MEnINGOENCEPHALOCELES of the sphenoid sinus are rare and most commonly occur as a result of trauma, iatrogenic injury, or skull base erosion from inflammatory or neoplastic disorders. Spontaneous lesions are exceedingly rare and have been theorized to result from either increased ICP or preformed developmental pathways.1,9,26 Progressive erosion of the skull base in patients with increased ICP and well-pneumatized sphenoid sinuses may result in focal areas of dehiscence and herniation of intracranial contents. Sometimes, an incompetent diaphragma sellae causes the suprasellar arachnoid cistern to prolapse inside the sellar cavity, a radiological condition termed “primary empty sella,” which is usually asymptomatic.22 “Empty sella syndrome” is the pathological variant of a radiologically verified empty sella.15 Errors in the embryological development of the sphenoid bone may also result in congenital defects of the skull base and may present in adulthood as an incidental neuroimaging finding of meningoencephalocele or symptomatically with CSF rhinorrhea and meningitis. The development of the sphenoid bone is complex and involves the fusion of multiple cartilaginous precursors into a single osseous structure. Incomplete fusion of the precursor of the greater wing of the sphenoid with the presphenoid and basisphenoid areas can result in a persistent channel termed the lateral
craniohyphengeal (Sternberg) canal. To date, reports of spontaneous, meningoencephalocele of the lateral sphenoid sinus remain limited given its rarity. In this paper, we present a multiinstitutional case series, describing the demographic characteristics, endoscopic technique, and outcome of therapy in 13 patients with this lesion.

**Methods**

This is a multiinstitutional retrospective review of patients undergoing endoscopic repair of spontaneous meningoencephalocele of the lateral sphenoid sinus between June 1996 and July 2005. Institutional review board approval was obtained prior to the review of the senior authors’ (V.K.A. and T.H.S.) series and was not available at the other participating institutions. Exclusion criteria included a history of trauma, hydrocephalus, and destructive or erosive lesions of the sphenoid sinus or skull base. These diagnoses were excluded based on patient history and preoperative MR imaging. The patients' demographic characteristics, nature and duration of presenting symptoms, and history of prior procedures were reviewed. The diagnostic tests, surgical approaches, and surgical adjuncts were also reviewed, including the use of image guidance, graft materials, and lumbar drains. Perioperative variables, including complications and postoperative outcomes at most recent follow-up, were used as the main outcome measures. Major complications were defined as vascular injury, neural injury, postoperative CSF leak, and postoperative meningitis. The follow-up evaluation of all patients included clinical history and endoscopic examination of the surgical cavity.

**Surgical Technique**

The endoscopic approaches that were used to approach the lateral sphenoid sinus included transnasal, transthamoid, and transpterygoid approaches. The technique of the senior authors (V.K.A. and T.H.S.) is briefly described here.

Intrathecal fluorescein is injected following the induction of general anesthesia and premedication with dexamethasone and diphenhydramine. This assists throughout the case in 1) identifying the area of the skull base defect, 2) gauging the volume of the CSF leakage, and 3) determining the effectiveness of the repair, as has been previously described. Image guidance is useful in various aspects of the procedure, including delineating the anatomy of the sphenoid sinus, anterior skull base, encephalocoele margins, optic nerve canal, and ICA. The identification of the ICA may also be aided by use of an endoscopic Doppler probe. Long-handled drills are used in the areas of dense bone in the various approaches involving the sphenoid sinus (sphenoid rostrum) and ptterygoid fossa (lacrimal bone, ptterygoid plate). Angled endoscopes are required for visualization of the encephalocoele in the lateral sphenoid wall. Placement of graft material is technically challenging and is aided by angled instruments including forceps, curettes, and probes.

A submucosal resection of the cartilaginous nasal septum and vomer is performed to provide graft material and to improve access to the sphenoid cavity both intraoperatively and at follow-up visits. Exposure of the meningoencephalocele and reconstruction of the skull base defect are achieved through 1 of 3 corridors based on the degree of lateral pneumatization. The corridors—in increasing order of lateral exposure—are the transnasal, transthamoid, and transtpterygoid approaches. The transnasal approach involves enlargement of the natural sphenoid ostia in the superior meatus. The creation of a common cavity incorporating the 2 ostia and the sphenoid rostrum is appropriate in patients with relatively little sphenoid pneumatization. The placement of angled endoscopes in the contralateral side allows for improved visualization of the lateral recess during surgery. The transthamoid approach involves anterior and posterior ethmoidectomy and creation of a large transthamoid sphenoidotomy. This may be combined with the transnasal approach and provides additional lateral exposure.

The transpterygoid approach may be required in cases of extensive lateral pneumatization (Fig. 1). Similar to the transthamoid approach, the procedure begins with an anterior and posterior ethmoidectomy and transthamoid sphenoidotomy. A maxillary antrostomy is performed and the mucosa of the posterior wall is reflected off the bone. The palatine bone is dissected and defined. The anterior perpendicular process of the palatine bone is removed and the sphenopalatine artery is mobilized. The posterior wall of the maxillary sinus adjoining the palatine bone is drilled and the sphenopalatine vascular bundle is identified, dissected, and either cauterized or ligated. Additional drilling of the palatine bone posterior to the sphenopalatine foramen exposes the ptterygomaxillary fossa. The remaining neurovasculature structures of the fossa are preserved and gently reflected laterally. The ptterygoid process is identified and drilled open to expose the lateral recess of the sphenoid.

The herniating tissue can either be preserved and pushed intracranially or coagulated with bipolar forceps and amputated flush with the osseous defect. Although the neural tissue is considered nonfunctional, the former approach may help minimize the briskness of the CSF leak in cases involving large lesions. The osseous skull base is then reconstructed with multiple layers including a fat/Gelfoam layer to fill the intracranial dead space, a nonporous fascia layer and a osseous buttress (septal vomer) placed as an onlay on top of the fascia, and tissue sealant placed around the wound edges. The fascia layer may be placed circumferentially underneath the osseous skull base as an underlay graft if the defect is small. Alternatively for larger defects, the fascia layer may be placed to cover the entire skull base defect on the sphenoid side of the lesion and then supported by an osseous graft which is placed on top of the fascia but sunk into the skull base defect. In this so-called “gasket-seal” closure, the fascia graft is intracranial in its central aspects but covers the sinonasal circumference of the skull base defect at the margins (Fig. 1F). The sphenoid cavity itself is not obliterated and is left open to promote physiological function and postoperative examination.
The medical records of 13 patients who met inclusion criteria were reviewed. The patients’ demographic characteristics are presented in Table 1. The mean age (± SD) of the cohort was 57.1 ± 14.3 years (range 36–78 years) at the time of surgery; 8 (62%) of the patients were women and 5 (38%) were men. The encephalocele was on the left side in 8 patients (62%) and on the right side in 5 (38%). There were no occurrences of bilateral lesions in this series. The majority of patients (11 patients, 85%) presented initially with unilateral clear rhinorrhea that was clinically consistent with CSF leakage. Beta-2 transferrin testing of the nasal fluid was performed in 3 cases (23.3%), with positive results in all 3. This testing was based on institutional availability. Other clinical manifestations included chronic headache in 10 patients (77%) and a history of meningitis in 2 (15%). The mean duration of symptoms at the time of surgery was 19 ± 31 months (range 3 weeks–9 years). Three patients (23%) had undergone a total of 6 prior procedures. All patients underwent a combination of diagnostic imaging studies including CT in 3 patients (23%), MR imaging (Fig. 2) in 13 (100%), and CT cisternography in 12 (92%).

The surgical variables and outcomes are described in Table 2. All patients underwent an endoscopic approach to the lateral sphenoid sinus as described in Methods. Five patients (38.5%) underwent a transnasal approach, 3 (23.1%) underwent a transpterygoid approach, and 5 (38.5%) underwent a transethmoid approach. Based on institutional availability at the time of the procedure, 7 patients (54%) underwent image-guided surgery using CT-based data sets. Intrathecal fluorescein injection was used in 9 patients (69%) (Fig. 3). As described in Table 1.
2, a variety of autologous and synthetic graft materials were used to repair the skull base defect. Tissue sealant was used in 12 patients (92%). A planned lumbar drain was used in 8 (62%). The mean duration of the procedure was 146 ± 28 minutes (range 110–200 minutes) and the mean duration of the hospitalization was 4.5 ± 1.7 days (range 2–7 days). At most recent follow-up (mean 4.7 ± 3.3 years after surgery, range 8 months–12.1 years), all patients were free of leaks, based on clinical history and endoscopic examination. Two patients (15%) had a post-operative CSF leak; one of the leaks stopped spontaneously and the other closed following revision endoscopic surgery. Both patients had initially undergone an image-guided, transethmoid approach and closure with conchal cartilage and temporalis fascia without the use of lumbar drainage. One patient (8%) experienced postoperative meningitis in the early postoperative period. This was treated successfully with intravenous antibiotics without sequelae. One patient experienced postoperative facial paresthesia.

**Discussion**

Meningoencephaloceles of the sphenoid sinus are rare and can arise from congenital, traumatic and erosive etiologies. The latter category includes locally destructive processes related to neoplastic and inflammatory diseases in addition to increased ICP. Meningoencephaloceles of the anterior skull base can also be classified based on the location as described by Van Nouhuys and Bruyn: sphenoorbital, sphenethmoid, transethmoid (cribiform), sphenomaxillary, and transsphenoid. Medial, parasellar lesions are relatively more common and may occur in conjunction with the “empty sella syndrome.” Meningoencephaloceles of the lateral recess are rarer and are more likely to occur in the 26–40% of patients that have extensive lateral pachymatization of the sphenoid sinus into pterygoid process. A defect in the thin bone of the middle cranial fossa in this area may result in CSF leakage and herniation of the temporal lobe into the sphenoid sinus.

Congenital meningoencephaloceles of the lateral sphenoid sinus are exceedingly rare and have been sporadically reported in the literature. These lesions theoretically arise from developmental errors that occur during the embryogenesis of the sphenoid bone (Fig. 4). Most of the sphenoid bone is formed from the ossification of cartilaginous precursors and only a small portion is formed from membranous bone. The cartilaginous precursors arise from mesoderm and neural crest cells beginning in the 4th week of development. These develop into 5 discrete, independent cartilaginous areas: the anterior and posterior sphenoid, lesser wing, greater wing, and pterygoid process. The individual portions undergo ossification from multiple endochondral ossification centers, beginning at the 13th week of fetal development, followed by fusion into a single bone. Incomplete fusion of the greater wings with the presphenoid and basisphenoid can result in a persistent tract, termed the lateral cranio-pharyngeal canal. This was first described in 1888 by Sternberg and is alternatively referred to as the Sternberg canal. This is differentiated from the central cranio-pharyngeal canal that arises from the midportion of the floor of the hypophyseal fossa and extends inferiorly to terminate behind the rostrum. Persistence of this canal may also result in a CSF leak and meningoencephalocele but is easily distinguished from the lateral canal by its midline location.

Evaluation of spontaneous lateral meningoencephaloceles is challenging and requires a thorough patient history and adjunctive diagnostic tools. The investigation focuses on potential inflammatory and neoplastic disorders and increased ICP. Additionally, any history of trauma, meningitis, rhinorrhea, and prior surgeries should be elic-

### TABLE 1: Summary of demographic and clinical data in 13 patients*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Pt Age at Op (yrs)</th>
<th>Sex</th>
<th>Clinical Presentation</th>
<th>Side of Leak</th>
<th>Prior Procedures</th>
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<tbody>
<tr>
<td>1</td>
<td>61</td>
<td>F</td>
<td>rhinorrhea, headache</td>
<td>lt</td>
<td>none</td>
</tr>
<tr>
<td>2</td>
<td>37</td>
<td>F</td>
<td>rhinorrhea, headache</td>
<td>lt</td>
<td>VP shunt</td>
</tr>
<tr>
<td>3</td>
<td>78</td>
<td>F</td>
<td>headache, meningitis</td>
<td>rt</td>
<td>none</td>
</tr>
<tr>
<td>4</td>
<td>44</td>
<td>F</td>
<td>rhinorrhea, headache</td>
<td>rt</td>
<td>none</td>
</tr>
<tr>
<td>5</td>
<td>36</td>
<td>F</td>
<td>rhinorrhea, headache</td>
<td>lt</td>
<td>none</td>
</tr>
<tr>
<td>6</td>
<td>66</td>
<td>F</td>
<td>headache, meningitis</td>
<td>lt</td>
<td>LP shunt</td>
</tr>
<tr>
<td>7</td>
<td>73</td>
<td>F</td>
<td>rhinorrhea, headache, meningitis</td>
<td>rt</td>
<td>none</td>
</tr>
<tr>
<td>8</td>
<td>44</td>
<td>F</td>
<td>rhinorrhea, headache</td>
<td>lt</td>
<td>none</td>
</tr>
<tr>
<td>9</td>
<td>59</td>
<td>M</td>
<td>rhinorrhea</td>
<td>lt</td>
<td>endoscopic closure × 3, craniotomy</td>
</tr>
<tr>
<td>10</td>
<td>56</td>
<td>M</td>
<td>rhinorrhea</td>
<td>lt</td>
<td>none</td>
</tr>
<tr>
<td>11</td>
<td>73</td>
<td>M</td>
<td>rhinorrhea</td>
<td>lt</td>
<td>none</td>
</tr>
<tr>
<td>12</td>
<td>47</td>
<td>M</td>
<td>rhinorrhea, headache</td>
<td>rt</td>
<td>none</td>
</tr>
<tr>
<td>13</td>
<td>68</td>
<td>M</td>
<td>rhinorrhea, headache</td>
<td>rt</td>
<td>none</td>
</tr>
</tbody>
</table>

* LP = lumboperitoneal; Pt = patient; VP = ventriculoperitoneal.
Endoscopic lateral sphenoid meningoencephalocele

The relative advantages and disadvantages of various diagnostic modalities, including biochemical (beta-2 transferrin) and imaging studies (CT, MR imaging, and cisternography), have been previously described. Identification on imaging of a dehiscence in the lateral sphenoid sinus with herniation of tissue from the temporal lobe is suggestive and is confirmed at the time of surgery.

The management of patients with spontaneous meningoencephalocele of the lateral sphenoid sinus is controversial in the literature. The relative efficacies of the various aspects of surgical repair, including surgical approaches, reconstruction methods, and surgical adjuncts (lumbar drainage, image guidance, fluorescein), are incompletely compared. Prior reports have described surgery for encephaloceles in this area through both craniotomy and endoscopic approaches.

Although craniotomy is associated with retraction of neurovascular structures, it has been advocated by some authors as allowing direct visualization and repair. The relative advantages of the endoscopic approach described in this manuscript include its noninvasive nature and excellent visualization. The use of both angled endoscopes and fluorescein enhances identification of the skull base defect in these cases, without any brain retraction. The utility of endoscopic approaches to this area is supported by other reports.

Al-Nashar et al. provided a detailed anatomical and surgical description of 7 CSF leaks (all successfully repaired) and 5 tumors of the lateral sphenoid sinus. In a recent series of 15 patients with meningoencephaloceles of the lateral sphenoid sinus, Castelnuovo et al. reported no postoperative CSF leaks following endoscopic closure with a multilayered graft.

The challenge of the endoscopic approach, however, is related to the lateral location of the meningoencephalocele. The different endoscopic approaches mentioned in this manuscript are each associated with relative advantages and disadvantages. Although considered relatively

**TABLE 2: Surgical variables and outcomes**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Approach</th>
<th>Graft Material</th>
<th>Lumbar Drain</th>
<th>Duration of Op (mins)</th>
<th>LOS (days)</th>
<th>Complications</th>
<th>Duration of FU (yrs)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>transnasal</td>
<td>NS, FL</td>
<td>no</td>
<td>120</td>
<td>2</td>
<td>perioperative meningitis</td>
<td>11.3</td>
</tr>
<tr>
<td>2</td>
<td>transnasal</td>
<td>NS, FL</td>
<td>yes</td>
<td>110</td>
<td>7</td>
<td></td>
<td>12.1</td>
</tr>
<tr>
<td>3</td>
<td>transnasal</td>
<td>NS, FL</td>
<td>no</td>
<td>130</td>
<td>3</td>
<td></td>
<td>0.7</td>
</tr>
<tr>
<td>4</td>
<td>transpterygoid</td>
<td>FL, MT</td>
<td>yes</td>
<td>160</td>
<td>4</td>
<td></td>
<td>5.0</td>
</tr>
<tr>
<td>5</td>
<td>transpterygoid</td>
<td>FL, MT</td>
<td>yes</td>
<td>150</td>
<td>5</td>
<td></td>
<td>5.8</td>
</tr>
<tr>
<td>6</td>
<td>transpterygoid</td>
<td>FL, MT</td>
<td>yes</td>
<td>180</td>
<td>5</td>
<td>facial paresthesia</td>
<td>3.0</td>
</tr>
<tr>
<td>7</td>
<td>transethmoid</td>
<td>NS, AF</td>
<td>yes</td>
<td>120</td>
<td>6</td>
<td></td>
<td>3.2</td>
</tr>
<tr>
<td>8</td>
<td>transethmoid</td>
<td>AC, TF</td>
<td>no</td>
<td>200</td>
<td>5</td>
<td>persistent leak requiring revision, endoscopic closure</td>
<td>3.6</td>
</tr>
<tr>
<td>9</td>
<td>transethmoid</td>
<td>AC, TF</td>
<td>yes</td>
<td>165</td>
<td>4</td>
<td></td>
<td>2.9</td>
</tr>
<tr>
<td>10</td>
<td>transethmoid</td>
<td>AC, TF</td>
<td>no</td>
<td>130</td>
<td>2</td>
<td>persistent leak that closed spontaneously</td>
<td>3.2</td>
</tr>
<tr>
<td>11</td>
<td>transethmoid</td>
<td>NS</td>
<td>no</td>
<td>180</td>
<td>3</td>
<td></td>
<td>3.3</td>
</tr>
<tr>
<td>12</td>
<td>transnasal</td>
<td>AF, DS</td>
<td>yes</td>
<td>130</td>
<td>6</td>
<td></td>
<td>4.3</td>
</tr>
<tr>
<td>13</td>
<td>transnasal</td>
<td>MT, DS</td>
<td>yes</td>
<td>125</td>
<td>7</td>
<td></td>
<td>3.2</td>
</tr>
</tbody>
</table>

* All patients were free of leak at follow-up. Abbreviations: AC = auricular cartilage; AF = autologous fat; DS = dural substitute; FL = fascia lata; FU = follow-up; LOS = length of hospital stay; MT = middle turbinate; NS = nasal septum (vomer); TF = temporalis fascia.
safe, these approaches do require significant experience in advanced endoscopic techniques. Additionally, there is potential for significant complications, including persistent CSF leak, neurovascular injuries, and meningitis, as noted in this paper.

Determination of the ideal approach is based on various factors, including the degree of lateral sphenoid pneumatization, location and size of the meningoencephalocele, and ability to perform an adequate skull base repair through a given exposure. In most cases, the final approach is not determined until the time of surgery. Given the relatively lower degree of complexity, the transnasal and transthyroid approaches are explored initially. However, these approaches may not provide adequate lateral exposure as supported by the occurrence of postoperative CSF leak in 2 patients who underwent the transthyroid exposure. The authors advocate the transpterygoid approach in far lateral cases given the improved visualization and direct access of this technique. The entire circumference of the lateral skull base can be identified and dissected and the wide operative field allows for adequate reconstruction.

The philosophy described in this current paper differs from prior reports that rely on a small sphenoidotomy and obliteration of the involved sinus with a fat graft without a specific skull base reconstruction. The issue of persistent CSF leak and mucocele formation associated with this approach has resulted in our advocating the creation of a large sphenoidotomy and a multilayered reconstruction as allowing for a more physiological and effective approach.

The current report represents a multiinstitutional series of patients presenting with spontaneous meningoencephalocele of the lateral sphenoid sinus. Our working hypothesis for the etiology of this lesion in this cohort is congenital persistence of the lateral cranioopharyngeal canal. This is based on the lateral location of a large canal, which differs from the small pits on the sphenoid roof that have been described in patients with increased ICP. Additionally, the lack of subjective and objective evidence for other etiologies supports the possibility of a congenital lesion. However, the possibility that this may be a variant of meningoencephalocele secondary to increased ICP is acknowledged and would be further evaluated by assessment of opening CSF pressure.

The large number of patients with this rare lesion presented in this report is based on the pooling of data from several centers over a 9-year period and allows for several important findings on descriptive analysis. Despite the possible developmental anomaly of the lesion, the majority of patients presented in adulthood, suggesting either a prolonged silent state with a spontaneous presenting event or the enlargement of the defect over a long period of time until herniation of a critical volume of tissue occurred. The latter phenomenon may result from the intracerebral pulsations pushing on the meningoencephalocele. The clinical presentations in our series included unilateral rhinorrhea, chronic headache, and meningitis, and the lesion was uniformly evident on imaging studies. The challenge in closing these defects is evidenced by the history of failed prior attempts in 25% of our patients. However, the 85% closure rate following the authors’ initial attempt and 100% final closure rate following revision endoscopic surgery in 1 patient highlights the efficacy.
of the endoscopic approach. Of note, the 2 patients who had persistent postoperative CSF leak had undergone a transethmoid approach without lumbar drainage.

The use of lumbar drainage in the management of patients with anterior skull base meningoencephalocele remains controversial. The theoretical advantages include decompression of the area of the reconstruction and the potential for a decrease in the incidence of postoperative CSF leaks. However, the impact on overall outcomes remains poorly described, and some patients in this series underwent successful closure without the use of lumbar drainage, supporting the primary importance of a robust closure and not postoperative lumbar drainage. The potential complications associated with lumbar drainage—including spinal headaches, overdrainage with cerebrospinal fluid (CSF) leak, and patient comorbidities associated with poor wound healing (chronic steroid use, diabetes, history of radiation therapy to the skull base). However, this is based largely on cumulative experience and not on adequately designed studies. Finally, the fact that lumbar drainage was not used in the 2 cases of postoperative CSF leak in our series raises the possibility that a higher rate of closure could be achieved with routine drainage.

There are several limitations to the current study. Descriptions of rare entities, including spontaneous meningoencephalocele of the lateral sphenoid sinus, are inherently limited by the small numbers of patients and inability to perform statistical analysis of study parameters. Additionally, the pooling of patients from multiple centers is associated with heterogeneity in the diagnostic and treatment algorithms. Finally, continued long-term follow-up is required to monitor for delayed leak in this cohort. Future case series are required to fully elucidate the clinical, diagnostic, and management issues associated with this disorder.

**Conclusions**

Errors in the complex embryological development of the sphenoid bone may result in focal areas of incomplete osseous fusion and herniation of intracranial contents into the sphenoid sinus. Spontaneous meningoencephalocele of the lateral sphenoid sinus may occur secondary to either increased ICP or persistence of the lateral craniohypophyseal canal. The clinical manifestations may be insidious and include CSF rhinorrhea, headache, and meningitis. The anatomical location in the lateral sphenoid recess presents unique challenges in the surgical approach to this lesion. The visualization and exposure afforded by the endoscopic approach allows for a high rate of successful closure. In many circumstances, a direct transnasal or transethmoid approach may be inadequate, in which case, the transpterygoid approach is preferable.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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**References**


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