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Cochlear Implantation



By **Anita Jeyakumar, MD**, pediatric otolaryngologist at Children's Hospital. This issue of *Pediatric Review* is intended for pediatricians, family physicians and all other interested medical professionals. For CME purposes, the author has no relevant financial relationships to disclose.

OBJECTIVES

At the end of this activity, the participant should be able:

1. Explain how a cochlear implant helps children with hearing difficulty
2. Describe which children are candidates for cochlear implantation
3. Discuss educational implications and medical needs of cochlear implant recipients

INTRODUCTION

An estimated 4,000 to 8,000 children are born in the United States each year with severe hearing impairment. A majority of the children are born to normal hearing parents. Pediatricians and parents need to understand the available options and be prepared to sort through the complex data and multiple options in order to arrive at the decision that is best for the family and their child.

So, what is a cochlear implant? Who is a candidate? Who is not a candidate? Why use cochlear implantation versus deaf education and American Sign Language? What are the educational implications? What is the quality of life in children with cochlear implants? Who pays? What are the special medical needs of a cochlear implant recipient?

WHAT IS A COCHLEAR IMPLANT?

A cochlear implant is a surgically implanted device that can provide a sense of hearing to a patient who is profoundly or severely hard of hearing (Figure 1). The quality of sound is different from natural hearing, with less sound information being received and processed by the brain. However, many patients are able to hear and understand

speech and environmental sounds. Newer devices and processing strategies allow recipients to hear better in noise, enjoy music and even use their implant processors while swimming.

WHO EXACTLY IS A COCHLEAR IMPLANT CANDIDATE?

That question doesn't have a simple, straightforward answer. It requires a multidisciplinary team to establish whether a child can benefit from a cochlear implant. The team consists of a cochlear implant surgeon (otolaryngologist), pediatric cochlear implant audiologist, pediatric audiologist, neuroradiologist, speech language pathologist, cochlear implant nurse, deaf educators, social workers and parent(s).

Cochlear implant candidacy criteria have evolved dramatically since multichannel implants were first approved for pediatric use by the FDA in 1990. Initially, only individuals with bilateral profound sensorineural hearing loss with no open set speech recognition were considered candidates for cochlear implantation. Over time, however, these criteria have become less stringent and we are now implanting individuals with greater amounts of residual hearing and pre-implant speech recognition scores.

Speech recognition outcomes have been climbing steadily since the earliest reports following multichannel implantation. Part of this improvement is attributed to advancements in technology both with the internal device as well as for the externally worn processors. Another portion of this improvement can be attributed to the advancements in speech coding strategies (e.g., how the incoming signal is transformed from the complex acoustic waveform to the electrical stimulation of auditory nerve fibers via the intracochlear electrodes).

Cochlear implant criteria for children are considerably more complicated than adults with many more elements of candidacy, including age, audiometric thresholds, auditory progress with hearing aids, speech recognition performance (for older children), and etiology, to name a few.

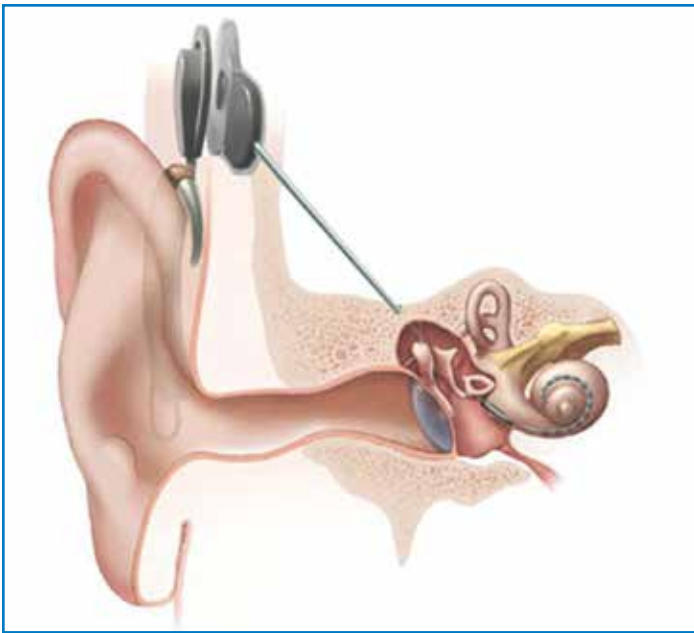


Figure 1. Cochlear implant with electrode array going to the cochlea

Starting with audiometric threshold, for children aged 12 to 24 months, the current criteria specify bilateral profound sensorineural hearing loss. It is important to note that this indication does not imply that children with less severe hearing loss do not benefit from cochlear implants.

Children have to go through a hearing aid trial before consideration of cochlear implantation. Children under two years of age, during the hearing aid trial, should be making at least month-to-month auditory progress as well as speech and language developmental progress. In other words, if a child has been fitted with hearing aids for three months, they should make at least three months of progress in auditory skills and speech/language development. If this is not the case for a child making full-time use of amplification and appropriate intervention, then a cochlear implant evaluation should be considered.

For children two years of age and older, current audiometric criteria for implantation are slightly more lenient, including bilateral severe-to-profound sensorineural hearing loss. Again, if a child has less severe sensorineural hearing loss and is not making auditory progress with full-time use of well-fitted hearing aids and recommended intervention, referral for a cochlear implant evaluation is appropriate.

WHO IS NOT A CANDIDATE?

Historically, children with multiple disabilities were not considered cochlear implant candidates. However, recent studies have shown that children with disabilities can have a significant improvement in their quality of life following cochlear implantation.

An absolute contraindication is a child who does

not have any cochlea or auditory nerve on either side. Anatomy can be determined by imaging, often a CT scan and/or MRI.

The lack of realistic expectations regarding the benefits of cochlear implantation and/or a lack of strong desire to develop enhanced oral communication skills poses a strong contraindication for implant surgery.

WHY COCHLEAR IMPLANTATION VERSUS DEAF EDUCATION AND AMERICAN SIGN LANGUAGE?

Cochlear implants are paradigmatic of a particular type of ethical dilemma in which advocacy groups have been at odds with children's parents or doctors. Fortunately, this has continued to evolve positively. Most deaf educators feel that aural communication is most effective for children with cochlear implantation. Also, advocacy groups are more often working with physicians and the families to help the family make a more informed decision.

WHAT ARE THE EDUCATIONAL IMPLICATIONS?

The costs associated with profound deafness are substantial. The expected lifetime cost to society for a child with prelingual onset of profound deafness without cochlear implantation exceeds \$1 million, largely because of special education and reduced work productivity. A 2000 Journal of the American Medical Association (JAMA) article summarized some early data very effectively, showing an overall cost reduction in children undergoing cochlear implantation. Including indirect costs, such as reduced educational expenses, cochlear implantation provided a savings of \$53,198 per child. The data is very similar to more recent data from other developed countries. Specifically, an international systematic review shows that while healthcare costs are high, savings in terms of indirect and quality of life costs are also significant.

WHAT IS THE QUALITY OF LIFE IN CHILDREN WITH COCHLEAR IMPLANTS?

Cochlear implants have been shown to literally transform the lives of recipients in a way that they have been labeled as the most successful and effective implantable prosthesis in terms of restoring function to recipients. A recent study from Dallas showed that children with cochlear implants experience quality of life similar to that of normal-hearing peers, and did not experience any of the isolation that has been reported in hearing-impaired counterparts.

WHO PAYS?

Most insurance companies and Medicaid will pay for a unilateral implant. At Children's Hospital in New Orleans,

however, if a child is assessed as being a candidate for a bilateral cochlear implant, an endowment can provide the funding for the second implant.

WHAT ARE THE SPECIAL MEDICAL NEEDS OF A COCHLEAR IMPLANT RECIPIENT?

Any child with hearing loss needs to be evaluated by specialists. However, if a child has been evaluated and received a cochlear implant, there are several medical and lifestyle considerations:

MRI: Individuals with cochlear implants are able to obtain MRIs, but have to take specific precautions. The MRI can potentially demagnetize the surgically implanted device or damage the metallic components.

Meningitis: Cochlear implant recipients can be more prone to pneumococcal meningitis, and have special protocols to get all the vaccinations before and after implantation. The protocols follow the CDC guidelines.

X-rays and CT scans: These are generally safe. The external processor should be removed before getting these tests, as the programs can be wiped out by the tests.

Head trauma: A blow to the head in the area of the implant can damage the implant and result in failure of the internal device. Children should take precautions with contact sports.

Static electricity: A discharge of static electricity directly to the external processor can degrade or damage the processor. Rarely, this can also damage the internal (implanted) device. Activities that can generate static are plastic slides, trampolines, balloons and plastic ball pits. Parents should ground themselves by touching the child's arm or leg before touching the processor.

Disposable zinc air batteries: These are used for the external processor, are a choking hazard and are to be kept in a tamper-proof case.

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Every 4.5 minutes, an American baby is born with a birth defect

Birth defects are a leading cause of infant and childhood mortality in the United States. There are many different kinds of birth defects, including congenital heart defects, cleft lip or palate, defects of the brain and spine, and a variety of genetic syndromes, such as Down syndrome. Some have only a minor and brief effect on a baby's health and some have life-threatening and/or life-long effects. These conditions also place considerable physical, emotional and economic burdens on individuals, families, and society at large. In fact, every 4 ½ minutes, a baby is born with a birth defect. More than 120,000 babies born with a birth defect (approximately 1 in 33 live births) are reported each year in the United States. In Louisiana, approximately 1,500 children are born with a birth defect each year. The risk for many types of birth defects can be reduced through healthy lifestyle choices and medical interventions before and during pregnancy. In honor of January as Birth Defects Prevention Month, Children's Hospital is actively focusing on raising awareness among healthcare professionals and the general public about the frequency with which birth defects occur and the steps that can be taken to prevent them.

In the United States, birth defects account for 20 per-

cent of infant deaths, as well as 6 to 15 percent of deaths in children up to age 14, according to the National Birth Defects Prevention Network (NBDPN). The most common type of birth defects, congenital heart defects, have a birth prevalence of about 1 in 100 births, and are a leading cause of infant mortality. Congenital cardiac and circulatory anomalies account for about one in three birth-defect related hospital stays and account for \$1.4 billion of the \$2.6 billion in annual hospital expenses attributed to birth defects, according to the Agency for Healthcare Research and Quality. Around 20 – 30 percent of people with congenital heart defects have other physical problems or developmental or cognitive disorders.

The medical costs (inpatient, outpatient and prescription drug expenses) for a child with an orofacial cleft are 500 to 800 percent higher than those for a non-affected child. The estimated overall lifetime expenses related to Down syndrome range from \$180,000 to \$880,000 (in 2013-adjusted-dollars) according to a 2008 study in Pediatrics. None of these numbers consider the loss of income and productivity, or the day-to-day emotional and physical stresses that affect both the patient and the primary caregivers.

The New England Journal of Medicine estimates that we know the cause of only about 30 percent of birth defects. While no known etiology has been identified for most birth defects, research continues to further our understanding of their causes. However, many preventive measures have been identified and raising awareness and universal utilization of these measures, both among the public and the healthcare community, can help to significantly limit the occurrence of birth defects. Maternal obesity, diabetes, lack of adequate folic acid/folate intake, smoking, alcohol consumption, infections, and many legal and illegal drugs have been clearly identified as major factors in determining the risk of birth defect occurrence.

Although not all birth defects can be prevented, the healthcare community can help all women (including teens) who are pregnant or could become pregnant to lower their risk of having a baby with a birth defect by encouraging them to follow some basic health guidelines throughout their reproductive years, including:

- Take 400mcg of folic acid daily from the beginning of menstruation through menopause.

- Eat a healthy diet and aim for a healthy weight.
- Keep diabetes under control.
- Get a medical checkup before pregnancy and address specific health issues including weight control, control of diabetes, and any medications taken.
- Stop smoking and avoid second-hand smoke.
- Stop drinking alcohol prior to pregnancy or as soon in pregnancy as possible.
- Do not take illegal drugs.
- Plan carefully. Use contraception if taking medications that increase the risk for birth defects.

You can make a difference in the lives of Gulf South families. Children's Hospital and The National Birth Defects Prevention Network encourage you to review this information and reach out to your patients.

For more information on Children's Hospital's birth defect related services, please visit www.chnola.org/services. The National Birth Defects Prevention Network may be visited at www.nbdpn.org.



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 - 4,000
 - 6,000
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- Multichannel implants were first approved for pediatric use by the FDA in _____.
 - 1985
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 - 2000
- True or False, cochlear implant criteria for children are considerably more complicated than for adults
 - True
 - False
- Children who are not candidates for cochlear implants include those _____.
 - With multiple disabilities
 - Whose families cannot afford the surgery
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