Surgical Treatment Efficacy of CSF-Venous Fistulas: Systematic Review

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Key words
- CSF-fistulae
- CSF–venous fistulae
- Hypotension
- SIH

Abbreviations and Acronyms
CSF: Cerebrospinal fluid
EBP: Epidural blood patch
MRI: Magnetic resonance imaging
SIH: Spontaneous intracranial hypotension

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BACKGROUND: **Cerebrospinal fluid (CSF)-venous fistula presents a pathologic connection between spinal subarachnoid space and adjacent epidural vein or veins. It is one of the 3 main causes of spontaneous intracranial hypotension along with dural defects and meningeal diverticulum. We performed a systematic review of the literature and analyzed individual participants' data focusing on clinical outcomes after different treatment modalities of CSF-venous fistula.**

METHODS: **Systematic review was conducted according to PRISMA recommendations. Literature search was performed in PubMed and Web of Science databases with following key phrases: "CSF-venous fistula", "Spontaneous intracranial hypotension". Overall, 97 articles were found during the initial search; 15 were included for the final analysis, with a total number of 137 patients.**

RESULTS: **Epidural blood patch (EBP) was performed as a first-line treatment in 37.1% of patients in individual data group, often not combined with fibrin glue (61.5%). Either partial (69.2%) or no resolution (30.8%) of symptoms was achieved after EBP injection. Nerve root ligation was the most common method of exclusion of CSF-venous fistula. Complete resolution of symptoms was achieved in 69.0% of patients, in 21.4% it was partial and in 9.5% no regress was found. Endovascular treatment was described only in 1 study.**

CONCLUSIONS: **Surgical ligation of fistula is a treatment of choice. In approximately 70% of patients complete long-term resolution of symptoms is achieved after surgery. Endovascular treatment and fibrin glue injections are prospective and evolving options, which require further investigation.**

INTRODUCTION

Cerebrospinal fluid (CSF)-venous fistula is a relatively new and undiscovered etiology of spontaneous intracranial hypotension (SIH). It was first described in 1974 in a patient with chronic post-lumbar puncture headache who underwent digital subtraction myelography.[1] Forty years later, CSF-venous fistula was proposed as a reason of SIH in a patient with orthostatic headaches.[2] CSF-venous fistula presents a pathological connection between spinal subarachnoid space and adjacent epidural vein or veins. It is one of the 3 main causes of SIH, along with dural defects and meningeal diverticulum. Although the prevalence of CSF-venous fistula is not well evaluated some authors think that it is the reason for SIH in 25% of the cases.[3]

Positional headaches and those associated with cough and Valsalva maneuver are common symptoms in patients with CSF-venous fistula. These patients most often have typical features of SIH on brain magnetic resonance imaging (MRI), including pachymeningeal enhancement, venous engorgement, pituitary enlargement, brain sagging, tonsillar herniation, and so forth. Because of the low incidence of the disease, however, algorithms of diagnosis and treatment are not established. The patients are usually managed conservatively as a first-line treatment. Epidural blood patches (EBPs) are commonly used either alone or combined with fibrin glue.

Open surgery with fistula ligation or coagulation is indicated in case of unsuccessful conservative treatment. Endovascular occlusion of draining vein is a relatively new and less common option.

We performed a systematic review of the literature and analyzed individual participants' data focusing on clinical outcomes after different treatment modalities of CSF-venous fistula (patches, surgical excision, and endovascular embolization).

METHODS

Systematic review was conducted according to PRISMA recommendations.[4] Literature search was performed in the PubMed and Web of Science databases with the following key phrases: "CSF-venous fistula", "Spontaneous intracranial hypotension".

The following algorithms for search in PubMed were used:

1) csf-venous[All Fields] AND ("fistula" [MeSH Terms] OR "fistula"[All Fields])

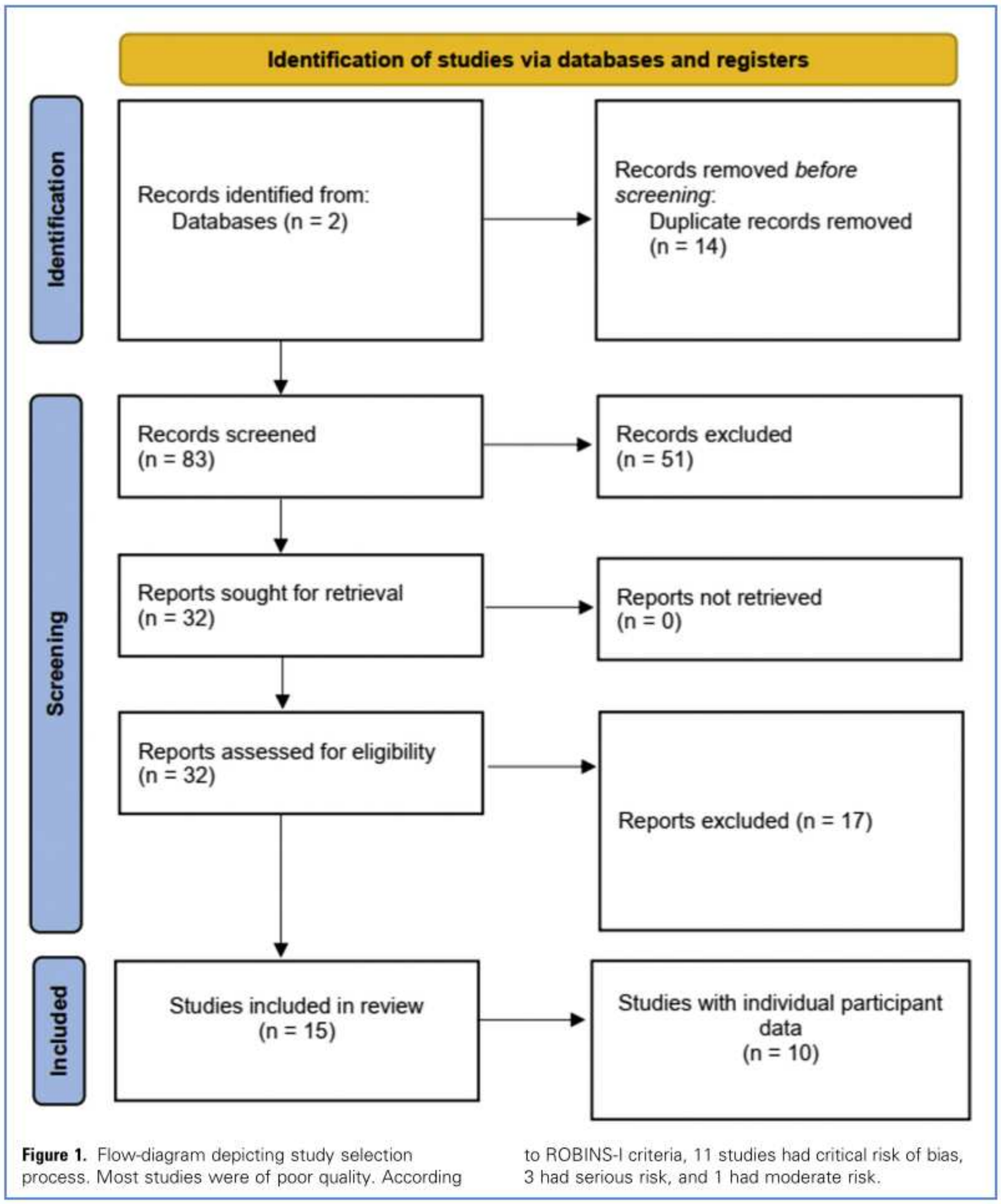

Figure 1. Flow-diagram depicting study selection process. Most studies were of poor quality. According to ROBINS-I criteria, 11 studies had critical risk of bias, 3 had serious risk, and 1 had moderate risk.

2) "intracranial hypotension"[MeSH Terms] OR ("intracranial"[All Fields] AND "hypotension"[All Fields]) OR "intracranial hypotension"[All Fields] OR ("spontaneous"[All Fields] AND "intracranial"[All Fields] AND "hypotension"[All Fields]) OR "spontaneous intracranial hypotension"[All Fields] AND csf-venous[All Fields]

We assumed the following inclusion criteria for studies: 1) published in English; 2) full access to the article is available; 3) information on treatment modality and outcomes are provided at least for 1 patient with CSF-venous fistula.

The following data were extracted from the studies: SIH symptoms and signs, duration of symptoms, brain sagging presence, treatment modality, number of EBPs, number of target EBP and fibrin glue injections, anatomical features of fistulas, resolution of symptoms, complications rate, rebound headache rate and duration, recurrence rate.

The ROBINS-I platform was applied to assess a quality of the studies.[5]

Statistical analysis was performed in jamovi, version 1.6.[6] The χ^2 test of association was used to compare clinical outcomes after different treatment modalities and to assess an influence of anatomical features and duration of symptoms on outcomes.

RESULTS

A total of 97 articles were found during the initial search; 15 were included for the final analysis,[2,3,7-19] with a total number of 137 patients. A flow diagram depicting the

Table 1. Symptoms of Patients With Available Individual Data

Symptoms	Rate
Headache (100%)	
Orthostatic	23/35 (65.7%)
Associated with cough or Valsalva maneuver	7/35 (20.0%)
Headache of second-half-of-the-day	1/35 (2.9%)
Nonspecified	4/35 (11.4%)
Dizziness	4/35 (11.4%)
Tinnitus	5/35 (14.3%)
Neck pain or stiffness	7/35 (20%)
Nausea/vomiting	3/35 (8.5%)
Disbalance	2/35 (5.7%)
Back pain	3/35 (8.5%)
Pain in lower/upper extremities	1/35 (2.9%)
Others	5/35 (14.3%)

study selection process is shown in Figure 1.

Individual participant data were available in 10 studies for a total number of 35 patients. Mean age was 55.4 years (range: 26–91 years), most of the patients were female (57.1%). Mean duration of symptoms before treatment was 63.9 months (range: 1–216 months).

Symptoms of patients with available individual data are shown in Table 1.

Headache was the dominant symptom in the initial sample of 135 patients (98.5%). In 23 patients (65.7%) with available individual data, headaches had orthostatic features. Their intensity was assessed by means of the HIT-6 scale in 2 studies[9,14] and by the MiDAS scale in 1 study.[11]

On brain MRI typical SIH features were found in almost all patients. Brain sagging was a dominant radiologic sign and was found in 88.6% of patients with available individual data. Different myelographic approaches were applied to diagnose and verify the location of CSF-venous fistulas: MR myelography, CT myelography, digital subtraction myelography, or selective spinal venography.

According to the included studies, CSF-venous fistulas localized commonly in lower segments of the thoracic spine. They were found at the T6 level or lower in 73.3% of the cases.

Treatment Outcomes

Data on treatment outcomes from different studies are shown in Table 2.

EBP was performed as a first-line treatment in 37.1% of patients in individual data group, often not combined with fibrin glue (61.5%). Either partial (69.2%) or no resolution (30.8%) of symptoms was achieved after EBP injection. In a group with partial resolution, EBP was combined with fibrin glue in more than a half of patients (55.6%). This combination was not used in any of the patients without resolution of symptoms. The difference between the 2 groups with and without fibrin glue approached the statistical significance, but it was not reached due to small sample size ($\chi^2 = 3.61$, $P = 0.057$).

In 101 patients (71.1%) from the general sample, EBP was applied initially, 59.4% of these patients achieved combined treatment with fibrin glue. In the study of Duvall et al., combined treatment did not bring any additional effect.[10] Only in 4 patients (3.96%) full resolution of symptoms was achieved after EBPs. In 26.5% of patients there was no resolution and in 69,4% resolution was partial.

The mean number of EBPs in individual data group was 3.2 ± 2.6 (range: 1–9). Multiple EBPs were not associated with better symptom resolution ($\chi^2 = 0.563$, $P = 0.453$). In 18 patients (17.8%) from the general group EBPs were applied more than once. In the study of Duvall et al., multiple and multilevel EBPs were used in 9 patients (22.5%).[10] Different clinical outcomes were observed in this group, from no effect to long-term full symptom resolution.

Information about target and non-target procedures was available for 7 patients, which was insufficient for statistical analysis. In 57.1% of patients, target EBPs were performed.

No complications were registered in the entire group after EBP procedure.

Fibrin glue was applied initially in 19 patients (13.4%), including 1 patient after unsuccessful EBP treatment. Among 16 patients who had symptoms before the procedure, 12 achieved full resolution of symptoms and 4 achieved partial resolution. In 14 patients from the individual data group, fibrin glue was the only treatment modality. In all of these patients, full resolution of symptoms was achieved. Target fibrin glue injections were applied in 13 patients with assistance of CT-navigation in the study of Mamlouk et al.[16] In 4 patients, the features of residual fistula were not seen on initial control CT myelography, which required 1 additional injection in 3 cases and 2 injections in 1 case. One of these fistulas had the highest diameter. It drained into venous plexus on 2 levels (T10-T11, T11-T12). Rebound syndrome and hypertensive headaches were registered in 4 patients. They lasted less than a month and did not require any additional treatment.

In 1 of 2 patients, hydration was applied as a first-line treatment in the study of Madhavan et al.[12] Full resolution of symptoms was achieved. However, symptoms recurred within 5 years. The patient declined additional treatment and was lost to follow-up.

A total of 107 patients underwent surgery either initially or after unsuccessful conservative treatment. Surgery was not performed in the following patients: 15 patients with full resolution of symptoms after fibrin glue injection, 3 patients after EBP, 5 patients after endovascular treatment, 9 patients were waiting for operation, 2 patients declined surgery.

Nerve root ligation was the most common method of exclusion of CSF-venous fistula. It was often combined with coagulation or clipping of draining veins. Fistula drained frequently into one vein (66,1%). Draining vein rarely had features of venous plexus (29.0%). In 8 patients fistula drained into vessels of venous malformation. Data on clinical outcomes were available for 84 patients after surgery. Complete resolution of symptoms was achieved in 69.0% of patients, in 21.4% it was partial and in 9.5% no regress was found.

Surgery was also performed in 20 patients from the individual data group. There was no difference in clinical outcomes between the 2 groups, with symptom duration greater than and less than 12 months ($\chi^2 = 0.636$, $P = 0.425$). In cases where the fistula drained into venous malformation, clinical outcomes were significantly worse ($\chi^2 = 6.35$, $P = 0.042$). Although there was a strong correlation between clinical outcomes and brain sagging resolution ($\chi^2 = 14.9$, $P < 0.001$) in

Table 2. Studies Included in Analysis and Relevant Data

Study	Sample Size	EBP Performed in	Stereotactic Fibrin Glue Injection	Surgical CSF Ligation	Resolution of Symptoms
Schievink et al., 2014[2]	3	2/3	1/3	2/3	100%
Kumar et al., 2016[8]	3	3/3	0/3	3/3	100%
Kranz et al., 2016[7]	3	3/3	0/3	3/3	100%
Shievink et al., 2016[11]	10	0	4/10	9/10	90%
Kranz et al., 2017[9]	22	22	0	10	59%
Duvall et al., 2019[10]	44	40/44	0	42/44	47%
Shievink et al., 2019[20]	3	2/3	0	3/3	66%
Madhavan et al., 2020[21]	2	0	0	1/2	50%
Chazen et al., 2020[13]	3	3/3	0	3/3	100%
Wang et al., 2020[14]	20	20/20	0	20/20	90%
Ortega-Porcayo et al., 2020[15]	1	0	0	1/1	0%
Mamlouk et al., 2021[16]	13	0	13/13	0	100%
Malinzak et al., 2021[19]	4	0	0	4/4	25%
Brinjikji et al., 2021[17]	5	0	0	5/5 (endovascular embolization)	80%
Shievink et al., 2021[18]	6	6/6	1/6	6/6	83%
Total	142	101	0	107	64%

the group with individual data, in 6 patients with complete resolution of symptoms MRI features of SIH either remained the same, or partial resolution was registered.

Mean follow-up period was 10.4 months (range: 1.5–60 months). Recurrence of symptoms was noticed in 6 patients after surgery (6.0%). No residual fistula was found in 2 patients, in 1 of these treatment proceeded with additional EBPs. New CSF-venous fistulas were found in 4 patients: 1–2 levels higher or 1 level lower.

Data on postoperative complications are scarce in the evaluated studies. Rebound syndrome was observed in 18 patients after surgery (16.8%). They regressed commonly within a month without additional treatment. Acetazolamide was administered for 2–3 weeks in selected patients. Other complications included paresthesia/hypesthesia in upper (2 patients) or lower extremities (2 patients), thoracic back pain (1 patient), upper extremity weakness (1 patient), transient lower extremity paraparesis (1 patient), transient urinary retention (1 patient), and abducens deficiency (1 patient).

Endovascular treatment was described only in study.[17] Spinal venography with

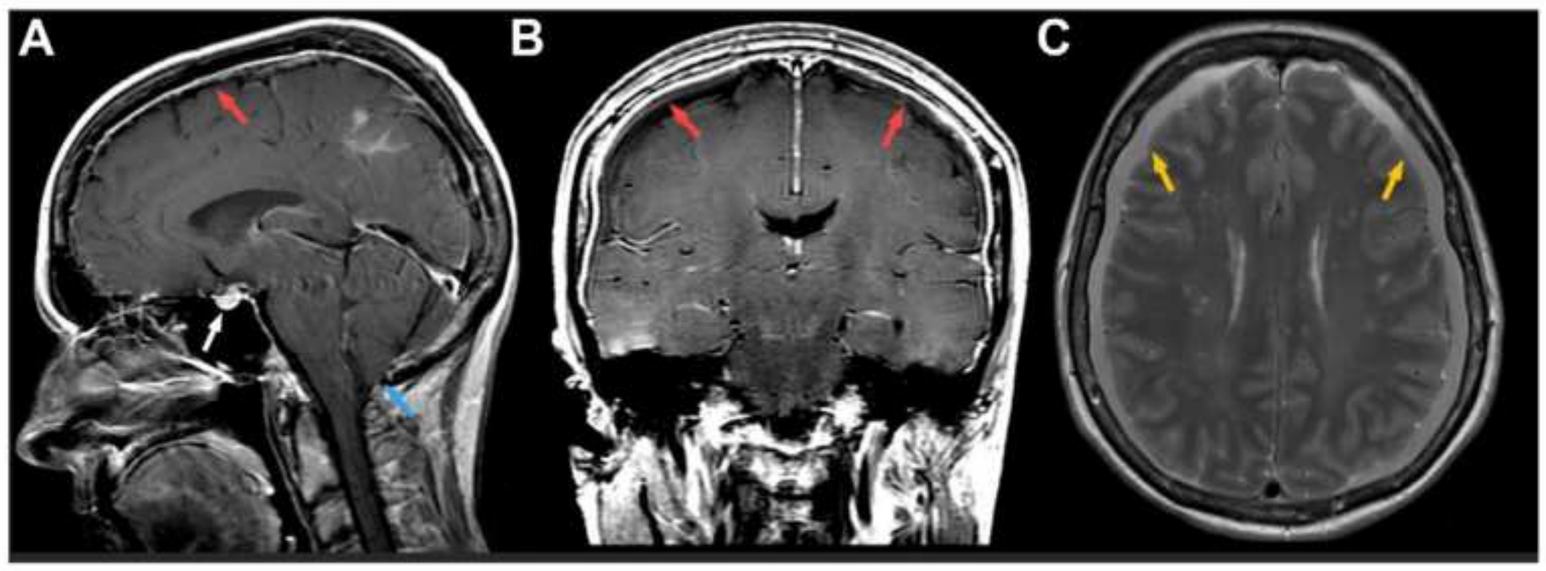

Figure 2. Brain MRI of a patient with classical signs of severe intracranial hypotension. (**A**) T1 + contrast, sagittal plane: pachymeningeal enhancement (*red arrow*), brain sagging (*blue arrow*), dislocation of cerebellar tonsils below foramen magnum with decreased volume of posterior fossae CSF cisterns. Enlargement of pituitary gland (*white arrow*). (**B**) T1 + contrast, frontal plane: pachymeningeal enhancement (*red arrow*) with bilateral subdural hygromas. (**C**) T2 axial plane: bilateral subdural hygromas (*yellow arrows*).

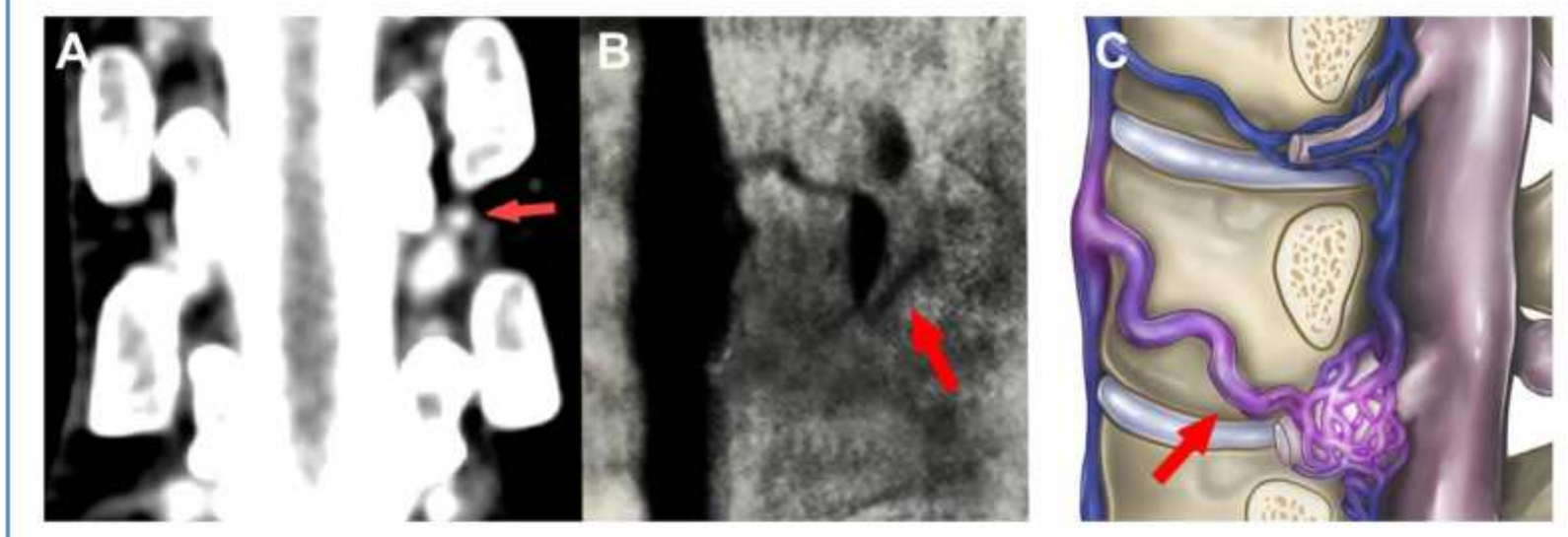

Figure 3. (**A**) CT myelography demonstrates contrast enhancement of radicular diverticula with paraspinal vein enhancement (*red arrow*). (**B**) Dynamic subtraction myelography shows contrast flow to the radicular diverticulum and associated paraspinal vein (*red arrow*). (**C**) Schematic presentation of venous CSF fistula.

selective Onyx embolization of draining veins was performed in 5 patients after azygos vein catheterization. No complications were registered. In 4 patients, complete resolution of symptoms was achieved (80%).

DISCUSSION

Herein we present systematic review of the literature focusing on treatment outcomes of CSF-venous fistulas. Two systematic reviews on SIH have been published.[22,23] Incidence of this disease is extremely low.

CSF-venous fistula causing SIH was first described in 2014 by Schievink et al.[2] Its pathophysiology is not well understood. Normally, CSF is drained at the dural recess of nerve roots by arachnoid granulations.[23] Fistulas occur as a rule at these dural recesses, where paraspinal veins are located.[24,25] Most CSF-venous fistulas are found in the thoracic spine, although cases at cervical and lumbar location are also reported.[26] In 80% of the cases, CSF-venous fistulas are associated with meningeal diverticula of spinal nerve roots.[9] Due to extension of a meningeal diverticulum, with time a pathologic connection between subarachnoid space and the paraspinal venous system can develop. In an upright position this can facilitate the flow of the CSF due to an increased gradient between CSF pressure and venous pressure. Increased CSF outflow leads to reduced intracranial pressure and symptoms of SIH.[27] Inflammation after trauma or intervention is also reported to be an etiologic factor, although further investigation is needed.[22]

Diagnosis and verification of CSF-venous fistula is complex and requires exclusion of other causes of SIH. MRI is highly sensitive in detecting of SIH features. However, up to 20% of patients with SIH can lack typical MRI signs.[22] In the study of Watanabe et al., SIH was suspected due to changes on brain MRI in 83% and on spinal MRI in 94% of the cases.[28] Pachymeningeal enhancement is the most common radiologic sign and is present on 75% of brain MRI scans (**Figure 2**). Other features include subdural blood or CSF collection (**Figure 2B** and **C**), brain sagging (**Figure 2A**), venous engorgement, compression of basal cisterns, decreased ventricular size, pituitary enlargement, flattening of chiasm and optic nerves, and anterior-posterior brainstem enlargement.

Typically, EBP is used as a first-line treatment in patients with SIH.[29] However, in cases of CSF-venous fistula the effect of the procedure is minimal and usually transient. Complete and persistent resolution of symptoms develops only in 4% of patients with CSF-venous fistulas, according to our analysis. Mechanisms underlying this effect is not well understood. Obstruction of pathologic CSF outflow is reported to be the main reason. It requires about 3 weeks for completion of chronic productive inflammation and fibrosis.[30] Occlusion of dural defect leads to immediate clinical improvement. As a rule, 10–20 mL of venous blood is injected during one procedure but 20–100 mL injection is also considered to be safe.[31] Clinical outcomes can be improved with fibrin glue injection, which was demonstrated by Mamlouk et al.[16] and increased efficacy up to 100%. However, such a high rate of complete resolution of symptoms with fibrin glue injection has not been demonstrated by other studies. (Shievink et al., 2016, 2021).[11,18]

EBP can be used not only as a therapeutic but also as a diagnostic tool in cases of CSF-venous fistula.[22] In cases of unsuccessful EBP, surgery should be considered.[31] Before surgery, the location of the pathology should be verified.

Dynamic subtraction myelography is a helpful diagnostic tool. It should be considered in patients with symptoms of SIH and large epidural spinal CSF collection on spinal MRI or CT myelogram (**Figure 3A**). This method has some limitations. First, results depend on movements of patients. They should be able to stop breathing comfortably in a lying position. General anesthesia is not necessary to achieve high-quality imaging. Duration of digital subtraction is 20–30 seconds, which is why CSF flow is too slow to be detected (**Figure 3B**).

Currently, surgical ligation of CSF-venous fistula is a standard of care.[22] However, endovascular procedures for these patients are evolving. This was demonstrated by Brinjikji et al. in a study in which transvenous Onyx embolization of draining veins was performed.[17] Unfortunately, reports on endovascular treatment of the disease are scarce in the literature. According to our analysis, the recurrence rate after treatment in all groups is approximately 6%, which is relatively low, considering that

experience with diagnosis and treatment is limited.

Study Limitations

Clinical application of the results of our analysis is limited due to the low quality of most studies and the small sample size. All included studies were retrospective, with high values of statistical and clinical heterogeneity, which is why a meta-analysis could not be conducted. Nevertheless, there is a rising interest in this problem, for which we lack knowledge about the underlying pathology, and have unsolved questions on diagnosis and treatment. These are prerequisites for analysis of existing experience described in literature.

CONCLUSIONS

CSF-venous fistula is occasionally encountered as a reason for SIH and requires timely diagnosis and treatment. Digital subtraction myelography is a helpful tool in diagnosis along with CT myelography and MR myelography. Surgical ligation of fistula is a treatment of choice. Approximately 70% of patients report that long-term resolution of symptoms is achieved after surgery. Endovascular treatment and fibrin glue injections are prospective and evolving options, which require further investigation.

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