American Journal of Emergency Medicine 47 (2021) 24–29

Contents lists available at ScienceDirect

American Journal of Emergency Medicine

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

Cerebral venous thrombosis: Diagnosis and management in the emergency department setting

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ARTICLE INFO

Article history:
Received 3 February 2021
Received in revised form 4 March 2021
Accepted 10 March 2021

Keywords:
Cerebral venous thrombosis
Thrombosis
Neurology

ABSTRACT

Introduction: Cerebral venous thrombosis (CVT) is an uncommon neurologic emergency associated with significant morbidity and mortality that can be difficult to differentiate from other conditions. It is important for the emergency clinician to be familiar with this disease as it requires a high index of suspicion, and early diagnosis and management can lead to improved outcomes.
Objective: This narrative review provides an evidence-based update concerning the presentation, evaluation, and management of CVT for the emergency clinician.
Discussion: CVT is due to thrombosis of the cerebral veins resulting in obstruction of venous outflow and increased intracranial pressure. Early recognition is important but difficult as the clinical presentation can mimic more common disease patterns. The most common patient population affected includes women under the age of 50. Risk factors for CVT include pregnancy, medications (oral contraceptives), inherited thrombophilia, prior venous thromboembolic event, malignancy, recent infection, and neurosurgery. CVT can present in a variety of ways, but the most common symptom is headache, followed by focal neurologic deficit, seizure, and altered mental status. Imaging studies such as computed tomography (CT) venography or magnetic resonance (MR) venography should be obtained in patients with concern for CVT, as non-contrast CT will be normal or have nonspecific findings in most patients. Treatment includes anticoagulation, treating seizures and elevated ICP aggressively, and neurosurgical or interventional radiology consultation in select cases.
Conclusions: CVT can be a challenging diagnosis. Knowledge of the risk factors, patient presentation, evaluation, and management can assist emergency clinicians.

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1. Introduction

Cerebral venous thrombosis (CVT) is an uncommon neurologic emergency that can lead to stroke, seizures, and death. CVT is defined by thrombosis of the intracranial veins and dural sinuses and has an estimated annual incidence of 0.3–1.5 cases/100,000 person-years, accounting for up to 1% of all strokes worldwide [1-3]. The disease is difficult to differentiate from other more common neurologic conditions due to its varied presentations. Delays in diagnosis are associated with higher morbidity, however, prompt recognition, diagnosis, and management can lead to improved outcomes [4]. Thus, it is important for the emergency clinician to be familiar with this disease.

2. Methods

This narrative review provides a focused overview of CVT for emergency clinicians. The authors searched PubMed for English language articles from 1990 to November 2020 using the keyword and Medical Subject Heading "cerebral venous thrombosis" for production of this narrative review, restricting results to retrospective and prospective studies, systematic reviews and meta-analyses, narrative reviews, and clinical guidelines. Initial literature search revealed 486 full text articles. Article inclusion was determined by author review and consensus based on clinical relevance to emergency department (ED) evaluation and management. When available, retrospective and prospective studies, systematic reviews/meta-analyses, and clinical guidelines were preferentially selected. A total of 57 articles were determined to be of relevance to emergency clinicians by author consensus and included in this narrative review. Of the 57 articles included in this review, there were 5 systematic reviews/meta-analyses, 2 guidelines from clinical societies, 7 RCTs, 24 observational studies, and 19 clinical reviews. Authors did not pool data for meta-analysis, as this is a narrative review.

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https://doi.org/10.1016/j.ajem.2021.03.040
0735-6757/Published by Elsevier Inc.

3. Discussion

3.1. Pathophysiology

The cerebral veins drain the capillary network that supplies the brain with blood (Fig. 1). The dural sinuses also drain the cerebrospinal fluid (CSF) via the arachnoid granulations and return the CSF into circulation through the blood stream [5]. An obstruction within this drainage system leads to increased venous pressure and reduced capillary perfusion pressure, which can result in ischemia, edema, elevated intracranial pressure (ICP), and even hemorrhagic infarction [1,5]. Two types of edema result from CVT: vasogenic and cytotoxic edema [6]. Vasogenic edema results from the elevated venous pressure that leads to disruption of the blood brain barrier and blood leakage across the interstitial membrane [6,7]. If the venous pressure remains elevated, subsequent ischemia of cerebral tissue disrupts oxygen dependent biochemical pathways and leads to cytotoxic edema [6,7]. Compression of the nerve fibers within the veins is thought to cause headache [5].

3.2. Epidemiology

Several multicenter cohort studies suggest a female predominance to CVT [8-10]. The International Study on Cerebral Venous Thrombosis (ISCVT), published in 2004, reported 74.5% of patients with CVT were women [9]. Additionally, this study found that 78% of patients with CVT were less than 50 years old [9]. A more recent cohort study of CVT among a Norwegian population found an equal prevalence of CVT among the sexes [11].

Approximately 85% of patients with CVT have at least one identifiable risk factor for thrombosis (Table 1) [12,13]. Oral contraceptive pill (OCP) use, pregnancy/peripartum status, obesity, and hypercoagulability are several of the most common risk factors, with OCPs increasing the risk of CVT six-fold [12,14]. Other risk factors include inflammatory conditions such as vasculitis or connective tissue disorders, sickle cell disease, head trauma, nephrotic syndrome, and dehydration [3,14]. Local infections of the head and neck, such as mastoiditis and sinusitis, have been reported to be associated with CVT, present in 8.2% of cases in one study [15]. Another important potential risk factor is recent surgery, particularly neurosurgery, which has been associated with up to 2% of CVT cases [9,12]. Malignancy has been reported to be associated with CVT in approximately 5% of cases, although some studies have noted that malignancy is a more common cause of CVT in patients over age 55, present in 25% of cases [15,16].

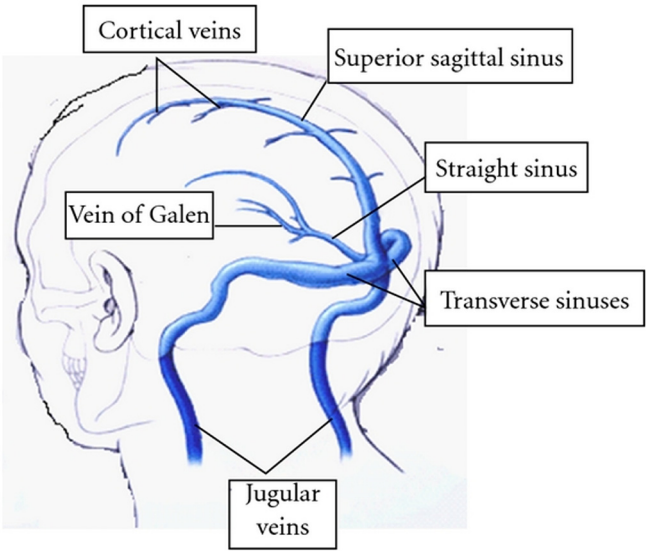

Fig. 1. Cerebral Venous Anatomy. Image Courtesy of Wikimedia Commons: https://commons.wikimedia.org/wiki/File:Major_venous_sinuses_and_their_tributaries.png

Table 1
Risk factors associated with CVT.

Risk factor	Examples
Medications	Oral contraceptives, hormone replacement therapy, glucocorticoids
Inherited Thrombophilia	Factor V Leiden, prothrombin 20210A, protein C deficiency
Acquired Hypercoagulability	Peripartum, malignancies, systemic lupus, nephrotic syndrome, inflammatory bowel disease, sickle cell disease
Infections	Mastoiditis, meningitis, sinusitis
Surgery	Craniotomy, indwelling ventricular catheter
Other	Obesity

3.3. History and examination

History should focus on the presence of these risk factors and medications that increase risk of thrombosis. In a study of risk factors for CVT, smoking, hypertension, and diabetes did not reach statistical significance, possibly reflecting the younger age of the population most affected by CVT [12]. Thus, the emergency clinician should consider CVT in patients with stroke-like symptoms without traditional stroke risk factors [12]. One study estimated OCP use among 54–71% of female patients with CVT, while another found OCP use in 10–73% of female patients with CVT [2,3,8,12]. Pregnancy and the first 6 weeks postpartum are higher risk periods for the development of CVT. One study estimated an odds ratio (OR) of 17.24 for pregnancy/post-partum period for CVT (95% confidence interval (CI) 6.83–44.04) [12], and CVT should be considered in patients with headache in this population. OCP use can also be easily missed if a thorough medication review is not performed. Additional risk factors identified for CVT include glucocorticoid use (OR 18.26; 95% CI 3.25–102.55), antiphospholipid syndrome (OR 6.98; 95% CI 2.06–23.6), factor V Leiden (OR 2.51; 95% CI 1.93–3.27), prothrombin 20210A (OR 5.53; 95% CI 3.98–7.69), and protein C deficiency (OR 10.74; 95% CI 3.07–37.65) [12]. One small study found a higher risk of CVT among patients at high-altitude, although many of these patients already had one hypercoagulable risk factor [17].

Patients present along a clinical spectrum based on the location of the thrombus, severity of ICP elevation, and presence of edema, making diagnosis difficult. Headache is the most common presenting complaint of CVT, occurring in 81–95% of patients, and focal neurologic deficits, seizures, and altered mental status can be other presenting features of CVT (Table 2) [2,8-11,17,18]. However, over 5% of patients will present without headache [18]. While headache is common in ED patients, several distinguishing factors may suggest CVT, as well as other diseases (Table 3). Additionally, while neurologic abnormalities are often utilized as a trigger for additional evaluation in patients presenting with headache, the VENOST study found that 25% of cases of CVT presented as isolated headache [8]. Unlike with subarachnoid hemorrhage, the headache in CVT tends to be more subacute, rather than sudden in

Table 2
Clinical presentations suggestive of CVT.

Clinical presentation
Headache plus sensory or motor deficit, oculomotor palsy, vision impairment, or papilledema
Atypical or severe headache in a patient with risk factor(s) for thrombosis
History of headache preceding a seizure, stroke, or altered mental status
Stroke in a young female patient or a patient without atherosclerotic risk factors
Acute stroke patient who develops a seizure
Stroke patient with neuroimaging that shows infarcts in multiple vascular territories

Table 3
Differential diagnosis and distinguishing features of CVT.

Disease	Distinguishing features
Meningitis	The classic triad of headache, fever, and neck stiffness is present in 44% of patients with meningitis [19]. Meningeal signs and symptoms, like neck stiffness, suggest meningitis, while focal neurologic deficits and papilledema should raise suspicion for CVT.
Subarachnoid Hemorrhage (SAH)	Headache, nausea/vomiting, neurologic deficits, and seizures can be the presenting symptoms of SAH or CVT [19,20]. SAH more commonly presents abruptly, with the headache reaching maximal intensity within a few minutes of onset, as opposed to headaches associated with CVT that typically develop over the course of days. Thunderclap headache is more common in SAH [19]. SAH also tends to occur in a slightly older patient population, with an average age of 57 years [21].
Acute Angle Closure Glaucoma	Headache, nausea/vomiting, and visual symptoms are common in acute angle closure glaucoma and CVT [19]. Abrupt onset (minutes to hours, rather than days), injected conjunctivae, blurry vision, and decreased pupillary reactivity increase likelihood of acute angle closure glaucoma.
Carotid/Vertebral Artery Dissection	Dissection can present as headache with neck pain and focal neurologic deficits. Like CVT, dissection can also present as isolated headache [19]. A history of trauma, neck manipulation, and sudden acceleration-deceleration should raise suspicion for vertebral or carotid dissection. Horner's syndrome, pulsatile tinnitus, and posterior circulation symptoms such as ataxia should also raise suspicion for dissection [19].
Temporal arteritis	Headache and visual symptoms can be the presenting symptoms of temporal arteritis, but additional features such as jaw claudication and fever can help distinguish it from CVT [19]. Additionally, temporal arteritis typically affects an older patient population (over 50 years), compared to patients with CVT (under 50 years) [19,22].
Idiopathic Intracranial Hypertension (IIH)	Headache, cranial nerve palsy, and papilledema can be present in IIH and CVT [8,23,24]. Neurologic deficits associated with IIH are more commonly abducens nerve palsy and visual field defects [23]. Other neurologic deficits in a patient with suspected IIH should also raise suspicion for CVT as the diagnosis. Several small studies found that MRI/MRV in patients with IIH was more likely to have signs of increased ICP on imaging, like flattening of the globe, an empty sella, or optic nerve tortuosity, compared to those with CVT [23,24]. Some studies have suggested that any patient with IIH should have outpatient follow up to determine the need for further evaluation for hypercoagulability [25].
Preeclampsia/Eclampsia	Headache, nausea/vomiting, and visual complaints in a pregnant or post-partum patient suggest preeclampsia or CVT [19]. Preeclampsia/eclampsia or HELLP syndrome should be considered in the setting of hypertension or laboratory signs of end organ damage such as proteinuria or liver function test abnormalities. The presence of focal neurologic deficits should raise suspicion for CVT.
Reversible Cerebral Vasoconstriction Syndrome (RCVS)	RCVS risk factors include peri-partum state, hypertension, and medications like triptans [8,26]. RCVS is an angiopathy characterized by abnormal vascular tone that leads to vasoconstriction and sometimes seizures and stroke [26,27]. Reoccurring thunderclap headache is characteristic of RVCS and uncommon in CVT [26]. Typical imaging findings include two or more areas of arterial narrowing on MRA [26].
Posterior Reversible Encephalopathy Syndrome (PRES)	PRES is associated with immunosuppression, end stage renal disease, and hypertension [27,28]. PRES results from endovascular dysfunction and elevated blood flow beyond the cerebral autoregulatory zones resulting in vasogenic edema. Depressed levels of consciousness and headache are more common in PRES. [28] Focal neurologic deficits can occur but are more rare in PRES compared to CVT [28]. Typical MRI findings in PRES are bilateral symmetric partial-occipital lesions [27,28].

Abbreviations: CVT – cerebral venous thrombosis, HELLP – hemolysis, elevated liver enzymes, low platelets, ICP – intracranial pressure, MRA – magnetic resonance angiography, MRI – magnetic resonance imaging, MRV - magnetic resonance venogram.

onset [3,8,18]. One study found that 57% of patients presented with subacute or chronic headaches, with symptoms progressing from 4 days to greater than 2 weeks, leaving 43% presenting acutely [8]. Another study reported 60% of patients presented acutely, although this study defined acute as 1–4 days after symptom onset [3]. While rare, there have been case reports of CVT presenting as maximal in onset, "thunderclap" headaches, which could prove problematic as clinicians may anchor on evaluating for subarachnoid hemorrhage [3].

Headache that is worsened with maneuvers that increase ICP (i.e., Valsalva maneuver, bending over, etc.) can be suggestive of CVT [3]. However, this is a non-specific finding and is common in other diseases that present with headache and increased ICP [3]. The location of headache in CVT is variable given the various locations of the venous sinuses, although it is controversial if the location of thrombus results in pain over the thrombus site [1]. Headaches can be unilateral, bilateral, frontal, or occipital [1,18]. A study evaluating the clinical features of CVT found that headache plus papilledema or headache plus seizure had a specificity of 97–99% for CVT, but sensitivity was only 7–10% [29]. Unfortunately, no combination of features demonstrated high enough sensitivity to definitively exclude the diagnosis [29]. Headache with a focal neurologic deficit or headache in the setting of recent neurologic surgery is also concerning for CVT [12].

Focal neurologic deficits, including cranial nerve palsies and visual field defects, occur in 31–68% of cases of CVT [6,9,17]. Studies have reported aphasia and other focal cortical functions, such as extremity weakness, as presenting symptoms in 13–16% of cases [8,9,11]. Papilledema is found in 28–57% of patients with CVT, and cranial nerve VI palsy may also occur due to elevated ICP [8-10]. Seizures occur in 23–44% of cases, which is markedly higher than in those patients with arterial ischemic stroke, where seizures occur in 2–9% of cases [1,8,9,11,17]. Thus, CVT is an important diagnostic consideration in patients presenting with seizure and focal neurologic deficit. In severe CVT with significant cortical involvement and increased ICP or thrombosis of the deep veins, the patient may present with altered mental status or encephalopathy, which occurs in 17–20% of cases [7]. The location of the thrombus can be associated with patient symptoms. However, most cases of CVT involve multiple locations and therefore may not present as a clear syndrome [15]. A summary of the frequency of different locations for CVT as well as associated symptoms is presented in Table 4.

3.4. Evaluation

3.4.1. Laboratory testing

Laboratory evaluation cannot be used to rule in or rule out CVT. D-dimer is often elevated in patients with CVT, but literature demonstrates a sensitivity of 82–94% [1-3,18]. The sensitivity is highest in patients with acute, extensive disease and lower in those with subacute or more focal thrombus. A recent prospective multicenter study proposed a predictive clinical score for the diagnosis of CVT (Table 5) [30]. The score ranges from 0 to 14 points (17 points if D-dimer is utilized). Low risk is 0–2 points, moderate risk is 3–5 points, and high risk is ≥6 points. D-dimer is not a mandatory part of the score but can be incorporated (D-dimer ≥500 μg/L is 3 points). The authors found that among the 186 patients with low probability for CVT (score 0–2), 11 patients, or 5.9%, were ultimately diagnosed with CVT [30]. Among this group of 11 patients who were low probability but diagnosed with CVT, all had a D-dimer >500 μg/L. [30] In this study the best CVT prediction model resulted from a CVT clinical score ≥ 6 and D-dimer ≥500 μg/L, with a sensitivity of 83% and specificity of 86.8% [30]. Although this is a promising step towards development of a useful tool for ruling out CVT, further studies are required [30]. Additionally, due to the poor specificity of D-

Table 4
Frequency and Associated Symptoms of Various Locations of CVT [9,10,15].

Location	Frequency (%)	Associated symptoms
Superior Sagittal Sinus	62–71	Headache Hemisensory loss Hemiparesis Seizures
Lateral Sinuses	31–47	Headache Aphasia (left sided) Seizures Mastoid pain
Straight Sinus including the Deep Venous System	10–18	Altered Mental Status Gaze palsy Coma
Cavernous Sinus	1–4	Eye pain Proptosis Cranial nerve III, IV, VI palsy

dimer, this test cannot be used in isolation to rule in CVT [18]. A review by the European Stroke Organization found that D-dimer was more likely to be falsely negative in CVT patients who presented with headache alone [16]. They also reported no difference in D-dimer levels in CVT patients with or without focal neurologic deficits [16]. The European Stroke Organization found that thrombophilia screening did not aid in the diagnosis or functional outcome of CVT patients and thus recommend against routine screening in patients suspected of CVT [16].

Lumbar puncture findings are similarly non-specific in CVT and are not definitively indicated for all patients with suspected CVT [7]. Increased opening pressure, pleocytosis, and increased red blood cells may be present [7]. A study of LP in CVT patients found that LP was normal in 44% of patients with CVT [31]. This study also found that the performance of an LP did not affect the prognosis of patients with CVT [31]. However, particularly among acutely ill patients, LP can play an important role in evaluating for life threatening diagnoses like bacterial meningitis or subarachnoid hemorrhage.

3.4.2. Imaging

CVT is primarily diagnosed by imaging [13,19,32]. The most common initial imaging modality obtained in the ED in patients with a suspected neurological condition (e.g., high-risk headaches, stroke, intracerebral hemorrhage, etc.) is non-contrast head computed tomography (CT). Non-contrast head CT possesses an important role in evaluating for other dangerous conditions such as intracerebral hemorrhage. However, this test has poor sensitivity and can be normal in 30–60% of patients with CVT [7,18]. Non-contrast head CT findings concerning for CVT include the "dense triangle sign" (thrombosis in the superior sagittal sinus) and the "cord sign" (thrombosis of cortical or deep veins) [3,7]. The cord sign is an area of increased attenuation in a dural sinus or vein (Fig. 2). This finding represents a clot, which appears hyper-attenuated in the acute phase, typically when symptoms have been present for less than one week. However, the clot will become iso- or hypo attenuated in the subacute and chronic phases [6,33]. Additionally, dehydration and elevated hematocrit may cause a false cord sign [34]. This sign may be found in 6.7–64.6% of cases [33-36]. Overall, the majority of non-contrast head CTs in CVT will be normal or have nonspecific signs of increased ICP, parenchymal abnormalities, or infarctions in multiple arterial distributions [2,3,7,18]. Thus, a normal non-contrast head CT should not be used to exclude the diagnosis of CVT, and in patients with severe headache or other findings concerning for CVT, further evaluation is necessary [32]. Hemorrhage may be present in approximately one third of patients, which is more commonly parenchymal and may occur in several arterial regions [32,34]. Head CT with intravenous (IV) contrast may reveal the "empty delta sign" (non-opacified thrombus surrounded by the collateral veins of the sinus wall after injection of contrast), which is consistent with a diagnosis of CVT but is present in only 29–35% of cases [6,34,37]. The empty delta sign may not be present in the acute phase, due to the hyperattenuated thrombus, but it may be present in the subacute phase [6].

CT or magnetic resonance imaging (MRI) with venous phase imaging is the recommended diagnostic modality and has high sensitivity and specificity. CT venography sensitivity and specificity approximate 95% for diagnosis of CVT and is readily available in most EDs [7,35]. MRI (without venous phase imaging) findings are dependent on the age of thrombus [6,7,38]. In the acute phase, 0–5 days after symptom onset, the thrombus will be predominately isointense on T1 weighted images and hypointense on T2-weighted images, resulting in the potential for missed diagnosis if MRI without venous phase imaging is used [6,34]. In the subacute phase, or days 6–15, the thrombus will be hyperintense on both T1 and T2 weighted images, thus making diagnosis easier radiographically [6,34]. After 15 days the thrombus can be isointense on T1 and T2 weighted images, again increasing the risk for missed diagnosis [6,34]. Because the duration of the CVT impacts MRI test characteristics, venous phase imaging is necessary. Similar to CT venography, magnetic resonance venography (MRV) displays high sensitivity and specificity for diagnosis [7,35]. While CT venography and MRV likely

Table 5
CVT Clinical Score.

Component	Points
Seizure(s) at presentation	4
Known thrombophilia	4
Oral contraception	2
Duration of symptoms >6 days	2
Worst headache ever	1
Focal neurologic deficit at presentation	1
[a]D-Dimer ≥500 μg/L	[a]3

Interpretation: Low risk – 0-2, Moderate risk – 3-5, High risk – greater than 6.
[a] D-dimer is not a mandatory part of the score, but if obtained, a D-dimer ≥500 μg/L is 3 points.

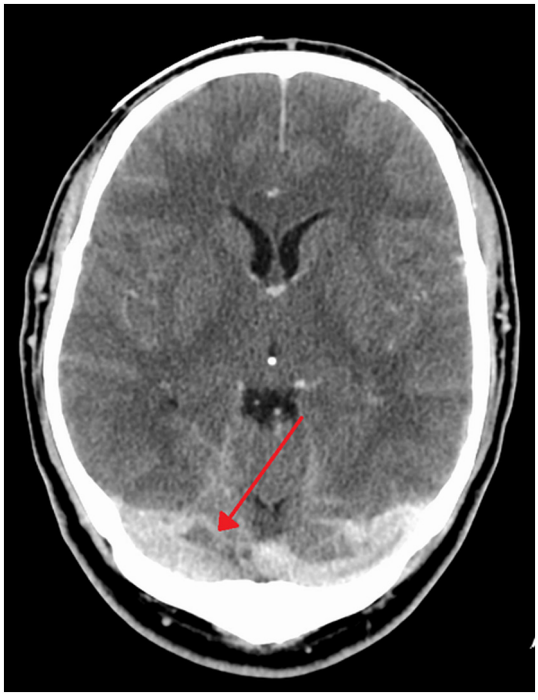

Fig. 2. Cord Sign on non-contrast head CT. Image Courtesy of Wikimedia Commons: https://commons.wikimedia.org/wiki/File:Duralvenoussinusthrombosis.png

perform similarly, MRV may be more effective for diagnosis in those with evidence of deep vein involvement (i.e., altered mental status).

3.5. Management

The overall goals of treatment of CVT are to treat the sequelae of CVT, prevent propagation of the clot, recanalize the occluded vessel, and prevent thrombosis elsewhere in the body.

3.5.1. Seizures

Important sequelae of CVT include seizures and increased ICP. Seizure is an independent prognostic risk factor for mortality [39]. Patients actively seizing should be treated with benzodiazepines. A retrospective study of seizures in CVT reported that lorazepam was the most commonly used medication to treat acute seizures from CVT [40]. Seizures in CVT are associated with supratentorial lesions [40]. European guidelines make a weak recommendation for patients with CVT who present with seizures and a supratentorial lesion to be started on an anti-epileptic drug to prevent early recurrent seizures, but they make no specific recommendation on agent choice or duration [16]. Seizure prophylaxis for all patients with CVT is not recommended.

3.5.2. ICP

Elevated ICP can be a devastating consequence of CVT. A major cause of death from CVT for patients is transtentorial herniation; thus, early management of ICP is critical [39]. There is little evidence to guide treatment of increased ICP in the setting of CVT. Therapeutic LP can be considered to reduce ICP, and although no randomized studies have demonstrated therapeutic benefit in CVT, studies have shown that LP is safe in patients with CVT who have isolated intracranial hypertension. LP is contraindicated in those with large lesions at risk for herniation [14,31]. Performing the LP must be balanced with the need for anticoagulation. European guidelines do not recommend acetazolamide as it has only be evaluated in small non-randomized studies with no clear benefit [16,41]. Steroids have not been shown to be beneficial, with one case-control study showing no difference in rates of death or dependence when compared with control [42]. They may be beneficial in those with an underlying inflammatory disorders such as systemic lupus erythematosus [16]. Although the most recent European Stroke Organization guidelines from 2017 did not comment on these strategies, older guidelines from the European Federation of Neurological Societies in 2010 recommended elevating the head of bead to 30 degrees, hyperventilation to 30-35 mmHg $PaCO_2$, and either hypertonic saline or mannitol to lower ICP based on expert opinion [16,43]. Emergent neurosurgical consultation is recommended in patients with CVT and signs of impending herniation, as small case series and observational data have shown that decompressive surgery can be life-saving, but there are little data to guide patient selection [44].

3.5.3. Anticoagulation

Anticoagulation is the mainstay of ED treatment of CVT with low molecular weight heparin (LMWH) or unfractionated heparin (UFH) to recanalize the occluded vein, prevent thrombus propagation, and treat the underlying prothrombotic state [45]. Intracerebral hemorrhage is not a contraindication to starting anticoagulation. Of the RCTs included in a Cochrane review evaluating anticoagulation for CVT, 25%–49% of patients had intracerebral hemorrhage prior to starting anticoagulation, and no patients developed new intracerebral hemorrhage (95% CI 0%–9%) [45]. It should be noted that RCTs of anticoagulation in CVT excluded patients with end stage renal disease, liver disease with evidence of synthetic dysfunction, recent gastrointestinal bleed, thrombocytopenia, and hypertension with a diastolic pressure greater than 110 mmHg [46]. LMWH may be more effective than UFH, unless the patient is clinically unstable or invasive intervention is planned. One RCT of 66 patients found LMWH reduced mortality (0% with LMWH) compared to UFH (19% mortality), as well as increased likelihood of complete recovery (88% LMWH versus 63% UFH) [47]. A case-control study found greater independence with LMWH versus UFH (92% versus 84%, OR 2.4) [48]. Warfarin or direct oral anticoagulants (DOACs) can be used for long term anticoagulation in CVT [49-51]. A meta-analysis including 6 studies published in 2020 found DOACs were as effective as warfarin in thrombus recanalization and functional outcomes [52]. Of note, in studies evaluating warfarin or DOACs for patients with CVT, LWMH was the initial anticoagulation agent [52]. Decisions concerning the long-term anticoagulation agent and duration of anticoagulation requires consideration of underlying thrombotic risk factors and should be made in conjunction with a specialist in neurology or hematology. The European guidelines recommend a period of 3 to 12 months [16]. The use of systemic thrombolytics has been described, but a systematic review found a risk of serious bleed of 11.5% and a risk of death of 7.7% with systemic thrombolysis. European guidelines currently do not recommend systemic thrombolysis [16,53]. The role of endovascular therapy for CVT has previously been complicated by the non-randomized nature of studies on endovascular therapy, with patients in the intervention group more critically ill [41,54]. However, a recent RCT of endovascular therapy showed no difference in mortality or modified Rankin Score (mRS) of 0–1 when compared with standard of care [55]. In this study, over 80% of patients in both groups had a mRS of 0–2 at discharge, considered a good outcome in most of the ischemic stroke literature, reflecting the overall good prognosis for the disease. However, a subset of patients with CVT may benefit from endovascular therapy, and thus the decision to intervene should be made in conjunction with neurology, neurosurgery, and interventional radiology if endovascular therapy is available [55]. Although overall the prognosis for CVT is favorable, there are potentially devastating consequences to this disease [56]. Clinically stable patients should be admitted for initiation of anticoagulation and neurology and hematology consultation. Patients with evidence of neurologic deterioration or those requiring neurosurgical or interventional radiology procedures should be admitted to an intensive care setting.

3.6. Prognosis

Patients with diagnosed CVT should be admitted for further evaluation of the underlying etiology. Mortality approximates 5%, with risk of permanent disability reaching 20%. The most important factors associated with poor prognosis include malignancy (OR 4.53), coma (OR 4.19), thrombosis of the deep venous system (OR 3.03), any mental status disturbance (OR 2.18), male gender (OR 1.60), and intracerebral hemorrhage (OR 1.42). In the absence of these factors, prognosis is favorable [16,57].

4. Conclusions

CVT is an uncommon cause of headache and stroke that primarily occurs in patients less than 50 years. Women are more commonly affected than men. Thrombus within the cerebral veins can cause increased ICP, cerebral edema, and ischemia. Patients may experience seizures, stroke, and altered mental status, leading to herniation and death. Early recognition is important but difficult as the clinical presentation can mimic more common disease patterns. Imaging studies such as CT venography or MRV should be obtained. Laboratory tests are unlikely to assist in these patients, though D-dimer has been investigated. Treatment includes anticoagulation. Seizures should be treated aggressively. Evidence of herniation requires emergent stabilization with lowering of ICP and emergent neurosurgical consultation.

Declaration of Competing Interest

No conflicts for any author.

Acknowledgements

AT, BL, KRS, and AK conceived the idea for this manuscript and contributed substantially to the writing and editing of the review. This manuscript did not utilize any grants or funding, and it has not been presented in abstract form. This clinical review has not been published, it is not under consideration for publication elsewhere, its publication is approved by all authors and tacitly or explicitly by the responsible authorities where the work was carried out, and that, if accepted, it will not be published elsewhere in the same form, in English or in any other language, including electronically without the written consent of the copyright-holder. This review does not reflect the views or opinions of the U.S. government, Department of Defense, U.S. Army, U.S. Air Force, or SAUSHEC EM Residency Program.

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