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Pediatric Auditory Brainstem Implantation: Surgical, Electrophysiological and Behavioral Outcomes

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Abstract

Objective—Demonstrate the safety of auditory brainstem implant (ABI) surgery and the subsequent development of auditory and spoken language skills in children without Neurofibromatosis type II (NFII).

Design—A prospective, single-subject observational study of ABI in children without NFII was undertaken at the University of North Carolina at Chapel Hill. Five children were enrolled under an Investigational Device Exemption (IDE) sponsored by the investigators. Over 3 years, patient demographics, medical/surgical findings, complications, device mapping, electrophysiological measures, audiological outcomes and speech and language measures were collected.

Results—Five children without NFII have received ABIs to date without permanent medical sequelae, although 2 children required treatment after surgery for temporary complications. All children wear their device daily and the benefits of sound awareness have developed slowly. Intra- and post-operative electrophysiological measures augmented surgical placement and device programming. The slow development of audition skills precipitated limited changes in speech production but had little impact upon growth in spoken language.

Conclusions—ABI surgery is safe in young children without NFII. Benefits from device use develop slowly and include sound awareness and the use of pattern and timing aspects of sound. These skills may augment progress in speech production but progress in language development is

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dependent upon visual communication. Further monitoring of this cohort is needed to better delineate the benefits of this intervention in this patient population.

Keywords

Auditory brainstem implant; pediatric outcomes

INTRODUCTION

The Auditory Brainstem Implant (ABI) is a neurostimulator used to enable auditory perception for individuals who are deaf due to absent or damaged cochleas and/or cochlear nerves, thereby precluding the use of a cochlear implant (CI). The ABI bypasses the cochlea and the auditory nerve and directly stimulates the cochlear nucleus in the auditory brainstem. The ABI was initially developed by William F. House for use in patients with Neurofibromatosis type II (NFII), who were undergoing surgery for tumor removal (Edgerton et al., 1982; Eisenberg et al., 1987, Brackmann et al., 1993; Shannon et al., 1993). ABI usage was further expanded to patients without NFII by other investigators (Colletti et al., 2001; 2002; 2004; 2005; Nevison et al., 2002; Sennaro lu et al., 2009). These studies demonstrate the device can variably enhance lip-reading and sound awareness with open-set word recognition being unusual (Colletti & Shannon, 2005). Colletti et al (2009) and others (Choi et al., 2011) have since demonstrated better results in the non-NFII, adult population. They hypothesized that NFII and/or its treatment results in pathological changes in the brainstem and cochlear nuclei that might impede good performance with an ABI.

In theory, young prelingually deaf children may be better candidates than adults for ABI since they have substantial developmental plasticity and have no auditory memory that requires precise matching of signals. Colletti & Zoccante (2008) reported on 26 children (aged 14 months to 16 years), implanted between 1996 and 2006. Etiology of deafness was related to anatomical abnormalities for most children but more than half had a variety of other disabilities that could impact outcomes. Results indicated that all children consistently used their devices and demonstrated gradual improvement in communication skills and functional use of sound in natural environments but only one post-linguistic child developed open-set speech perception. Sennaro lu et al. (2009) also implanted 6 children with severe inner ear malformations. These children gained basic auditory function and were able to recognize and discriminate environmental sounds and two developed limited open-set discrimination. In follow up, these clinicians reported on 21 children with ABIs and concluded that the presence of additional handicaps is the most important factor limiting language development for children (Sennaro lu et al, 2011). A more inclusive and recent report of 60 children from the same implant center in Turkey indicated that of the 35 children with several years of device use, a statistically significant relationship existed between hearing threshold, speech intelligibility and language outcomes. Among 35 children, 29 (83%) had closed set discrimination and 12 (34%) developed open set discrimination above 50% (Sennaro lu, et. al, 2016).

After extensive experience with the use of CIs in children with severe inner ear malformations and/or cochlear nerve deficiency (Buchman et al, 2011), we developed

interest in the use of the ABI for these children in the US. A feasibility study was designed and developed at the University of North Carolina at Chapel Hill (UNC-CH) in 2012 and an investigational device exemption (IDE) was obtained to implant 10 children with carefully defined inclusion criteria. To date, 5 children have received an ABI at UNC-CH and have been followed for up to 3 years. This report describes clinical outcomes for this group and summarizes the surgical/medical, electrophysiology, audiology, and speech and language findings. The specific aims for the study were:

Aim 1: Demonstrate the safety of ABI surgery in children.

Aim 2: Demonstrate the development of sound awareness and improved speech understanding among children implanted with the ABI when compared to their baseline skills.

Aim 3: Demonstrate the development of oral language skills following the use of the ABI that were not evident prior to its use.

METHODS

The Biomedical Institutional Review Board (IRB) at the UNC-CH approved this study protocol.

Subjects

Children without NFII (18 months to 5 years of age) were included to minimize the adverse effects of auditory deprivation on speech perception, speech production and language. The lower limit of 18 months allowed for a CI at 1 year and at least 6 months of CI experience prior to considering the ABI. Children were excluded if they demonstrated known developmental or cognitive delays that could affect outcomes. More than 25 children were pre-screened and 13 children were evaluated on site. Five children determined to have age appropriate cognitive and developmental skills, as assessed by an experienced speech language pathologist (SLP), were selected for inclusion in this study.

Age at ABI surgery was between 2 and 5 years. There were 3 females and 2 males. All children were diagnosed with cochlear nerve deficiency (CND) based on review of imaging by the team surgeons. Two of the children were diagnosed with CHARGE syndrome based on the presence of 3/5 characteristic features (Hsu, et al 2014) but demonstrated normal cognition. The other three had no other significant medical diagnoses beyond hearing loss.

Three of 5 children had previous CIs (2 unilateral and 1 bilateral). The two children with unilateral CIs were not wearing their devices due to lack of sound awareness or intolerance. For S1, the CI was removed to place the ABI because he had a shunt on the opposite side. For S5, CI device use was reintroduced and reprogrammed about 6 months after ABI surgery. One bilateral CI recipient, S4, used two devices consistently before and then after the ABI was programmed. She was the oldest child enrolled who also had the longest history of CI use. Prior to ABI surgery with her bilateral CIs, she was able to detect sound and identify some suprasegmental cues with her CI, but she had plateaued in progress with speech production and auditory discrimination skills after 4 years of intensive therapy.

Parents were advised to supplement their child's communication with a visual system to support language growth while working on auditory skills development. Because the children lived in 5 different states, the educational environments and services they received were unique and beyond the control of this investigation. In some states, limited communication mode options were available. All families received support from the team SLP and audiologist either through direct therapy, tele-therapy or in-service to local educators.

See Table 1 for communication mode choices and further demographic details.

Surgery

In all cases, the Nucleus 24 Multichannel ABI (Cochlear Corp., Sydney AU) was placed using a retrosigmoid craniotomy. This approach provided direct access to the lateral recess of the 4th ventricle and region of the cochlear nucleus. It has previously been documented to be safe for this use (Colletti et al., 2010) and has been used extensively in adults and children at our institution for other purposes. Surgery was carried out under general endotracheal anesthesia without paralytic agents, which was administered by a pediatric anesthesiologist. Cranial nerve electromyography monitoring consisted of bipolar, needle electrodes placed in the ipsilateral facial muscles (orbicularis oris and oculi-CN VII), soft palate (CN IX-X), and trapezius (CN XI). Appropriate monopolar needle electrodes served as ground and stimulating electrodes and were placed in the pre-sternal subcutaneous skin. The NIM-Response® 3.0 (Medtronic, Minneapolis, MN) nerve integrity monitor was used to continuously monitor EMG activity under both spontaneous conditions, as well as under evoked testing.

A standard retrosigmoid craniotomy based upon the sigmoid and transverse sinuses was performed. The mastoid air-cells, if present were covered with either bone wax or dry DuraPair (Medtronic). Dural incisions were made in cruciate fashion. Under microscopic view, CSF was drained from cisterna magna and the lateral cerebellopontine angle cistern. After cerebellar relaxation, the lower cranial nerves were identified, and the cranial nerve IX and choroid plexus emanating from the Foramen of Luschka were viewed with gentle retraction of the cerebellar flocculus. The facial nerve, ventral to the lateral recess and cochleovestibular nerve and/or its remnant, as well as the lower cranial nerves exiting inferiorly were identified and confirmed with stimulation. The choroid plexus was retracted posterior and superior to reveal the cochlear nucleus prominence on anterior wall.

Prior to inserting the electrode, the receiver-stimulator of the Nucleus 24 Multichannel ABI was seated in a depression on an appropriate site on the skull, based on methylene blue markings placed prior to incision. The receiver-stimulator was immobilized with periosteal sutures and a trough for electrode immobilization created if skull thickness permitted. Next, the array was advanced into the recess with the array facing the nuclear prominence. Teflon® felt was used to secure the array at the level of the lateral recess. The separate ground, ball electrode was advanced under the temporalis muscle periosteum in a standard fashion.

Intraoperative testing

The intraoperative electrically evoked auditory brainstem response (eABR) testing was undertaken to provide electrophysiological information concerning device placement and functionality. The set-up and paradigm for this testing are detailed elsewhere (Nevison, 2006). The number of averages was dictated by the quality of the response and was typically set to 500–1000. With the onset of stimulation, the morphology of the response was anticipated to appear as 1, 2 or 3 positive peaks with latencies of about .6 – .8 milliseconds with possible second peak at around 1.2 – 1.9 milliseconds and a third peak if present occurring approximately a millisecond later. When responses were observed, pulse polarity was reversed to confirm the neural basis. Stimulation and recording started with wide bipolar stimulation between stimulating electrodes transverse across the length of the device with subsequent pairings honing in on more spatially localized stimulation, as dictated by the pattern of results and available testing time. In four of the five pediatric cases, clear responses were recorded at the first placement of the paddle. Figure 1 demonstrates the orientation of the device relative to the cranial nerves with both right and left side device placement. When pairs of electrodes were stimulated, as described, consideration of non-auditory cranial nerve excitation was given by monitoring facial (CNVII), glossopharyngeal (CNIX), and vagus (CNX) nerve stimulation through activation of the facial nerve monitor and changes in respiration and heart rate.

At 3 weeks post-operatively, the child was sedated for suture removal and eABR using the identical montage to gain information that would circumvent non-auditory stimulation at the time of the initial stimulation. While sedated, monopolar stimulation (300- μ Sec/phase and 220 clinical units) was used to determine if facial or vagal nerve responses occurred.

Device activation and programming

Initial device stimulation took place in a hospital setting with pediatric anesthesia support. As with CIs in young children, behavioral observation and conditioned behavioral audiometry techniques were used to determine electrical thresholds (T) and comfortable (C) listening levels. The goal was to activate the maximal number of electrodes that would yield the best detection audiogram without non-auditory stimulation (e.g. facial contraction, throat tickle, cough, shoulder activation, extremity sensations, head and body tilt). Starting with electrodes that elicited robust intraoperative eABR, stimulation commenced using monopolar grounding and an ascending approach, until a behavioral response was observed. Frequency bands were assigned to electrodes per the standard allocation table for the number of active electrodes. After all channels were explored, subsequent programming sessions took place at the pediatric cochlear implant clinic.

During the first few months of device use, families stayed locally for several days at a time for programming sessions. Each child was trained to participate in a conditioned response and was closely observed for aversive responses. Generally, the programming sessions were similar to CI recipients using a progressive mapping approach that included making incremental changes to charge [nanoCoulombs (nC)] levels over time, allowing for comfortable adaptation to sound, and monitoring for sound detection through sound booth

testing. The children were seen for device programming at regular intervals during the first 6 months and then every 6 months thereafter.

Assessment of Audition, Speech Perception and Speech and Language

Standard audiometric techniques were used to measure sound field hearing thresholds with the ABI activated. Visual reinforcement audiometry was used until the child was old enough to participate in conditioned play audiometry. Stimuli included live-voice speech and warbled tones or narrow-band noise at 125, 250, 500, 1000, 2000, 4000, and 6000 Hz. A speech perception assessment battery was chosen that included parental report of hearing; four or twelve choice closed set identification of word pattern and single words; and open-set word understanding as measured by common tests used with the CI population. Speech and language assessment was completed pre- and at 6-month post-operative intervals using a battery of age-appropriate measures. Appendix A (see Supplemental Digital Content 1, which describes test materials) includes a summary of the speech perception, speech production and language assessment battery used in this study.

Electrophysiological Measures of Electrically-evoked Event Related Potentials (eERP)

eERPs in children with ABIs have been described in previous studies (He et al., 2015, 2016). Only a brief summary is included in this report. In short, the stimulus was a train of biphasic, charge-balanced electrical pulses. The ABI speech processor was bypassed and the electrical stimulus was directly delivered to individual ABI electrodes using a research interface. Pulse duration and stimulating electrode were selected for each subject based on their programming maps.

Electroencephalographic (EEG) activity was recorded using a Neuroscan system (version 4.4), a SynAmp² amplifier (Compumedics, Charlotte, NC) and disposable surface electrodes on the scalp. The EEG was sampled at 1000 Hz, baseline corrected, amplified with a gain of 10, online filtered between 0.1–100 Hz prior to averaging. The artifact rejection threshold was $\pm 100 - 150 \mu\text{V}$. For every subject, at least two replications of 100 artifact-free sweeps were recorded for each stimulation condition. These replications were offline filtered between 1–30 Hz and averaged prior to data analysis.

In these five children with ABIs, we 1) investigated the feasibility of measuring the onset eERP and the electrically evoked auditory change complex (eACC); 2) examined the association between onset eERP morphology and auditory/non-auditory stimulation; and 3) evaluated the possibility of using the onset eERP to determine the auditory detection threshold (T level) for individual stimulating electrodes.

Results

Surgery—There were no cases of cranial nerve or dural sinus injury, cerebellar swelling or contusion, subarachnoid hemorrhage or brainstem injury. In all subjects, the choroid plexus and the lateral recess of the 4th ventricle (Foramen of Luschka) were identified without difficulty. The electrode array was advanced in to the recess fully in all cases except S2. In that case, the last row of 3 (of 21) electrodes was visible at the recess entrance. Only this

case had no intra-operative electrophysiological responses recorded. Postoperative CT scans were taken on the first night after surgery in all cases as shown in Figure 2.

Postoperatively, S1 and S2 experienced complications while no significant issues were encountered with the other 3 children, all being discharged within 4 days of surgery. S1 is a child with CHARGE syndrome and known subglottic stenosis with baseline stridor (albeit mild). Following surgery, stridor and cough induced a pseudomeningocele at the operative site followed by CSF wound leakage requiring a second surgery to reinforce the closure and placement of a lumbar drainage. Once resolved, no other medical issues were encountered. For S2, surgery and the immediate post-operative period were uneventful. On day 10, she developed a fever, WBC 24.9 and meningeal signs necessitating lumbar puncture. CSF analysis revealed 1060 nucleated cells/mm³ (80% neutrophils), protein 65 mg/dl and glucose 51 mg/dl. Presumptive diagnosis was bacterial meningitis although gram stain and all cultures were negative throughout (i.e. aseptic meningitis). She was treated with 6 weeks of intravenous vancomycin and ceftriaxone with resolution.

Intra-operative electrophysiological measures—Intra-operative eABRs were recorded in all subjects except for S2. These eABR responses consist of up to three vertex positive peaks within the first 4 ms after the stimulus, which is consistent with those reported in literature (O'Driscoll et al., 2011). For S2, no eABR responses were recorded despite multiple attempts using different recording systems. Factors accounting for the lack of response in this subject remain unknown. Figure 3 shows intra-operatively recorded traces for the electrode pair of 8 □ 3 in S3. These responses were recorded at a stimulation level of 150 clinical units. Robust eABR responses could be easily identified in all traces. These eABRs showed two vertex positive peaks: P1 and P2. Peak latency for the P1 and P2 was around 1 ms and 2.7 ms, respectively.

Post-operative electrophysiology—Post-operatively, both the onset eERP and the eACC to temporal gaps could be recorded. Morphological characteristics of the onset eERP showed substantial variations among subjects, as well as across stimulating electrodes within the same subject. Signs of non-auditory stimulation were only observed when recorded eERPs showed multiphasic morphologies with large amplitudes (i.e. Type II response, He et al., 2016). Compared with the onset eERP, the eACC showed less inter- and intra-subject variations in morphological characteristics. Figure 4 shows objective thresholds determined using eERP recordings and behavioral T levels measured using clinical procedures in four subjects (i.e. S1, S3, S4 and S5). For each subject, one or more electrodes were tested. Results of the one-tailed Spearman Rank correlation test showed a statistically significant correlation between thresholds determined using these two procedures ($\rho=0.89$, $p<0.01$). Overall these results showed a robust correlation between T levels measured using behavioral procedures and electrophysiological measures of the onset eERP. Finally, the onset eERP had good test-retest reliability for results recorded in two test sessions with an inter-session interval of up to six months. However, poor repeatability was recently observed in some children at longer time intervals due to changes in morphological characteristics of the onset eERP. Potential factors accounting for these changes might include cortical maturation, change in stimulation level and possible electrode migration. These newly

observed long-term changes in the onset eERP are reported in a companion paper (He et al., submitted for publication).

Device programming—The process of programming the speech processor was dynamic and variable for each child, depending on his or her ability to cooperate and provide reliable information during behavioral testing. Over all cases, as many as 20 and as few as 10 electrodes were activated and then the number of active channels decreased over time. This occurred either because non-auditory behaviors were observed or reported by the child or because behavioral audiograms suggested poor or absent detection at corresponding frequencies. As might be anticipated based on the electrode and neural tissue interface, active electrodes tended to be adjacent and localized to an area of the paddle. The relationship between pulsewidth (PW) and current level (CL), which determines the overall amount of charge (nC), was systematically altered to find the lowest PW that resulted in desirable sound detection levels and no non-auditory stimulation. This was achieved for two children (S1 and S3) by using variable PW; the remaining three children had a fixed setting ranging from 100 to 300 PW. As electrodes were eliminated, children had between 3 and 13 active electrodes from the 20 available in the SPeak coding strategy. During the course of the study, a relationship between active electrodes, behavioral levels and eERP onset responses was revealed and this information was used to corroborate electrode selection and setting decisions (He et al., 2016). For example, a lack of eERP on a particular electrode aligned with a poor detection response in the region of the assigned frequency band. Once that channel information was assigned to another viable electrode, detection improved. Program parameters became relatively stable for most children between 6 and 9 months post activation. However, S2 did not exhibit a behavioral response during the first year after device activation. For her, a program was created with all electrodes active using high charge levels. By her 18-month follow up visit, parental reports suggested she was beginning to detect sound. Through a process of channel elimination and behavioral testing, three electrodes were eventually determined to provide optimal detection results. Figure 5 shows the best-aided audiogram for each child and the number of active electrodes, along with program parameter details. Subjects 4 and 5, who also have a CI, did not wear the CI initially but resumed use after detection responses became stable with the ABI. CI parameters were not changed for S4 but modifications were made for S5 compared to original settings.

Audition and speech perception outcomes—The time required to obtain the first evidence of sound detection in the sound booth with the ABI varied from 2 weeks (S1) to 18 months (S2). The three children with previous use of CIs had variable experience with sound (S1, S4, S5). Eventually, all five children participated in reliable sound field audiometric assessment indicating sound detection across octave frequencies between 125 Hz and 6 K Hz in the range of 20 to 50 dB HL.

Demonstration of meaningful use of sound, as measured by parental questionnaire, evolved slowly for each child and no child reached a ceiling on the Infant-Toddler- Meaningful Auditory Integration Scale (IT-MAIS). In contrast, children with CIs often demonstrate change on similar measures within months 3–6 of CI use (Osberger, et al., 1997). Scores

fluctuated over time for S1–4 and remained low for S5. This variability may be evidence of the fact that change occurred slowly and perception of benefit was influenced by parent projection, which is a disadvantage of parent reported scales. No child was able to progress in the hierarchy of speech perception skills beyond closed set measures. No child in the present study demonstrated open set speech perception during formal assessment using the ABI alone. See Table 2 for individual scores over time.

Speech and language outcomes—One of the aims in this study was to demonstrate the development of oral language skills following the use of the ABI. Results of the articulation test, *Identifying Early Phonological Needs in Children with Hearing Loss (IEPN)*, are shown in Table 3. All subjects were observed to vocalize at will, with 80% vocalizing on demand. Syllable development is shown in 80% of the subjects by 2.5 years post-ABI.

S1 demonstrated an increase in consonant development at 2 years and by 3 years, appropriate voicing of consonants with 90% accuracy. Manner and place of consonants is 56% and 58% accurate, respectively. He includes initial and final consonants when producing words, but not necessarily the proper consonant sound. Central and back vowels are developed, but front vowels are limited.

S2 vocalizes on demand and produces sounds that are visually accessible (/b/, /p/, /w/). Development of vowels and non-visible consonants has not developed 3 years post activation.

S3 has a repaired cleft palate and vocalizing on a sustained breath or producing plosive sounds has been difficult due to structural impairments and poor motor kinesthetic control. Although the IEPN shows minimal growth for S3, his production of syllable patterns in frequently used words (i.e. no, mama, up-up-up) has increased. Central vowels and approximations of the consonants /p/, /m/, /b/ and /n/ have been observed.

S4 was able to imitate approximations of initial consonants /f/, /m/, /b/, /h/, and /ʃ/ as well as the vowels /u, I/ prior to ABI surgery. After 2.5 years of use, she imitated syllable structures and marked initial and final consonant placement with an appropriate or a substituted consonant most of the time. Appropriate vowel and consonant production continues to improve.

S5 vocalizes but has not developed any speech at 2 years of use. Although she spontaneously vocalizes, vocalization on demand has not been observed.

Language was assessed using the *Oral and Written Language Scales 2 (OWLS 2)*. This test evaluates expressive and receptive language skills and also yields a composite score; however, it is not normed for children with hearing loss. Standard scores from 85 to 115 suggest language skills within normal limits for children with normal hearing. The test was administered following standardized administration procedures using spoken language. Visual cues from lip reading were available but Cued Speech or Sign Language was not used to obtain these scores. Table 4 shows S1 to have the highest standard score for receptive language, but his expressive language is severely delayed. S2, S3 and S4 have spoken

language scores in the severely delay range. S5 obtained a raw score of 0. The standard scores shown are a statistical extrapolation from the overall distribution of raw scores rather than a reflection of spoken language skills (Carrow-Woolfolk, 2011).

Since all children were using visual support for language, these scores cannot be considered a measure of language competence in their typical communication mode and underestimate language skills. However, for the purpose of this study with the aim of assessing the potential to develop oral language, this standardized test was used.

Although formal testing of the children's visual coded language was not conducted in this study, informal observations reveal that all of the subjects have delays in their language system; however, all are using a visual system in a functional approach to communicate with others.

DISCUSSION

This feasibility study demonstrates the surgical procedure for ABI placement in young children (2–5 years old) is safe, though not without the potential for complications, when performed by experienced surgeons in a supportive, tertiary care environment. Five children with a variety of temporal bone morphologies and CND were successfully implanted and managed without long-term medical complications, to date. Four of 5 children had robust intraoperative responses on eABR testing, implying auditory stimulation and appropriate device placement. Behavioral testing revealed relatively good audibility for 4 subjects and with one (S2) developing delayed audibility after 1 year of device use. eERP testing showed good correlation with behavioral thresholds and some evidence of temporal processing by the auditory system. Unfortunately, the temporal resolution for gap detection following ABI is far less than that observed for children with normal auditory systems. These findings may underlie the poor speech perception development over time, despite nearly 3 years of device use. We will continue to follow their progress closely to better understand their ultimate potential.

At the onset of the study it was our intention to include children who were congenitally deaf as well as those with post-linguistic onset of deafness. Given the known importance of previous experience with sound and spoken language exposure for CI outcomes, we hypothesized that children with delayed onset deafness, perhaps as a result of a sudden traumatic injury, meningitis, or progressive loss of CI function due to cochlear ossification/fibrosis without the potential for re-implantation, would have more potential to use the novel signal. Despite our best efforts, no post- or peri-linguistically deafened children were identified. Other investigators however, have implanted such children with encouraging results (T. Roland, J. Golfinos, and W. Shapiro, personal communication, January, 2017). It should be noted that all of the children in this study are cognitively and developmentally on track, with the exception of their communication skills. A majority of children who may not be candidates for a CI due to anatomy issues also often have other developmental concerns that may result in even less favorable results than reported here.

In general, reliable ABI use in this relatively young group of pre-linguistic children resulted in sound detection and awareness when in a controlled environment but limited speech understanding in a closed set environment and no open-set speech perception. One could argue that these findings relate to the children's underlying anatomic and physiologic substrate. Maybe the auditory system development was quite limited or non-existent prior to ABI surgery and electrical stimulation with the ABI was not sufficient for inducing auditory development within the period of observation. This certainly was the case for S2, where no cochlear remnants were apparent and no eABR was recorded in the OR, despite good electrode placement. Another plausible explanation is that the ABI, in its current format, fails to provide the temporal and spectral variation needed to induce auditory system development and maturation. While the underpinnings of this finding might relate to coding strategies, electrode/neural interface shortcomings, or unique auditory system structure and function, the poor speech perception and production results supports this notion. While some investigators have demonstrated open set speech perception following ABI in pre-linguistic children, this has been the exception rather than the rule (Noij et al, 2015).

As a medical intervention, the potential risk profile for ABI surgery is higher than that of CI surgery. In fact, S1 and S2 experienced perioperative complications rarely seen following CI. For S1, CSF leakage was likely related to perioperative airway obstruction from his underlying subglottic stenosis that was exacerbated by endotracheal intubation. The resulting abnormally high CSF pressures played some role in the observed wound leakage. Being our first case in the series also might have contributed to the complication. The latter explanation seems less likely given our significant experience with intracranial surgery and ABI placement in adults. Either way, future ABI surgery in similar children should account for this potential problem. S2 developed perioperative signs and symptoms of meningitis 10 days following surgery despite the usage of appropriate perioperative antibiotics and steroids. This complication seems far less preventable and is expected to occur in some proportion of all intracranial surgeries. While no permanent medical issues resulted in this cohort, we remain wary of the potential for significant perioperative complications and the unknown long-term consequences. Specifically, perioperative risks for all intracranial procedures include bleeding with the potential for permanent neural dysfunction (i.e. stroke), infection, and hydrocephalus. Device related issues that are specific to ABI might include electrode paddle migration or the need for revision for device failure. We will continue to monitor these children should such problems become evident.

Given the fact that ABI in this young, pre-linguistic group of children resulted in sound awareness alone with limited speech perception or spoken language development, it might be argued that the risk to benefit relationship is not favorable enough to warrant further ABI placement in similar patients. It was always our intention at UNC-CH to implant only 5 pre-linguistic children, to observe their development, prior to considering others for this surgery. Of interest, many of the parents are happy that they chose to pursue the ABI and many feel that sound awareness alone was sufficient for considering the risk. Should the children in the present study develop greater skills in the future, the risk to benefit profile would surely change and need to be reconsidered.

Intra-operative eABR measures validated the electrode paddle placement during surgery and guided device programming decisions. Stimulation and programming of the speech processors commenced conservatively and in an environment of careful observation for aversive effects. The creation of these programs required more time relative to CI programming but resulted in a similar outcome of consistent detection of sound across frequencies in the sound booth. Sennaro lu and colleagues (2016) observed no relationship between the number of active electrodes and hearing thresholds or language outcome in their larger set of pediatric ABI recipients. In this cohort, there was some association with number of channels and pure tone average (PTA), as shown in Figure 5. The child with the most active electrodes also demonstrated the lowest PTA and the child with the fewest active electrodes had the highest PTA. Overall charge levels were calculated using a formula to consider pulse width and current level for the CI24M receiver and then the average charge per channel was determined. Measures varied from S2, who was diagnosed with Michel Aplasia and required the greatest charge per channel, and S4 and S5 who had normal cochlear anatomy and required the least charge per channel. All recipients demonstrated improved sound detection over time that stabilized with extended device use.

The audiologic and mapping paradigm for children with ABIs requires special considerations that are time consuming, tedious, and not apparent for the routine CI patient. First, when compared to CI patients, behavioral observation during mapping requires the consideration of non-auditory stimulation, which at times can be subtle and difficult to detect. Moreover, given the unreliable and potentially aversive relationship between neighboring electrodes, comprehensive electrode interrogation must be utilized rather than more traditional interpolation techniques. Toh and Luxford (2008) reported non-auditory side effects (NASE) in 42% of multichannel ABI users. Colletti, et al, (2010) reported that ipsilateral body tingle was observed in 2 children (6.9%), facial nerve stimulation in 2 (6.9 %), dizziness in 1 (3.4%), headache in 1 (3.4%), and throat tingle/tickle in 1 (3.4%) child. These side effects were reportedly alleviated with selective electrode deactivation. In the present study, NASE were observed in 3 children and were persistent overtime despite reprogramming. It remains unclear why these findings are different than those previously reported. It may relate to our use of electrophysiological techniques to confirm auditory stimulation and the ability of our children to report NASEs.

The robust correlation observed between auditory detection thresholds measured using eERPs and those measured with behavioral procedures is one of the most auspicious findings of the study to date. The atypical eERP morphology evoked by non-auditory stimulation is similarly noteworthy. Specifically, our results showed that the Type I onset eERP was not associated with NASE in any patient (He et al., 2016). In contrast, an association between the Type II onset eERPs and NASE were observed in several cases (He et al., 2016). This finding, if confirmed by results of future studies, would be extremely useful during device programming, as it will allow clinicians to better select active ABI electrodes for individual patients at an early post-implant stage. Further work in this area is clearly warranted, as it portends an objective approach that may improve the accuracy and efficiency of the mapping process.

After about 3 years of use, the ABI does provide reliable sound detection in a structured environment but, so far, limited ability to afford improved speech understanding and guarded perceived benefit by the parents to provide meaningful use of sound for communication. This may change with continued use. Other reports of speech perception benefits for ABI recipients have indicated that a longer post implant period of use is needed before measurable benefits are observed; some open set word understanding may emerge as the pool of pediatric recipients increases (Noij, et. al, 2015). The test battery chosen in the current protocol includes typical assessment measures used with children with CIs. However, speech perception assessment following this model proved to be overly ambitious. The lack of spoken language experience and minimal segmental cues afforded by the ABI resulted in floor affects. It is important to note, however, that sound awareness skills alone were reported to improve quality of life per parental account. Unfortunately, a quality of life measure that is sensitive to sound awareness was absent in this study. All of the children were using visual support for communication in the form of Cued Speech or Sign Language; therefore, opportunities to observe use of sound only for communication were limited.

Comparing children with CND treated with ABI to those treated with a CI is worth considering, given the different risk profile for the 2 surgical procedures. We have previously reported outcomes of 50 children with CND who have received a CI (Teagle, et al 2015; Buchman, et al 2011). In those studies, 38% (19/50) either did not obtain useful auditory stimulation or obtained insufficient benefit to continue consistent device use and became non-users. The remaining 62% (31/50) obtained sound awareness from their implants. Of this group, slightly more than half obtain pattern and duration cues that supplement communication and about 16% actually achieved some limited (~35% correct) open set word understanding. All but 1 relied on visual support for communication. Comparatively, results from the present study suggest a higher success rate for gaining sound detection with ABI (100%), similar limited benefit for detecting the supra-segmental features of sound, and very limited or no measurable open set word understanding. As the duration of use is much longer amongst the CI study cohort, future comparisons between these groups will continue to be useful.

With regard to oral language skills development, changes in speech production skills were measurable in 4 of 5 children. By providing sound awareness, the ABI recipients demonstrated development of supra-segmental qualities for duration, intensity and pitch in their speech production that may have led to improved vocal quality and intelligibility with context. In the area of language development, tests chosen were not sensitive to the slow and small changes that occurred over time. The OWLS 2 was chosen due to the ability to compare standard scores from the age of 3 years until the age of 21 years. In addition, previous multi-center studies used this test for cochlear implant subjects (Fink, et.al, 2007). However this test was too global and did not capture the minute changes that occurred from one test date to another. This paucity of appropriate speech perception and language assessment materials became evident as the study progressed and has initiated the development and use of other assessment and monitoring tools that may be more sensitive to behavioral changes in children who are learning to use auditory information.

Visual support of language was recommended to all parents at the initiation of the study due to the previously reported findings and concern for creating further language delays due to auditory deprivation. However, the importance of structured opportunities to work on listening skills was also strongly recommended and implemented for most of the children. Those who participated in regular auditory therapy (S1, S3 and S4) demonstrated improvement during therapy sessions but these skills appeared to have been “green-housed” to the therapy room. Robbins (1994) describes this effect as the overtraining of certain skills in isolation that fail to generalize to impact broader communication skills. Although one subject developed functional spoken language with the use of Cued Speech and the ABI, development of spoken language without a visual system within this study has not materialized. All subjects need a visual system to communicate expressively and to understand others.

Finally, from a clinical perspective, one of the most important findings from this investigation was the need for high-level communication among team members. From candidate selection to device placement and programming, and ongoing assessment and habilitation, consistent communication and consideration of each child’s course of progress was felt to be essential. Additionally, the experience of this study confirmed that all auditory implants are not equal. CIs are one of the most effective treatment options for sensory loss in modern medical history. Our knowledge of the cochlea, its structures and neural interface to the brain, has been made more transparent by the advent of CIs. Potential exists to learn more about higher auditory structures and function through the use of implantable technology but current devices and practices are not sufficient to span this breach in knowledge.

At this time, it seems apparent that the ABI device and processing strategies we have available provide limited timing and spectral cues, which do not afford resolution of speech at a natural conversational rate. There is a need to better understand cortical neural encoding and processing of temporal and spectral information provided by electrical stimulation in children with ABIs and how these cues relate to speech production and pattern perception. Further studies focusing on these directions are warranted.

CONCLUSIONS

Given the surgical risks, complexity of device programming, and our limited clinical experience, except in the case of a completely isolated or absent cochlea, CI should continue to be pre-requisite to obtaining an ABI. The children included in this study wear their devices consistently. Achievement of functional listening skills includes sound awareness, increased vocalization and use of pattern and timing features of sound. Progress in speech production appears to have been supplemented by sound detection and temporal and intensity cues. Progress in language is dependent upon visual communication. Electrophysiological measures corroborate behavioral measures and supplement speech processor programming decisions. Continued monitoring of this initial cohort of children will determine if greater benefits are observed over time. Improved technology coupled with the development of objective techniques for reliably placing and programming ABIs may be needed before use of these devices becomes mainstream.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Nerve Proximity by Side

Figure 1.

Schematic of the Cochlear Nucleus ABI24M electrode pad. Orientation of the electrodes relative to the cochlear nucleus varies depending on right or left side placement. This illustrates the potential for other cranial nerve stimulation. Image courtesy of Cochlear Americas, ©2017.

Figure 2.

Post-operative scans for each of the five pediatric ABI recipients.

- A) Subject 1. Post-operative CT scan was obtained showing ABI in good location on the left side and no evidence of intracranial hemorrhage
- B) Subject 2. Absent cochlea bilaterally. ABI paddle in expected position on left side.
- C) Subject 3. Cochleae appeared isolated from the internal auditory canal and thus cochlear implantation was not undertaken. ABI on right side.
- D) Subject 4. CI seen in cochlear on left ear, ABI placed on right side.
- E) Subject 5. ABI on right side, cochlear implant in left ear. Artifact from the ABI internal receiver obscures imaging.

Figure 3.

Intra-operative eABRs recorded for the electrode pair of 8 and 3 in S3. Solid and dashed lines indicate replications recorded for the same stimulation condition. Black and dark grey lines indicate responses recorded using biphasic pulses with reversed polarity. Vertex positive peaks are labeled for these traces.

Figure 4.

Objective and behavioral T Levels measured in four subjects. Each symbol represents results measured for one electrode. Results measured for different electrodes in the same subject are indicated using the same type of symbols. The dashed line presents result of linear regression.

Figure 5. Audiograms, active electrode configurations, ABI program parameters for 5 subjects. Subjects 4 and 5 continue to use a CI in the contralateral ear and CI responses are designated as CL. All children were programmed in a SPeak coding strategy with variable numbers of active electrodes, current levels and pulse width. Average charge per channel [nanoCoulombs (nC)] was calculated by converting clinical units used in the software using the formula $(100^{(\text{clinical level}/255)} \times 17.5) \times (\text{pulse width}/1,000)$.

Table 1

Demographic information

	S1	S2	S3	S4	S5
Age at ABI	3.3	2.5	3.5	5.5	2.1
Gender	M	F	M	F	F
ABI Side	L	L	R	R	R
Etiology/Imaging	CHARGE/Hypoplastic Type III cochleas	Michel Aplasia/Absent cochleas	CHARGE/Patent cochleas isolated from IAC	UNK CND Patent cochleas, small IAC	UNK CND Patent cochleas, small IAC
CI Previous/Current	Yes/No	No/No	No/No	Yes/Yes	Yes/Yes
Length of Use	3.5 yrs	3 yrs	2.5 yrs	2 yrs	2 yrs
Communication Mode	Cued Speech	TC ASL	TC Signed English	TC Signed English	TC ASL

Demographic information for five children who received a Cochlear Nucleus ABI.

CHARGE includes the cluster of these features: Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and development, and Ear abnormalities and deafness.

UNK CND= Unknown Cochlear Nerve Deficiency.

Communication mode varied for each child based on support available in their location. Total Communication (TC), implies the use of all forms of communication, including lip reading, gesture, and simultaneous voicing of spoken language. With American Sign Language (ASL) signed words may not match spoken word order or phrasing. With Cued Speech and Signed English, visual cues supplement and follow a spoken language format.

Table 2

Speech Perception scores over time

Subject	Tests	Pre-ABI	6 months	12 months	18 months	24 months	30 months	36 months
S1	IT MAIS	3%	22%	43%	65%	32%	20%	42%
	ESP	CNT	CNT	(LV) 25% pattern 58% monosyll	(LV) 100% pattern 75% monosyll 67% spondee	(LV) 100% pattern 75% monosyll 67% spondee	(ST) 66% pattern 66% word 25% spondee 25% monosyll	(ST) 70% pattern 62% word 41% spondee 50% monosyll
	5 F PTA		30 dB HL	30 dB HL	30 dB HL	42 dB HL	40 dB HL	31 dB HL
S2	IT MAIS	0%	3%	28%	20%	12%	35%	72%
	ESP	CNT	CNT	CNT	CNT	CNT	(LV) 25% pattern	(LV) 25% pattern
	5 F PTA		NR	NR	48 dB HL	50 dB HL	35 dB HL	35 dB HL
S3	IT MAIS	0%	30%	52%	67%	70%	62%	32%
	ESP		CNT	CNT	(LV) 91% pattern 33% monosyll 25% spondee	(LV) 83% pattern 25% monosyll (ST) 66% pattern 41% word	(ST) 37% pattern 20% word 8% spondee 8% monosyll	(ST) 45% pattern 29% word 8% spondee 8% monosyll
	5 F PTA		42 dB HL	30 dB HL	28 dB HL	32 dB HL	30 dB HL	29 dB HL
S4	IT MAIS	75%	70%	50%	55%	55%	42%	
	ESP	(ST) 29% pattern 16% word 29% spondee 29% monosyll	(ST) 46% pattern 29% word 16% spondee 8% monosyll	(ST) 50% pattern 45% word 25% spondee 25% monosyll	(ST) 33% pattern 16% word 8% spondee 12% monosyll	(ST) 45% pattern 41% word 25% spondee 16% monosyll	(ST) 50% pattern 37% word 20% spondee 20% monosyll	
	5 F PTA	RE 57 dB HL LE 41 dB HL	34 dB HL	31 dB HL	23 dB HL	33 dB HL	29 dB HL	
S5	IT MAIS	0%	5%	2%	5%	5%	5%	
	ESP	CNT	CNT	CNT	CNT	CNT	CNT	
	5 F PTA	CNT	56 dB HL	34 dB HL	34 dB HL	32 dB HL	37 dB HL	

The Infant-Toddler Meaningful Auditory Integration Scale (IT MAIS) is questionnaire collected from caregivers based on 10 probes of auditory behaviors. The Early Speech Perception Test (ESP) is presented as a 4 choice test in the Low Verbal version (LV) or as a 12 choice test in the Standard (ST) version. Pure tone average (PTA) is based on five frequencies (125, 250, 1K, 2K, 4K Hz). 36 month data is not yet available for S4 and S5.

Table 3

Longitudinal results of *Identifying Early Phonological Needs in Children with Hearing Loss* measured in all five subjects.

Test Interval	Vocalize on Demand	Syllables	Initial Consonants	Final Consonants	Vowels	Manner Cues	Place Cues	Voice Cues
1 yr	100%	40%	63%	29%	12%	18%	20%	16%
2 yrs.	100%	80%	95%	64%	31%	54%	58%	67%
S1	3 yrs.	100%	100%	94%	53%	56%	58%	90%
	1 yr.	CNT	CNT	CNT	CNT	CNT	CNT	CNT
	2 yrs.	100%	40%	66%	0%	12%	24%	41%
S2	3 yrs.	100%	80%	75%	0%	26%	27%	25%
	1 yr.	CNT	CNT	CNT	CNT	CNT	CNT	CNT
	2 yrs.	CNT	CNT	CNT	CNT	CNT	CNT	CNT
S3	2.5 yrs.	44%	16%	11%	20%	4%	6%	0%
	1 yr.	100%	71%	11%	28%	35%	28%	41%
S4	2 yrs.	100%	38%	41%	17%	25%	21%	20%
	1 yr.	CNT	CNT	CNT	CNT	CNT	CNT	CNT
S5	2 yrs.	CNT	CNT	CNT	CNT	CNT	CNT	CNT

CNT: cannot be tested

This test is used to assess a child's use of sounds in single word productions. The result reflects the percentage correct in single words and is scored based on production of voicing, manner and place. Totals represent an average of each section.

Table 4

Oral and Written Language Scales 2 (OWLS 2)

Subject Number	Last Interval Tested	Listening Comprehension Standard Score	Oral Expression Standard Score	Oral Composite Standard Score
S1	3	77	48	60
S2	3	46	50	45
S3	2.5	40	40	40
S4	2.0	40	40	40
S5	2.0	62	58	58

This test assesses receptive (understanding of) and expressive (use of) language for children and young adults aged 3 through 21 years. Standard scores are based on a mean of 100 and standard deviation of 15.