Am J Otolaryngol 41 (2020) 102560

Contents lists available at ScienceDirect

Am J Otolaryngol

journal homepage: www.elsevier.com/locate/amjoto

Surgical management of middle cranial fossa bone defects: meningoencephalic herniation and cerebrospinal fluid leaks

Elena Hernandez-Montero*, Erika Caballero, Luis García-Ibanez

Instituto de Otología Garcia-Ibanez, C/Dr. Roux 91, Bajos, Barcelona 08017, Spain

ARTICLE INFO

Keywords:
Meningoencephalic herniation
Cerebrospinal fluid (CSF) leak
Temporal bone defect
Middle cranial Fossa

ABSTRACT

Along the medical practice of an Otologist he/she will face middle cranial fossa (MCF) bone defects. The purpose of this study is to contribute to the understanding of this possible life threatening condition, and to share and discuss our management approach. A literature review is also presented.

Study design: Retrospective case series at García-Ibáñez Otology and Skull base private center referral. METHODS: This study is based on the analysis of data collected from 19 cases of temporal bone meningoencephalic herniations surgically treated from 2006 to 2018. The follow-up ranged from 18 to 162 months with a mean average of 44.5 months.

Main findings: Meningoencephalic herniations were divided into four etiologic groups: spontaneous (24.8%), secondary to chronic otitis media (21.8%), iatrogenic (45.9%), and posttraumatic (7.5%). Different surgical techniques were used as treatment: transmastoid (TM) approach (27.8%), MCF approach (27.8%), combined technique (transmastoid plus minicraniotomy, 3%), and middle ear obliteration with blind sac closure of the external auditory canal (41.4%).

Conclusions: Variables like bilateral hearing level, size and location of the bone defect and existence of CSF leak should be analyzed to select the safest and most effective closing surgical approach.

1. Introduction

Tegmen tympani and mastoid tegmen are components of the temporal bone, being the roof of the middle ear and floor of the MCF respectively. The bone defects of the tegmen tympani and mastoid tegmen may predispose to the development of meningoceles, meningoencephaloceles and cerebrospinal fluid (CSF) leaks [1,2] Spontaneous CSF leak and encephaloceles, although uncommon, are important cause of conductive hearing loss, otoliquorrhea, aural fullness and middle ear effusion [3].

Although thinning of the tegmen cortex appears to be common in autopsy studies, ranging from 15% to 34% [4], the occurrence of spontaneous CSF leak is infrequent [1]. Several mechanisms have been identified for the thinning of the bone tegmen, including congenital defects, chronic middle ear disease, intracranial hypertension, trauma, cerebrospinal pulsation against the skull base, obesity, iatrogenic and idiopathic [1,2]. Many authors advocate the presence of granulations as the most important factor, compromising both tegmen and dura, while cholesteatoma may cause local ischemia and dural enzymatic degradation [5,6].

Temporal bone MEH requires early surgical repair, because the rates of meningitis, encephalitis and brain abscesses are considerably increased, threatening patients' life [5,7].

2. Material and methods

This study describes a series of 18 patients (19 ears) treated at García-Ibáñez Otology Institute between 2006 and October 2018, with surgical repair of the tegmen defect and temporal bone MEH and their pathogenesis, clinical presentation and surgical approach. A literature review is also performed.

The variables analyzed were age, sex, side, clinical presentation, etiology, preoperative and postoperative Pure Tone Audiometry (PTA) and contralateral ear condition.

We grouped the tegmen defect into four categories based on the etiology: Spontaneous, chronic otitis media/Cholesteatoma (COM/Ch), traumatic and iatrogenic.

Intraoperatively we collected the following data: surgical approach,

Abbreviation: MCF, Middle Cranial Fossa; MEH, Meningoencephalic Herniations; COM/Ch, Chronic Otitis Media/Cholesteatoma; TM, Transmastoid; CSF, Cerebro Spinal Fluid

* Corresponding author at: Av. Josep Tarradellas 19-21; 6-2, Barcelona 08029, Spain.
E-mail address: ehernandez.hv@gencat.cat (E. Hernandez-Montero).

https://doi.org/10.1016/j.amjoto.2020.102560
Received 5 April 2020

Table 1
Clinical presentation.

	Spontaneous	COM/Ch
Hearing loss	5 (83.33%)	11 (84.61%)
Fullness	2 (33.33%)	2 (15.38%)
Sudden	1(16.6%)	0
Vertigo	2 (33.33%)	4 (30.76%)
Unsteadiness	1 (16,67%)	0
Otorrhea	0	7 (53.84%%)
Otoliquorrhea	4 (66.6%)	2 (15.38%)
Tinnitus	0	2 (15.38%)
Meningitis	0	1 (7.69%)
Duration of symptoms in months	24	28,7

size of the defect, materials used for repair, associated middle ear surgeries, complications, recurrences and follow-up.

All patients had an oto-neurological and facial nerve assessment preoperatively.

3. Results

Average age was 48 years, ranging 21–68 y.o. There were 12 males (66. 3%) and 6 females (33.3%). 15 cases affected right ears (78.94%) and 4 cases (21.05%) left ears. Average body mass index (BMI) was 35.4 kg/m^2 (class II obesity).

Clinical presentation is summarized in Table 1.

The etiology was spontaneous in 6 patients (31.58%) and related to COM/Ch in 13 patients (68.42%). One patient had a history of abrupt Valsalva maneuver, but none had suffered cranial trauma or iatrogenic events.

Considering etiology, 13 cases (68.42%) had a history of COM/Ch: one of these cases had not been operated before. 3 cases presented supracochlear cholesteatoma; 2 of them with superior canal dehiscence (SCDS). This last finding (SCDS) was also present in 3 patients who had cholesteatomas that did not contact the superior canal and in one patient with idiopathic tegmen defect.

13 cases were referred from other centers with a history of previous otologic surgery: 9 cases had one previous timpanoplasty (69.21%); 2 cases (15.38%) had been submitted to timpanoplasty twice; 2 cases (15.38%) had myringotomy and tube placement because of false middle ear otitis with effusion. It is not possible to prove if some of them were iatrogenic. 2 cases (15.38%) presented recurrent cholesteatoma.

The interval between initial symptoms and diagnosis of the MEH was 2.6 years, ranging from 2 months to 10 years. The interval between previous surgery and surgery of the defect was three years; ranging from one to five years.

Otomicroscopic findings at first consultation are collected in Table 2.

Table 2
Otomicroscopic findings at first consultation. (EAC: External Auditory Canal; VT: Ventilation Tube).

	Spontaneous N:6	Cholesteatoma/CMO N: 13
Normal otomicroscopy	0	1
Atical retraction	0	4
Middle ear with content	0	1
Pulsatile mass	0	2
Atical scab	0	2
Otoliquorrhea	3	1
Otorrhea	0	2
Epidermal pearls	0	4
Polyps in EAC	0	2
Exostosis	3	0
Flaccid eardrum	2	0
VT	2	0

Table 3
Intraoperative findings of tegmen defects.

Meningocele + CSF leak	6 patients
Tegmen defect + CSF leak	5 patients
Only tegmen defect	5 patients
Meningocele	1 patient
Meningoencephalic herniation	2 patients

Preoperative Pure Tone Audiometry (PTA) showed conductive hearing loss in 10 cases (52.63%), mixed hearing loss in 8 cases (42.10%) and sensorineural hearing loss in 1 case (5.26%): this one was a profound hearing loss.

Location of the defect was mastoid tegmen in 2 cases, (10.53%), tegmen tympani in 8 (41.11%), and both in 9 (47.37%).

The size of the defect was less than 1 cm in 5 cases (26.32%), all of them with CSF leak, defect 1 cm to-2 cm in 9 cases (47.37%) and more than 2 cm in 5 cases (26.32%). The biggest defect found was 4 cm in 1 case.

Table 3 shows intraoperative findings of the tegmen defects:

Concerning the surgical approach, we performed 1 case (5.26%) through TM approach, 7 through MCF approach (36.84%) and 11 (57.89%) using a combined approach.

The materials used for reconstruction are summarized in Table 4.

We found 4 patients with bilateral defects of the tegmen in our series (Fig. 1). Amongst these 8 ears (42.10%), 3 of them were of idiopathic origin, presenting tegmen defects with CSF leak. 1 had a meningocele. 2 cases presented small tegmen defects (less than 1 cm) without CSF leak who are under control. Another patient had a bilateral meningocele due to COM, which had been tried to be repaired before unsuccessfully: one of the ears has already been re-operated and the other one will be planned soon.

Postoperative PTA showed profound sensorineural hearing loss in 3 cases (15.8%): in all of them resection of a supracochlear cholesteatoma was practiced. In 6 cases (31.6%) the conductive hearing loss improved and worsened or did not have changes in 3 cases (15.8%); mixed hearing loss improved in 2 cases (10.53%) and worsened in 4 cases (31.6%).

Follow–up was achieved postoperatively with high-resolution Computed Tomography (CT) at 6 and 12 months. No recurrence of the tegmen defect was detected. Only 1 patient had recurrent cholesteatoma.

The algorithm of treatment proposed by the authors of this paper is:

- Bone tegmen defects smaller than 1 cm without CSF leak do not need repair
- Bone tegmen defects between 1 and 2 cm with or without fistula or smaller than 1 cm with CSF leak are repaired through a TM approach
- Bone tegmen defects bigger than 2 cm with or without CSF leak are repaired through a combined approach.
- Any bone tegmen defect with or without CSF leak, previously unsucessfully repaired: combined approach.

The choice of the surgical approach depends on the etiology, location and size of the bone defect, presence of CSF leak, chronic middle ear infection and hearing level.

Table 4
Materials used for repairing. F: Temporalis fascia; C: Cartilage; B: Bone.

	Spontaneous	CMO/Ch
Fascia (F)	2 (33.33%)	2 (15.38%)
Cartilage (C)	1 (16.67%)	3 (23.08%)
C + F	3 (50%)	6 (46.15%)
C + F + B	0	2 (15.38%)

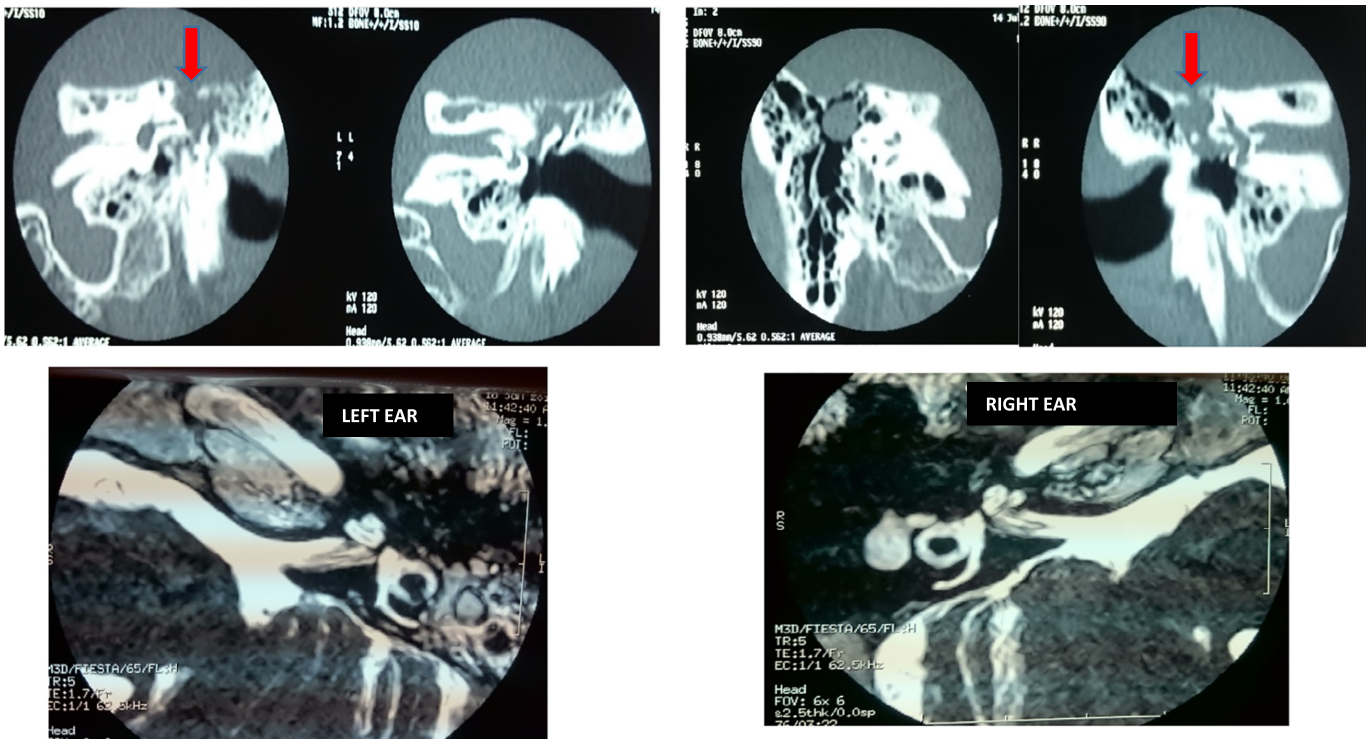

Fig. 1. Coronal CT and axial T2 weighted MRI showing: bilateral mastoid tegmen defect (arrow). Right ear: pediculated meningoencephalocele Left ear: mastoid effussion.

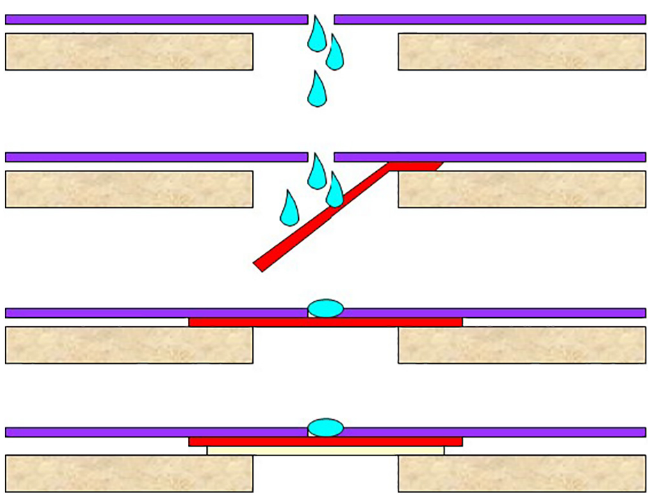

Fig. 2. Scheme of multilayer reparation technique with a transmastoid approach.

The **TM approach** consists in dissecting the temporal dura 0.5 cm all around the bone defect with the help of a blunt instrument to create a space between dura and bone, so that the materials chosen for the closure can be introduced and stabilized in different layers (Fig. 2).

In **the middle fossa mini-craniotomy approach** a 2 by 3 cm window is created in the squamous portion of the temporal bone, centered in the root of the zygoma. Temporal dura is dissected from the floor of the middle cranial fossa to expose the tegmen defect completely. The selected materials are introduced and stabilized with fibrin glue. This approach is optimal for tympanic defects and avoids ossicular chain manipulation.

The **combined approach** consists in the association of the two above described procedures. It is the preferred approach for the biggest defects, meningoceles, meningoencephaloceles and failed cases (Fig. 3).

The materials we used for closure depended on the size of the defect and the presence or not of CSF leak: for defects smaller than 1 cm we used temporal fascia; for defects between 1 and 2 cm we used temporal fascia and auricular cartilage; those bigger than 2 cm. were repaired by using auricular cartilage, bone from the craniotomy and temporal fascia. When CSF leak is associated, even in defects smaller than 1 cm, a multilayer technique is indicated. The adherence of the materials to the brain tissue is favored by a fibrin glue.

4. Discussion

The thinning of the tegmen cortex appears to be common in autopsy studies, ranging from 15% to 34% [4] but the occurrence of the tegmen tympani or mastoid defect, associated to CSF leaks, meningocephaloceles and meningoceles in the middle ear or mastoid is rare [5,6]. Often insidious in onset, usually only occurs ipsilateral though Kenning TJ [1] found that 43% of cases were bilateral, which is consistent with our series (42.10%). Other authors report a bilaterality of 8.3% and 15% [2,8].

Spontaneous CSF leak and encephaloceles are an uncommon but important cause of conductive hearing loss, CSF otorrhea, aural fullness and middle ear effusion [3,9]. The diagnosis of defect tegmen tympani or mastoid was preoperative in all of our cases, except for one case who had an admission for meningitis for medical treatment.

Several mechanisms have been proposed for the thinning of the bone tegmen, including congenital defects, intracranial hypertension, Obstructive Sleep Apnea [9] trauma, iatrogenic, idiopathic, presence of aberrant arachnoid granulations [10], cholesteatoma and chronic middle ear disease causing local ischemia and dural enzymatic degradation [5,6] obesity, etc. Average body mass index (BMI) in our series was 35.4 kg/m^2 which stands for obesity.

The use of preoperative lumbar drain (LD) is controversial and there is currently no consensus about it. In addition, LD has been associated with increased rates up to 12.3% including meningitis, pneumocephalus, persistent headache, uncal herniation and lumbar

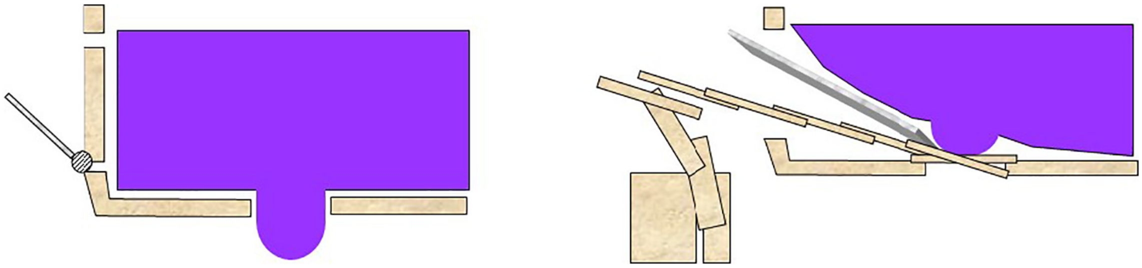

Fig. 3. Scheme of multilayer reparation technique with a combined approach.

radiculopathy [11]. We do not use LD routinely in our patients.

The most prevalent etiology in our series was COM/Ch, similar to what Grinblat G et al. [12] report. Sanna et al. [2] communicated traumatic etiology as the most frequent one in their series. This fact could be explained because we are a private referral center where traumatic emergencies are not usually attended; for Carlson et al. [8] the spontaneous etiology was the most prevalent.

The cranial base should be examined radiologically with high-resolution computer tomography with axial and coronal sections to define location and size of the bone defect, but it does not allow seeing the continuity of the soft tissue with the tegmen defect. Besides, it does not distinguish amongst granulation, cholesteatoma, cholesterol granuloma or other soft tissue masses inside the middle ear cavity [1,2,8,13,19].

MRI is the ideal method for identifying hernia brain tissue in middle ear cavity; herniated meningoencephalic tissue is seen as a mass isointense to brain in all sequences. Cholesteatoma is seen hypointense in T1 and hyperintense in T2 and cholesterol granuloma appears hyperintense in both T1 and T2. Contrast administration enhances only granulation tissue [14]. Diffusion weighted sequences allow cholesteatoma detection [15].

Audiometric results were variable. This fact is explained because the hearing loss is affected by both the chronic ear disease and the MCF defect and not only due to the latter. Conductive and mixed hearing loss are the most frequent types of hearing loss in patients with tegmen defect [2,12].

The prevalence of SCDS has been reported to be 30 times higher in patients with spontaneous CSF leak [3,8,16]. A high rate of concomitant tegmen dehiscence (up to 76%) in patients with SCDS, support of a congenital theory representing two different manifestations of a lateral skull base disorder [17]. We found 6 cases of SCDS in the same ear of the tegmen defect and/or in the contralateral one.

The recurrence rates of leaks after transmastoid approach range from 6 to 20% [18,19] reason why some authors prefer the mini-craniotomy approach to have a better evaluation of the entire tegmen, since up to 50% of patients present multiple tegmen defects [20]. The combined approach was the one we used the most, representing in our practice the most effective closure method. We did not perform middle ear obliteration in any of our patients.

Besides, the multilayer closure is usually necessary. Autologous materials such as fascia, bone grafts, cartilage, bone pate, and vascularized flaps have the advantages of availability, low risk of extrusion or infection; some authors use bone pate, vascularized flaps and other materials that increased risk of infection and cost [2,21,22].

Sanna et al. [2] found a closing rate of 75.4% if only a single layer was placed and a 100% closing rate with multilayer closure. We systematically use a multilayer technique and achieve a 100% closing rate, comparable to previously published reports [1,2,8].

5. Conclusions

The bone tegmen defects are potentially life threatening and require early surgical repair. The surgical procedure is safe and, by selecting the right approach and using a multilayer technique, closure rates of 100% are the norm.

Author contribution

Hernandez Elena: Conceptualization; Writing-review&editing.
Caballero Erika: Methodology; Data curation; Writing-original draft.
Garcia-ibanez Luis: Supervision; Project administration; Software.

Declaration of competing interest

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

[1] Kenning TJ, Willcox TO, Artz GJ, Schiffmacher P, Farrell CJ, Evans JJ. Surgical management of temporal meningoencephaloceles, cerebrospinal fluid leaks, and intracranial hypertension: treatment paradigm and outcomes. Neurosurg Focus 2012;32(6):E6.
[2] Sanna M, Fois P, Paolo F, Russo A, Falcioni M. Management of meningoencephalic herniation of the temporal bone: personal experience and literature review. Laryngoscope 2009;119(8):1579–85.
[3] Kutz JW, Johnson AK, Wick CC. Surgical management of spontaneous cerebrospinal fistulas and encephaloceles of the temporal bone. Laryngoscope 2018;128(9):2170–7.
[4] Pachauri R, Nagaonkar S, Kirtane MV. Spontaneous CSF otorrhoea - presenting as conductive deafness. Indian J Otolaryngol Head Neck Surg Off Publ Assoc Otolaryngol India 2002;54(1):51–4.
[5] Mosnier I, Fiky LE, Shahidi A, Sterkers O. Brain herniation and chronic otitis media: diagnosis and surgical management. Clin Otolaryngol Allied Sci 2000;25(5):385–91.
[6] Manolidis S. Dural herniations, encephaloceles: an index of neglected chronic otitis media and further complications. Am J Otolaryngol 2002;23(4):203–8.
[7] Sdano MT, Pensak ML. Temporal bone encephaloceles. Curr Opin Otolaryngol Head Neck Surg 2005;13(5):287–9.
[8] Carlson ML, Copeland WR, Driscoll CL, Link MJ, Haynes DS, Thompson RC, et al. Temporal bone encephalocele and cerebrospinal fluid fistula repair utilizing the middle cranial fossa or combined mastoid-middle cranial fossa approach. J Neurosurg 2013;119(5):1314–22.
[9] Nelson RF, Roche JP, Gantz BJ, Hansen MR. Middle cranial Fossa (MCF) approach without the use of lumbar drain for the management of spontaneous cerebral spinal fluid (CSF) leaks. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2016;37(10):1625–9.
[10] Gacek RR, Gacek MR, Tart R. Adult spontaneous cerebrospinal fluid otorrhea: diagnosis and management. Am J Otol 1999;20(6):770–6.
[11] Stokken J, Recinos PF, Woodard T, Sindwani R. The utility of lumbar drains in modern endoscopic skull base surgery. Curr Opin Otolaryngol Head Neck Surg 2015;23(1):78–82.
[12] Grinblat G, Dandinarasaiah M, Prasad SC, Piras G, Piccirillo E, Fulcheri A, et al. Temporal bone meningo-encephalic-herniation: etiological categorization and surgical strategy. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2018;39(3):320–32.
[13] Souliere CR, Langman AW. Combined mastoid/middle cranial fossa repair of temporal bone encephalocele. Skull Base Surg 1998;8(4):185–9.
[14] Bovo R, Ceruti S, Padovani R, Martini A. Temporal bone brain herniation. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2006;27(4):576–7.
[15] De Foer B, Vercruysse J-P, Bernaerts A, Deckers F, Pouillon M, Somers T, et al. Detection of postoperative residual cholesteatoma with non-echo-planar diffusion-weighted magnetic resonance imaging. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2008;29(4):513–7.
[16] Allen KP, Perez CL, Kutz JW, Gerecci D, Roland PS, Isaacson B. Elevated intracranial pressure in patients with spontaneous cerebrospinal fluid otorrhea. Laryngoscope 2014;124(1):251–4.
[17] Stevens SM, Hock K, Samy RN, Pensak ML. Are patients with spontaneous CSF

otorrhea and superior canal dehiscence congenitally predisposed to their disorders? Otolaryngol–Head Neck Surg Off J Am Acad Otolaryngol-Head Neck Surg 2018;159(3):543–52.

[18] Kim L, Wisely CE, Dodson EE. Transmastoid approach to spontaneous temporal bone cerebrospinal fluid leaks: hearing improvement and success of repair. Otolaryngol–Head Neck Surg Off J Am Acad Otolaryngol-Head Neck Surg 2014;150(3):472–8.

[19] Leonetti JP, Marzo S, Anderson D, Origitano T, Vukas DD. Spontaneous transtemporal CSF leakage: a study of 51 cases. Ear Nose Throat J 2005;84(11):700, 702–4, 706.

[20] Gubbels SP, Selden NR, Delashaw JB, McMenomey SO. Spontaneous middle fossa encephalocele and cerebrospinal fluid leakage: diagnosis and management. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2007;28(8):1131–9.

[21] Sanna M, Dispenza F, Flanagan S, De Stefano A, Falcioni M. Management of chronic otitis by middle ear obliteration with blind sac closure of the external auditory canal. Otol Neurotol Off Publ Am Otol Soc Am Neurotol Soc Eur Acad Otol Neurotol 2008;29(1):19–22.

[22] Schurr PH. Endaural cerebral hernia. Br J Surg 1960;47:414–7.