Pediatric Cochlear Implantation

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Cochlear implantation represents the only successful prosthetic replacement of special sensory neural function in humans. The remarkable outcomes in children who have received early intervention for their hearing loss attests to its success. This article will answer some of the questions I am frequently asked about this technology.

1. **What is the magnitude of the problem cochlear implantation addresses?** Every year 3 out of every 1000 newborns have a major hearing impairment, making hearing loss the most common sensory deficit in children. This means that 33 babies are born every day with hearing loss. Of these, 95% are born to normal hearing parents. Since hearing is essential for the development of spoken language, these children face a massive hurdle to effective communication, even within their own families. Prior to cochlear implantation, a child with profound hearing loss who was confined to the traditional deaf education system would, on average, graduate from high school with a 3rd grade reading level (Project Hope Survey 1991). They faced three times the rate of unemployment as the hearing population and 70% relied on public assistance. The overall cost to society for deaf education and special public services is enormous, estimated at up to $1 million per child over the course of their life.

2. **How do cochlear implants work?** Cochlear implants are highly complex electrical stimulators. In one sense they are akin to vagal nerve stimulators (used for seizures) and to deep brain stimulators (used for Parkinson's disease), inciting neural function through electrical stimulation. However, they are far more complex because they are attempting to represent, in real time, the complex spatial, temporal, and frequency characteristics of sound via electrical stimulation. Cochlear implantation has taught us a great deal about the physics of sound and how the normal human auditory system works. The cochlea is tonotopically organized (the low frequencies are represented in the apical turn and high frequencies in the basal turn). Yet the various sounds around us are so complex, particularly human speech, that representing each one to the cochlea in a distinctive way through finely tuned electrical stimulation has developed into a discipline unto itself. Engineers specializing in psychoacoustics have developed "processing strategies"; complex algorithms that tell a cochlear implant how to stimulate the cochlea in such a way (at up to 31,500 pulses per second distributed over 22 channels along the length of the cochlea) so that each sound incites a distinctive combination of neural impulses. The "brains" of the implant are contained in the external sound processor worn on the ear, similar in appearance to a behind-the-ear hearing aid. The internal implant, consisting of a receiver, electronics package, and stimulating electrode, simply carries out its commands.

3. **What are the indications for cochlear implantation in children?** A cochlear implant is indicated for severe to profound sensorineural hearing loss in children. In other words it takes the place of a poorly functioning cochlea if hearing aids are unable to provide adequate hearing for a child's optimal development. In the vast majority of children with sensorineural hearing loss (congenital and acquired) the problem is poorly functioning cochlear hair cells. Since the spiral ganglion nerve endings supplying these hair cells are still present, they can be electrically stimulated, which is then perceived by the auditory cortex as sound. Cochlear implants are not needed when there is a conductive hearing loss (i.e. ear canal atresia with normal cochlear function) or when hearing aids work effectively for development of speech and language as treatment for lesser degrees of hearing loss .

4. **Who are good candidates?** The current mantra of pediatric cochlear implantation is, "**the younger the better.**" Cochlear implants have also taught us a great deal about cortical brain development in children, particularly with regard to sensory input. When a child is born, they have yet to lay down an efficient network of higher cortical neural projections necessary for cognitive processing of sensory input. This cortical development occurs postnatally, but only in response

to effective auditory input and only for a limited period of time. In the first 3 years of a child's life the brain is maximally "plastic." After this age central neural plasticity begins to decline and it is greatly diminished after 7 years of age. The first 3 years of life have come to be known as the "critical period" when central neural plasticity is at its maximum. Every day, week, and month after birth that a child is without sound they are losing irreplaceable time for optimal auditory outcomes. Currently the FDA criteria allows implantation at 12 months of age. The trend however is to proceed even earlier (down to 6 months) in order to capitalize on this limited window of opportunity. Late presentation (after 3 years of age) is associated with poorer outcome. Early intervention has produced astounding outcomes in children with profound hearing loss. They frequently meet or surpass age norms for receptive and expressive language and usually can be mainstreamed in school by the 1st grade. This results in a huge cost savings to our special education system. It also highlights the value of universal newborn hearing screening (NBHS) towards the goal of early diagnosis.

5. **How well does someone hear with a cochlear implant?** Because there are many facets of hearing function that we measure (speech discrimination, sound localization, tone sensitivity, etc) it is difficult to convey this answer in one statement. To generalize, cochlear implants can transform a child's profound hearing loss to a mild hearing loss. Children with a cochlear implant are not able to function in all adverse sound environments (high background noise) as well as normal hearing children, but their remaining impairments are manageable. Auditory performance has been improved even further by **bilateral** cochlear implantation, which has now become the standard of care.

6. **Should parents wait for future technology, or more advanced implants?** (See response to question 4 above.) The answer is **absolutely not**. Children must be implanted as young as possible. The current limiting factor for a child is not the state of technology but their own limited time window of opportunity for auditory cortical development.

7. **What is the most important take home message for pediatricians with regard to pediatric hearing loss?** Cochlear implants can change the entire course of a hearing impaired child's life, but only if they are diagnosed and present for treatment early. Newborn hearing screening has been very helpful in this regard, but has a false negative rate averaging 11%. This means that of the children born with hearing loss 1 out of 10 will be missed by newborn screening and be thought to have normal hearing. Therefore it is important that pediatricians be quick to order further testing on any child in who even the slightest concern for hearing loss has been raised, either by the parents or healthcare givers, regardless of the results of NBHS.

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