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State of the art cerebrospinal fluid leak and encephalocele repair

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The advent of endoscopic sinus surgery has revolutionized the treatment of cerebrospinal fluid (CSF) leaks. Defects traditionally treated with an open approach have now been successfully repaired using either an endoscopic or combined endoscopic/open approach. This process has resulted in decreased operative morbidity and shorter lengths of stay. Various approaches will be described to treat CSF leaks based on their location within the sinonasal cavity. This article is designed to describe the techniques of endoscopic repair of CSF leaks and determine when an open approach will be needed, either as an adjunctive procedure or as the sole treatment of a CSF leak. When appropriate, schematic illustrations have been included to facilitate reader understanding and orientation.

The surgical approach to cerebrospinal fluid (CSF) leak repair has undergone significant evolution throughout the past century. Although multiple technical advancements have contributed to this evolution, perhaps none is so important as the development of endoscopic transnasal surgery. In 1926, Dandy described a closure of a cranio-nasal fistula using a frontal craniotomy approach. This approach was associated with a 60% to 80% success rate and became the standard of care for the period. Through the next 20 years, attempts at less invasive interventions led to the first extracranial approach described by Dohlman in 1948, through a naso-orbital incision. In 1952, Hirsch presaged the era of modern, minimally invasive surgery using a transnasal approach. Subsequent technical advancements in surgical instrumentation led to the development of endoscopic approaches for CSF leak repair in the early 1980s. These approaches have been refined over the past 20 years, and have become the standard of care for the treatment of most leaks and, in experienced hands, are associated with close to a 90% success rate.[1]

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CSF physiology

CSF is normally produced at a rate of 350-500 mL/d. Of this fluid, 70% is produced by the choroid plexus located in the lateral, third, and fourth ventricles, while the remaining 30% of CSF is produced by a combination of capillary ultrafiltration (18%) and water metabolism (12%). Normal CSF flow proceeds from the lateral ventricles through the third and fourth ventricle, finally entering the subarachnoid space surrounding the brain and spinal cord via the foramina of Luschka and Magendie. CSF absorption occurs at the arachnoid villi, which act as valves, directing CSF into the dural sinuses under a pressure gradient of 1.5-7 cm H_2O. The dynamic relationship between CSF production and absorption contributes to a normal intracranial pressure (ICP) of 5-15 cm H_2O, which is subject to a high degree of variability based on age, activity, time of day, and other factors.[1]

Diagnostic work-up and preoperative planning

CSF leaks may be categorized according to a variety of schema. For the purposes of appropriate preoperative work-up and planning, it is useful to classify CSF leaks into those that are spontaneous and those that are potentiated by known or suspected skull base defects. These 2 groups have

represented distinct clinical entities with important implications in operative approach and postoperative treatment.

Potentiated CSF leaks occur as a result of a preexisting dehiscence in the skull base, which may result from a variety of processes. Traumatic CSF leaks occur in the setting of blunt or penetrating trauma, each of which produces idiosyncratic types of injury. One to three percent of blunt head trauma produces a thin skull base fracture, which may be associated with a small dural tear. CSF leaks occur within 48 hours to 3 months of the initial insult[2] and are usually treated conservatively because 70% will close spontaneously. However, these patients must be followed closely because a 30% to 40% risk of ascending meningitis has been found in long-term follow-up.[1] Penetrating head injury may be accidental or iatrogenic in etiology. Although accidental penetrating trauma may result in significant defects in the skull base, iatrogenic injuries are usually less than 2 cm in size.

Iatrogenic CSF leaks are most commonly associated with endoscopic sinus and neurologic surgery. Injuries associated with functional endoscopic sinus surgery are usually localized to the lateral lamella of the cribriform plate and the posterior ethmoid roof. The lateral lamella tends to be thin and, thus, is vulnerable to injury, especially when resecting the middle turbinate close to the skull base. Posterior ethmoid injuries can occur in the setting of a medially and superiorly pneumatized maxillary sinus. This results in a relative reduction in posterior ethmoid height and tends to direct the surgical approach more superiorly, leading to potential inadvertent penetration of the skull base. Transsphenoidal hypophysectomy is the most common neurosurgical procedure associated with CSF leak, secondary to the potential for injury to the sellar diaphragm.[3]

Embryologic malformations may result in large skull base defects with associated CSF leaks and/or encephaloceles. Common locations include the cribriform plate adjacent to the vertical attachment of the middle turbinate, the foramen cecum, and posteriorly along the superior and lateral sphenoid walls. These patients may have multiple areas of dehiscence and represent a rare but difficult population to treat.

Historically, spontaneous CSF leaks and encephaloceles were classified as those for which no etiology could be found. Within the past decade, it has become clear that these patients typically have increased ICPs and may represent a variant of benign intracranial hypertension.[1] These patients can be identified by a syndrome similar to those with benign intracranial hypertension, more specifically, middle-aged, obese females with pulsatile tinnitus, and pressure type headaches. Preoperative imaging may reveal an empty sella, enlarged arachnoid granulations, and dilated skull base foramina. The pathologically increased ICP is transmitted via pulsatile hydrostatic forces to the weakest areas of the skull base, including the cribriform plate, ethmoid roof, and lateral sphenoid recess. This results in CSF leaks, which are usually multiple, associated with a high degree of encephalocele formation, and recur at a higher rate than those of any other etiology (25% to 87% vs 10%).[3-5] Thus, within this patient population, particular attention must be directed toward postoperative treatment of ICP to reduce the rate of recurrence.

Neoplasm represents an interesting clinical entity that can straddle the classification between spontaneous and potentiated CSF leaks. Tumors may result in skull base dehiscence by direct extension or erosion. In addition, radiation and chemotherapy aimed at treatment of the tumor may result in a devascularized skull base defect that may be particularly difficult to repair. Conversely, intracranial masses may obstruct CSF flow, thereby resulting in increased elevated ICP and its concomitant effect on the skull base.

Even if the etiology of a given CSF leak is known, diagnosis and localization remain a clinical challenge. Several tests are available to assist in this endeavor; however, each has specific limitations based on location and rate of leak. The diagnosis of a CSF leak may be established on clinical presentation and by testing nasal secretions for beta-2 transferrin. Patients typically present with unilateral clear rhinorrhea, which may be associated with symptoms of sinusitis and nasal obstruction, headache after nose blowing (resulting from pneumocephalus), and recurrent meningitis. Beta 2-transferrin is a protein found only in CSF, perilymph, and aqueous humor, and provides a sensitive and specific test for the presence of CSF.

Beta-trace protein is the second most abundant protein in CSF and has been defined as prostaglandin D synthase[6,7] Although it was described in 1961,[8] its description in the literature and clinical use have been limited. It has been used in Europe, mainly Austria and Switzerland, as an alternative to beta 2-transferrin in the biochemical detection of CSF. It has 2 advantages over beta 2-transferrin: (1) a decreased processing cost and (2) rapid determination (within 15-20 minutes). However, it is limited in that it is present in very minute quantities in normal serum and requires a slightly larger volume of sample when compared with beta 2-transferrin tested with the Sebia Hydrogel 6 assay.[7] Presently, it is only used in the United States for research purposes only.[6] As a CSF marker, it shares a sensitivity and specificity similar to that of beta 2-transferrin. The combination of its decreased cost and rapid availability makes it an attractive alternative to beta 2-transferrin. Only time will tell the role of this substance in the future.

High-resolution computerized tomography (CT) is of enormous value in the clinical work-up and is almost always indicated. Although this test cannot be used to diagnose an active CSF leak, it can delineate suspicious skull base defects and may be used in conjunction with an image guidance system to direct endoscopic surgical intervention. In contrast, several studies exist that may be used to diagnose and localize simultaneously a CSF leak. Radioactive cisternograms use the strategic placement of intranasal pledgets after the intrathecal injection of a radioactive marker. The location of a CSF leak is then extrapolated from an analysis of the pledgets, which were able to absorb the radioactive marker. Although this test is helpful in the diagnosis of slow or intermittent leaks, its overall use is limited by the fact that secretions tend to mix throughout the nasal cavity, and, thus, the test can only reliably localize a leak to the right or left side.[1]

CT cisternograms also require the instillation of intrathecal contrast but carry the advantage of higher accuracy in