The Management of Spontaneous Otogenic CSF Leaks: A Presentation of Cases and Review of Literature

Meghan N. Wilson¹ Lawrence M. Simon¹ Moises A. Arriaga¹ Daniel W. Nuss¹ James A. Lin²

¹Department of Otolaryngology Head and Neck Surgery, Louisiana State University, New Orleans, Louisiana, United States

²Department of Otolaryngology Head and Neck Surgery, University of Kansas, Kansas City, Kansas, United States

Address for correspondence Meghan Wilson, MD, Department of Otolaryngology, 533 Bolivar Street, Suite 566, New Orleans, LA 70112, United States (e-mail: mwils5@lsuhsc.edu).

J Neurol Surg B 2014;75:117–124.

Abstract

Objective The types of otogenic cerebrospinal fluid (CSF) fistulae were previously classified into defects through, adjacent to, or distal to the otic capsule. This article presents cases of the three different types of spontaneous CSF fistulae and reviews pertinent literature. We examine the management of the different types of otogenic CSF leaks with modern audiovestibular testing, imaging, and surgical techniques.

Design Case series and review of the literature.

Setting Academic tertiary neurotologic referral practice.

Participants Four patients identified through a retrospective search.

Main outcome measures Resolution of CSF leak and absence of meningitis.

Results Surgical intervention was performed on the four cases described in this series; none had a return of CSF otorrhea in the postoperative period or meningitis.

Conclusions Otogenic CSF fistulae may lead to life-threatening infection and in congenital forms are typically not diagnosed unless meningitis has occurred. Rapid and proper recognition, work-up, and treatment of such leaks decrease the risk of permanent neurologic sequelae as well as recurrent meningitis.

Keywords

- CSF fistula
- CSF otorrhea
- cochlear malformation

Introduction

Spontaneous otogenic cerebrospinal fluid (CSF) leak is a potentially life-threatening problem because meningitis risk is relatively high. Otogenic cerebrospinal fistulae may be challenging to identify, particularly if a patient has not been afflicted with meningitis. Congenital deformities of the temporal bone include an anatomical connection from the subarachnoid space to the middle ear leading to a physiologic CSF leak. Such leaks may present as childhood meningitis with or without unilateral serous otitis media or drainage through a perforated eardrum or tympanostomy tube. Adult patients, more commonly, may develop spontaneous otogenic CSF leaks through the middle or posterior fossa dural

plates and may be diagnosed with a high index of suspicion before a bout of intracranial infection.

The types of CSF fistulae were previously classified by Neely into defects through, adjacent to, or distal to the otic capsule.¹ The management of each type of CSF fistula varies and is affected by factors such as the functional status of the ear, age-related limitations, and potential for concomitant anatomical anomalies. Those type 1 leaks that occur through the inner ear most often occur through a non- or poor-functioning inner ear.² A poorly functioning ear allows for decisive and aggressive obliteration of the middle, external, and potentially inner ear, especially in the setting of life-threatening infection. Perhaps the rarest CSF leaks are the type 2 leaks that occur near or adjacent to an anatomically

received

June 16, 2013

accepted

September 17, 2013

published online

December 11, 2013

© 2014 Georg Thieme Verlag KG
Stuttgart · New York

DOI <http://dx.doi.org/10.1055/s-0033-1359304>.
ISSN 2193-6331.

and functionally normal inner ear through a congenital dehiscence. Diagnosis is often difficult as is control of these type 2 leaks if preservation of hearing and normal anatomy is a goal. Type 3 leaks that are typically adult in onset are likely acquired and in the setting of benign intracranial hypertension.³ Because adults have mature immune systems and usually are more descriptive and likely to seek attention for otologic symptoms than children, these type 3 leaks may be diagnosed before an episode of intracranial infection. The diffuse nature of intracranial hypertension along the surfaces of the temporal bone may be why such adult-onset leaks tend to be multiple. Repair of a type 3 spontaneous otogenic CSF leak requires not only wide radiographic and surgical examination of potential defect locations, but also CSF reduction, shunting, and/or diversion as a possible adjunctive therapy.

Case Studies

Case 1

A 9-month-old male infant with a history of bilateral myringotomy and tubes for “chronic serous otitis media” at 4 months of age presented to the emergency department with fever, lethargy, and drainage from the right ear. His auditory brainstem response (ABR) study performed shortly after birth demonstrated a profound sensorineural hearing loss on the right with normal hearing on the left. After he had his tubes placed, he had no drainage from either ear until 3 days before admission when he developed profuse, watery right otorrhea that worsened with crying. Within a day of otorrhea onset, he developed fever and lethargy and was seen by his pediatrician who initiated ofloxacin and Augmentin. Despite the treatment, his condition deteriorated with nausea and vomiting, and he was brought to the emergency department where he was noted to have a temperature of 103°F, a white blood cell count of 11,700, an erythrocyte sedimentation rate of 113 mm/hour, and a C-reactive protein level of 139 mg/L. He was lethargic, and examination of the left ear showed a patent tympanostomy tube, clear tympanic membrane, and no drainage. Examination of the right ear showed copious clear fluid draining from the patent tympanostomy tube with increased drainage during crying spells.

Computed tomography (CT) scan demonstrated a cochlear malformation on the right consistent with an incomplete partition according to the classification system by Jackler, Luxford, and House.⁴ His right inner consisted only of a basal turn, a dilated vestibule, and a bony channel linking the basal turn through the inferior petrous apex and clivus to the nasopharynx (► Fig. 1). There was no appreciable rhinorrhea; therefore, the bony connection with the nasopharynx was not considered a physiologic fistula. Additionally, the superior and posterior semicircular canals appeared to be one large fused canal rather than two separate canals. No abnormalities were seen on the left. Magnetic resonance imaging (MRI) was obtained as well that showed no intracranial abnormalities and no evidence of encephalocele.

Lumbar puncture was performed and turbid fluid was obtained. The patient was treated with 14 days of intravenous (IV) vancomycin and ceftriaxone. The patient’s lethargy,

emesis, and fevers resolved soon after initiation of antibiotics. The drainage from his right ear also ceased. The final culture from the lumbar puncture grew coagulase-negative staphylococcus.

On hospital day 18, the patient was taken to the operating room for ear canal closure and middle ear and Eustachian tube obliteration because he had poor hearing and was believed to be at risk for recurrent meningitis despite cessation of the leak. A modified posterior auricular incision was made to avoid traumatizing his facial nerve at the mastoid tip, and the ear canal was transected at the bony cartilaginous junction. The ear canal skin was removed, and the middle ear was inspected. Given the patient’s age and size, a formal canalplasty was not performed because his bony tympanic ring was incompletely developed inferiorly. He was noted to have thickened mucosa in the middle ear, but no active CSF leak was identified. Although the promontory was identified, his incudostapedial joint was not readily visible. After removing all of the middle ear mucosa and external auditory canal skin, the eustachian tube was closed with Surgicel (Ethicon, Inc., Somerville, NJ) and bone wax. The middle ear was packed with bone wax, and then a piece of temporalis muscle was placed in the bony external auditory canal (EAC). The lateral EAC was closed in two layers. Given that the eustachian tube is the “final common pathway” for potential CSF otorrhorrhea, we opted not to open his mastoid because the surgical result is isolation of the middle ear and mastoid from aerodigestive tract contamination. The patient was observed for 2 days prior to discharge and had no recurrence of fever and no evidence of leakage.

The patient was seen in the clinic 2 days after discharge for a fever of 103°F at home. The patient was sent to the emergency department at that time for further evaluation. He had no evidence of CSF drainage, and his repeat lumbar puncture was not consistent with bacterial meningitis. He was treated with 10 more days of IV antibiotics and his fevers resolved. There have been no more bouts of meningitis or recurrent CSF otorrhea or rhinorrhea at least 19 months after his surgery. In light of his bacterial meningitis history, we closely followed his contralateral, ear, and speech development which has been progressing normally as of last follow-up.

Case 2

An 8-year-old girl with a history of chromosomal deletions compatible with life and developmental delay presented to the emergency department with fever and lethargy. As in our first case, the patient was noted at birth and through several ABRs test to have profound sensorineural hearing loss on the right side with moderate hearing loss on the left side. Also, she had a distant history of tympanostomy tube placement followed by recent onset right-sided otorrhea.

Prior to examination by our service, she had a CT scan and a lumbar puncture consistent with bacterial meningitis. Of note, her examination did reveal her to have bilateral mild microtia (type 1) with very small ear canals⁵ with some drainage from her right ear canal. She was very noncompliant with examination. Her CT scan demonstrated an inner ear consistent with cochlear hypoplasia according to the

Fig. 1 (A–D) Axial and (E, F) coronal noncontrasted computed tomography of case 1, showing an abnormal right cochlea consisting of only a basal turn (arrow).

Jackler classification⁴ (→ **Fig. 2**). Her malformation included a right mega vestibule, dysplastic semicircular canals, and a laterally placed vertical facial nerve (→ **Fig. 3**). Treatment was initiated with IV vancomycin and ceftriaxone. On hospital day 2, the patient developed profuse clear drainage from the right ear, and both neurosurgery and otolaryngology were consulted. Neurosurgery placed a lumbar drain for presumptive CSF leak leading to resolution of the right-sided otorrhea; however, ~ 24 hours after placement of the lumbar drain, the patient had an acute change in mental status and increased irritability. A head CT scan was immediately performed and demonstrated extensive pneumocephalus (→ **Fig. 4**), and the lumbar drain was subsequently removed. The patient developed recurrent otorrhea from the right ear after drain removal, and we made the decision with neurosurgery to surgically obliterate the CSF fistula given the failure of conservative measures.

We initially considered a modified Rambo-type closure of the ear canal given her laterally displaced facial nerve⁶ but opted to instead perform a traditional ear canal closure by cleanly transecting the ear canal lateral to the bony cartilaginous junction, separating the lateral ear canal skin from the ear canal cartilage, and everting the skin laterally with closure of a deeper musculo-periosteal layer as described elsewhere.⁷ After ear canal closure, the medial ear canal skin and tympanic membrane were removed as were the incus followed by the malleus. A CSF leak through a fixed stapes footplate was identified. The fixed stapes suprastructure was an impediment to firm packing of the oval window; therefore, it was removed and a CSF gusher was encountered. The vestibule was then packed with a large piece of temporalis fascia and allowed to reexpand within the vestibule with its edges tucked behind the medial rim of the oval window. This slowed the leak considerably allowing for bone wax to be

Fig. 2 (A) Axial and (B, C) coronal noncontrast computed tomography scan of case 2, showing an poorly developed right cochlea (arrow) with only one and a half turns and opacification of the mastoid air cells on the right side.

sculpted over and around the round window completely sealing it off. A prolonged Valsalva was performed to ensure control of the fistula. Subsequently, the eustachian tube was occluded with Surgicel and bone wax, and the middle ear was packed with bone wax. Postoperatively, the patient was observed in the intensive care unit for 2 days; a lumbar drain was not used. She had no recurrence of fever and no evidence of CSF leak in the follow-up period. Facial nerve monitoring was used throughout the entire case without incident, and facial nerve function remained intact postoperatively. Postoperative ABR contradicted previous outside ABRs, revealing normal hearing on the contralateral left side. She has had no recurrence of meningitis in at least 9 months since surgical repair.

Fig. 4 Noncontrast axial computed tomography scan of case 2 showing extensive pneumocephalus that developed after placement of a lumbar drain.

Case 3

A 12-year-old girl with a prior history of pneumococcal meningitis presented to the emergency department complaining of fever, neck pain, emesis, and dizziness that began abruptly that morning. She had no history of recent upper respiratory illness and no history of acute otitis media or otorrhea, although she did have left ear fullness. She was afebrile in the emergency department but she had a white blood cell count of 16.5K with a left shift. Lumbar puncture yielded an opening pressure of 42 mm H₂O, and turbid fluid was obtained. She was admitted and treated immediately with IV vancomycin and ceftriaxone. For further work-up of her recurrent meningitis, a CT scan and MRI were performed and showed a hypoplastic left mastoid with partial opacification of the mastoid air cells (►Fig. 5). Inner ears were grossly normal with no obvious bony dehiscence through the temporal bones from subarachnoid space to mucosal surfaces. Final culture from the recent lumbar puncture also grew *Streptococcus pneumoniae*. Her meningitis symptoms resolved after 10 days of IV antibiotics. She was evaluated by our service after discharge, and she had normal eardrums, cranial nerves, tuning forks examination, and audiogram.

Fig. 3 (A–C) Coronal computed tomography scan showing laterally displaced facial nerve (arrows) of the patient in case 2.

Fig. 5 Computed tomography scan of case 3 demonstrating decrease aeration of the left mastoid and no major abnormality of the inner ear or middle ear.

Although the CT scan and MRI did not show a location of CSF leakage, we believed that otogenic CSF fistulae was responsible for the recurrent meningitis, with the left ear as the offending side given the partial left mastoid opacification and fullness. In an attempt to prevent future bouts of meningitis, the patient was taken to the operating room for exploration of the mastoid and middle ear and patching of potential fistula(e). On making the postauricular incision and the vascular strip, clear fluid percolated through a pneumatized posterior bony ear canal. Suspecting a tegmen or posterior fossa arachnoid granulation, complete mastoidectomy and exposure of the epitympanum was performed but revealed no potential sites of CSF leak. On Valsalva, fluid was noted to accumulate from the middle ear, although there were no areas of exposed dural surfaces noted. The oval and round windows were inspected and noted to be clear of leakage; however, fluid was seen coming from air cells in the hypotympanum, anterior to the area of the jugular bulb, consistent with a persistent Hyrtl (tympanomeningeal) fissure. The hypotympanic air cell trabeculae were curetted away to allow a more regular surface to pack. The area of leakage was then filled with temporalis muscle and fibrillar Surgicel (Ethicon, Inc., Somerville, NJ). A layer of hydroxyapatite cement was placed and then additional temporalis muscle. There was no evidence of spinal fluid at the conclusion with a Valsalva maneuver, but to divert CSF away from the closure site, lumbar drainage was performed for 3 days and the patient was discharged home on postoperative day 4 with no evidence of recurrent CSF leakage. Close examination of the patient's preoperative CT scan does reveal a potential bony dehiscence along the area of Hyrtl fissure that proved to be physiologically patent (►Fig. 6). The patient returned 7 months later with ipsilateral otitis externa for which she was readmitted given her past history, but this was the sole adverse event 12 months after her surgery.

Case 4

A 52-year-old obese woman with history of multiple sets of tympanostomy tubes for chronic otitis media and left-sided conductive hearing loss presented to our otolaryngology clinic. Although she had no tubes in place on our initial

Fig. 6 Axial computed tomography scan at high resolution demonstrating persistent Hyrtl fissure (arrows) allowing communication between posterior fossa and middle ear space.

evaluation, she did note profuse nocturnal left otorrhea while she had her last left tube. The patient reported no rhinorrhea or abnormal taste, and no history of meningitis. On our examination, we did note a left serous effusion. Because of her history suspicious for CSF otorrhea, a CT scan of the temporal bones was ordered demonstrating evidence of tegmen mastoideum dehiscence (►Fig. 7). An MRI was also ordered that did not reveal evidence of encephalocele. With her history and radiographic evidence of cranial dehiscence, we performed a tympanocentesis to sample the effusion for β -2 transferrin with perioperative systemic antibiotics. The results were positive, confirming a left-sided otogenic CSF fistula.

Although the patient described no history of meningitis, her options of observation versus repair of the leak were presented to her to decrease the potential for intracranial infection. She opted for repair, and a middle cranial fossa approach was performed to explore and repair the entire tegmen given the potential for multiple leak sites.^{8,9} The patient was noted to have a tegmen defect and, despite an apparently negative MRI, a small encephalocele. The defect was repaired with DuraMatrix and DuraSeal. Postoperatively, a lumbar drain was left in place for 3 days. The patient has had no evidence of CSF drainage at least 5 months after the surgical repair.

Discussion

We have presented four cases of spontaneous otogenic CSF leaks with each category as described by Neely in 1985.¹ Our first two cases are representative type 1 leaks that occurred through nonfunctioning deformed inner ears; our third case represented a type 2 leak that occurred adjacent to a normal inner ear through a Hyrtl fissure. Our fourth case was an adult-onset type 3 leak that occurred away from the inner ear through a tegmen mastoideum dehiscence.

Fig. 7 (A, B) Coronal noncontrasted computed tomography scan showing tegmen dehiscence (arrows) and underlying mastoid opacification.

Type 1 leaks have been the subject of debate in the past, particularly with the eponymous Mondini defect. Streeter previously described the details of inner ear embryology.¹⁰ Inner ear development begins during the 4th week of gestation and continues until approximately gestational week 25. The membranous cochlea begins to form at day 22 as the otic placode, which then invaginates by day 30. The bony and membranous cochlea coils from base to apex between weeks 5 and 25. The mesenchyme turns into cartilage, and then ossification begins at week 18.

Cock, in 1838,¹¹ was the first to describe the association of meningitis with a direct communication between the internal auditory meatus and a deformed inner ear. This association was further defined by Phelps and Michels.¹² Ohlms et al describes the 39 cases of CSF otorrhea and inner ear malformation published up to 1989.¹³ Of these patients, 60% were < 4 years of age; 75% were < 10 years. All required a surgical procedure to correct the problems, and 30% required two or more surgeries. Phelps et al² described 20 patients with congenital malformations; 8 of these patients had unilateral involvement, and 12 had bilateral malformations. Patients with bilateral malformations were noted to have an increased incidence of meningitis over those with unilateral anomalies. The development of CT and MRI scans in more recent years has allowed a more detailed and specific classification of inner ear malformations and has provided a clearer understanding of the pathways in which spontaneous CSF fistula develops.

Malformations of the inner ear can lead to close associations or communications between the perilymphatic spaces and the intracranial subarachnoid space. Schuknecht¹⁴ suggests in his study that the most likely route for CSF leakage in malformed ears is through the lamina cribrosa, noting that affected ears exhibited very thin bone between the cochlea and internal auditory meatus. Normally, the subarachnoid space extends into the internal auditory canal as far as the lateral fundus where it is separated from the perilymph by the bony lamina cribrosa. The most common pathway for the CSF to enter the middle ear is through the oval window via a

dehiscence in the stapes footplate, as described here in case 2.^{14–16}

The classic, or true “Mondini” malformation, described in 1791,¹⁷ is a cochlea with only 1.5 turns with a normal basal turn and dilated apical sac. Some hearing is usually preserved in a true Mondini deformity. Although controversial, studies have shown no increased risk of CSF leak and meningitis in patients with a true Mondini deformity than the general population.^{14,17,18} Phelps et al strenuously argued that the Mondini classification was being used as a blanket term encompassing a wide range of inner ear malformations, thus falsely elevating the risk of meningitis with the so-called Mondini deformity.¹² Agreeing with Schuknecht, Phelps and Lloyd described an increased likelihood of CSF otorrhea and meningitis in those patients with “a dilated basal turn, often associated with a tapering internal auditory meatus, and primary anacusis.”¹⁸ Our case reports on patients 1 and 2 clearly demonstrate the scenario described by Phelps et al with abnormal basal turns, tapering internal auditory canals, and presumed primary congenital deafness on the affected side.

Work-up of any child with an acquired or congenital sensorineural hearing loss in one or both ears should, at the very least eventually include a high-resolution scan to examine inner ear morphology, although the timing of such a study is debatable. In the case of a documented acquired hearing loss, we argue that a contrasted dedicated internal auditory canal MRI with additional high-resolution T2 sequences is preferred to identify any central lesions (i.e., tumors) as well as to examine the morphology of the inner ear to determine if the patient may have an enlarged vestibular aqueduct or a meningitis-prone cochlear deformity. In a congenital sensorineural hearing loss, either a temporal bone CT scan or a dedicated internal auditory canal MRI may be ordered for close examination of the inner ear. The high-resolution MRI scanning in the case of congenital bilateral profound sensorineural hearing loss may be more suitable because it allows identification of eighth nerve deficiency if cochlear implantation is under consideration.

Numerous surgical options have been presented for the management of congenital perilymphatic or CSF fistula. For type 1 defects through the otic capsule, many authors in the past have recommended stapedectomy and packing of the vestibule. Tyagi et al¹⁹ note that packing the vestibule with muscle or fascia has a 30 to 60% failure rate.^{13,20,21} They describe a multiple layer repair including packing of the vestibule with muscle, fascia, and glue and reinforcing the repair with a pedicled temporalis muscle graft. Intraoperative and postoperative lumbar drainage was used to decrease the pressure and flow of CSF during and after repair. They used this technique in the repair of leaks of four children and had no recurrences over 18-month follow-up. We also recommend an multiple layer repair and have found bone wax to provide good results in reinforcing the repair. In addition to the multilayered closure, we also performed blind sac closure of the external auditory canal as used in several skull base approaches. This allows more vigorous obliteration of the eustachian tube and middle ear space with no concern for violation of the tympanic membrane. Not only does the skull base closure technique allow obliteration of the inner ear route, but it also effectively separates the middle ear from aerodigestive tract contamination. By obliterating the middle ear space and the eustachian tube, one effectively separates the mastoid from the remaining aerodigestive tract; therefore, a mastoidectomy is neither required nor performed.

Stevenson et al²² suggested leaving the stapes in situ and placing grafts around it to seal the oval window niche if possible to prevent the possibility of the CSF gusher that may lead to difficulty in packing the vestibule such as what we encountered in case 2.^{13,23} They particularly recommend this technique in a severely dysplastic sac where the size of the vestibule and medial wall defects can lead to difficulties in packing. Conforming the packing around an intact and either mobile or fixed stapes suprastructure may be difficult to perform securely. This problem as well as a CSF gusher was encountered in case 2, but the CSF leak was still controlled.

Other congenital routes of CSF leakage in such cases may be a patent cochlear aqueduct, patent Hyrtl fissure (see case 3), a dilated fallopian canal,²⁴ or petromastoid canal.²⁰ Hyrtl fissure is a transient landmark in fetal temporal bone development. It is normally obliterated during the ossification process by gestational week 24. When it persists, it allows a communication between the posterior cranial fossa and the middle ear.^{25,26} As seen in case 3, this communication can lead to the development of meningitis. Rich et al describe a case of CSF leak from a Hyrtl fistula²⁶ in which a 5-year-old boy with a history of meningitis with purulent otitis media was found to have CSF leakage from an air cell 2 mm inferior and anterior to the round window that was packed with muscle to seal the leak. A recurrent leak developed 15 months after the initial surgery reexploration was performed; this cell was widened, allowing visualization of the entire tract, and then packed with muscle and bone wax. The patient had no additional leakage after the second procedure. In addition, Gacek and Leipzig described a case of patent Hyrtl fissure in 1979²⁵ with a meningoencephalocele present in the fissure. These type 2 leaks often present diagnostic and treatment

dilemmas because the inner ears are normal in function and radiographic appearance with a subtle visible route of leakage.

Sealing a type 2 defect should include an attempt at maintaining normal ear physiology including a patent eustachian tube, eardrum, and ossicular chain. As in case 3, these can be achieved, but the defect closure is certainly less secure, and CSF diversion should be more strongly considered to aid in the healing process. Should a more conservative attempt at type 2 leak closure fail, a more aggressive closure with sacrifice of external and middle ear physiology and preservation of inner function should be considered given the life-threatening nature of meningitis. Finally, there may be cases in which inner ear function has to be sacrificed to obliterate the CSF fistula definitively, but this should be a last resort. In either case of ear function sacrifice, a bone-anchored implant may aid in rehabilitation of the affected side.

Because type 1 and 2 leaks are congenital, their presentation is often during childhood; but most type 3 defects are presumably acquired and often present during adulthood. The etiology of the skull base defects in type 3 leaks is undetermined, but increased intracranial pressure, normal pressure CSF pulsations, and aberrant arachnoid granulations²⁷ have been described as possible etiologies. Elevated intracranial pressure is a common theme in many of these adult-onset leaks and currently the most favored etiology for all adult-onset spontaneous CSF fistulae. The diffuse nature of elevated intracranial pressure may explain why the location of these defects may be multiple.

The surgical management of type 3 otogenic CSF leaks is different from management of types 1 and 2 leaks. Work-up should include a CT scan and audiogram at the minimum. MRI and additional imaging including CT or MRI cisternography is considered if any abnormalities are detected on CT scan. Given the diffuse nature of elevated intracranial pressure, multiple skull base defects are relatively frequent. The defects tend to occur more along the middle fossa floor.^{9,19} When considering repair of an acquired spontaneous temporal bone CSF leak, the subtemporal approach provides a better field of view of multiple holes in the skull base and facile access above the tympanic cavity as well as medial to the inner ear relative to a transmastoid approach. A repair from above may also be physically more secure because it allows easy placement of a rigid graft larger than the defect with no chance it can fall through it; therefore, many authorities prefer repair of spontaneous middle fossa defects from above, and we strongly agree with this approach. The addition of a mastoid exploration and middle fossa repair adds the possibility of posterior fossa dural defect repair with little additional time and risk.

Conclusions

Otogenic CSF fistulae can occur through, adjacent to, and distant to the otic capsule. Each type of CSF fistula requires an individualized diagnostic and therapeutic strategy with regard to preoperative work-up, adjunctive lumbar drainage, type of surgical procedure, and postoperative functional expectations. Identifying the type of fistula and recognizing

potential pitfalls in treatment methods is essential to safe and effective treatment.

Notes

Presented as a poster presentation, North American Skull Base Society Annual Meeting; February 17–19, 2012; Las Vegas, NV.

References

- Neely JG. Classification of spontaneous cerebrospinal fluid middle ear effusion: review of forty-nine cases. *Otolaryngol Head Neck Surg* 1985;93(5):625–634
- Phelps PD, King A, Michaels L. Cochlear dysplasia and meningitis. *Am J Otol* 1994;15(4):551–557
- Brainard L, Chen DA, Aziz KM, Hillman TA. Association of benign intracranial hypertension and spontaneous encephalocele with cerebrospinal fluid leak. *Otol Neurotol* 2012;33(9):1621–1624
- Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope* 1987;97(3 Pt 2, Suppl 40):2–14
- Aguilar EF III. Auricular reconstruction of congenital microtia (grade III). *Laryngoscope* 1996;106(12 Pt 2, Suppl 82):1–26
- Meyerhoff WL, Stringer SP, Roland PS. Rambo procedure: modification and application. *Laryngoscope* 1988;98(7):795–796
- Chen DA. Dural herniation and CSF leaks. In: Brackman DE, Shelton C, Arriaga MA, eds. *Otologic Surgery*. 3rd ed. Philadelphia, PA: Saunders; 2010:245–251
- Kutz JW Jr, Husain IA, Isaacson B, Roland PS. Management of spontaneous cerebrospinal fluid otorrhea. *Laryngoscope* 2008;118(12):2195–2199
- Gubbels SP, Selden NR, Delashaw JB Jr, McMenomey SO. Spontaneous middle fossa encephalocele and cerebrospinal fluid leakage: diagnosis and management. *Otol Neurotol* 2007;28(8):1131–1139
- Streeter GL. On the development of the membranous labyrinth and the acoustic and facial nerves in the human embryo. *Am J Anat* 1906;6:139–165
- Cock E. The pathology of congenital deafness. *Guys Hosp Rep* 1838;7:289–307
- Phelps PD, Michaels L. The common cavity congenital deformity of the inner ear. An important precursor of meningitis described in 1838. *ORL J Otorhinolaryngol Relat Spec* 1995;57(4):228–231
- Ohlms LA, Edwards MS, Mason EO, Igarashi M, Alford BR, Smith RJ. Recurrent meningitis and Mondini dysplasia. *Arch Otolaryngol Head Neck Surg* 1990;116(5):608–612
- Schuknecht HF. Mondini dysplasia: a clinical and pathological study. *Ann Otol Rhinol Laryngol* 1980;89(Suppl 65):1–23
- Barcz DV, Wood RP II, Stears J, Jafek BW, Shields M. Subarachnoid space: middle ear pathways and recurrent meningitis. *Am J Otol* 1985;6(2):157–163
- Herther C, Schindler RA. Mondini's dysplasia with recurrent meningitis. *Laryngoscope* 1985;95(6):655–658
- Mondini C. Anatomica surdi nati sectio: bononiensi scientiarum et artium instituto atque academia commentarii. Bononiae 1791; 7:419–428
- Phelps PD, Lloyd GAS. Mondini dysplasia is not associated with meningitis and cerebrospinal fluid fistula. *Arch Otolaryngol Head Neck Surg* 1991;117(8):931–932
- Tyagi I, Syal R, Goyal A. Cerebrospinal fluid otorrhinorrhoea due to inner-ear malformations: clinical presentation and new perspectives in management. *J Laryngol Otol* 2005;119(9):714–718
- Spetzler RF, Wilson CB. Management of recurrent CSF rhinorrhea of the middle and posterior fossa. *J Neurosurg* 1978;49(3):393–397
- Park TS, Hoffman HJ, Humphreys RP, Chuang SH. Spontaneous cerebrospinal fluid otorrhea in association with a congenital defect of the cochlear aqueduct and Mondini dysplasia. *Neurosurgery* 1982;11(3):356–362
- Stevenson DS, Proops DW, Phelps PD. Severe cochlear dysplasia causing recurrent meningitis: a surgical lesson. *J Laryngol Otol* 1993;107(8):726–729
- Kimitsuki T, Inamitsu M, Komune S, Komiyama S. Congenital malformation of the inner ear associated with recurrent meningitis. *Eur Arch Otorhinolaryngol* 1999;256(Suppl 1):S11–S14
- Teufert KB, Slattery WH. Cerebrospinal fluid leak of the fallopian canal. *Ear Nose Throat J* 2013;92(3):E20–E23
- Gacek RR, Leipzig B. Congenital cerebrospinal otorrhea. *Ann Otol Rhinol Laryngol* 1979;88(3 Pt 1):358–365
- Rich PM, Graham J, Phelps PD. Hyrtl's fissure. *Otol Neurotol* 2002;23(4):476–482
- Propst EJ, Blaser S, Gordon KA, Harrison RV, Papsin BC. Temporal bone findings on computed tomography imaging in branchio-otorenal syndrome. *Laryngoscope* 2005;115(10):1855–1862