

Ventriculoperitoneal Shunt as Treatment for Perilymphatic Fistula: A Report of Six Cases

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Abstract: We report six cases of perilymphatic fistula in patients who received ventriculoperitoneal shunts as part of their final mode of therapy. The last of our 6 patients actually received a ventriculoperitoneal shunt as her initial mode of therapy. All but one had benign intracranial hypertension. All six felt better (less disequilibrium, tinnitus, and pressure and occasional hearing improvement) after LP with removal of 15–20 ml of cerebrospinal fluid.

Key Words: benign intracranial hypertension; displacement analyzer; LP; perilymphatic fistula; pseudotumor cerebri; tympanic membrane ventriculoperitoneal shunt

During the last 25 years, the author (DJW) has been intrigued by the diagnosis and treatment of perilymphatic fistula (PLF), initially in post-stapedectomy patients and later in patients whose PLFs were most likely initiated by a sudden change in middle-ear or spinal fluid pressure [1–8]. The treatment of such patients can be nothing more than a simple débridement around and reinforcement of the oval windows (OW) or round windows (RW) with soft tissue (fascia, fat, perichondrium, or loose areolar tissue). Over the years, however, in many patients with PLF, the disorder tended to recur after treatment (at periods varying from weeks to years). Occasionally, multiple breakdowns would occur in a single patient. This experience led the author to perform a lumbar puncture (LP) on one such patient (patient 1) only to find that the patient had benign intracranial hypertension (BIH) or pseudotumor cerebri. Draining 15 ml of cerebrospinal fluid (CSF) led to almost immediate relief of this patient's vertigo, bobbing oscillopsia, and tinnitus. After multiple attempts at "classic" PLF repair, he ultimately

was cured by the placement of a ventriculoperitoneal (VP) shunt. The authors subsequently began to perform diagnostic LPs on other similar patients suspected of having PLF, and that in turn led us to the diagnosis of BIH in more patients suspected of having a diagnosis of PLF. We found that lowering CSF pressure, whether it was normal or elevated, could lead to a temporary remission of symptoms that might last from hours to months. The use of a VP shunt followed naturally as a modality to treat these desperate patients.

Finally, the most recent addition of tympanic membrane displacement (TMD) technology (pioneered by Dr. Robert Marchbanks in Southampton, England) [9] has helped us to indirectly measure CSF pressure in the presence of an open cochlear aqueduct (CA) (briefly discussed here). Marchbanks's studies confirmed those of Wodyka [10], indicating that the CA had the potential of being open in all decades of life, although in decreasing percentages as humans advance through the decades. To some extent, this work was also confirmed by the temporal bone analysis of Gopen et. al. [11], although they were not able to demonstrate decreasing patency with advancing age.

METHODS

The authors selected six patients who presented at the Dartmouth Hitchcock Medical Center (DHMC), Lebanon, NH, during the last 18 years. These cases particularly illustrate how a change in CSF pressure can affect

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hearing, balance, tinnitus, and aural fullness (and, occasionally, deep ear pain) in patients with active PLFs. In all but one case (the last of the six) standard methods of PLF repair failed. These cases are presented here with comments regarding each.

CASE REPORTS

Patient 1

LL, a 42-year-old man, presented in 1985 with a history of intermittent disequilibrium for 6 years and daily disequilibrium for 3 years, which worsened with exercise. Reports of this patient's condition had appeared in part on three previous occasions [6–8]. His hearing was normal; on the Quix test, eyes-closed turning test (ECTT) of Singelton [12], eyes-closed walking test (ECWT) of Weider [8], and Fukuda high-stepping test with eyes closed, left drift was demonstrated. One hour after 40 mg of intramuscular (IM) furosemide (Lasix) was given, results on all the foregoing tests became normal. The authors' belief is that Lasix, when given intramuscularly or intravenously, lowers CSF pressure and, therefore, perilymphatic fluid pressure, permitting a temporary "normalization" in inner-ear function in patients with a PLF [13–15]. It basically stops inner-ear fluid flow and, therefore, the sense of imbalance.

Soon after presentation, LL was taken to surgery, where a left exploratory tympanotomy was performed. A fistula was identified at the superior end of the OW and was repaired. LL experienced approximately 6 weeks of symptom-free living, the first time in 6 years in which he had not been dizzy or experienced imbalance. At the end of the 6-week period, his symptoms returned without obvious cause. He had two OW repairs during the next 6 months, each one being successful for approximately 1 month. Finally, an endolymphatic shunt combined with repeat RW and OW reinforcement was performed, and LL again became asymptomatic for a period of some 6 weeks. Finally, at the end of 1 year and partially out of mutual frustration on the part of patient and surgeon, a left vestibular nerve section was performed, and LL remained asymptomatic with respect to balance for 5 years. He continued to experience some pulsatile tinnitus in the left ear when recumbent. In December 1992, he again complained of a sudden return of constant imbalance that increased with activity. His hearing was unchanged (right ear hearing normal, left ear unchanged since vestibular nerve section). He had bilateral tinnitus. He described the sound as a "motor" in his left ear, which worsened when lying down. He had a high-frequency steady ring in the right ear. Neurological tests were such that all Romberg-type tests (Quix, ECTT, ECWT, Fukuda) were positive for

the right, unoperated ear. Again, after 40 mg of IM Lasix, all Romberg-type test results normalized, and platform posturography improved dramatically. Tinnitus diminished as well. This positive effect lasted approximately 8 hours, as is typical.

Six weeks of bedrest was recommended, after which LL became asymptomatic for 6 months. When one day he bent over to tie his shoes, the symptoms immediately returned and did not abate after 2 more weeks of bedrest. Therefore, his right middle ear was explored, and an OW fistula was identified. Both windows were reinforced with loose areolar tissue. He appreciated immediate normalization of his balance, which lasted 1 month. Approximately 6 weeks later, the ear was reexplored, and both windows were reinforced once more. Again, he became asymptomatic for approximately 5 weeks, after which time symptoms returned.

An LP was performed in February 1994. The opening pressure was 280 mm of water. Fifteen milliliters of CSF were drained. LL experienced immediate and profound relief of symptoms (including imbalance, tinnitus, head pressure, motorlike tinnitus when recumbent, and bobbing oscillopsia). It should be noted that LL had bobbing oscillopsia despite good vestibular function in his right ear.

After some discussion, an LP shunt was performed in March 1994 [6]. This worked wonderfully until LL's dentist put him in the Trendelenburg position, at which time all symptoms returned immediately. The patient actually was aware of a "pop" in his ear when he was moved into this position. In June 1996, a CA blockade was performed by way of a posterior fossa approach [5,7]. This procedure led to 6 months of vertigo-free living. Then symptoms gradually returned. In July 1999, a VP shunt was performed, and the LP shunt was removed. With this procedure, LL's feeling of intracranial pressure disappeared, his tinnitus diminished, and his balance improved to near normal and remains so.

Comment

What seems quite clear in this patient is that minute changes or increases of CSF pressure led to recurrence of his symptoms. In retrospect, he probably had BIH for many years, and it was this increase of CSF pressure that led to the frequent recurrences of his cochlear and vestibular symptoms and easy breakdown of his PLF repairs. The so-called events that led to his recurrences were relatively minor. We felt that the increase in CSF pressure and, secondarily, perilymph pressure led to a decrease in compliance of the OW and RW and, consequently, ease of repeated "breakthrough" or recurrent PLFs. Having previous experience with LP shunting in two additional patients [6] and experiencing long-term failure plus some problems with low-pressure

headaches, the authors have resorted to VP shunting in such situations.

Patient 2

JT [7,8], a 64-year-old science teacher and part-time maintenance man, experienced in January 1995 a sudden increase in left-ear tinnitus (machine-like hum) and mild vertigo with increased physical activity. His first mild episode of vertigo occurred after lifting a heavy machine. A few weeks later, while hammering, he experienced a sudden further increase in tinnitus and decrease in hearing in his left ear plus nausea and vertigo. His audiogram demonstrated a 30-dB low-frequency sensorineural hearing loss with a discrimination of 84%. In June 1995, his referring physician explored the left middle ear and found and repaired what he believed to be an OW PLF. However, JT experienced no reduction in his symptoms. He was, therefore, referred to me (DJW). His Quix test, ECWT, and Fukuda test all were positive for the left ear and became straight (normal) 1 hour after he received 40 mg of IM Lasix. Two weeks later, this dramatic response to IM Lasix was demonstrated graphically by platform posturography. In addition to JT's improvement in balance, his motorlike hum in the left ear also diminished dramatically. Symptomatic improvement lasted for 3 days. We elected to obtain audiograms with the patient upright and then with his head down on a tilt board. JT demonstrated random changes in pure-tone hearing when lying supine on a tilt board with the head down. In November 1995, a left middle-ear exploration was performed. An LP was performed just prior to surgery, and his opening CSF pressure was, to our surprise, 270 mm of water. A lumbar drain was left in for 4 days. The patient became completely asymptomatic.

After 6 weeks of resting and feeling completely free of any imbalance, JT returned to work. His hearing improved, and vertigo and tinnitus diminished. Then, while lifting an air conditioner into position, he experienced a pop in his left ear, and all aural symptoms returned in full. Thinking another middle-ear PLF repair to be a temporary cure at best, we elected to perform a posterior fossa CA blockade [7,8]. After his surgery, JT experienced a sudden decrease in hearing threshold in the nonoperated right ear, which returned to normal after approximately 3 weeks. This outcome was reported by Walstead [16,17] and others performing acoustic tumor surgery. JT's CA blockade was successful and remained so for approximately 6 months, after which time symptoms again gradually returned.

On September 19, 1996, a VP shunt was performed. All aural symptoms greatly diminished. In November 1996, hypothyroidism was diagnosed, and JT was placed

on thyroid replacement therapy, at which time his remaining mild ataxia was eliminated. He continues to be followed up yearly and has remained asymptomatic.

Comment

This patient's onset of symptoms constituted a recognizable event for the onset of PLF. His initially unrecognized BIH was most likely a factor in initiation of his original symptoms, the failure of his first repair, and the ultimate failure of his second repair. This case demonstrates the value of performing a diagnostic LP in such cases.

Additionally, the decrease in hearing in JT's "good ear" at the time of his posterior fossa CA blockade provided evidence that CAs may be widely patent into the sixth decade [9-11] and that, if they are open, they may be more open than those found in the general population. Such patients probably are more prone to OW and RW tearing when confronted with a CSF pressure surge.

Patient 3

JD was a 36-year-old mildly obese woman who presented to DHMC on May 20, 1999. Her chief complaint was imbalance dating back to a motor vehicle accident in 1993. Her car was hit head-on at a mild angle. When the oncoming car hit her car's driver-side door and pushed her vehicle into a snow bank, she suffered mild whiplash. She broke the impact with her right arm. Since that time, she had experienced vertigo with physical stress and also with positional change (rolling to the right), much like a person with benign paroxysmal positional vertigo (BPPV). Her hearing was normal, with no tinnitus and no aural pressure or pain. She was motion-intolerant in elevators and occasionally experienced car sickness. Other doctors had tried to treat her imbalance with meclizine, scopolamine patches, and antidepressants. JD had also been referred to psychiatry, and some questioned whether financial compensation was a factor in her failure to improve. Her neurological examination result was almost normal, with a question of her going to the left on a Quix test and occasionally on the ECWT. In early May 1996, two Epley maneuvers [18,19] were performed from right to left. She experienced significant reduction in symptoms and felt almost normal for several months. Then, while walking down a set of stairs, she missed the bottom step and landed hard on the floor with both feet. At that time, she experienced an immediate return of symptoms, which became more clearly defined. Whenever she rolled to the right, she became dizzy, and she began having vertigo on physical stress or during any significant exercise. In April 1997, her symptoms became more severe. She was having great difficulty in driving:

She could not tell whether her car or the car next to her was moving. The author (DJW) placed the hand-held vibrator (used to perform Epley maneuvers) on her right mastoid, and she became destabilized [20]. Her transtympanic electrocochleogram result was normal. She performed platform posturography poorly and experienced increased nausea after Lasix administration. Electronystagmography results showed that she had occasional spontaneous left-beating nystagmus. She had a mild, subjectively positive fistula test outcome in her right ear.

In January 1998, JD's symptoms increased dramatically, and she became totally disabled. Her ECWT and Fukuda high-stepping test went to the right, and a decision was made to explore the right ear. Having diagnosed BIH in three previous patients with similar histories and symptoms, the author (DJW) requested that an LP be performed just before surgery to determine her CSF pressure. As the result was 290 mm of water, a decision was made to put in a lumbar drain. The right ear was explored, and the OW and RW were reinforced; no fistula was seen at the time of surgery. (Our belief is that if a lumbar drain is placed preoperatively, a PLF will probably not be seen at the time of surgery.) The next morning, she awoke and said, "It's the first time I've felt normal in 5 years." Some mild head pressure (which she had been experiencing but about which she had not complained) was completely gone, and her balance was perfect. The drain was left in for 11 days. JD experienced 4 symptom-free years despite no attempt to treat her BIH during this period. She felt no symptoms with respect to her eyes or headaches.

Then, in the spring of 2002, she fell and injured her back. Her symptoms returned. LPs were performed in the early summer of 2002 and in February 2003. Both times, CSF pressure was elevated in the range of 280–290 mm of water. Each time she had an LP, 15 ml of fluid was withdrawn, and each time her vertiginous symptoms diminished for a while but did not abate. In August 2003, a decision was made to perform a VP shunt. This procedure resulted in complete resolution of all symptoms. She did not have the right ear reexplored. The presumption was that healing of the OW PLF occurred when her CSF pressure was lowered to normal levels. Her shunt was placed 16 months ago. At the time of this writing, she is asymptomatic with respect to vertigo and head pressure.

Comment

The authors believe that JD most probably had increased CSF pressure at the time of her motor vehicle accident and that her BIH possibly made her more vulnerable to the development of her "trauma-induced PLF." We believe that BIH in the presence of an open

CA reduces the compliance of the OW annular ligament and the RW membrane, rendering them more susceptible to tearing. Her symptoms early on were so mild that few physicians believed her problem to be real, a common occurrence for people with this entity. Her vestibular symptoms were dramatically reduced every time CSF pressure was reduced. Ultimately, her "cure" necessitated a permanent lowering of CSF pressure by means of a VP shunt.

Patient 4

PN, a 34-year-old woman, presented to DHMC in May 1996 with a 2-month history of vertigo on exertion, high-pitched tinnitus, and intermittent right-ear pain that occurred for perhaps 10 minutes four to five times daily. PN actually had a 7-year history of vertigo that began when she picked up off the floor a woman who had slipped out of a wheelchair. Dr. Dennis Fitzgerald [21] of Washington, DC, diagnosed and repaired a right OW PLF in 1992. Her vertigo was reduced to 50% of what it had been, and she was able to return to work, although still limited in what she could do owing to balance disorder. In 1995, she and her husband moved to New Hampshire. During the 2 months before her visit to the author (DJW), her symptoms increased dramatically such that she was severely limited in her ability to hold down her job as a file clerk. Her main complaints were intermittent right-ear pain, vertigo on exercise, and right-ear tinnitus. Physical examination revealed that she had positive fistula test results bilaterally (stronger on the left). She veered to the left on her Quix test, Fukuda high-stepping test, ECTT, and ECWT. One hour after being given 40 mg of IM Lasix, all of the foregoing test results reverted to normal. Bilateral PLFs were suspected.

In August 1996, PN underwent bilateral exploratory tympanotomies. She had an obvious PW PLF in her left ear, with a fairly dramatic free flow of fluid that reaccumulated after each suctioning. Many adhesions surrounded the OW in the right ear, with a less obvious PLF. The OW and RW were reinforced bilaterally with loose areolar tissue. After surgery, her vertigo, aural pressure, tinnitus, and aural pain immediately disappeared. The duration of normalcy was 4 weeks, after which time symptoms returned approximately 1 hour after she awakened one morning. PN experienced fullness and tinnitus in her right ear, the ear originally repaired in 1992. She had a positive fistula test result on the right and a negative result on the left. All Romberg-type tests veered to the right (although to the left previously) and again normalized after 40 mg of IM Lasix.

In November 1996, a stress transtympanic electrocochleogram [8,22] was performed. At rest, her

summating potential–action potential ratio was normal in the right ear at less than 30% but, with a Valsalva maneuver, it rose to approximately 80%. This rise in summating potential–action potential ratio occurred five consecutive times with the Valsalva maneuver. This outcome indicated to us that the CA was most likely widely patent and that any increase in CSF pressure was capable of an immediate and dramatic effect on the patient's cochlear fluid pressure. In December 1996, a right endolymphatic shunt with repeat reinforcement of the OW was performed. PN experienced complete abatement of all symptoms. Her good result lasted for approximately 3 months until she underwent back surgery for a ruptured lumbar disk. Symptoms returned immediately on awakening from surgery. The feeling was that the postoperative nausea and some vomiting caused the recurrence.

On October 1, 1999, an LP revealed an opening pressure of 285 mm of water. Fifteen milliliters of CSF were removed, after which all aural symptoms abated for 2 days. On November 19, 1999, a VP shunt was placed. PN immediately became asymptomatic and remained so until the early summer of 2002, when symptoms returned. We discovered at that time that her shunt was malfunctioning. A valve was replaced, and again she experienced symptom-free living for another 18 months. In September 2003, she was given a diagnosis of an abdominal abscess thought to be secondary to her VP shunt. The shunt was removed, and the abscess was drained. Despite shunt removal, PN has remained asymptomatic with respect to balance, aural pressure, tinnitus reduction, and hearing. She is, however, experiencing some very mild head pressure, although it is not constant.

Comment

PN's initial event was fairly classic for the initiation of a PLF. It went undiagnosed for 2 years before her initial diagnosis and PLF repair by Fitzgerald [21]. Her lack of complete cure and relative ease of relapse most likely were due to an unrecognized, underlying BIH and a probable PLF in the other ear. Once the BIH was remedied by a VP shunt, PN's recurrent PLFs ceased to be a problem. As this case demonstrates, VP shunts are not without problems of their own.

Patient 5

SS presented as a 35-year-old woman who had been hit on the head with a swing in 1984. After this event, she had a history of daily imbalance that increased with physical activity. She had fluctuating hearing in the low frequencies in her right ear. She also had an increase in tinnitus and pressure in the same ear. She presented to

DHMC in the fall of 1990. In addition to her low-frequency hearing loss in the right ear, she had a bilateral, symmetrical, high-frequency sensorineural hearing loss. Her right ear discrimination was 92%. Discrimination in the left ear was 100%. A neurological examination elicited a positive fistula test result in her right ear. Her Quix test, Fukuda test, and ECWT all demonstrated right drift. All reverted to normal 1 hour after she received 40 mg of IM Lasix. In June 1990, an exploratory tympanotomy of the right ear was performed. An OW PLF was found and repaired; actually, both windows were reinforced. Her vertigo completely resolved. Tinnitus became much reduced, and her low-frequency hearing became slightly improved and stable. She remained completely asymptomatic for 5.5 years.

All symptoms returned in January 1996 secondary to nose blowing and jumping rope. SS experienced an increase in tinnitus, aural pressure, and imbalance. Her low-frequency hearing loss now began to fluctuate once again. She was reexplored in February 1996 and, once again, an OW fistula was thought to be present and was reinforced with loose areolar tissue. SS again experienced complete symptomatic recovery. In January 2000, 4 years after her revised repair, she stood up under a fixed iron beam, hitting her head hard, and all symptoms returned. These resolved with bedrest, and she remained asymptomatic between January and July 2000, at which time symptoms returned. Despite another attempt at bedrest, SS continued to experience constant vertigo, tinnitus, pressure, imbalance, and low-frequency hearing loss.

An LP was performed in January 2001. Her opening pressure was 180 mm of water; closing pressure was 120 mm of water after removal of 15 ml of CSF. One hour later, she had no vertigo, no tinnitus, and no aural fullness, and all her Romberg-type test results reverted to normal. SS remained completely asymptomatic for 9 months or until the fall of 2002. An important note is that no middle-ear surgery—only a spinal tap with reduction of CSF pressure—was performed.

Because of the return of symptoms in the fall of 2002, LP was repeated in December, and 20 ml of CSF was removed. Symptoms greatly diminished but did not completely disappear. SS attempted to continue to teach school and did so until the end of March 2003, when symptoms became intolerable. At this time, she was offered a repeat middle-ear exploration, but she was also offered a VP shunt. After much discussion, a VP shunt was performed on June 16, 2003, despite normal CSF pressure. SS experienced immediate relief from tinnitus, head pressure, and aural fullness. Low-frequency hearing thresholds improved slightly as well. Thus far, she has remained completely symptom-free (16 months).

Comment

SS is the first patient in our series to undergo a VP shunt for recurrent PLF in the face of normal CSF pressure. Again, this patient clearly shows that an increase of CSF pressure transmitted through a presumably patent CA was capable of causing a recurrent perilymph leak and its associated symptoms of hearing loss, tinnitus, and aural pressure. For some reason, she also experienced a feeling of intracranial pressure, despite normal CSF pressures. We believe that the VP shunt placement in this particular patient allowed the PLF to "self-seal," as had the LP in January 2000. We believe the value of the VP shunt lies in (1) the general lowering of CSF pressure and (2) preventing extreme pressure surges caused by lifting, sneezing, nose blowing, and the like.

Patient 6

On June 8, 2001, BA, a 55-year-old woman, was referred to me (DJW) by a colleague for symptoms of BPPV and to rule out a PLF. Whenever she lay on her left side, she became vertiginous. Her BPPV began in the fall of 2000 and responded to Epley maneuvers [18,19] at that time. She was involved in an automobile accident on January 11, 2002, when her car was rear-ended at some 30 mph while stopped at a traffic light. A couple of months after this accident, her BPPV began again. She did respond to Epley maneuvers [18,19] going from left to right but only for brief periods (2–7 days). At the time of her initial visit, she became vertiginous when rolling to the left and, occasionally, when bending over and righting herself again. Additional problems were hypertension, obesity (5'3", 300 lb.), snoring, and sleep apnea.

During this initial office visit, BA had clear signs of BPPV with geotropic rotary nystagmus with the left ear down. The author (DJW) performed two Epley maneuvers [18,19] from left to right with a small hand-held vibrator on BA's left mastoid. After this procedure, she could lie in the Halpike left position with no vertigo. Also during this visit, strategies to treat sleep apnea were discussed.

The patient returned to my office on June 29, 2001. Symptoms of BPPV had returned after only a few days. The author performed two more Epley maneuvers from left to right at this visit. BA then experienced 3 weeks symptom-free from any vertigo before return of the identical symptom complex described earlier. Between our initial visit and January 2003, the author saw BA approximately 13 times, during which he performed Epley maneuvers for presumed BPPV. After each set of maneuvers, BA experienced a period of being nearly symptom-free followed by return of symptoms. (Her tem-

porary positive experience after canal repositioning maneuvers was similar to that experienced by JD, patient 3.)

Gradually, new information began to surface. BA was beginning to become terribly carsick for the first time in her life. This occasionally happened even when she was driving. She could not read for very long without getting sick or feeling nauseated. She became dizzy walking down the aisles of large department stores, was having occasional bad headaches, and developed a chronic feeling of mild pressure in her head. We began more seriously to entertain a diagnosis of PLF.

On May 27, 2003, BA underwent platform posturography. It was followed immediately by an injection of 40 mg IM Lasix, and 1 hour later posturography was repeated. After administration of Lasix, the patient did show slight improvement in some of her tests, but the results were ambiguous. On September 17, 2003, an LP was performed. Her opening pressure was 210 mm of water (mild elevation). After we drained 20 ml of CSF, her "head pressure" (not previously mentioned by the patient) diminished, her tinnitus diminished to the point of almost disappearing, her balance became normal, and she had no further symptoms of positional vertigo. Hearing subjectively improved in both ears, and this improved state lasted for 3 days. She was astounded, as for the first time since her automobile collision, she felt truly normal.

TMD testing in her left ear [9] confirmed or also indicated elevated CSF pressure. TMD testing is nearly identical to a tympanogram with the exception that the pressure-sensing mechanism is much more sensitive. The pressure-sensing device can detect as little as a nanoliter of pressure change (a thousandth of a millionth of a liter). An airtight seal is obtained in the ear canal, and a sound sufficient to produce a stapedius reflex is elicited. The stapedius muscle will pull the stapes toward or away from the inner ear, depending on its resting position. This small inward or outward motion of the stapes footplate will, in turn, cause inward or outward displacement of the tympanic membrane, which will be depicted on a computer monitor and can be printed. If the CA is functionally patent in a patient with normal CSF pressure in the sitting position, the perilymph will flow toward the CSF, the stapedius footplate will "relax" into the inner ear, and a stapedius reflex will pull the footplate laterally and displace the TM in the same direction. When a patient lies down for 15 minutes, the CSF will become more elevated in the head, and CSF will move toward the inner ear and displace the TM laterally; then a stapedius reflex will pull the footplate inward and secondarily pull the TM medially. In a patient with BIH (such as BA), the footplate will rest laterally even in the sitting position, as was the situation when her left ear was tested. BA demonstrated

a biphasic pattern when the right ear was tested (sitting and recumbent), indicating that the CA on the right side was most likely not open.

The combination of (1) a good response to the removal of CSF with respect to balance, head pressure, and tinnitus; (2) the realization that she had BIH (LP and TMD results); and (3) poor success with the Epley maneuver with respect to her symptoms of BPPV led us to believe that she had a PLF in her left ear.

Because of previous poor experiences in repairing PLFs by the direct middle-ear approach in obese patients with BIH (patients 1 and 4 being prime examples), we decided to go directly to VP shunting and to make no attempt to repair the middle-ear PLF. We had also had experience with patients 1–5, whose symptoms of PLF were ultimately cured by VP shunts without repeating middle-ear explorations and OW and RW reinforcement. Therefore, on December 3, 2003, BA underwent a VP shunt. She experienced 3 excellent asymptomatic months (no imbalance; tinnitus reduction; decrease in feeling of aural and intracranial pressure; and no BPPV). On March 9, 2004, her shunt had to be removed because of an abdominal infection at the shunt site. She remained asymptomatic for 2 weeks after shunt removal, and then symptoms returned. Her VP shunt was replaced in June 2004, and she remains asymptomatic at the time of this writing.

Comment

As stated, we doubt that a simple middle-ear exploration and repair in a person with known BIH and a high probability of a PLF would be curative for an indefinite period.

DISCUSSION

All these patients (with the exception of patient 1) had an obvious event that prestaged their symptoms of vertigo and, in patient 1, events terminated all postoperative vertigo-free intervals. Bedrest, often suggested as an effective treatment, had been tried in several cases but was helpful only transiently in patients 1 and 5. The author (DJW) has seldom found bedrest to be helpful in promoting a long-term cure unless the case is immediately diagnosed and bedrest is suggested within a week to 10 days of the event or onset of symptoms.

Until approximately 3 years ago, the author (DJW) would perform a simple exploratory tympanotomy with reinforcement of the OW and RW as an initial procedure on a patient suspected of having a PLF. Currently, simple OW and RW reinforcements are being coupled with an LP, with the drainage of 15–20 ml of CSF at the time of surgical exploration. When an LP is performed before the middle-ear exploration, a PLF will

seldom be identified during surgery, as the reduction in CSF pressure will most likely lower perilymph pressure to the extent that a leak cannot be visualized. Occasionally, a lumbar drain will be left in for several days to a week, with the intent of permitting initial healing of a PLF during a period of low perilymphatic pressure.

This author (DJW) agrees with Fitzgerald [21] that patient history and preoperative evaluation define the diagnosis of PLF to the extent that one does not necessarily have to identify it at surgery in order to proceed to reinforcing both windows. Hearing is seldom negatively affected more than a few decibels, and surgeon and patient are usually rewarded by hearing stabilization and reduction of tinnitus, pressure, vertigo and, occasionally, pain. As stated, the defining characteristic of a patient with PLF is constant mild instability reliably made worse by physical stress (stress that elevates the CSF).

Recurring PLF has been a major problem for the authors and their patients. The senior author (DJW) is impressed with the ease with which a PLF can recur. Events leading to recurrence are often trivial as compared to the event that initiated the initial onset of symptoms. One might ask why. One reason might be that the initial “explosive event” leading to the onset of symptoms most likely is generated by a CSF “pressure head” capable of destroying the protective filtering mechanism of arachnoid tissue that fills the CA. When this happens, CSF fluid might flow more freely through the CA into the scala vestibula. Once this arachnoid mesh filter has been violated, it may never be fully restored or it may take many months without excessive surges of CSF pressure to be restored. This may be the situation in patient 4, who had a VP shunt for several years, had it removed out of necessity, and now no longer requires it. Certainly clinically this seems to be the outcome.

If a patient is in a cycle in which a simple middle-ear PLF repair will not stay put over time, the treating medical team may need to resort to CA blockade [7,23] or a VP shunt (the latter seemingly being less risky and possibly more long-lasting) for more permanent resolution of the problem. Acetazolamide has been used by the author (DJW) [3,4] to reduce CSF pressure medically, although with only temporary good results and poor patient compliance.

Additionally, only recently [6] has the diagnosis of BIH been made in patients with PLF, and indeed, it may play an etiological role in PLF. One needs to read and study the works of Sismanis [24–26] to appreciate more fully the relationships of hearing, balance, and tinnitus to BIH and the treatment of BIH, VP shunting may be required for long-term cure in patients with this disorder. This has been our experience.

Only during the last 18 months have the authors resorted to VP shunting in a patient (patient 5) with

normal CSF pressure. Time will tell whether this modality is warranted in such cases, but all patients who have received this therapy thus far have been most grateful.

Finally, the senior author (DJW) believes that consideration of offering an endolymphatic shunt still has a place (as in patients 1 and 4). The rationale for such a procedure in the case of a patient with a PLF is probably twofold. First, many (if not all) patients with an active PLF probably have a secondary endolymphatic hydrops [27]. An endolymphatic shunt has the potential to correct this disorder in some cases [3,4,28]. Second, performing an endolymphatic shunt or sac decompression may increase the overall compliance of the inner ear, theoretically taking some of the stress away from a CSF pressure surge against the RW and OW.

In the last case presented, we made a decision to go straight to a VP shunt rather than attempting an OW and RW reinforcement as a first option. The patient's CSF pressure was only slightly elevated at 210 mm of water, causing the stapes footplate to rest laterally in the sitting position. (The stapes reflex pulled the stapes and the ear drum inward, as demonstrated by TMD testing [9].) Two factors led to making the decision for a VP shunt as an initial procedure: One was BA's dramatic improvement (complete normalcy with respect to balance, hearing, and absence of tinnitus plus a decreased feeling of "head pressure") after her LP; the second was her excessive weight (300 lb.). In this author's [DJW] experience, obese people most likely have greater CSF pressure surges in simply bending over or straining to get up. Additionally, BA currently has untreated sleep apnea that may also be a risk factor for the development of PLF.

In summary, the authors have presented six cases demonstrating the various methods of diagnosing and treating PLF. Diagnostic modalities range from a good history and office Romberg testing to stress electrocochleogram and platform posturography before and after Lasix administration to, most recently, LP for determining CSF pressure and the effect on a patient's symptoms of removing 15–20 ml CSF. Treatment ranges from simple reinforcement of the OW and RW with loose areolar tissue to coupling that method occasionally with an endolymphatic shunt and, more recently, with an LP with drainage of 15–20 ml CSF or the placement of a lumbar drain to lower CSF pressure. A more definitive therapeutic option for the patients presented here proved to be the placement of a VP shunt.

CONCLUSIONS

Patients who present with (or, occasionally, without) a defining event (explosive or, occasionally, implosive)

and complaining of daily instability with or without hearing loss, aural pressure, tinnitus, and sometimes deep ear pain most likely have a PLF. Their imbalance is nearly always worsened by increased physical stress (any body function that increases CSF pressure). The diagnosis is strongly suggested when these entities improve or disappear on reduction of CSF pressure (administration of Lasix or an osmotic diuretic of some other kind or by an LP and fluid drainage). Additionally, the performance of an LP can reveal the diagnosis of BIH or pseudotumor cerebri that often is present in the absence of ocular signs and significant headache (as has been our experience). Treatment ranges from simple OW and RW reinforcement to the performance of a VP shunt, which, in cases of standard treatment failure, has offered a more permanent solution for some of our patients, particularly those with BIH.

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