Published in final edited form as:

Otol Neurotol. 2017 February ; 38(2): 212–220. doi:10.1097/MAO.0000000000001287.

Initial Results of a Safety and Feasibility Study of Auditory Brainstem Implantation in Congenitally Deaf Children

Eric P. Wilkinson, MD,

House Clinic/Huntington Medical Research Institutes

Laurie S. Eisenberg, PhD,

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Mark D. Krieger, MD,

Children's Hospital of Los Angeles

Marc S. Schwartz, MD,

House Clinic/Huntington Medical Research Institutes

Margaret Winter, MS,

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Jamie L. Glater, AuD,

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Amy S. Martinez, MA,

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Laurel M. Fisher, PhD, and

Address Correspondence to Eric P. Wilkinson, House Clinic/Huntington Medical Research Institutes; 2100 W. Third St. #111, Los Angeles, CA 90057; ewilkinson@houseclinic.com.

The Los Angeles Pediatric ABI Team

House Clinic/Huntington Medical Research Institutes

Eric P. Wilkinson

Marc S. Schwartz

Children's Hospital of Los Angeles

Mark D. Krieger

Tena L. Rosser

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Laurie S. Eisenberg

Laurel M. Fisher

Jamie L. Glater

Dianne H. Ganguly

Amy S. Martinez

Debra K. Schrader

Robert V. Shannon

Margaret Winter

Team Consultants

Danielle Dzubak

Carren J. Stika

Financial Disclosure: The team discloses research support (ABI devices) from Cochlear Americas for this research study.

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

Robert V. Shannon, PhD

Caruso Department of Otolaryngology-Head and Neck Surgery, Keck School of Medicine of the University of Southern California

the Los Angeles Pediatric ABI Team

Abstract

Objective—To determine the safety and feasibility of the auditory brainstem implant (ABI) in congenitally deaf children with cochlear aplasia and/or cochlear nerve deficiency.

Study Design—Phase I feasibility clinical trial of surgery in 10 children, ages 2 to 5 years, over a 3 year period.

Setting—Tertiary children's hospital and university-based pediatric speech/language/hearing center.

Intervention(s)—ABI implantation and post-surgical programming.

Main outcome measure(s)—The primary outcome measure is the number and type of adverse events during ABI surgery and post-surgical follow-up, including behavioral mapping of the device. The secondary outcome measure is access to and early integration of sound.

Results—To date, nine children are enrolled. Five children have successfully undergone ABI surgery and postoperative behavioral programming. Three children were screen failures, and one child is currently undergoing candidacy evaluation. Expected adverse events have been documented in three of the five children who received the ABI. One child experienced a cerebral spinal fluid (CSF) leak, which resolved with lumbar drainage. One child demonstrated vestibular side effects during device programming, which resolved by deactivating one electrode. One child experienced postoperative vomiting resulting in an abdominal radiograph. Four children have completed their 1-year followup and have speech detection thresholds of 30 to 35 dB HL. Scores on the IT-MAIS/MAIS range from 8 to 31 (out of a total of 40), and the children are demonstrating some ability to discriminate between closed-sets words that differ by number of syllables (pattern perception).

Conclusions—ABI surgery and device activation appears to be safe and feasible in this preliminary cohort.

Introduction

The auditory brainstem implant (ABI) is a surgically implanted prosthetic device that directly stimulates the cochlear nucleus complex in deaf patients who are unable to benefit from a cochlear implant (CI). The ABI was originally developed at the House Ear Institute by Drs. William House and William Hitselberger for patients with neurofibromatosis type 2 (NF2)(1,2). Patients with NF2 develop bilateral vestibular schwannomas, and surgical treatment compromises the cochleovestibular nerve, causing profound hearing loss. For these patients, cochlear implantation is not viable.

During ABI surgery in NF2, the device is introduced into the lateral recess of the fourth ventricle and placed over the area of the ventral and dorsal cochlear nuclei after tumor removal(3). The ABI is designed to stimulate auditory neural structures within the cochlear nucleus and convey sound to the auditory cortex. The Nucleus ABI received marketing approval by the U.S. Food and Drug Administration (FDA) in 2000 for individuals with NF2, 12 years of age and older.(2)

Indications for ABI have expanded for use in non-NF2 adults and children with cochlear nerve deficiency (hypoplasia or aplasia of the cochlear nerve) and cochlear aplasia (absence of the cochlea). Professor Vittorio Colletti in Italy began reporting on ABI implantation in adults without NF2 as well as children who were not candidates for cochlear implants in the early 2000s.(4,5) Although auditory outcomes vary widely, overall results of the non-NF2 ABI recipients have exceeded those reported for the NF2 ABI population. In some cases, open-set speech recognition has been reported.(6) One hypothesis for this encouraging finding suggests that concurrent tumor resection obscures important landmarks for electrode array placement, which is not the case for non-NF2 patients.(4) Conversely, many pediatric ABI candidates exhibit associated cognitive disabilities in addition to congenital deafness, which interfere with auditory skill development.(6)

Although use of the ABI in young children has increased globally, prior to 2012, no pediatric ABI surgeries had been performed in the United States for the non-NF2 indication. In December 2012, a multidisciplinary team from Los Angeles was cleared by the U.S. Food and Drug Administration (FDA) to conduct a Phase I feasibility clinical trial to obtain preliminary data on the safety of the ABI in young children between the ages of 2 and 5 years. In this report, we present 1-year follow-up data on the first four children to undergo an ABI in our trial.

Methods/Study Design

With oversight from the FDA under an Investigational Device Exemption (IDE) and funding from the National Institute on Deafness and Other Communication Disorders (NIDCD), a proposed Phase I feasibility clinical trial was reviewed by the Investigational Review Board of Children's Hospital Los Angeles (CHLA) and a Data Safety Monitoring Board (DSMB) composed of experts in related fields.(7)

Participants and Inclusion Criteria

The study is limited to surgical implantation of 10 children from age 2–5 with congenital bilateral deafness due to cochlear nerve hypoplasia or aplasia, cochlear aplasia, or cochlear ossification. A CI trial is mandatory prior to enrollment in the study unless the child's cochlear anatomy precludes CI insertion. Patients with radiographic evidence of cochlear aplasia or cochlear ossification may be enrolled in the trial without having CI experience. Full inclusion and exclusion criteria are listed in Table 1.(8,9)

Prior to official enrollment into the trial, IRB approved record review by the ABI team takes place, to exclude candidates with obvious eligibility issues. Records include CT and MRI imaging, as well as medical, audiological and educational reports. With approval by the

team, the family is invited to provide informed consent, and undergo the formal evaluation. Formal evaluation for candidacy after enrollment entails medical, radiological, audiological, speech and language, and psychological assessment, and screen failures may still occur during the formal evaluation process. The screening/baseline and postoperative measures are shown in Table 2.

To date, nine children have enrolled in the study, and their demographics are shown in Table 3. Their ages at time of enrollment ranged from 2 years, 3 months to 4 years, 11 months. Four of the children had received a CI (one bilaterally) prior to enrollment in the ABI trial. Five of the nine children met inclusion criteria and underwent ABI surgery with postoperative programming. The ninth child has been enrolled in the trial, but is still undergoing candidacy evaluation. Three children did not progress to surgery: one child was excluded for medical reasons, the family of a second child voluntarily withdrew from the study prior to surgery, and the third child was found to be progressing satisfactorily with the CI at the time of the evaluation.

ABI Surgery

The ABI approved by the FDA (Nucleus ABI24M ABI) was implanted in four of the five children (Cases #1, 2, 3, and 5). The fifth implanted child (Case #8) was implanted with the Cochlear Profile 541 ABI (Cochlear Americas, Denver, CO) after the FDA approved an IDE modification. It should be noted that at the time of this report, the Cochlear 541 ABI is not approved in the United States outside of IDE studies.

Surgical Protocol

Center for Disease Control (CDC) recommended vaccines Prevnar-13 and Pneumovax-23 are administered prior to surgery at the requisite intervals. Long-acting paralytic induction agents are avoided for monitoring purposes. Intravenous antibiotics are administered, typically vancomycin. Appropriate monitoring lines are placed, including blood pressure arterial line and electrocardiogram leads. Peripheral IV catheters and a urinary catheter are placed. The patient is intubated for the procedure. The patient is administered dexamethasone, mannitol, and furosemide for brain relaxation purposes. Lumbar drainage, while optional, is typically performed. The surgical team positions the patient either in the lateral or supine position depending on patient anatomy.

The head is secured in a Mayfield head holder or pin fixation. The abdomen is prepped for abdominal fat harvest, the postauricular region is shaved to expose a proper area of the scalp for retrosigmoid craniotomy, and craniotomy is performed.

If an indwelling CI is in place on the side of the ABI surgery, it is explanted. A seat and trough for the ABI device and tie-down suture holes are created. At this point, removal of spinal fluid for brain relaxation, may be performed. The ABI is inserted (Figure 1), and electrically evoked auditory brainstem responses (eABR) are obtained by the electrophysiologist to confirm proper position of the implant.(10–15) After the team decides that the implant is in the desired position and will not be moved any further, Teflon felt is placed to secure the device in position. Abdominal subcutaneous fat is harvested.

Closure of the craniotomy is performed in layers. Primary dural closure is performed including interposition of pieces of muscle to assist with a watertight closure, and collagen dural matrix and fibrin glue may also be used. Abdominal fat is used to bolster the closure, and the muscle and skin flaps are closed in layers. A firm gauze wrap is then applied for the first 48 hours. The patient is then extubated and taken to the intensive care unit. Lumbar drainage is maintained for 48 hours, then the drain is clamped and removed 24 hours later if there is no wound leakage. Perioperative antibiotic coverage is continued while the drain is in place. Following discharge, the skin sutures are removed after 10 days.

Sedated Device Activation

Four weeks after the surgical procedure, the patient is returned to the operating room where an IV infusion of propofol is begun for light sedation. The patient is kept under cardiopulmonary monitoring. The electrophysiologist places scalp electrodes to measure eABR and each electrode is tested to ensure no adverse cardiopulmonary, myogenic stimulation, or other non-auditory side effect (NASE) is observed during device stimulation. Initial programming of the electrodes is based on the eABR waveforms obtained at this sedated device activation.

Feasibility Clinical Trial Endpoints

The primary endpoint is a safety endpoint: The count of unexpected serious adverse events related to both the surgery and NASE during device stimulation. The distinction between adverse events related to the surgery and those related to device use is critical to understanding the risks of the ABI in young children. The risks of a retrosigmoid craniotomy in children are largely known and experienced surgical teams have procedures in place to mitigate these adverse events. NASE related to ABI stimulation are largely known from the adult NF2 ABI literature. The published accounts of pediatric non-NF2 NASE detail a smaller number of these types of adverse events. Any unexpected surgical or NASE would increase the risk profile of an ABI in young children.

Behavioral Activation/Programming and Follow-up Evaluation

The pediatric audiologists on the team measure detection levels for each viable electrode and program the ABI processor using standard techniques recommended for young children. Because of the potential NASEs, pediatric advanced life support (PALS) emergency equipment and personnel are on standby. Two additional secondary, preliminary efficacy, endpoints are considered: (1) sound detection on an audiogram at .25, .5, 1, 2, 3, 4 kHz by 12 months after surgery of no greater than 50 dB HL, and (2) a structured parent interview using the Meaningful Auditory Integration Scale (MAIS)(16) and its infant-toddler version (IT-MAIS).(17). Specific parent-reported behaviors which fulfill preliminary efficacy are that the child: 1) wears the device consistently, 2) demonstrates changes in vocal behavior, and 3) alerts to environmental sounds.

Results

Eighth nerve findings in the cerebellopontine angle and lateral recess reflected significant anatomical variability. These findings, as well as adverse events requiring intervention, are shown in Table 4. There were no unexpected device-related adverse events. There were no instances of meningitis, skin flap complications, device failure, seizure activity, cerebral hemorrhage, or mortality. A full table of all adverse events of all grades is included in an online supplement.

Four children have completed the 1-year evaluation and their audiological results are shown in Figure 2. The individual electrodes programmed are also shown, with some variability in the number, but particularly the placement on the electrode array among the four children.

Soundfield and speech detection thresholds indicate access to most of the sounds of speech with the ABI activated. Although soundfield thresholds hover around 40 to 45 dB HL (with exception of the high frequencies for Case #3), the speech detection thresholds (SDTs) range from 30 to 35 dB HL.

Total scores on the IT-MAIS/MAIS, as reported by the parents at the 1-year evaluation, range from 8/40 for Case #1 to 31/40 for Case #5. These scores indicate that variability is high and progress slow as the children begin developing rudimentary auditory skills. More specifically, these scores show that all four children are consistently using the device throughout the day, and Cases #1 and #2 demonstrate increased vocal behavior with the ABI activated. Only Case #5 is frequently alerting to environmental sounds. None of the children shows evidence of spontaneously differentiating between two talkers or between speech and nonspeech sounds.

The Low-Verbal Version of the Early Speech Perception (ESP) Test(18) has been attempted for each child. Although three of the four children show evidence of some pattern perception by their ability to differentiate between words with different numbers of syllables, only Case #5 passed Category 2 Pattern Perception.

Discussion

Though preliminary, our results show that with careful patient selection and proper attention to operative technique, ABI surgery and subsequent device programming in children can be performed safely. No patient experienced any long-term sequelae.

The patient who experienced the CSF leak was treated immediately with lumbar drainage, which resolved the leak with no aftereffects. CSF leak has been the most frequently reported significant complication in pediatric ABI surgery(8,19,20), occurring in up to 8.5% of cases. Nearly all centers in the world performing this procedure have experienced CSF leaks.(21) It should be noted that in the development and modification of our surgical protocol, we have added the option for lumbar subarachnoid drainage, and have begun to perform it routinely, similar to prophylactic drainage in acoustic neuroma surgery in adults. This decision was supported by our DSMB. Perioperative lumbar drainage has been shown to be useful in preventing CSF leak after retrosigmoid approach.(22) Laing and Moffat (23) showed that

intracranial CSF pressure rises immediately after posterior fossa surgery and normalizes after 48 hours, with impaired cerebrospinal fluid absorption theorized to be the cause. Elevation in CSF pressure can result in CSF egress around the tract of the ABI electrode cable into the subgaleal space, resulting in pseudomeningocele and CSF leak. Complications of pseudomeningocele include skin wound breakdown, meningitis, and need for further surgery. In our opinion, CSF leak must be vigilantly prevented and lumbar drainage should be strongly considered.

Despite use of pressure dressings, nearly 10% of pediatric ABI patients may have a CSF leak postoperatively. Pressure dressings have been shown to be ineffective in preventing hematoma formation, and some authors have advocated discontinuing their routine use as unnecessary and potentially detrimental. (24–26) One study showed that surgeons are unable to accurately assess pressure to prevent magnet migration with dressings during MRI (27). It is therefore difficult to support the routine use of tight pressure dressings over an implanted device in a child when this could result in a pressure ulcer and devastating wound complication. We have used dressings for 48 hours to provide initial counterpressure to prevent pseudomeningocele as the lumbar drain is utilized, but discontinue dressings after that point.

The experience of the team is paramount to the success of pediatric ABI implantation. A previous consensus statement on ABI surgery outlines the team personnel necessary to ensure 1) minimization of complications and 2) proper programming of the device and rehabilitation of the child with the ABI.(8) The surgical team should have experience in three areas: CI surgery, vestibular schwannoma surgery, and adult ABI surgery.(8) The variation in anatomy of the cerebellopontine angle, cranial nerves, and lateral recess in pediatric patients showing congenital inner ear malformations may be very unfamiliar to surgeons not frequently performing posterior fossa surgery. Despite our report only including a small number of cases, we have found significant variability in the anatomy. In two cases, a membrane over the lateral recess required sharp dissection to identify the cochlear nucleus complex. In all cases, knowledge of anatomic variability near the lateral recess of the fourth ventricle has been critical to the safe dissection and device placement of the ABI.

We chose the lower age limit of 2 years for this feasibility study. The age was chosen by a consensus of the surgeons and researchers(20) as an age when critical factors such as patient weight and blood volume, temporal bone anatomy, scalp thickness, speed of brainstem development(28), and intracranial anatomy(29) would be developed sufficiently to minimize risk both from the surgery (e.g., skin breakdown) and the device itself, (e.g., hard failure due to falls). Young patients with cochlear nerve deficiency frequently have some balance or coordination difficulty, which improves with physical therapy and age.

With the goal of minimizing risk to the patient and maximizing less invasive options, we advocate CI in children with normal cochleas with possible cochlear nerve deficiency, particularly when a hypoplastic nerve is visualized or there is a patent bony cochlear nerve canal (BCNC) of at least 1mm.(30) The risk profile for a CI does not include the same potential complications as a craniotomy for ABI. The decision to place a CI in a patient with

cochlear nerve deficiency is not simple, and should be individualized. Currently, in smaller series, MRI imaging and electrophysiology are not predictive of outcome with CI, with many reports of children performing sufficiently with CI in these cases to avoid ABI.(31,32)

There are data to support an ABI after CI failure in cochlear nerve deficiency. In a retrospective cohort study by Colletti, (33) a group of children were studied who initially underwent CI and failed to progress with auditory perceptual skills. Before CI, all of the children had severe to profound sensorineural hearing loss and a diagnosis of cochlear nerve deficiency. Explantation of the cochlear implant and simultaneous ABI on the same side was performed, and all patients demonstrated an absent cochlear nerve at surgery. Performance as measured by the Categories of Auditory Performance(34) (CAP) scale was significantly improved after ABI.

In our study, we did not perform round window evoked response ABR (eABR) preoperatively. Many of our patients were implanted at outside centers with CI before being enrolled in our study, which makes assessment with all available electrophysiological techniques difficult, including those performed under general anesthesia for preoperative assessment. Additionally, we elected not to remove the CI in cases of unilateral CI implantation, in favor of ABI implantation on the contralateral side. As such, we did not investigate the CI side further with round window eABR.

European data demonstrate that children with other disabilities, including severe syndromic associations and associated cognitive deficits, perform poorly.(8,33,35) Of course, benefits of ABI implantation must be weighed against the risk of surgery in this group of patients in whom craniotomy is performed solely for ABI placement. Based on potential complications, we do not feel that the risk/benefit analysis supports implantation of this group of children.

We consider the team approach essential for pediatric ABI implantation(8) due to complicating factors specific to young children, including the possibility of non-auditory sensations. Their limited ability provide feedback about electrical stimulation mandates assessment by experienced pediatric audiologists. Speech-language pathologists and educational specialists provide critical guidance to the families and care providers of children with ABI.

Many families, when confronting the diagnosis of cochlear nerve deficiency, and the possibility that CI may not provide the expected audiological benefits, seek out an ABI as an option that, in their perception, may provide similar outcomes as CI. There can be a misconception that ABI, as a newer technology, is an improved technology over CI. It is critical that the pediatric ABI team and referring caregivers educate and reinforce to the family that ABI outcomes are in most cases inferior to outcomes with CI. Development of language, specifically the use of sign language communication, is critical to programming and rehabilitation, and families should demonstrate adequate development of skills with sign language as part of the process toward ABI. Family involvement is critical; in the NF2 population of patients 12–18 years of age, family support was highly predictive of success with the device.(36) Early referral of patients to a pediatric ABI center should be made when a child is undergoing CI with abnormal anatomy. Counseling to parents and caregivers on

language, speech, educational, and developmental issues must occur early in the ABI process to prepare them for what is to come.

Our audiological outcomes are consistent with other studies regarding the rate of progress of children with ABI. Progress is slow and incremental. The number of active electrodes in our patients are very similar to other reported series in the literature.(21,37,38) All centers have experienced complications; in our center we had one CSF leak repaired with lumbar drainage, but have had no device failures and have had very limited non-auditory side effects (NASE), limited to vestibular effects seen in one patient requiring one electrode to be removed from the clinical map. In our center, even with lumbar drainage in place, patients can be managed on the hospital ward instead of the ICU. However, at our children's hospital, the floor census is frequently so high that patients eligible for transfer to the hospital floor will frequently be kept in the ICU on floor status due to bed availability. For this reason, we have categorized the number of "effective ICU days" due to this (see Table 3). In a safety study, we are not actively engaged in minimizing hospital stays, but rather optimizing a surgical and postoperative protocol to maximize patient safety.

It is anticipated that ABI outcomes will occur along the mid-to-lower percentiles of longitudinal series of CI patients.(39,40) For example, the 1-year data in the present study indicate lower scores on the IT-MAIS/MAIS and ESP than the 1-year post CI mean scores for 42 pairs (CI and normally hearing) of children assessed in the national Childhood Development after Cochlear Implantation (CDaCI) study.(41) Young(32) reported that in some cases of CI in cochlear nerve deficiency, development of adequate responses may take several years. ABI outcomes may in many cases follow a similar trend.

It is interesting that Case #5 is demonstrating the most rapid progress of the four children. This child did not receive the ABI until nearly 5 years of age and had not received a CI due to cochlear aplasia. Further, this child is in a school for deaf, communicating via American Sign Language (although receiving additional speech therapy). We can only speculate at this point about the reasons for these promising findings and wait to see if this trajectory is maintained over time.

The ABI in children involves placement of a device that will stimulate the central nervous system for the patient's lifetime. Histological sectioning of both sides of the brainstem from a post-lingually deafened adult patient who used an ABI daily for 15 years was reported in 2012. (42) This individual demonstrated very high levels of speech recognition performance up until the time of his death from complications of NF2. The report showed intact, comparable populations of auditory neurons on both the implanted and unimplanted sides of the brainstem. This finding suggests that long-term electrical stimulation of the cochlear nucleus is feasible and is unlikely to result in additional damage to neural tissue. This study has several limitations. The most significant limitation is the small number of patients making it difficult to generalize the results to larger patient populations. As a clinical trial focused on safety and feasibility, detailed surgical and audiological protocols are open to optimization during the trial to maximize patient safety, and individual center preferences and choices in the interest of patient safety may make comparisons with other centers

difficult. While efficacy of the device can be studied within the limited followup that we currently have, long-term followup is not available.

Conclusion

Preliminary results from this safety trial indicate that ABI surgery and device programming may be performed safely in young children when carried out by a competent multidisciplinary team. The team approach is essential for all aspects of decision-making throughout the trial. Careful patient selection is of the highest importance in evaluating the eventual long-term risk vs. benefit profile of this procedure. When determining candidacy for an ABI, medical and cognitive comorbidities should be minimized. Moreover, emphasis on visual communication by both parent and child ensures that language remains a priority. Prevention of complications by meticulous wound closure and perioperative care is critical to prevent the most frequent complication of CSF leak. The role of proper informed consent, reasonable family expectations and family involvement cannot be overstated.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

NIH Grant #U01DC013031 (NIDCD)

IRB CHLA #CCI-13-00094

[ClinicalTrials.gov](https://clinicaltrials.gov/ct2/show/study/NCT02102256) #NCT02102256

We would like to thank the families of the patients enrolled in our trial for their commitment and trust as we endeavor to establish the feasibility of this technology in the pediatric population.

References

1. Brackmann DE, Hitselberger WE, Nelson RA, et al. Auditory brainstem implant: I. Issues in surgical implantation. *Otolaryngol Head Neck Surg.* 1993; 108:624–33. [PubMed: 8515999]
2. Schwartz MS, Otto SR, Shannon RV, Hitselberger WE, Brackmann DE. Auditory brainstem implants. *Neurotherapeutics.* 2008; 5:128–36. [PubMed: 18164492]
3. Fayad JN, Otto SR, Brackmann DE. Auditory brainstem implants: surgical aspects. *Adv Otorhinolaryngol.* 2006; 64:144–53. [PubMed: 16891840]
4. Colletti V. Auditory outcomes in tumor vs. nontumor patients fitted with auditory brainstem implants. *Adv Otorhinolaryngol.* 2006; 64:167–85. [PubMed: 16891842]
5. Colletti V, Carner M, Miorelli V, Guida M, Colletti L, Fiorino F. Auditory brainstem implant (ABI): new frontiers in adults and children. *Otolaryngol Head Neck Surg.* 2005; 133:126–38. [PubMed: 16025066]
6. Colletti L, Shannon RV, Colletti V. The development of auditory perception in children after auditory brainstem implantation. *Audiol Neurotol.* 2014; 19:386–94. [PubMed: 25377987]
7. Fisher LM, Eisenberg LS, Krieger M, Wilkinson EP, Shannon RV, Los Angeles Pediatric ABIT. Regulatory and funding strategies to develop a safety study of an auditory brainstem implant in young children who are deaf. *Ther Innov Regul Sci.* 2015; 49:659–65. [PubMed: 26366332]
8. Sennaroglu L, Colletti V, Manrique M, et al. Auditory brainstem implantation in children and non-neurofibromatosis type 2 patients: a consensus statement. *Otol Neurotol.* 2011; 32:187–91. [PubMed: 21224730]

9. Buchman, CA., Teagle, HF. An early feasibility study of the safety and efficacy of the Nucleus 24 auditory brainstem implant in children with cochlear or cochlear nerve disorders not resulting from neurofibromatosis type II [ClinicalTrials.gov identifier: NCT01904448]. Available at: <https://clinicaltrials.gov/ct2/show/NCT01904448>. Accessed August 1, 2016
10. Nevison B. A guide to the positioning of brainstem implants using intraoperative electrical auditory brainstem responses. *Adv Otorhinolaryngol.* 2006; 64:154–66. [PubMed: 16891841]
11. O'Driscoll M, El-Deredy W, Atas A, Sennaroglu G, Sennaroglu L, Ramsden RT. Brain stem responses evoked by stimulation with an auditory brain stem implant in children with cochlear nerve aplasia or hypoplasia. *Ear Hear.* 2011; 32:300–12. [PubMed: 21150625]
12. O'Driscoll M, El-Deredy W, Ramsden RT. Brain stem responses evoked by stimulation of the mature cochlear nucleus with an auditory brain stem implant. *Ear Hear.* 2011; 32:286–99. [PubMed: 21157353]
13. Waring MD. Properties of auditory brainstem responses evoked by intra-operative electrical stimulation of the cochlear nucleus in human subjects. *Electroencephalogr Clin Neurophysiol.* 1996; 100:538–48. [PubMed: 8980418]
14. Waring MD. Intraoperative electrophysiologic monitoring to assist placement of auditory brain stem implant. *Ann Otol Rhinol Laryngol Suppl.* 1995; 166:33–6. [PubMed: 7668693]
15. Waring MD. Auditory brain-stem responses evoked by electrical stimulation of the cochlear nucleus in human subjects. *Electroencephalogr Clin Neurophysiol.* 1995; 96:338–47. [PubMed: 7635078]
16. Robbins AM, Renshaw JJ, Berry SW. Evaluating meaningful auditory integration in profoundly hearing-impaired children. *Am J Otol.* 1991; 12(Suppl):144–50.
17. Zimmerman-Phillips S, Robbins AM, Osberger MJ. Assessing cochlear implant benefit in very young children. *Ann Otol Rhinol Laryngol Suppl.* 2000; 185:42–3. [PubMed: 11140998]
18. Moog, JS., Geers, AE. Early Speech Perception Test for profoundly hearing-impaired Children. St. Louis, MO: Central Institute for the Deaf; 1990.
19. Noij KS, Kozin ED, Sethi R, et al. Systematic Review of Nontumor Pediatric Auditory Brainstem Implant Outcomes. *Otolaryngol Head Neck Surg.* 2015; 153:739–50. [PubMed: 26227469]
20. Colletti V, Shannon RV, Carner M, Veronese S, Colletti L. Complications in auditory brainstem implant surgery in adults and children. *Otol Neurotol.* 2010; 31:558–64. [PubMed: 20393378]
21. Couloigner V, Gratacap M, Ambert-Dahan E, et al. A report of three cases and review of auditory brainstem implants in children. *Neurochirurgie.* 2014; 60:17–26. [PubMed: 24656883]
22. Bien AG, Bowdino B, Moore G, Leibrock L. Utilization of preoperative cerebrospinal fluid drain in skull base surgery. *Skull Base.* 2007; 17:133–9. [PubMed: 17768443]
23. Laing RJ, Smielewski P, Czosnyka M, Quaranta N, Moffat DA. A study of perioperative lumbar cerebrospinal fluid pressure in patients undergoing acoustic neuroma surgery. *Skull Base Surg.* 2000; 10:179–85. [PubMed: 17171145]
24. Gurgel RK, Oghalai JS. Are mastoid pressure dressings necessary after otologic surgery to prevent postoperative hematomas? *Laryngoscope.* 2012; 122:485–6. [PubMed: 22354627]
25. Khan I, Mohamad S, Ansari S, Iyer A. Are head bandages really required after middle-ear surgery? A systematic review. *J Laryngol Otol.* 2015; 129:740–3. [PubMed: 26134601]
26. Luo SD, Su CY, Wu CM, Hwang CF. Mastoid pressure dressing for cochlear implantation—is it necessary? *Int J Pediatr Otorhinolaryngol.* 2009; 73:857–60. [PubMed: 19362744]
27. Cuda D, Murri A, Succo G. Focused tight dressing does not prevent cochlear implant magnet migration under 1.5 Tesla MRI. *Acta otorhinolaryngologica Italica: organo ufficiale della Societa italiana di otorinolaringologia e chirurgia cervico-facciale.* 2013; 33:133–6. [PubMed: 23853406]
28. Moore JK, Ponton CW, Eggermont JJ, Wu BJ, Huang JQ. Perinatal maturation of the auditory brain stem response: changes in path length and conduction velocity. *Ear Hear.* 1996; 17:411–8. [PubMed: 8909889]
29. Courchesne E, Chisum HJ, Townsend J, et al. Normal brain development and aging: quantitative analysis at in vivo MR imaging in healthy volunteers. *Radiology.* 2000; 216:672–82. [PubMed: 10966694]
30. Adunka OF, Jewells V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. *Otol Neurotol.* 2007; 28:597–604. [PubMed: 17667769]

31. Song MH, Kim SC, Kim J, Chang JW, Lee WS, Choi JY. The cochleovestibular nerve identified during auditory brainstem implantation in patients with narrow internal auditory canals: can preoperative evaluation predict cochleovestibular nerve deficiency? *Laryngoscope*. 2011; 121:1773–9. [PubMed: 21792968]
32. Young NM, Kim FM, Ryan ME, Tournis E, Yaras S. Pediatric cochlear implantation of children with eighth nerve deficiency. *Int J Pediatr Otorhinolaryngol*. 2012; 76:1442–8. [PubMed: 22921779]
33. Colletti L, Wilkinson EP, Colletti V. Auditory brainstem implantation after unsuccessful cochlear implantation of children with clinical diagnosis of cochlear nerve deficiency. *Ann Otol Rhinol Laryngol*. 2013; 122:605–12. [PubMed: 24294682]
34. Archbold S, Lutman ME, Marshall DH. Categories of Auditory Performance. *Ann Otol Rhinol Laryngol Suppl*. 1995; 166:312–4. [PubMed: 7668685]
35. Freeman SR, Stivaros SM, Ramsden RT, et al. The management of cochlear nerve deficiency. *Cochlear Implants Int*. 2013; 14(Suppl 4):S27–31.
36. Otto SR, Brackmann DE, Hitselberger W. Auditory brainstem implantation in 12- to 18-year-olds. *Arch Otolaryngol Head Neck Surg*. 2004; 130:656–9. [PubMed: 15148193]
37. Puram SV, Barber SR, Kozin ED, et al. Outcomes following Pediatric Auditory Brainstem Implant Surgery: Early Experiences in a North American Center. *Otolaryngol Head Neck Surg*. 2016; 155:133–8. [PubMed: 27095049]
38. Lundin K, Stillesjo F, Nyberg G, Rask-Andersen H. Experiences from Auditory Brainstem Implantation (ABIs) in four paediatric patients. *Cochlear Implants Int*. 2016; 17:109–15. [PubMed: 26841821]
39. Barnard JM, Fisher LM, Johnson KC, et al. A Prospective Longitudinal Study of U.S. Children Unable to Achieve Open-Set Speech Recognition 5 Years After Cochlear Implantation. *Otol Neurotol*. 2015; 36:985–92. [PubMed: 25700015]
40. Eisenberg LS, Johnson KC, Martinez AS, Visser-Dumont L, Ganguly DH, Still JF. Studies in pediatric hearing loss at the House Research Institute. *J Am Acad Audiol*. 2012; 23:412–21. [PubMed: 22668762]
41. Eisenberg LS, Johnson KC, Martinez AS, et al. Speech recognition at 1-year follow-up in the childhood development after cochlear implantation study: Methods and preliminary findings. *Audiol Neurotol*. 2006; 11:259–68. [PubMed: 16699259]
42. Otto SR, Moore J, Linthicum F, Hitselberger W, Brackmann D, Shannon RV. Histopathological analysis of a 15-year user of an auditory brainstem implant. *Laryngoscope*. 2012; 122:645–8. [PubMed: 22253008]

Figure 1. Surgical Field in a Right Ear. Panel A: Preparation of the ABI electrode paddle for insertion. The polyethylene wings are trimmed for proper sizing in the pediatric lateral recess. Panel B: The ABI paddle is inserted into the lateral recess. Note the absence of the eighth nerve, and presence of the lower cranial nerves. VII=Facial nerve; AI=Anterior Inferior Cerebellar Artery (AICA); LCN=Lower Cranial Nerves.

Figure 2.

Electrode maps and initial auditory perception results. Shaded electrodes are active while white electrodes are inactive.

Table 1**Study Inclusion and Exclusion Criteria.**

Inclusion Criteria	
1	Age 24 months to 60 months at implantation, or up to 72 months in a child who has demonstrated only sound detection with a cochlear implant
2	Bilateral profound SNHL as documented through behavioral assessments
3	Thin slice MRI and CT imaging demonstrates, on one or both sides: <ul style="list-style-type: none"> • Cochlear nerve hypoplasia/aplasia OR • Cochlear aplasia OR • Cochlear ossification, post meningitis
4	Lack of audiological benefit with cochlear implant after 3 to 6 month appropriate amplification trial, waived in post-meningitis patients with CT evidence of severe ossification and in patients with cochlear aplasia.
5	Strong family support, to include language proficiency of both parents in the child's primary mode of communication and primary educational mode of communication, whether that is English or another form of visual support for communication such as American Sign Language, Signed English or Cued Speech.
6	Reasonable expectations from parents, to be determined informally by clinicians and psychologist, to include: <ul style="list-style-type: none"> • understanding of benefits and limitations of auditory brainstem implant • understanding of parental role in rehabilitation • understanding that the child may not be successful in developing spoken language as a primary mode of communication or in developing sufficient oral language skills to allow them to make maximum academic progress in an oral environment
7	The child's parents or legal guardian has proficiency in spoken and written English to enable completion of study-related questionnaires.
8	Informed consent from the child's parents/legal guardian.
Exclusion Criteria	
1	Cognitive and/or developmental delays which would be expected to interfere with the child's ability to cooperate in testing and/or programming of the device, in developing speech and oral language, or which would make an implant and subsequent emphasis on aural/oral communication not in the child's best interest
2	Anomaly/pathology involving the brainstem and/or cortex
3	Retrocochlear pathology resulting from NF2, or other types of cranial nerve/brainstem tumors
4	Co-existing medical conditions that require irradiation of the brainstem or brainstem tumors
5	Any medical contraindication precluding safe administration of general anesthesia, such as <ul style="list-style-type: none"> • Cardiopulmonary or • Renal disease
6	Otological conditions which contraindicate surgery <ul style="list-style-type: none"> • Active middle ear infection • Tympanic membrane perforation
7	Parents or legal guardians who are unable or unwilling to comply with the protocol

Table 2

Screening/Baseline and Postoperative Testing Protocol.

Interval	Medical	Audiology	Speech/Language	Education	Psychology
Screening/Baseline	<ul style="list-style-type: none">Physical exam	<ul style="list-style-type: none">	<ul style="list-style-type: none">Speech production	Review educational services	<ul style="list-style-type: none">Adaptive behavior
	<ul style="list-style-type: none">Neurotology exam	<ul style="list-style-type: none">	<ul style="list-style-type: none">Expressive/receptive language[†]		<ul style="list-style-type: none">Early non-verbal cognitive skills
	<ul style="list-style-type: none">Review of CT and MRI imaging with neuroradiologist	<ul style="list-style-type: none">IT-MAIS[*]	<ul style="list-style-type: none">Vocabulary[‡]		
		<ul style="list-style-type: none">MAIS^{**}	<ul style="list-style-type: none">Shared book reading		
			<ul style="list-style-type: none">Oral motor mechanism screening		
1 week, 3 & 12 months post surgery Neurotology exam					
1, 3, 6, 9, 12 months post activation		<ul style="list-style-type: none">		Review:	
		<ul style="list-style-type: none">Audiometric thresholds		<ul style="list-style-type: none">	<ul style="list-style-type: none">IFSP/IEP
		<ul style="list-style-type: none">Speech detection thresholds		<ul style="list-style-type: none">	<ul style="list-style-type: none">Progress reports
		<ul style="list-style-type: none">IT-MAIS[*]			
		<ul style="list-style-type: none">MAIS^{**}			
12, 24, 36 months post activation		<ul style="list-style-type: none">ESP^{***}			
			<ul style="list-style-type: none">Speech production		<ul style="list-style-type: none">Adaptive behavior
			<ul style="list-style-type: none">Expressive/receptive language		<ul style="list-style-type: none">Early non-verbal cognitive skills
			<ul style="list-style-type: none">Vocabulary		
			<ul style="list-style-type: none">Shared book reading		

^{*} Infant-Toddler Meaningful Auditory Integration Scale

^{**} Meaningful Auditory Integration Scale

^{***} Early Speech Perception Test

[†] The entire team informally evaluates the parents and child on their visual communication skills during the prescreening and candidacy stages of the trial. Those parents and children who show little to no signing skills are not enrolled into the trial, but are counseled about seeking out sign language training services. Those children who advance to the candidacy stage are assessed on informal and formal language measures by a speech-language pathologist skilled in manual communication. Testing is conducted using simultaneous communication (SimCom, also known as Total Communication).

Table 3

Patient Characteristics and Demographics.

Case #	Gender	Diagnosis	Imaging Findings (CT/MRI)	ABI Surgery	Age at ABI surgery (#or enrollment if no surgery)	Previous CI	Age at CI surgery	Contralateral CI left in place	ABI side	Length of Hospital Stay (days)	ICU Stay, Days (Effective Days)*	Number of Active Electrodes	Reason for Inactivated Electrodes
1	M	CND	SCC hypoplasia, hypoplastic IAC, CND	Yes	3y	Bilateral	1y9m	Left	Right	7	2	13	NAD
2	F	CND	Cochlea/SCCs normal; Isolated CND	Yes	3y	Unilateral	2y3m	Yes	Right	5	1	11	NAD
3	F	CND	SCC hypoplasia, hypoplastic IAC, CND	Yes	2y3m	Unilateral	1y	Yes	Left	4	1	9	NAD, VSE (1 electrode)
4	F	Cochlear hypoplasia/CND	Severe cochlear hypoplasia, hypoplastic IAC, CND	No	* 3y5m	No	n/a	n/a	n/a	n/a	n/a	n/a	n/a
5	M	Cochlear aplasia	Cochlear aplasia, SCC hypoplasia	Yes	4y11m	No	n/a	n/a	Right	4	1	11	NAD
6	M	CND	Cochlea/SCCs normal; Isolated CND	No	*2y11m	Bilateral	1y7m	n/a	n/a	n/a	n/a	n/a	n/a
7	F	CND	IP1 with possible CND	No	*2y4m	Unilateral	11m/2y2m	n/a	n/a	n/a	n/a	n/a	n/a
8	M	CND	IP1 with hypoplastic IAC, CND	Yes	4y	Unilateral	1y4m	Yes	Right	5	1	12	NAD
9	M	CND/Cochlear malformation/Microtia/Atresia	Atresia of EAC/middle ear. Absent stapes/round window, hypoplastic IAC, CND	In process	*2y10m	No	n/a	n/a	n/a	n/a	n/a	n/a	n/a

CND=Cochlear Nerve Deficiency

NAD=No Auditory Detection

VSE= Vestibular Side Effect

SCC=Semicircular canal

IP1=Incomplete partition type 1

* Effective Days in ICU refers to # of days requiring ICU care. In some cases patients were maintained for extra time in ICU due to lack of floor bed availability

Table 4

Surgical Anatomical Findings and Perioperative Adverse Events Requiring Intervention.

Case #	Vestibulocochlear Nerve Findings	Lateral Recess Anatomy	Perioperative Adverse Events Requiring Intervention *
1	absent	imperforate	<ul style="list-style-type: none"> CSF leak (CI explanted on same side as ABI), lumbar drain placed
2	absent	widely patent	None
3	absent	covered by thin arachnoid membrane	<ul style="list-style-type: none"> Perioperative anemia; blood transfusion Vestibular NASE on one electrode; electrode removed from map
5	hypoplastic – smaller than facial nerve	flocculus adherent to rhomboid lip	<ul style="list-style-type: none"> Influenza 5 weeks after surgery; delayed sedated activation
8	two distinct tiny nerves posterior to facial nerve, each originating from distinct areas of the petrous ridge	covered by thin arachnoid membrane	<ul style="list-style-type: none"> Preoperative iron deficiency anemia requiring iron therapy Postoperative nausea/vomiting; abdominal radiograph

NASE = Nonauditory Side Effect

* For a full table of all adverse events of all grades, see the table in the online supplement