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Hearing Preservation in Pediatric Cochlear Implant – Is There a Role?

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Introduction

Congenital hearing loss affects approximately 1 in 500 live births, making it the most common birth defect. Adequate hearing is essential in order for children to develop normal spoken language and thus also plays a subsequent role in the development of reading comprehension, two fundamental skills without which individuals may be at a disadvantage in terms of societal integration, education, and employment. Early detection of congenital hearing loss is possible though the routine use of newborn hearing screening which can utilize auditory brain stem response and otoacoustic emission testing. Early detection of hearing loss is critical to appropriate rehabilitation because the reorganization of neural pathways that would otherwise be dedicated to auditory input, takes place at a young age in children with profound sensorineural hearing loss, and these pathways become reassigned to other tasks[1,2].

At later ages, if hearing is restored, the reassignment of these co-opted pathways to their original purpose of auditory processing becomes difficult. Thus congenitally deaf children who receive cochlear implants later in life do not achieve the same speech and language abilities as children who are implanted at younger ages[3]. The younger a deaf child is rehabilitated, the closer to normal their auditory development will be.

Ironically, children with bilateral partial deafness pose a more difficult challenge with regards to aural rehabilitation than those with bilateral profound sensorineural hearing loss. Most children with partial deafness have sharply sloping audiograms with lower hearing thresholds at low frequency and higher thresholds at high frequency. Accordingly, although deficits in the low-frequency range, if any, may be overcome with conventional hearing aids, high frequency hearing loss in the severe to profound range is challenging to aid via standard acoustic means particularly when it is isolated. Severe high-frequency deficits can be well rehabilitated through electrical stimulation (i.e. cochlear implantation); however, conventional cochlear implantation carries with it a risk of cochlear trauma, which can in turn lead to a loss of residual hearing. This risk of eliminating the residual native cochlear function along with moderate word recognition scores have traditionally prevented children with severely sloping hearing loss from being candidates for cochlear implantation. Unfortunately, despite residual low-frequency hearing and maximal acoustic amplification, these children still have significant difficulty with speech recognition and demonstrate verbal stigmata in consonant pronunciation when they speak. Clearly a better means of rehabilitation is needed.

Combined Electrical and Acoustic Stimulation

The tonotopic organization of the cochlea and the evolution of surgical techniques in cochlear implantation together allow for a relatively straightforward solution to the dilemma of the partially deaf or severely sloping hearing loss. The hair cells at the basal turn of the cochlea are tuned to higher frequencies, while hair cells nearer the apex of the cochlea are tuned to progressively lower frequencies. Accordingly, the patient with severely sloping hearing loss requires electrical stimulation of the spiral ganglion solely at the basal region. Approach to the cochlea via the facial recess allows implantation of a cochlear implant electrode, either via the round window or a cochleostomy, starting at the basal turn. Deeper insertion of the electrode will allow more apical electrical stimulation. Thus partial insertion of a conventional cochlear implant electrode or full insertion of a shorter electrode will provide the necessary electrical stimulation to the basal region only. This theoretically leaves the apical cochlea untouched and still able to receive acoustic signals transmitted normally via the traveling wave of the basilar membrane. In this fashion, an individual may receive combined electrical and acoustic stimulation (EAS).

It is likely that for many years even conventional cochlear implant recipients have benefited from combined EAS. Advances in surgical technique and electrode design have provided for rates of partial hearing preservation as high as 90% and of complete hearing preservation in 45% of conventional cochlear implant recipients[4]. The preservation of residual hearing in conventional cochlear implantation is not a matter of mere abstract interest. Even though maximal electrical stimulation is provided equally to both groups, those individuals with preserved residual hearing after conventional cochlear implantation fare better in speech perception tests than those who lost all residual hearing as a result of implantation[5]. This is potentially in part because those individuals with residual low-frequency hearing are supplementing the electrical signals provided by the implant with acoustic ones.

Intentionally combined EAS is currently achieved using short electrode arrays (10-20mm). The microphone and speech processor unit of the cochlear implant is coupled to a conventional hearing aid to form a hybrid device that delivers electric and acoustic signals. The short arrays are inserted into the basal turn of the cochlea but stop short of the apical regions of the cochlea where native hair cells are functional. Implantation with short electrode devices has been extremely successful in preserving residual hearing. One multicenter adult study in the United States preserved hearing in 96% of individuals and the average postoperative hearing levels were within 10 dB of preoperative levels (range 0-30 dB)[6]. Coupling the short electrode implant with a conventional hearing aid in these patients resulted in 79% monosyllabic word recognition scores. This method of EAS still carries the risk of cochlear trauma and elimination or reduction of residual hearing. A number of groups have been studying and advocated for a number of modifications to standard cochlear implantation techniques in an effort to maximize hearing preservation. For example, these include perioperative administration of steroids, the use of the round window for electrode insertion or 'soft' cochleostomy, the lateral placement of the electrode rather than perimodiolar, the use of hyaluronic acid to remove blood and bone dust from the cochlea, the use of an off-stylet technique, and changes in electrode design. A consensus on the method(s) most likely to preserve residual hearing however remains a matter of study and debate.

EAS Results in Children

A large experience with short electrode EAS devices in adults can be found in both North America and Europe, however up until relatively recently, the majority of the clinical experience with EAS devices in implanted in children has been in Europe. Combined stimulation devices were originally developed by von Ilberg[7] and colleagues and later expanded by Skarzynski and colleagues, who also implanted the first EAS devices in children[8]. In general, it is worth noting that results in terms of word recognition scores and hearing preservation can vary greatly across studies[9-11]. When comparing such studies, the most important variable that affects postoperative outcomes appears to be preoperative hearing levels, with better preoperative hearing levels typically, and understandably, leading to better postoperative word recognition scores. Better postoperative hearing levels[11], shorter duration of high-frequency hearing loss[12], and younger age[13] are also correlated with better postoperative speech recognition.

Because EAS is designed to enhance word recognition, the standard by which success of these procedures is measured is monosyllabic word recognition scores in quiet and noisy environments. In the largest series of EAS published to date, word recognition scores for children improved from 33% to 64% in quiet and 5% to 36% in noisy conditions[14]. A marked difference was seen between those children with normal low-frequency hearing and those with impaired, but aidable, low-frequency hearing. Children with normal preoperative low-frequency hearing showed both greater postoperative speech comprehension and greater improvements in these scores. These results are comparable to those seen in adults, who also show statistically significant differences in postoperative word recognition between individuals who require low-frequency acoustic amplification and those who do not[15].

The performance of individuals with hybrid or EAS devices improves over time, a phenomenon that is well studied in traditional candidates receiving conventional cochlear implantation[16]. Word recognition scores in quiet improves for up to 6 six months post implantation while performance on word recognition in noise can improve for up to 2 years after surgery. This difference in timeline to plateau for skills in quiet versus noise is important to consider when monitoring postoperative results as well as during pre-operative counseling of patients. Additionally, previous studies have found that a small but consistent subset of children will not be able to meaningfully participate in word recognition testing[8,14]. Accordingly, qualitative and surrogate measures of patient progress, such as school performance, behavior, and speech quality, may need to be employed rather than established quantitative measures.

Additional important considerations include loss of residual hearing in those individuals intended for or previously benefiting from EAS. A small but consistent proportion (approximately 5%) of individuals who undergo short electrode implantation will lose their residual low-frequency hearing, either by the time of their first postoperative audiogram or within the first 3 to 6 months after surgery. This begs the question of whether the short electrode implant can provide adequate electrical stimulation to allow for word comprehension in the absence of acoustic supplementation. Thankfully, the answer to this question appears to be "Yes." In individuals who lost residual hearing and were therefore forced to rely solely on electrical stimulation from the short electrode, word recognition scores were very similar to those who benefited from EAS[15]. This may be because of preserved low-frequency hearing in the contralateral ear, which allows the patient bimodal hearing (acoustic in one ear and electric in the other) that is functionally equivalent to unilateral EAS. Previous work has suggested that the two scenarios are equivalent with respect to word recognition[17]. Interestingly, this study also suggested that bimodal hearing with a conventional electrode in the implanted ear was superior to bimodal hearing with a short electrode. It is important to bear in mind that the electrode used in this study was very short (10 mm) and that the same disadvantage may not apply to longer "short" electrodes that are typically16 to 20 mm[15,18]. It is important to note that in other studies, the very short electrode implant performs similarly to the full array with respect to word recognition (reviewed by Turner[19]).

In addition to hearing in noise, temporal cues afforded by residual hearing may allow for better appreciation of music. Thus patients with EAS are expected to fare better in this arena than individuals with conventional cochlear implants, and this stands as an additional reason to strive for the preservation of residual hearing. Standard cochlear implant recipients struggle with tasks that are solely pitch dependent and lacking in rhythmic cues[20,21]. Tone discrimination is easily accomplished in normal hearing individuals when tones differ by 1%; however cochlear implant recipients are unable to discriminate between tones that differ by 36% or more[20]. Furthermore, although able to appreciate some of the more subjective properties of music, standard cochlear implant users still incorrectly identify tonal emotional qualities of music significantly more frequently than their normal hearing peers[22].

EAS makes a significant difference with respect to music appreciation and the performance of pitch-based tasks. Hybrid implant users perform at near-normal levels in the identification of melody-only popular songs, while standard cochlear implant users fare poorly at this task[23]. Not surprisingly, in the same study, patients benefiting from combined EAS performed very well on instrument identification tests provided the instrument registered in the lower frequencies where native cochlear function was present, but performed poorly if the instrument tested was played in the higher frequencies. EAS participants nonetheless performed better than conventional cochlear implant users.

Criteria for EAS Implantation in Children

Criteria for consideration of hybrid or EAS devices in children mirrors that of adults:

1. Stability of hearing. Children with unstable hearing or progressive hearing loss should generally not be considered for short electrode EAS. Although given the promising results mentioned above concerning the adequacy of the short electrode devices, we feel that enough of a difference between the conventional arrays and at least the shortest hybrid array exists to avoid placing this implant in a child that appears to be on a trajectory towards complete sensorineural hearing loss. This criterion is of even higher importance in children because the rate of hearing loss in children is more variable and unpredictable than in adults[24].

2. Low-frequency hearing. At least partial hearing should be present in order to make EAS useful. Generally this consists of pure-tone thresholds of 60 dB or greater at 250, 500, and sometimes 1000 Hz.

3. Other factors. Children and their families must be willing and able to participate in the same rigorous rehabilitation regimens and follow-up appointments as those required of conventional cochlear implant recipients. Accordingly, preoperative counseling and screening are as important for EAS candidates as for conventional cochlear implant candidates.

Future Possibilities with EAS in Children

The possibility of reliable hearing preservation with short electrode EAS devices in children will allow a number of advances in childhood cochlear implantation. One of these is the preservation of cochlear architecture, even if there is no functional significance currently. An oft stated argument against bilateral cochlear implantation is that both cochlea will suffer irreversible damage and hence be unsuitable for any newer technology that might arise in the long life most children with cochlear implants have ahead of them. This point is somewhat moot given that the window of opportunity a child has to appropriately program his or her auditory pathway is far smaller than the time it takes for a significant technological advancements to come to market; however, the short electrode implant may provide bilateral stimulation and appropriate organization of auditory pathways while simultaneously preserving a large portion of the cochlea for future technology. Under the banner of this philosophy, Gantz and colleagues have begun a trial of bilateral infantile cochlear implantation with a long, conventional electrode on one side and a short electrode on the other[25]. These children appear to perform as well as bilateral conventionally implanted children with respect to speech perception and speech and language acquisition. Of note, all of the children participating in this study have congenital complete sensorineural hearing loss.

It is common for children with congenital severe to profound sensorineural hearing loss to have some residual hearing. With bilateral implantation using a conventional electrode on the worse-hearing ear and a short electrode on the better-hearing ear, we might expect these children to develop speech and language without the typical audible stigmata of a steeply sloping hearing loss. One would expect these individuals to outperform both bilateral conventionally implanted individuals, conventionally aided patients with steeply sloping hearing losses, and perhaps even steeply sloping hearing loss patients with short electrode EAS devices who were implanted postlingually. Indeed, given the promising results of the Iowa study using one short electrode and one conventional electrode in congenitally deaf children, perhaps all children with severe-to-profound hearing loss should be implanted in this fashion. If residual hearing exists and persists, we may expect the child to outperform a bilateral conventionally implanted individual, while if residual hearing disappears or does not exist, then the results of the study suggest that the child will perform equally to a child with bilateral conventional implants. The effects of electrically remapping frequencies that were previously acoustically delivered in the event of progressive hearing loss has not been studied, however, and is a likely next step in the development of future "short-long" implant programs.

Conclusion

The development of the short array cochlear implant, its coupling to a conventional hearing aid, and the ability to preserve hearing with this electrode is an innovative solution to the problem of aural rehabilitation in children with steeply sloping high frequency hearing loss. Current studies are promising with respect to the ability of EAS devices to provide advantages in speech-in-noise comprehension and aspects of music appreciation. Future work will further illuminate the role of EAS devices in bilateral implantation, either in conjunction with another EAS device or with conventional or slim, full length electrodes.

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