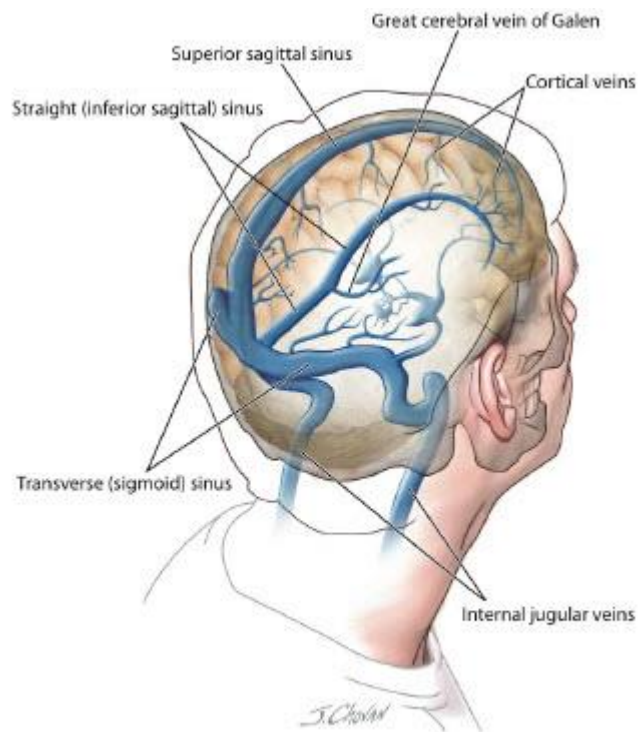


Cerebral vein and cerebral venous sinus thrombosis

Cerebral vein and cerebral venous sinus thromboses are blood clots that form in the veins that drain the blood from the brain called the sinuses and cerebral veins. They can lead to severe headaches, confusion, and stroke-like symptoms. They may lead to bleeding into the surrounding brain tissues. The clots can be triggered by infections of the ear, face, or neck, by medications containing estrogen, pregnancy, or dehydration. They can also be caused by clotting disorders. Sometimes the cause is unknown.

The diagnosis is uncommon. It can take a special MRI or CT scan (called MR venogram or CT venogram) to make the diagnosis.

The first line of treatment is the blood thinner medication heparin or enoxaparin. After the initial treatment with heparin the patient will start taking warfarin which is another blood thinning medication. The length of treatment with warfarin depends on what caused the clot. If the cause was a temporary trigger such as an infection, pregnancy or use of medications containing estrogen, then treatment would last 3-6 months. If the cause was not identified or if the patient has is a strong clotting disorder, then treatment may last 6-12 months.



Other names for cerebral vein and cerebral venous sinus thrombosis

- Cerebral venous thrombosis (CVT)
- Cerebral vein thrombosis
- Cerebral venous and sinus thrombosis,
- Cerebral venous sinus thrombosis (CVST)
- Cerebral sinovenous thrombosis (CSVST)
- Cerebral vein and dural sinus thrombosis
- Sinus and cerebral vein thrombosis

How common is cerebral vein and cerebral venous sinus thrombosis?

Cerebral vein and cerebral venous sinus thrombosis is an uncommon type of clot. Only about 5,000 people in the U.S. are diagnosed with it per year (compared to nearly a million people every year with deep vein thrombosis and pulmonary embolism, and about 800,000 people every year with strokes).

Who develops it? What are the causes?

The most common causes for cerebral vein and cerebral venous sinus thrombosis in adults are:

- Medications that contain estrogen such as birth control pills, patch or ring and estrogen replacement therapy.
- Elevated levels of estrogen in pregnancy and post-pregnancy period.
- Certain medications, for example tamoxifen and chemotherapy drugs.
- Cancer.
- Inherited clotting disorders such as factor V Leiden, prothrombin 20210 mutation, and deficiency of protein C, S or antithrombin.
- Acquired clotting disorders such as antiphospholipid antibodies.

Sometimes, no obvious cause is identified, in spite of an extensive laboratory work-up.

How does cerebral vein and cerebral venous sinus thrombosis form?

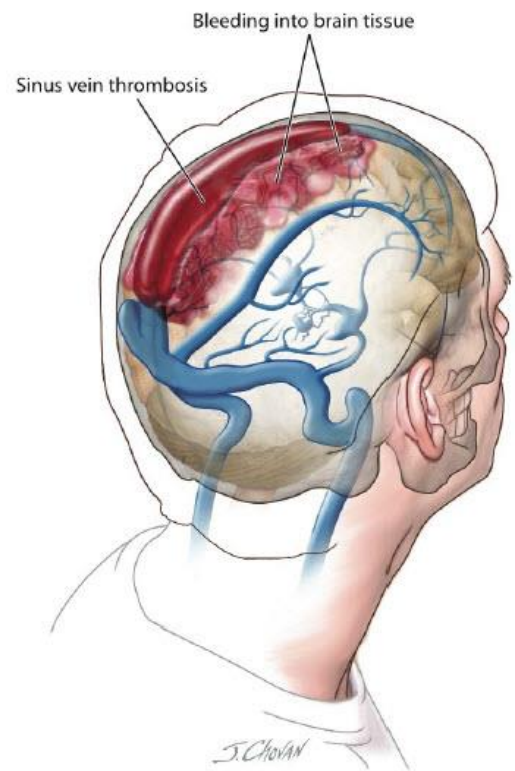
Normally, blood is transported through arteries into the brain, where it delivers oxygen and nutrients. Once the blood has done its job, it collects into small veins (=cerebral veins) that drain into large veins, called sinus veins. The sinus veins lead to the jugular veins in the neck, which carry the blood back to the heart (see image above).

For clarification: These sinus veins have nothing in common (except for the name “sinus”) with the sinuses of the face on both sides of the nose and above the eyes which can get infected, leading to sinusitis.

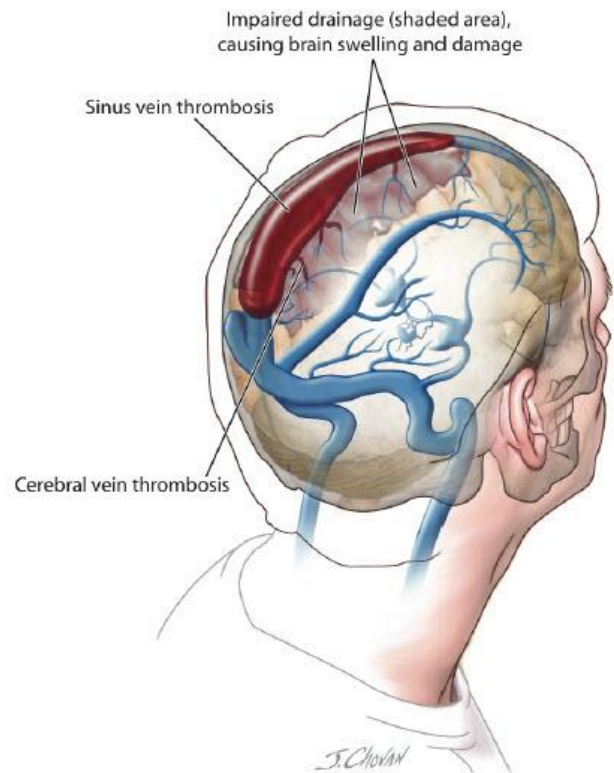
How does the clot affect the brain?

The clot blocks the blood flow in the veins which leads to a backup of blood flow and increasing blood pressure in the blood vessels just before the obstruction (see image). This is like water in front of a dam.

The increased pressure leads to swelling of part of the brain, which results in headaches. The pressure can damage the brain tissue, leading to a stroke, causing symptoms such as numbness or weakness in the arm or legs and trouble with speaking, seeing and walking. This type of stroke is sometimes called a venous infarction.



The increased pressure can also lead to a rupture of a blood vessel and bleeding into the brain. In medical terms this is called “cerebral hemorrhage”. (see image). It is like water in a reservoir overflowing into the surroundings or like a ruptured dam. Other names for this condition are “venous hemorrhagic infarction” or “venous hemorrhagic stroke”. Cerebral hemorrhage can lead to further damage of brain tissue. About one-third of patients with cerebral vein and cerebral venous sinus thrombosis have such bleeding.



What are the symptoms?

Symptoms from sinus and cerebral vein clots depend on the location and extension of the clot and vary from patient to patient. The most common symptom is a severe headache, often the worst headache that a patient has ever had. It can be of sudden onset, develop over a few hours, or develop over a few days. Nausea and vomiting may occur, as may blurred vision.

Other symptoms that may also occur include:

- Seizures
- Speech impairment
- Left body or right numbness
- Numbness or weakness of an arm, a leg, or both
- Confusion
- A decreased level of alertness

Symptoms may be exactly what people think of as occurring in a stroke. A very extensive blood clot may lead to loss of consciousness and death.

How is it diagnosed?

The diagnosis of cerebral vein and cerebral venous sinus thrombosis can be easily missed because it requires the correct imaging X-ray test. The routine CT or MRI tests that are often done to evaluate a stroke or brain bleeds are often normal in cases of cerebral vein and cerebral venous sinus thrombosis. Also, a plain X-ray of the head or skull is not helpful. **The appropriate test to do is an MRI venogram (=MRV) or CT venogram (=CTV).**

To avoid missing the diagnosis the health care professional who evaluates the patient with severe headache or stroke-like symptoms needs to think about cerebral vein and cerebral venous sinus thrombosis and order the required imaging tests described above. The health care professional should have an increased level of suspicion that a patient may have a sinus or cerebral vein thrombosis if the patient has one of the following:

1. The worst headache ever
2. Risk factors for blood clots, such as being on estrogens or progestin contraceptives, being pregnant or having delivered in the preceding 3 months
3. Personal or family history of blood clots
4. Known clotting disorder

Clotting disorder work-up

In unexplained cerebral vein and cerebral venous sinus thrombosis, the patient will need to undergo a series of tests to look for a strong clotting disorder. This may influence the length of treatment with blood thinners.

What is the treatment for cerebral vein and cerebral venous sinus thrombosis?

Patients with an acute clot are admitted to the hospital. If symptoms are severe, patients will be admitted to a stroke or intensive care unit. The immediate treatment consists of giving blood thinning medications (anticoagulants). In the first few days these are either heparin into the veins (intravenously), or

injections of low molecular weight heparin (Enoxaparin, Dalteparin, Tinazparin, Lovenox®, Framin®, Innohep®) under the skin (subcutaneously). The purpose of giving blood thinners is to prevent the existing clots from getting bigger and prevent new clots from forming. The body's own clot-dissolving system then slowly, over weeks and months, works on dissolving the existing clots. Even patients that have bleeding into the brain need treatment with blood thinners in order to prevent new clots from forming. In these cases the doctors must pay very close attention so the bleed does not get worse.

Once the patient has been stable for a few days, an oral blood thinner - warfarin (brand name Coumadin®) - is started. A key question is how long a patient needs to be on warfarin. The American Stroke Association published clear guidelines for treatment in a statement for healthcare professionals in 2011 (see reference 1 at the end).

- **If the clot was associated with a temporary risk factor**, such as an infection or trauma, a period of 3-6 months is typically sufficient.
- **Patients who have strong clotting disorders** have a high risk of developing another clot and need long-term (often lifelong) treatment with warfarin.
- **Patients with an unprovoked clot** (this means that the cause was not identified) receive warfarin treatment for a period of 6-12 months.

Finding leftover clots (scar tissue, recanalized clots) after a few months of treatment with blood thinners typically **does not** influence the decision regarding how long to treat with blood thinners.

What is my risk to develop another clot after treatment is over?

The risk of having another sinus or cerebral vein thrombosis, once a patient has stopped warfarin is relatively low in a person who does not have a strong clotting disorder and their whose clot was unprovoked.

Having sinus or cerebral vein thrombosis increases the risk for developing other vein clots, mostly deep vein thrombosis (DVT) of the legs and pulmonary

embolism (PE). For this reason is it very important that patients who stop warfarin know the symptoms of DVT and PE and be on the lookout for them, to seek medical attention early if such symptoms occur.

Symptoms of Deep Vein Thrombosis (DVT)

- Swelling of the leg or along a vein in the leg
- Pain or tenderness in the leg, which you may feel only when standing or walking
- Increased warmth in the area of the leg that's swollen or painful
- Red or discolored skin on the leg

Symptoms of Pulmonary Embolism (PE)

Shortness of breath

- Rapid heart rate
- Unexplained cough and/or coughing up blood
- Chest pain, especially with deep breathing

What can I expect after treatment is over?

Once a decision is made to stop warfarin, a baseline imaging study (MRV or CTV) is often done. The purpose of this is to have a baseline or reference study, in case new symptoms come up in the future. If one has a baseline study for comparison, it is easier to tell new clot from a previous clot. In approximately 80 out of 100 patients (80%) the clots partially or completely dissolve and the veins become clear again. 20 out of 100 patients (20%) will have chronic occlusion (partial blockage) of the sinus or cerebral veins where the clot had occurred. Clinically this does not seem to matter and does not mean that the patient will have chronic long lasting symptoms. This is another reason why routine follow-up MRV or CTV X-ray studies are not needed.

What is my long term outlook?

In the acute setting some patients with extensive clot or associated bleed may suffer disabling symptoms or die. However, the majority of patients recover completely. Almost 80% of patients fully recover, but it may take several weeks

or months to get back to normal. Headaches and seizures may persist for some time.

What else do I need to know?

If you have had a sinus and/or cerebral vein thrombosis these are the important points to know:

- What caused your clot and if you have any risk factors for developing sinus and/or cerebral vein thrombosis. If you do not have a known cause or risk factor did you have appropriate laboratory work-up to look for a clotting disorder?
- How long you should be on warfarin. Make sure your health care professional is aware of the published management consensus statement (Listed as Reference 1 at the end of this document). You may want to print it out and provide it to them.
- You must avoid estrogen therapy or progestin pill or injections in the future. Mirena® IUD is probably safe.
- It is OK for most women to become pregnant in the future. Blood thinner therapy should be considered during pregnancy and for several weeks (6-12 weeks) after delivery.
- You have an increased risk for developing other blood clots such as deep vein thrombosis (DVT) and pulmonary embolism (PE).
 - Take steps to reduce your risk for developing DVT and PE. The steps include: maintaining a normal weight, stopping smoking and being physically active.
 - Know the symptoms of DVT and PE (on page 7) so that you can seek treatment as soon as possible if these develop.
 - Let your healthcare providers know of your health history if you are having major surgery, admitted to a hospital or if you become pregnant. You will need blood thinning therapy to prevent developing a DVT or PE during these high risk situations.

What do my family members need to know?

Your siblings, children and parents need to know they have a slightly higher risk for blood clots, because you had a clot. They do not need screening (with MRV or CTV scan and blood work) unless you were found to have a strong inherited clotting disorder. In that case it might be appropriate to consider testing them for the same clotting disorder. This is a complex issue that doctors will make for individual patients.

References and Guidelines

1. Saposnik G et al. *Diagnosis and management of cerebral venous thrombosis*. A statement for healthcare professionals from the American Heart Association/ American Stroke Association. *Stroke* 2011;42 :1158-92
2. Bruno M et al. *Venous Thromboembolic Events After Cerebral Vein Thrombosis*. *Stroke*. 2010;41:1901-1906.

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Reviewer: Eric Adelman MD, Margaret McDermott MD

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