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Cochlear implantation in late childhood and adolescence: is there such a thing as 'too late'?

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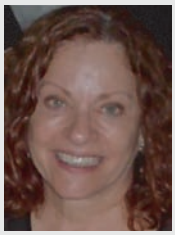

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"Although the associated performance may not parallel 'normal' hearing age-matched individuals or those implanted at a younger age, late cochlear implantation in prelingually deaf individuals provides a communication benefit, which should not be negated."

Cochlear implantation has significantly positively, impacted the lives of deaf individuals internationally. The device that converts acoustic energy into electrical stimulation bypassing endogenously dysfunctional cochlear structures has gained widespread popularity in the aural rehabilitation of patients with severe to profound sensorineural hearing loss. The performance of patients following cochlear implantation can be broadly variable, dependent on a number of factors, including age of onset of deafness, duration of deafness, age of implantation, etiology of deafness, use of hearing aids, mode of communication, cognitive ability, motivation and psychosocial support system to name a few [1–4].

With regard to cochlear implant candidacy, it is widely accepted that individuals with congenital deafness implanted in early childhood and individuals with acquired deafness of a short duration have the best potential for cochlear implant performance in terms of speech recognition and word discrimination. However, controversy remains surrounding the utility of later cochlear implantation in congenitally deaf individuals, namely cochlear implantation in late childhood and adolescence. Is there, in fact, a well-defined window of opportunity for implantation of congenitally deaf children, or can congenitally deaf children obtain benefit from cochlear implantation if implanted at an older age?

There have been numerous studies that have revealed that early cochlear implantation in prelingually deaf children is advantageous for a number of reasons. Cochlear implantation has been found to be safe and efficacious in children below the age of 12 months, and rates of major and minor complications are equivalent to older children and adults [5,6]. In addition, these early implanted children have been shown to develop age-appropriate auditory perception and oral language skills [5]. When comparing children implanted between 12 and 36 months of age, children who received implants at a younger age have been found to acquire auditory skills nearer to those of their peers with normal hearing at a younger age [7]. Adolescents who had undergone cochlear implantation in early childhood have been found to report strong social skills and high self-esteem on analysis [8]. The vast majority also report strong identification with the hearing community or mixed identification with both the deaf and hearing community [8]. In addition, in a similar study, the overwhelming preponderance of adolescents implanted in early childhood was reported to be completely mainstreamed by high school [9].

Keywords: adolescent • childhood • cochlear implantation • cochlear implant performance • congenital deafness • prelingual deafness

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There remains some controversy within the literature regarding a sensitive period for cochlear implantation in congenitally and prelingually deaf children. During the first years of life, external stimulation is critical for the normal neurological development of critical functional networks and neural connections, which support behavioral learning [10,11]. Although there is a high degree of neuroplasticity resulting in later synaptogenesis, auditory deprivation has been found to result in abnormal or delayed maturation within the auditory cortex [12,13]. Although studies have revealed no age of implantation-dependent difference in electrically evoked compound action potential of the auditory nerve and brainstem following cochlear implantation [14], differences have been reported in higher order function along the auditory pathway. Studies of cortical auditory-evoked potential P1 peak latency time reveal critical differences dependent on the age of implantation. In normal hearing individuals, with age the latency between presentation of auditory stimulus and cortical auditory-evoked peak potential P1 decreases with time, indicating a shorter time period necessary for cortical response to auditory stimulus. Prelingually deaf children implanted before the age of 3.5 years exhibit normal cortical P1 latencies within 6 months of implant use [10]. Alternatively, children implanted above the age of 7 years exhibit abnormal P1 latency, which does not reach normal latencies following implant use, and it has been suggested that these differences may be critical for optimal speech and language development [10]. Additional studies have investigated this concept of a critical point as represented by performance on speech perception testing. Harrison *et al*. investigated the impact of age of implantation on cochlear implant performance in an attempt to identify a critical point for implantation in prelingually deaf children [15]. In this study of 82 children implanted between 2 years and 13 years of age, cochlear implant performance was assessed as a function of age [15]. Children implanted at 5 years of age and younger were found to outperform their older peers in all phoneme and word speech perception tasks with children implanted at 2 years of age exceeding all other groups [15]. When corrected for duration of deafness, the optimal split was found to be at age 4.4 or 5.6 years based on Glendonald Auditory Screening Procedure word and Test of Auditory Comprehension scores, respectively [15]. Although these findings support the assertion that early cochlear implantation yields the best cochlear implant performance, they also show benefit in those implanted at a later age.

> **"Although studies have revealed no age of implantation-dependent difference in electrically evoked compound action potential of the auditory nerve and brainstem following cochlear implantation … differences have been reported in higher order function along the auditory pathway."**

With newborn screening programs becoming more prevalent, it is often possible to identify children with congenital deafness early, allowing for cochlear implantation within the first year of life. However, newborn hearing screening programs are not universal. Within the USA alone, the vast majority of states have newborn hearing screening programs, although certain states allow parents to opt out of hearing screening [101]. As such, many children with congenital deafness may not be identified. In areas with recently implemented screening programs, later diagnosis may occur in children born prior to implementation. In addition, children may be lost to follow-up, lack resources/social support for intervention or present with acquired prelingual deafness, which may lead to later diagnosis. The question arises, "How should these children be managed?" There is a window of time documented wherein cochlear implantation is of increased benefit in children with prelingual deafness. However, is there a window after which cochlear implantation is of negligible benefit in the congenitally or prelingually deaf individuals? The literature appears to support increased auditory and linguistic benefit following early implantation but does not support a total lack of benefit following cochlear implantation at a later age.

There are emerging data supporting the benefit of late cochlear implantation in individuals with prelingual deafness. In recent data from the University of Iowa comparing children implanted before the age of 2 years with children implanted from age 2 to 4 years, children within the older group perform equally to their early implantation counterparts by >3 years of cochlear implant use [16]. Caprosecco *et al*. presented cochlear implant outcomes of 38 individuals with bilateral sensorineural hearing loss diagnosed before the age of 3 years who underwent cochlear implantation at 14 years of age or older [17]. Within their review, the majority of patients had a confirmed diagnosis of severe to profound prelingual deafness (prior to age 3 years). Ten patients had a diagnosis of sensorineural hearing loss prior to the age of 3 years with progression (severe to profound sensorineural hearing loss) thereafter. Within this study, all patients improved beyond sound detection. A total of 53% of patients attained open set speech recognition with scores of >30% on City University of New York Sentences and Central Institute for the Deaf Everyday Sentence Lists testing. Of these patients, 21% attained scores of greater than 90% on both City University of New York Sentences and Central Institute for the Deaf Everyday Sentence Lists testing. In addition, 90% of patients reported that their ability to understand speech with visual cues was 'much improved' following cochlear implantation, and 85% of patients felt that the implant had a positive or somewhat positive effect on their employment. Furthermore, 70% of patients reported use of the telephone following implantation. A total of 81% reported using their cochlear implant for greater than 8 h per day. Three factors were found to have a significant impact on cochlear implant performance within this population: mode of communication in childhood (oral vs total communication or American Sign Language), stable versus progressive hearing loss and time without use of a hearing aid on the implanted ear. Patients who used oral communication, had progressive hearing loss and wore a hearing aid on the implanted ear were more likely to obtain better speech perception outcomes.

Similarly, in a review by Zeitler *et al*. of 67 patients with prelingual deafness, post-operative cochlear implant performance was retrospectively studied [18]. The mean age at cochlear implantation was 12.9 years with an associated mean duration of deafness of 11.5 years, representing 61% of patients with

congenital sensorineural hearing loss. Within this series, significant improvement in speech perception scores for both Hearing in Noise sentence test and Consonant-Nucleus-Consonant monosyllabic word test scores were noted at 12 months post-operatively with mean score changes of 51.1 and 32.2%, respectively ($p < 0.001$). The mean changes in Hearing in Noise sentence test and Consonant-Nucleus-Consonant monosyllabic word test scores at the end of study were 60% and 38.7%, respectively ($p < 0.001$), indicating that the majority of improvement in performance occurs within the first year of cochlear implant use. Like the study by Caprosecco *et al.*, adolescents with progressive deafness and those using oral communication have significantly better cochlear implant performance than age-matched peers [17].

> "Although these findings support the assertion that early cochlear implantation yields the best cochlear implant performance, they also show benefit in those implanted at a later age."

In a review by Santarelli *et al.* of 18 prelingually deaf-implanted adolescents and young adults (age 13–30 years), cochlear implant performance at 6 months, and 1, 2 and 3 years was reported [19]. Although cochlear implant performance was found to be lower than previously published data within the literature obtained from early implanted congenitally deaf patients, phoneme identification, word and sentence identification scores improved significantly following cochlear implantation in these late implanted patients. In this study, word and sentence recognition scores continued to reveal significant improvement beyond the 1 year postimplantation interval. Mean values of approximately 10% preoperatively improved to 38.7, 51.6 and 65.6% postoperatively for disyllabic, trisyllabic and sentence scores, respectively.

Similar findings have been reported in cochlear implantation of prelingually deaf adults. Prelingually deaf adults who underwent cochlear implantation exhibited significant improvement in open set speech perception (sentence) scores following implantation with a mean score change from 7.0 to 46.7 ($p < 0.05$) [20]. Improved performance in this population was found to positively correlate with higher preimplantation thresholds and negatively correlate with the primary use of sign language for communication preimplantation [20].

According to current definitions of success and metrics utilized to assess success, late cochlear implantation in prelingually deaf individuals has been demonstrated to be of benefit. However, one may argue that within this population a more muted definition of benefit should be utilized. In the study by Caprosecco *et al.*, although open set speech recognition of >90% was not attained in the vast majority of patients, >90% of patients reported improvement in communication ability [17]. The main benefits of cochlear implantation in this population may be to aid sound awareness, lip reading and overall communication ability as well as the possibility of speech understanding as shown in several published articles. Although the associated performance may not parallel 'normal' hearing age-matched individuals or those implanted at a younger age, late cochlear implantation in prelingually deaf individuals provides a communication benefit, which should not be negated. Given that these recipients report that implantation positively impacts their lives, ability to communicate, opportunities for employment and thus quality of life, cochlear implantation in this population should be considered a viable option for patients with preoperative counseling, realistic expectations and positive motivation.

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