

Supplemental Table I. Summary of all clinical questions and related recommendations

Part	Section/Topic	Clinical question	Quality of evidence	Recommendation	Level of recommendation	Additional information
Part I. Diagnostic recommendation	Neuroimaging	In patients suspected of CVT should MR venography versus digital subtraction angiography (DSA) be used to diagnose CVT?	Very low	MR venography can be used as a reliable alternative to DSA for the confirmation of the diagnosis of CVT in patients with suspected CVT	Weak	
		In patient with suspected CVT should CT venography versus digital subtraction angiography be used to diagnose CVT?	Very low	CT venography can be used as a reliable alternative to DSA for the diagnosis of CVT in patients with suspected CVT	Weak	
		In patients suspected of CVT, should CT venography versus MRI and MR venography be used to diagnose CVT?	Very low	CT venography can be used as a reliable alternative to MR venography for confirming the diagnosis of CVT in patients with suspected CVT	Weak	
	D-dimer	In patients suspected of acute cerebral venous thrombosis, should D-dimer be measured before neuroimaging to diagnose CVT?	Low	Measurement of D-dimer before neuroimaging is recommended in patients with suspected CVT, except in those with isolated headache or prolonged duration of symptoms	Weak	
	Screening for thrombophilia	In patients with CVT, does a policy of screening for thrombophilia prevents recurrent venous thrombosis, reduces death and improves functional outcome?	Very low	Thrombophilia screening is not recommended to reduce death, improve functional outcome or prevent recurrent venous thrombosis in patients with CVT	Weak	
	Malignancy screening	In patients with CVT, does screening for an occult malignancy (including haematological malignancies) improves outcome?	Very low	Routine screening for occult malignancy in patients with CVT is not recommended to improve outcome.	Weak	

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Part II. Therapeutic recommendations	Acute anticoagulant treatment	in patients with acute cerebral venous thrombosis, does anticoagulation improve clinical outcome compared to no anticoagulation?	Moderate	Treatment of adult patients with acute CVT with heparin in therapeutic dosage is recommended, including in those with intracerebral haemorrhage at baseline.	Strong	No recommendation can be given on the treatment of children with CVT.
	Type of heparin in acute CVT	In patients with acute cerebral venous thrombosis does low-molecular weight heparin (LMWH) improve clinical outcome compared to unfractionated heparin (UFH)?	Low	Treatment of patients with acute CVT with LMWH instead of UFH is recommended. This does not apply to patients with contraindication for LMWH or situations where fast reversal of the anticoagulant effect is required (e.g. patients who have to undergo neurosurgical intervention).	Weak	
	Thrombolysis and thrombectomy in acute CVT	Does thrombolysis improve clinical outcome compared to anticoagulation in patients with acute cerebral venous thrombosis?	Very low	No recommendation	Uncertain	We suggest not using thrombolysis in acute CVT patients with a pre-treatment low risk of poor outcome

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Part II. Therapeutic recommendations	Duration of anticoagulation	For patients with CVT, does treatment with long term anticoagulation \geq 6 months improve outcome, compared with treatment with short-term anticoagulation (<6months)?	Very low	Using oral anticoagulants (vitamin K antagonists) for a variable period (3-12 months) after CVT is recommended to prevent recurrent CVT and other venous thromboembolic events	Weak	Patients with recurrent venous thrombosis or with an associated prothrombotic condition with a high thrombotic risk may need permanent anticoagulation. We suggest following specific recommendations for the prevention of recurrent venous thromboembolic events in those conditions.
		For patients with previous CVT, does treatment with long term anticoagulation reduce recurrence of venous thrombotic events, compared with treatment with short-term anticoagulation?	Very low			
	New oral anticoagulants	In patients with cerebral venous thrombosis, does treatment with new oral anticoagulants (factor Xa or thrombin inhibitors) improve clinical outcome, reduce major haemorrhagic complications and reduce thrombotic recurrences, compared to conventional anticoagulation (heparin and vitamin K antagonists)	Very low	Use new oral anticoagulants for the treatment of CVT is not recommended, especially during the acute phase.	Weak	
	Therapeutic lumbar puncture	For patients with acute CVT and symptoms or signs of increased intracranial pressure, does therapeutic lumbar puncture (LP) improve outcome, compared with standard treatment?	Very low	No recommendation	Uncertain	Therapeutic LP may be considered in patients with cerebral venous thrombosis and signs of intracranial hypertension, because of a potential beneficial effect on visual loss and/or headache, whenever its safety profile is acceptable.
For patients with previous CVT and symptoms or signs of increased intracranial pressure, does therapeutic LP improve headache or visual disturbances?		Very low	No recommendation			

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Part II. Therapeutic recommendations	Acetazolamide and diuretics	For patients with acute CVT and symptoms or signs of increased intracranial pressure, does treatment with carbonic anhydrase inhibitors improve outcome, compared with standard treatment?	Low	We suggest not using acetazolamide in patients with acute CVT to prevent death or to improve functional outcome	Weak	In isolated intracranial hypertension secondary to CVT, causing severe headaches or threatening vision, acetazolamide may be considered if its safety profile is acceptable
		For patients with previous CVT and symptoms or signs of increased intracranial pressure, does treatment with carbonic anhydrase inhibitors improve headache or visual disturbances?	Very low	No recommendation		
	Steroids	For patients with acute CVT and symptoms or signs of increased intracranial pressure, does treatment with steroids improve outcome, compared with standard treatment?	Low	Steroids in patients with acute CVT are not recommended to prevent death or to improve functional outcome	Weak	We suggest to use steroids in patients with acute CVT and Behçet's disease and other inflammatory diseases to improve outcome
	Shunt	For patients with acute or recent CVT and parenchymal lesion(s) with impending herniation does shunting (without other surgical treatment) improve outcome, compared with standard treatment?	Very low	Routine shunting (without other surgical treatment) in patients with acute CVT and impending brain herniation due to parenchymal lesions is not recommended to prevent death	Weak	
		For patients with acute or recent CVT and hydrocephalus does shunting (without other surgical treatment) improve outcome, compared with standard treatment?	Very low	No recommendation	Uncertain	

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Part II. Therapeutic recommendations	Decompressive Surgery	For patients with acute CVT and parenchymal lesion(s) with impending herniation, does decompressive surgery (hemispherectomy or hematoma evacuation) improve outcome, compared with conservative treatment?	Low	Decompressive surgery for patients with acute CVT and parenchymal lesion(s) with impending herniation is recommended to prevent death	Strong	
	Prevention of seizures and anti-epileptic drugs (AEDs)	In patients with acute or recent CVT do antiepileptic drugs prevent acute and recent post-CVT seizures, compared with no antiepileptic treatment?	Low	Antiepileptic drugs in patients with acute CVT with supratentorial lesions and seizures are recommended to prevent early recurrent seizures	Weak	
		In patients with acute or recent CVT do antiepileptic drugs prevent remote post-CVT seizures, compared with no antiepileptic treatment?	Very low	No recommendation	Uncertain	
	Cerebral Venous Thrombosis during Pregnancy	In pregnant and puerperal women with CVT, does the use of anticoagulant therapy improve the outcome without causing major risks to mother and foetus?	Low	Therapy with subcutaneous LMWH in pregnant and puerperal patients with acute CVT is recommended.	Weak	
	Contraceptive use after cerebral venous thrombosis	In women with prior CVT does used of combined oral hormonal contraception increase the risk of recurrent CVT or other VTE?	Very low	Women in fertile age and prior CVT should be informed about the risks of combined hormonal contraception and advised against its use.	Weak	

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Part II. Therapeutic recommendations	Safety of pregnancy following CVT	In females with previous history of CVT is a policy of not contraindicating future pregnancies associated with recurrence of CVT or other venous thromboembolic events (VTEs) (lower or upper limb deep vein thrombosis, pulmonary embolism, abdominal or pelvic venous thrombosis) and unfavourable pregnancy outcome?	Low	For all women with prior history of CVT, we suggest to inform on the absolute and relative risks of venous thrombotic events and abortion during subsequent pregnancies and to not contraindicate future pregnancies based only in the past history of CVT	Weak	
		For pregnant women with previous history of CVT, does prophylaxis with antithrombotic drugs reduce the risk of thromboembolic events or affect pregnancy outcome?	Low	Prophylaxis with sc LMWH during pregnancy/puerperium is recommended for pregnant women with previous history of CVT and without contraindication for prophylaxis or indication for anticoagulation in therapeutic dosage.	Weak	