INTRODUCTION

The advent of the cochlear implant has brought the world of sound to a growing population of patients. The number of patients who are candidates for cochlear implantation has increased steadily as we have worked to find new ways to incorporate cochlear implantation in the care of patients. Dr. Fishman wrote, “The minimum requirement for cochlear implantation is the presence of an implantable cavity and proximity to stimulable neural elements whose projections connect to the auditory cortex.” (1) This quote by Dr. Fishman emphasizes the role of cochlear implantation especially in patients who have difficult anatomy or underlying pathology that make cochlear implantation more difficult. The question is who fits these criteria for cochlear implantation? Second, who doesn't fit this criterion? And lastly, what makes the patient difficult to implant?

ANATOMY AND EMBRYOLOGY

In order to better understand both the role of cochlear implantation the process in which it works, we must first understand the anatomy and embryology the ear better. To quickly summarize the physiological cause of hearing, sound waves travel through the ear canal vibrating the tympanic membrane. This then causes the ossicular chain to vibrate. The stapes attached the oval window causes a hydraulic like motion pushing fluid waves to the cochlea. This causes motion of the basilar membrane which is then sensed by the inner hair cells. Inner hair cell motion then activates or inhibits chemical transmitter release. The nearby neurons are then activated based on these changes and transmit the signal to the central auditory complex. This is what creates sound in the normal working ear. (2)

The ear starts as a formation of the otic placode which is an ectodermal thickening in the third gestational week. This is the beginning of the combined cochlear and vestibular membranous labyrinthine system. The placode envaginates from the surface and forms the otocyst in roughly the fourth gestational week. There are 3 infolds that develop in the fifth week. These include the primordial
endolymphatic sac and duct, the utricle and semicircular canals, and the saccule and cochlea. These relationships are important understand as the innervation of these portions of the inner ear are based on this embryological formation. The cochlear duct begins to grow from a primordial blood starting at the basal region spiraling apically to reach its full 2-1/2-2-3/4 turns. This generally starts occurring at the sixth week of gestation advancing around the eighth to 10th week. The Organ of Corti completes its formation at about the 25th week. The semicircular canals then form as 3 small folded evaginations on the primordial vestibular appendage. The centers compress and fuse to form the semicircular duct structures in the adult ear. These canals continued to enlarge and complete their formation starting with the superior, the posterior, and finally the lateral canal. The otic capsule develops from a cartilaginous precursor which ossifies during development. The cochleovestibular nerves and ganglia develop with the membranous labyrinth and cochleovestibular end organs. These ganglia are of neural crest origin and migrate between the epithelial basement membrane of the otic vesicle. (1)

**Goals of Cochlear Implantation**

In order to better understand how one is able to implant cochlear devices in those difficult ears, one must understand the principal goals of cochlear implantation. The principal cause of hearing loss in most people is damage to the sensory hair cells. This sever the connection to the central auditory system. The function of cochlear implant is to bypass the missing or damaged hair cells and stimulate directly the surviving neurons. Histological analysis has shown that peripheral neurons of a spinal ganglion cells generally undergo retrograde degeneration. However, the cell bodies of the spinal ganglion cells are much more robust and remain intact even years after hearing loss. (Wilson B) The cochlear implantation is used to excite the nodes around the in the spiral ganglion cells. There is question to how many surviving cells are left in the cochlea depending upon the pathology. Most of these have shown that the surviving cell counts very from location to location and from cochlea to cochlea. While the minimum number of auditory neurons necessary for speech recognition is unclear, most experts agree that it is minimal for speech recognition. Multiple histological analyses have been performed studying the ganglion cell count and various disorders. The normal cochlea has between 29,030 8000 and ganglion cells. Patients with labyrinthis ossificans can have between 6000-28,000 ganglion cells. Patients with Mondi deformity can have 7000-16,000. (2)

Although discussion of the surgical approach to cochlear implantation in the normal ear is beyond the scope of this lecture, cochlear implantation is generally undertaken through a standard transmastoid facial recess approach. The cochleostomy is generally performed anterior inferior to the round window, and the cochlear implant is inserted into the Scala Tympani. As will be discussed later, surgical approaches have to be altered based on the anatomy of various disorders.

**Cochleovestibular Malformation**

With the advent of high-resolution computed tomography, cochleovestibular malformations have been diagnosed more frequently, and newer classifications have been made to better evaluate and diagnose these disorders. There is much controversy in the literature, especially regarding the Mondi any
deformity. Jackler et al in 1987 proposal widely accepted classification wheeze today. However, in recent years this term has come to virtually mean any congenital malformation of the osseous labyrinth. Controversy still remains on the true definition of the Mondi deformity. Most experts would agree that the classic Mondi deformity is described as a cochlea with 1-1/2 turns, dilated vestibule, and enlarged vestibular aqueduct. Although cochleovestibular malformations are relatively rare in the general population, they are found in approximately 20% of all cases of congenital sensorineural hearing loss. There are multiple classification systems, and recently a group from Turkey redefined classifications scheme based on the understanding of relevant embryology. (3) The scheme is based on the chronological development of the ear and the various cochleovestibular malformations that occur on this chronological order. The Michel deformity generally occurs in the third week of gestation resulting in the absence of a cochleovestibular system. Cochlear Agenesis occurs later in week 3. The common cavity deformity generally occurs in the fourth week of gestation. In the six-week, the incomplete partition type 1 occurs. The classic Mondi deformity, or incomplete partition type II, occurs at the seventh week of gestation. (3) Generally, it is believed that a unilateral cochleovestibular malformation is secondary to a nongenetic cause while bilateral cochleovestibular malformations or often related to genetic association. There are several genetic associations with cochleovestibular abnormalities. These include fibroblast growth factor-3 and Nkx5 to 1 homeobox gene which is responsible for the development of the semicircular canals, and Pax-2 to which is responsible for the development of the cochlea.

Evaluation process for patients with any dysplasia or cochleovestibular malformation is generally started with a formal hearing evaluation and/or high-resolution computed tomography. Most of these inner ear dysplasia are easily identifiable on CT imaging. Again, up to 20% of patients with a congenital sensorineural hearing loss will have osseous abnormalities on CT imaging, and the percentage of patients require cochlear implant with an osseous dysplasia is higher than in the general population. (4) MRI is also useful in evaluating cochlear patency and the absence or presence of the cochlear nerve. T2 MRI imaging is often useful in evaluating the patency and septations within the cavity. Sagittal views of the internal auditory canal using CISS or Fiesta sequence can ascertain the absence or presence of the cochlear nerve.

Patients diagnosed with a congenital sensorineural hearing loss and cochleovestibular malformation are regarded as cochlear implant candidates. However, what contraindications can occur when implanting this subset of patients? What complications can occur with implanting patients with cochleovestibular malformations? There are several general considerations in the surgical management. The type of malformation generally dictates the surgical approach for implantation. With the exception of Michel's dysplasia, all cochleovestibular malformations can be implanted, and most can be approached with the standard trans-mastoid facial recess-posterior tympanostomy approach. The electrode choice can differ between the types of cochlear malformation. Many malformed cochleae do not have the neuroepithelium in normal location. This is the target for the cochlear implantation. With few exceptions, normal electrode arrays are used. After implantation, device programming can be more
challenging. Frequent adjustment in the program is necessary in these patients as stimulation of the facial nerve is more common. This can require deactivation of the electrodes in question. The performance, however, between a patient with a malformed cochlea and a patient with a normal cochlea is very similar. In a retrospective matched-pairs analysis by Eisenmann et al of 17 patients with malformed cochlea as compared with 17 patients with normal cochleae, the study revealed similar outcomes after 24 months. The only difference was that patients with malformations required a longer period of time to reach the end point. (5) Dr. Papsin evaluated 298 children who were implanted between January of 1992 to January of 2002. He grouped these children according to the deformity and looked at performance based on different closed and opened set speech scores. He found that patients with incomplete partition are more likely to have progressive onset of hearing loss as compared to other groups. He also found that these children consistently had higher scores on their speech scores. Dr. Papsin used both opened and closed set speech scores to evaluate the outcomes. He found that hypoplastic cochleae and common cavity deformities were related generally with worse performance than with patients with incomplete partition. He theorized that an anatomic and physiological basis was behind this decreased performance. In those patients, he theorized that there were fewer active electrodes to implant, a narrower dynamic range, impaired neural synchronicity, and the lack of general topical organization in the cochlea. (4)

The common cavity deformity generally lacks a central modiolus which is where the cochlear implant should be placed. Histological analysis shows that common cavity has rudimentary forms of known normal cochlear structures which are equivalent to the Organ of Corti and stria vascularis. It also shows that there is auditory neural tissue which lies peripherally in the common cavity. The surgical approach to cochlear implantation in most cochleovestibular malformations is similar to patients with normal cochlear anatomy. However, the common cavity deformity is often addressed in a separate technique. Surgery is undertaken with continuous facial nerve monitoring. A standard mastoidectomy with subperiosteal pockets is made for the receiver stimulator package, and exposure of the posterior surface the cavity is undertaken at the antrum with a drill. A cochleostomy is then performed at this point at the common cavity. Some authors recommend intraoperative imaging guidance during insertion in these patients. There is controversy on whether obliterating the external meatus or performing canal wall down mastoidectomy is necessary in these cases. Most authors implant these patients with a straight array with concentric band electrodes allowing for the outer wall to be maximally contacted. (6)

**Complications in Cochleovestibular Malformation**

Complications from cochlear implantation are relatively rare; however, there are increased risks with cochleovestibular malformations based on the altered anatomy. Often complications of cochlear implantation include: CSF leak, internal auditory canal implantation, and facial nerve complications. CSF leak is more common in these patients often due to lack of a bony cochlear lamina cribosa. This is often missing or replaced by a fibrous band. The defects in the modiolus and the lamina cribosa at the fundus of the internal auditory canal are often responsible for the mixing of perilymph and CSF. This then leads to an increased risk of CSF leak during cochlear implantation and afterwards. (6) While,
cavity malformations generally have the highest risk of CSF or perilymph leakage, the degree of dysplasia often does not correlate with the risk of CSF leakage. Management is generally started preoperatively by rating the risk through evaluation of the preoperative imaging. Contrary to common logic, a wider cochleostomy allows for more packing material to help decrease the chance of CSF leak. During surgery performing a Valsalva maneuver with the patient in head down and confirming cessation of leakage can aid in ensuring any leak is repaired. Rarely, in the management of CSF leak, a lumbar drain may need to be placed. With the risk of CSF leak, there is an increased risk of meningitis postoperatively. While all patients undergoing cochlear implantation should undergo preoperative pneumococcal vaccination, it is imperative that these patients receive their preoperative pneumococcal vaccination. Internal auditory canal insertion is also a risk of performing cochlear implantation in patients with cochleovestibular malformations. Again, the altered anatomy in these patients leads to a higher risk. This is often secondary to the absence of the cochlear lamina cribosa. Unrecognized insertion into the internal auditory canal can lead to CSF leak, vertigo, facial nerve stimulation, and suboptimal hearing outcomes. The most common anomaly for intermeatal electrode placement is the X-linked deafness associated cochlear anomaly. (6)

15% of patients with cochleovestibular anomalies have an aberrant facial nerve course. Typically, the nerve is anterior from the first genu down the promontory to the round window. Occasionally the nerve can be found split with one branch in its normal position and another branch in an abnormal position. Coelho and Rolando recommend drilling along the tegmen to the antrum and identifying the incus. Once incus is removed the facial nerve is generally identified along its horizontal segment. The authors have found that this is one of the more consistent landmarks for the facial nerve. They recommend using this procedure in all patients with CHARGE syndrome and Branchio-Oto-Renal syndrome. (6)

**Conclusions**

Cochleovestibular malformations have been increasing in their incidence secondary to an increased roll in high-resolution imaging. As stated before, up to 20% of patients with sensorineural hearing loss have an osseous abnormality. It is important to understand the anatomical and physiological aspects that separate these malformations from the normal cochlea especially in regards to goals of cochlear implantation. Also, one should understand the surgical anatomy and possible complications in undergoing these procedures. Good outcomes are achieved in patients with cochleovestibular malformations when successfully implanted.

**Labyrinthitis Ossificans**

Bacterial meningitis is the most common cause of postnatal profound hearing loss and children, with an incidence of 6-16% of the patients. 5 to 7 percent of patients with bacterial meningitis will go on to develop bilateral severe to profound hearing loss. (7) The bacterial infection often enters the cochlea from the subarachnoid space to the cochlear aqueduct. The cochlear aqueduct generally has failed to obliterate in these patients. The aqueduct runs through the petrous bone from the posterior fossa and
generally terminates laterally in the Scala Tympani close to the round window and the basal turn of the cochlea. The infection generally spreads to the cochlea rapidly and often within hours of diagnosis. Due to the anatomical relationship of the cochlear aqueduct, infections arise apically from the basal turn damaging intra-cochlear structures. These are generally profound and irreversible damage. Endostial inflammation causes new bone formation within the cochlear lumen. There is no predictable pattern for the amount of osteoneogenesis that occurs as there is a lot of variability from case to case. The osteoneogenesis often occurs rapidly however. Animal studies have shown that ossification can occur within 72 hours of the meningitis. Ossification can occur through multiple pathways. They can occur through meninoogenic, tympanogenic, or hematogenic processes. Each of these processes indirectly destroys spiral ganglion cells. Evaluation and diagnosis in these patients early on is critical. However, this is often difficult in these patients. These patients are often critical and require further stabilization before audiological and radiographic evaluation can occur. Once the patients are stabilized, audiological evaluation and amplification should be performed. Serial audiometric testing should be performed in these patients, and any deterioration to severe or profound sensorineural hearing loss should prompt consideration immediate implantation. This deterioration often reflects the progressive fibrosis and ossification that occurs after meningitis. Once initial diagnoses evaluation is undertaken, preoperative imaging is often required. High-resolution computed tomography is necessary to evaluate the bony anatomy of the temporal bone. T2-weighted MRI is more sensitive method of assessing intra-cochlear fluid signal and assessing for blockage. Also, using contrast enhanced T1 weighted MRI, imaging can detect early fibrosis in the cochlea. Labyrinthitis ossificans is generally graded by 3 grades. Grade 0 is a normal cochlea. Grade 1 ossification is generally sclerosis without any narrowing. Grade 2 ossification is sclerosis with narrowing, and a grade 3 classification is a complete or partial obliteration of the cochlea. (6) Young et al retrospectively evaluated high-resolution CT imaging and correlated with her surgical findings. They found that lateral canal ossification suggested further ossification of the cochlea in 70% of the cases which led to a 77% sensitivity, while ossification the cochlea on high-resolution CT imaging only identified surgical ossification at the basal turn in 45% of the patients with the sensitivity of only 50%. (8) Isaacson et al presented to series of 45 children with post-meningitic hearing loss and evaluated the accuracy of MRI for predicting intraoperative cochlear obstruction. They found that the sensitivity for MRI in these patients reached 94% while the specificity was 87.5% leading to a positive predicted value of 94.1%. (7)

**Surgical Technique**

As described earlier, the ossification of the cochlea inflicts damage to both the cochlear duct and to the spiral ganglion cells. As with normal cochlear implant insertion, full electrode insertion into the Scala Tympani is the ultimate goal with electrode activation of the spiral ganglion cells and the cochlea. With labyrinthitis ossificans, there is an increased difficulty in inserting a full complement of electrodes. The question is what to do when this is not possible. In most instances of limited ossification, drilling can continue along the basal turn until a lumen is identified. When there is osteoid and fibrous tissue in the duct, this can be removed with picks, rasps, or drills. If a patent lumen is found before or at the descending turn of the cochlea, a full insertion is performed. Most authors believe in the judicious use
of a test electrode or depth gauge to aid with insertion. If the entire basal turn is ossified, a basal turn drill-out can be performed. This is described as using the 1 mm diamond drill, drilling in the anterior medial direction for no more than 9.5 mm or when the external auditory canal is visualized. (6) This generally allows for the placement of enough electrodes to get multichannel performance in the patient. If the standard cochleostomy reveals obstruction even after drill-out to the pars ascendus, then drilling can be extended superiorly posterior to the oval window where the Scala Vestibuli is then identified. The cochleostomy is enlarged and filled with glycerin which clears the blood and bone debris. The electrode is then inserted into the Scala Vestibuli and the cochleostomy is packed. Lin et al reviewed patients who underwent Scala Vestibuli insertion and found all of their patients improved communication. (9)

Total ossification the cochlea can be difficult to manage. There are several described approaches in these patients. Cohen and Waltman have described a short insertion tunnel technique for insertion. Gantz has described a total drill-out technique. Balkany et al have described a technique that combines an inferior trough along with an open drill-out technique which allows for fixation points and more electrodes to be utilized in these patients. A standard trans-mastoid approach along with a tympanomeatal flap is undertaken. The incus and malleus are then removed allowing for visualization of the round window, the stapes superstructure, the cochleariform process, and oval window. This allows for the identification of the relationship of the basal turn of the cochlea with the surrounding structures. Through the facial recess, an 8 millimeter long tunnel is then drilled out from the cochleostomy. This is followed anteriorly with a 1.5 mm diamond drill. Drilling is discontinued if any bleeding occurs. A 1 mm diamond drill is then used as the tunnel is unroofed at approximately 4 mm. The abnormal bone of the basal turn is then followed around the modiolus was using the carotid canal, cochleariform process, and oval window as landmarks. The electrode is then passed through the facial recess, into the tunnel, and into the trough. Further fixation of electrodes occurs in the split incus bridge and with bone wedges in the trough. (10)

Double or split array electrodes in cochlear implantation have been developed based on the positive correlation between the number of electrodes implanted and speech recognition scores. These are generally performed with 2 cochleostomies. A basal array is placed in the inferior basal-turned tunnel. The fossa incudis, incus, and stapes superstructure are then removed. A second cochleostomy is then placed in the in the second turn by drilling adjacent to the anterior oval window ligament, and the array is inserted there. (6)

Performance

A study from London looked to evaluate the speech performance and matched groups with ossified cochlea as compared to non-ossified copious. Using the Manchester spoken language development scale (MSLDS), and the categories auditory performance scale (CAPS), the group was able to perform performance studies. They were able to find the speech performance was very similar using both scales in comparison of ossified versus non-ossified cochleae. The study was able to come up with
an algorithm for the surgical management of ossification. After evaluation of the preoperative imaging, attempts at Scala Tympani cochleostomy are performed. If obstruction is encountered, the basal turn was then explored to the pars ascendus. If a patent lumen is found, a test electrode is then placed. If it is successful, a cochlear implant is inserted into the Scala Tympani. However, if the test electrode is unsuccessfully inserted, the surgeons recommend an attempted insertion into the Scala Vestibuli. If exploration of the basal turn is unable to find a patent lumen, a cochleostomy is drilled to evaluate the patency of the Scala Vestibuli. If the Scala Vestibuli is patent, insertion of the cochlear implant is placed through the Scala Vestibuli. If the Scala Vestibuli is obstructed, the authors recommend several options including a double array, compressed array, basal-turn-only insertion, or total drill-out as described above. (11)

**Conclusions**

Bacterial meningitis can lead to a severe nonreversible hearing loss. It is the most common cause of postnatal hearing loss. It can also lead to rapid irreversible changes in the cochlea which can then lead to further difficulty if implantation is required. A thorough investigation of the cochlear anatomy using high-resolution imaging including CT and MRI and is important in the workup these patients. Again, understanding the anatomy, different surgical options and algorithm is important in the surgical management of these patients.

**Otosclerosis**

Otosclerosis is a disorder of bone reabsorption of the petrous bone followed by replacement with thick irregular sclerotic bone. This is most commonly found at the location around the oval window referred to as fenestral otosclerosis. 10% of patients with otosclerosis will have foci affecting the otic capsule. (12) This was originally referred to as retro-fenestral otosclerosis. Originally described by House and Sheehy, Far Advanced Otosclerosis is a severe mixed hearing loss. This definition has been changed in recent years to advanced otosclerosis. This includes patients with a sensorineural hearing loss and a diminished speech discrimination score. The mainstay of treatment for patients with otosclerosis remains a stapedectomy. This is a relatively simple and an inexpensive procedure for patients. Even in patients with advanced otosclerosis with a mixed hearing loss, a surgical correction of the conductive component can be effective. However, in severe mixed hearing loss, a stapedectomy is often unsatisfactory for the hearing goals of the patient. Treatment with a cochlear implant results in excellent hearing in these patients. However, it is expensive and requires an experienced surgeon.

**Grading**

There multiple grading systems available to classify otosclerosis. These are based on computed tomography imaging. The Rotteveel classification system is a 3 grade system. Grade 1 otosclerosis is solely fenestral. Grade 2a otosclerosis is a retro-fenestral otosclerosis which presents as a double ring or halo effect on imaging. Grade 2b is defined as a narrowed basal turn, and Grade 2c is defined as both retro-fenestral and a narrowed basal turn. Grade 3 otosclerosis is a diffuse confluent retro-fenestral involvement. (13)
Surgical Considerations

Considerations in cochlear implantation in patients with otosclerosis generally include the fenestral and basal turn ossification. Often these patients need extra drilling, and many only have partial electrode insertion. One must also be prepared to perform a Scala Vestibuli insertion. 80-100% of patients with basal turn ossification will have a partial electrode insertion or misplacement during surgery. Often, the changes associated with otosclerosis can lead to a false passage where the cochlear implant is inserted into a false lumen. Also, obliteration of the Scala Tympani will require Scala Vestibuli insertion in up to 25% of cases. (12)

Complications

A large review from the Netherlands looked at the outcomes of cochlear implantation in patients with otosclerosis. Surgical difficulties and complications from multiple studies were collated. Anywhere from 0-75% of patients had facial nerve stimulation after cochlear implantation. Between 2-25% the patients had insertion into the Scala Vestibuli. Extra drilling was required and 13-60% of patients. (12)

Advanced Otosclerosis Algorithm

Merkus et al have described an algorithm for the management of advanced otosclerosis. This is based on the maximum speech discrimination scores for the patient, the computed tomography grading classification, and the air-bone gap in the audiogram. Patients with a maximum described speech discrimination score less than 30% should undergo cochlear implantation. The patient with a maximum speech discrimination score between 30-50% should be evaluated further CT scan findings. The patients with a CT scan grade of type 2c or 3 should undergo cochlear implantation. Those patients that do not have the CT findings but have an air-bone gap greater than 30 dB should undergo stapedectomy. Those patients that do not have an air-bone gap greater than 30 dB consider cochlear implantation. In the patient with a maximum speech discrimination score of 50% to 70%, CT imaging should be evaluated. In a patient with a CT scan showing a type 2c or 3, the patient should undergo cochlear implantation. If the CT scan does not show these findings and the patient has an air-bone gap greater than 30 dB, the patient should undergo stapedectomy. Inversely, if the air-bone gap is less than 30 dB, the patient should undergo hearing amplification with close follow. (12)

Performance

Performance scores in compiled by Merkus et al found a 100% increase in hearing after cochlear implantation through all studies. Word discrimination scores after surgery were found to be anywhere from 45% to 98%. (12)

Otosclerosis Conclusions

Cochlear implantation should be considered in patients with advanced otosclerosis. Using the algorithm described above, patients should be offered a cochlear implantation. Performance improvement had been found in all studies. Complication rates from otosclerosis cochlear implantation
are similar to those in patients with normal cochlear anatomy. It is important to understand the approach and pitfalls in implantation in the background of retro-fenestral otosclerosis. (12)

Conclusions

The role of cochlear implantation and improving the hearing of our patients is increasing as we understand the patient populations that would benefit best. Studies have shown that patients with cochleovestibular malformations, labyrinthitis ossificans, and otosclerosis should be added to this list. The anatomy and physiological process for each of these creates for more difficult surgical approach and can lead complications that one must be aware. The only absolute contraindications to implantation of a cochlear implant are the absence of the cochlea or the cochlear nerve. In general, incomplete partition implant recipients generally do the best of those with cochleovestibular malformations. Both CT and MRI are being recommended as imaging modalities in patients with cochleovestibular anomalies as well as those with a history of bacterial meningitis. T2-weighted MRI is sensitive that detecting intracochlear fluid which can be helpful in evaluating for the patency and the septations in the cochlea. While the mainstay of treatment for patients with otosclerosis continues to be the stapedotomy, cochlear implantation may service important adjunct in the treatment of the hearing loss in patients with advanced otosclerosis.

Bibliography


