and rehabilitation specialists, appropriate imaging capacity, assistance from an experienced ABI team (perhaps on-site assistance for first few ABI procedures and remote assistance for activation and programming), dedication and funding to continue the program, and assistance from a manufacturer. Ideally the center will serve that country and surrounding countries. It will be ideal if the neighboring countries also speak similar language.

Keeping in mind these considerations it would seem logical that there is a lot of benefit in confining the procedure to a few select centers in each region or country for now so that enough expertise and data can be collected, analyzed and conclusions drawn.

In UK at a national level this means that there should be strong governance around identification of centres that provide this service. In the UK, NHS England undertook a detailed appraisal process before reaching the decision to formally fund 2 such centres – London and Manchester. These centres serve a population of around 60 million. This can be extrapolated by other countries as an indicator of the number of centres needed.

Experienced centres should provide mentorship and support for new centres whether in the developed or developing countries. This must include training of local expertise not just in surgery and anaesthesia but assessment, case identification and habilitation. In the early stages of this process, the mentoring team should join the local team to maximise safety for the child and positive outcomes. The implant companies should provide appropriate intra-operative and habilitation clinical and scientist support.

It is also important to use ABI from both implant companies at the same time. Experience showed that, it can be very problematic for a country if one centre depends on only one company and that company experiences a problem in supplying the implants. Therefore, it is advisable to use implants from both companies so that if a company experiences a problem the intervention will not stop in that country, region or the continent.

According to the experience of the new center, patient load a new center can be developed for that region or continent over the years. This needs to be planned in a very responsible manner.

1. Sennaroglu, L., et al., *Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement.* Otol Neurotol, 2011. **32**(2): p. 187-91.
2. Sennaroglu, L. and M.D. Bajin, *Classification and Current Management of Inner Ear Malformations.* Balkan Med J, 2017. **34**(5): p. 397-411.
3. Waring, M.D., *Refractory properties of auditory brain-stem responses evoked by electrical stimulation of human cochlear nucleus: evidence of neural generators.* Electroencephalogr Clin Neurophysiol, 1998. **108**(4): p. 331-44.

Current status of pediatric auditory brainstem implantation in inner ear malformations; Consensus statement of the 3rd pediatric ABI meeting

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