Fully Awakening the Auditory System: Bilateral Cochlear Implants in Children

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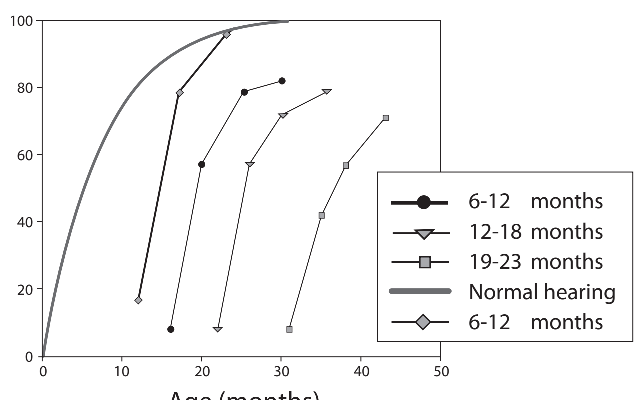
Cochlear implantation has revolutionized the treatment of severe to profound SNHL in children. Over the two decades since their mainstream adoption, unilateral implants have demonstrated their ability to provide deaf patients with the ability to hear sound and communicate effectively. Bilateral cochlear implantation (BCI) offers the potential benefits of improved speech development and the promise of improved sound localization. We will review the state-of-the-art in pediatric BCI and focus on the importance of early implantation, its safety, and cost effectiveness.

Plasticity

To place this discussion it is important first to realize the importance of early detection and implantation of severe to profoundly deaf children. Early implantation is the single most important determinant of speech and language outcome and independently has the greatest impact yet demonstrated on improving outcome. In **Figure 1** you can clearly see that as the age at unilateral implantation decreases the rate of improvement steepens and the eventual peak of performance increases. In the children implanted under the age of one in this study, the score on this test reaches the level we would expect in normal hearing children at around age two!

Figure 1. Performance on the IT-MAIS test in groups of unilaterally implanted children at varying ages of implantation. (SickKids unpublished data)

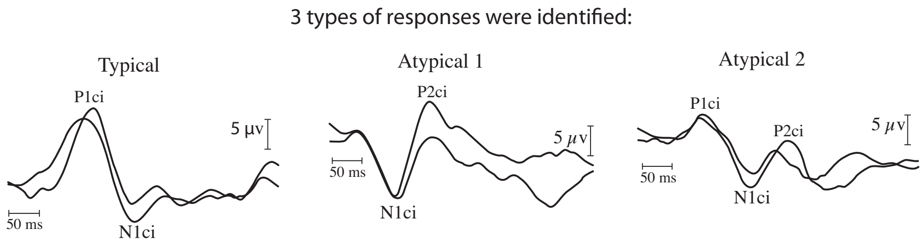


To further place the discussion in context it is important to review some key electrophysiology concepts with regards to auditory pathway development. With brainstem response audiometry (ABR) and electrical evoked brainstem response audiometry (eABR) it is possible to examine the brainstems' and early cortical responses to sound stimuli in normal and BCI patients. A sound stimulus can be applied to one ear and the latencies in waves I, III, and IV of the ABR and eABR can be observed and recorded. In children with BCI, these waves and their corresponding latencies can then be compared to the waves on the contralateral side. Most importantly, wave V corresponds to the response of the lateral lemniscus, and this is the location where inputs from the left and right are compared in the brainstem for the first time. Studies have been conducted at our institution examining differences in the development of wave V in children who were bilaterally implanted sequentially (BCIse) and those who were implanted simultaneously (BCIsi). The results demonstrate that patients who were implanted simultaneously have wave V responses that are symmetrical bilaterally. This symmetrical electrophysiologic response theoretically represents 'fusion' of the auditory information from both ears. Interestingly, in children with BCIse with a long delay between implantations, it has been found that these two wave V signals occur at very different latencies and never match – meaning that such patients might never develop binaural fusion, and may have difficulty extracting information from their complex auditory environments.

Detailed measurements were performed on children after receiving implants simultaneously, with a short delay, and with long sequential implantation. Our data demonstrated that simultaneously implanted children "fused" their wave V at initial device activation, while those with short delay did not fuse until around nine month's time. Long sequentially implanted patients began to show trends toward fusion approximately three years after their first implant, but none of these patients have truly fused as of yet. This might significantly reduce the advantage of providing the second implant to these individuals.

Cortical responses were also measured in BCI users at device activation. Three types of cortical responses were identified and were termed 'normal', corresponding to a waveform consistent with that from a normal hearing individual, and 'atypical 1' and 'atypical 2' (see **Figure 2**). Initial findings demonstrated that older patients and those with longer duration of deafness were more likely to produce an atypical response. Furthermore, in younger patients there likely exists an optimal period of plasticity in the auditory cortex and implantation within this "critical period" might allow optimal development of the processes required for normal auditory processing. In theory a key advantage clinically would be that if binaural fusion could actually take place the listener would be more capable of discerning a signal and extracting it from the background noise. This of course would be a tremendous advantage to children in a classroom setting.

Figure 2: Cortical Responses in children with cochlear implants demonstrating the three commonly found types of cortical responses identified.



In children with BCI it has been demonstrated that compared to the first implanted ear alone in quiet, speech perception in the bilateral condition is improved for those patients implanted either simultaneously or with short delay. Our sequentially implanted patients, on the other hand, demonstrate a very poor performance in signal to noise ratio and are particularly poor when noise is supplied to the first implanted ear. Clearly, these patients are relying more heavily on their first implanted side and are not taking advantage of a binaurally fused image.

Towards Improved Outcomes – Bilateral Cochlear Implantation in Children

In 2005 a prospective study at our institution was begun looking at children with simultaneous bilateral implants. Over 100 children have been enrolled to date. This group was compared to BCIse with short intervals between implants (<1 year) and BCIse with >3 years between sequential implants. Our first analysis of these groups focused on surgical safety and close examination has demonstrated that simultaneous BCI is as safe as unilateral implantation. Specifically there are no differences in pain control, days of hospitalization, or post-operative complications. The same anesthesia and surgical technique is used for BCI as is used for unilateral implant surgery. The only additions to the surgical procedure are that we have to turn the head after the first implant is placed and the wound closed and perform an identical contralateral procedure. Originally this surgery was performed on older children, but as discussed above younger and younger children were implanted and now we attempt to implant all children before the age of one year (if we are certain of the audiological thresholds), including those with developmental delay or even cochlear anomalies.

The impact of cochlear implants on the vestibular system is an area that remains under active study. It is known that 35% of children with SNHL demonstrate a concomitant vestibular dysfunction. Interestingly, a group of BCI patients were studied and compared to normal hearing and SNHL patients. Our cohort did not demonstrate significant post operative balance dysfunction and had levels of dysfunction equal to what we would expect in SNHL patients.

While the balance function in the BCI patients was below the normal group, it was clear that the implanted children had compensatory mechanisms that were active and at least equal to those of normal hearing children. Both groups demonstrated the same benefit from having their eyes open versus closed in several tests of vestibular function.

Localization

The discussion of plasticity of the auditory cortex above indicated that an advantage in speech in noise is conferred with early simultaneous implantation. Further potential benefits that may result from a fused binaural image are seen when special unmasking is examined. Spatial unmasking refers to the patient's ability to discriminate the locations of sounds presented from either side or the center. When this was examined we demonstrated that compared to normal listeners, BCIsi patients no significant difference. It is important to remember these patients are still quite young (approximately five years of age), but nonetheless performed well at this basic localization task. It is hoped that as the BCIsi cohort gets old enough to test formally in more advanced paradigms of localization, they will show significant benefit compared to unilaterally and BCIse patients.

Cost Effectiveness

The cost-effectiveness of a second cochlear implant has still not been convincingly demonstrated. What has been demonstrated is that there is significant outcome benefit from BCIsi versus BCIse and unilateral implantation but not to the degree that cost-effectiveness can be determined. On the other hand, the cost of performing BCIsi is significantly less that performing BCIse surgery. BCIsi is performed at a substantially decreased cost when compared to performing two separate surgical procedures. This strongly supports performing simultaneous surgery in centers capable of safely carrying out the procedure. If there is cost-effectiveness in the BCIsi group, it will likely be best demonstrated in the patients demonstrating significant outcome benefits and having the lowest health care costs.

Imperfect Improvements

While BCI has delivered many improvements for hearing and language, there is still much on-going research and development. With existing BCI technologies, the auditory system is not yet being fully awakened – children continue to have difficulty, particularly with sound localization and speech in noise.

The major issue facing children with cochlear implants today is hearing in noisy environments such as the classroom. Theoretically, bilateral implants and a fused binaural auditory system could allow extraction of the signal from the noise to take place. This ability has not yet been reliably demonstrated in bilaterally implanted animals or humans except in substantially controlled laboratory settings. Nonetheless, bilaterally implanted children report reduced head shadow and improved ability to identify the source of sounds. Many report improved speech perception in noisy environments as well but the data demonstrating this ability has not yet conclusively been reported. Our studies on localization and lateralization in BCIsi patients demonstrate that they have some improved localization ability compared to the BCIse group but are still nowhere near the results obtained in normally hearing children. The BCIsi group is still quite young and further studies will be required to delineate any differences more precisely.

Conclusions

The developing auditory system is plastic within sensitive periods. BCIsi and BCIse are safely performed operations and are optimally aimed at establishing binaural fusion which theoretically would result in improved psychophysical outcome. A complete team with excellent clinical support personnel and therapists is essential for achieving success with a cochlear implant program. The cost-effectiveness of BCI is greatest when implants are placed simultaneously as a result of decreased surgical and health care related cost. While bilateral implants offer advantages over unilateral implantation, there are still strides to be made in improving speech in noise and sound localization. By implementing the above state-of-art findings and with continued research into the areas where improvements are needed the promise of fully awakening the auditory system with simultaneous bilateral cochlear implantation is close at hand.

Recommended readings

1. Papsin BC, Gordon KA: Cochlear implantation in a child with severe to profound deafness. N Engl J Med 2007: 357(23): pp 2380-7.
2. Gordon KA, Valero J, Papsin BC: Auditory brainstem activity in children with 9-30 months of bilateral cochlear implant use. Hear Res 2007: 233(1-2): pp 97-107.
3. Papsin BC, Gordon KA: Should bilateral cochlear implants be the standard of care in children with bilateral deafness? Current Opinions in Otolaryngology - Head and Neck Surgery 2008: 16(1): pp 69-74.
4. Ramsden JD, Papsin BC, Leung R, James A, Gordon KA: Bilateral simultaneous cochlear implantation in children: Our first 50 cases. Laryngoscope 2009: 119(12): pp 2444-8. utcome. Otol Neurotol 2009: 30 (4): pp 488-495.
5. Salloum CA, Valero J, Wong DD, Papsin BC, van Hoesel R, Gordon KA: Lateralization of interimplant timing and level differences in children who use bilateral cochlear implants. Ear Hear 2010: 31(4): pp 441-456.