Spontaneous sphenoid lateral recess cerebrospinal fluid leaks arise from intracranial hypertension, not Sternberg's canal

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Background: **Spontaneous cerebrospinal fluid (CSF) leaks/encephaloceles are proven to be associated with intracranial hypertension by objective measurements of CSF pressure during or following endoscopic repair. A common area of involvement is a pneumatized lateral recess of the sphenoid (LRS) sinus, where prolonged intracranial pressures lead to arachnoid pits and subsequent development of skull-base defects. Even though the LRS is never present at birth, a "congenital" cause of these leaks due to a persistent Sternberg's (lateral craniopharyngeal) canal continues to be erroneously perpetuated in the literature. The objective of this study was to eliminate the myths defining these leaks as congenital in nature.**

Methods: **Evaluation of LRS CSF leaks present within a multiinstitutional case series was performed. Data regarding demographics, body mass index (BMI), radiologic evaluation of intracranial hypertension, and direct intracranial pressure measurements (when available) were collected.**

Results: **Data evaluation identified 77 LRS CSF leaks in 59 patients (mean age 52 years). Obesity was present in 83% of individuals (mean BMI 36) and 81% were females. Radiologic evidence of intracranial hypertension (eg, empty sella, dilated optic nerve sheaths, and scalloped/attenuated bone) was present on 96% of preoperative computed tomography (CT) and/or magnetic resonance imaging (MRI) scans. Opening or postsurgical lumbar drain or ventriculostomy pressure measurements were elevated in 95% of patients (mean 27.7; range, 9–50 cmH_2O).**

Conclusion: **This study provides objective evidence that LRS CSF leaks are secondary to erosions from intracranial hypertension and refutes the myth regarding a congenital origin from Sternberg's canal.** Copyright © 2014 ARS-AAOA.

Key Words:
Sternberg's canal; lateral craniopharyngeal canal; CSF leak; spontaneous; sphenoid sinus; lateral recess; endoscopic surgery; intracranial hypertension

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The successful treatment of spontaneous cerebrospinal fluid (CSF) leaks and encephaloceles has been recognized as contingent upon the management of increased intracranial pressure (ICP). Traditionally, endoscopic repair success rates for idiopathic or spontaneous CSF leaks have ranged from 25% to 87% over time, with success rates increasing to over 90% with management of increased ICP.[1–4] Clinical symptoms of headache and pulsatile tinnitus, patient demographics such as obesity and female gender, and radiographic associations with prolonged intracranial hypertension (eg, empty sella, attenuation or scalloping of skull base, and arachnoid pits) increases the suspicion of elevated ICP. Intracranial hypertension is often confirmed by opening pressure during lumbar puncture or ventriculostomy placement at time of endoscopic repair or during postoperative ICP evaluations.[4] The literature varies regarding the most frequent location of spontaneous CSF leaks, including the cribriform plate, the pneumatized lateral recess of the sphenoid (LRS) sinus, or posterior table of the frontal sinus.[4–6]

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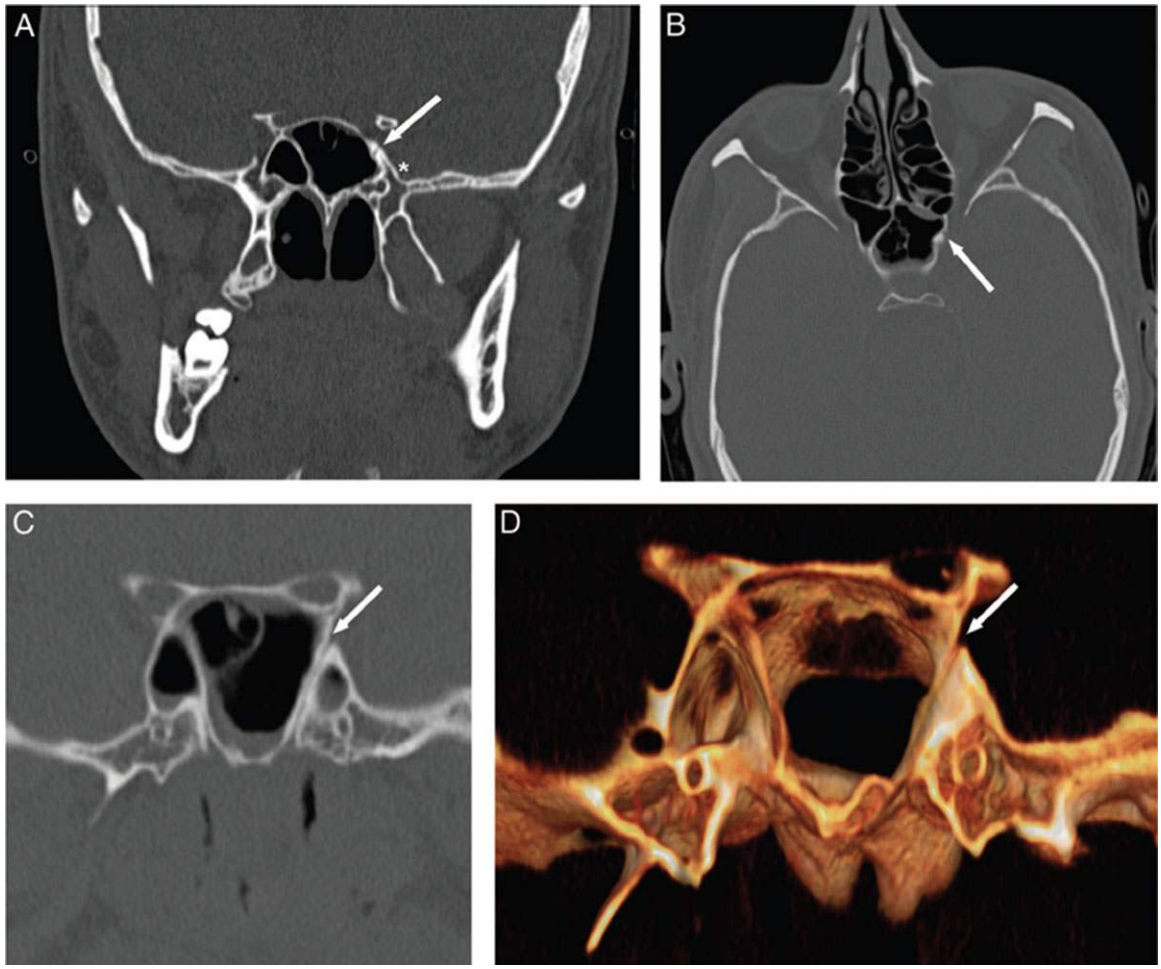

FIGURE 1. High-resolution CT scan of a patient with imaging findings consistent with Sternberg's description of the lateral craniopharyngeal canal. (A) Coronal and (B) axial images reveal a bony defect (arrows) located medial to the foramen rotundum and superior orbital fissure that opens in the region of the cavernous sinus. On (C) oblique CT and (D) three-dimensional reconstruction a canal is visualized in this area. The location of the second division of the trigeminal nerve (*) is highlighted on (A) the coronal image (reproduced with permission from Ref. 10). CT = computed tomography.

Despite findings of elevated ICP in the majority of patients with LRS encephaloceles/CSF leaks and the lack of sphenoid sinus pneumatization at birth, a congenital origin of CSF leaks due to persistent lateral craniopharyngeal canal (Sternberg's canal) continues to be erroneously propagated in the literature.[7,8] Maximilian Sternberg first posited the existence of a *lateral* craniopharyngeal canal in 1888 due to a failure of fusion in the alisphenoid, basisphenoid, and presphenoid ossification centers.[9] As originally described, the canal extends from the junction of the body of the sphenoid bone and the posterior root of the lesser sphenoid wing just medial to the superior orbital fissure inferiorly to connect with either the pharynx at the processus vaginalis or into the lateral wall of the sphenoid sinus. Although cadaver studies have claimed the canal is present in up to 4% of cadaveric skulls, only 1 defect/canal consistent with Sternberg's description was identified in 1000 sphenoid bones on evaluation of consecutive computed tomography(CT) scans (Fig. 1).[10] Because the canal is located medial to the superior orbital fissure, encephaloceles and CSF leaks lateral to the maxillary nerve (cranial nerve V2) cannot be considered to be the etiology of LRS.[9] A review of manuscripts describing management of "Sternberg's" CSF leaks published since our clarification in 2009 involved mostly middle-aged females with concomitant empty sellas and arachnoid pits, and all defects are lateral to V2.[7,11–18]

The purpose of this study was to provide objective evidence supporting intracranial hypertension as the etiology of LRS CSF leaks and encephaloceles and to eliminate the myths defining these leaks as congenital in nature.

Materials and methods

Institutional Review Board approval at the University of Alabama-Birmingham, Medical University of South Carolina, and University of Pennsylvania was obtained prior to data collection. Spontaneous LRS CSF leaks and encephaloceles managed by 3 rhinologists (RJS, JNP, and BAW) were included in the analysis. Data regarding age, gender, body mass index (BMI), radiologic evaluation of intracranial hypertension, and direct intracranial pressure measurements by lumbar puncture or ventriculostomy (when available)

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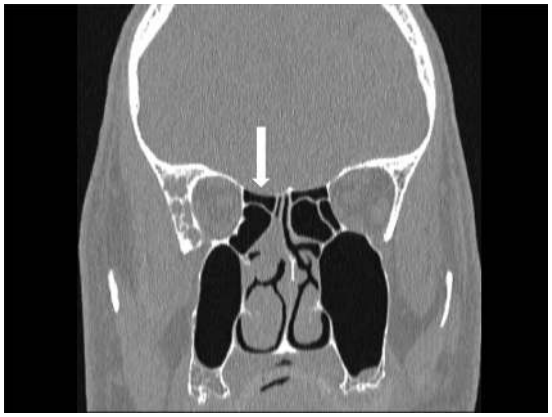

FIGURE 2. Attenuation of the posterior ethmoid roof (arrow) is noted in a patient with bilateral LRS encephaloceles. Note the very thin and even absent bone. LRS = lateral recess of sphenoid.

were collected. Radiological evaluation of CT and magnetic resonance imaging (MRI) scans consisted of attenuated anterior or middle cranial fossa bone, arachnoid pits (at least 2 mm deep), skull-base defects, empty sella, dural ectasia (eg, dilated Meckel's cave), and abnormalities of the optic nerve sheath complex (eg, vertical tortuosity, dilated optic nerve sheaths, and scleral flattening). Attenuation of cranial bone was identified as no visible bone on CT bone algorithm windows at a separately identifiable location from the skull-base defect (Fig. 2).

When lumbar puncture (or ventriculostomy) was performed, the patient was placed in the lateral decubitus position and the neurosurgical or anesthesiology team obtained an opening pressure immediately after insertion. In some cases, intracranial pressure monitoring was performed via lumbar (or ventricular) drain, as described.[4,19,20] CSF was diverted at an average of 10 mL/hour until the 2nd to 4th postoperative day (depending on surgeon preference) when the drain was clamped for 4 to 6 hours to obtain equilibrated ICP measurements. ICP was measured with manometry attached to the lumber drain (or by ventriculostomy apparatus) and zeroed at the spine (or external auditory canal) in the lateral decubitus position with the body level. For this study, the opening pressure or the postclamping recording (in cmH_2O) was used because postclamping pressures were not available in all cases of lumbar puncture due to institutional and surgeon variability.

Results

A total of 59 patients (mean age 50; range, 20–70 years) with 77 cerebrospinal fluid leaks/encephaloceles in the LRS were identified (18 patients with bilateral defects). Including individuals with bilateral LRS defects, 25 of 59 patients exhibited multiple CSF leaks. Demographics are listed in Table 1. Obesity was present in 83% of individuals (mean BMI 37.7; range, 26.6–60 kg^2/m) and 81% were females. Encephaloceles were present in 95%. Radiographic signs of prolonged intracranial hypertension were present in 96% of patients with arachnoid pits (93%), anterior cranial fossa (ACF) skull-base attenuation (80%), and empty sella (75%) as the most common findings. Opening or postsurgical lumbar drain or ventriculostomy pressure measurements (normal ICP 5–15 cmH_2O) were elevated in 95% of patients (mean 27.7 ± 8.2; range, 9–50 cmH_2O).

TABLE 1. Patient demographics

Age (years), mean (range)	50 (20–70)
Gender, *n* (%)	
Female	49 (83)
Male	10 (17)
Body mass index (kg/m^2), mean (range)	37.7 (26.6–60)
Encephalocele, *n* (%)	56 (95)
Multiple defects, *n* (%)	
Bilateral LRS	18 (31)
Other sites	7 (12)
Radiologic findings, *n* (%)	
Arachnoid pits	55 (93)
Empty sella[a]	44 (75)
Attenuation of skull base	47 (80)
Dural ectasia[a]	15 (25)
Optic nerve dilation[a]	2 (3)
Intracranial pressure ($cm\ H_2O$), mean ± SD (range)	27.7 ± 8.2 (9–50)

[a]MRI in 44 patients.
LRS = lateral recess of sphenoid; MRI = magnetic resonance imaging; SD = standard deviation.

Discussion

The data provided in this study demonstrates that the vast majority of patients presenting with LRS CSF leaks/encephaloceles have radiographic signs and objective CSF pressure measurements that support intracranial hypertension as the underlying etiology. Preoperative imaging had characteristic findings of empty sella, arachnoid pits, and skull-base attenuation in the majority of patients, similar in incidence to populations of spontaneous CSF leaks due to elevated ICP. Demographics of this population mirror patients with idiopathic intracranial hypertension—predominantly obese, middle-aged females—but are also similar to the spontaneous CSF leak population as a whole.[4,21–23] Arachnoid pits would be expected to be present in 25% of the population,[10] but were identified (often multiple and extensive) in 93% of patients in this study (Fig. 3). Thus, development of lateral sphenoid CSF leaks is likely the result of lateral recess pneumatization, attenuated sphenoid sinus recess roof and skull base, and the development of arachnoid pits from underlying intracranial hypertension.[24,25]

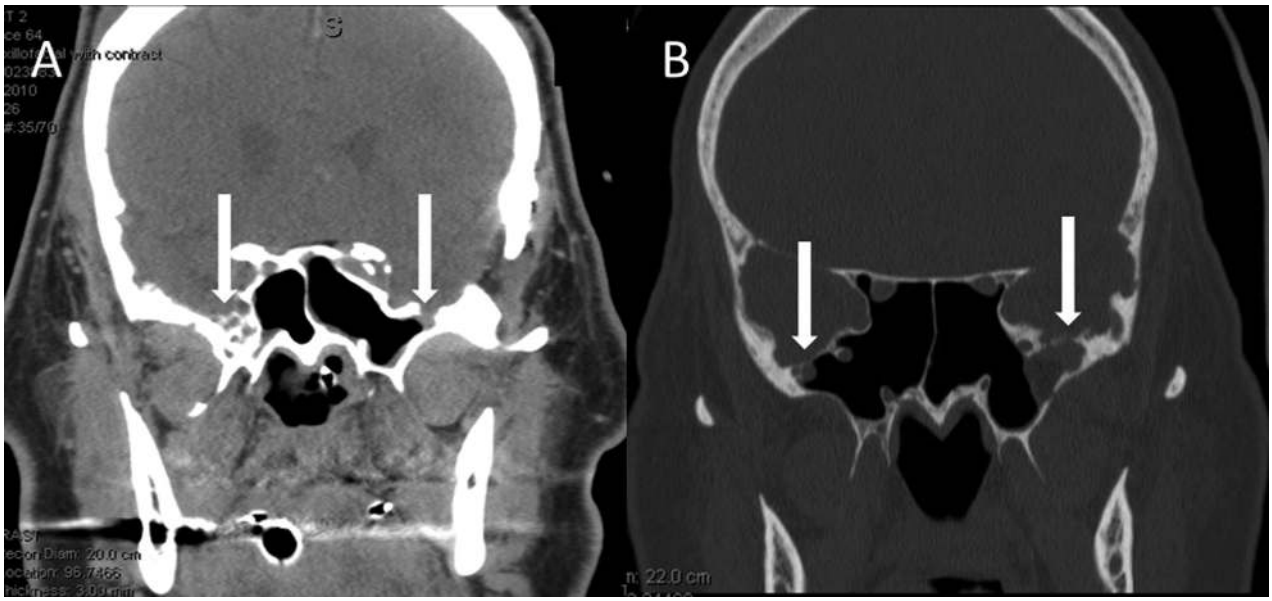

FIGURE 3. Coronal CT scans of 2 patients with bilateral arachnoid pits with subsequent CSF leak and encephalocele development. (A) Note how this patient has multiple pits on the right, but because the lateral recess did not fully pneumatize, he did not develop a CSF leak on that side. (B) This patient has arachnoid pits present all over the middle cranial fossa dura with several defects on the left and a single defect on the right. CT = computed tomography. CSF = cerebrospinal fluid; CT = computed tomography.

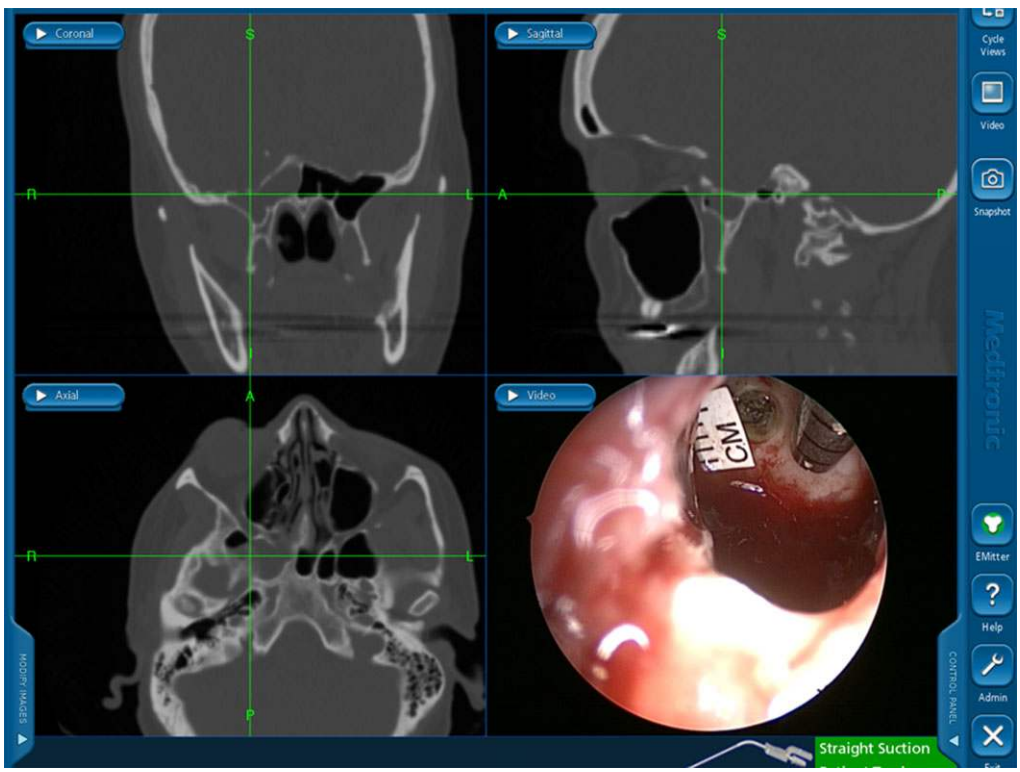

FIGURE 4. Triplanar CT imaging with endoscopic view through a transpterygoid approach following resection of the encephalocele in a morbidly obese female. Note the location lateral to V2. CT = computed tomography; V2 = maxillary nerve (cranial nerve V2).

Numerous case reports and series have propagated an alternative explanation—a congenital etiology—behind the development of LRS CSF leaks due to a persistent "Sternberg's canal." The first argument against a "congenital" origin is that a newborn sphenoid bone has no pneumatization. Congenital transsphenoidal encephaloceles may occur; however, they are always located in the midline without aeration of the surrounding sphenoid sinus.[26] Because congenital means "present at birth" and pneumatization occurs with growth and age, any CSF leak or

encephalocele within a lateral recess of the sphenoid sinus is also, by definition, acquired and not congenital. While pneumatization into a patent canal is a possible consideration for the development of such a leak, the canal as originally described originated medial to the superior orbital fissure (and V2) and thus cannot be considered the etiology of LRS leaks that occur lateral to the maxillary nerve (Fig. 4).[9] In this location, the canal originates at the cavernous sinus dura and an encephalocele must not only transverse the cavernous sinus, but also penetrate 2 layers of dura mater before exiting the skull base. As is currently reported in the literature, the majority of LRS encephaloceles resulting from a persistent lateral craniopharyngeal canal were found lateral to V2, which conflicts directly with the original description. Furthermore, several works reporting supposed Sternberg's canal encephaloceles have radiographic images of patient examples that contain empty sella and arachnoid pits that were not identified by the primary authors.[7,11–18] There is also a high incidence of middle-aged females and obesity in these case series, but underlying ICP was not measured.

The occurrence of multiple leaks in the current series also supports chronically elevated CSF pressure as the underlying etiology for LRS skull-base defects. Even if one were to presume that defects in the LRS are secondary to Sternberg's canal and not intracranial hypertension, a much higher incidence of anterior and middle cranial fossa encephaloceles/CSF leaks would be expected at nerve foramina that are congenitally present (eg, cribriform plate) as they are universal areas of weakness in the skull base. Thus, development of an encephalocele/CSF leak must require some other factor such as antecedent trauma or elevated ICP, even if the sphenoid sinus pneumatized to incorporate a canal.

Conclusion

This study provides objective evidence that LRS CSF leaks are secondary to intracranial hypertension and refutes the myth regarding a congenital origin from Sternberg's canal as the etiology of these leaks. Although defects in embryologic development might predispose a weakness in the sphenoid bone to the development of a spontaneous CSF leak, the presence of a theoretical "congenital" variant, especially given the medial location of the canal (as originally described) and the direct and indirect evidence of high ICP presented here, should not be listed as an underlying cause of LRS defects and should be purged from the literature.

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