Transmastoid Repair of Spontaneous Cerebrospinal Fluid Leaks

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Abstract

Objective Determine the efficacy of using a purely transmastoid approach for the repair of spontaneous cerebrospinal fluid (CSF) leaks and further elucidate the relationship of elevated body mass index (BMI) and skull base thickness in our patient population.

Method We conducted a retrospective chart review of patients treated for spontaneous temporal bone CSF leaks at our tertiary care institution from the years 2006 to 2015. Cases were categorized as primary or secondary. We analyzed success rates, length of stay, use of lumbar drains, BMIs, and rates of meningitis. Skull base thickness was compared with BMI in each case.

Results We identified 26 primary operations for spontaneous CSF leaks and 7 secondary operations. Twenty-three of 33 repairs were performed via the transmastoid approach alone with an 87% success rate (20/23). Of the 10 repairs including a middle cranial fossa (MCF) or combined MCF-transmastoid approach, 2 failed for an 80% success rate (8/10). Five transmastoid repairs underwent placement of a lumbar drain versus all 10 repairs employing an intracranial exposure. Average length of stay for those undergoing a transmastoid approach (1.7 days) was significantly shorter than for patients undergoing a MCF repair (6.3 days). Four patients presented with meningitis. Average BMI was 35.3. No correlation was established between BMI and skull base thickness ($R^2 = 0.00011$).

Conclusion The transmastoid approach is effective in the majority of cases and prevents the need for an intracranial operation, resulting in lower morbidity and a shorter length of stay. We believe that this is the preferred primary approach in most patients with spontaneous CSF leaks.

Keywords

- ▶ encephalocele
- ▶ middle cranial fossa
- ▶ body mass index
- ▶ length of stay
- ▶ lumbar drains
- ▶ transmastoid
- ▶ spontaneous cerebrospinal fluid leak

Introduction

Cerebrospinal fluid (CSF) leaks of the temporal bone, while rare, continue to present a management challenge to the skull base surgeon. Historically, most CSF leaks have been associated with trauma or infection;^{1,2} however, in recent years there has been an increased incidence of spontaneous leaks.^{3,4}

Various theories exist for the etiology of spontaneous leaks. The congenital defect theory postulates that a specific aberrant communication between the subarachnoid space and the temporal bone occurs from abnormal development.⁵ Gacek et al proposed the theory of arachnoid granulations protruding through the tegmen.^{6,7} Regardless of predisposing factor, adult-onset spontaneous CSF otorrhea is now

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thought to arise most often from chronically elevated intracranial pressure (ICP) thinning the tegmen and resulting in bony dehiscence. Obesity has become increasingly common and may lead to intracranial hypertension.^{5,8–13} The association between body mass index (BMI) and skull base thickness is still not clear, but it is postulated that increased ICP is responsible for skull base dehiscence in these patients.^{14,15}

Unlike CSF leaks caused by trauma, spontaneous CSF leaks rarely resolve without treatment, making this a surgical disease.¹³ CSF leaks can be repaired via a middle cranial fossa (MCF), transmastoid, or combined MCF-transmastoid approach. Advantages of the MCF approach are enhanced exposure to the tegmen, preservation of hearing by avoiding contact with middle ear structures, and ease of performing complex multilayered repairs, especially for cases with large or multifocal defects. The advantages of the transmastoid approach are that this is an easier technique to perform, offers simultaneous access to both tegmen and posterior fossa plate, and avoids the risks of craniotomy and temporal lobe retraction. Most neurologists and neurosurgeons agree that an intracranial exposure may be necessary for most cases involving large defects, contracted mastoids offering minimal exposure, history of prior failed repairs, and risk factors for poor healing such as a history of prior radiation.^{16,17} Nevertheless, many skull base surgeons advocate a MCF or combined MCF-transmastoid approach for primary repair of most spontaneous CSF leaks to reduce recurrence rates.¹⁶ The ability to inspect the entire middle fossa floor and to perform a more robust multilayer repair favors this approach. However, despite good outcomes with these techniques, some prefer using a more limited transmastoid approach to avoid an intracranial procedure. Hence, there remains disagreement as to the preferred method of managing spontaneous CSF leaks.

A review of the literature reveals a scarcity of reports on the outcomes of an isolated transmastoid approach.^{3,5} We believe that the transmastoid approach offers an effective method for repair of most spontaneous CSF leaks of the temporal bone, with shorter hospital stays. In this study, we attempt to support this viewpoint with a review of our experience. In addition, we hypothesize that BMI is inversely correlated to skull base thickness in our cohort of patients with spontaneous CSF leak.

Methods

After obtaining approval by Mount Sinai's Institutional Review Board, a retrospective chart review was conducted for all cases of spontaneous CSF otorrhea treated by a single neurotologist (ES) from 2006 to 2015. We recorded demographic data, BMI, presentation, perioperative imaging and audiometry, surgical approach, presence or absence of intraoperative encephalocele and/or CSF leak, site of dehiscence, type of repair, use of lumbar drain, postoperative complications, and length of hospital stay (LOS).

When available, perioperative imaging was reviewed. Lateral skull base thickness was measured using a previously described and validated protocol by Stevens et al.^{18,19}

We measured the tegmen mastoideum at its thickest and thinnest segments, and a fixed midpoint between the posterior semicircular canal and the cortical squamosa on coronal computed tomography (CT) views demonstrated the lateral-most curvature of the posterior semicircular canal (see ►Fig. 1). Similarly, three measurements were made of the tegmen tympani using standard coronal cuts and using a fixed point directly cephalad to the ossicles (see ►Fig. 2). Due to reports of lower inter-rater reliability in the ipsilateral side, we decided to obtain measurements of the contralateral side for each patient.^{18,19} Hence, overall skull base thickness

Fig. 1 Coronal view demonstrating the lateral-most curvature of the posterior semicircular canal. Fixed tegmen measurement at midpoint between the posterior semicircular canal and the cortical squamosa along with thickest and thinnest segments.

Fig. 2 Coronal view of tegmen tympani best depicting the incus, malleus, and two turns of the cochlea. Fixed tegmen measurement directly cephalad to ossicles along with thickest and thinnest segments.

was determined by averaging all six measurements including both tegmen areas. A correlation model was employed to determine the relationship between BMI and skull base thickness.

Postoperative follow-up for each patient was reviewed until the last evaluation on record. The average length of follow-up was 33 months and ranged from 2 to 96 months. Data regarding CSF leak recurrence was gathered and repair failure was defined as any documentation of a recurrent leak at any time after the reconstruction. Overall surgical success rates were calculated for each of the three different approaches used: transmastoid, MCF, and combined MCF-transmastoid approach. LOS was determined by counting hospital days until discharge from day of operation. A *t*-test was used to compare LOS between the transmastoid alone group and those undergoing a transcranial repair.

Surgical Approach

The majority of cases at our institution were repaired using a transmastoid approach, which is our preferred method of repair for all cases with isolated tegmen defects, including those involving encephaloceles (►Fig. 3). This procedure entails a standard postauricular incision and canal wall up mastoidectomy with transmastoid atticotomy. Once the tegmen defect is isolated, healthy dural margins are exposed with gentle blunt elevation of dura in the epidural plane. If an

encephalocele is found, it is fulgurated at its stalk and excised. A multilayered repair is performed by placing a graft of cartilage-perichondrium from the cymbum in the epidural space, with perichondrium against the dural defect. Temporalis fascia is then applied as an overlay graft and held in place with fibrin glue. Fascia is also draped over the aditus to avoid fat herniation into the middle ear cavity. The mastoid cavity is then obliterated with an abdominal fat graft. Fibrin glue is placed between the fat layers. A watertight periosteal closure is performed and a mastoid dressing applied. This dressing is kept in place for the first 48 hours while the patient follows strict CSF leak precautions.

Cases involving difficult-to-access anterior defects, prior failed repairs, and history of prior radiation are preferably repaired using a transcranial approach with or without a mastoidectomy. A linear incision is made in the preauricular line. If a combined approach is to be used, the postauricular incision is joined to this. A limited temporal craniotomy is performed, centered over the defect location, and the temporalis muscle is elevated. An extradural approach is used to expose the dehiscence tegmen, and if an encephalocele is present, this is fulgurated at the stalk with bipolar cautery and excised. The dural defect is repaired using a dural substitute such as acellular dermis (AlloDerm). A calvarial bone graft from the craniotomy bone flap is wedged into the bony defect with a fascial graft underneath. When there is a space between bone and dura, a graft of temporalis muscle or abdominal fat is also placed. Fibrin glue is also used as an added seal. In combined approaches, the mastoid cavity is obliterated with fat and fibrin glue in a similar fashion as with transmastoid repairs. The craniotomy defect is closed with titanium mesh, and the skin is reapproximated in a watertight fashion. If a lumbar drain is used, CSF is removed at 10 cc/hour for 2 days.

Results

A total of 33 cases of spontaneous CSF otorrhea repairs were identified in 28 patients, with a mean follow-up of 32 months. Thirteen patients (46%) were female and 13 patients had left-sided defects. The average age for the group was 52. Twenty-three cases were repaired using a transmastoid approach, six had a middle cranial fossa approach, and four had a combined approach. All but one of the transmastoid repairs were primary procedures. Seven of 10 cases using a transcranial approach were secondary repairs for prior repair failures. Three of the transmastoid repairs failed, for an overall success rate of 87% (20/23). The MCF and the combined MCF-transmastoid approaches collectively had two repair failures for an overall 80% (8/10) success rate. ►Table 1 summarizes the demographics, defect type and location, surgical approach, and success rate for our cohort of patients.

The majority of patients presented with hearing loss and a persistent middle ear effusion. Fifteen patients had tympanostomy tubes inserted by the referring physician and presented with profuse clear otorrhea. Four of the 28 patients (14%) presented with meningitis. In most cases when

Fig. 3 Left ear transmastoid approach for repair of spontaneous CSF leak with encephalocele (blue arrow) secondary to tegmen defect.

Table 1 Summary of results

Demographics				Operative findings and treatment					Follow-Up			
No.	Age	Sex	BMI	Defect location	Approach	Encephalocoele	Lumbar drain	LOS (days)	Secondary repair	Success		
1	52	F	31.4	L single tegmen mastoidea	TM	Yes	Yes	8		No		
	54	F	31.4	L multiple tegmen defects	MCF	Missing	Yes	Missing	Yes	Yes		
2	45	M	38	L single tegmen mastoidea/tympani	TM	Yes	No	1		No		
3	43	M	43	L single tegmen mastoidea	TM	Yes	No	1		No		
	43	M	43	L single tegmen mastoidea	MCF	Yes	Yes	4	Yes	No		
4	55	F	27.3	L single tegmen mastoidea	TM	Yes	No	0		Yes		
5	58	M	35.6	L single tegmen mastoidea	TM	Yes	No	0		Yes		
6	73	F	29.4	L single tegmen mastoidea	TM	Yes	No	2		Yes		
7	13	M	Unclear	R single tegmen mastoidea	TM	No	No	0		Yes		
8	64	M	42.2	R single tegmen mastoidea	TM	Yes	Yes	2		Yes		
9	46	M	41.7	L single tegmen mastoidea	TM	Yes	No	1		Yes		
10	59	M	28.7	L single tegmen tympani	TM	Yes	No	1		Yes		
11	63	M	57.9	R single tegmen mastoidea	TM	Yes	Yes	4		Yes		
12	66	M	31.5	R single tegmen tympani	TM	Yes	No	2		Yes		
13	76	F	44.6	R multiple tegmen mast/posterior plate	TM	Yes	Yes	4		Yes		
14	33	M	35.1	L single tegmen mastoidea	TM	Yes	No	2		Yes		
15	41	F	49.8	R single tegmen mastoidea	TM	No	No	0		Yes		
16	47	F	37.6	R single posterior plate	TM	No	No	1		Yes		
	54	F	37.6	R single posterior plate	TM-MCF	Yes	Yes	3	Yes	Yes		
17	57	M	37.8	L multiple tegmen mastoidea/tympani	TM	No	Yes	6		Yes		
	58	M	38.8	R single tegmen mastoidea/tympani	TM	Yes	No	3	Yes	Yes		
18	50	M	34.2	R single tegmen mastoidea	TM	Yes	No	0		Yes		
19	72	M	30.6	L single tegmen mastoidea	TM	Yes	No	0		Yes		
20	68	F	33.5	R single tegmen tympani	TM	Yes	No	1		Yes		
21	58	F	26.1	L single tegmen mastoidea	TM	Yes	No	0		Yes		
22	36	M	27.5	R single tegmen mastoidea	TM	No	No	1		Yes		
	39	M	27.5	R single tegmen mastoidea	TM-MCF	Yes	Yes	5	Yes	Yes		
23	40	F	30	L single petrous apex	TM-MCF	No	Yes	13		No		

Table 1 (Continued)

Demographics			Operative findings and treatment					Follow-Up		
No.	Age	Sex	BMI	Defect location	Approach	Encephalocele	Lumbar drain	LOS (days)	Secondary repair	Success
24	57	M	28.2	R single tegmen mastoidea	TM-MCF	Yes	Yes	3		Yes
25	52	F	27.2	L multiple tegmen defects	MCF	Yes	Yes	19		Yes
26	39	F	32.8	L multiple tegmen mastoidea/tympani	TM-MCF	Yes	Yes	5		Yes
27	53	F	40	R single tegmen mastoidea	TM-MCF	Yes	Yes	3	Yes	Yes
28	51	F	23.7	R multiple tegmen defects	MCF	Yes	Yes	2	Yes	Yes

Abbreviation: LOS, length of stay; MCF-TM, middle cranial fossa-transmastoid.

otorrhea or rhinorrhea was present, the diagnosis was established by β -2-transferrin assay of the fluid. For most cases, the diagnosis was confirmed on preoperative imaging. CT demonstrated a tegmen dehiscence in the majority of cases, and magnetic resonance imaging (MRI) demonstrated an encephalocele when it was present. There were 24 confirmed encephaloceles at the time of surgery.

Eight patients with transmastoid repair had pre- and postoperative audiograms. In these, average preoperative air-bone gap (ABG) was 29 dB (dB), and the average postoperative ABG was 18 dB. Four of these 8 patients had improvement, and 2 had complete closure, of their ABG. The remaining 4 patients with available studies had postoperative audiogram within 10 dB of preoperative values. The limited number of studies precluded any significant statistical analysis.

Five patients with transmastoid repairs also had lumbar drain placement, and one patient had a lumbar tap immediately prior to surgery. All 10 cases with transcranial repair underwent placement of a lumbar drain. A lumbar drain was placed if the intraoperative CSF leak was profuse or the case was a revision. Mean LOS for the transmastoid and transcranial repairs were 1.6 and 6.3 days, respectively, and this difference was statistically significant ($p < 0.05$, Student's t -test). Nevertheless, a subgroup analysis of five patients who underwent transmastoid repairs with the addition of a lumbar drain demonstrated a mean LOS of 4.8 days, which was not significantly different from that of the transcranial repairs. Overall, the mean LOS was 5.8 days when a drain was used versus 0.9 days without drain placement ($p = 0.001$, Student's t -test), thus lumbar drains likely accounted for the extended LOS. Unfortunately, we did not routinely measure opening pressures, but we generally keep these drains for 48 hours postoperatively in those patients.

Perioperative imaging was available for review on 19 of the 28 patients. Average contralateral skull base thickness and BMI for this group were 1.8 mm and 36.9, respectively. There was no significant correlation between BMI and skull base thickness ($R^2 = 0.00011$). A line of best fit can be seen in ►Fig. 4. Average BMI of the entire cohort was 34.9. A 75% success rate was achieved when pooling both primary and secondary repairs involving patients with BMI equal to

Fig. 4 A line of best fit demonstrating no significant correlation between BMI and skull base thickness ($R^2 = 0.00011$).

or greater than 40. On the contrary, an 86% success rate was achieved when pooling both primary and secondary repairs involving patients with BMI < 40. This difference did not achieve statistical significance.

Discussion

Although prior autopsy studies have demonstrated a high incidence (15–34%) of tegmen defects in random temporal bone specimens, encephaloceles and spontaneous CSF leaks are rare.²⁰ Nevertheless, clinically evident spontaneous CSF otorrhea now appears to be increasing in prevalence as reflected by the current literature.^{3,4} There is an established correlation between spontaneous CSF leaks and obesity, which may explain the rising incidence in the United States.^{8–12,20} The underlying pathogenic mechanism may be occult intracranial hypertension. It is believed that obesity leads to increased intra-abdominal and pleural pressures, resulting in poor venous drainage from the intracranial cavity.⁸ Obstructive sleep apnea, a frequent comorbidity in these patients, can also lead to transient spikes in ICP during apneic episodes.^{11,18} The average BMI in our patients was 35.3, which is significant for morbid obesity according to the World Health Organization.^{21,22}

Chronic intracranial hypertension may result in erosion of the skull base and tegmen dehiscence. Prior studies have demonstrated that, independent of BMI, patients with spontaneous CSF otorrhea have diffuse calvarial thinning in comparison to paired controls.¹⁴ Stevens et al have also reported that patients with skull base thickness of <0.9 mm were more likely to develop recurrence or need a ventriculoperitoneal shunt.¹⁹ In our study, there was no significant correlation between BMI and contralateral skull base thickness, but this result may have been influenced by our sample size.

Controversy persists regarding the best approach for repair of spontaneous CSF leaks.¹⁶ Jackson et al concluded that small tegmen defects are amenable to a transmastoid approach, while large defects should undergo intracranial exposure; however, their series included cases of chronic otitis media or prior surgery as well as spontaneous leaks.²³ Most reports still reveal a preference for MCF approach for the repair of spontaneous CSF leaks,^{4,16} because an intracranial exposure offers total control over the floor of the middle cranial fossa.^{4,9,24} In an attempt to make a smaller craniotomy, proponents of the combined MCF-transmastoid approach argue for a limited subtemporal craniotomy centered over the dehiscence after first identifying the site via mastoidectomy.¹⁶ While this approach may lessen the risk of intracranial complications, there have been reports of postoperative seizures, aphasia, and epidural bleeding with the MCF exposure.^{4,16,23} A combined approach also adds to operative time.

There have been fewer publications on the use of the transmastoid approach alone. In 1995, May et al reviewed 11 cases of spontaneous CSF otorrhea repaired via a transmastoid approach, and only one patient developed a recurrence after 2 years of follow-up.²⁵ In their separate series,

Rao et al and Oliaei et al achieved an 80% and 90% success rate using a transmastoid alone repair to manage 11 cases, respectively.^{26,27} Kari and Mattox reviewed 56 consecutive cases of lateral CSF leak repairs, 33 of which were spontaneous. Although the exact numbers are not provided, the authors report that most of the 56 cases were repaired using a transmastoid approach alone and only one recurrence was reported in the series.³ Kim et al reported on 16 consecutive cases of spontaneous temporal bone CSF leaks repaired via a transmastoid approach alone, with a 93% success rate and a median follow-up of 9 months. They also reported no significant postoperative hearing impairment, with a mean postoperative pure tone average of 29 (± 11 SD) dB and air–bone gap < 12 dB in 14 patients with adequate audiometric data.⁵

Our series presents one of the largest case series favoring the use of the isolated transmastoid approach for the repair of spontaneous temporal bone CSF leaks. Our technique is similar to that described by Semaan et al where a transmastoid extradural-intracranial multilayered repair is performed; however, we also obliterate the mastoid cavity with an abdominal fat graft.²⁸ We believe that our success rate of 86% supports this as the best initial approach to correct the skull base defect and avoid a craniotomy. This approach does not preclude revision surgery using a MCF exposure. Although we observed a lower overall repair success rate using MCF, this does not imply that MCF is inferior to transmastoid approach. These MCF cases were the more difficult cases, including 7/10 secondary repairs after prior failed attempts. We do not question the fact that the MCF approach has the highest chance of success, but we do advocate a transmastoid approach in uncomplicated cases because of its lower potential morbidity and shorter LOS.

Lumbar drains are not routinely used, but can be employed in high-flow leaks or revision cases (in our series, drains were placed in 21% of the transmastoid versus 100% of transcranial cases). We observed a significantly shorter LOS of 1.6 days using a transmastoid approach versus 6.3 days using a transcranial exposure, so this was a more cost-effective approach in our series. However, other authors have reported shorter hospital stays for patients undergoing intracranial procedures, and this may vary between institutions.¹⁶ Placement of lumbar drains likely explains the difference in hospital stay observed in our series, and the surgeon's judgment plays an important role on the use of drains. Despite adequate surgical treatment, two of our cases failed MCF repair and required a ventriculoperitoneal shunt. One of these patients had evidence of obstructive hydrocephalus, while the other had benign intracranial hypertension with elevated ICPs >20 mm Hg. This subset of patients should undergo multidisciplinary evaluation by neuro-ophthalmology, neurosurgery, and neurology.²⁶

Furthermore, although not analyzed in our study, a transmastoid repair is likely to require less intraoperative time than an intracranial repair, especially a combined MCF-transmastoid repair. Considering that a majority of our patients were obese, with average BMI of 35.3, the need for prolonged anesthesia time is an important factor in this population.

As in prior reports, most of our patients with spontaneous CSF otorrhea had preoperative conductive hearing loss, which was usually secondary to middle ear effusion.²⁸ Although we had incomplete data, our observed rate of postoperative hearing loss was in keeping with the available literature on hearing outcomes following transmastoid repair.^{5,27} Most patients in our series had good hearing outcomes—four out of eight patients had improvement in ABG, and the remainder had minimal to no change in their hearing. It is possible that the fat graft impeded the conductive mechanism; it is also possible that middle ear fibrosis played a role.

In conclusion, we believe that the transmastoid approach is the preferred approach for the primary repair of most spontaneous lateral skull base CSF leaks. Most tegmen defects in our series occurred at the tegmen mastoideum (18/23), and even those that occurred at the tegmen tympani were accessible through the mastoid. It is still prudent to select a MCF or combined MCF-transmastoid approach for cases with inaccessible anterior defects, for prior failed cases, and for cases with prior radiation.²⁹

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