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A case report and narrative review of literature of spontaneous intracranial hypotension during the postpartum period

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Abstract

Introduction: **Spontaneous intracranial hypotension (SIH) is defined as the occurrence of orthostatic headache resulting from spontaneous cerebrospinal fluid (CSF) leak. Orthostatic headaches are not always related to SIH. When symptoms declare in the postpartum period after neuraxial anesthesia, postdural puncture headache (PDPH) is more often suspected. We present here the first case of SIH occurring 3 months after vaginal delivery with epidural anesthesia.**
Methods: **For the writing of our case, we conducted a Pubmed search of SIH in association with the postpartum period. We then proceeded to make a narrative review of literature of SIH using the same search engine.**
Results: **SIH has an annual incidence of 5/100 000 and middle-aged women are the most affected. Clinical presentation consists of orthostatic headache in more than 90% of patients. Meningeal irritation and cochlear-vestibular symptoms are other frequently reported signs. Known risk factors are connective tissue disease and trivial trauma. There are no clear evidences regarding the appropriate imaging and treatment modalities in SIH. Common practice suggest the realization of brain MRI with diffuse pachymeningeal enhancement being the most pathognomonic sign. Treatment usually consists of bed rest for a limited period of time followed by one or more blind lumbar epidural blood patches (EBP) if necessary. Other treatment options include targeted EBP and surgery.**
Discussion and conclusion: **SIH is often underdiagnosed and misdiagnosed. Anesthetists in particular are not familiar with the pathology. Only two cases of postpartum SIH are reported in literature. This review highlights SIH as a possible cause of postpartum headache and the need for healthcare providers to recognize the syndrome.**

Keywords: **Spontaneous intracranial hypotension, orthostatic headache, postpartum headache.**

Introduction

Spontaneous intracranial hypotension (SIH) is a clinical condition characterized by postural headaches resulting from cerebrospinal fluid (CSF) leak. The leakage is by definition spontaneous which excludes iatrogenic causes like history of a dural puncture[1].

The syndrome is not uncommon and has been widely reported over the last few years, as shown by the growing number of publications. However, there is still lack of knowledge of the pathology which can lead to misdiagnosis and underdiagnosis. A recent survey of 227 healthcare professionals in the UK conducted in 2021 showed that only half on anesthesiologists had been involved in the care of the pathology[2].

Orthostatic headaches are not always related to SIH or an intracranial hypotension syndrome. In fact, during pregnancy and the postpartum period which can extend up to 6 months after delivery, SIH is considered very rare. When symptoms occur within the first days following an epidural or spinal procedure, post-dural puncture headaches (PDPH) are more often suspected.

Diagnosis of SIH during the postpartum period may therefore be challenging especially when

The patient has given written consent for publication.
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epidural anesthesia was previously performed. Here we present the first case of spontaneous intracranial hypotension occurring three months after delivery with epidural anesthesia.

For better understanding we proceeded to make a narrative review of literature of SIH with focus on its implications during the peripartum period.

Methods

For the writing of our case, we first conducted a search in literature of case reports describing spontaneous intracranial hypotension in the postpartum period. We used the keywords spontaneous intracranial hypotension and postpartum in the Pubmed search engine. We obtained written patient consent and used CARE guidelines to report our case.

We then conducted a narrative review of literature. Following keywords were used in the same search engine: spontaneous intracranial hypotension in association with postpartum period", pregnancy", delivery", labor", epidural anesthesia", spinal anesthesia", etiology", definition", symptoms", lactation", diagnosis", differential diagnosis", complications and treatment". We also searched for postpartum headache and post dural puncture headache to make evidence-based discussion and conclusions. Preferences were made for systematic reviews of literature and meta-analysis.

All articles were selected based on if they were written in English and relevance to our case. Researches were made in May 2021 and extended in December 2022 without any limitation regarding publication date.

Spontaneous intracranial hypotension 3 months after vaginal delivery with epidural anesthesia: a case report

Introduction

SIH results from a CSF leakage and usually consists of orthostatic headaches presenting with no history of dural puncture3. Diagnosis during the postpartum period may be challenging especially when epidural anesthesia was previously performed.

Narrative

We relate the case of a 31-year-old woman who presented to the emergency room with headache. She had given birth 3 months earlier under epidural anesthesia without complication, a simple uterine revision for placental remnants having been performed in the hours following delivery.

The headaches started the day before her admission and gradually increased. They were described as holocranial, predominantly frontal, bilateral and pulsatile. They also had a major positional component, disappearing completely in the supine position and increasing when moving to the sitting and standing positions. She also complained of nausea and had experienced episodes of vomiting while standing. There was no recent trauma to report. She was breastfeeding, but reported a gradual decrease in lactation over the last few days.

A complete neurological examination performed in the emergency room was reassuring. Routine blood tests showed no abnormalities. The patient was not pyretic and had no nuchal stiffness. A CT-scan was performed to exclude any intracranial process requiring urgent management, which was unremarkable. An initial diagnosis of SIH of undetermined origin was made and weighed against a possible diagnosis of post-dural puncture headache (PDPH), although the latter seemed unlikely given the 3 month delay post epidural anesthesia. The patient was discharged with conservative treatment including level-1 analgesics in combination with caffeine. She was also advised to consult the anesthesiology department so that an epidural blood patch (EBP) could be performed if the symptoms did not improve within a few days. Due to increased orthostatic headaches and vomiting, the patient was referred to the anesthetists for an EBP the following day. Given the probable hypothesis of SIH, a blind EBP was performed at the lumbar L4-L5 level under radioscopy and 20mL of autologous blood was injected into the epidural space after location and injection of contrast agent. The patient remained in the prone position for 2 hours after the injection and a gradual return to the upright position was achieved. The symptoms were rapidly ameliorated during the following hours. Unfortunately, this improvement was only temporary and orthostatic headache and nausea recurred over the next few days, although to a lesser extent. The patient also complained of increased nuchal pain. MRI imaging of the spinal cord was performed 5 days after the first admission to the emergency room. It revealed a spinal cord with a homogeneous signal and normal morphology. A collection of liquid signal was demonstrated in the epidural space, predominantly in the thoracic region and especially posteriorly, with a tendency for the dural sac to collapse and present irregular contours, especially in the lower thoracic region (Figure 1). She was admitted a week later, 15 days after initially presenting to the emergency room, for a second EBP at the L3-L4 lumbar level. She did not report headache after that.

She gave birth to a second child two years later, also under spontaneous vaginal delivery but no epidural anesthesia was performed this time. There

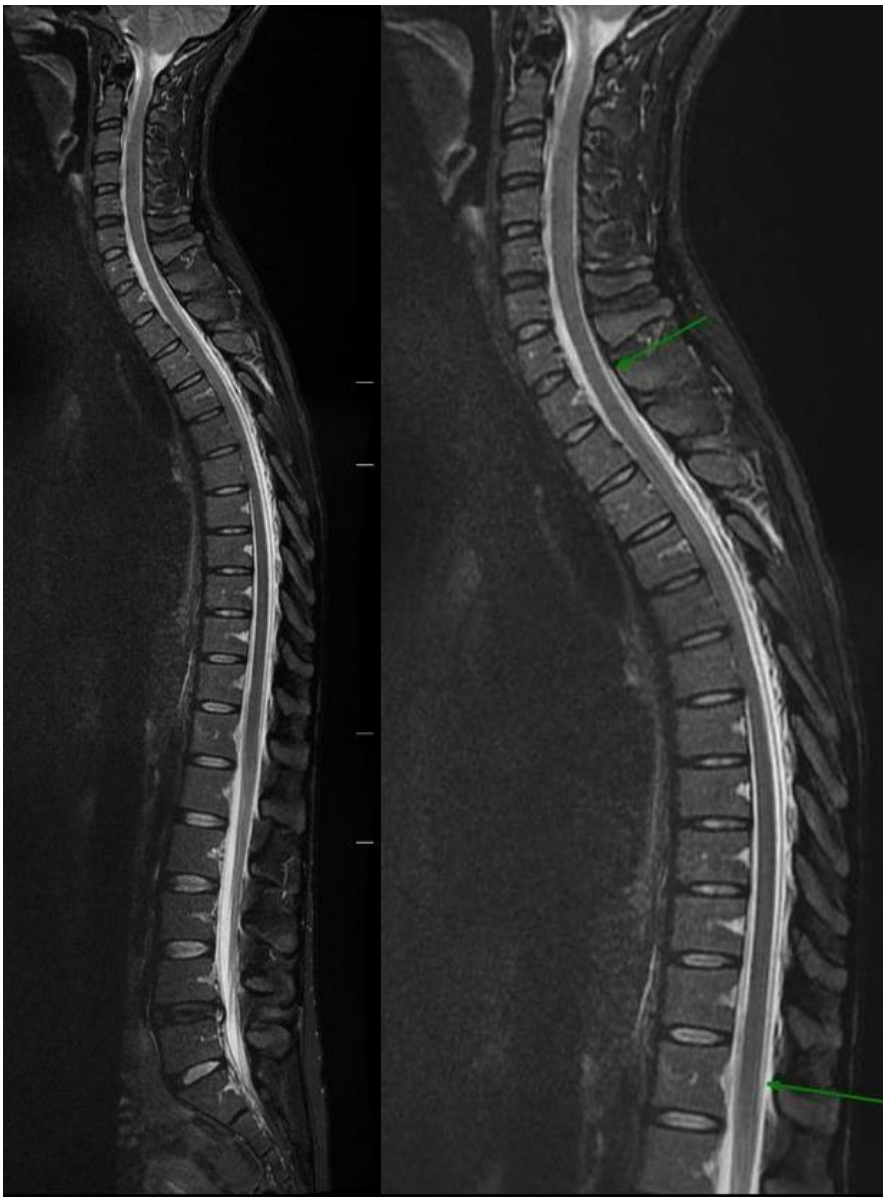

Fig. 1 — Spine MRI images performed 5 days after patient's first admission to the emergency department. Sagittal STIR (Short Tau Inversion Recovery) sequence of full spine MRI showing a collection of liquid signal in the epidural space (arrow), predominantly in the thoracic region and especially posteriorly, with a tendency for the dural sac to collapse and present irregular contours, especially in the lower thoracic region. Evidence of an extrathecal CSF leak (SLEC) without identification of the leakage site.

was no complication or headache to be reported during pregnancy and at 6-months follow-up after delivery.

Discussion

This is to our knowledge the first case of SIH occurring 3 months after delivery with epidural anesthesia. Few cases of SIH in literature have been reported during pregnancy and in the postpartum period but with no history of epidural anesthesia[1-4].

Headaches during the postpartum period are frequent. During the first week and in the presence of a postural component associated with a history of epidural anesthesia they are often attributed to PDPH. Other diagnosis should be considered after. Although symptoms resulting from CSF volume depletion are similar in PDPH and SIH, etiology, site of leakage, diagnosis and treatment differs. Ineffective conservative treatment and need for more than one EBP were in favor of SIH in our case. Moreover, the great height and relative hyperlaxity of the patient suggesting a potential connective tissue disorder of Ehlers Danlos type, more frequently associated with a SIH, should be highlighted here, as well as the fact that the patient had to stop breastfeeding due to a drastic drop in lactation a few days before the onset of symptoms, evoking a potential alteration of prolactin secretion also associated with SIH[4]. Furthermore, while not directly related to SIH, epidural anesthesia

in combination with hormonal changes of the pregnancy might have caused a dural weakness that did not spontaneously resolve because of an underlying connective tissue disorder[5].

Because persistent CSF leakage might lead to serious complications like cerebral venous sinus thrombosis (CVST) and subdural hematoma, early recognition and treatment are important. This is particularly true in SIH as the leaks can recur, be multi-level and EBP be less effective. Evidence of SIH should lead to investigation for a connective tissue disorder and a cautious follow-up.

Results

Definition and epidemiology

SIH also referred to as Aliquorrhea - meaning no CSF - was described for the first time by the neurologist Schaltenbrand in 1938. However, there is mention of Low-pressure CSF headache as soon as 1898 after the administration of spinal anesthetic via lumbar puncture by the anesthesiologist Bier[6].

International Classification of Headache Disorders, 3rd edition (ICHD-3, 2018) provides definition and diagnostic criteria for low-pressure CSF headache related to SIH[7]. It can be simplified as orthostatic headaches developing in temporal relation with objective data of low CSF pressure (< 6 cmH2O) or evidence of CSF leaking on imagery or leading to their discovery. The absence of a procedure known to cause CSF leakage and no evidence of a better diagnosis are also required[3].

SIH is considered a frequent and increasing cause of postural headache with an annual incidence ranging from 1 case per 50 000 in the 1990's to 5 cases per 100 000 nowadays and there may be even more. This is attributed for the most part to under- and misdiagnosis of CSF leakage especially when there is no history of spinal tap[8].

Middle-aged women are the most affected at a rate of 1.5 to 2:1 compared to men. Peak incidence is around the 4th decade of life but there have been case reports of patients from 2 to 88 years old[9].

Physiopathology, symtoms and complications

Anatomical and physiological changes occurring in SIH can be explained using the Monroe-Kellie doctrine which stipulates that the intracranial space contains a constant volume of blood, brain and CSF and that a change in volume of one or two components induces a reciprocal alteration in volume of the others. Therefore, compensatory venous dilatation associated with CSF volume depletion is a common finding.

On the other hand, brain volume is usually thought as invariable or increased due to brain swelling as the consequence of venous stagnation. A recent study comparing brain MRI changes before and after successful treatment found conflicting results illustrated by a decreased total brain volume when symptoms were presents. The author proposed reduced free water redistribution in the brain parenchyma as an explanation[10].

Clinical presentation consists of a wide variety of symptoms, ranging from minor to debilitating. They are listed from most to least common[9]:

- Orthostatic headache: a headache that worsens when standing upright, throughout the course of the day or after Valsalva maneuvers and usually resolves within 30 minutes in the recumbent position. They can be diffuse or more commonly located in the frontal or occipital area but rarely unilateral. The mechanism involved is loss of CSF buoyancy causing traction on the dura with the posterior fossa being highly sensitive and compensatory venous dilatation in the pain-sensitive structures. They are reported in more than 90% of cases and considered the initial and classical manifestation of SIH[11] ;
- Signs of meningeal irritation: nausea and vomiting (50%), posterior neck pain and stiffness (40%) and photo- or phonophobia (5%) ;
- Cochlear-vestibular signs: tinnitus (20%), dizziness (15%) and hearing changes (10%) that can be best explained by transmission of low CSF pressure to the perilymph resulting in a compensatory expansion of the endolymphatic compartment. Stretching of the cranial nerves is an alternative theory[12] ;
- Visual signs: diplopia (5%), visual blurring or superior binasal visual field cut are also the result of stretching of the cranial nerves.

All manifestations cited above can present a postural component. Other clinical presentations are rather considered rare (< 5%) or described in case reports:

- Other signs of cranial nerve involvement: facial numbness, pain or weakness, paresthesia and dysgeusia ;
- Other types of headache: including thunderclap headache or paradoxical headache exacerbated when lying down[13] ;
- Back pain: sometimes localized at the site of the leak but generally not[14] ;
- Radicular upper limb symptoms ;
- Hyperprolactinemia and galactorrhea as result of downward traction on the pituitary stalk[15].

Average onset of symptoms before diagnosis is 30 days but has been described as long as 20 years[9]. With chronicity, postural component might become less obvious and headache continuous. Additional symptoms could arise:

- Anxiety and depression ;
- Central diabetes insipidus[16] ;
- Parkinsonism and ataxia ;
- Frontotemporal dementia: clinical entity characterized by apathy, a disinhibited behavior and reduced MMSE scores that can be reversible in up to 70% of cases after treatment of SIH[17] ;
- Other cognitive impairments ;
- Leptomeningeal hemosiderosis[13] ;
- Epilepsy.

If left untreated for a long period of time and/or in dramatic CSF volume depletion as observed in important CSF leaks caused by surgery, SIH can lead to serious complications. Severe brain sagging and squeezing, especially in the brain stem, can cause diencephalic herniation and formation of subdural hygromas as well as rupture of bridging veins. Symptoms may present as follows[18]:

- Decreased level of consciousness and coma ;
- Subdural hematomas ;
- Cortical vein and sinus thrombosis[13] ;
- Cerebral vasoconstriction syndrome ;
- Cerebellar hemorrhage.

Etiology

Spontaneous CSF leakage occurs through the dura mater and is typically located at the nerve root of the cervicothoracic junction or the thoracic spine. Subsequent anatomical changes leading to symptoms of intracranial hypotension are now thought to be the consequence of CSF volume depletion rather than a fall in CSF pressure as previously suggested. The underlying mechanism is still not completely understood and some authors even state that the dural leak may not be the cause but the effect of epidural hypotension[13].

Known factors involved in the development of dural defect are structural and can be associated with a minor traumatic event. Etiology may for the most part be multifactorial and remains widely unknown.

Structural abnormalities generating dural weakness are hereditary, congenital or acquired. This includes connective tissue disorders, bone spurs, degenerative disc herniation, and meningeal diverticular or even complete absence of dura around a nerve root[12]. Bariatric surgery or malnutrition have also been cited as possible risk factors[16].

Documented connective tissue disorders such as Marfan syndrome, Ehlers-Danlos Syndrome, autosomal dominant polycystic kidney disease are reported in 5% of cases (less frequently : familial osteosclerosis, neurofibromatosis). Evidence of an underlying connective tissue disorder in the medical history or at physical examination can be found in as much as 60% of cases: 1 in 5 patient presenting SIH have Marfanoid features and 2 in 5 joint hypermobility. Personal or family history of retinal detachment at an early age also predisposes to connective tissue disorder[3-19].

Trivial trauma defined as minor daily events including coughing, positional changes, sports, sexual intercourse or even child birth are reported in one-third of patients[20].

Although described as rare during the peripartum period, anatomical and physiological changes occurring during pregnancy may lead to SIH. Degradation of collagen and reduction of elastin observed before delivery in association with epidural or spinal anesthesia could contribute to dural weakness[5].

Diagnosis

According to the ICDH-3, diagnosis relies on recognition of clinical presentation and is confirmed by imagery and/or lumbar puncture[7].

Brain imaging

Brain MRI

MRI of the brain obtained with and without gadolinium injection is the most specific diagnostic tool but is only moderately sensitive: findings are normal in about 20% of patients. Head MRI abnormalities can be summarized by the acronym SEEPS"[8]:

- Subdural fluid collection: subdural hematoma and hygromas ;
- Enhancement of meninges: diffuse, smooth pachymeningeal enhancement (DPGE) and always without leptomeningeal involvement. Consequence of dural veins dilatation according to the Monroe-Kellie doctrine[21]. Pathognomonic sign (present in 3 out of 4 patients) and seen on average for 15 weeks after onset of symptoms (decrement in MRI findings with time)[11] ;
- Engorgement of veins: dilated cerebral venous sinuses ;
- Pituitary hyperemia ;
- Sagging of the brain: decreased size in ventricles and obliteration of the basal cisterns, increased anterior-posterior diameter of the brainstem, cerebellar tonsillar herniation, inferior displacement of the optic chiasm and reduced optic nerve sheath diameter and thickness[14]. Consequence of the downward displacement of the brain from low CSF buoyancy and related to the severity of symptoms. This sign is very specific[22].

There are no quantifiable criteria for head MRI abnormalities. An 18-points ranking score, known as the Bern score, has been proposed[23].

Brain CT

CT of the brain is of limited use as no reliable sign of SIH are demonstrable and it is usually normal.

However, it remains an important tool in the emergency department setting where MRI is not readily available or in patients who cannot undergo MRI due to contraindications, particularly in ruling out other serious conditions[8].

Subdural fluid collection, cerebellar tonsillar herniation, ventricular collapse and hyperdensity at the level of the tentorium sylvian fissure (as a sign of subarachnoid pseudo hemorrhage) are among the observable abnormalities but they are neither sensitive nor specific[16].

Spinal imaging

The aim of spinal imaging is to exhibit the CSF leak or its indirect signs.

CSF leakage occurs for the majority at the cervicothoracic junction or at the thoracic spine and is multi-level 25% of the time. 4 types of CSF leaks have been described[23]:

- Type 1: ventral and posterolateral dural tears ;
- Type 2: proximal nerve root sleeve tear, typically meningeal diverticulum ;
- Type 3: CSF-venous fistula (initially described in 2014 but a growing number) ;
- Type 4: distal nerve root sleeve tear.

Noninvasive and invasive techniques are currently in use and serve different purposes.

Noninvasive techniques

Noninvasive imaging of the spine essentially consists of spinal MRI as CT is of limited benefit in CSF study. It relies on the identification of indirect signs of CSF leakage and is poorly sensitive in determining the localization of such a leak. In approximately 10% of patients, extradural CSF accumulation that can be seen at C1-C2 is an extension of a lower leakage and a false localizing sign[24].

Spinal MRI

MRI of the spine is considered complementary to brain imagery. T2 fat suppression sequence allows for visualization of spinal longitudinal extra dural fluid (SLEC) in about 50-75% of patients, type 3 and 4 leaks being SLEC-negative"[23]. Engorgement of spinal epidural venous plexuses and ventral shifting of the spinal cord in the thoracic area are also frequently reported[25]. Rarely, MRI can show spinal pachymeningeal enhancement or meningeal diverticula[16].

Invasive techniques

The following techniques are classified as invasive because intrathecal injection is required. Dural puncture might present the risk of worsening intracranial hypotension. However, it has only been reported by 5% of patients, was generally mild and cerebral herniation has never been reported[22].

They are the only reliable tools in precisely locating the leaks and therefore can be used for diagnostic and therapeutic purposes. Some of the following reasons might explain why some leaks are not identified[6]:

- Low-flow leaks and CSF-venous fistulas are subtle ;
- Imaging technique is not adequate ;
- Intermittent or postural leaks are not present in the recumbent position.

CT myelography

Intrathecal injection of iodinated contrast followed by thin-cut CT of the entire spine is the gold standard for precise location of a CSF leak. Images can be acquired in two different ways[13]:

- Conventional: contrast is injected under fluoroscopic guidance, table is titled and redistribution to the cervical region is observed. The patient is then transported to the CT scanner;
- Dynamic: injection is performed in the CT scanner which permits immediate acquisition and localization of high-flow leaks. Radiation exposition is higher.

MRI myelography

It is considered as reliable as CT myelography but intrathecal gadolinium is an off-label use. MRI myelography can be considered when low-flow leaks are not localized on CT myelography or to locate ventral dural tears and CSF-venous fistulas[8].

Radioisotope cisternography

Radioisotope cisternography consists of intrathecal injection of Indium-111 followed by sequential scanning at various intervals up to 24-48 hours. It can be useful when diagnosis is uncertain and myelography normal. Absence of radioactivity over cerebral convexities, which generally does not extend beyond the basal cisterns, is a characteristic sign. Display of radioactivity in the kidneys in less than 4h, explained by extravasation of the radioisotope in the para-spinal space and early venous resorption, is commonly observed as well[16].

Lumbar puncture

A CSF opening pressure < 60mm H2O is one of the diagnostic criteria in the ICHD-37. However, normal or even elevated CSF opening pressures are reported in up to one third of patients. In addition, CSF analysis is widely variable and can either be normal or pathological. Lymphocytic pleiocytosis, elevated protein count and xantochromia are often seen. What remains constant is that cytology and

microbiological analysis are always normal and glucose is never low[14].

Differential diagnosis

In this section and for the relevance of our case report, we focused on the differential diagnosis of headaches as classified by the ICDH-3 with special focus on headaches presenting with a postural component and/or in the peripartum period. They may be categorized as primary or secondary based on the evidence of an underlying cause. Headaches can also be classified as either acute or chronic based on the timing of symptoms, with the latter requiring a minimum 3-month duration.

SIH can be classified as a secondary headache presenting with a postural component. There are few articles in literature about its prevalence in the peripartum period and references about the syndrome primarily consist of case reports.

Orthostatic headaches

As stated before, not all headaches related to SIH are orthostatic, and not all orthostatic headaches are caused by SIH.

Primary headaches

Cervicogenic headaches, which are common, might also present a postural component.

New daily persistent headache (NDPH) which refers to chronic headache with an acute onset is another type of primary headache. Some patients experiencing chronic orthostatic headaches as a result of SIH in whom imaging did not reveal any direct or indirect signs of CSF leakage might be wrongly classified as having NDPH[6].

Secondary headaches

When an individual presents with an orthostatic headache following a spinal or epidural procedure, the primary diagnosis that should be considered is PDPH. This condition will be further developed in the upcoming section.

Subarachnoid hemorrhage, carotid or vertebral artery dissection, CVST, benign intracranial hypertension, posttraumatic headache and meningitis have to be taken into account when dealing with postural headaches[6].

Other causes of secondary postural headaches are worth to be mentioned:

- Postural tachycardia syndrome (POTS) is a form of dysautonomia mainly characterized by a chronic headache with a postural component, dizziness and blurred vision. POTS may be associated with Ehlers-Danlos Syndrome[16].
- Craniocervical instability (CCI) is a condition where the craniocervical junction is unstable, resulting in non-pathological deformation of the brainstem, upper spinal cord, and cerebellum. CCI is most commonly observed in individuals with hereditary connective tissue disorders like Ehlers-Danlos Syndrome[6].
- Syndrome of the trephined is a consequence of the surgical treatment for Chiari malformation that may lead to an increase compliance of the dural sac without a CSF leak[21].

Headaches in the peripartum period

The term peripartum period can be defined as the immediate time before and after child birth. It typically starts at the 28th week of pregnancy and ends 6 weeks after delivery. The occurrence of headaches in the first six weeks after delivery is known as postpartum headache[1]. However, Goldszmidt et al found that 39% of women experienced headaches within 3 months of giving birth and late postpartum period can extend up to 6 months[26].

Primary headaches

Primary headaches are frequently reported in the general population and can also be observed during the peripartum period. They mainly consist of migraine and tension-type headaches[4].

Secondary headaches

Pre-eclampsia, eclampsia and CVST are major causes of secondary headaches in the peripartum period4. Other rare causes of headache during pregnancy and the postpartum period include posterior reversible leukoencephalopathy, epileptic seizures, subarachnoid hemorrhage, carotid or vertebral artery dissection, posttraumatic headache, and meningitis6. These types of headaches usually do not exhibit any postural component.

The leading cause of orthostatic headache in the early postpartum period is PDPH, accounting for approximately 5% of all postpartum headaches4. The headache presenting with a postural component is secondary to a dural leak, similar to SIH. However, in opposition to SIH, the leak is not spontaneous but the consequence of a dural puncture either inadvertent during epidural anesthesia at the time of labor or expected during spinal anesthesia for cesarean section. It can also be the result of a lumbar puncture.

The most recent definition from the ICDH-3 describes PDPH as a headache that occurs within five days of a dural puncture and is caused by leakage of cerebrospinal fluid through the puncture site. It resolves spontaneously within two weeks after the puncture or after an EBP has been applied"[7]. In obstetrical anesthesia, the incidence rate of an accidental dural puncture (ADP) varies from 0.4 to 6% after epidural anesthesia[27]. Among

these accidental punctures, it is estimated that approximately 52% to 85% of the patients concerned will present a PDPH syndrome[28]. The time of onset of symptoms as described by the ICDH-3 varies greatly. In 66% of cases, headaches occur within two days after the procedure and it is estimated that 90% occur within three days[29]. In addition, other cases of PDPH revealed more than a week after the dural puncture have been reported up to 14 days after the procedure[30].

Treatment

No randomized-controlled trial has assessed SIH treatment, and the current data on its outcomes predominantly relies on retrospective studies with limited patient enrollment[6].

Conservative measures

Conservative therapy including bed rest, analgesics and hydration is first-line treatment. Keeping the body in a supine position relieves pressure at the site of CSF leakage and thereby can help restoring normal brain volume and alleviating symptoms[18].

Efficacy is still a topic of debate and comes at a high social and economic cost to patients6. While some suggest that the majority of cases resolve spontaneously[11], other research has shown that as few as 15% experienced an improvement within 2 weeks from symptom onset with conservative treatment[16]. Furthermore, patients may experience debilitating symptoms or may have already exhausted the conservative approach due to delayed diagnosis[12]. A recent meta-analysis showed that conservative treatment was attempted for an average of 7 to 9 weeks and was successful in 28% of patients. A significant proportion of patients treated with conservative therapy remained symptomatic at 2 years[9].

The common practice is to consider the use of EBP when 1) orthostatic headaches persist after 14 days of bed rest or 2) symptoms are causing significant impairment.

Epidural blood patch

EBP with autologous blood is considered the mainstay treatment in SIH. Two potential action mechanisms have been proposed:

- An almost immediate effect associated to a temporary increased pressure in the epidural space transmitted to the intradural space[3] ;
- A latent effect associated to an inflammatory reaction sealing the site of the leak[31].

EBP can be performed blindly at the lumbar level or targeted at the site of leakage. As stated before, CSF leakage in SIH occur more frequently at the cervico-thoracic junction and the thoracic spine.

Blind EBP

Advocates for blind EBP suggest that the site of the leak is not influential because the results are primary due to the first effect or the inversion of gradient between the epidural (maintained negative by lymphatic drainage in the inferior vena cava system) and the dural compartment[32]. Furthermore, blood is thought to spread diffusely including up to the cervical area of the spine. This assumption was confirmed by an MRI study in one patient showing that 10 mL of blood injected at the lumbar area was found up to the cranium 1h after[22]. Cho et al also studied the spread of a mixture of blood and contrast agent. Spinal CT scans performed immediately after EBP showed that the injection of blood could cover up to 12 levels from the primary site of injection[33].

The success rate obtained after the first EBP in SIH ranges from 30% to more than 60%17-25[34]. Patients can also relapse after initial treatment and successive EBP may be needed to achieve complete symptom relief. There are case reports of up to seven EBP11. The evolution of MRI findings does not correlate with the efficacy of EBP[35]. EBP success rate is significantly lower than that of PDPH where it is generally thought to be close to 90%. This could be explained by[17]:

- Untargeted EBP at the site of the leak ;
- The anatomy of the leak not being a simple hole;
- Leaks can be located in the ventral part of the spinal cord, the nerve root sleeves or consists of a CSF-venous fistula[36] ;
- Leaks are multi-level in 1 out of 4 patients ;
- Time interval before treatment which is generally early in PDPH but longer in SIH as it is often misdiagnosed. This can lead to compensatory mechanisms of chronic CSF volume depletion like changes in CSF production rate[6].

The volume of blood required for an EBP at the lumbar level is unknown. Reported volumes used range from 10 to 50mL. Overall, the maximum volume injected is limited by local back pain, development of radiculopathy or headache[12].

Some measures have been proposed to achieve better success:

- Higher volume of blood, up to 100mL[37] ;
- Prone, lateral or decubitus position associated with Trendelenburg position for one hour after EBP to allow diffusion of blood12 or even up to 24 hours if the leak is located at the cervical spine. Trendelenburg position 30 minutes before the procedure and during the procedure has also been suggested ;
- Blind EBP placed at the thoracic region as leaks occur more frequently at the thoracic spine or the cervico-thoracic junction[37] ;

- EBP performed at two different sites at the same time[12] ;
- Eviction of physical exercise and Valsalva maneuvers for 7 days to avoid a sudden increase in intracranial pressure that might dislodge a blood clot ;
- Strict bed rest for 24 hours[16] ;
- Fluoroscopic guidance to confirm that the tip of the Tuohy needle is adequately placed in the epidural space[33] ;
- Repeated EBP have a cumulative effect[11].

When there is no symptom relief, a second EBP can be performed generally 7 days after the first one[16]. An interval of less than 5 days is contraindicated because of the high volume of blood injected and the risk of developing a rebound intracranial hypertension. However it is possible that a long gap may decrease the chances of success[37].

Lumbar EBP is generally regarded as a safe procedure, with most complications being mild and temporary. Blood leaks into the subcutaneous tissues and intrathecal injection of blood, epidural infection, subdural hematoma, cauda equina syndrome, neck stiffness and back or radicular pain are among them[15].

Targeted EBP

There is currently no consensus on whether targeted or blind EBP is the best approach[33]. Arguments for targeted EBP rely on the hypothesis that sealing of the leak would occur if blood is injected at the same level (second mechanism). Different research studies have reported conflicting findings regarding the relationship between the location of CSF leakage and the response to EBP injection. While some studies found no significant difference between the site of CSF leakage and the location of EBP, others reported statistical differences, particularly when the CSF leak is located in the cervical spine.

Correia et al reported that targeted CT guided EBP is more effective, with symptom relief achieved after the first attempt in71 to 87% based on the reviewed cases[15]. However, Lee et al retrospectively studied 62 patients who underwent EBP between 2010 and 2015 in a single hospital and found no statistical difference in clinical response regarding EBP distance from the leakage site[38]. Moreover, outcomes after 5 years seem to be unrelated to the site of delivery as well[34]. In a retrospective study of 116 patients, Ahn et al compared the complete recovery rates following first targeted EBP versus two-site blind EBP and did not find any significant difference between the two approaches either[39].

Targeted EBP should be performed using fluoroscopic guidance to confirm the adequate level of delivery. Adding contrast to the autologous blood to carry out epidurography or performing spinal CT scans after the procedure might help showing that the patched blood appropriately covers the site of the leak[33]. Overall, tolerated volume for cervical and thoracic EBP is lower compared to lumbar EBP[12].

Performing an EBP at the thoracic and cervical level is technically challenging compared to the lumbar level and is associated with higher risk of procedural complications[34]. Technical difficulties involve the restricted epidural space and the uncertainty of the precise location of CSF leaks[33]. Radiation exposure is another notable disadvantage that should be taken into account.

To summarize, it is usually recommended that when two or three attempts at a lumbar EBP of appropriate volume executed by a skilled operator have failed, CT-myelography imaging the site of the leak should be performed and fluoroscopy-guided targeted EBP undertaken. If symptom relief is still not achieved, multidisciplinary reassessment and other approaches like EBP using fibrin glue or surgery have to be considered[16].

Reappearance of headache following successful treatment may indicate a recurrence of the leak but if the symptoms have changed rebound transient intracranial hypertension, subdural hematoma and dural venous sinus thrombosis should be considered. Recurrence occurs in 10% of patients regardless of treatment[12].

Surgery

Surgery may be effective in selected cases and in patients who failed to respond to conservative treatment and repeated EBP. Sobczyk et al analyzed case reports and series of patients treated with surgery from 1998 to 2022 and found an overall success rate ranging from 82.6 to 100%. Surgical approach requires the exact site of the CSF leak and its direction to be known[40]. Surgical techniques described in literature include suturing dural defects, ligating diverticula or healing with gel foam and fibrin glue[3].

CSF-venous fistula also known as type 3 CSF leak responds poorly to EBP compared to other types of CSF leak. In a retrospective study of 44 patients presenting with SIH secondary to CSF-venous fistulas, Duvall et al demonstrated that only 10% showed clinical improvement lasting 3 months after EBP. Surgery consisting of ligation of the thoracic nerve root and its associated vein was attempted in 42 patients, 80% of whom presented enhanced medical condition. Diagnosis may also

be challenging since no extradural CSF is found on conventional spinal imaging (SLEC-negative) and intrathecal injected contrast is rather seen in an epidural vein[36].

Others

Other treatments have been cited in literature with limited or no effectiveness:

- Oral or intravenous caffeine ;
- Oral theophylline[18] ;
- Oral mineralocorticoids and corticosteroids[16] ;
- Intravenous administration of factor XIII[18];
- Vitamin A[41] ;
- Abdominal binder[16] ;
- Intrathecal or epidural infusion of crystalloids or colloids[12].

EBP with fibrin glue has also been considered and has shown encouraging results. However, it requires to be injected at the exact site of the leak as fibrin glue doesn't diffuse as well as blood. Rare anaphylactic reactions have been reported[12].

More recently, greater occipital nerve blockade has been proposed in association with EBP based on its demonstrated efficacy in reducing headaches associated with PDPH after caesarean section[42].

Discussion

SIH is not a rare entity. However, the syndrome is still not fully understood and remains unknown by a lot of specialists including anesthesiologists. The given name of the pathology itself and ICDH-3 diagnostic criteria are misleading since not all patients exhibit low CSF opening pressure. Mokri proposed the alternative term of spontaneous CSF hypovolemia"[43].

Orthostatic headache is the most common manifestation. The postural component may not be as evident and there may be a delay in the onset of symptoms when standing upright especially when headache becomes chronic. Meningeal irritation and cochlear-vestibular symptoms are other frequently reported signs[11,12].

Association of dural weakness with a trivial trauma may precipitate the dural leak. Connective tissue disorders like Marfan syndrome, Ehlers-Danlos Syndrome, autosomal dominant polycystic kidney disease and underlying connective tissue disorders are among the precipitating factors of dural vulnerability. In fact, discovery of a spontaneous CSF leak should lead to investigation such as analysis of collagen and elastin fibers in skin biopsies especially if suspected at physical examination[44].

Brain MRI with gadolinium injection is the imagery of choice but diagnosis should not be excluded based on normal findings[6]. Recognition of MRI signs might be the reason for the growing diagnosis of the syndrome[22]. Brain MRI findings indicative of intracranial hypotension, as described by the SEEPS acronym, may also be present in patients experiencing PDPH. However, since these patients undergo imaging less frequently or at an earlier stage of the disease, the observed abnormalities are more likely to be associated with acute cerebral changes, such as venous sinus distension, rather than chronic cerebral changes like brain sagging or subdural fluid collection[45]. Spinal MRI is complementary to diagnosis but finding of SLEC is not indicative of a spinal fluid leak at the same location. Referring to our case report, a CT scan of the brain was performed initially, and although this is not the recommended imaging modality for SIH diagnosis, it was considered appropriate because the patient presented in the emergency room. Ultimately, the diagnosis of SIH was based on the patient's clinical presentation.

Conservative treatment consisting mainly of bed rest and analgesics should be attempted for a maximum of 14 days. After that or in the presence of debilitating symptoms, blind lumbar EBP using autologous blood should be performed. Blind EBP provides a chance for prompt and economical treatment, and is considered a safe method. Although patients may not experience instant and complete relief, they typically observe an improvement in symptoms. To diagnose SIH, EBP has been proposed as a diagnostic tool and the relief of headache within 72 hours was included in the diagnostic criteria as specified in the ICDH-2 (2004) guidelines[19].

It is important to note that headaches can recur in SIH even after successful EBP and it should not be viewed as a negative prognostic sign. A second or third blind EBP should be conducted after a minimum interval of 5 days. If necessary, invasive imaging such as CT myelography may be conducted to identify the precise location of the leakage, which can facilitate targeted EBP under fluoroscopic guidance. Surgery may also be considered as an alternative treatment in appropriate cases.

The use of invasive imaging has been progressively abandoned due to the lack of significant advantages of targeted EBP[32]. Invasive imaging may only be necessary for diagnostic purposes if both brain and spinal MRI are negative or in cases where repeated blind EBP have been unsuccessful. CT-myelography and radio-isotope cisternography are the preferred imaging modalities. Lumbar puncture should be avoided because of its high degree of variability, particularly if the diagnosis has already been established through suggestive symptoms and imaging.

Orthostatic headaches, especially in the postpartum period, are not always related to SIH. When symptoms occur within the first days following an epidural or dural procedure, PDPH is more often suspected, and in the peripartum setting, pre-eclampsia, eclampsia, CVST, migraine or tension headaches should be evaluated and excluded beforehand.

In the late postpartum period, and particularly more than 3 months after an uncomplicated delivery, other hypotheses should be considered. Primary headache disorders such as new daily persistent headache and secondary causes of SIH like subarachnoid hemorrhage, carotid or vertebral artery dissection, CVST, posttraumatic headache and meningitis should be set aside in the first instance. POTS and cervicogenic headache also have a similar symptomatology and can be confused with SIH. In our case, given the abrupt and orthostatic nature of the symptoms (headache, nausea and vomiting in the standing position), the absence of prior trauma, and the delay in the onset of symptoms after the epidural procedure, SIH was quickly suggested.

SIH is rarely described during pregnancy and even less in the postpartum period. A PubMed search found eleven relevant cases in literature of SIH occurring during pregnancy and only two cases in the postpartum period, up to one month after delivery.

Concerns of SIH implication during pregnancy include differential diagnosis with severe pathology like CVST or less severe primary headache-types, adequate treatment and imaging modalities, and appropriate mode of delivery. They are discussed in the following case reports:

- Asakura et al reported the first case of SIH occurring during pregnancy. The woman was admitted at 8 weeks gestation and received conservative treatment. She was discharged after a month when symptoms reduced[46] ;
- Hashmi reported a case of rapid clinical improvement to high-dose oral glucocorticoids in a woman in her 10th week of pregnancy after failure of conservative treatment[47] ;
- Singh et al stated that conservative treatment should be tried for a longer time before considering EBP in pregnancy after their patient failed to respond to EBP but symptoms relieved after further conservative treatment[48] ;
- Grange et al reported the case of a woman who benefitted from two consecutive EBP during pregnancy with completely recovery. She then received spinal anesthesia for caesarean section indicated by transverse presentation of the fetus and previous cesarean section without any recurrence of symptoms at 6 months postpartum[49];
- Reihani et al reported a case of SIH initially misdiagnosed as CVST. Diagnosis of SIH was confirmed by MRI and CT of the brain and decision was made to end pregnancy by cesarean section because labor pain would have been unbearable. The authors noted that the patient's symptoms had improved after delivery without invasive treatment and therefore pregnancy may have contributed to the worsening of symptoms[50];
- McGrath et al presented a case of reoccurrence of SIH in early pregnancy 5 years after presenting with the syndrome during the first pregnancy. Both cases were successfully treated by EBP and the patient presented no symptoms in between. They discussed the role of pregnancy as a risk factor for SIH and differential diagnosis of headache in association with nausea and vomiting of the first term[51] ;
- Ferrante et al, a team of neurologists and gynecologists, presented a case series of 5 parturients affected by SIH, one improving after conservative treatment and the remaining necessitating lumbar EBP. They highlighted the risk of gadolinium MRI in pregnancy because of the accumulation of the contrast agent in the amniotic fluid and fetal tissue. Despite the potential risks associated with performing neuroimaging, Ferrante et al concluded that the inherent risks of the disease outweigh the risks of the procedure. They also recommended natural birth even if three of their patients underwent cesarean section by their own choice[52].

Two cases of postpartum SIH are reported in literature. Both happened during the first month after delivery. We discussed them in the following segment and made comparison to our case in Table I.

Albayram et al (2008) reported the case of a 29-year-old woman who presented with SIH 4 days after vaginal delivery without epidural anesthesia. In addition to orthostatic headache, she also demonstrated ceasing of lactation similar to our case. Although more often associated with galactorrhea, the authors explained the reduction in lactation by an impaired venous drainage of hypophysis caused by SIH which led to a dysfunction of the pituitary gland and a decreased outflow of hypophysis hormones. They also mentioned that stress and pain could have played a role. She benefitted from a blind lumbar EBP and fully recovered at a 6 months follow-up[4].

Van Sonderen et al (2013) reported the case of a 25-year-old who presented with orthostatic headache 4 weeks after vaginal delivery without epidural anesthesia. The patient was wrongly diagnosed with migraine based on normal brain MRI results and was discharged. She presented again 2 weeks later with

Table I. — Case reports of SIH occurring in the postpartum period.

	Albayram S, 2008	*Van Sonderen A, 2013*	Our case
Age and parity	29-year-old, G2P2	25-year-old	31-year-old, G1P1
Comorbidity	No	No	No
Mode of delivery	Uncomplicated vaginal delivery	Uncomplicated vaginal delivery	Uncomplicated vaginal delivery
Neuraxial anesthesia	No	No	Epidural anesthesia
Onset	4 days PP	4 weeks PP	3 months PP
Clinical presentation	Orthostatic headache, neck and right arm pain, truncal ataxia during standing and ceasing of lactation	Bifrontal orthostatic headache, photophobia, nausea and one episode of vomiting Diplopia at 6 weeks PP	Bilateral holocranial orthostatic headache, nausea, vomiting and ceasing of lactation
Clinical examination	Normal	Normal at 4 weeks PP Right-sided abducens nerve palsy at 6 weeks PP	Normal
Diagnosis	Brain MRI with gadolinium: DPGE Lumbar puncture : decreased CSF opening pressure (2cmH2O) CT-cisternography: leak of contrast at the lower cervical and upper thoracic region	Brain MRI with gadolinium at 4 weeks PP: normal Brain MRI with gadolinium at 6 weeks PP: bilateral subdural hygromas around the cerebrum and cerebellum Lumbar puncture at 6 weeks PP: CSF opening pressure unmeasurable	Cranial CT-scan in the emergency room: normal Spinal MRI 5 days after first admission: collection of liquid signal in the epidural space at the thoracic region
Treatment	Full bed rest with LMWH during investigation stopped after CVST diagnostic ruled out EBP because unbearable headache	Analgesics for suspicion of first migraine attack Readmitted 2 weeks later because symptoms severity increased ; SIH diagnosis was made and treated with EBP	Analgesics and caffeine for one day Readmitted the next day for blind EBP Second blind EBP at day 15
EBP	L2-L3 fluoroscopy-guided with 50mL of autologous blood	At 6 weeks PP but no more information	First EBP: L4-L5 level with 20mL of autologous blood Second EBP : L3-L4 level with 20mL of autologous blood
Follow-up	At 5 days: symptoms decreased and lactation recovered At 6 months : full recovery and normal brain MRI	Slow but full recovery in the following weeks	Full recovery after the second EBP Second pregnancy 2 years later with no recurrence of SIH

G = gravity ; P = parity ; PP = postpartum ; DPGE = diffuse pachymeningeal enhancement ; LMWH = low molecular weight heparin ; EBP = epidural blood patch.

diplopia and a second brain MRI was performed confirming SIH diagnosis. She underwent a blind EBP and recovered the following weeks. The authors suggested that SIH might be considered when dealing with postpartum headaches[1].

In our case discussion, we explored the differential diagnosis of SIH with PDPH, and considered the possibility of a link between neuraxial anesthesia and the development of SIH. As there is little existing literature on this subject, we drew conclusions based on the following case reports:

- An et al reported the case of a 56-year-old man who received lower thoracic epidural anesthesia for appendectomy and experienced orthostatic headache the next day. PDPH diagnosis was made and the patient was discharged with conservative treatment. Headaches persisted for 3 months and a lumbar EBP was performed with uncomplete resolution of symptoms. He was diagnosed 5 days later with SIH when spine MRI and CT myelography showed multiple meningeal diverticulum in the cervicothoracic junction and a C5-C6 spinal dural tear. A targeted EBP in the upper thoracic region was done and symptoms decreased. The authors concluded that epidural anesthesia might have triggered SIH based on studies showing that injection of saline in epidural space provoked rapid rise of pressure in the epidural and subarachnoid space , speculating that this rapid rise in pressure might induce the dural sac to tear[53] ;
- Lee et al reported the case of a patient who presented orthostatic headache for a year after spinal anesthesia for knee arthroscopy surgery. PDPH was diagnosed and conservative treatment was administered for a week but proved ineffective. Subsequently, a lumbar EBP was performed which led to partial improvement.

However, the patient's headache worsened. An MRI myelography was conducted, revealing CSF leakage at the T4-T7 levels. The patient then received fluoroscopy-guided targeted EBP at the T8-T9 level resulting in an 80% reduction of symptoms. A second targeted EBP at the same level a week later led to complete resolution of symptoms[54].

These are the takeaways from these case reports:

- SIH can occur during the peripartum period and pregnancy might play a role ;
- Although there is no evidence in terms of treatment during pregnancy, blind EBP can be performed safely and showed clinical improvement in multiple patients ;
- There is no clear evidence on the use of imaging during pregnancy, but gadolinium-enhanced brain MRI can be considered if SIH is suspected, despite the potential risk to the fetus ;
- Vaginal delivery is recommended in patients presenting with SIH during pregnancy, and epidural anesthesia is not contraindicated ;
- SIH should be considered as a possible cause of postpartum headache ;
- SIH in the postpartum period may lead to a reduction in lactation ;
- Epidural anesthesia may trigger SIH.

Another important consideration is that the physiological enlargement of the pituitary gland, which is commonly observed on MRI during pregnancy, should not be misinterpreted as pituitary hyperemia seen in SIH[52].

Severe complications in SIH are relatively rare and typically manifest at a later stage of the disease. Complications such as CVST are observed in less than 1% of cases. However, given that the peripartum is a known risk factor for thromboembolic events, there is a possibility that the incidence of CVST may be higher in the peripartum period. It's important to remain vigilant for such complications, especially in high-risk patients, and to promptly manage and treat them as appropriate[52].

Limitations

We conducted a literature review using a narrative approach. Although we primarily relied on meta-analysis to draw our conclusions, we also included case reports that described the peripartum period. Furthermore, there have been no randomized controlled trials that have assessed treatment and imaging modalities. As a result, our review predominantly relied on expert opinions.

While the postpartum period is strictly defined as six weeks after delivery, our case report occurred three months after giving birth. Despite this, we feel that it holds relevance to our work as anesthesiologists.

Conclusions

SIH is not as rare as previously thought and might as well present during the peripartum period. We believe that it should be taken into consideration when dealing with postpartum headaches even when suspecting PDPH after an inadvertent dural puncture when performing epidural anesthesia during labor. Onset of symptoms later than one week after the procedure, absence of clinical improvement after conservative treatment, uncomplete response after one EBP or relapse after initial improvement as well as clinical history of connective tissue disorder or physical examination suggesting such should raise suspicion. This is of crucial importance regarding the debilitating symptoms, possible recurrence and the potential severe complications of SIH. Multidisciplinary approach and neurological follow-up of these patients are therefore essential.

SIH is often underdiagnosed and misdiagnosed. Anesthetists in particular are not familiar with the pathology. However, they play a critical role in the management of SIH due to their expertise in performing EBP. This review highlights the lack of resources in literature about SIH in the peri- and postpartum period. Further investigations are needed to investigate the pathogenesis of SIH in this setting as well as the optimal treatment modalities.

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