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Early Communication Development of Children with Auditory Brainstem Implants

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Abstract

The auditory brainstem implant (ABI) is an auditory sensory device that is surgically placed on the cochlear nucleus of the brainstem for individuals who are deaf but unable to benefit from a cochlear implant (CI) due to anatomical abnormalities of the cochlea and/or eighth nerve, specific disease processes, or temporal bone fractures. In the United States, the Food and Drug Administration has authorized a Phase I clinical trial to determine safety and feasibility of the ABI in up to 10 eligible young children who are deaf and either derived no benefit from the CI or were anatomically unable to receive a CI. In this paper, we describe the study protocol and the children who have enrolled in the study thus far. In addition, we report the scores on speech perception, speech production, and language (spoken and signed) for five children with 1–3 years of assessment post-ABI activation. To date, the results indicate that spoken communication skills are slow to develop and that visual communication remains essential for post-ABI intervention.

Implantable auditory technology has become the standard of care for children born with severe to profound sensorineural hearing loss. Today, multichannel cochlear implants (CI) provide access to speech, enabling many recipients to become proficient in spoken communication (Geers, Mitchell, Warner-Czyz, Wang, & Eisenberg, 2017; Niparko et al., 2010). Despite these findings, large variability in outcomes is characteristic of this population. Not all children achieve such high levels of success for a variety of reasons, including later age at implantation, limited pre-CI residual hearing, and low socioeconomic status (Niparko et al., 2010). The most likely impediments to success with the CI are associated with developmental delays and/or additional disabilities (Barnard et al., 2015; Cruz et al., 2012; Johnson, Wiley, & Meinzen-Derr, 2016), as well as anatomical malformation of the inner ear and neural structures (Birman, Powell, Gibson, & Elliott, 2016; Buchman et al., 2011; Young, Kim, Ryan, Tournis, & Yaras, 2012). Specific to those children identified with abnormal anatomical structures, the auditory brainstem implant (ABI) has become a relatively recent prosthetic option. The ABI is advocated for deaf individuals who are unable to benefit from a CI due to such conditions as neurofibromatosis type 2 (NF2), cochlear nerve deficiency or aplasia, temporal bone fracture, and severely malformed or ossified cochleae (Birman et al., 2016; Buchman et al., 2011; Colletti, Shannon, & Colletti, 2014; Noij et al., 2015; Shannon, 2015).

The external components of the ABI are essentially the same as those for the CI, but the internal components differ both in design and in surgical placement. Instead of insertion through the scala tympani of the cochlea similar to a CI electrode array, an ABI electrode paddle is placed directly on the cochlear nucleus of the brainstem. Thus, ABI surgery is substantially more invasive and complex than CI surgery (Aaron, Kari, Friedman, & Niparko, 2016; Wilkinson et al., 2017). Furthermore, electrical stimulation of the cochlear nucleus may produce non-auditory side effects, minor and major, due to its close proximity to other anatomical structures within the brainstem. As

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should be evident, ABI surgery and device programming are of higher risk to the child than the CI and requires a highly experienced multidisciplinary team.

Although the use of ABIs in young children is of recent origin in the United States, the first ABI surgery occurred in 1979 when William House and William Hitselberger implanted an electrode on the cochlear nucleus of an adult woman with NF2 during surgical removal of an acoustic tumor (Edgerton, House, & Hitselberger, 1982). Removal of these types of tumors (vestibular schwannomas) often requires severing the auditory nerve, precluding use of a CI. Following clinical trials of the ABI in patients with NF2, the U.S. Food and Drug Administration (FDA) approved the device in 2000 for use in individuals with NF2 (Shannon, 2015).

Around the time that the FDA was approving the ABI in the United States, Professor Vittorio Colletti and his team in Verona, Italy began exploring the ABI in non-NF2 adults and children (Colletti et al., 2002, 2005). Results in large samples of children are shown to vary. Whereas children with acquired deafness have shown the most promising results with the ABI, children born deaf who have significant additional disabilities or developmental delays have shown little to no progress with this device (Colletti et al., 2014; Sennaroglu et al., 2016). It has been estimated that 2.1% of pediatric CI candidates meet eligibility for an ABI (Kaplan et al., 2015), which in most cases is determined by radiographic imaging during the pre-CI evaluation.

Commencing in 2012, several FDA-sponsored Phase I trials were initiated in the United States to investigate the safety and feasibility of the Nucleus ABI in young non-NF2 children (Fisher et al., 2015; Puram et al., 2016; Teagle, Henderson, He, Ewend, & Buchman, 2018; Wilkinson et al., 2017). We recently described one such trial and presented 1-year results for four children and 3-month results for one child with the ABI (Wilkinson et al., 2017). To summarize the early findings, surgery and programming of the ABI processor were relatively safe with no occurrence of unexpected adverse events. However, progress in auditory skill development was incremental, with most children demonstrating sound detection thresholds of 40–50 dB HL and emerging auditory pattern perception skills (differentiation between words that differ by number of syllables).

The current report extends the Wilkinson et al. (2017) findings for these same five study participants, with results now spanning 1–3 years of follow-up assessments. More children have enrolled in the study than actually proceeded to surgery with an ABI, thus, we also describe demographic data for all 10 of the children currently enrolled in the study. However, the main thrust of this paper is to examine early efficacy for the five children with at least 1 year of ABI use, emphasizing their functional use of sound with the ABI to assist in spoken communication development. Because visual language is a necessary requirement for enrollment into the Los Angeles trial, the extent to which these children can also develop functional use of sound to support spoken communication will be a compelling indicator of device effectiveness.

Methods

Study Enrollment

This is an IRB-approved study with oversight from the FDA. At the time of this writing, the Phase I trial remains open for enrollment to children ages 2–5 years born with bilateral profound sensorineural hearing loss who demonstrate only sound detection with a CI, or up to 6 years for children with acquired deafness (e.g., meningitis with ossification or temporal bone fracture). The provisions of the study set a cap of 10 surgeries. Many children who are surgical candidates for ABI present with developmentally and/or medically challenging conditions. Cognitive and/or developmental delays may interfere with the child's ability to cooperate in testing and programming of the device. In addition, if children are medically fragile, it may place them at higher surgical risk. Accordingly, we restricted enrollment to "typically" developing children with cochlear nerve deficiency, aplasia, cochlear malformations, or acquired deafness due to meningitis or temporal bone fracture. In reference to the Colletti et al. (2014) findings, eligible children who are typically developing have the potential to achieve moderate to high levels of auditory skill development with the ABI. Full inclusion and exclusion criteria for this trial will be found in Wilkinson et al. (2017).

Upon receiving a referral or study inquiry, the research coordinator speaks with prospective participants about the study criteria. When there is clear evidence of exclusionary criteria (e.g., significant cognitive delay, children above 6 years of age, evidence of significant detection benefit with the CI), the family is informed that their child does not meet the specified criteria. If the family voices continued interest in an ABI, the research coordinator refers the family to other possible sources or study sites for information. When there is no obvious evidence of exclusionary obstacles, the research coordinator requests preliminary medical, radiological images, audiological, and educational records for review. If the record review indicates no issues of major concern, the family is invited to enroll in the study to learn more and to have their child evaluated.

As part of the formal candidacy process, parents are consented into the study and their child participates in a multiday evaluation consisting of medical examination and comprehensive assessment of hearing, speech, and language development. In addition, the educational specialist on the team reviews the child's intervention/educational services and a clinical psychologist evaluates the child's adaptive behavior and nonverbal cognitive skills. One of the many requirements for inclusion into the study is a 3- to 6-month trial with a CI prior to ABI surgery, whenever medically safe and possible to do so. This requirement is important because there is evidence of children with apparent cochlear nerve deficiency demonstrating some level of auditory skill with the CI (Birman et al., 2016; Young, Kim, Ryan, Tournis, & Yaras, 2012).

Children who qualify for and undergo ABI surgery participate in follow-up testing at regular intervals for a 3-year period. Figure 1 provides an overview of the testing timeline and protocols for each of the appointments the children attend during their enrollment in the study. For children who do not proceed to an ABI, all follow-up care and services revert to their local providers. Annual post-ABI evaluations for the implanted children consist of device programming, speech perception, speech production, language, early emergent literacy, and adaptive functioning assessments. The educational specialist also meets with the children and their parent(s) and reviews each child's educational service plan annually for 3 years.

Participants

Ten children have enrolled in the study to date, of whom six have proceeded to ABI. The youngest child enrolled was 2 years, 3 months of age and the oldest, 4 years, 11 months. All 10 participants are bilaterally, congenitally deaf as a result of structural

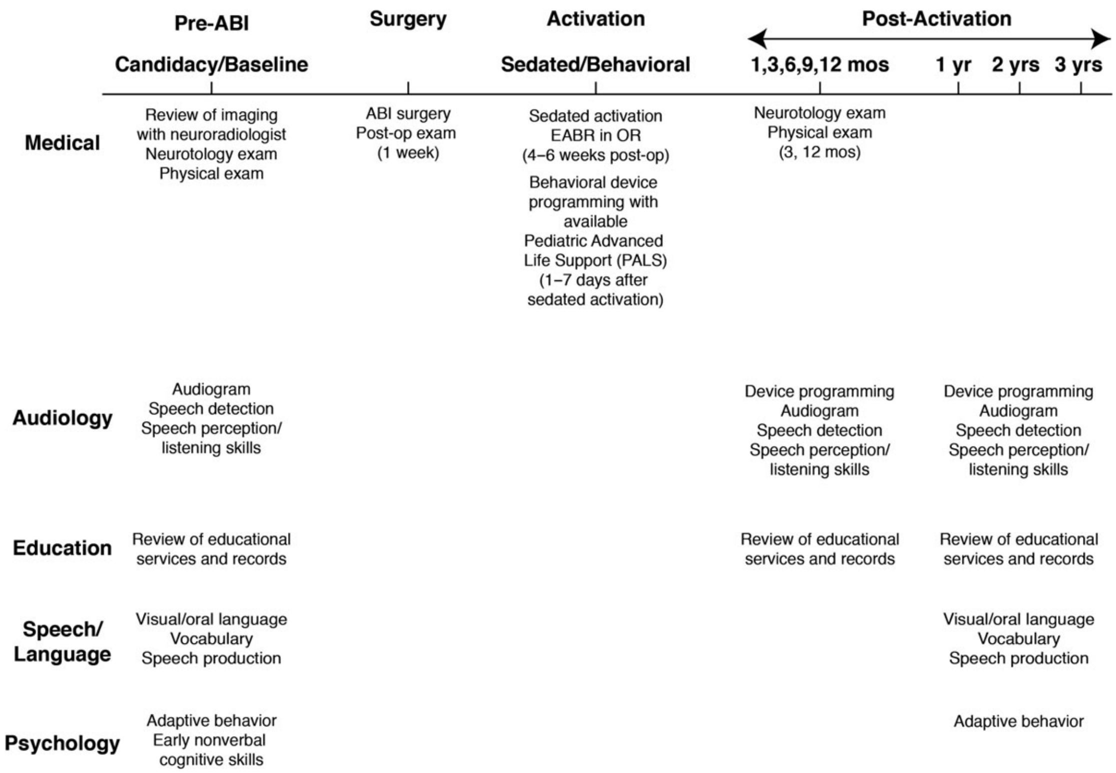

Figure 1 Protocol for the Los Angeles pediatric auditory brainstem implant (ABI) clinical trial.

Table 1 Demographic information for all 10 participants enrolled in the pediatric auditory brainstem (ABI) clinical trial, of whom 6 underwent ABI surgery

Case	Gender	Diagnosis	ABI surgery	Age at ABI surgery	Previous CI	Age at CI surgery	ABI side
1	M	Cochlear nerve deficiency	Yes	3 y	Bilateral	1y9m	Right
2	F	Cochlear nerve deficiency	Yes	3 y	Unilateral	2y3m	Right
3	F	Cochlear nerve deficiency	Yes	2y3m	Unilateral	1 y	Left
4	F	Cochlear aplasia	No	*3y5m	No	N/A	N/A
5	M	Cochlear aplasia	Yes	4y11m	No	N/A	Right
6	M	Cochlear nerve deficiency	No	*2y11m	Bilateral	11m/2y2m	N/A
7	F	Cochlear nerve deficiency	No	*2y4m	Unilateral	1y7m	N/A
8	M	Cochlear nerve deficiency	Yes	4 y	Unilateral	1y4m	Right
9	M	Cochlear nerve deficiency/cochlear malformation	No	*2y10m	No	N/A	N/A
10	M	Cochlear nerve deficiency	Yes	3y8m	Unilateral	1y9m	Right

CI = cochlear implant; M = male; F = female; y = years; m = months; * = age at study enrollment if no ABI; N/A = not applicable.

abnormalities unrelated to NF2. Anatomically, study inclusion criteria include cochlear nerve deficiency or aplasia, cochlear aplasia, or cochlear ossification. Families must demonstrate reasonable expectations and understand that visual communication will remain a necessity with an ABI. Children must not demonstrate cognitive or developmental delays or have other medical contraindications.

Table 1 presents the demographics of the 10 participants who thus far have been consented and evaluated under the study protocol. The table indicates the six participants who received an ABI. All participants used a visual means of communication (e.g., American Sign Language or ASL, manually coded English, or cued speech) upon enrollment. Seven of the 10 participants had received CIs (two bilateral, five unilateral). Of these seven, only one child (Case 7) consistently used and demonstrated measurable responses to sounds. For the other three participants, CIs were contraindicated.

Reasons for exclusion varied among the four participants who did not receive an ABI, but generally were due to medical conditions or developmental circumstances. Of note, Case 7, the child with measurable responses using a CI, did not qualify for an ABI based on demonstrated skills and progress with the CI. Despite our intent to enroll typically developing candidates, several of the six ABI recipients exhibited vestibular and/or attentional deficits, impulsivity, and distractibility.

ABI Device Programming

Pediatric audiologists on the ABI team perform the initial device activation approximately 4 weeks after surgery. The first activation occurs in an outpatient setting, while the child is under light sedation with cardiopulmonary monitoring. Thereafter, all programming happens in a clinical setting. During the activation and early programming sessions, the audiologists carefully observe the children for nonauditory side effects that may occur, such as coughing, gagging, eye-rubbing or unsteadiness. Subsequent programming is done at 1-, 3-, 6-, 9-, and 12-months post initial-ABI stimulation, then annually thereafter unless needs arise sooner.

When programming the ABI, techniques used are similar to those for children with CIs but with notable differences. Unlike a CI, where stimulation levels on adjacent channels can be used to interpolate levels of nearby electrodes, children with ABIs must provide detection thresholds for each electrode. Electrodes that elicit nonauditory responses are deactivated. As with standard techniques, children respond using visually reinforced audiometry or conditioned play as they are able.

The audiologists verify the children's hearing thresholds with the ABI at each programming session using audiometric soundfield testing. Sound detection thresholds are measured at 0.25, 0.5, 1, 2, and 4 kHz using warble-tone or narrow-band stimuli, with speech presented by monitored live voice. The children's thresholds currently range from 30 to 40 dB HL, with speech detection/awareness thresholds at about 25 dB HL. In addition, the audiologists interview the parent using the Meaningful Auditory Integration Scale (MAIS) (Robbins, Renshaw, & Berry, 1991), or for younger children the Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS) (Zimmerman-Phillips, Robbins, & Osberger, 2000). Parents answer questions about their child's daily use of the ABI, his or her responses to sounds, and the development of vocal behaviors. Soundfield detection thresholds with the ABI activated and results from the parent interviews are not presented in this report; the 1-year results from these measures can be found in Wilkinson et al. (2017).

Assessment Materials

Whereas overriding goals of the candidacy evaluation include determining children's compatibility with the ABI study criteria and establishing a baseline of abilities, post-ABI an important goal is assessing progress. Presented in this report are ABI findings specific to speech perception, speech production, and language for the five children with 1–3 years of ABI use. The assessment tools are described below and in Table 2.

Speech perception

Speech perception testing was initiated at 3-to-6 months post-ABI activation, and at yearly follow-up intervals. The battery consists of closed- and open-set tests. The closed-set tests involve picture-pointing tasks that incorporate measures of pattern perception (i.e., differentiating words that differ in number of syllables), word identification, and sentence identification. The specific measures include the Early Speech Perception test (ESP), the Test of Auditory Comprehension (TAC), and the Pediatric Speech Intelligibility test (PSI). The one open-set measure, Lexical Neighborhood Test (LNT), consists of lexically easy and hard monosyllabic words that require the child to repeat and/or sign what she or he hears. Table 2 provides descriptions of these four tests. Participants progress on an individual basis from easy to more difficult tests in a hierarchical fashion to determine skill development over the 3-year follow-up protocol.

Speech production

Speech development was assessed using observation of the child interacting with the parent, and by use of the Identifying Early Phonological Needs in Children with Hearing Impairment test (IEPN), a word-level speech production measure. The IEPN was selected as the primary speech measure because it is designed specifically for children with hearing loss who are low verbal and still developing speech. For the test, children view a picture of an object and name or imitate the name of the object. Testing generally was initiated at 1- or 2-years post, once the child was able to provide the necessary spontaneous or imitative verbal response. Table 2 describes the feature-based scoring of the IEPN.

Language

All study participants undergo a language evaluation as part of the candidacy evaluation. In addition, children who receive the ABI undergo a communication skills assessment annually, near their 1-, 2-, and 3-year postactivation dates. The tests administered during the candidacy or annual evaluations vary based on the children's overall abilities and age at each test point. The focus of testing post-ABI is to assess change in English language development, orally and when supported visually using manually coded English. Change is measured not only based on standardized clinical measures but also with respect to functional improvements—gains noticed by parents on a more daily basis. For this reason, evaluative measures are comprised of direct clinician-administered assessments, parent-report based questionnaires or interviews, videotaped parent-child interactions (e.g., during shared book reading), observation, and review of educational records or school-based assessments (e.g., completed by the child's teacher or speech-language pathologist).

Table 2 describes the two clinician-administered language measures for which results are reported (Preschool Language Scales, 5th ed. or PLS-5, and the Peabody Picture Vocabulary Test, 4th ed. or PPVT-4). Raw scores were obtained for spoken and visual language (Simultaneous Communication) separately. Raw scores, as opposed to standard scores, are reported so that direct comparison can be made using spoken and visual language. Standard scores are based on spoken language norms and not visual, so they are avoided here. We recognize that the scores may be slightly inflated for signed versus spoken administration because of the iconic nature of some signs.

Prior to testing with the standardized language measures, children were observed with their parent while reading a book or playing with toys. Based on this observational data and information from baseline or previous annual testing, starting points for the standardized measures were determined. Because most participants were at the prelinguistic or early linguistic stage of development for spoken language, but at the linguistic level (i.e., word combinations, sentences, or conversational language) with visual language, there was usually clear separation in the starting points (basal) for spoken language and sign-based skills. When this was the case, testing began using spoken language, beginning with item number one of the standardized measure. Testing continued with spoken language until a ceiling was established. A basal was then established using Simultaneous Communication (spoken language paired with manually coded English), progressing until a ceiling was reached to obtain a visual language score. When there was overlap between a child's spoken language skills and sign skills, individual test items were first presented verbally and then presented using Simultaneous Communication until ceilings were established in each modality.

Results

Thus far, for every three children enrolled and evaluated in the study, two children have actually gone on to receive the ABI.

Table 2 Test measures for speech perception, speech production, and language

Measure	Description	Features
Speech perception		
Early Speech Perception Test (**ESP**; Moog & Geers, 1990).	Designed to assess pattern perception, spondaic and monosyllabic word identification. Performance is classified into one of four categories representing increasing levels of auditory skill. Low verbal and standard scoring versions	**Age:** 2–3 years and older **Administration:** Live-voice **Response format**: Closed-set picture pointing
Test of Auditory Comprehension (**TAC**; Trammell et al., 1981)	10 subtests assessing a continuum of auditory tasks from perception of environmental and speech stimuli to comprehension of simple and complex stories presented in quiet and competing speech noise	**Age:** 4–12 years **Administration:** Recording **Response format:** Closed-set picture pointing
Pediatric Speech Intelligibility Test (**PSI**; Jerger, Lewis, Hawkins & Jerger, 1980).	Assesses child's ability to identify words and sentences depicted on a picture plate from a closed set of five alternatives. Presented in quiet and, if child meets criteria, in a range of message-to-competition ratios	**Age**: 3 years and older **Administration**: Recording **Response format**: Closed-set picture pointing
Lexical Neighborhood Test (**LNT**; Kirk, Pisoni, & Osberger, 1995).	Assesses open-set spoken word recognition for children with hearing loss. Comprised of two 50-item monosyllabic word lists; 25 lexically "easy" and 25 lexically "hard" words	**Age**: 4 years and older **Administration**: Recording **Response Format**: Open-set word repetition
Speech production and language		
Identifying Early Phonological Needs in Children with Hearing Impairment (**IEPN**; Paden & Brown, 1992).	Word-level speech measure designed for children with hearing loss. Children spontaneously or imitatively name an object or picture they are shown. Scores indicate the overall correctness of phonemes and how well children produce targeted phoneme features. It rates accuracy of word patterns (e.g., stress and syllable number), basic vowel areas (e.g., tongue height and place), and consonant features (e.g., place, manner, voicing). Broadly summarized, phoneme productions can earn from 0 points (if the designated feature is not produced), up to 3 points (for correct pronunciations). Scores show percent accuracy. Normative scores are not provided	**Age**: Age independent **Administration**: Clinician-elicited responses using toy props or pictures **Response format**: Spontaneous or imitative speech
Preschool Language Scales 5th ed. (**PLS-5**; Zimmerman, Steiner & Pond, 2011).	Evaluates language using toys, pictures, and conversation. It has two subscales: auditory comprehension and expressive communication (expressed as composite raw scores for this study). It assesses precursors for language, progressing to understanding and use of grammatical markers, emergent literacy skills, theory of mind, and other high-level communication skills.	**Age:** Birth—7 years **Administration:** Clinician-elicited responses using play and direct questioning **Response format:** Pointing, selecting items, and answering questions
Peabody Picture Vocabulary Test 4th ed. (**PPVT-4**; Dunn & Dunn, 2007).	Assesses understanding of words using a four-choice picture-pointing task.	**Age:** 2.5 years–90+ years **Administration:** Clinician-elicited **Response format:** Closed-set picture pointing

Results reported here are from five children (out of the total of six) who received an ABI and who have completed at a minimum the 1-year post-ABI protocol. Four of the five children (Cases 1, 2, 3, and 5) have completed the 3-year post-ABI protocol and one child (Case 8) completed the 1-year post-test interval. Data are not included for the sixth child (Case 10), who has less than 1 year of ABI use.

Two of the five children (Cases 2 and 8) entered the trial using manually coded English and are enrolled in educational programs that use Simultaneous Communication (also referred to as total communication or TC). Two children (Cases 3 and 5) came into the study using ASL. After receiving the ABI, when transitioning from birth-to-three services to school-age services, one of these two children (Case 3), enrolled in an educational program that uses Simultaneous Communication. Case 5 transitioned from an ASL program to a Simultaneous Communication program just prior to the 3-year evaluation. Case 1 entered the study using Quebec Sign Language (LSQ) the French equivalent of ASL, and continues to be educated using LSQ.

Because this study was not designed or powered for inferential statistics, individual test scores are presented in bar graphs at each test interval. Missing data on graphs indicate that not all children could be assessed on all measures at each test interval due to age or skill-level constraints.

Speech Perception

Closed-set tests (ESP, TAC, and PSI)

Individual scores for the ESP and TAC are presented in Figures 2 and 3, with no graphic display for the PSI. Recall that the tests are hierarchical, with specific age and skill-level indications for advancing to more challenging tasks. If the test was not administered because the child was either too young or the task too difficult, data would not be displayed for that follow-up test interval.

The ESP is administered using live voice. Post-ABI activation, the children demonstrated varying levels of pattern perception (one versus two versus three syllables) on this closed-set measure. At 1-year post, all children were at level 1; they had not reached level 2 pattern perception. Two of the five children achieved mastery on pattern perception at 2- to 3-years post-ABI activation.

On the TAC, using recorded stimuli, Case 2 passed subtest 2 (linguistic/human versus nonlinguistic/environmental) at 2-years post-ABI. At 3-years post, this same child reached subtest 4 (core noun vocabulary). A second child (Case 5) passed subtest 2 at 3-years post-ABI.

Case 2 progressed to the PSI for assessing closed-set sentence identification. The child was able to identify sentences with 100% accuracy by 2-years post-ABI activation with the stimuli presented live voice, auditory-only. By 3-years post, Case 2 attained a score of 80% for recorded stimuli.

Open-set test (LNT)

By 3 years post, Case 2 was the only child to progress to open-set word recognition testing using live-voice testing. For the lexically easy and hard words, auditory-only scores were 20% and 36%, respectively. Phoneme scores, as expected, were higher

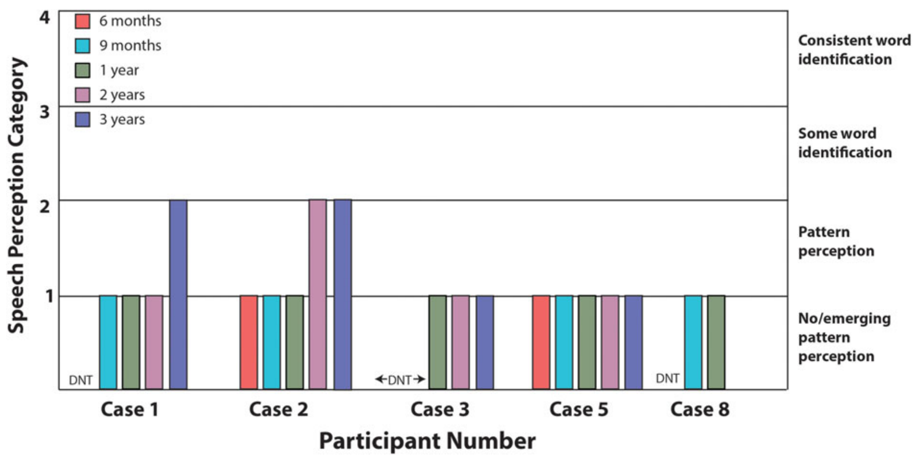

Figure 2 Individual category scores reached on the Early Speech Perception (ESP) Test for Cases 1, 2, 3, 5, and 8. DNT = did not test.

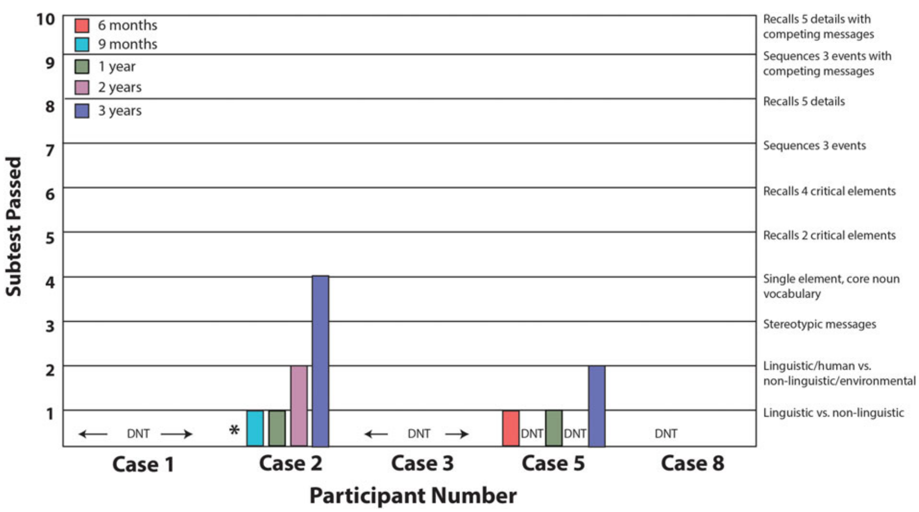

Figure 3 Individual subtest levels reached on the Test of Auditory Comprehension (TAC) for Cases 2 and 5. DNT = did not test; * = tested but did not meet criterion to pass Subtest 1.

than the word scores (44% for easy and 60% for hard). The higher word and phoneme scores for hard relative to easy stimuli may be explained by practice effects (easy words were presented first).

To summarize the speech perception findings, only one child (Case 2) demonstrated successive gains in auditory skill development with use of the ABI. This child is the only one thus far to demonstrate progression toward open-set speech recognition.

Speech Production (IEPN)

The composite scores for the IEPN post-ABI can be viewed in Figure 4 for the four participants who are exposed to English in at least some form—oral or visual. Composite percent-correct scores for production of word pattern, vowel area, and consonant features in imitated words are displayed for each child at intervals when testing could be accomplished. Reasons for not testing included time constraints (Case 2, at year 1), a child not having the necessary verbal response (Case 3, at year 1), or the test interval not being reached (Case 8).

Word patterns

All children had started producing basic word patterns by 1- or 2-years post-ABI. By the 2-year post point, all four children matched syllable number for one versus two syllable words with 40–100% accuracy. In addition, even if not the correct consonant target children had started including a word-initial consonant sound when needed 67–96% of the time. Inclusion of word-final consonants varied greatly (from 6% to 94%) at 1- or 2-years post. Use of intonation or proper stress has been slower to develop, with children obtaining scores of 0–30% correct in this regard irrespective of length post-ABI.

Basic vowel areas

Three children (Cases 2, 5, and 8) started producing vowel features, beyond neutral vowels (e.g., /ə/) by 1- or 2-years post-ABI. The fourth child (Case 3) reached this point at year 3. Within-child accuracy tended to range greatly across vowel features. For example, after 3 years of ABI use, Case 2 demonstrated vowel feature accuracy ranging from 47% (for high-front vowels) to 93% (for low-back vowels). Also at 3-years post, Case 3 demonstrated scores of 0% accuracy (for mid-low-back vowels and diphthongs) up to 47% (for high-back vowel sounds).

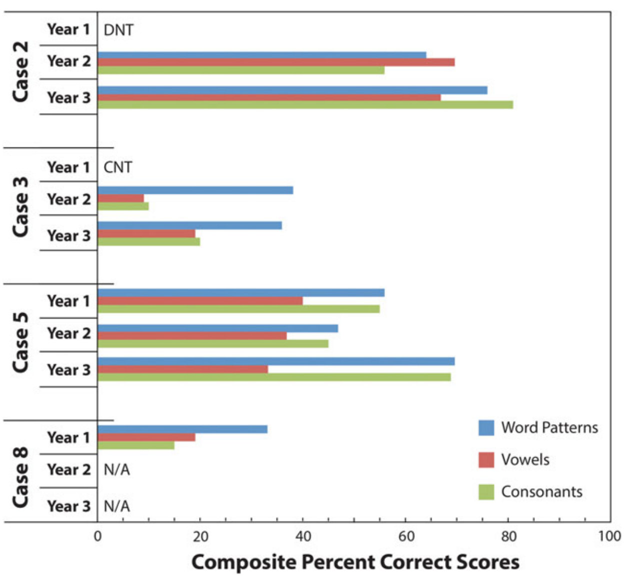

Figure 4 Individual percent composite scores for word patterns, vowels, and consonants on the Identifying Early Phonological Needs in Children with Hearing Impairment (IEPN) for Cases 2, 3, 5, and 8. DNT = did not test; CNT= could not test; N/A = not applicable.

Basic consonant features (manner, place, voicing)

By 2-years post-ABI activation, two children (Cases 2 and 5) demonstrated development across all consonant manner features (stop, nasal, fricative and liquid). With 1 or 2 years of ABI use, the other two children (Cases 3 and 8) were producing the nasal and liquid manner of production, but not stops or frication. One of these latter two children (Case 3) subsequently demonstrated use of frication at the 3-year post-ABI test interval. For consonant place, whereas all children were producing at least some front (e.g., "b, m, w") and mid (e.g., "t, d, n, l") sounds, only one of the four children (Case 2), demonstrated use of back sounds (e.g., "k, g, ng"). This child scored 19% correct for back sounds at the 2-year post evaluation and 90% accuracy at 3-years post-ABI. With respect to voicing control, overall children demonstrated greater accuracy for voiced versus unvoiced sounds. Irrespective of time post-ABI, children accurately imitated voicing for voiced consonants 60–90% of the time. Accurate production of devoicing for the unvoiced feature ranged from 0% (for Case 8 at 1-year post) to 100% of the time (for Case 2 at 3-years post).

Of the four children, only one child (Case 2) has demonstrated systematic improvements in speech production over time. At 3-years post-ABI, this child is beginning to produce words, common phrases, and simple sentences that others understand with context or if familiar with the child.

One child (Case 5) demonstrated a slight decrement in speech production skills at 2-years post across all features (word patterns, vowels, and consonants). At the 2-year post-ABI evaluation, this child's parent reported a reduction in ABI use. The child was removing the ABI at the end of every school day, reporting sounds were too loud. The child demonstrated more sound omissions when imitating words at 2-years post than at 1-year post, resulting in lower scores for the IEPN. At the 3-year post evaluation, the child had resumed full-time device use and reported no loudness issues. IEPN scores improved, with overall scores at 3-years post surpassing 1-year post scores.

Language

PLS-5

Figure 5 displays PLS-5 composite raw scores for spoken language (Auditory Oral) and visual language (TC/Simultaneous Communication) for Cases 2, 3, 5, and 8. Whereas pre-ABI scores for the other measures are not discussed, they are presented here to demonstrate the baseline language skills of these children at entry into the study. In addition, they serve to compare scores with a nonqualifying ABI candidate with 9 months of CI use. It will be seen on Figure 5 that spoken language scores were not always obtained at baseline. At that point, children lacked oral language experience. Use of voice was limited and parents reported no use of spoken language or responses to sound. When administered pre-ABI, children typically scored at the 6-month level or below on this measure (Cases 2 and 8). For comparison to a nonqualifying ABI candidate, Figure 5 also displays the pre-ABI language results for Case 7 (far right on the figure). This case presented for ABI candidacy evaluation, but did not qualify based on demonstrated benefit using a CI. Having had 9 months of CI use, this child was showing substantial skills with

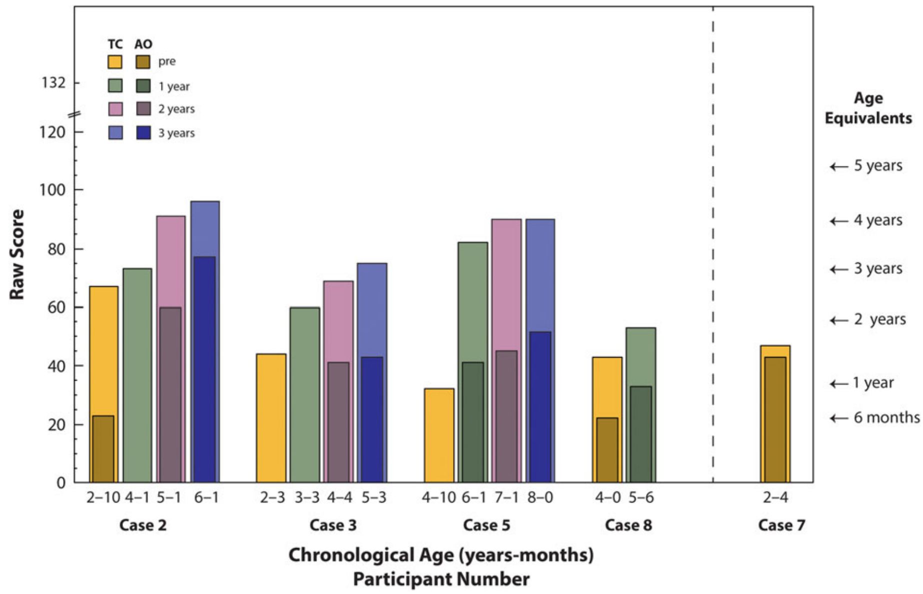

Figure 5 Composite (Auditory Comprehension and Expressive Communication) raw scores (left vertical axis) and age equivalents (right vertical axis) are displayed for the Preschool Language Scales (PLS-5) for Cases 2, 3, 5, and 8. As a comparison, the bars to the far right display the scores for Case 7, a nonqualifying auditory brainstem implant (ABI) candidate using a cochlear implant (CI). The outer bars represent scores for total communication administration. The inner shaded bars represent scores for verbal administration. TC = total communication; AO = auditory oral.

a minimal gap between demonstrated oral and visual language skills.

Post-ABI, the children generally demonstrated incremental progress at each post-test interval for oral and manual modalities. For manual administration, the increasing height in the outer bars showed that visual language was improving over time. For oral administration, the inner shaded bars indicate that at 1-year post, oral skills were typically still very limited. For those children who could be tested in the oral modality, skills were generally near the 1-year level. This is consistent with prelinguistic use of gestures, but also emerging attention to name or other environmental sounds.

At 2-years post, one child (Case 2) surpassed the 2-year level on the PLS-5 when assessed in the auditory oral modality. To reach this level of mastery, a child must be able to name and/or point to objects. It is notable that Case 2 was the only child who substantially exceeded Case 7's oral language score with the CI. At 3-years post, Case 2 continued to show a trajectory of growth for spoken language, demonstrating year-to-year gains. Visual communication skills improved but not to the extent of oral skills, narrowing the gap between the two modalities for this child. Also at 3-years post, Case 5 was starting to use speech reading information for highly contextualized communication.

PPVT-4

Scores for the PPVT-4 are shown in Figure 6 for Cases 2, 3, 5, and 8. As can be seen, when the test was presented with sign, age-equivalent scores for Cases 2, 3 and 5 approximated what might be expected based on the children's chronological age. However, for Case 5, scores were seen to drop at 2-years post and were still lagging behind age expectations at 3-years post. Verbal administration yielded very low age-equivalent scores for three of the four cases. One child (Case 5) showed some growth at 3-years post, but overall spoken vocabulary was still limited. Case 2 is the only child to demonstrate an age-equivalent of 2 years, 6 months at 2-years post and 3 years, 7 months at 3-years post (i.e., comparable to time with the ABI, but well below chronological age). Similar to the PLS-5, this is an encouraging outcome because it suggests that this child is showing year-by-year growth in receptive vocabulary when the stimuli are presented verbally.

Discussion

Gains in communication skill development for five pediatric ABI recipients have remained incremental and slow to generalize even for those children with 3 years of experience with the device. With respect to speech perception outcomes, the children displayed varying levels of pattern perception and closed-set word/sentence identification, with one child progressing to open-set word recognition. Small improvements were also shown for speech production and spoken language for three children, with moderate gains for a fourth child. Direct speech and language testing was not possible for Case 1 (a non-English user). However, based on observation, parent-report and review of the child's educational progress, outcomes appeared to resemble those for the majority of participants. The child's oral skills were developing, but slowly. Sign skills (LSQ) were advancing steadily.

These relatively early findings underscore the limitations of the ABI for children born with profound hearing loss who are unable to benefit from a CI. Similar findings have been reported by two other pediatric ABI centers conducting clinical trials in the United States (Puram et al., 2016; Teagle et al., 2018), although results from large samples of children reported from outside of the United States appear to be promising (Colletti et al., 2014; Sennaroglu et al., 2016). It is therefore encouraging that one child in our series (Case 2) has shown greater gains in the acquisition of speech perception and spoken communication skills when compared to the four other children. This child is beginning to communicate with family and peers using spoken language.

The other children have demonstrated varying degrees of progress in their speech production or language skills whether

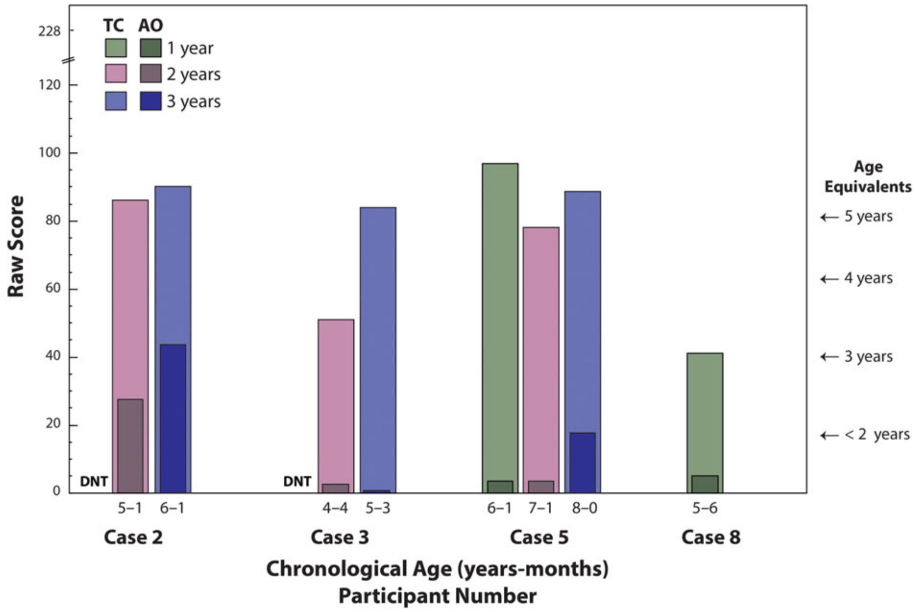

Figure 6 Individual raw scores (left vertical axis) and age equivalents in years (right vertical axis) are displayed for the Peabody Picture Vocabulary Test (PPVT-4) for Cases 2, 3, 5, and 8. The outer bars represent scores for total communication administration. The inner shaded bars represent scores for verbal administration. TC = total communication; AO = auditory oral; DNT = did not test.

tests were administered verbally or with manually coded English. In all cases, reliance on speech reading has been essential. In spite of varying degrees of progress in auditory and oral skill development, on the whole these children demonstrate steady visual language gains, some with skills approximating age-level expectations. These are encouraging results, but the fact remains that children with ABIs demonstrate slower progress than children with CIs in the early acquisition of spoken language (Niparko et al., 2010), and will likely remain reliant on visual communication (i.e., ASL, manually coded English, and cued speech). A small percentage may develop oral language skills with reliance on speech reading.

Predicting which combination of factors will promote a faster rate of progress with the ABI is not straightforward, although etiological factors may be predictive. From a longitudinal study tracking 64 children with ABIs, Colletti et al. (2014) reported that the highest categories of auditory skill, such as open-set speech recognition, were reached by those children with acquired deafness (i.e., ossified cochleae following meningitis or temporal bone fracture). In contrast, those children born deaf with additional complex needs generally did not progress beyond sound detection. The children in the middle range of the sample were those born deaf but who did not exhibit additional disabilities or developmental delays (i.e., typically developing). This group generally achieved closed-set speech identification with eventual recognition of common phrases. The children enrolled in our trial fit the description of those falling in the middle range.

Many of the ABI candidates demonstrated vestibular deficits as evidenced by late walking or need for occupational or physical therapy during infancy. Children who are born without an eighth nerve or one diminished in size likely have no or reduced vestibular input for the typical development of balance and postural control, along with other early motor behaviors (Beraneck, Lambert, & Sadeghi, 2014). Motor and balance deficits can hinder early language growth (Iverson, 2010). Also noted, some children demonstrated attentional deficits, impulsivity or distractibility. There is evidence that lack of vestibular input may generalize to a reduced ability to sustain attention and self-regulate (Iverson, 2010; Karasik, Tamis-LeMonda, & Adolph, 2011). The exact impact of vestibular or motor deficits on auditory learning is unclear but such deficits could conceivably slow accommodation to electrical stimulation with the ABI.

In addition to etiological factors, a possible explanation for why children with ABIs may underperform in comparison to children with CIs is the relatively older ages at time of enrollment into the trial. The youngest age for inclusion in the Los Angeles trial was set at 2 years to minimize potential surgical risks. The youngest child enrolled in the trial was 2 years, 3 months of age, whereas the oldest child was just under 5 years of age. The FDA criterion for minimal implantable age for CI is 12 months, with many children undergoing CI surgery at even younger ages. Moreover, most children eligible for CIs are born with varying levels of residual hearing (primarily in the low frequencies) and are able to take advantage of limited auditory stimulation with hearing aids prior to CI surgery. Due to a complete absence of acoustic hearing and an inability to use a CI, ABI candidates experience true auditory deprivation and for relatively long periods. ABI recipients therefore may be at a disadvantage, lacking sound input during a sensitive period for auditory development (Sharma, Dorman, & Spahr, 2002).

Another reason why many children with ABIs may not achieve performance levels on par with children with CIs is the impoverished signal delivered by the device and the limited number of electrodes that may be safely activated. Audiologists are able to program the ABI and validate the device settings through soundfield detection thresholds (Teagle et al., 2018; Wilkinson et al., 2017). However, spectral and temporal resolving abilities with the ABI are unknown at this time.

In many ways, outcomes with the ABI are reminiscent of the early 1980s when the first-generation commercial CI (single-channel system) was undergoing pediatric FDA trials. In that trial, children had been implanted at older ages relative to current standards and generally achieved pattern perception and linguistic versus nonlinguistic identification (Thielemeir, Tonokawa, Petersen, & Eisenberg, 1985). Despite these rudimentary auditory skills, a small number of children with single-electrode CIs eventually went on to attain open-set word recognition (Berliner et al., 1989; Geers & Moog, 1988). We had earlier observed this same finding in two children with ABIs who were implanted in Europe prior to the initiation of FDA trials (Eisenberg, Johnson, Martinez, Visser-Dumont, Ganguly, & Still, 2012). Those two children received the ABI at relatively older ages (1 year, 11 months; 2 years, 11 months) by today's CI standards. Nevertheless, they were performing in the low average range on measures of speech perception after 2–3 years of ABI use when matched to a sample of CI participants implanted at similar ages. The fact that Case 2 in the current trial was demonstrating a similar level of skill is an encouraging indicator of efficacy.

Anecdotally we have noticed that although the highest performing children are making measurable gains in speech perception and spoken language, their articulation development is comparably delayed. Parents also report that their children's speech has been slow to develop and remains difficult to understand, particularly for unfamiliar listeners. Findings from the IEPN post-ABI are consistent with the clinical observation and parent-report of slow development of speech over time. This is different from findings that children with CIs show measurable gains across all features on the IEPN (word patterns, vowels, and consonants) within 6 months of device use (Brown & McDowall, 1999). It is also in contrast to findings that children with CI's often start to babble within 6 months of implantation (Fagan, 2015; Moeller, et al., 2007), and start becoming intelligible within 3-years of cochlear implantation (Blamey et al., 2001; Flipsen & Colvard, 2006).

An essential component to emerge from this study is the need for ongoing counseling with families and interventionists. A dilemma faced by the ABI team is how best to optimize auditory-based intervention with the ABI while minimizing possible detriments to visual language development. It is possible that "type" of visual input (i.e., ASL versus signed English/cued speech) plays an important role for successful intervention with the ABI. ASL is a different language than English, thus it does not support development of English structures. It is possible that visual access to manually coded English may more easily facilitate the transition from signed to spoken communication following ABI intervention. The one child in our study who demonstrates emergent spoken communication (Case 2) came into the study using manually coded English, specifically Signed Exact English, which marks English morphology. The child also interacted consistently with both parents using Signed Exact English. A form of visual modality that codes for English morphology (e.g., Signed Exact English, cued speech) could represent the most viable language option for potential ABI recipients seeking to learn English. Research by Giezen, Baker, & Escudero (2014) would support combining visual and spoken language to facilitate auditory learning. This notion is further supported by the fact that the other three children in the trial (Cases 3, 5, and 8) who communicate via Simultaneous Communication, but not Signed Exact English, are not progressing in their auditory and spoken language skills to the same extent as Case 2.

Two limitations to this study are noted. The first limitation is the restricted enrollment to a subset of potential ABI candidates that are "typically developing". Findings become commensurately limited when attempting to generalize to the broader population. Although restricted enrollment criteria can be viewed as a short falling, with a population and outcomes that already are shown to be quite variable, the intent was to focus on a group that may be more likely to demonstrate progress.

The second limitation is the uncontrolled factor of therapeutic and educational services or placements. Most families live too far from the ABI center to participate in weekly therapy or attend a uniform educational program, which would provide continuity in habilitative services for the study participants. Instead, therapy and educational services differ greatly across the participants. To mitigate this limitation, the ABI team provides guidance to the children's local providers. For most providers, having a child with an ABI is a new experience and they are seeking information and suggestions.

To conclude, the ABI may be a viable option for children born with malformed cochleae and/or deficient auditory nerves who show no demonstrable benefit from a CI. Despite greater surgical and programming risks in comparison to CIs, adverse effects with the ABI are minimal if carried out by experienced surgeons and pediatric audiologists. Team members from related disciplines in radiology, speech-language pathology, education of the deaf, and psychology are essential for conducting the multiple assessments required to determine candidacy and post-ABI follow-up. The ultimate benefits of this technology for the majority of eligible pediatric candidates are not entirely known at this stage. Indications from the literature of children acquiring some of the basic skills needed to support spoken communication point to the potential of the ABI. At the present time, because spoken communication outcomes cannot be predicted or routinely expected, the importance of maintaining a visual mode of communication remains a high priority for these children both pre- and post-ABI intervention. Moreover, many years of intervention should be expected.

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Conflict of interest

Eric P. Wilkinson receives research funding from Cochlear Corporation and MedEl Corporation. Consultant for Nurotron Biotechnology House Clinic and Huntington Medical Research Institutes.

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References

Aaron, K. A., Kari, E., Friedman, R. A., & Niparko, J. K. (2016). Cochlear implants and auditory brainstem implants for children: Surgical considerations. In Eisenberg L. S. (Ed.), *Clinical management of children with cochlear implants* (2nd Ed., pp. 69–103). San Diego, CA: Plural Publishing.

Barnard, J. M., Fisher, L. M., Johnson, K. C., Eisenberg, L. S., Wang, N.-Y., & Quittner, A. L.…the CDaCI Investigative Team (2015). A prospective, longitudinal study of US children unable to achieve open-set speech recognition five years after cochlear implantation. *Otology & Neurotology*, *36*, 985–992. doi:10.1097/MAO.0000000000000723

Beraneck, M., Lambert, F. M., & Sadeghi, S. G. (2014). Functional development of the vestibular system: Sensorimotor pathways for stabilization of gaze and posture. In Romand R., & Varela-Nieto I. (Eds.), *Development of auditory and vestibular systems* (4th Ed., pp. 449–487). Paris, France: Academic Press, Elsevier.

Berliner, K. I., Tonokawa, L. L., Dye, L. M., & House, W. F. (1989). Open-set speech recognition in children with a single-channel cochlear implant. *Ear and Hearing*, *10*, 237–242. doi:10.1097/00003446-198908000-00005

Birman, C. S., Powell, H. R. F., Gibson, W. P. R., & Elliott, E. J. (2016). Cochlear implant outcomes in cochlea nerve aplasia. *Otology & Neurotology*, *37*, 438–445. doi:10.1097/MAO.0000000000000997

Blamey, P. J., Sarant, J. Z., Paatsch, L. E., Barry, J. G., Bow, C. P., Wales, R. J.…Tooher, R. (2001). Relationships among speech perception, production, language, hearing loss, and age in children with impaired hearing. *Journal of Speech, Language, and Hearing Research*, *44*, 264–285. doi:10.1044/1092-4388(2001/022)

Brown, C., & McDowall, D. W. (1999). Speech production results in children implanted with the Clarion implant. *Annals of Otology, Rhinology, and Laryngology*, *108*(Suppl. 4), 110–112. doi:10.1177/000348949910805422

Buchman, C. A., Teagle, H. F., Roush, P. A., Park, L. R., Hatch, D., Woodard, J.…Adunka, O. F. (2011). Cochlear implantation in children with labyrinthine anomalies and cochlear nerve deficiency: Implications for auditory brainstem implantation. *Laryngoscope*, *121*, 1979–1988. doi:10.1002/lary.22032

Colletti, L., Shannon, R. V., & Colletti, V. (2014). The development of auditory perception in children after auditory brainstem implantation. *Audiology and Neurotology*, *19*, 386–394. doi:10.1159/000363684

Colletti, V., Carner, M., Fiorino, F., Sacchetto, L., Miorelli, V., Orsi, A.…Pacini, L. (2002). Hearing restoration with auditory brainstem implant in three children with cochlear nerve aplasia. *Otology & Neurotology*, *23*, 682–693. doi:10.1097/00129492-200209000-00014

Colletti, V., Carner, M., Miorelli, V., Guida, M., Colletti, L., & Fiorino, F. (2005). Auditory brainstem implant (ABI): New frontiers in adults and children. *Otolaryngology-Head & Neck Surgery*, *133*, 126–138. doi:10.1016/j.otohns.2005.03.022

Cruz, I., Vicaria, I., Wang, N.-Y., Niparko, J., & Quittner, A. L., the CDaCI Investigative Team (2012). Language and behavioral outcomes in children with developmental disabilities using cochlear implants. *Otology & Neurotology*, *33*, 751–760. doi:10.1097/MAO.0b013e3182595309

Dunn, L. M., & Dunn, D. M. (2007). *Peabody picture vocabulary test* (4th ed.). Minneapolis, MN: NCS Pearson, Inc.

Edgerton, B. J., House, W. F., & Hitselberger, W. E. (1982). Hearing by cochlear nucleus stimulation in humans. *Annals of Otology, Rhinology, and Laryngology*, *91*(Suppl. 91), 117–124.

Eisenberg, L. S., Johnson, K. C., Martinez, A. S., Visser-Dumont, L., Ganguly, D. H., & Still, J. F. (2012). Studies in pediatric hearing loss at the House Research Institute. *Journal of the American Academy of Audiology*, *23*, 412–421. doi:10.3766/jaaa.23.6.4

Fagan, M. K. (2015). Why repetition? Repetitive babbling, auditory feedback, and cochlear implantation. *Journal of Experimental Child Psychology*, *137*, 125–136. doi:10.1016/j.jecp.2015.04.005

Fisher, L. M., Eisenberg, L. S., Krieger, M., Wilkinson, E. P., & Shannon, R. V. (2015). Regulatory and funding strategies to develop a safety study of an auditory brainstem implant in young children who are deaf. *Therapeutic Innovation and Regulatory Science*, *49*, 659–665. doi:10.1177/2168479015599559

Flipsen, P., Jr, & Colvard, L. (2006). Intelligibility of conversational speech produced by children with cochlear implants. *Journal of Communication Disorders*, *39*, 93–108. doi:10.1016/j.jcomdis.2005.11.001

Geers, A. E., Mitchell, C. M., Warner-Czyz, A., Wang, N.-Y., & Eisenberg, L. S., the CDaCI Investigative Team (2017). Early sign language exposure and cochlear implantation benefits. *Pediatrics*, *140*, pii e20163489e. doi:10.1542/peds.2016-3489

Geers, A. E., & Moog, J. S. (1988). Predicting long-term benefits from single-channel cochlear implants in profoundly hearing-impaired children. *American Journal of Otology*, *9*, 169–176.

Giezen, M. R., Baker, A. E., & Escudero, P. (2014). Relationships between spoken word and sign processing in children with cochlear implants. *Journal of Deaf Studies and Deaf Education*, *19*, 107–125. doi:10.1093/deafed/ent040

Iverson, J. M. (2010). Developing language in a developing body: The relationship between motor development and language development. *Journal of Child Language*, *37*, 229–261. doi:10.1017/S0305000909990432

Jerger, S., Lewis, S., Hawkins, J., & Jerger, J. (1980). Pediatric speech intelligibility test. I. Generation of test materials. *International Journal of Pediatric Otorhinolaryngology*, *2*, 217–230. doi:10.1016/0165-5876(80)90047-6

Johnson, K. C., Wiley, S., & Meinzen-Derr, J. (2016). Cochlear implantation in children with additional disabilities. In Eisenberg L. S. (Ed.), *Clinical management of children with cochlear implants* (2nd Ed., (pp. 737–789). San Diego, CA: Plural Publishing.

Kaplan, A. B., Kozin, E. D., Puram, S. V., Owoc, M. S., Shah, P. V., Hight, A. E.…Lee, D. J. (2015). Auditory brainstem implant candidacy in the United States in children 0-17 years old. *International Journal of Pediatric Otolaryngology*, *79*, 310–315. doi:10.1016/j.ijporl.2014.11.023

Karasik, L. B., Tamis-LeMonda, C. S., & Adolph, K. E. (2011). Transition from crawling to walking and infants' actions with objects and people. *Child Development*, *82*, 1199–1209. doi:10.1111/j.1467-8624.2011.01595.x

Kirk, K. I., Pisoni, D. B., & Osberger, M. J. (1995). Lexical effects on spoken word recognition by pediatric cochlear implant users. *Ear and Hearing*, *16*, 470–481. doi:10.1097/00003446-199510000-00004

Moeller, M. P., Hoover, B., Putman, C., Arbataitis, K., Bohnenkamp, G., Peterson, B.…Stelmachowicz, P. (2007). Vocalizations of infants with hearing loss compared with infants with normal hearing: Part I–phonetic development. *Ear and Hearing*, *28*, 605–627. doi:10.1097/AUD.0b013e31812564ab

Moog, J. S., & Geers, A. (1990). *Early speech perception test for profoundly hearing-impaired children*. St. Louis, MO: Central Institute for the Deaf.

Niparko, J. K., Tobey, E. A., Thal, D. J., Eisenberg, L. S., Wang, N.-Y., Quittner, A. L., & Fink, N. E. (2010). Spoken language development in children following cochlear implantation. *Journal of the American Medical Association*, *303*, 1498–1506. doi:10.1001/jama.2010.451

Noij, S. V. K. S., Kozin, E., Sethi, R., Shah, P., Kaplan, A. B., Herrmann, B.…Lee, D. (2015). Systematic review of nontumor pediatric auditory brainstem implant outcomes. *Otolaryngology-Head and Neck Surgery*, *153*, 739–750. doi:10.1177/0194599815596929

Paden, E., & Brown, C. (1992). *Identifying early phonological needs in children with hearing loss*. Durham, NC: MED-EL Group.

Puram, S. V., Barber, S. R., Kozin, E. D., Shah, P., Remenschneider, A., Herrmann, B. S.…Lee, D. J. (2016). Outcomes following pediatric auditory brainstem implant surgery: Early experiences in a North American center. *Otolaryngology-Head & Neck Surgery*, *155*, 133–138. doi:10.1177/0194599816637599

Robbins, A. M., Renshaw, J. J., & Berry, S. W. (1991). Evaluating meaningful auditory integration in profoundly hearing-impaired children. *American Journal of Otology*, *12*(Suppl.), 144–150.

Sennaroglu, L., Sennaroglu, G., Yucel, E., Bilginer, B., Atay, G., Demir Bajin, M.…Ziyal, I. (2016). Long-term results of ABI in children with severe inner ear malformations. *Otology & Neurotology*, *37*, 865–872. doi:10.1097/MAO.0000000000001050

Shannon, R. V. (2015). Auditory implant research at the House Ear Institute 1989-2013. *Hearing Research*, *322*, 57–66. doi:10.1016/j.heares.2014.11.003

Sharma, A., Dorman, M. F., & Spahr, A. J. (2002). A sensitive period for the development of the central auditory system in children with cochlear implants: Implications for age of implantation. *Ear and Hearing*, *23*, 532–539. doi:10.1097/01.AUD.0000042223.62381.01

Teagle, H. F. B., Henderson, L., He, S., Ewend, M. G., & Buchman, C. A. (2018). Pediatric auditory brainstem implantation: Surgical, electrophysiolog, and behavioral outcomes. *Ear and Hearing*, *39*, 326–336. doi:10.1097/AUD.0000000000000501

Thielemeir, M. A., Tonokawa, L. L., Petersen, B., & Eisenberg, L. S. (1985). Audiological results in children with a cochlear implant. *Ear and Hearing*, *6*(Suppl.), 27S–35S. doi:10.1097/00003446-19850500-00007

Trammell, J., Farrar, C., Francis, J., Owens, S., Schepard, D., Thies, T.…Faist, L. (1981). *Test of auditory comprehension*. North Hollywood, CA: Forworks.

Wilkinson, E. P., Eisenberg, L. S., Krieger, M. D., Schwartz, M. S., Winter, M., & Glater, J. L.…the Los Angeles Pediatric ABI Team (2017). Initial results of a safety and feasibility study of auditory brainstem implantation in congenitally deaf children. *Otology & Neurotology*, *38*, 212–220. doi:10.1097/MAO.0000000000001287

Young, N. M., Kim, F. M., Ryan, M. E., Tournis, E., & Yaras, S. (2012). Pediatric cochlear implantation of children with eighth nerve deficiency. *International Journal of Pedeiatric Otorhinolaryngology*, *76*, 1442–1448. doi:10.1016/j.ijporl.2012.06.019

Zimmerman, I. L., Steiner, V. G., & Pond, R. E. (2011). *Preschool language scales* (5th ed.). San Antonio, TX: Pearson.

Zimmerman-Phillips, S., Robbins, A. M., & Osberger, M. J. (2000). Assessing cochlear implant benefit in very young children. *Annals of Otology, Rhinology, and Laryngology*, *109*(Suppl. 185), 42–43. doi.org/10.1177/0003489400109S1217