Auditory Development in Children who Use Cochlear Implants
——Past Experience Guides Present Research——

Karen A. Gordon1,2)

1) Archie's Cochlear Implant Laboratory, The Hospital for Sick Children, Toronto, Canada
2) Department of Otolaryngology-Head and Neck Surgery, University of Toronto, Toronto, Canada

Cochlear implants have been able to restore hearing to children who are deaf1,2). These devices take the place of the dysfunctional cochlea by stimulating the auditory nerve with electrical pulses. The auditory pathways then carry this information to the brain. In this way, acoustic sound is represented by electrical pulses and interpreted by the central auditory system so that children can learn to hear and understand what they've heard.

Cochlear implants promote development in the auditory pathways3-5) and outcomes of cochlear implantation can be excellent. There are many reports of children using cochlear implants who achieve age-appropriate spoken communication skills6-8) and we have shown that their ability to sustain the voicing of a vowel sound (/a/) improves with implant use9). Of course, these outcomes are variable and, indeed, voicing remains less consistent than in normal hearing peers. Many studies have asked how we might predict which children will benefit most from cochlear implant use. Many factors have been suggested to play a role in how children will learn to understand spoken language with their cochlear implant. Most important appears to be the age at onset of deafness and age at cochlear implantation7,10-14). Auditory deprivation during potentially sensitive periods in early development leaves the auditory thalamo-cortex vulnerable to being taken over by other inputs15,16). These non-auditory inputs are often sensory; visual inputs are the most obvious invader of the deprived auditory cortex because of the increased reliance of vision when hearing is severe to profoundly impaired16,17). Thus, it is prudent to limit the duration of deafness in children. Because many children have congenital onset of deafness, this means providing cochlear implants at a young age. In our centre, we routinely provide cochlear implants to children with early onset deafness at ages as young as 8 months. Although it makes sense to provide a child with sound as soon as possible18), is it currently unclear whether implantation during infancy will provide better outcomes than waiting until the baby is over 12 months19). It is evident, however, that implantation before 2 years of age is important8,13). This is not to say that no child implanted at an older age will learn to hear speech. Indeed, older children who successfully achieved oral language skills, with normal hearing or through consistent hearing aid use, can be successful cochlear implant users20,21).

A number of other factors in addition to age, duration of deafness, and speech and language skills, should be assessed to determine cochlear implant candidacy. The degree of deafness must be known because, with most currently used implants, the residual hearing is at risk of being eliminated. Also,
implantation might be more challenging if the cochlea is malformed. This is assessed through imaging using Computer Tomography (CT) scans and Magnetic Resonance Imaging (MRI) which are also used to determine any abnormalities of the auditory nerve. A viable auditory nerve is imperative for cochlear implant function. The child must be healthy enough to withstand the operation and must have sufficient cognitive abilities to provide reliable behavioral responses to sensory input. This can be determined using somatosensory stimulation (e.g. vibration) prior to cochlear implantation. Reliable behavioral responses are necessary so that the child will be able to indicate whether the device ever provides noxious stimulation. We also need to ensure that the child and family have access to therapy which will emphasize listening and oral communication and that the family has the means and resources available to participate in this therapy. The family (and child if appropriate) should also have realistic expectations for what the cochlear implant can provide. The cochlear implant cannot restore normal hearing and this comes with long term implications for their academic and social lives.

**Research goals for future cochlear implant users**

Current research aims to capitalize on the advances already made through current cochlear implant technologies. If our ultimate objective is to establish normal hearing skills for children who are deaf through cochlear implantation, we will need to address a number of issues. I will discuss 3 major areas in which research is ongoing:

1) The cause and onset of childhood deafness and consequential effects on the central auditory system remain unclear in most children.

2) Current cochlear implants cannot fully represent the frequency, intensity, and timing information carried in complex acoustic sounds such as speech and music

3) Cochlear implants have traditionally been provided unilaterally which eliminates binaural processing.

**Childhood deafness**

It is clear that the duration of deafness during development will result in reorganization of the thalmo-cortical pathways which may be impossible to reverse through cochlear implant use. However, little is known about the etiology or onset of deafness and how these factors affect the immature auditory pathways. Changes in the central auditory system resulting from the type and/or onset of deafness in children could affect how the cochlear implant is able to stimulate the auditory nerve and the more central pathways in order to provide hearing.

Imaging and genetic testing have dramatically improved our ability to detect the origin of deafness in children. In a group of 72 children, we found that approximately 15% had cochlear abnormalities (including enlarged vestibular aqueducts which are of little concern for cochlear implantation). Another approximately 20% of these children had severe homozygous mutations of GJB-2 which normally codes for the Connexin 26 protein (important for gap junctions in the cochlea). Another 5 children (7%) were diagnosed with cytomegalovirus based on MRI. This means that the etiology of deafness was unknown in the majority (58%) of children in this group. Furthermore, we cannot be sure of the onset of deafness in any of these children. Neonatal hearing screening helps to determine whether hearing loss is present at birth but we do not know whether the auditory system was able to receive any input during gestation. Normal hearing in humans begins in the third gestational trimester and this prenatal stimulation is likely important for auditory development. Because of the variability in etiology of deafness and the often unknown onset of deafness in children, the integrity of the auditory pathways which are to be stimulated by the cochlear implant remains unknown.

Current efforts to understand the effects of deafness on the auditory system have used Positron Emission Tomography (PET) and MRI prior to cochlear implantation. These studies have found some interesting correlations between the resting state of the brain and speech perception scores.
obtained after cochlear implantation. In children, a loss of hypometabolism in the deaf auditory cortex correlated with poor post-implant performance and was interpreted as a permanent and abnormal takeover of the auditory brain by other inputs (cross-modal plasticity)\(^1\). In adults, post-implant speech perception improved with increases in activity in left pre-frontal cortex and decreased activity in right Heschl’s gyrus and posterior superior temporal sulcus\(^2\). These findings suggest that there is a complex network of cortical function which can be affected by deafness and which is important to cochlear implant use.

We have assessed the functional effects of deafness on the auditory pathways by recording electrophysiological measures at the beginning stages of cochlear implant use prior to chronic stimulation. We reported that the auditory brainstem response evoked by cochlear implant stimulation is very robust in children who are deaf\(^2\) indicating that these pathways remain viable and can respond in a recognizable pattern immediately upon electrical stimulation. Moreover, there were no significant effects of duration of deafness in children on amplitude or latencies of the auditory brainstem response peaks\(^2\) which means that degeneration in the auditory brainstem pathways measured by this technique is too subtle to be detected using these measures. More recently, we have studied the effects of deafness on cortical responses. Unlike the auditory brainstem responses, cortical responses in children at initial cochlear implant use show a very high degree of variability\(^2\). This suggests that there are multiple effects of deafness in the immature brain. We isolated 3 main types of cortical responses in this group of children. They were mostly young at the time of cochlear implantation and age did not help to predict the type of response recorded. On the other hand, we found a more uniform response type in children whose deafness was due to severe biallelic GJB-2 mutations\(^2\). In comparison, auditory nerve responses in children whose deafness was of unknown etiology were larger in amplitude when evoked by an electrode at the apical end of the cochlear implant array than a one at the basal end.

In sum, the central auditory system changes during the period of deafness and these changes could depend upon the etiology and/or onset of deafness. It will be important to identify deafness-induced changes in the auditory system if we are to optimally stimulate this system using electrical pulses from a cochlear implant.

**Limits of current devices**

The interaction between the cochlear implant device and the central auditory system will also depend upon what auditory information the cochlear implant is able to provide. The implant can deliver sufficient frequency and intensity information over time in order to represent speech. This we know because children and adults learn to recognize speech sounds with their cochlear implants. However, the implant can only provide a crude representation of complex sounds and this could compromise hearing abilities. In order to study this issue more closely, we have been assessing the limits of hearing through a cochlear implant in children.

To do this, we have asked children to listen for the emotion carried in speech or music rather than asking about the content of the sound. Music can be happy or sad depending upon the the mode (major vs. minor key) and tempo (fastness and slowness of a piece of music) in which it is played. In western music, major keys and fast tempos are considered happy whereas minor keys and more slow tempos are sad. Using a previously published test of music\(^2\), we asked children using unilateral cochlear implants whether short musical excerpts were happy or sad. Of 32 pieces of music, children using cochlear implants were able to accurately determine whether they were happy or sad 78% of the time. This was significantly better than chance (50%) but also significantly poorer than the performance of normally hearing children who found this task very easy to do (97% accuracy).
We also used the DANVA test\textsuperscript{29} to assess whether children using cochlear implants could hear emotion carried in speech. In this test, the children heard the same sentence ('I’m going out of the room but I’ll be back later”) spoken in a happy, sad, angry or fearful voice. Children using cochlear implants had a more difficult time distinguishing between these emotions (50% accuracy) than normal hearing children (79% accuracy). This finding indicated that, whilst cochlear implants convey sufficient information about the content of spoken language, they are less able to provide subtle cues carried in speech such as the emotion of the speaker.

In sum, we need to provide cochlear implant users with better representation of the acoustic input so that they can have better access to subtle cues available in speech and music. Future cochlear implant designs and speech processing strategies will perhaps address some of these concerns.

**Unilateral input**

Cochlear implant users are often tested in ideal listening environments (ie. in the soundbooth) where there is no noise and the acoustic input is coming from one place. In reality, however, they are listening in noisy situations, to multiple talkers who may move around. This is particularly true for children who need to hear in group situations such as the classroom and playground. Normal hearing individuals can use binaural cues to help in these situations; sounds coming from any direction other than directly in front or behind will reach each of the two ears at a slightly different time or level. The auditory pathways are able to detect these fine differences in timing and level between the ears and allow the listener to identify the direction of the sound. Binaural processing thus allows for sound to be organized in space. These binaural cues are helpful for hearing in noise but children provided with one cochlear implant do not have access to them. Cochlear implantation of both ears is consequently being done in an effort to restore binaural hearing abilities.

Bilateral cochlear implantation has helped children who are deaf in both ears to localize sound better than they could with a single cochlear implant\textsuperscript{30–33} and to better perceive speech in noise\textsuperscript{34–40}. However, outcomes are highly variable and remain poorer than normal hearing peers. We have therefore been exploring effects of bilateral cochlear implantation on development of the central auditory pathways in children.

We were first interested in the ability to stimulate the auditory pathways with the second cochlear implant. Because unilateral cochlear implantation promotes development of the auditory brainstem\textsuperscript{4–25} and thalamo-cortical pathways\textsuperscript{3,5}, we expected that pathways stimulated by the newly implanted ear would be more immature than those with prior cochlear implant experience. This mismatch in development was confirmed in the auditory brainstem\textsuperscript{41,42}. Brainstem responses evoked by the newly implanted side showed delayed peak and interpeak latencies relative to the responses from the first implant in children who had used that first cochlear implant alone for several months or years. In comparison, children who received bilateral cochlear implants simultaneously (in the same surgery) showed no mismatches in auditory brainstem function. Early results suggested that these differences in response latencies were resolved after 9 months of bilateral cochlear implant use in children who had only short periods of unilateral cochlear implant use (<12 months) but not in children with longer unilateral cochlear implant exposure\textsuperscript{41,43}. The mismatch in auditory brainstem timing indicates that unilateral cochlear implant use sets up an asymmetry of function in the auditory brainstem which could have consequences for binaural processing. We are currently exploring these issues further by examining developmental plasticity in the thalamo-cortex using a novel beamformer to isolate specific cortical generators involved in hearing with two cochlear implants\textsuperscript{44–46}. Our aim is to determine whether there is a critical period for binaural hearing and whether binaural processing can be elicited through bilateral cochlear implantation.

We have also been using behavioral measures to assess binaural hearing in children using bilateral cochlear implants. In a recent study, we asked
children who used one cochlear implant for a number of years prior to receiving a second device to listen to electrical pulses delivered to both cochlear implants. These pulses were delivered slowly (11 pulses per second) and could be offset in time or current level. These stimuli were randomized along with presentations in which bilateral pulses were of equal current level with no timing differences and with stimulation to either the left or right implant alone. We asked the children to tell us whether they heard these inputs as coming from the middle of their head, the left side, right side, or both sides at once. A control group of children with normal hearing were included; these children listened to similar cues in acoustic clicks which were also presented at 11 per second. The children using cochlear implants were able to perform the task after some training as demonstrated by accurate identification of unilateral presentations from the left or right cochlear implants. They were also able to perceive differences in level of bilateral presentations. Stimuli in which higher current levels were provided by the second implant were perceived as coming from that side. As the bilateral input became more heavily weighted in level from the first implanted side, the children indicated that the input shifted to that side. Thus, children using bilateral cochlear implants were able to make use of inter-implant level differences. On the other hand, these children were not able to hear differences in timing between implants despite the fact that these cues were easily distinguished by the children with normal hearing. We also noted that the children using bilateral cochlear implants rarely indicated that they heard bilateral input as coming from the middle and, occasionally, they indicated that they heard sounds from both implants separately. None of the children with normal hearing gave the latter response. Based on these results, we suggest that children who receive bilateral cochlear implants after long periods of unilateral cochlear implant use have better access to interaural level cues than timing cues.

In sum, bilateral cochlear implants are currently providing children with greater access to binaural cues than they had with one cochlear implant. This provides benefit for sound localization and hearing in noise. However, these abilities are poorer than normal perhaps reflecting asymmetry in function in the bilateral auditory pathways in children who had previous unilateral cochlear implant exposure and/or abnormal binaural processing (particularly of interaural timing cues). Future work will explore whether the provision of bilateral cochlear implants simultaneously allows for better development of binaural processing in children.

Conclusions

Current research strives to improve upon current knowledge with an aim to provide children who are deaf with as normal hearing as possible. To meet this objective, we will need to better understand deafness in childhood, to improve upon current cochlear implant technologies, and to provide bilateral implants where appropriate.

References


