Clinical Presentation and Epidemiology of Spontaneous Cerebrospinal Fluid Leaks

Chad Purcell, MD[a,*], Elisa A. Illing, MD[b]

KEYWORDS

• Cerebrospinal fluid leak • Epidemiology • Idiopathic intracranial hypertension
• Obesity • Obstructive sleep apnea

KEY POINTS

- Spontaneous cerebrospinal fluid (CSF) leaks are an increasingly common type of CSF leak seen by skull base surgeons.
- Spontaneous CSF leaks are strongly associated with idiopathic intracranial hypertension (IIH), obstructive sleep apnea, and elevated body mass index.
- Clinicians should be aware that CSF leaks often present with nonspecific symptoms, an should have raised suspicion with unilateral symptoms or findings.
- Some patients with thinning of the skull base are particularly susceptible to spontaneous CSF leaks.

INTRODUCTION

Cerebrospinal fluid (CSF) leaks reflect an abnormal communication between the subarachnoid space and the extracranial environment. Egress of CSF suggests a defect in the skull base and dura mater, with most common leak site being the anterior or lateral skull base, or the spine. Nontraumatic CSF leaks were first categorized by Ommaya in 1964 as "high" or "normal" pressure leaks.[1] Those with high pressure occurred due to elevated intracranial pressure (ICP) from intracranial masses and hydrocephalus, while normal pressure CSF leaks occurred due to congenital skull base defects or focal thinning of the skull base. Subsequently, numerous publications described patients with CSF otorrhea and rhinorrhea without a history of trauma, intracranial

[a] Division of Otolaryngology, Queen's University, Kingston, Ontario, Canada; [b] Department of Otolaryngology, Indiana University School of Medicine, 1130 West Michigan Street, Fesler Hall 400, Indianapolis, IN 46202, USA
* Corresponding author. Kingston Health Sciences Centre, 144 Brock Street, Hotel Dieu Hospital - Murray Building Office 15, Kingston, Ontario K7L5G2, Canada.
E-mail address: purcellrhinology@gmail.com

Otolaryngol Clin N Am 59 (2026) 497–508
https://doi.org/10.1016/j.otc.2026.01.003 **oto.theclinics.com**

Abbreviations	
BMI	body mass index
CKD	chronic kidney disease
CSF	cerebrospinal fluid
ICP	intracranial pressure
IIH	idiopathic intracranial hypertension
OSA	obstructive sleep apnea

masses, or hydrocephalus in the setting of intracranial hypertension of idiopathic origin.[2–5]

Friedman first described the diagnostic criteria for idiopathic intracranial hypertension (IIH) in 2002, which include findings of papilledema, abducens nerve palsy, and increased lumbar puncture opening pressure.[6] These criteria were revised in 2013 to include imaging findings such as empty sella, transverse venous sinus stenosis, flattening of posterior globe, and optic nerve tortuosity on MRI.[6,7] Although many patients may have underlying IIH as a contributing etiology to the spontaneous CSF leak, many may not have classic diagnostic criteria while leaking.[8] Clinicians should aggregate history, physical examination, audiometric findings, beta-2 transferrin testing, and imaging to aid in diagnosing spontaneous CSF leaks and their sources. With expanded clinical awareness, improved diagnostic tools, and worsening of the obesity epidemic, the incidence of spontaneous CSF leaks has risen from 3% to 5% in the 1990s to 14% to 46% in the 2000s.[9] Given the increasing incidence and lifetime risk of meningitis, ranging from 10% to 25% for these patients, it is of utmost importance that clinicians stay aware and knowledgeable about the diagnosis and treatment of spontaneous CSF leaks.[9,10]

INCIDENCE AND RISK FACTORS FOR SPONTANEOUS CEREBROSPINAL FLUID LEAKS

Incidence

The exact incidence of spontaneous CSF leaks is uncertain due to underdiagnosis. Historically, they represented less than 5% of CSF leaks.[1] However, contemporary series report that 20% to 46% of surgically managed CSF leaks are spontaneous.[4,9] This trend likely reflects both improved recognition and a true rise in incidence, correlated with global increases in obesity and IIH.[11]

Demographics

- *Age:* Most occur in the fourth to sixth decades of life[12]
- *Sex:* Female predominance (especially with IIH as underlying etiology)[5]
- *Body mass index (BMI):* Strong association with obesity (average BMI 35–38 kg/m^2)
- *Ethnicity:* Limited data support association

Similar to the demographics of patients with idiopathic intracranial hypertension, patients with spontaneous CSF leaks are generally obese (BMI 35–38 kg/m^2), middle-aged women (~80% of spontaneous CSF leak patients overall) who present with spontaneous clear rhinorrhea or otorrhea.[13] Patients diagnosed with spontaneous CSF leaks tend to be in their fourth or fifth decade of life, which is notably older than those with IIH, where peak incidence is in the third decade of life.[3,4,8,9,11,13,14] One proposed explanation for the difference in age of presentation is that CSF otorrhea or rhinorrhea may provide a release of ICP, which could otherwise manifest in findings associated with IIH. Multiple defects have been reported in over 31% of patients with spontaneous CSF leaks.[15] Multifocal and recurrent leaks are more common in

patients with higher BMI and obstructive sleep apnea (OSA), and recurrence rates are more common in these cases unless the underlying intracranial hypertension is controlled.[16–18]

Risk Factors

As shown in **Table 1**, spontaneous CSF leaks can be related to specific set of predisposing patient and disease-state factors. Both anatomic and physiologic processes likely contribute to increasing a patient's risk of spontaneous CSF leakage. Anatomic factors that predispose individuals to CSF leaks include congenital defects, connective tissue disorders, or decreased functional arachnoid granulations. Physiologic mechanisms contributing to spontaneous CSF leaks relate to obesity and metabolic disorders resulting in intracranial hypertension, chronic sinusitis conditions that cause bony remodeling of the skull base, as well as recurrent Valsalva maneuvers, such as chronic vomiting, weightlifting, or untreated OSA.[12,27–29]

Congenital Defects

Although most patients will have demographic characteristics consistent with metabolic syndrome, obesity, and IIH, some will not. This should raise suspicion for alternative etiologies such as any congenital defects in the skull base. Case series exist in the literature for congenital inner ear dysplasia resulting in CSF otorrhea, and for clival defects such as ecchordosis physaliphora and canalis basilaris medianus in CSF rhinorrhea cases.[21–24] Sternberg's canal is often referenced as a possible congenital defect of the sphenoid that could result in a spontaneous CSF leak, although it is disputed as being a consequence of IIH rather than the primary cause of the spontaneous leak.[20,30]

Connective Tissue Disorders

Some connective tissue disorders are associated with meningeal abnormalities. Meningeal diverticula are known to occur in Marfan's syndrome, neurofibromatosis, autosomal dominant polycystic kidney disease, and familial osteosclerosis.[26] Connective tissue disorders such as Marfan syndrome and Ehlers-Danlos syndromes are present in a minority of patients with spontaneous CSF leaks, although evidence

Table 1
Risk factors for spontaneous cerebrospinal fluid leak

IIH	*Strong* (Most Common Risk Factor; Skull Base Thinning from Chronic Pressure)[9,19]
Obesity (as IIH risk factor)	*Strong*[6,8,12]
Congenital defect	Reported association[20–24]
Connective tissue disorder (Ehlers–Danlos syndrome, Marfan syndrome)	Reported association[25,26]
Chronic sinusitis	Reported association. Bony remodeling in certain situations (allergic fungal rhinosinusitis, mucoceles) recognized as a contributor to osseous thinning at leak sites[3]
Abnormal bony metabolism	Reported association. Chronic kidney disease, hyperparathyroidism, and Paget's disease
Recurrent Valsalva	Strong association with OSA, chronic vomiting, and aggressive weightlifting

supports these patients are predisposed to dural weakness.[26,31] For patients with spontaneous CSF leak with a history of aortic dilatation, arterial dissection, lens dislocation, or family history of connective tissue disorder, an evaluation for a connective tissue disorder should be considered.[31]

Obesity and Metabolic Factors

Patients with obesity are at higher risk for IIH and subsequent CSF leaks given the elevated intra-abdominal pressure, poorer cardiac function, and diminished venous return of blood flow from the brain.[32] CSF reabsorption occurs through the arachnoid granulations into the venous sinuses of the dura mater. Arachnoid granulations were first suggested as potential source of spontaneous CSF leaks in 1990. Gacek[33] proposed that CSF pulsations within arachnoid granulations can slowly erode the surrounding bone. Both obesity and OSA increase the intra-abdominal and venous pressure, which increases the brain's dural sinus pressure. As a result, CSF may be unable to drain effectively from these arachnoid granulations.[34] This could create a build-up of CSF within the cisterns, elevating ICP. Obstruction of arachnoid granulation resulting in a backup of CSF has been proposed as one potential mechanism for the association of CSF leaks with IIH.[35] Other potential mechanisms for the development of IIH are discussed in article "Idiopathic intracranial hypertension." One retrospective study found that young women with IIH had fewer MRI-identified arachnoid granulations than age-matched women or men without IIH, which could explain the increased incidence of IIH in women.[35]

Idiopathic Intracranial Hypertension

Among those patients with chronically elevated ICP, few develop a spontaneous CSF leak. However, many patients with spontaneous CSF leak are found to have IIH.[36] This suggests that elevated ICP is only a contributing factor to the generation of spontaneous CSF leaks, albeit an important one. Skull base defects and associated CSF leaks are most likely to occur in areas of thin bone or natural dehiscence, such as the cribriform plate, lateral recess of the sphenoid, or tegmen tympani.[9,19,37,38] These are locations that also have firm dural adherence to the bone. The combination of elevated ICP with chronic CSF pulsations results in thinning and erosion of the thinner areas of the skull base, formation of meningoencephaloceles, and eventually a rupture in the arachnoid membrane CSF leak.[39] Diagnosing and treating IIH may be challenging if patients develop a CSF leak, as the leaking fluid provides a pressure-lowering "overflow valve" that masks the classic symptoms of raised ICP: headache, pulsatile tinnitus, and vision loss.[39] This is demonstrated in the well-reported scarcity of papilledema for patients with spontaneous CSF leaks.[14,40,41] Rather, radiologic findings of empty sella, tortuous optic nerve sheaths, flattening of posterior orbit, arachnoid pits, and transverse venous sinus stenosis, along with a clinical history of obesity, OSA, and a remote history of symptoms of IIH may help the clinician diagnose an actively leaking patient with IIH as underlying etiology.[36] Diagnosing IIH, even if not preoperatively, is imperative to long-term successful CSF leak repair, prevention of future CSF leaks, and deterrence of sequelae of uncontrolled intracranial hypertension. Several studies demonstrate recurrent CSF leaks without acetazolamide management, or with ventriculoperitoneal shunt failure.[42–44] IIH diagnosis, management, and treatment are discussed in detail in the article "Idiopathic intracranial hypertension."

Chronic Sinusitis

Bony remodeling can occur in sinonasal inflammatory disease or after facial traumas/infections. Particularly, in cases of allergic fungal rhinosinusitis, bony erosion of the

skull base can be extensive, leading to cranial neuropathies.[45–48] Case reports demonstrate increased risks of spontaneous CSF leaks in cases of skull base remodeling; however, these remain the minority of cases.[49] The etiology of the leak is not always certain to be due to the inflammatory process itself or surgical intervention to treat the underlying sinusitis and obstruction. Nonetheless, patients should be screened for spontaneous CSF leaks at the time of presentation, and the risks of CSF leak with operative intervention should be discussed during the informed consent process.

Abnormal Bony Metabolism

In patients with disease states that result in aberrant bony metabolism, skull base erosion may occur in the absence of intracranial hypertension. In cases of chronic kidney disease (CKD), patients may develop secondary hyperparathyroidism and subsequent skull base erosion that may results in spontaneous CSF leak.[50] Patients with CKD have impaired vitamin D metabolism, phosphate retention, and elevated parathyroid hormone, which result in cortical bone erosion.[51] A 2017 study from the United States National Inpatient Sample database demonstrated that nearly 2% of patients with CSF rhinorrhea had a history of CKD.[52] Similarly, in patients with Paget's disease and hyperparathyroidism, comparable presentations may occur, though rare.

Obstructive Sleep Apnea (Recurrent Valsalva Maneuvers)

Likely related to the obesity epidemic in the past 3 decades, OSA has been strongly associated with spontaneous CSF leaks. In spontaneous CSF leaks of the temporal bone, rates of OSA have been noted to range from 46% to 83% of patients with CSF leak undergoing perioperative polysomnography testing.[53,54] During apneic events of OSA, ICPs are known to spike, likely contributing to skull base erosion with time.[55,56] This phenomenon is supported by differences noted in calvarial and skull base thickness between those with obesity alone and obesity with OSA. In one study, the temporal bone calvarial thickness was decreased by 23% in patients with OSA, even when controlling for obesity. This bone thinning was not present in the extracranial zygoma, implying that ICP differences likely contribute to eventual CSF leaks, rather than obesity alone. The transient pressure elevations seen with OSA may also make the diagnosis of IIH difficult, as these spikes are limited to apneic events and are absent during waking hours. Thus, traditional methods to diagnose IIH may not provide insight into the etiology of the CSF leak, and polysomnography should be considered as an essential diagnostic tool in these patients.[54] Furthermore, OSA is associated with multiple other comorbid conditions, including but not limited to cardiovascular disease, stroke, pulmonary disease, chronic fatigue, and all-cause mortality.[57–60] This highlights the importance of not overlooking the overall health of patients with OSA.

CLINICAL PRESENTATION

Clinicians should be aware of the constellation of symptoms that patients may describe with spontaneous CSF leaks to avoid misdiagnosis and delays in treatment. The first steps should be focused history and physical examination (**Fig. 1**), followed by appropriate imaging and possibly laboratory testing.

A focused history should include symptoms, comorbidities, history of any surgical interventions or trauma history, social history, including physical activities, and occupational history. Patients with spontaneous CSF leaks may describe a history of symptoms that mirror IIH symptoms. These include headaches (60%), pulsatile tinnitus

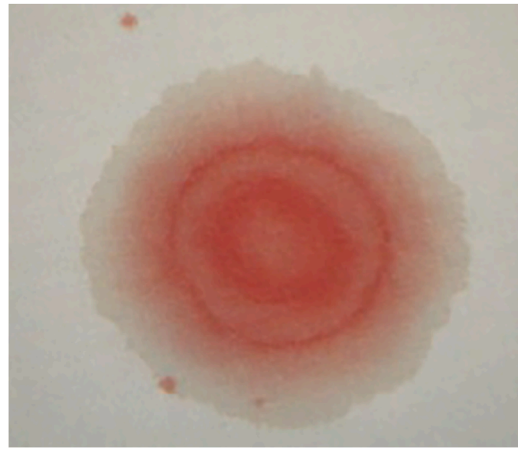

Fig. 1. Halo sign; cerebrospinal fluid and blood separation from secretion sample on tissue paper.

(20%), and diplopia (5%), which are all less prevalent in actively leaking patients than reported among patients with IIH alone (90%, 60%, and 30%, respectively).[61] Notably, most patient with spontaneous cranial CSF leak do not endorse symptoms of intracranial hypotension. This is due to the CSF pressure gradient where CSF pressure is negative above the cervical spine, and positive below. If the ICP exceeds atmospheric pressure at the level of the skull base where a dehiscence is present, a CSF leak will ensue. Leakage of CSF will offload pressure and offset ICP elevation, and no intracranial hypotension will occur. Rather than a headache, patients may describe rhinorrhea or otorrhea.[36] Patients with CSF leak may experience worsened headaches with transient ICP elevations when leaning forward, lying flat, performing Valsalva maneuvers, or with exertion, which may also correlate to worsened otorrhea or rhinorrhea.

Lateral Skull Base Location

For patients with lateral skull-base CSF leaks, presenting symptoms may be vague and difficult to distinguish from those of chronic otitis media. Multiple studies show that hearing loss, aural fullness, and otorrhea remain the most common presenting symptoms of lateral CSF leaks.[33,62–64] Determining the history of recurrent ear infections, any history of surgical intervention for otologic conditions, and an audiogram are helpful for the evaluation of patients presenting with unilateral middle ear effusion. If a tympanostomy tube has been placed, collection of any otorrhea for β2 transferrin testing is a helpful tool for diagnosis. Fluid can also be aspirated through the tympanic membrane using a 25 gauge needle and 1 mL syringe to send for β2 transferrin testing, which has a sensitivity of 99% and specificity of 97%.[65] Rarely, patients with lateral skull base leaks may present with rhinorrhea, if the fluid traverses the Eustachian tube into the nasopharynx and nasal cavity. CSF otorrhea most commonly arises from a defect in the lateral skull-base at the tegmen tympani (47.6%) and tegmen mastoideum (47.2%), while posterior fossa defects (1.9%) and zygomatic root defects (1.4%) are less commonly reported.[66]

Anterior Skull Base Location

For patients with anterior skull base CSF leaks, persistent, “leaky faucet” nasal or nasopharyngeal drainage is described, often with a salty or metallic taste.[67] CSF is

a clear, watery fluid with salt content similar to plasma. These features help distinguish CSF from sticky nasal secretions, which have a more thick, stringy, mucinous consistency. It can be difficult to distinguish nasal secretions from CSF based on consistency and appearance alone, especially in cases where small volumes of CSF mix with sinonasal secretions. Thus, the collection of fluid for β2 transferrin testing is recommended.[68] Patients rarely report changes in their sense of smell with spontaneous CSF leaks, in contrast to many with inflammatory sinonasal disease. Anosmia is uncommon unless the cribriform plate or ethmoid is significantly or bilaterally disrupted. Reports have been published of patients with ethmoid roof defects who presented with anosmia in addition to rhinorrhea, but these remain the minority.[18] Additionally, complicating the presentation of symptoms is the increased prevalence of aspiration pneumonitis in patients with higher flow spontaneous CSF leaks. Reported in one study as high as 25%, the nonspecific symptoms of cough, sputum production, and shortness of breath may be difficult to discern from other reactive airway diseases commonly concomitant with inflammatory sinonasal disease.[69]

The most frequent locations of spontaneous CSF leaks include the cribriform plate, the pneumatized lateral recess of the sphenoid sinus, the ethmoid roof, or the posterior table of the frontal sinus.[30] Traditionally, the cribriform plate has been considered the most common site of anterior skull base CSF leaks because of its delicate, porous bone attached to a thin bony lateral lamella that fuses with the fovea ethmoidalis. This fragile, downward-angled structure creates a weak spot for spontaneous or provoked leaks. However, recent prevalence estimates suggest that the epidemiology may be changing, with CSF leaks most commonly originating from the sphenoid sinus (41.1%), followed by the cribriform plate (25.4%), and ethmoid skull base (20.4%).[70] The shift in epidemiology toward a preference for the lateral sphenoid location may be due to both anatomic predisposition and changes in the weight of US population over time. Anatomically, pneumatization of the lateral recess of the sphenoid is reported in 91% of patients with spontaneous CSF leaks, compared to only 23% to 43% in control patients.[71] CSF leaks related to the lateral recess of the sphenoid sinus usually show radiographic signs of increased ICP, rather than congenital defects from encephaloceles of Sternberg's canal.[30] With an ever-rising rate of obesity in the United States and worldwide, the lateral recess of the sphenoid may become increasingly common as a site of spontaneous CSF leaks.

Encephaloceles/Meningoceles

Herniation of the meninges alone (meningocele) or both the meninges and brain (meningoencephalocele/encephalocele) through a bone or dural defect at the skull base creates a direct communication that may present as CSF rhinorrhea or otorrhea.[72] Spontaneous CSF fistulas in the skull base are linked to higher rates of coexisting meningoceles and/or encephaloceles on radiographic evaluation, with rates reaching up to 50% among patients with CSF leaks and increased ICP.[73,74] One study reviewed 79 patients with IIH and 76 control subjects. Meningoceles were found in 11% of patients with IIH, compared with 0% in control subjects ($P < .003$).[75] A systematic review examining the association between obesity and spontaneous temporal bone CSF leaks found that most patients had a single skull base defect and encephaloceles. There are also small studies showing a possible association between obesity and meningoceles/encephaloceles, including one study that compared 17 spontaneous CSF leaks to 44 nonspontaneous leaks, in which the mean BMI was significantly higher in the spontaneous CSF leak group ($P < .001$).[76] Taken together, findings of encephaloceles or meningoceles without a clear etiology should raise clinical suspicion for IIH and prompt a detailed evaluation.

Meningitis

Occasionally, patients with spontaneous CSF leaks may present with a history of meningitis, or active concerns for meningitis (15%).[8,70] Symptoms of active meningitis include alterations in mental status, fever, neck stiffness, light sensitivity, nausea, and vomiting. Clinicians should have elevated suspicion for CSF leak in patients who have a history of recurrent meningitis.[77] Low-flow CSF leaks may present with recurrent meningitis instead of drainage from the nose.[9]

SUMMARY

Spontaneous CSF leaks represent a rare but increasingly recognized and distinct clinical entity with characteristic epidemiologic and clinical patterns. Patients who experience spontaneous CSF leaks, particularly on a multisite or recurrent basis, tend to be middle-aged, women with obesity and may have associated conditions such as IIH and OSA. Patients tend to present with symptoms that can help localize the source of the leak. Anterior leaks often arise from the cribriform plate or sphenoid sinus, while lateral leaks originate from the tegmen tympani. Patients may describe ear or nasal symptoms, which may indicate the potential CSF leak source. Symptoms may include clear rhinorrhea or otorrhea, pulsatile tinnitus, muffled hearing, vertigo, headaches, neck pain, and signs of meningitis. Further research is needed to clarify the pathogenesis and natural history of spontaneous leaks to identify predictive features and develop prevention strategies and methods of early detection.

CLINICS CARE POINTS

- The reported incidence of spontaneous CSF leaks is increasing, as is the proportional to the obesity epidemic and prevalence of OSA.
- Patients presenting with unilateral clear rhinorrhea or otorrhea/effusion should be assessed or investigated for possible CSF leak.
- Patients at risk of spontaneous CSF leak should be counseled about their risk and modifiable risk factors.
- Spontaneous CSF leaks and IIH share similar demographic profile, predominantly affecting individuals and women with obesity.

REFERENCES

1. Ommaya AK. Cerebrospinal fluid rhinorrhea. Neurology 1964;14:106–13.
2. Schlosser RJ, Bolger WE. Nasal cerebrospinal fluid leaks. J Otolaryngol 2002;31-(Suppl 1):S28–37.
3. Schlosser RJ, Bolger WE. Nasal cerebrospinal fluid leaks: critical review and surgical considerations. Laryngoscope 2004;114(2):255–65.
4. Schlosser RJ, Wilensky EM, Grady MS, et al. Elevated intracranial pressures in spontaneous cerebrospinal fluid leaks. Am J Rhinol 2003;17(4):191–5.
5. Schlosser RJ, Woodworth BA, Wilensky EM, et al. Spontaneous cerebrospinal fluid leaks: a variant of benign intracranial hypertension. Ann Otol Rhinol Laryngol 2006;115(7):495–500.
6. Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology 2002;59(10):1492–5.

7. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the Pseudotumor cerebri syndrome in adults and children. Neurology 2013;81(13):1159–65.
8. Bidot S, Levy JM, Saindane AM, et al. Spontaneous skull base cerebrospinal fluid leaks and their relationship to idiopathic intracranial hypertension. Am J Rhinol Allergy 2021;35(1):36–43.
9. Wise SK, Schlosser RJ. Evaluation of spontaneous nasal cerebrospinal fluid leaks. Curr Opin Otolaryngol Head Neck Surg 2007;15(1):28–34.
10. Hegazy HM, Carrau RL, Snyderman CH, et al. Transnasal endoscopic repair of cerebrospinal fluid rhinorrhea: a meta-analysis. Laryngoscope 2000;110(7):1166–72.
11. Chaaban MR, Illing E, Riley KO, et al. Spontaneous cerebrospinal fluid leak repair: a five-year prospective evaluation. Laryngoscope 2014;124(1):70–5.
12. Nelson RF, Gantz BJ, Hansen MR. The rising incidence of spontaneous cerebrospinal fluid leaks in the United States and the association with obesity and obstructive sleep apnea. Otol Neurotol 2015;36(3):476–80.
13. Stevens SM, Rizk HG, Golnik K, et al. Idiopathic intracranial hypertension: contemporary review and implications for the otolaryngologist. Laryngoscope 2018;128(1):248–56.
14. Yang Z, Wang B, Wang C, et al. Primary spontaneous cerebrospinal fluid rhinorrhea: a symptom of idiopathic intracranial hypertension? J Neurosurg 2011; 115(1):165–70.
15. Schlosser RJ, Bolger WE. Management of multiple spontaneous nasal meningoencephaloceles. Laryngoscope 2002;112(6):980–5.
16. Dallan I, Cambi C, Emanuelli E, et al. Multiple spontaneous skull base cerebrospinal fluid leaks: some insights from an international retrospective collaborative study. Eur Arch Otorhinolaryngol 2020;277(12):3357–63.
17. Filip P, Patel EA, Khalife S, et al. Multiple skull base defects in the setting of spontaneous cerebrospinal fluid rhinorrhea; a dual institution view. Am J Otolaryngol 2024;45(1):104048.
18. Schwartz N, Brown KD, Sasaki-Adams D, et al. Multifocal skull base defects with associated spontaneous cerebrospinal fluid leaks. Otol Neurotol 2021;42(5): e624–8.
19. Woodworth BA, Prince A, Chiu AG, et al. Spontaneous CSF leaks: a paradigm for definitive repair and management of intracranial hypertension. Otolaryngol Head Neck Surg 2008;138(6):715–20.
20. Castelnuovo P, Dallan I, Pistochini A, et al. Endonasal endoscopic repair of Sternberg's canal cerebrospinal fluid leaks. Laryngoscope 2007;117(2):345–9.
21. Georgalas C, Terzakis D, Tsikna M, et al. Ecchordosis physaliphora: a cautionary tale. J Laryngol Otol 2020;134(1):46–51.
22. Hasegawa H, Van Gompel JJ, Choby G, et al. Unrecognized notochordal lesions as a likely cause of idiopathic clival cerebrospinal fluid leaks. Clin Neurol Neurosurg 2023;224:107562.
23. Khairy S, Almubarak AO, Aloraidi A, et al. Canalis basalis medianus with cerebrospinal fluid leak: rare presentation and literature review. Br J Neurosurg 2019; 33(4):432–3.
24. Wang B, Dai WJ, Cheng XT, et al. Cerebrospinal fluid otorrhea secondary to congenital inner ear dysplasia: diagnosis and management of 18 cases. J Zhejiang Univ - Sci B 2019;20(2):156–63.
25. Schievink WI, Gordon OK, Tourje J. Connective tissue disorders with spontaneous spinal cerebrospinal fluid leaks and intracranial hypotension: a prospective study. Neurosurgery 2004;54(1):65–70 [discussion: 70–1].

26. Mokri B, Maher CO, Sencakova D. Spontaneous CSF leaks: underlying disorder of connective tissue. Neurology 2002;58(5):814–6.
27. Bakhsheshian J, Hwang MS, Friedman M. Association between obstructive sleep apnea and spontaneous cerebrospinal fluid leaks: a systematic review and meta-analysis. JAMA Otolaryngol Head Neck Surg 2015;141(8):733–8.
28. Vivas EX, Mccall A, Raz Y, et al. ICP, BMI, surgical repair, and CSF diversion in patients presenting with spontaneous CSF otorrhea. Otol Neurotol 2014;35(2):344–7.
29. Davis J, Yanny I, Chatu S, et al. Spontaneous cerebrospinal fluid leak following a pilates class: a case report. J Med Case Rep 2014;8:456.
30. Illing E, Schlosser RJ, Palmer JN, et al. Spontaneous sphenoid lateral recess cerebrospinal fluid leaks arise from intracranial hypertension, not Sternberg's canal. Int Forum Allergy Rhinol 2014;4(3):246–50.
31. Reinstein E, Pariani M, Bannykh S, et al. Connective tissue spectrum abnormalities associated with spontaneous cerebrospinal fluid leaks: a prospective study. Eur J Hum Genet 2013;21(4):386–90.
32. Stucken EZ, Selesnick SH, Brown KD. The role of obesity in spontaneous temporal bone encephaloceles and CSF leak. Otol Neurotol 2012;33(8):1412–7.
33. Gacek RR. Arachnoid granulation cerebrospinal fluid otorrhea. Ann Otol Rhinol Laryngol 1990;99(11):854–62.
34. Davson H,SM. Physiology of the CSF and blood-brain barriers. Boca Raton: CRC Press; 1996.
35. Huda F, Abdelmonem A, Dehghani Firouzabadi F, et al. The role of arachnoid granulations in idiopathic intracranial hypertension. NeuroRadiol J 2023;36(6):651–6.
36. Bidot S, Levy JM, Saindane AM, et al. Do most patients with a spontaneous cerebrospinal fluid leak have idiopathic intracranial hypertension? J Neuro Ophthalmol 2019;39(4):487–95.
37. Banks CA, Palmer JN, Chiu AG, et al. Endoscopic closure of CSF rhinorrhea: 193 cases over 21 years. Otolaryngol Head Neck Surg 2009;140(6):826–33.
38. Castelnuovo P, Mauri S, Locatelli D, et al. Endoscopic repair of cerebrospinal fluid rhinorrhea: learning from our failures. Am J Rhinol 2001;15(5):333–42.
39. Quatre R, Attye A, Righini CA, et al. Spontaneous cerebrospinal fluid rhinorrhea: association with body weight and imaging data. J Neurol Surg B Skull Base 2017;78(5):419–24.
40. Aaron G, Doyle J, Vaphiades MS, et al. Increased intracranial pressure in spontaneous CSF leak patients is not associated with papilledema. Otolaryngol Head Neck Surg 2014;151(6):1061–6.
41. Fyrmpas G, Konstantinidis I, Selviaridis P, et al. Management of spontaneous cerebrospinal fluid leaks of the sphenoid sinus: our experience. J Laryngol Otol 2014;128(9):797–802.
42. Elshanawany A, Mohammad F. Spontaneous cerebrospinal fluid rhinorrhea as a primary presentation of idiopathic intracranial hypertension, management strategies, and clinical outcome. Surg Neurol Int 2024;15:458.
43. Teachey W, Grayson J, Cho DY, et al. Intervention for elevated intracranial pressure improves success rate after repair of spontaneous cerebrospinal fluid leaks. Laryngoscope 2017;127(9):2011–6.
44. Mokri B. Intracranial hypertension after treatment of spontaneous cerebrospinal fluid leaks. Mayo Clin Proc 2002;77(11):1241–6.

45. Illing EA, Dunlap Q, Woodworth BA. Outcomes of pressure-induced cranial neuropathies from allergic fungal rhinosinusitis. Otolaryngol Head Neck Surg 2015; 152(3):541–5.
46. Marfani MS, Jawaid MA, Shaikh SM, et al. Allergic fungal rhinosinusitis with skull base and orbital erosion. J Laryngol Otol 2010;124(2):161–5.
47. Reitzen SD, Lebowitz RA, Jacobs JB. Allergic fungal sinusitis with extensive bone erosion of the clivus presenting with diplopia. J Laryngol Otol 2009;123(7):817–9.
48. White LC, Jang DW, Yelvertan JC, et al. Bony erosion patterns in patients with allergic fungal sinusitis. Am J Rhinol Allergy 2015;29(4):243–5.
49. Pizzo LJ, Mishler KE. Frontoethmoid mucocele with CSF leak. Ear Nose Throat J 1984;63(11):571–3.
50. Jyothsna BP, Kumar KV, Unni NV, et al. Spontaneous cerebrospinal fluid rhinorrhea in end stage renal disease. Indian J Nephrol 2021;31(3):296–8.
51. Moe SM, Sprague SM. Chapter 54: Chronic kidney disease – mineral bone disorder. In: Taal MW, Chertow GM, Marsden PA, et al, editors. Brenner and rector's the kidney. 9th edition. Philadelphia: Elsevier; 2012. p. 2021–58.
52. Marchiano E, Carniol ET, Guzman DE, et al. An analysis of patients treated for cerebrospinal fluid rhinorrhea in the United States from 2002 to 2010. J Neurol Surg B Skull Base 2017;78(1):18–23.
53. Nelson RF, Roche JP, Gantz BJ, et al. Middle Cranial Fossa (MCF) approach without the use of lumbar drain for the management of spontaneous Cerebral Spinal Fluid (CSF) leaks. Otol Neurotol 2016;37(10):1625–9.
54. Rabbani CC, Saltagi MZ, Manchanda SK, et al. Prevalence of obstructive sleep apnea (OSA) in spontaneous cerebrospinal fluid (CSF) leaks: a prospective cohort study. Otol Neurotol 2018;39(6):e475–80.
55. Jennum P, Borgesen SE. Intracranial pressure and obstructive sleep apnea. Chest 1989;95(2):279–83.
56. Sugita Y, Iijima S, Teshima Y, et al. Marked episodic elevation of cerebrospinal fluid pressure during nocturnal sleep in patients with sleep apnea hypersomnia syndrome. Electroencephalogr Clin Neurophysiol 1985;60(3):214–9.
57. Young T, Skatrud J, Peppard PE. Risk factors for obstructive sleep apnea in adults. JAMA 2004;291(16):2013–6.
58. Farrell PC, Richards G. Recognition and treatment of sleep-disordered breathing: an important component of chronic disease management. J Transl Med 2017; 15(1):114.
59. Young T, Palta M, Dempsey J, et al. Burden of sleep apnea: rationale, design, and major findings of the Wisconsin sleep cohort study. Wis Med J 2009;108(5): 246–9.
60. Force USPST, Bibbins-Domingo K, Grossman DC, et al. Screening for obstructive sleep apnea in adults: US preventive services task force recommendation statement. JAMA 2017;317(4):407–14.
61. Bidot S, Bruce BB. Update on the diagnosis and treatment of idiopathic intracranial hypertension. Semin Neurol 2015;35(5):527–38.
62. Brown NE, Grundfast KM, Jabre A, et al. Diagnosis and management of spontaneous cerebrospinal fluid-middle ear effusion and otorrhea. Laryngoscope 2004; 114(5):800–5.
63. Kutz JW Jr, Husain IA, Isaacson B, et al. Management of spontaneous cerebrospinal fluid otorrhea. Laryngoscope 2008;118(12):2195–9.
64. Leonetti JP, Marzo S, Anderson D, et al. Spontaneous transtemporal CSF leakage: a study of 51 cases. Ear Nose Throat J 2005;84(11):700, 702–4, 706.

65. Warnecke A, Averbeck T, Wurster U, et al. Diagnostic relevance of beta2-transferrin for the detection of cerebrospinal fluid fistulas. Arch Otolaryngol Head Neck Surg 2004;130(10):1178–84.
66. Vasudevan SS, Albornoz V, Schwab S, et al. Global prevalence, recurrence, and risk factors associated with spontaneous CSF Otorrhea-A meta-analysis. Otolaryngol Head Neck Surg 2025;173(4):797–811.
67. Wang EW, Vandergrift WA 3rd, Schlosser RJ. Spontaneous CSF leaks. Otolaryngol Clin North Am 2011;44(4):845–56, vii.
68. Oakley GM, Alt JA, Schlosser RJ, et al. Diagnosis of cerebrospinal fluid rhinorrhea: an evidence-based review with recommendations. Int Forum Allergy Rhinol 2016;6(1):8–16.
69. Tang R, Zhou J, Zhang S, et al. Increased prevalence of aspiration pneumonitis in spontaneous cerebrospinal fluid leaks. Int Forum Allergy Rhinol 2024;14(5):1002–5.
70. Hong CS, Kundishora AJ, Elsamadicy AA, et al. A unique subset: idiopathic intracranial hypertension presenting as spontaneous CSF leak of the anterior skull base. J Neurol Surg B Skull Base 2022;83(2):105–15.
71. Shetty PG, Shroff MM, Fatterpekar GM, et al. A retrospective analysis of spontaneous sphenoid sinus fistula: MR and CT findings. AJNR Am J Neuroradiol 2000;21(2):337–42.
72. Pérez MA, Bialer OY, Bruce BB, et al. Primary spontaneous cerebrospinal fluid leaks and idiopathic intracranial hypertension. J Neuro Ophthalmol 2013;33(4):330.
73. Alonso RC, de la Peña MJ, Caicoya AG, et al. Spontaneous skull base meningoencephaloceles and cerebrospinal fluid fistulas. Radiographics 2013;33(2):553–70.
74. Silver RI, Moonis G, Schlosser RJ, et al. Radiographic signs of elevated intracranial pressure in idiopathic cerebrospinal fluid leaks: a possible presentation of idiopathic intracranial hypertension. Am J Rhinol 2007;21(3):257–61.
75. Bialer OY, Rueda MP, Bruce BB, et al. Meningoceles in idiopathic intracranial hypertension. AJR Am J Roentgenol 2014;202(3):608–13. PMID: 24555598.
76. Holzmann D, Wild C. Obesity as a risk factor for primary spontaneous rhinoliquorrhea. Arch Otolaryngol Head Neck Surg 2003;129(3):324–6.
77. de Macedo Filho L, Machado CCP, Mendes GBB, et al. Spontaneous rhinorrhea and idiopathic intracranial hypertension: a complex and challenging association. Neurol Neurochir Pol 2024;58(1):31–7.