Preoperative computed tomography may fail to detect patients at risk for perilymph gusher

Michael D. McFadden, MD; Jason G. Wilmoth, MD; Anthony A. Mancuso, MD; Patrick J. Antonelli, MD

Abstract
Perilymph gusher (PG) is a very rare occurrence that can lead to an adverse outcome during inner ear surgery. In the absence of a family history of X-linked mixed deafness syndrome, surgeons may have difficulty determining if a patient is at risk preoperatively. Radiographic imaging is often performed in an attempt to identify such a possibility, but there are few data to support the value of negative studies. We conducted a retrospective study of 3 cases of PG in which findings on preoperative high-resolution computed tomography (CT) of the temporal bone had been interpreted as normal. We reviewed these CTs to discern if they did in fact demonstrate any abnormalities that might have indicated a risk of PG, and we found that the original radiologist had missed a dilated internal auditory canal and a deformity of the cochlear modiolus in the affected ear of 1 of these patients. No abnormality was detected on review of the CTs of the other 2 patients. Therefore, we conclude that negative CT findings do not necessarily rule out a risk of PG.

Introduction
Perilymph gusher (PG) is a rare complication of inner ear surgery. In a study of 2,405 patients who had undergone stapedectomy, Ginsberg et al reported that PG occurred in 0.5%.1 Stapedectomy is believed to pose a greater risk of PG in children than in adults, but it is still rare in children.2,3 When PG has been observed, most cases have occurred in males who had a mixed hearing loss (an X-linked trait), and most were associated with dysplasias of the otic capsule—specifically, dilation of the fundus of the internal auditory canal and a defect at the base of the modiolus.4–8 Because PGs may compromise surgical outcomes, preoperative detection is desirable.5,9,10 Although PGs have been reported in patients with radiographically normal temporal bones,9 most reports predate modern imaging techniques.11,12 In this article, we describe our retrospective review of the temporal bone findings in 3 patients who had experienced PG after their preoperative CTs had been reported as radiographically normal.

Patients and methods
After obtaining clearance from our institutional review board, we reviewed the case histories and preoperative temporal bone high-resolution CT studies of 3 consecutively occurring cases of PG in patients who had been treated by or later referred to the senior author (P.J.A.). These images were independently examined by a neuroradiologist (A.A.M.), a general otolaryngologist (M.D.M.), and a neurotologist (P.J.A.), who looked for anomalies associated with PG. Such anomalies include a dilated internal auditory canal fundus, a dilated vestibule, an enlarged cochlear aqueduct (specifically, the otic segment), an enlarged vestibular aqueduct, and cochlear dysplasia.

Results
Our examination of the CTs of patients 1 and 2 revealed no anomalies that would indicate a risk of PG. However, the CT of patient 3 did demonstrate the presence of a dilated internal auditory canal fundus and a deficient modiolar base. Patient 3’s cochlear aqueduct was normal; her vestibular aqueduct on the operative side was not visible.

Patient 1. A 34-year-old man with a history of a pre-lingual, bilateral, profound sensorineural hearing loss (SNHL) had been referred to our institution for possible placement of a cochlear implant. He had no known risk factors for hearing loss, and its etiology was unknown.
Notably, his family history was negative for hearing loss. Audiometry confirmed the profound SNHL (figure 1, A). CT revealed no inner ear dysplasia (figure 1, B).

The patient was taken to the operating room for a left cochlear implant. Upon creation of a cochleostomy immediately anterior and inferior to the round window, a brisk flow of clear fluid immediately erupted. The electrode array was inserted fully. The cochleostomy was packed tightly with periosteum, resulting in cessation of cerebrospinal fluid (CSF) flow.

The patient was admitted for bed rest, observation, and treatment with stool softeners. No CSF otorhinorrhea was observed for 24 hours, and the patient was discharged home. Five years postoperatively, he was successfully using his implant, and he exhibited no evidence of CSF leak or meningitis.

Patient 2. A 5-year-old boy had been referred to our institution for evaluation and treatment of bilateral conductive hearing loss after an outside otologist had performed left middle ear exploration and malleus mobilization. The patient had no personal otologic risk factors for hearing loss, and his family history was negative. Audiometry confirmed a purely conductive hearing loss bilaterally (figure 2, A). CT revealed no inner ear dysplasia (figure 2, B).

The patient was taken to the operating room for revision surgery. Intraoperatively, the patient was noted to

Figure 1. Patient 1. A: Preoperative audiometry confirms the SNHL in the left ear. B: Axial CTs of the temporal bones reveal a normal internal auditory canal and modiolus (top) and a normal basal turn of the cochlea and cochlear aqueduct (bottom).

Figure 2. Patient 2. A: Prestapedotomy audiometry confirms the conductive hearing loss in the left ear. B: Axial CTs (top row) of the temporal bones demonstrate a normal internal auditory canal and modiolus. Axial (bottom left) and coronal (bottom right) CTs reveal that the left cochlear aqueduct (arrows) is normal.
have a fixation of the stapes footplate. As a stapedotomy was performed with a CO\textsubscript{2} laser, a brisk flow of CSF rushed through the 0.15-mm laser fenestration. The fluid was evacuated until the flow diminished sufficiently to allow for expansion of the stapedotomy with a microdrill and reconstruction with a fluoroplastic-platinum piston over a fascia graft. The graft was secured with gelatin sponge, and the CSF flow abated. The patient was admitted for observation and CSF drainage via a lumbar drain. No CSF otorhinorrhea was noted, and the patient was discharged after 48 hours. Eight years postoperatively, his left conductive hearing loss persisted, but he had no appreciable SNHL. The patient was fitted with hearing aids, and he was able to hear without difficulty.

Patient 3. A 61-year-old woman had presented to another institution with a long-standing history of mixed hearing loss on the left. She had no known personal otologic risk factors for hearing loss; because she had been adopted, her family history was unknown. Audiometry confirmed the mixed hearing loss (figure 3, A), and radiographic imaging was reported as normal (figure 3, B). The patient underwent a left stapedotomy. Upon fenestration of the footplate, a brisk flow of CSF was noted. The middle ear was packed with gelatin sponge. The stapes prosthesis was not successfully placed.

The patient was admitted, and lumbar drain was placed for 3 days. The mixed hearing loss persisted postoperatively, but no CSF leak had recurred during 3 years of follow-up. During that time, the patient was able to hear without difficulty with the assistance of a hearing aid. Her records were submitted to the senior author for consultation following the stapedotomy, but no additional surgery was performed.

**Discussion**

The underlying anomaly in PG is an abnormal communication between the subarachnoid space and the perilymphatic space. Much debate has occurred regarding the exact site of communication between the subarachnoid and perilymphatic spaces. Most authors have described inner ear dysplasias in which an abnormal communication existed between the internal auditory canal and the vestibule or cochlea.\textsuperscript{4-8,10,12-14}

The association between PG and X-linked mixed deafness syndrome has been recognized for many years.\textsuperscript{15,16} The radiologic findings associated with this syndrome have been established as a bulbous dilation of the internal auditory canal, an incomplete separation of the basal turn of the cochlea from the internal auditory canal, and a widened first and second part of the intratemporal portion of the facial nerve canal.\textsuperscript{5,8,10} Female heterozygotes have demonstrated milder radiologic abnormalities, most noticeably dilated internal auditory canals. Our patient 3 likely fell into this category, but a definitive determination is not possible without further genetic studies. To our knowledge, the internal auditory canal has not been established as a viable route for a pathologic communication of CSF into the inner ear in radiographically normal temporal bones.

The cochlear aqueduct has also been implicated as a possible route of abnormal communication between the subarachnoid and perilymphatic spaces,\textsuperscript{13,17} but its role in PG has been called into question.\textsuperscript{18} For example, Park et al attributed 3 cases of spontaneous CSF fistula with recurrent meningitis to abnormally patent cochlear aqueducts.\textsuperscript{17} However, 2 of those 3 cases involved controlling the CSF leak by packing the internal auditory canal in ears with Mondini dysplasias; the other case involved an infracochlear cystic...
lesion that was merely in close proximity to the cochlear aqueduct. The cochlear aqueduct rarely if ever exceeds 2 mm at its narrowest otic segment. The lumen of the cochlear aqueduct is normally filled with soft tissue, but a lumen remains into adulthood. In 1978, Wlodyka reported the results of his histologic examination of the cochlear aqueducts in 250 temporal bone specimens. His examination included measurement of stain permeation through the aqueduct, as well as exposure of the aqueduct after infiltration with fluid plastic. He concluded that the patency of the cochlear aqueduct decreased with age, but he did not attempt to quantify how much patency would be required to allow for unimpeded flow of CSF into the perilymphatic space. Circumstances such as red blood cells filling the cochlear aqueduct after subarachnoid hemorrhage, the presence of red blood cells filling the cochlear aqueduct after subarachnoid hemorrhage, hearing loss after lumbar puncture, and the presence of fluorescein behind the round window after instillation via lumbar puncture suggest that some degree of patency remains in a small but significant percentage of adults.

PG is rare overall, and very rare in patients undergoing stapedectomy for otosclerosis; it is believed to be slightly more common in patients who have congenital malformations of the inner ear, including congenital stapes fixation. Notably, our patient 2 fell into this latter category. Unfortunately, in view of the sporadic nature of PG in these instances, no recent series have been reported in the literature. Likewise, the rare nature of the sporadic form of PG means that it is difficult to make statistically meaningful comparisons against normal temporal bones, either radiographically or histologically. Our MEDLINE review of the pertinent literature failed to identify any other study in which modern high-resolution CT was used in an attempt to identify patients with this abnormality, so a meta-analysis of documented cases of PG is not possible.

In our study, patient 3 had been incorrectly reported as radiographically normal despite the presence of CT abnormalities consistent with X-linked mixed deafness syndrome. The temporal bones of patients 1 and 2 demonstrated no radiographically apparent inner ear dysplasia or widening of the cochlear aqueduct that would indicate the precise focus of abnormal communication between the subarachnoid and perilymphatic spaces. Thus, the absence of radiographic anomalies does not rule out the possibility of PG. Because stapedial reflexes have been reported as usually—although not universally—in tact in patients with X-linked PG, stapedial reflex screening may also be considered. Based on our experience and others reported in the literature, a suspicion of PG should be heightened when evaluating young males who have had a lifelong conductive or mixed hearing loss.

Surgeons should be prepared to manage a PG intraoperatively because there are no known means of reliably predicting its occurrence. Patients with congenital deformities of the inner ear should be counseled about the possibility of this complication before they undergo stapedectomy or cochleostomy.

References