Current Trends in Pediatric Cochlear Implantation

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Introduction

Since cochlear implants (CI) were introduced for clinical use in the 1960's, they have undergone many advances, and are likely the best-performing neural prosthesis available today. The most recent data from the National Institutes of Health (NIH) indicates that worldwide, more than 300,000 individuals have been implanted, with a majority of patients being children. From the initial single-channel array, they have evolved into multichannel devices that can deliver intelligible speech and even musical information to some patients. The efforts of several disciplines, including engineering, acoustics, otolaryngology, and audiology have led to advances in hardware design, surgical technique, and signal processing for continued development of CI technology.

In the past 50 years, since the first patients received commercial CI devices, implantation criteria for both children and adults have undergone many changes. Initially the technology was reserved only for patients with bilateral profound deafness. However, due to improved function, safety, and the increasing benefits of cochlear implantation, CIs are now available to many patients with less severe forms of hearing handicap. While the current Food and Drug Administration (FDA) criteria for children have remained unchanged for more than a decade, the premise that profound bilateral sensorineural hearing loss is a critical and sometimes the sole criterion for the indication of CI in children is being challenged. This chapter will review the recent developments and current state of pediatric cochlear implantation.

Current Criteria for Pediatric Cochlear Implantation

Based on FDA guidelines in the United States, the criteria for CI in children remains based on documented audiologic thresholds and auditory progress. In general, for children less than 2 years of age, CI is indicated for bilateral profound sensorineural hearing loss (SNHL). For children older than 2 years, the criterion is lowered to those with severe to profound SNHL. Speech recognitions scores of 30% or less are also used to qualify a child for implantation. In addition, there needs to be a 3 to 6 month trial with properly fitted hearing aids and demonstration of little or no progress.

Unilateral vs. Bilateral CI for Children

Unilateral vs. bilateral implantation in children has been the subject of much debate. There are obvious added costs of a 2nd implant and additional risks of surgery on both sides. However, there is building evidence to support bilateral CI in children, and it is increasingly considered the standard of care for pediatric patients (National Institute on Deafness and other Communication Disorders, 2011). While children with unilateral CI perform well in quiet controlled environments (Sarant *et al.*, 2001), the results of testing in sound booths do not reflect common listening

environment for children in everyday situations such as classrooms, gymnasiums, or cafeterias. To support this, studies have demonstrated some delays in language development in children with unilateral CIs (Geers 2002; Tobey *et al.*, 2003). Innate binaural hearing has the advantages of sound localization and superior speech understanding in the presence of background noise. While the benefits of bilateral CI use in children for sound localization has been demonstrated in some (Litovsky *et al.* 2006a), many children show no sound localization ability with their implants (Gavin *et al.*, 2007). However, bilateral implants have a clear advantage in children over unilateral in the area of speech perception in quiet (Scherf *et al.* 2007) and noisy environments (Litovsky *et al.* 2006b). More recently, studies examining long term vocabulary and language outcomes have shown a clear advantage in children with bilateral CI as compared to unilateral (Sarant *et al.*, 2014). Based on these and other studies, the default in our program is to offer bilateral cochlear implants in children unless there are other factors to recommend otherwise.

CI in Children with Malformed Inner Ears

Malformations of the inner ear are common in children with SNHL and can account for the etiology in up 35% of patients with congenital hearing loss (Rachovitsas *et al.*, 2012). Initially, cochlear implantation in children with inner ear malformation was contraindicated due to the increased risks of surgery and decreased potential functional outcomes (Weber *et al.*, 1998). In addition, anatomical studies revealed diminished spiral ganglion cells in these patients (Monsell *et al.*, 1987). Numerous studies have reported outcome results after CI in children with inner ear malformations. Some patients with inner ear malformation do not fare as well as those with normal anatomy, particularly those with total semicircular canal aplasia, cochlear nerve hypoplasia, cochlear nerve aplasia, or common cavity (Buchman *et al.*, 2004). However, with careful preoperative planning, most children with inner ear anomalies can undergo cochlear implantation safely without significantly increased risks and those with certain malformations. Children with cochlear and vestibular dysplasia receive some benefit and children with enlarged vestibular aqueduct, demonstrate functional speech outcomes similar to children with normal inner ears (Lee *et al.*, 2010; Lee *et al.*, 2014). Children with inner ear malformations require additional precautions and careful preoperative counseling but can safely be implanted with good outcomes in certain patients.

Hearing Preservation CI in Children

Partial deafness with severe to profound loss in high frequencies and reasonable to good hearing in the low to mid frequencies, also known as a ski slope hearing loss, is more often found in the older adult population and commonly seen in presbycusis. Based on initial criteria, these patients were not candidates for implantation. However, such patients often have poor speech discrimination and many do not perform well with hearing aids (Hogan and Turner, 1998). Numerous studies in adults have demonstrated the ability to preserve low frequency acoustic hearing (Reviewed in Huarte and Roland, 2014). Accordingly, the use of electrical and acoustic stimulation (EAS) has been shown to provide superior speech perception in adults (Adunka *et al.*, 2013). While this pattern of hearing loss is less common in children, a recent study shows that complete hearing preservation can

be achieved in a majority of pediatric patients and some hearing can be preserved in all children (Skarzynski *et al.*, 2016). In addition, it has also been shown that children have the highest potential for having preserved hearing following CI surgery (Anagiotos *et al.*, 2015). Further investigation of hearing preservation in children undergoing CI surgery may lead to this becoming a standard option opening the benefits of improved function to a greater population of children with hearing loss in the future.

CI Children with Single-sided Deafness

Recently there has been increasing investigation of CI as a potential treatment option for single-sided deafness (SSD), particularly when associated with incapacitating tinnitus (Van de Heyning *et al.*, 2008). While the literature on this continues to evolve, there are studies that demonstrated sound localization and speech perception benefits in adults undergoing CI for SSD (Tavora-Vieira *et al.*, 2015; Zeitler *et al.*, 2015). While studies in children are extremely limited, there is data that showed rapid development of speech discrimination in the implanted ear, improvements in sound localization and speech perception in noise, as well as a high degree of patient satisfaction, hinting towards potential benefits in a wider population (Vlastarakos *et al.*, 2014). Further investigation along this line may open the benefits of CI to children with SSD in the future.

Summary

Cochlear Implants have dramatically changed the lives of countless children with hearing loss who otherwise would have gone thru life without the sense of sound and limited to lip reading and non-verbal communication. Currently recommendations are for children to receive bilateral cochlear implants unless anatomical or other limitations would indicate otherwise. Inner ear anomalies are no longer considered a contraindication for CI in children, and some children with abnormal anatomy, particularly enlarged vestibular aqueducts do quite well with implants. Children with partial deafness who maintain some low frequency in many cases perform better with a CI a hearing aid, opening the benefits of implants to another population of children with hearing impairment. While evaluation of the benefits of CI for SSD is still in its infancy in children, further investigation in this area may demonstrates that implanting children with unilateral hearing loss may also receive substantial benefit from this game changing device.

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