RESEARCH PAPER

Neurologia i Neurochirurgia Polska
Polish Journal of Neurology and Neurosurgery
2024, Volume 58, no. 1, pages: 54–59
DOI: 10.5603/pjnns.97522

ISSN: 0028-3843, e-ISSN: 1897-4260

LEADING TOPIC

Leading Topic Editor: Olga P. Fermo, MD, Department of Neurology, Mayo Clinic, Jacksonville, Florida, United States

Recurrence of cerebrospinal fluid-venous fistulas at different spinal levels following transvenous embolisation or blood/fibrin glue patching

Roaa Zayat[1], Olga P. Fermo[1], Thien J. Huynh[2]

[1]Department of Neurology, Mayo Clinic, Jacksonville, Florida, United States
[2]Department of Radiology, Mayo Clinic, Jacksonville, Florida, United States

ABSTRACT

Aim of the study. This study presents cases of recurrent cerebrospinal fluid-venous fistulas (CVFs) de novo at a different spinal level following successful treatment of initial CVFs. The aim was to highlight this rarely described phenomenon and report the clinical and imaging features after initial treatment, providing insights into the dynamics of recurrent CVFs.

Clinical rationale for the study. Understanding the course of CVFs post-treatment is crucial for optimising patient management, especially when symptoms persist or recur.

Material and methods. We performed a retrospective chart review of all patients with recurrent CVFs at a different level after treatment of their initial CVF at our institution. Clinical and imaging records were reviewed and summarised, including Bern score features on brain magnetic resonance imaging (MRI) before and after treatment.

Results. Four patients with five recurrent CVFs were identified. Recurrent or persistent symptoms encouraged subsequent brain MRI scans, which revealed different outcomes: i.e. persistence, or improvement, or complete resolution of abnormal findings. Initial positive responses included improvement of the pachymeningeal enhancement and venous sinus distension. These improvements were reversed when recurrent symptoms arose, which was also correlated with changes in the Bern score.

Conclusions and clinical implications. Recognising the factors of CVF recurrence is crucial for comprehensive management. This study underlines the significance of repeated evaluation of persistent or recurring symptoms of CSF leak after treatment for CVFs.

Keywords: cerebrospinal fluid-venous fistula, spontaneous intracranial hypotension, Bern score, recurrence, myelography, embolisation, patch

(*Neurol Neurochir Pol 2024; 58 (1): 54–59*)

Introduction

Cerebrospinal fluid-venous fistulas (CVF), also known as type III cerebrospinal fluid (CSF) leaks, are spontaneous abnormal connections that form between the spinal subarachnoid space and a paraspinal vein. This results in unregulated CSF volume loss and spontaneous intracranial hypotension (SIH). The diagnosis of SIH is often established via an MRI of the brain showing diffuse smooth pachymeningeal enhancement and brainstem sagging secondary to insufficient CSF volume [1–3]. Bern SIH score is a radiological scoring system that predicts the probability of identifying a spinal CSF leak source associated with SIH based on six findings on brain MRI including pachymeningeal enhancement, effaced suprasellar

Address for correspondence: Thien J. Huynh, MD, Mayo Clinic, 4500 San Pablo Rd., Jacksonville, FL 32224, USA; e-mail: Huynh.Thien@mayo.edu
Received: 21.09.2023 Accepted: 14.12.2023 Early publication date: 31.01.2024

cistern, venous sinus distension, decreased mamillopontine distance, effaced prepontine cistern, and the presence of subdural fluid collections [4] (Tab. 1). Identification and localisation of CVFs requires advanced myelographic techniques including lateral decubitus dynamic computerised tomography myelography (CTM) or digital subtraction myelography (DSM) [2, 3]. CVFs can be treated via surgical ligation, fibrin glue/blood patching, or, the most recently devised method, transvenous embolisation.

De novo recurrence of CVF at a new level after treatment has been recently described after surgery [5], but it has not been well described following transvenous embolisation or fibrin patching. Furthermore, only a few cases of recurrent CVF have been described. The purpose of our study is to report four cases of recurrent CVF at new levels after successfully treated initial CVF, highlighting clinical and imaging features after treatment and prior to the discovery of a new CVF.

Clinical rationale for the study

Recurrent CVFs pose a rarely discussed clinical challenge. Understanding the dynamics and clinical and imaging features of this phenomenon will guide more effective diagnostic and management strategies, and ensure optimal patient care.

Material and methods

Institutional Review Board (IRB) approval for this study was obtained. A retrospective chart review of all patients with CVFs at our institution was performed. The charts of patients who had recurrent CVF at a different level after treatment for their initial CVF were reviewed to collect the clinical and imaging features (Tab. 2). The successful treatment of their initial fistulas was proved by the absence of the initial CVF upon repeat myelography.

Table 1. Bern SIH score

Feature	Point(s)	Scoring	
Engorgement of venous sinuses	2	≤ 2	Low
Pachymeningeal enhancement	2		
Suprasellar cistern ≤ 4 mm	2	3–4	Intermediate
Subdural fluid collections	1		
Prepontine cistern ≤ 5 mm	1	≥ 5	High
Mamillopontine distance ≤ 6.5 mm	1		

Table 2. Characteristics of initial and recurrent CSF-venous fistula

Age/ /sex	Initial CVF level and side	Treatment	Symptoms improved post--treatment?	Time to symptom recurrence	Follow-up brain imaging findings	Recurrent evel
Current study						
68M	R T5	Embo	Partial	4 w	1 m: partially improved (initial Bern 5 to 3 at follow-up) 3 m: subtle worsening dural enhancement, improved from baseline (Bern 3)	R T7
60F	L T9	Embo	Yes	15 m	3 to 15 m: progressive resolution of dural enhancement and brain sagging (initial Bern 8, then 4 at 3 months, 2 at 12 and 15 months) 19 m: worsening dural enhancement, brain sagging, venous distension (Bern 8)	L T6
78M	R T6	Fibrin Patch	No	10 d	1 m: unchanged (initial Bern 4 and unchanged at follow-up)	R T9
50F	R T7	Embo	Yes	4 m	1 m: partially improved (initial Bern 8, improved to 5)	R T12
					7 m: worsened, still improved from baseline (Bern 6)	
	R T12	Embo	No	Immediately	1 m: partially improved (Bern 5) 7 m: similar to previous (Bern 5)	R T8
Malinzak et al. 2021 [5]						
60F	R T6	Surgery	Yes	3 m	3 m: unchanged	R T5 (6 m)
67M	L T10	Surgery	Yes	5 m	2 m: partially improved 5 m: worsened, subdurals	L T9 (24 m)
56F	L T11	Surgery	Yes	4 m	15 m: partially improved	L T8 (13 m)
51F	L T7	Surgery	Yes	1.5 m	2 m: unchanged	L T10 (10 m)

d — days; F — female; m — months; M — male; R — right; L — left; T — thoracic; w — weeks

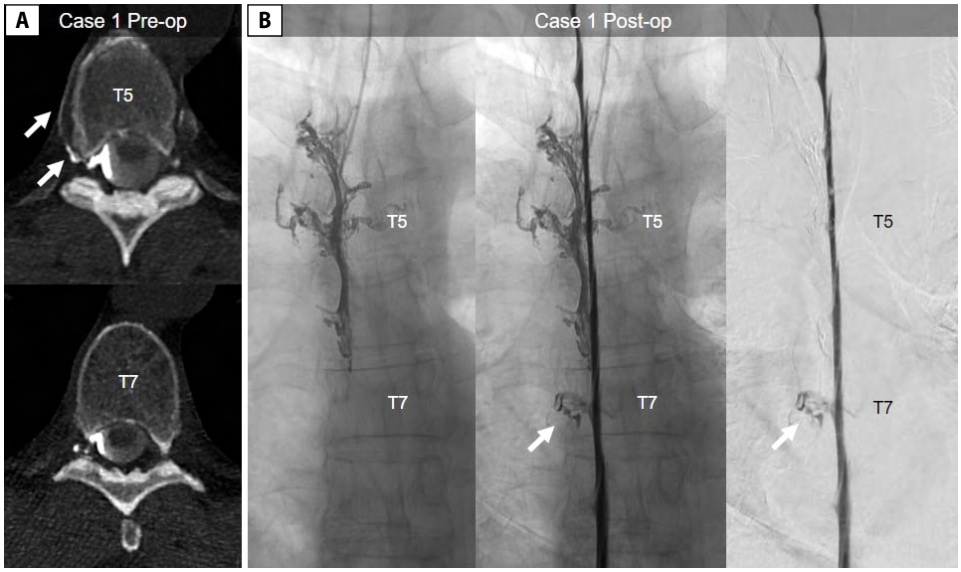

Figure 1. Patient 1. **A.** Pre-embolisation of T5 CVF: right lateral decubitus CTM showing CVF at right T5 (arrow) with no evidence of CVF at right T7 level. **B.** Post-embolisation of T5 CVF: lateral decubitus DSM showing new CVF at right T7 level (arrows) with no evidence of residual leak at embolised right T5 level

Results

A total of 42 patients with CVFs were identified at our institution. After the first treatment, either with fibrin glue/blood patching or transvenous embolisation, 10 patients had residual or recurrent symptoms.

Four of these 10 patients had new CVF at a new level confirmed on myelography, and the details of these four are presented in this research paper.

Five patients had suspected or confirmed residual CVF at the initial level, and one patient had further work-up pending.

Patient 1

A 68-year-old male presented with a 1.5-year history of progressive neck pain, frontal headache, tinnitus, imbalance and photosensitivity that are Valsalva-induced and exacerbated when in an upright position or coughing. Brain MRI showed thick smooth pachymeningeal enhancement, venous distension, and effaced prepontine cistern (Bern score 5). Right lateral decubitus CTM identified a CVF at the right T5 level, treated with transvenous embolisation (Fig. 1). The symptoms improved slightly over four weeks, but gradually returned. Brain MRI four weeks after treatment showed improved but persistent thin pachymeningeal enhancement, improved venous distension, and persistent prepontine cistern effacement (Bern score 3). There was concern for persistent CVF surrounding the embolisation cast, and a transforaminal fibrin glue patch was performed at T5 three months post-embolisation. This however did not improve the symptoms. Repeat brain MRI at five months showed subtly worsened pachymeningeal enhancement without venous distension (Bern score 3); there was a slight increase in pituitary size although the suprasellar cistern remained > 4 mm in height. At eight months post-treatment, lateral decubitus DSM showed new CVF at the right T7 level without residual leak at the previously embolised level at right T5. Additional embolisation of the right T7 CVF resulted in partial improvement of symptoms. A brain MRI four months after the last treatment showed slight improvement in pachymeningeal enhancement and decreased size of the pituitary gland (Bern score 3).

Patient 2

A 60-year-old female presented for evaluation of a 5-year history of orthostatic headaches exacerbated by Valsalva, laughing, and coughing. Brain MRI showed brainstem sagging, pachymeningeal enhancement, venous sinus distension, decreased mamillopontine distance, effaced prepontine cistern, and effaced suprasellar cistern (Bern score 8). Lateral decubitus dynamic CTM showed clear CVF at left T9 level (Fig. 2). Transvenous embolisation resulted in a complete resolution of symptoms. Brain MRI after three months revealed resolution of the pachymeningeal enhancement and venous distension (Bern score 4). At 15 months following the procedure, she experienced recurrence of new daily headaches akin to the ones prior to treatment. A repeat brain MRI at 15 months did not show clear new worsening of SIH features compared to an MRI brain at 1 year post-procedure (Bern score 2). The patient's symptoms however further worsened and follow-up brain MRI 19 months after procedure showed worsening SIH features including recurrent pachymeningeal enhancement, brain sagging, and venous distention (Bern score 8). A lateral

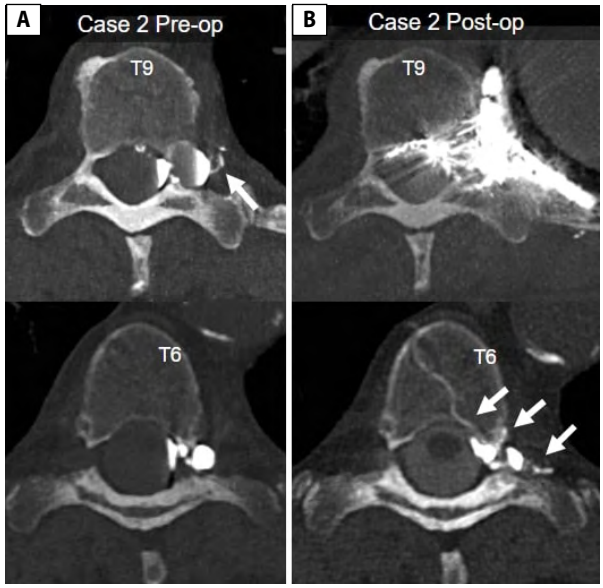

Figure 2. Patient 2. **A.** Pre-embolisation of left T9 CVF: Lateral decubitus dynamic CTM showing left T9 CVF (arrow) with no evidence of CVF at T6 level. **B.** Post-embolisation of left T9 CVF: lateral decubitus dynamic CTM showing new CVF at left T6 level with a connection to basivertebral vein (arrows). Embolisation cast at left T9 also noticed

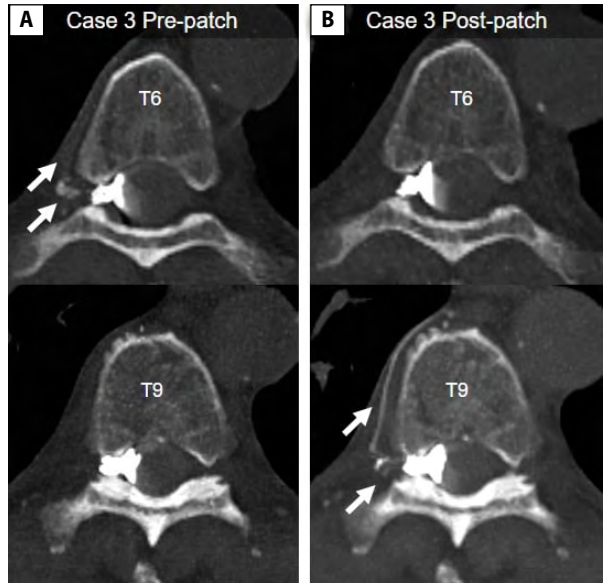

Figure 3. Patient 3. **A.** Pre-patching of right T6 CVF: lateral decubitus dynamic CTM showing CVF at right T6 level before treatment (arrows). **B.** Post-patching of right T6 CVF: lateral decubitus dynamic CTM showing a new CVF at right T9 level (arrows), with no evidence of previously treated CVF at T6 level

decubitus dynamic CTM clearly identified a new CVF at left T6 level with no residual leaking at the previously embolised level at left T9 (Fig. 2). Embolisation of this new CVF was performed, and the patient reported significant improvement of her leak-related headaches, which was sustained at 3-month follow-up. MRI brain at 3-months post-second treatment showed marked improvement with only mild residual reduced mamillopontine and prepontine distances (Bern score 2).

Patient 3

A 78-year-old male presented with new persistent daily bitemporal headaches, most prominent when turning his head briskly from side to side. Brain MRI was significant for diffuse, smooth pachymeningeal thickening/enhancement and right venous sinus distension (Bern score 4). Multiple blood patches yielded only a transient response, and MRI findings were unchanged from the previous exam (Bern score 4). A conventional myelogram performed at another hospital demonstrated potential leak sites at ventral T10-11, and left T3-4. Targeted fibrin glue/blood patching resulted in resolution of the headache, and a follow-up brain MRI showed decreased pachymeningeal thickening (Bern score 4). About three years later, the patient started experiencing similar symptoms. Brain MRI showed worsened, mild, and diffuse dural thickening/enhancement and right venous sinus distension (Bern score 4). Right lateral decubitus dynamic CTM at our institution showed a new right T6 CVF (Figure 3). Two sequential targeted CT-guided epidural blood and fibrin glue patches were performed at right T6. Each however yielded only a transient benefit for 2–3 weeks.

Repeat brain MRI 1.5 months after the second patch showed persistent dural thickening/enhancement and venous distension (Bern score 4). Repeat right lateral decubitus dynamic CTM showed a new CVF arising from the right T9 level (Fig. 3). Notably, the previous right T6 CVF was no longer visualised, indicating successful treatment. The patient responded well to CT-guided right T9 transforaminal epidural fibrin glue patch for only one month. Brain MRI one month post-treatment showed stable smooth dural thickening/enhancement in the supratentorial region with no interval change from the previous study (Bern score 4). According to patient preference, additional fibrin patching has been scheduled.

Patient 4

A 50-year-old female presented with a 3-year history of progressive pressure-like orthostatic headaches associated with neck pain, nausea, vomiting, blurred vision, dizziness, disorientation, muffled hearing, tinnitus and imbalance. Brain MRI showed typical stigmata of SIH: i.e. severe brain sagging, pachymeningeal enhancement/thickening, venous sinus distension, effaced suprasellar cistern, decreased mamillopontine distance, and effaced prepontine cistern (Bern score 8). Lateral decubitus CTM showed CVF at the right T7 level (Fig. 4) which was treated with venous embolisation.This resulted in complete resolution of CSF leak-induced symptoms. Brain MRI showed improvement of pachymeningeal thickening and normal mamillopontine distance (Bern score 5). After four months, she started having orthostatic headaches again, associated with nausea and vomiting. Brain MRI showed

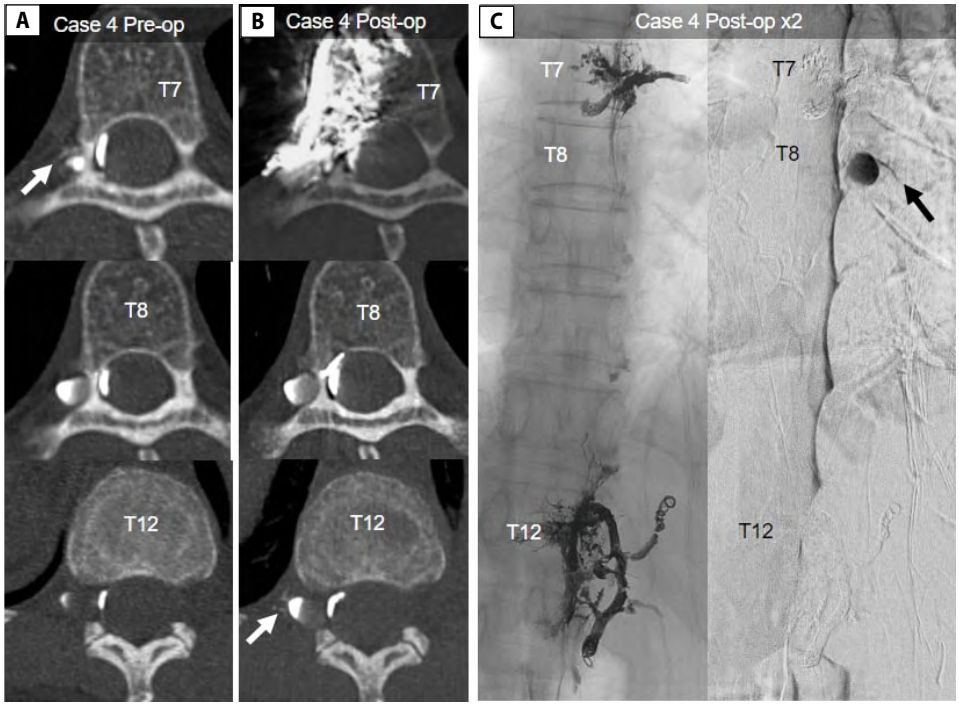

Figure 4. Patient 4. **A.** Pre-embolisation of right T7 CVF: lateral decubitus dynamic CTM showing CVF at right T7 level before treatment (arrow) with no evidence of CVF at right T8 or right T12. **B.** Post-embolisation of right T7 CVF: lateral decubitus dynamic CTM showing a new CVF at right T12 level (arrow). Embolisation cast at right T7 is also noticed. **C.** Post-embolisation of right T7 and T12 CVFs: lateral decubitus DSM showing new CVF at right T8 level (arrow) with embolisation casts seen at right T7 and right T12 levels

recurrence of a reduced mamillopontine distance compared to the previous scan (Bern score 6). Lateral decubitus CTM visualised clear CVF arising at the right T12 level with no residual leaking at the previously embolised level at right T7 (Fig. 4). Embolisation of the new CVF did not result in alleviation of her symptoms, follow-up brain MRI showed normal mamillopontine distance with persistent venous distenion (Bern score 5). A trial of CT-guided right T7 and T12 fibrin glue patch was performed, to which her symptoms responded well, but for only a couple of weeks. Subsequent brain MRI showed persistent venous sinus distension again (Bern score 5). Another lateral decubitus DSM clearly visualised CVF arising at right T8 level (Fig. 4), for which she underwent a CT-guided right T8 fibrin glue/blood patch. No leaking was seen on the previously embolised levels of right T7 and right T12. The symptoms improved for only one week. Six days after the patch, pituitary engorgement and venous distension on brain MRI were persistent with only subtle improvements (Bern score 5).

Discussion

We present four patients with recurrent CSF-venous fistulas, at separate levels from the initially treated fistula, after the effective treatment of the initial fistula by transvenous embolisation or fibrin glue/blood patching. Treatment success of the initial fistula was proven by the absence of the initial CVF upon repeated lateral decubitus myelography. These patients experienced either persistent or recurrent symptoms either shortly after treatment or months or even years afterwards, which prompted a repeat brain MRI followed by myelography. On the initial post-treatment MRI scans, the previously observed abnormal findings were unchanged or improved. We observed that the signs which first responded to treatment were pachymeningeal enhancement/thickening, venous sinus distension, and low mamillopontine distance. Importantly, these signs were also noted to deteriorate or reappear on MRI scans when patients began to manifest recurrent symptoms. In two of the patients, the Bern score exhibited an improvement after successful treatment, followed by a deterioration upon the exacerbation or recurrence of symptoms. This observation could open the doors towards investigating the reliability of the Bern score in identifying recurrent CSF leaks, specifically those related to recurrent CVF, and in guiding subsequent diagnostic steps such as repeat myelography.

In other words, a re-increase in Bern score after initial treatment, or a persistently high Bern score, may prompt consideration for repeat myelography evaluating for the possibility of a new CVF.

Malinzak et al. first documented a series of four cases involving the emergence of new CVFs after the surgical ligation of initial fistulas (Tab. 2). Their findings were similar to ours. The new CVFs were detected at distinct levels from the original ones.

Brain MRI demonstrated either a deterioration or lack of change of abnormal findings when symptom recurrence was reported by the patients. Additionally, during the transient resolution of symptoms following surgical ligation, resolution of pachymeningeal enhancement was documented in two of their patients. They suggested the presence of multilevel fistulous connections between CSF and veins in particular types of CVFs. As a result, the ligation of one connection could potentially prompt other connections to compensate for drainage [5]. Another hypothesis is the plausibility of baseline intracranial hypertension originating from underlying disorders such as Chiari malformation or obstructive sleep apnoea (OSA). This, in turn, might potentiate the development of decompressive CSF leaks. Herein, whenever a leak site is subjected to embolisation, ligation, or patching, a new site of leakage may arise in compensation.

These insights underline the importance of evaluating patients with recurrent leaks for conditions like underlying OSA and other disorders that could potentially increase intracranial pressure.

Clinical implications/future directions

This study demonstrates that new CVFs may underlie patients' reports of persistent or recurrent CSF leak symptoms after initial CVF treatment, particularly when their brain MRI scans show persistent or worsening abnormalities. Repeat myelography may be required in some patients, despite the successful treatment of initial CVFs.

Article information

Data availability statement: *The authors state that original contributions presented in the study are included in the article (and as Supplementary Material, if applicable) and that further inquiries can be directed to the corresponding author.*

Ethics statement: *IRB approval was obtained. Mayo Clinic Institutional Reviewer, IRB#: 21-005449.*

Authors' contributions: *RZ: design of study, data collection, data interpretation and analysis, drafting, revision and approval of final manuscript; OPF: design of study, data collection, data interpretation and analysis, drafting, revision and approval of final manuscript; TJH: design of study, data collection, data interpretation and analysis, drafting, revision and approval of final manuscript.*

Funding: *None*

Conflicts of interest: *The authors declare there is no conflict of interest.*

References

1. Roytman M, Salama G, Robbins MS, et al. CSF-venous fistula. Curr Pain Headache Rep. 2021; 25(1): 5, doi: 10.1007/s11916-020-00921-4, indexed in Pubmed: 33475890.
2. Schievink WI, Maya M, Prasad RS, et al. Spontaneous spinal cerebrospinal fluid-venous fistulas in patients with orthostatic headaches and normal conventional brain and spine imaging. Headache. 2021; 61(2): 387–391, doi: 10.1111/head.14048, indexed in Pubmed: 33484155.
3. Sobczyk P, Bojarski P, Sobstyl M. Surgical management of spontaneous intracranial hypotension syndrome: a literature review. Neurol Neurochir Pol. 2023; 57(2): 151–159, doi: 10.5603/PJNNS.a2022.0076, indexed in Pubmed: 36511484.
4. Dobrocky T, Grunder L, Breiding PS, et al. Assessing spinal cerebrospinal fluid leaks in spontaneous intracranial hypotension with a scoring system based on brain magnetic resonance imaging findings. JAMA Neurol. 2019; 76(5): 580–587, doi: 10.1001/jamaneurol.2018.4921, indexed in Pubmed: 30776059.
5. Malinzak MD, Kranz PG, Gray L, et al. Postsurgical Recurrence of CSF-Venous Fistulas in Spontaneous Intracranial Hypotension. Neurol Clin Pract. 2021; 11(3): e356–e358, doi: 10.1212/CPJ.0000000000001061, indexed in Pubmed: 34484913.