Cochlear Implantation of the Common Cavity Malformation may be Performed During or Before CSF Leak Repair

Benjamin R. Roman, MD1, Daniel H. Coelho, MD2, J. Thomas Roland, Jr., MD1

1Department of Otolaryngology, Head and Neck Surgery, New York University School of Medicine, New York, NY.
2Department of Otolaryngology, MCV/Virginia Commonwealth University School of Medicine, Richmond, VA USA

INTRODUCTION

- Congenital malformations of the inner ear result in sensorineural hearing loss due to an abnormal membranous or membranous and bony labyrinth.
- When osseous cochlear abnormalities exist, patients are at increased risk of cerebrospinal fluid (CSF) leak and meningitis.
- Certain malformations are especially prone to these complications, including the common cavity malformation.1,5 (TABLE 1) Aggressive management of CSF leaks is necessary to prevent meningitis in these cases.
- Traditional approaches have used the patient’s own tissue to seal the defect or the final pathway to the middle ear.
- However, these approaches may preclude, or at best significantly interfere with later cochlear implantation of an already distorted cochlea.
- We present a case in which a cochlear implant was used as part of the material to seal a CSF leak, and suggest a paradigm shift in the management of these patients.

CASE REPORT

- A 2 year old female with congenital deafness and bilateral common cavity malformations had previously undergone cochlear implantation on the left side at 16 months.
- During surgery, it had been noted that there was CSF pulsation through the cochleostomy, which was securely plugged with the patient’s peristeum after electrode insertion. Bilateral PE tubes were placed. Thereafter she was beginning to develop oral language.
- Five months later she was treated for Streptococcus pneumoniae meningitis at an outside hospital. No evidence of otitis media or CSF leak was seen. She received a full course of IV antibiotics and recovered without sequelae.
- Five months after being treated for meningitis, the patient again presented with clinical signs of meningitis, confirmed by CSF culture to be due to Haemophilus influenzae, non type B.
- Examination revealed a serous effusion in the left middle ear, and some fluid present in the right middle ear. Within 48 hours it became apparent that there was a CSF leak with clear fluid draining from the right PE tube.
- Temporal bone CT scan confirmed a properly placed CI electrode in the left ear, an effusion consistent with otitis media on the left, and also raised suspicion that there was a bony defect anterior to the staples footplate on the right. (FIGURE 1)
- The decision was made to proceed with CSF leak repair and simultaneous cochlear implantation.
- In the OR, a standard mastoidectomy and facial recess approach to the right middle ear was used. Findings correlated with the CT scan, with an arachnoid bleb and active CSF leakage seen in an area of dehiscence in the anterior half of the oval window footplate. (FIGURE 2)
- Insertion of the electrode was accomplished with a cochleostomy on the inferior aspect of the common cavity (there was no discernable round window) and the use of fluoroscopy to ensure a single turn placement without intrameatal cannulation or electrode kinking or bending.7 The cochleostomy was tightly packed with peristeum.
- The staples footplate was removed and the CSF leak was repaired with peristeum packing. The vestibule was tightly packed, and was followed by muscle medial to the malleus to hold pressure on the defect.
- The patient did well and was discharged on post-operative day #3 without evidence of otitis media or CSF leak bilaterally. She completed a total of 2 weeks of appropriate antibiotics via a peripherally inserted central cathether (PICC).
- Of note, she had received all appropriate vaccinations in a timely fashion, including all doses of Hib vaccine and all four doses of the pneumococcal conjugate vaccine (PCV-Prevnar 7) before the first episode of meningitis. The pneumococcal polysaccharide vaccine (PPV Pneumovax 23) was administered during this hospitalization.
- One year postoperatively, she has a mild hearing loss on the left and a mild to moderate hearing loss on the right. She is developing language, following simple directions, responding to her name and environmental sounds, and imitating names, common words and phrases. She has not shown any evidence of CSF leak or meningitis, despite several episodes of otitis media that were treated in the routine manner.

DISCUSSION

- A CSF leak can lead to life-threatening meningitis and must be surgically repaired.
- Traditionally, the areas of communication between the subarachnoid space and the middle ear are identified, and the inner ear and defect are packed with muscle and fascia. Occasionally, total petrosectomy and middle ear obliteration with abdominal fat grafts may be necessary.
- Such procedures make later cochlear implantation difficult or impossible, even in experienced hands.3,4
- These measures were not necessary for the case reported here. Given the language benefit she derived from her first implant on the left, and the potential for further benefit on the right, the decision was made to perform a cochlear implantation at the time of the CSF leak repair on the right. Cochlear implantation at a later date might not have been possible due to fibrosis and scarring caused by the repair.
- Post-implantation intracochlear/intracavity fibrosis may be responsible for sealing off any potential sites of either CSF egress or bacterial ingress.
- Awareness of the risk of meningitis in children with cochlear implants has grown over the last decade.8,9 In response, the CDC has developed guidelines for vaccination against Streptococcus pneumoniae and Haemophilus influenza type B, including use of the 7-valent 23-valent pneumococcal polysaccharide vaccine (PPV23) (Pneumovax pneumococcal conjugate vaccine (PCV7) (Prevnar7), and the @, as well as the Hib vaccine.10
- However, proper immunization does not fully eliminate the risk of meningitis, as demonstrated in this case. Children under two years old who have not had the 23-valent vaccine are still susceptible to meningitis. Children under two years old who have not had the 23-valent vaccine are still susceptible to meningitis, as evidenced in this case by the development of non type B Haemophilus influenza.
- In children who have inner ear malformations that can lead to CSF leaks, the additional risk of meningitis that vaccination strategies apply even more rigorously, both before and after implantation.
- In addition to immunization awareness, changes in devices and surgical techniques have resulted in a decreased incidence of meningitis associated with cochlear implantation over the last decade.8,9

CONCLUSIONS

- We argue here that children with inner ear malformations leading to CSF leaks may require additional surgical modifications to prevent meningitis.
- For those candidates without a CSF leak or meningitis, we suggest that potential sites of CSF leak be closed during cochlear implantation surgery.
- Furthermore, we suggest that earlier cochlear implantation may be warranted in these children in an attempt to prevent meningitis before it occurs while simultaneously aiding hearing.

REFERENCES

4. Reefhuis J, Honein MA, Whitney CG, et al. Risk of bacterial meningitis in children with cochlear implants has grown over the last decade.8,9 In response, the CDC has developed guidelines for vaccination against Streptococcus pneumoniae and Haemophilus influenza type B, including use of the 7-valent 23-valent pneumococcal polysaccharide vaccine (PPV23) (Pneumovax pneumococcal conjugate vaccine (PCV7) (Prevnar7), and the @, as well as the Hib vaccine.10
8. Reefhuis J, Honein MA, Whitney CG, et al. Risk of bacterial meningitis in children with cochlear implants has grown over the last decade.8,9 In response, the CDC has developed guidelines for vaccination against Streptococcus pneumoniae and Haemophilus influenza type B, including use of the 7-valent 23-valent pneumococcal polysaccharide vaccine (PPV23) (Pneumovax pneumococcal conjugate vaccine (PCV7) (Prevnar7), and the @, as well as the Hib vaccine.10