Overview:
Hearing allows us to be conscious of what is going on around us. It is always working to warn us of danger. Most importantly, hearing permits communication. Hearing loss affects 28 million Americans (1 out every 10 people), and 1 in 1000 children are born deaf. The isolation from society and learning that occur in patients with untreated hearing loss leads to developmental delays. It is critically important to diagnose and treat hearing loss as quickly as possible.

Sensorineural hearing loss is primarily a disease of the auditory portion of the inner ear, the cochlea. Hair cells are the sensory receptor cells and are the most important cell in the inner ear. They convert the mechanical sound waves into neural information for transmission to the brain. Hair cells can not regenerate. When they are lost, a permanent hearing loss is the result. There are many different causes of sensorineural hearing loss. In children, the most common causes are prematurity, infections around the time of birth, genetic predisposition, or developmental abnormalities.

Evaluation:
When a child is first identified with sensorineural hearing loss, the medical evaluation should be based upon trying to identify the reason for the hearing loss. Occasionally, it is possible to find a treatable cause of the hearing loss so progression can be prevented. Also, it is important to identify problems with other organ systems that may have occurred in combination with the hearing loss. Finally, other family members may be at risk of hearing loss and can be identified and treated at an early age.

We have a standard screening protocol for all children referred to our center with sensorineural hearing loss. After careful medical and audiometric examinations, we often order several tests to understand the disease process as best as possible. These include a CT scan of the temporal bone to look for inner ear malformations, an MRI scan of the brain to evaluate the auditory nerve and to look for co-existent central nervous system abnormalities, a renal ultrasound to rule-out kidney malformations, and an electrocardiogram to look for heart conduction problems. We draw blood to look for genetic and infectious causes of hearing loss. We also routinely recommend families for a genetics consultation and ophthalmologic consultation.

Treatment:
Most children are started on hearing aids immediately. If a child has a mild or moderate sensorineural hearing loss, hearing aids, assistive listening devices and hearing and speech/language therapy may be the only treatment needed. Periodic hearing tests will be performed by a pediatric audiologist to monitor the degree of hearing loss as well as your child’s progress in developing communication skills.
Good hearing is critical for children to learn to talk. Speech and language therapy by a speech and language pathologist or a specialized teacher of the hard of hearing is an important component of a well-rounded treatment regimen.

**Cochlear Implantation:**
Children with severe to profound sensorineural hearing loss are considered for cochlear implantation. A cochlear implant is an electronic device that is implanted behind the ear under the skin. An electrode is inserted into the inner ear, and restores hearing in patients who have severe to profound hearing loss. A cochlear implant bypasses the normal sound conduction mechanism through the external, middle, and inner ear, and directly stimulates the auditory nerve. An implant does not restore or create normal hearing. Instead, under the appropriate conditions, it can give a deaf person a useful auditory understanding of the environment and help him or her to understand speech. Cochlear implants were approved by the Food and Drug Administration in the mid-1980s and are covered by most private insurance policies, Medi-Cal, and California Children’s Services (CCS).

**Making the Decision for Cochlear Implantation:**
In order to be considered for a cochlear implant, patients have to have lost so much hearing that a hearing aid is of little to no benefit. The most common children who benefit from a cochlear implant are those who are born deaf. However, the only way to determine whether or not a patient would benefit from cochlear implant is to be evaluated by the implant team of physicians, audiologists, and therapists. A cochlear implant evaluation is a very involved process involving several members of our cochlear implant team (see below). The patient and family will be counseled extensively and be examined by a pediatric audiologist, a pediatric speech pathologist, and a psychologist. Other physicians may also be involved if clinically indicated. The cochlear implant team members then meet to decide whether they think the child could benefit from a cochlear implant. If the cochlear implant team agrees that a cochlear implant has a good chance of being helpful, this procedure is offered as an option to the parents/guardians. If they desire to proceed in this way, surgery is then scheduled.

**Cochlear Implant Technologies:**
A cochlear implant is made up of two parts. The external device consists of a microphone behind the ear that hears the sounds in the environment. These sounds are then digitized and processed by a small computer called a speech processor. Small speech processing units can be worn behind the ear while larger ones have to be clipped on to the patient’s clothing. Historically, the bigger the processor, the more capability it had to select speech signals out of the environment, ultimately improving hearing. However, computer technology has continued to improve so that behind-the-ear speech processors now provide essentially equivalent results.

The signals from the speech processor are sent to the implanted part of the device though the skin using a magnet. The implanted part is an electronic device that is put under the skin behind the ear. An electrode connected to the device is inserted into the auditory portion of the inner ear, the cochlea. The electrode is simply a bundle of tiny wires that have open contacts spread out along the length of the cochlea. Thus, the electrical signals can be sent to different areas of the cochlea and represent different frequency sounds. State-of-the-art
cochlear implant devices now have 12 to 22 electrodes which stimulate the auditory nerve. These multi-channel implants have the advantage of stimulating many different nerve fibers individually, thereby transmitting more detailed information to the brain. The more information that reaches the brain, the greater the patient's ability to understand what is happening in his/her environment.

**Benefits of a Cochlear Implant:**
If hearing aids do not provide sufficient benefit for oral language development, a cochlear implant may be recommended. A cochlear implant allows a deaf patient to hear. Hearing through an implant sounds different than normal hearing, but it allows many people to communicate fully with oral communication in person and over the phone. Untreated severe or profound hearing loss in young children has significant deleterious effects on speech and language development. Because a child learns much about his/her world by listening, a cochlear implant can provide significant improvements in a child's ability to learn to communicate. Cochlear implants have also benefited school age and older children as well to help them develop and improve their communication skills and achieve their educational goals. Most, although not all, children who receive a cochlear implant end up being mainstreamed in community schools and do quite well with their peers.

**Alternatives to a Cochlear Implantation:**
For children, once the diagnosis of severe hearing loss is made, parents may chose to have their child taught in either a spoken language, a manual language such as American Sign Language (ASL), or a combination of both (Total Communication). Sign language is a way to teach children language and how to communicate without having to hear. As well, they can develop close relationships with other people in the deaf community.

**Cochlear Implant Surgery:**
This is a routine type of surgery for physicians trained in ear surgery. It takes about one and a half to two hours and involves making an incision behind the ear. The mastoid bone behind the ear is drilled to visualize the inner ear. The body of the device is countersunk into the skull and the electrode is inserted into the inner ear. The incision is then closed and a dressing applied. In most cases, the child is kept in the hospital one night for observation. It is not particularly painful, and patients can resume their normal activities within 2-3 days typically. The implant is not turned on until the incision has fully healed, about 4 weeks after surgery.

Many parents desire bilateral cochlear implants for their child (one for each ear). Although both can be placed in a single operation, usually the second is placed 3 or more months after the first one is placed. This reduces the length of time the child is under anesthesia and allows the child to recover from the first surgery. As well, it offers the opportunity to see how the child responds to the cochlear implant before committing to another one.

**Risks of Cochlear Implantation Surgery:**
Although cochlear implantation is almost always safe, complications are possible, just as with any kind of surgery. Inserting a cochlear implant often destroys any residual hearing in the operated ear. Thus, one can not turn back. The standard surgical risks of a cochlear implant
are all uncommon. These include: bleeding, infection, device malfunction, electrode or device migration, facial nerve weakness, ringing in the ear, dizziness, and poor hearing result.

One important risk of a cochlear implant is meningitis (infection of the fluid around the brain). This is very rare (reported to be 91 cases out of 60,000 patients with cochlear implants). However, 17 of these patients have died. These patients who had meningitis tended to have several predisposing risk factors including congenital inner ear malformations, a previous history of meningitis, immune system dysfunction, age less than 5 years, and a history of recurrent ear infections. Because the cochlear implant runs between the middle and inner ear, bacteria within the middle ear may tract along the implant into the normally sterile inner ear. The inner ear has connections with the brain, through which the infection may spread. All patients who will receive a cochlear implant have to be immunized pre-operatively against some common bacteria that cause meningitis.

An additional consideration is learning to interpret the sounds created by an implant. This process takes time and practice. Speech-language pathologists, teachers of the hard of hearing, or educational audiologists are the professionals frequently involved in this learning process. Teacher and parental involvement are paramount. Not everyone performs at the same level with a cochlear implant. Prior to implantation, all of these factors need to be discussed and understood.

The Cochlear Implant Team:
The pediatric cochlear implant team is part of the Children’s Hearing Center at LPCH, a combined program at Stanford University and Lucile Packard Children’s Hospital. The team is dedicated towards the evaluation and management of pediatric patients with profound sensorineural hearing loss. This team works closely together to provide individualized, yet highly specialized, care for this diverse and complex patient population. Each team member gets to know each patient and their family quite closely during the cochlear implant evaluation process. The members of the cochlear implant team are listed below.

**Otolaryngologist - Head and Neck Surgeon.** This is a surgeon who has done residency training in Ear, Nose, and Throat (ENT) and who has also undergone fellowship training either in surgery of the Otolgy, Neurotology, and Skull Base Surgery or in Pediatric Otolaryngology. This physician performs the medical evaluation and begins the hearing loss evaluation. If the decision made by the family and the entire cochlear implant team is to proceed with cochlear implantation, this physician will perform the surgery.

**Cochlear Implant Program Coordinator.** The program coordinator tracks all patients undergoing cochlear implant evaluation, and works very closely with the patient and their family. All tests and evaluations results are organized together for presentation at the weekly cochlear implant team meeting. The coordinator spends a great deal of time counseling the family. This includes teaching what a cochlear implant is and what it can be expected to do. Additionally, family members need to understand the degree of time commitment and hard work it will take on their part to achieve an optimal long-term result. Often, the coordinator will
learn about the child's school environment by talking with their teachers or making a school visit. Visits to the home are sometimes required.

**Pediatric Audiologists.** This is a doctoral level hearing professional with expertise in evaluating and treating children with hearing loss. The audiologists who work with cochlear implantation are specially trained in this area. Pre-operatively, they will determine the nature and severity of the hearing loss and provide extensive counseling regarding hearing loss and the communication and educational choices available to you and your child. Their evaluations include diagnostic hearing assessments, as well as hearing aid consultations, fittings and maintenance. Finally, an auditory skills and hearing aid verification evaluation is performed to determine the child's ability to attend to and integrate sound using conventional amplification.

During the surgery, the audiologist is present in the operating room. Immediately before and after the cochlear implant is placed, they interface it to an external computer to verify that the internal electronics are working. If there is a device malfunction, a backup device is on-hand so that it can be replaced before closing the wound.

After surgery, the audiologist has a long-term commitment to help the patient use the cochlear implant to the best of their abilities. Four weeks after surgery, they activate the device and begin initial programming. Repeated visits will be needed to determine what works best for each individual patient. This involves similar evaluations to those performed at the time of diagnosis or when a hearing aid was assessed. These assessments are used to set the current levels for each electrode that will provide a tolerable, yet audible, level of electrical stimulation for the patient. The audiologist also obtains behavioral measurements of the child's response to sound using the implant and monitors and manages the hearing loss in the implanted and opposite ear.

**Speech, Language, and Learning Center (Speech Pathology).** Speech, Language, & Learning provides evaluation, management, and consultation for infants, children, adolescents, and young adults with hearing impairments who are in the candidacy process for a cochlear implant. An initial evaluation is performed to determine the child's performance levels in receptive and expressive language, speech, and audition. The tests used during the initial evaluation are selected to fit the needs of the patient. Following the evaluation, results are shared with the cochlear implant team and parents. Occasionally, these services may include conferences with parents, teachers and other professionals. Recommendations are made and a report is written. Reevaluations are also conducted to document progress after cochlear implantation. As a result of the initial evaluation, the speech-language pathologist may recommend speech therapy services for the patient. These regular sessions are designed to help the patient overcome or compensate for any problems identified in the evaluation related to the hearing loss. The sessions, usually held individually, are planned with activities appropriate for the patient based on the results of the evaluation. Not all patients seen for evaluation are enrolled in therapy with us. Other recommendations may be made, such as a referral for outside community services.

**Psychology.** All children who are being considered for a cochlear implant will undergo extensive testing to assess their ability to learn to use an implant. This is because children
with difficulty concentrating on tasks or learning may need additional support services after surgery to allow them to grow to their fullest potential. Testing of the parents is also performed, because parental involvement in the child's education is critical to achieving good outcomes. These tests are performed at regular intervals after implantation has been performed to allow us to adapt our rehabilitation strategy to the child's needs.

**Genetics.** Genetic mutations are responsible for over half of the cases of childhood hearing loss. Children seen in the Children's Hearing Center are evaluated by a geneticist specializing in pediatrics to look for syndromic features. This is so that other potential medical problems can be identified. Additionally, genetic counseling will be performed to help the parents understand their risk of having other children with hearing loss.

**Developmental Pediatrics.** Some children being evaluated for cochlear implantation require an assessment by a pediatrician with expertise in neuro-developmental disability. This evaluation looks at the overall general pediatric, developmental, and psychosocial condition of the child, in addition to whether there is any evidence of a neurological or genetic syndrome. It is very important to estimate each child's future neurological status, as this can affect their ability to use a cochlear implant.

**Ophthalmology.** It is important that children with one sensory deficit get evaluated for other sensory deficits. The effects of more than one problem on childhood development are severe. As well, some syndromes involve both hearing and visual defects. Children seen at the Children's Hearing Center are evaluated by ophthalmologists skilled in the diagnosis of subtle ocular changes related to hearing loss.

**Research coordinator.** Stanford is a research-intensive institution and the Children's Hearing Center at LPCH integrates excellent clinical care with basic, translational, and clinical research programs. We have a research coordinator who will be contacting you to discuss opportunities for you and your child to be involved in our ongoing research studies. While there is certainly no requirement to participate, we have found that most families do so because they get to learn more about how their child is progressing. As well, the advances lead to improved developmental outcomes for all children.