Chapter 104 – Endoscopic Management of Cerebrospinal Fluid Leaks

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A cerebrospinal fluid (CSF) leak implies a fistula between the subarachnoid space and the sinona-sal tract. Patients with CSF leaks have symptoms such as clear nasal discharge and headache or complications such as pneumocephalus, meningitis, or a brain abscess. Immediate identification plus repair of a CSF fistula avoid the development of life-threatening complications.[1,2]

CSF leaks can be classified in a number of ways. Etiology, anatomic site, age of the patient, and underlying intracranial pressure are the most common classification systems used. Etiologic classification can be subdivided into traumatic and nontraumatic in origin. In the traumatic category, leaks can be either accidental or iatrogenic. Most CSF leaks are caused by trauma-induced fractures of the cranial base or by injuries produced during endoscopic sinus surgery or cranial base surgery. CSF leaks are diagnosed in 3% of patients suffering closed head injuries and in up to 30% of patients with a fracture of the skull base.[3] Similarly, iatrogenic trauma such as that caused by traditional cranial base surgery and endoscopic sinus surgery is associated with CSF leaks. Endoscopic sinus surgery remains one of the most common causes of CSF rhinorrhea despite an overall incidence of 1% in patients undergoing such surgery.[4–21] Nontraumatic CSF leaks may occur secondary to tumors of the skull base, congenital malformations, or high-pressure hydrocephalus (HPH). The etiology of HPH includes tumors, trauma, infections, and hemorrhagic cerebrovascular accidents (CVAs). Occasionally, no cause is identified for the HPH and it is therefore deemed “idiopathic.”

Multiple techniques have been reported for the repair of a CSF fistula. In 1952, Hirsch described the first endonasal repair of CSF rhinorrhea with a septal flap.[22] Subsequently, Montgomery described his experience with septal flaps through an external nasal approach to treat CSF rhinorrhea.[23] In 1976, McCabe reported his experience with the use of osteomucoperiosteal flaps from the septum or middle turbinate through an external ethmoidectomy approach.[24] In 1989, a publication of McCabe's updated experience reported a 100% closure rate with follow-ups of 1.6 to 22 years.[25] The use of these and other local flaps to treat CSF fistulas via an endoscopic approach was subsequently reported.[13,26]

In 1985, Calcaterra[27] described the use of free muscle or fascia grafts to treat CSF fistulas via an external ethmoidectomy approach, and in 1989, Papay and colleagues[28] described an endoscopic technique. Multiple publications followed with reports of the use of free grafts or local flaps, or both, for the repair of CSF fistulas. Recently, a vascularized nasoseptal flap has been devised to reconstruct skull base de-fects of various causes, and early follow-up data have revealed a remarkable improvement in CSF fistula rates after expanded endonasal approaches to the skull base.[29]

The availability of biologic material and the experience and familiarity of the operating surgeon with various techniques are the most important factors influencing the choice of which technique is used during endoscopic endonasal closure of CSF fistulas. Most techniques yield similar results in experienced hands, as confirmed by a meta-analysis of the literature by Hegazy and coworkers.[1] After analyzing all reports in the English literature, these authors found no significant differences in outcome regardless of which surgical technique or which homologous material was used for the repair.

PATIENT SELECTION

Diagnosis plus management of a CSF leak involve three critical steps: distinguishing a CSF leak from other sources of rhinorrhea, locating the fistula, and ruling out high intracranial pressure secondary to altered CSF dynamics.

Clinical diagnosis of a CSF leak is based on a history of clear, watery nasal drainage that is generally unilateral and commonly associated with headache. Increased rhinorrhea when the patient leans over, tilts the head forward, or performs a Valsalva maneuver further suggests the presence of a CSF leak. A history of tumor, trauma, or previous surgery involving the paranasal sinuses or cranial base heightens the level of suspicion. Occasionally, a life-threatening complication arising from the CSF leak, such as pneumocephalus, brain abscess, or ascending bacterial meningitis, may be the initial manifestation.

Conditions such as vasomotor rhinitis and sympathetic denervation can cause profuse rhinorrhea that may be confused with a CSF leak. Nasal irrigation fluid used during endoscopic sinus surgery or as part of postoperative care may accumulate in the paranasal sinuses and later manifest as postoperative rhinorrhea. Thus, biochemical testing is indicated to confirm the true nature of the nasal drainage. CSF will be high in glucose and low in protein. However, normal nasal discharge has been shown to be falsely positive for glucose in 45% to 75% of cases.[30] β_2-Transferrin is a protein found in CSF, aqueous humor, and perilymph, but not in blood or nasal secretions; therefore, β_2-transferrin is a reliable chemical marker of CSF leakage.[31–35] In addition to β_2-transferrin, a new chemical marker called β-trace protein is being explored for the diagnosis of CSF fistula.[36]

PREOPERATIVE PLANNING

When CSF rhinorrhea develops after sinus surgery, the endoscopic surgeon typically has an impression about the possible site of injury to the cranial base (i.e., site of the CSF fistula).

Therefore, a thorough endoscopic office examination of the nasal cavity may identify the site of leakage. Fistulas with low-pressure leaks are difficult to identify, especially in the presence of postoperative tissue edema and blood clots.

Intrathecal injection of contrast material and radioactive tracers has been advocated to confirm a CSF leak and identify the site of origin. Intrathecal fluorescein may be used to aid in the diagnosis and localization of a CSF leak. During this test, 0.5 mL of fluorescein at a concentration not greater than 5% is diluted with 10 mL of CSF obtained through lumbar puncture and then injected intrathecally. Fluorescein is neurotoxic, so a low-concentration, low-volume injection is mandatory to avoid the neurologic complications associated with higher concentrations. After intrathecal injection of the fluorescein solution, the CSF leak may be visualized with a nasal endoscope.[31–35,37] Under Wood's lamp (i.e., black light), fluorescein will appear as bright yellow-green. The yellowish color of fluorescein may also be identified without the need for special lighting.

The logistics of performing a lumbar puncture in an outpatient setting and medicolegal considerations (the fluorescein drug package insert includes an advisory warning against intrathecal use) have discouraged us from using this technique. Intrathecal injections are rarely critical to identification of a CSF fistula. Exceptional patients are those with multiple fistulas, such as those patients who have suffered skull base fractures, and the rare patient with repeated episodes of meningitis in the absence of an apparent cranial base defect or other predisposing factors such as rhinosinusitis or otitis media.

Others have advocated intrathecal injection of air, which can “bubble” out at the fistula site and therefore aid in identification.[37] Air, however, is an irritant to the brain and may induce seizures. Normal saline solution may be injected into the intrathecal space to increase the pressure within the subarachnoid space and thus aid in identification of the leak.

Scintigraphy with indium-111 has been advocated for the identification of a CSF leak. Radiotracing is a very sensitive test; however, it is associated with a high false-positive rate[31] and has a poor resolution that precludes establishing the specific point of leakage. In general, we do not advocate scintigraphy. In our practice, the presence of a CSF leak is ascertained with β_2-transferrin electrophoresis, and its site of origin is identified by computed tomography (CT) scanning, magnetic resonance imaging (MRI), or CT cisternography, with or without endoscopy (Fig. 104-1).

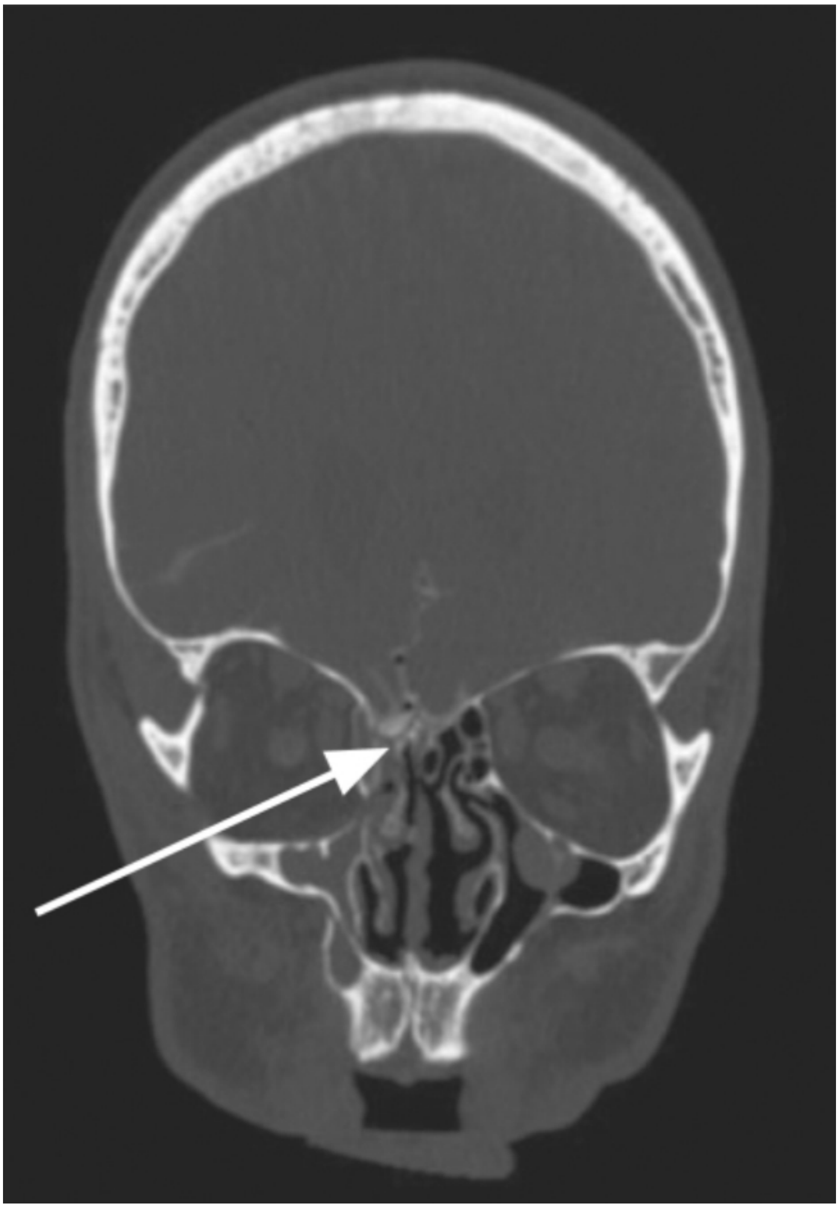

Figure 104-1 Coronal CT cisternogram demonstrating a right cribriform defect leading to a post-traumatic cerebrospinal fluid fistula *(arrow)*.

An important consideration in a patient with spontaneous rhinorrhea or rhinorrhea after skull base trauma is to distinguish whether the CSF rhinorrhea is arising from the sinonasal tract or from other sites such as the middle ear or mastoid (Fig. 104-2). A CSF fistula in the temporal bone may drain into the nose through the eustachian tube. Imaging is critical to locate the site of the fistula. Radiographically, high-resolution computed tomography (HRCT) can be used to identify a cranial base defect. HRCT is our preferred initial imaging study to aid in identification of the site of injury and its extent (Fig. 104-3). HRCT with contrast enhancement also provides information about the possibility of intracranial complications that occur in the setting of acute trauma (iatrogenic or accidental), such as hematoma or brain contusion. HRCT with views taken at a plane perpendicular to the suspected site of injury better evaluates the integrity of the bony wall in question. Coronal CT views are best to evaluate defects of the cribriform plate, fovea ethmoidalis, or planum sphenoidale, whereas axial views are superior for evaluation of the posterior wall of the frontal or sphenoid sinuses. We use MRI to ascertain the contents of a meningocele or meningoencephalocele (Fig. 104-4).

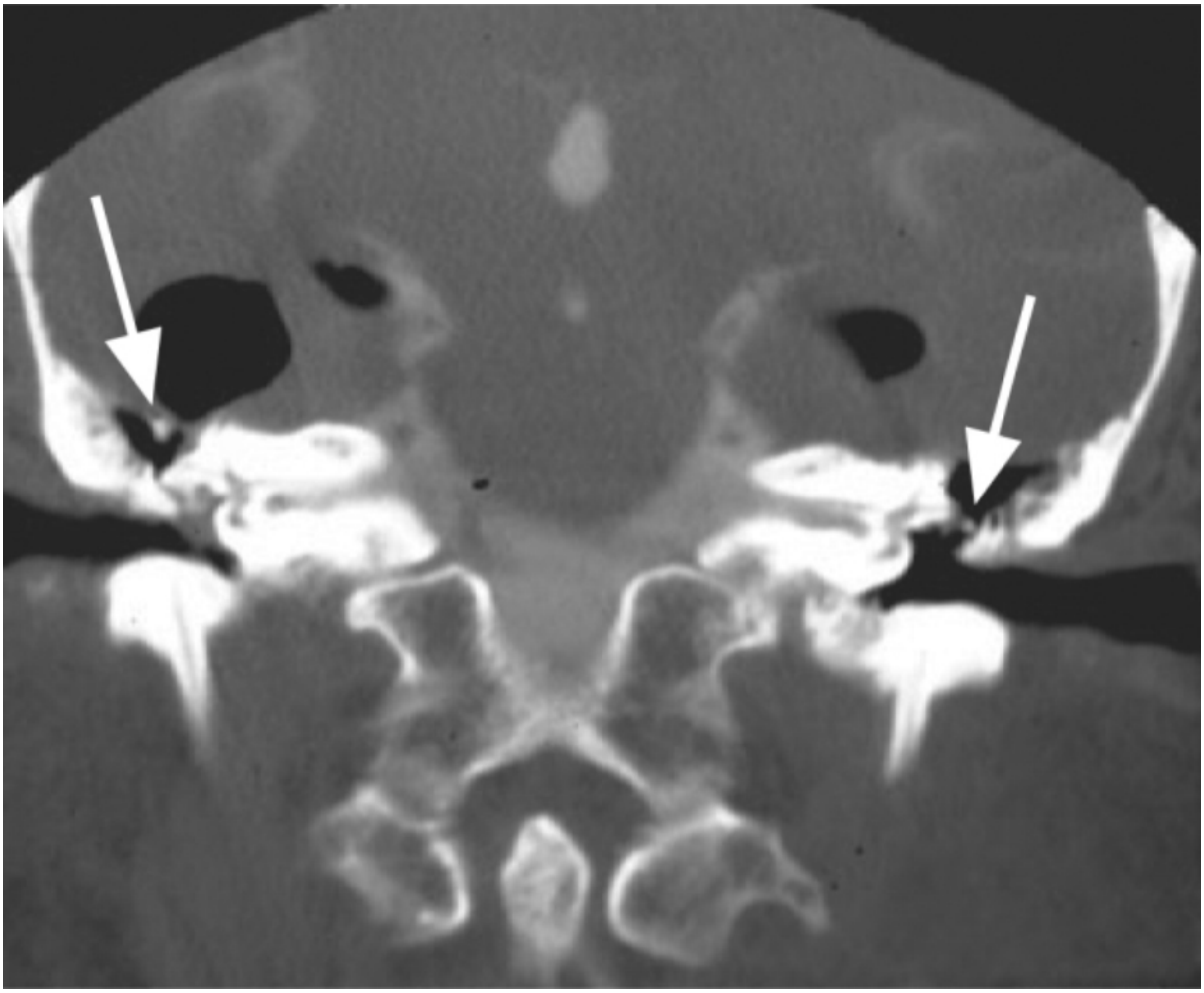

Figure 104-2 Coronal CT cisternogram demonstrating bilateral tegmen defects with contrast material penetrating through existing defects *(arrows).*

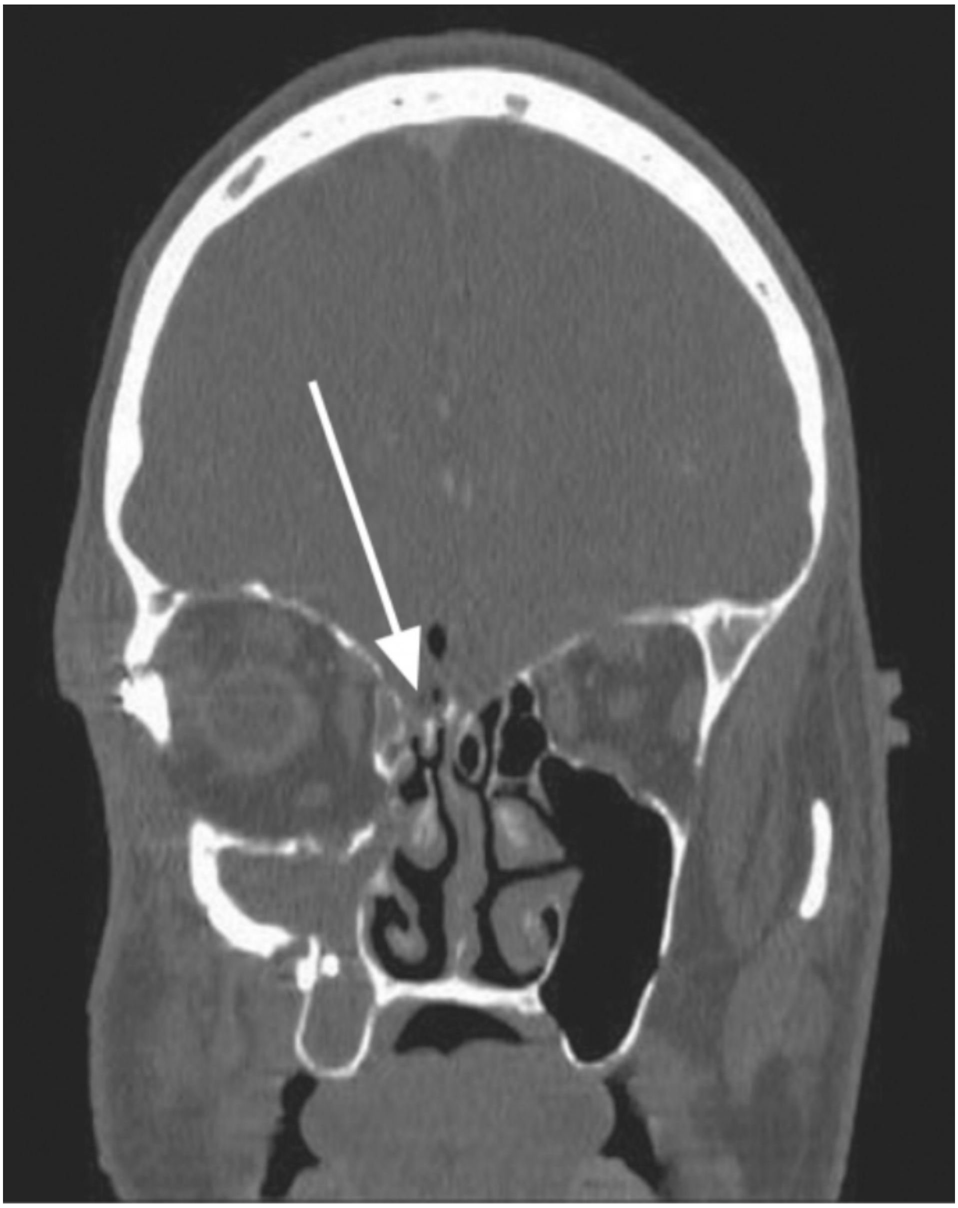

Figure 104-3 Coronal CT scan demonstrating a defect in the right cribriform plate after trauma as the cause of the cerebrospinal fluid fistula *(arrow)*.

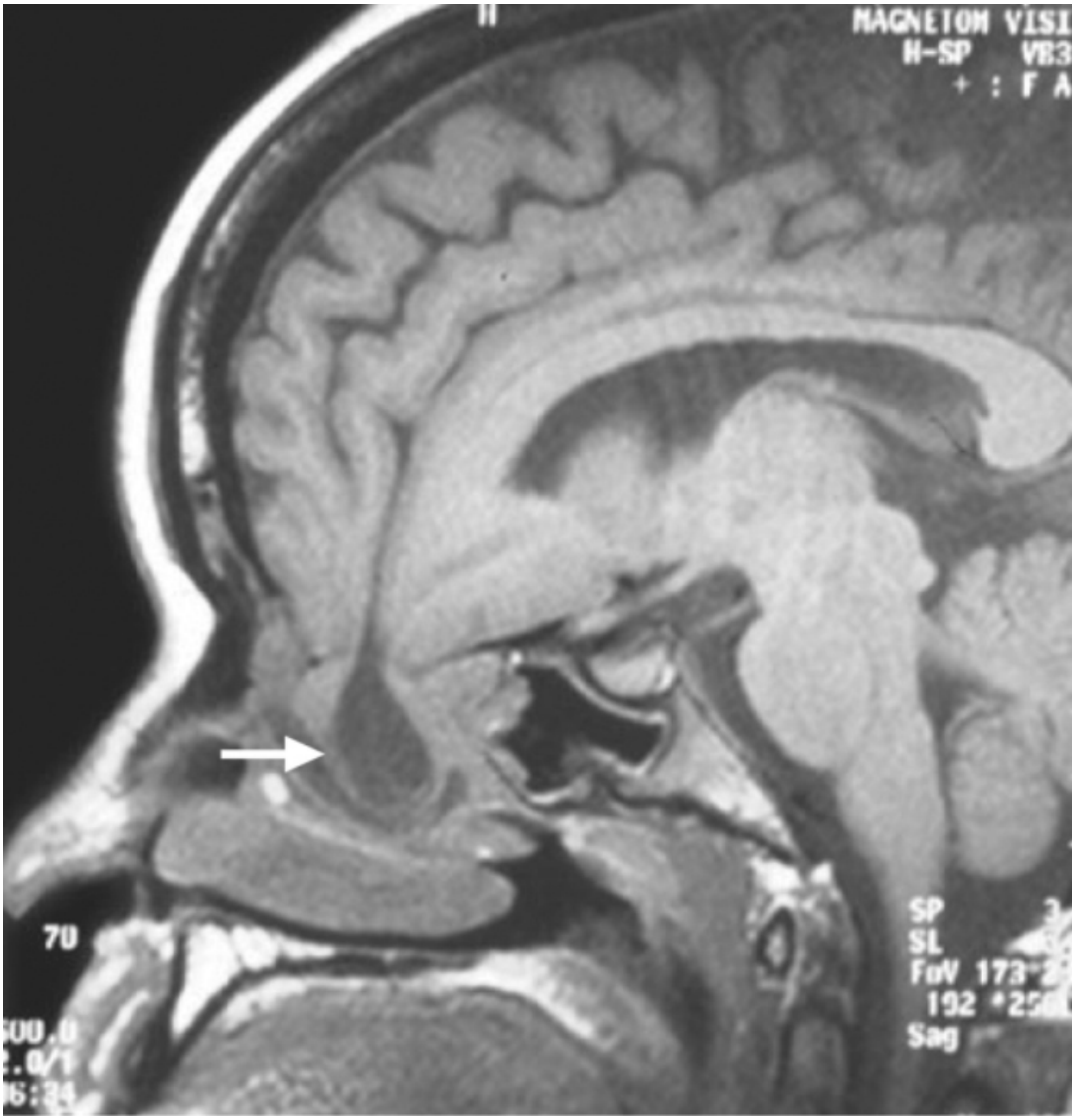

Figure 104-4 Sagittal magnetic resonance image demonstrating large meningoencephaloceles of the anterior cranial base *(arrow)*.

HRCT, however, may not identify small areas of surgical trauma or linear nondisplaced fractures. In this case, HRCT can be used in conjunction with intrathecal injection of contrast material to identify the fistula. CT cisternography has been documented to be both sensitive and reliable. Water-soluble nonionic contrast agents with less associated toxicity, headache, nausea, and arachnoiditis have replaced metrizamide. Identification of the fistula with contrast-enhanced studies, however, requires the presence of an active leak. Intermittent leaks that are temporarily sealed by swelling, inflammation, or brain herniation may yield a false-negative result. Intrathecal injection of saline solution to increase the CSF pressure, “a saline challenge,” enhances the sensitivity of the test.

Others have suggested MR cisternography to complement the information offered by HRCT without the need for an intrathecal contrast agent.[38] In our experience, however, this technique has yielded inconsistent results.

SURGICAL TECHNIQUE

Ethmoid Sinus Roof and Cribriform Plate

If a CSF leak is suspected during endoscopic sinus surgery, the overlying mucosa should be reflected away to closely examine the area and determine the extent of injury. An inlay or onlay free tissue graft may be used to patch the site of injury.

Fascia lata, temporalis muscle, abdominal fat, septal or middle turbinate mucosa or composite grafts, periosteum, and perichondrium are all suitable grafting materials. Whenever possible, the dural edges are undermined with a small elevator and the edges of the graft are tucked between the dura and bone (i.e., epidural inlay graft).

Alternatively, the dura may be separated from the brain, and the inlay graft may be placed in the subdural space. When an inlay graft is not possible because of technical difficulties, because the fistula involves a linear fracture that does not expose the dural defect, or because dissection of the dura may place neurovascular structures at risk, the graft is placed as an onlay over the defect (outside the cranial cavity). Free muscle or fat grafts can also be used as a dumbbell–type plug. Fibrin glue, platelet-rich serum, or other biologic glue may be used to increase the adhesiveness of the muscle or fascia graft.

The graft is supported in place with layers of Gelfoam or Gelfilm (Upjohn Co., Kalamazoo, MI), followed by packing of bacitracin-impregnated gauze or sponge packing. The use of Gelfoam or Gelfilm (or both) prevents adherence of the packing material to the graft, thereby preventing accidental avulsion when the packing is removed 3 to 7 days after surgery.

Alternatively, a vascularized tissue flap may be fashioned with mucoperichondrium from the middle turbinate or septum and harvested transnasally. If the fistula involves the cribriform plate, the mucosa and bone of the lateral aspect of the middle turbinate are removed and the remaining mucoperiosteal flap is rotated to cover the defect or cover a muscle or fascia free graft. Similarly, the mucosa and bone of the lateral aspect of the middle turbinate may be removed to cover a defect of the fovea ethmoidalis. An additional option includes removal of mucosa from a portion of the ipsilateral septum, removal of the corresponding septal cartilage or perpendicular plate of the ethmoid, and rotation of the contralateral mucoperichondrial flap to cover the defect. The flap is then supported by Gelfoam or Gelfilm (or both) and bacitracin-impregnated gauze. This type of septal flap is the preferred flap for defects of the planum sphenoidale that may not be reached by middle turbinate flaps (i.e., defects that are posterior to the posterior attachment of the middle turbinate). Recently, we have been using the Hadad-Bassasteguy flap, which consists of a septal mucoperichondrial/mucoperiosteal flap based on the posterior septal artery. The entire mucoperichondrium/mucoperiosteum may be harvested on one side to cover very large defects of the skull base.

Exposure of the entire defect is essential. In fact, exposure of the defect may be a more important factor in determining success of the repair than its size or site.[39]

Sphenoid Sinus

The sphenoid sinus can be approached transseptally or, preferably, through a direct endoscopic approach such as the one described for pituitary surgery. CSF leaks involving the sphenoid sinus are amenable to a repair that includes obliteration of the sinus. After proper identification of surgical landmarks such as the carotid canal, optic nerve canal, and optic carotid recesses and establishing the extent of the defect identifying the leak, the sinus mucosa is thoroughly removed. Onlay or inlay free tissue grafts are placed and the sinus is then obliterated with abdominal free fat. Anteriorly, the exposed fat is covered with a layer of Surgicel followed by Gelfilm, and the nose is packed with a ½-inch nasal strip that has been impregnated with antibiotic ointment.

An intraoperative CSF leak arising from the sella turcica during pituitary surgery can most often be repaired by obliterating the sella with free fat and reconstructing the floor of the sella with a free bone or cartilage graft harvested during the endoscopic approach.[41] Thus, it does not require postoperative packing or removal of the entire mucoperiosteum of the sinus. The previously described Hadad flap has been very effective in repairing these defects.

The risk of accidental injury to adjacent neurovascular structures is a critical consideration when dealing with defects at the lateral wall of the sphenoid sinus. Adequate visualization of this area is mandatory. Leaks that arise in the lateral recess present technical difficulties as a result of poor visualization after a traditional sphenoidotomy (Fig. 104-5). Repair often requires ligation of the sphenopalatine artery and extension of the sphenoidotomy toward the pterygopalatine fossa.[42] The transpterygoid approach can be combined with endoscopic medial maxillectomy as necessary to allow ade quate range of motion and visualization of even the most lateral of sphenoid defects. Through this extended lateral approach to the sphenoid sinus the endoscopic surgeon is now able to use a zero-degree scope and straight instruments to replace angled scopes and working around corners (Fig. 104-6).

Figure 104-5 Coronal CT scan of the sphenoid sinus depicting the limit of visualization of a traditional sphenoidotomy *(green dots)*. The lateral recess of the sphenoid sinus can be visualized with a transpterygoid approach *(red dots)*. The *diagonal arrows* point to the medial pterygoid plates. The *vertical arrow* points to a bone defect of the right middle cranial fossa.

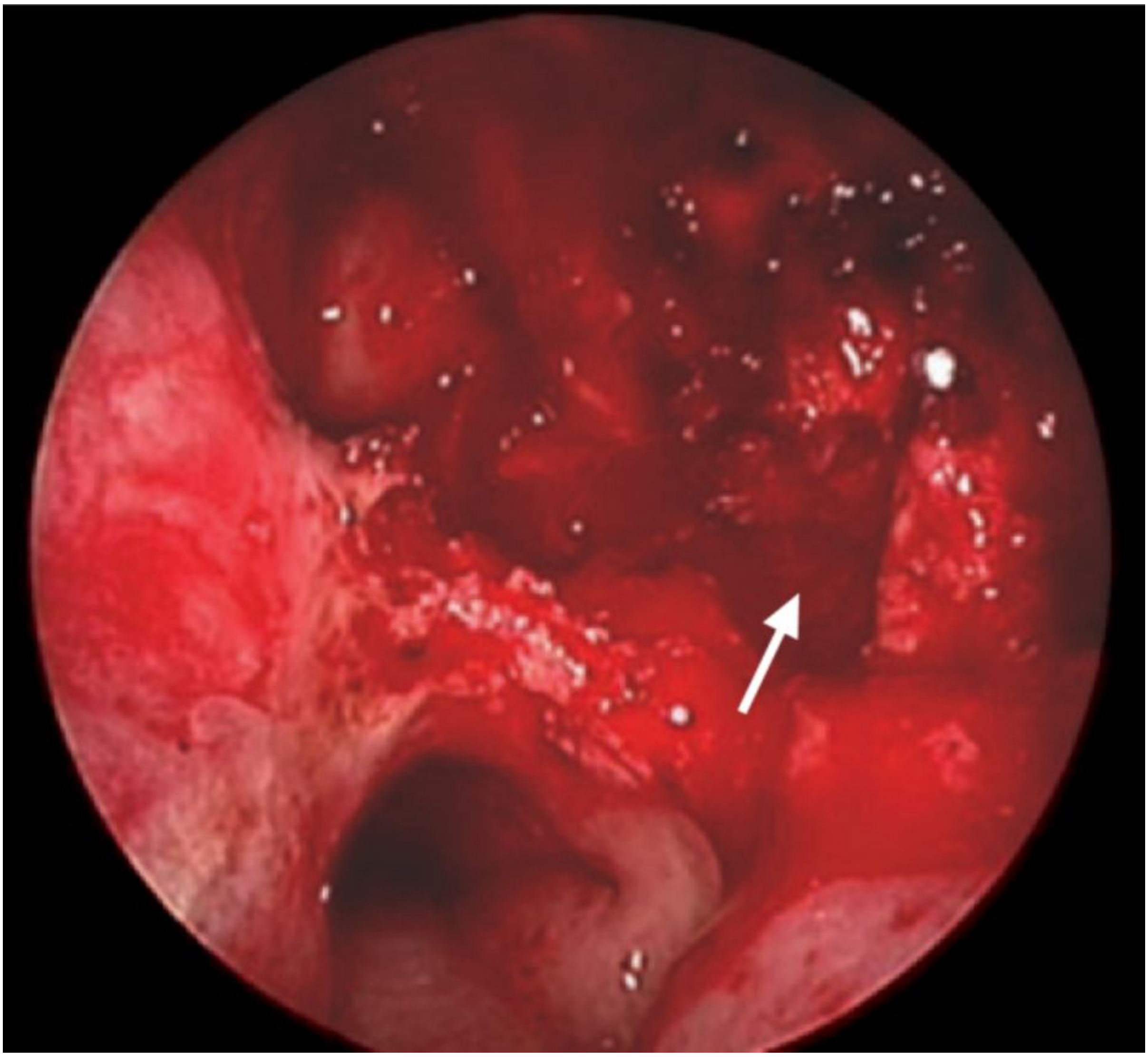

Figure 104-6 Endoscopic view using a zero-degree scope of the left lateral recess of the sphenoid sinus via a transpterygoid approach *(arrow)*.

Frontal Sinus

CSF leaks involving the frontal sinus are usually repaired via a transfrontal sinus approach. The Draf III approach,[43] which involves medial widening of the frontal recesses and removal of the superior nasal septum and inferior frontal sinus septum, may provide exposure of leaks around the frontonasal recess, thus allowing endoscopic repair.

POSTOPERATIVE MANAGEMENT

As previously mentioned, the general principles of managing a CSF fistula include adjunctive measures that may facilitate healing of the repair, including avoidance of activities that raise intracranial pressure, such as straining, leaning forward, or lifting weights greater than 15 pounds. Other measures include bed rest, stool softeners, 30- to 45-degree elevation of the head of the bed, sneezing with an open mouth, and absolute avoidance of nose blowing. “Deep extubation” to prevent straining and coughing is used, and positive pressure mask ventilation is contraindicated.

The use of prophylactic antibiotics for the prevention of meningitis in patients with CSF fistulas con-tinues to be controversial. However, antibiotics are warranted when the patient has an active sinus infection. In patients with indwelling lumbar catheters, prophylactic antibiotics are routinely administered, and although the concept is not universally accepted, antibiotics are believed to decrease the incidence of catheter-related infections.[44] The routine use of antibiotics for traumatic CSF leaks is not of proven efficacy, however, and may select resistant bacteria.[45] Nonetheless, we do favor the use of perioperative prophylactic antibiotics during repair of the CSF leak. Antibiotics are continued until the nasal packing is removed.

A postoperative CT scan without contrast enhancement in the first 24 hours after surgery is important to rule out evidence of intracranial bleeding, parenchymal injury, or tension pneumocephalus. We favor a routine CT scan of the brain, even in the absence of any neurologic deficit.

In our practice, we work in conjunction with a neurosurgical team during the repairs; although we do not consider it necessary in all cases, neurosurgical consultation provides an important perspective on intraoperative and postoperative management, especially on the need for a CSF drain or a shunt. A lumbar drain with a designated amount of CSF removed daily based on CSF production is helpful to control intracranial pressure, but it is used only for patients suspected of having HPH. Overdrainage should be avoided because it creates negative intracranial pressure (i.e., suction effect) that may result in pneumocephalus and promote bacterial contamination of the CSF with resultant meningitis.

High-Pressure Hydrocephalus

Normal intracranial pressure ranges from 5 to 15 cm H_2O when the patient is in the supine position. However, intracranial pressure increases with positional changes, with the Valsalva maneuver, and during rapid eye movement (REM) sleep. Increased intracranial pressure, or HPH, is defined as sustained or intermittent pressure of 20 to 30 cm H_2O.[4–8] Post-traumatic, postsurgical, and postinfectious hydrocephalus can result from obstruction of the arachnoid villi, thus preventing adequate CSF reabsorption with subsequent increased intraventricular pressure. In post-CVA, post-traumatic and postsurgical hydrocephalus, the obstruction arises from subarachnoid hemorrhage, whereas in the case of infection, inflammatory changes are responsible for obstructing the arachnoid villi (Fig. 104-7).[46–51]

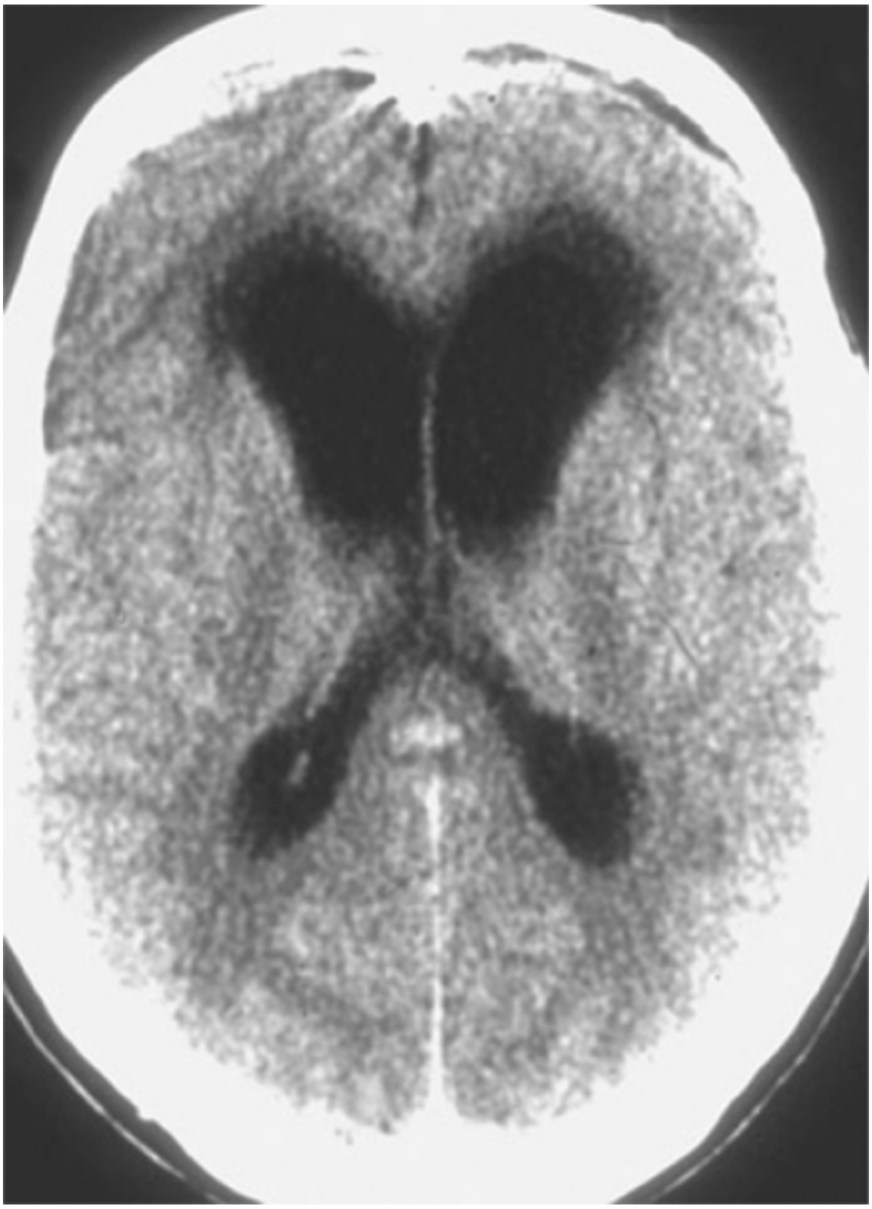

Figure 104-7 Axial CT scan of the head in a patient with high-pressure hydrocephalus and ventriculomegaly of the temporal horns.

Radiation therapy causes vascular changes in the brain and elevation of CSF protein, scarring, and adhesions, all of which may contribute to impairment of CSF reabsorption. Likewise, meningitis causes scarring of the CSF cisterns and absorptive sites, thus leading to chronic hydrocephalus.[46–48]

Patients with recurrent CSF leaks, a past history of head or cranial base trauma (or both), and a history of cranial base surgery and those with "spontaneous" CSF leaks are considered at high risk for HPH and are enrolled in the following protocol (Fig. 104-8).[52] The CSF leak isrepaired via an endonasal endoscopic approach by following surgical principles such as adequate exposure, hemostasis, preparation of the fistula for grafting, and stabilization of the graft. A lumbar spinal drain is inserted intraoperatively or immediately after surgery. Three to 5 days postoperatively, the lumbar spinal drain is clamped, and if no CSF leak is identified for 24 hours, the drain is removed. Twenty-four to 48 hours later, a lumbar puncture is performed to measure CSF opening pressure. A ventriculoperitoneal (VP) shunt is recommended if CSF opening pressure is elevated. IF CSF opening pressure is normal, the patient is discharged home and monitored in our outpatient offices.

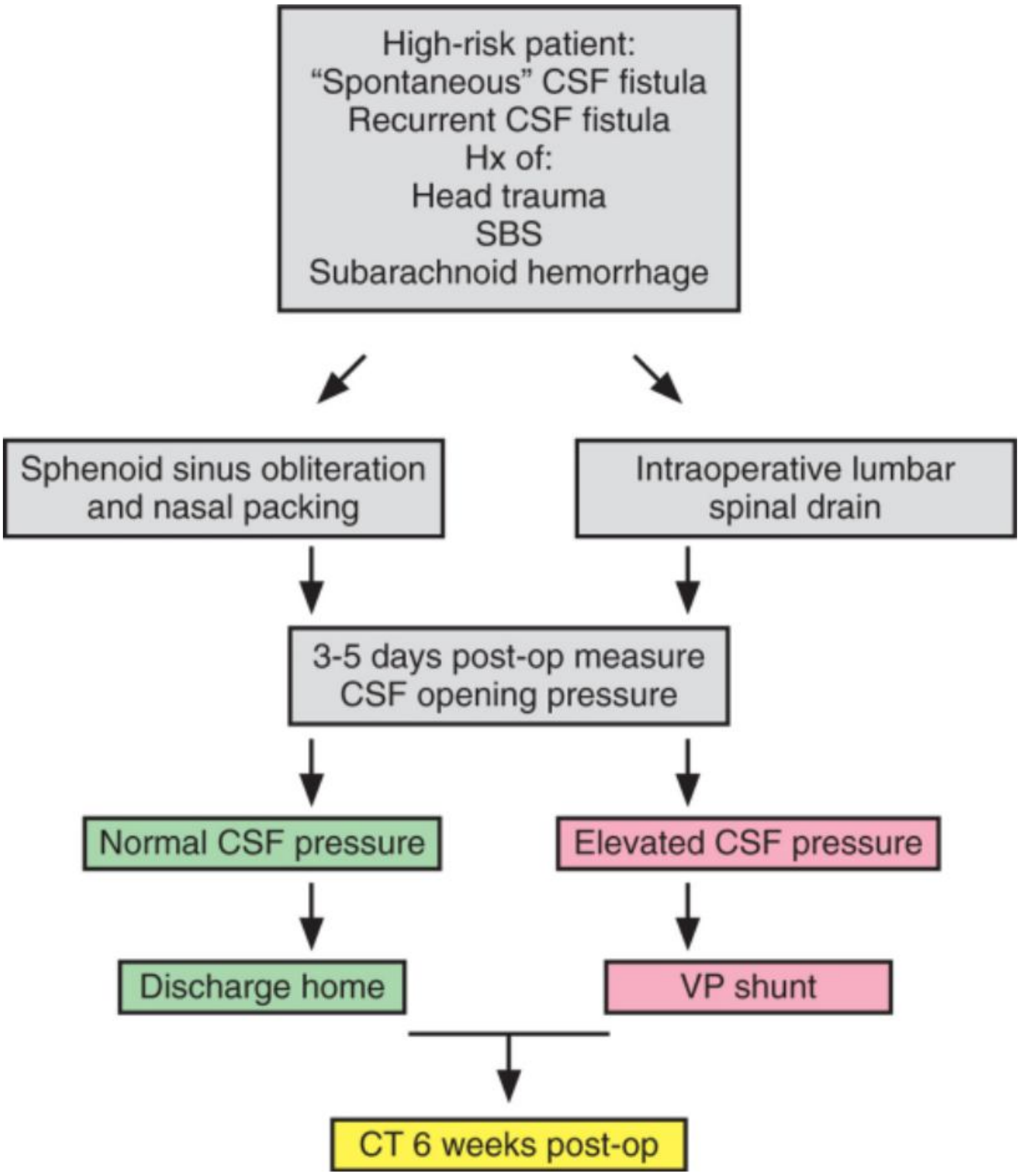

Figure 104-8 Management algorithm for patients at high risk for high-pressure hydrocephalus and failure of repair of a cerebrospinal fluid (CSF) fistula. CT, computed tomography; Hx, history; SBS, skull base surgery; VP, ventriculoperitoneal.

Patients with CSF leaks in the sphenoid sinus are treated by obliteration of the sinus with abdominal fat, which is then stabilized with nasal packing. A postoperative lumbar spinal drain is not required. A lumbar puncture is performed 3 to 5 days postoperatively to measure opening pressure and establish the need for a VP shunt.

In patients with normal CSF opening pressure, a CT scan of the brain is performed 6 weeks after surgical closure. Ventriculomegaly of the temporal horns or transependymal edema will identify patients with HPH who may have escaped identification (false negative).

CSF leaks in patients who did not have any of the previously mentioned high-risk factors are repaired endoscopically and without the use of a spinal drain.

Nasal irrigation with nasal saline solution plus gentle débridement of crusting is started 1 week postoperatively.

PEARLS

- Early identification plus repair of CSF fistulas avoids the development of life-threatening complications.
- Most traumatic CSF leaks will heal with conservative management.
- Endoscopic repair is as successful as open repair given localization of the leak.
- β_2-Transferrin is an extremely reliable marker for CSF leak because it is found only in CSF, perilymph, and vitreous humor.
- A CT cisternogram with water-soluble contrast material is a very useful localizing study in cases in which intrathecal fluorescein is not used.

PITFALLS

- Traumatic CSF fistulas after skull base fracture are often multifocal in nature.
- Failure of CSF fistula repair is frequently a result of inadequate exposure versus inadequate repair.
- High-pressure hydrocephalus is a factor in failure of CSF fistula repair.
- Intrathecal administration of an inappropriate concentration of fluorescein can lead to neurologic sequelae.
- Positive pressure ventilation should be avoided in the immediate postoperative period to prevent pneumocephalus.