Surgical Outcomes and Postoperative Management in Spontaneous Cerebrospinal Fluid Rhinorrhea

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Abstract

Background The etiology of spontaneous cerebrospinal fluid (CSF) rhinorrhea remains unknown, though emerging evidence suggests that this likely represents a variant of idiopathic intracranial hypertension. Long-term success rates for repair and postoperative management strategies remain variable.

Methods Retrospective review of patients undergoing surgical management of spontaneous CSF rhinorrhea was conducted over a 5-year period. Analysis was performed to correlate recurrence with demographics and perioperative variables. Lumbar puncture usage and neuro-ophthalmological examinations in clinical management were also assessed.

Results Forty-eight patients were included in the study. The mean age was 51.4 years with 94% females. Leaks were most commonly located in the sphenoid (43.8%) and cribriform region (33.3%). The most common findings on magnetic resonance imaging were empty sella (48%) and Meckel's cave diverticula (24%). Nine patients (18.8%) had recurrent CSF leaks. Six occurred >1 month postoperatively. Three had repeat endoscopic repairs, two received ventriculoperitoneal shunts, and one was managed with a lumbar drain, with overall success rate of 93.8%. Acetazolamide was utilized in 19 cases (39.6%) postoperatively. Overall, 59% of patients had elevated opening pressures on postoperative lumbar puncture ($n = 32$). Neuro-ophthalmology evaluated 28 patients; 25% had visual field deficits, and 7.1% had papilledema.

Keywords

- ▶ endoscopy
- ▶ cerebrospinal fluid
- ▶ leak
- ▶ rhinorrhea
- ▶ spontaneous
- ▶ skull base
- ▶ intracranial hypertension

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Conclusions Management of spontaneous CSF leaks remains a significant challenge. Endoscopic repair is successful in most patients with little morbidity; however, postoperative management remains inconsistent, and further studies are warranted to establish consensus on post-surgical care. The association with elevated intracranial pressure and visual field deficits/papilledema suggests opening pressures, and neuro-ophthalmologic evaluation should be considered in the treatment algorithm.

Introduction

Spontaneous cerebrospinal fluid (CSF) leaks occur rarely in the adult population and are associated with increased risk of meningitis.¹ Although large series have assessed management and outcomes of CSF leaks due to various etiologies, including skull base tumor removal, trauma, and iatrogenic causes, few series exist specifically evaluating outcomes in patients with spontaneous CSF rhinorrhea.² Though the exact pathogenesis of spontaneous CSF leaks and meningoencephaloceles remains unknown, it is apparent that select patients are influenced by increased intracranial pressure (ICP), commonly due to idiopathic intracranial hypertension (IIH). Emerging data also suggests that patients with elevated ICP may also develop diplopia and reduced vision with increased blind spots and disc edema.³

Previous cohorts of patients with CSF rhinorrhea have demonstrated that the average ICP elevated up to 32 cm H₂O after leak repair supporting ICP as a potential cause for spontaneous CSF leaks.^{4–6} Studies in the lateral temporal bone have shown evidence of opening pressures >20 cm H₂O in over 30 to 60% of patients who underwent repair for spontaneous otogenic leaks.^{7,8} Further, radiographic evidence of elevated ICP was present in 50 to 85% of patients.^{6,7} Thus, elevated ICP is often present though not universally in all patients.

Strategies to reduce ICP include medical (i.e., acetazolamide, and topiramate) and surgical (ventriculoperitoneal [VP] shunt) therapies. The use of acetazolamide has been demonstrated to decrease ICP in the short term.⁹ However, long-term effectiveness of acetazolamide is unknown. Moreover, both IIH and CSF leaks have been linked to obesity, and there are reports of laparoscopic gastric banding as an adjunct in preventing recurrent CSF rhinorrhea.¹⁰ A recent Cochrane review failed to generate an evidence-based management strategy for IIH.¹¹

The present series adds to the growing body of literature regarding treatment of spontaneous CSF leaks. Surgical outcomes and postoperative management, including opening pressures and neuro-ophthalmology evaluation, are reported to better ascertain the long-term care strategy.

Methods

Patients undergoing operative repair for spontaneous CSF rhinorrhea between January 2008 and December 2013 were included in this retrospective analysis. Patients were in-

cluded if they were adults (age >18 years) and had spontaneous CSF rhinorrhea after excluding all known causes, previous trauma, prior sinonasal or skull base surgery, or known neoplastic process at the anterior skull base. Approval was obtained from the University of Texas Southwestern Medical Center Investigational Review Board prior to commencing the review.

Data collected included patient demographics, body mass index (BMI), patient symptoms, radiographic findings, and intraoperative factors, such as method of reconstruction, use of acetazolamide, lumbar drain, and number of days of inpatient bed rest. Additional long-term postoperative factors, including neuro-ophthalmology evaluation and ICP were recorded if available.

Descriptive statistics of the study population were performed with mean and standard deviation calculated for continuous variables and frequencies for categorical variables. Various patient characteristics and perioperative factors were correlated with recurrence of CSF leak with two-sample *t*-tests for continuous variables and chi-square test for categorical variables. Similarly, proportionality and *t*-tests were used to determine factors related to duration of bed rest in the patient population. Univariate analysis was performed using SAS 9.4 (SAS Institute Inc., Cary, North Carolina). Multivariate analysis with logistic regression of select variables was also performed to create a predictive model for recurrent CSF leak from the patient data.

Results

Forty-eight patients with spontaneous CSF rhinorrhea were included in the study. Patient demographics, clinical characteristics, and preoperative symptomatology are summarized in ►Table 1. The mean duration of CSF rhinorrhea was 43 weeks at the time of initial presentation. The most common presenting symptom other than rhinorrhea was headaches (50%). Visual field deficits were rare findings prior to surgical repair of the CSF leak (4%). Overall, patients had been followed up for a mean of 1.49 years (range: 0–6.6 years) with nine patients having recurrent CSF rhinorrhea (9/48, 18.8%). Of these, six occurred >1 month post-operatively. Three had repeat transnasal endoscopic repairs, two received a VP shunt, three were lost to follow up, and one received a lumbar drain, followed by a shunt. Both patients who received a VP shunt had recurrent leaks <1 month after primary repair. Overall, success rate with additional interventions was 93.8% (45/48).

Table 1 Patient demographics and preoperative factors stratified by recurrence in CSF leak after repair

Variable	No recurrence no. (%)	Recurrence no. (%)	Total	Odds ratio	p-Value
Mean age (years) ^a	53.1 ± 9.6	44.3 ± 8.1	51.4 ± 9.9	n/a	0.01
Gender (female)	37 (95%)	8 (89%)	45 (94%)	2.3 (0.2–28)	0.50
Mean duration before treatment (weeks) ^a	30.6 ± 30.4	91.3 ± 173.9	43.0 ± 83.5	n/a	0.05
Mean weight (kg) ^a	89.6 ± 17.7	112.6 ± 18.8	93.9 ± 19.9	n/a	0.001
Mean height (m) ^a	1.6 ± 0.1	1.6 ± 0.1	1.6 ± 0.1	n/a	0.37
Mean BMI ^a	34.4 ± 6.4	41.9 ± 5.9	35.8 ± 6.9	n/a	0.003
Headaches ^b	18 (46%)	6 (67%)	24 (50%)	2.3 (0.5–11)	0.27
Pulsatile tinnitus ^b	1 (2.6%)	1 (11%)	2 (4%)	4.8 (0.3–84)	0.25
Visual field defects ^b	2 (5%)	0	2 (4%)	n/a	0.49
History of meningitis ^b	6 (15%)	0	6 (14%)	n/a	0.21
History of brain abscess ^b	1 (2.6%)	0	1 (2%)	n/a	0.63

Abbreviation: BMI, body mass index.

^aContinuous variables presented as mean ± standard deviation.

^bBinary categorical variables documented by history during the preoperative encounter.

All patients received a compute tomography (CT) scan for perioperative planning and intraoperative image guidance. The most common location of the leak was in the cribriform plate region (►Table 2). No spontaneous leaks in the frontal sinus were noted in this case series. Fifty-six percent of patients had leaks on the left side of the skull base. Magnetic resonance imaging (MRI) was available for review in 25 patients. The most common abnormalities noted were an empty sella (48%) followed by Meckel's cave diverticula (24%) (►Table 3).

All surgical repairs were performed utilizing image-guided transnasal endoscopic repair. A multitude of tissue arrays were utilized for the skull base reconstruction as dictated by surgeon preference (►Table 4). Free mucosal graft and nasoseptal flap was utilized as a component of the reconstruction in 27 (56%) and 11 (23%) cases, respectively. A multilayered reconstruction approach was employed in 22 patients (46%). A lumbar drain was used selectively in 13 patients (27%).

Multivariate analysis was performed utilizing select data common to all patients included in the study. A predictive model of the odds of recurrence via a multivariate logistic regression was created (►Table 5). Overall, female gender,

increased BMI, use of multilayer closure, lumbar drain, and use of acetazolamide was associated with an increase in the odds ratio for CSF leak recurrence. Meanwhile, increasing age and number of days at bed rest were associated with a decrease in the odds ratio. Nevertheless, only age and BMI were statistically significant in this model.

Patients spent a mean of 1.2 days on bed rest during the postoperative period. Interestingly, the average days of bed rest among patients who did not have recurrent leaks was 1 day longer than those who had recurrent leaks (1.4 vs 0.3 days, $p = 0.02$). There is limited data on the optimal period for bed rest after CSF leak repair with most surgeons at our institution preferring 0 to 3 days. Among those with bed rest ≥ 2 days, 91% of patients had multilayer CSF leak closure. In patients who had bed rest for ≤ 1 day, the proportion of multilayer closure was 4% (difference of 87%, $p < 0.001$). Similarly, the use of lumbar drains was associated with a higher proportion of bed rest ≥ 2 days (48% vs 8%, $p = 0.002$). No statistical difference was found between bed rest and BMI or age ($p = 0.25$ and $p = 0.076$, respectively).

Long-term outpatient treatment with acetazolamide was utilized in 40% ($n = 19$) of patients with 6% starting this modality immediately postoperatively. Thirty-two patients

Table 2 Location of CSF leak by odds of recurrence

Location ^a	No recurrence	Recurrence	Total	Odds ratio	p-Value
Cribriform	15 (38%)	1 (11%)	16 (33%)	0.2 (0.02–1.8)	0.18
Ethmoid	5 (13%)	3 (33%)	8 (17%)	3.4 (0.6–18.1)	0.15
Central Sphenoid	12 (31%)	1 (11%)	13 (27%)	0.3 (0.03–2.5)	0.26
Sphenoid LTR	6 (15%)	2 (22%)	8 (17%)	1.6 (0.3–9.5)	0.62
Sphenoid-ethmoid	0 (0%)	2 (22%)	2 (4%)	26 (1.1–605)	0.04

Abbreviation: LTR, lateral pterygoid recess.

^aOne patient with unknown leak location that did not recur.

Table 3 MRI findings by odds of recurrence

Finding	No recurrence	Recurrence	Total	Odds ratio	p-Value
Empty sella	11 (52%)	1 (25%)	12 (48%)	0.3 (0.02–3.4)	0.32
Meckel's cave diverticula	5 (26%)	1 (16%)	6 (24%)	1.3 (0.1–18)	0.84
Optic nerve sheath dilation	3 (16%)	0 (0%)	3 (12%)	n/a	0.45
Tortuous/elongated optic nerve	1 (5%)	0 (0%)	1 (4%)	n/a	0.68
Flattened sclera	1 (5%)	0 (0%)	1 (4%)	n/a	0.68
Reverse cupping of optic disc	0 (0%)	0 (0%)	0 (0%)	n/a	n/a
Slit-like ventricle	0 (0%)	0 (0%)	0 (0%)	n/a	n/a

Abbreviation: MRI, magnetic resonance imaging.

Table 4 Perioperative factors associated with odds of recurrence

	No recurrence	Recurrence	Total	Odds ratio	p-Value
Bone graft	8 (21%)	1 (11%)	9 (19%)	0.5 (0.1–4.5)	0.51
Nasoseptal flap	10 (26%)	1 (11%)	11 (23%)	0.4 (0.04–3.3)	0.35
Free mucosal graft	24 (62%)	3 (33%)	27 (56%)	0.3 (0.1–1.4)	0.12
Temporalis fascia	4 (10%)	3 (33%)	7 (15%)	4.4 (0.8–25)	0.08
Acellular dermis	19 (49%)	1 (11%)	20 (42%)	0.1 (0.01–1.2)	0.04
Abdominal fat	1 (3%)	0 (0%)	1 (2%)	n/a	0.63
Multilayer reconstruction	21 (54%)	1 (11%)	22 (46%)	0.1 (0.01–0.9)	0.02
Lumbar drain	11 (28%)	2 (22%)	13 (27%)	0.7 (0.1–4.1)	0.72
Acetazolamide use (perioperative)	2 (5%)	1 (11%)	3 (6%)	2.3 (0.2–28.7)	0.50
Acetazolamide use (outpatient)	14 (36%)	5 (56%)	19 (40%)	2.2 (0.5–9.7)	0.28
VP shunt	0	3 (33%)	3 (6%)	Infinity	0.0002
Bed rest (days)	1.4 ± 1.2	0.3 ± 1.0	1.2 ± 1.2	n/a	0.02

Abbreviation: VP, ventriculoperitoneal shunt.

underwent postoperative lumbar puncture performed on average 170 days (median: 56 days, range: 0–1998 days) after repair. Overall, 59% of patients had elevated opening pressures (>20 cm H₂O) with mean ICP of 22.5 ± 6.4 cm H₂O. Additionally, 62% (16/26) of patients without recurrence had evidence of persistent increased ICP. Neuro-ophthalmology

evaluated 28 patients; seven of which had objective visual field deficits, and two had papilledema. Evaluation occurred on average 96 days after surgery (median: 75.5 days, range: 22–191 days). Mean CSF pressures of patients with papilledema was higher than of those without this finding (35.5 ± 4.9 vs 21.7 ± 5.4 , $p = 0.002$).

Table 5 Select multivariate analysis – logistic regression model

	Recurrence odds ratio	Standard error	p-Value	95% CI lower limit	95% CI upper limit
Age	0.83	0.07	0.041	0.70	0.99
Gender (F)	31.3	73.84	0.145	0.31	3203
BMI	1.22	0.12	0.05	1.00	1.49
Multilayer closure	1.85	5.58	0.84	0.01	678
Lumbar drain	33.4	78.6	0.14	0.33	3367
Days of bed rest	0.18	0.28	0.27	0.01	3.85
Acetazolamide long-term use	4.12	5.55	0.29	0.29	57.7

Abbreviation: CI, confidence interval.

Discussion

Several important observations stem from this analysis. Spontaneous CSF leaks in this study occurred in patients who were primarily middle age, female, and overweight/obese, which is consistent with previous studies.^{6,12–15} Similarly, the most common symptoms in this series were headache and meningitis, along with rhinorrhea, mirrored in previous studies.⁶ Classical clinical signs of IIH include headache and visual symptoms, such as transient obscurations, tinnitus, papilledema, and diplopia.¹⁶ Yet most patients had only headaches as the leak likely provided a release valve of the increased ICP. An empty sella was the most common concurrent MRI finding, suggesting history of underlying increased ICP.¹⁷

The most common leak location was in the cribriform plate and sphenoid sinus. This is also reflective of previous studies.^{18,19} Leak location can occasionally be difficult to classify as pneumatizing patterns in the sinuses are unique among individuals and change with time. In particular, the sphenoid sinus pneumatization can extend quite lateral by increased pneumatization of the pterygoid process, but this was not found to be statistically more likely to be associated with a recurrence of CSF leak. However, leaks in more than one sinus may be associated with an increased chance of recurrence ($p = 0.04$ in the cases of sphenoid–ethmoid leaks). Leaks can span more than one sinus either in a contiguous fashion or be isolated in two discrete locations although this was not characterized well in this study. Meanwhile frontal sinus leaks were not found in this study, but have been described in the literature.²⁰ Defect size was not documented in this study and warrants further study. It can be difficult to definitively see defects on radiologic imaging, and objectively measuring defect size intraoperatively can be similarly difficult.

Many reconstructive techniques have been previously described in the literature; however, there is a paucity of data delineating success rates by reconstructive method for patients with spontaneous CSF fluid leaks. The largest single institution series did not note differences in recurrence rates based on reconstructive method so long as a multilayer closure with free mucosal or pedicled nasoseptal flap overlay was performed. In their series, all recurrent CSF leaks were due to unrecognized elevated ICP or inability to control ICP with medications, thus requiring VP shunting for definitive control.⁶

In the present series, multiple surgeons performed surgical repairs, and thus, technique and perioperative management were not uniform across all surgeons. This afforded the unique opportunity to compare multiple closure techniques as outlined in **Table 4**. On univariate analysis, use of an acellular dermis ($p = 0.04$) and multilayer closure ($p = 0.02$) were found to be favorable factors in repair of CSF rhinorrhea. Sample size limited the ability to detect differences in reconstructive technique on multivariate analysis.

It is well established that most etiologies of CSF leaks (i.e., neoplastic, iatrogenic, and traumatic) have low recurrence rates of <10%. However, spontaneous CSF leaks are known to

have higher rates in the range of 25 to 87%.²¹ Perioperative attempts to reduce recurrence include use of lumbar drains, bed rest, and acetazolamide. The use of lumbar drains and acetazolamide in this study was not found to be significantly associated with recurrence on univariate analysis. The decision for lumbar drain placement and use of acetazolamide was individualized for each patient. Several factors were involved in the decision-making to use lumbar drain during surgery or acetazolamide postoperatively, including elevated BMI, high-flow leak, large defect (>2 cm), history of previous CSF leak, and difficult reconstruction. Due to the retrospective nature of the study, the degree that these factors contributed to the decision cannot be determined and merits future study.

The reasons for lumbar drain placement, timing of drain removal, and immediate acetazolamide use was primarily due to surgeon preference. A previous study has reported the methodical use of lumbar drains in 36 patients.⁹ The protocol involved clamping the lumbar drain prior to removal and measuring the CSF pressure. If the pressure was above 15 cm H₂O, acetazolamide was given prior to removal of the drain. Success rate of 94.4% was reported. Unfortunately, there was no control group without lumbar drains for comparison. The routine use of lumbar drain in spontaneous CSF leak repair remains controversial and should be discouraged given potential risk of complications, including infection, dislodgement, post-spinal headaches, and, rarely retained catheter fragment.

A subsequent study looked at 46 patients with spontaneous CSF leaks, including 21 patients who presented with recurrence after primary surgical intervention at outside institutions.⁶ All had evidence of elevated ICP noted at the time of re-repair with a mean ICP of 32.3 ± 9.0 cm H₂O. CSF pressures were aggressively managed post repair with 50% of patients on acetazolamide and 41% of patients treated with VP shunt. Three patients had recurrence after the discontinuation of acetazolamide. Long-term acetazolamide use was not associated with a decrease in recurrence rate in the present study ($p = 0.28$). The long-term effects of acetazolamide are unknown as studies fail to follow up ICP with extended acetazolamide use.²² A recent Cochrane review concluded that there was insufficient evidence to support the efficacy of acetazolamide in IIH.²³

The mean CSF pressure among patients with recurrence (22.2 ± 7.1 cm H₂O) and those without (22.5 ± 6.4 , $p = 0.90$) were not statistically significant. Although limited by small sample size, it is clear that neither all patients with spontaneous CSF leak have IIH nor do all patients with IIH develop spontaneous CSF leaks. The overall incidence of IIH has been estimated as 0.9 per 100,000/year in the general population and 1.6 per 100,000 women/year.²⁴ The incidence increases as the patient's weight increases from his/her ideal weight.²⁵ Population level estimates for incidence of spontaneous CSF leaks are lacking. If one assumes that all patients in the metropolitan area (population of 6,784,499 in 2013)²⁶ come only to the author's institution for CSF rhinorrhea, the incidence of spontaneous CSF rhinorrhea would be crudely estimated to be 0.14 per 100,000/year, well below

the estimate of the overall population incidence. If a previous study from the author's institution on lateral skull base spontaneous CSF leaks (38 patients) were included in the estimate, the incidence for spontaneous CSF leaks would be 0.25 per 100,000/year. This is likely an underestimation as it does not consider other centers in the metropolitan area that may treat this condition, although no other tertiary care referral center with an otolaryngology residency exist in the metropolitan area. Yet, it also does not account for the population outside the metropolitan area that may travel to seek care at the authors' center, which would decrease the incidence. Nevertheless, some association does exist between IIH, lateral, and anterior skull base CSF leaks.¹⁶

Recent studies have raised the possibility of obstructive sleep apnea as a risk factor for spontaneous CSF leaks. In a meta-analysis of three studies, patients with spontaneous CSF leaks were 2.85 times more likely than those with non-spontaneous CSF leaks to have obstructive sleep apnea (OSA).²⁷ Possible pathophysiologic mechanisms include hypoxia- and hypercapnia-induced elevation in ICP that occurs during sleep.²⁸ This may explain why some patients with spontaneous leaks have normal ICP on lumbar puncture, as the elevation in ICP may only be present during sleep. The use of continuous positive airway pressure (CPAP) for the treatment of sleep apnea may itself predispose people to elevated ICP, as demonstrated in a study where healthy patients were exposed to CPAP. In this study, subjects were found to have transiently elevated CSF pressures while using CPAP. Interestingly, acetazolamide has been found to not only decrease ICP, but also change cortical activity during sleep and have beneficial effects on oxygen saturation and sleep quality.²⁹

Acetazolamide has also been used to treat papilledema seen with increased ICP. In this series, 28 of the patients received neuro-ophthalmologic evaluations, most often due to headaches after the leak repair. Of these, two (7%) patients had papilledema, and although the presence of papilledema was statistically associated with an increase in mean CSF pressures, it is difficult to establish if papilledema by itself is a risk factor for recurrent CSF rhinorrhea given limited sample size. Previous studies have only found a modest association between ICP and higher grades of papilledema.³⁰ Aaron et al³¹ examined the presence of perioperative papilledema in spontaneous CSF leaks. Of 16 patients, none had papilledema. In a separate control cohort of IIH patients without CSF leaks, all had papilledema. The authors hypothesized that in patients with spontaneous CSF rhinorrhea, the leak acted as a release valve reducing the incidence of papilledema.³¹ Therefore, it is a possibility that patients may develop worsening vision after repair. Indeed, this may be the case as 25% of patients evaluated had evidence of visual field deficits, and 7% of patients developed frank papilledema after repair in this study.

Exact prescriptive recommendations for postoperative treatment algorithm cannot be obtained from the present study. This would require inclusion of greater number of patients, preferably in a multicenter prospective clinical trial. Nonetheless, the data suggest overall high success rate with adjunct usage of acetazolamide with a smaller

subset requiring VP shunting to achieve definitive control of CSF rhinorrhea. This carefully selective approach may obviate the need for VP shunting in most patients and, thus, avoid the long-term risks associated with this modality. The data also attests to the utility of ophthalmologic evaluation in the postoperative period to rule out adjunct visual abnormalities. Importantly, it underscores the need to view the repair of spontaneous CSF rhinorrhea in the overall context of the patient with a distinct pathophysiology likely due to elevated ICP. It also highlights the need for close collaboration with neurology and neuro-ophthalmology to proactively monitor ICP and for potential ophthalmologic sequelae in this group of patients.

Conclusion

The present study represents a large experience with intraoperative and postoperative management of patients presenting with spontaneous CSF rhinorrhea. Endoscopic repair was successful in majority of patients, though many require adjunct acetazolamide to control elevated ICP. Only a small subset of patients required adjunct VP shunting for persistent leaks after repair. Long-term ICP monitoring and neuro-ophthalmologic evaluation were employed in a substantial number of cases. Nonetheless, the postoperative management strategy lacks standardization, and further studies are warranted to establish consensus on operative and postoperative management of this important, emerging disease entity.

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Conflicts of Interest

None.

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