Neurology

Cerebral venous sinus thrombosis

patient information

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What is cerebral venous sinus thrombosis?

Blood carries oxygen, nutrients and waste through your body via blood vessels. Arteries transport oxygen-rich blood and nutrients to all parts of the body, and the veins drain blood that is low in oxygen and contains waste products. With thrombosis, blood clots form in the veins (venous thrombosis) or in the arteries (arterial thrombosis). These blood clots block the blood vessel, which causes a disturbance in the blood flow.

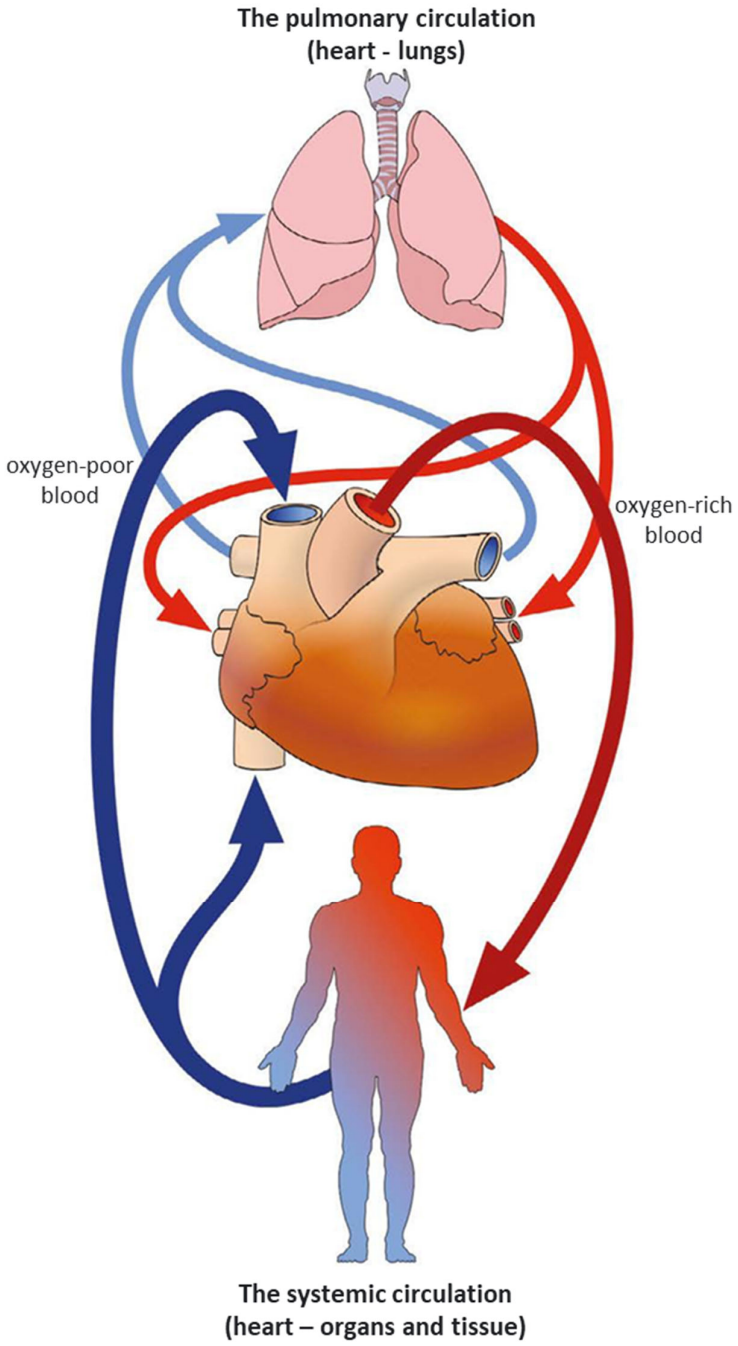
Venous thrombosis can occur in various parts of the body, such as in the deep veins of the legs. In that case, we call this a deep vein thrombosis. If the blockage of the vein occurs in the brain, this is called cerebral venous sinus thrombosis. Cerebral venous sinus thrombosis is a special type of stroke. Arteries in the brain can also get blocked, which causes an acute cerebral infarction. In this flyer, we discuss cerebral venous sinus thrombosis: blood clots in the draining vessels of the brain, that drain waste products and blood low in oxygen from the brain.

When your doctor has diagnosed you with cerebral venous sinus thrombosis, you probably have many questions. What are the causes and the consequences? And which treatments can you get? With this flyer, we would like to give you more information. This flyer is assembled by physicians from the department of Neurology of Amsterdam UMC in The Netherlands, a former patient who has suffered this disease, the Dutch Heart Foundation and the Dutch Thrombosis Foundation.

How does a healthy heart and circulatory system work?

What does the circulatory system look like?

Our body needs oxygen and nutrients to be able to function. These substances reach the muscles and organs in the body through blood. In the lungs, the blood receives oxygen. The left side of the heart pumps this oxygen-rich blood through the arteries to all parts of the body. In the capillaries (the smallest branches of the arteries) the blood delivers oxygen and nutrients to the organs and absorbs carbon dioxide and waste products. The waste products are removed from our body through the urine and the stool. The oxygen-poor, carbon dioxide-rich blood flows through the veins back to the right side of the heart and to the lungs. In the lungs, the carbon dioxide leaves our body and the blood takes up oxygen again. The circulatory system ensures that our body absorbs the right substances in the right places and that these substances will be drained again.



Blood clotting

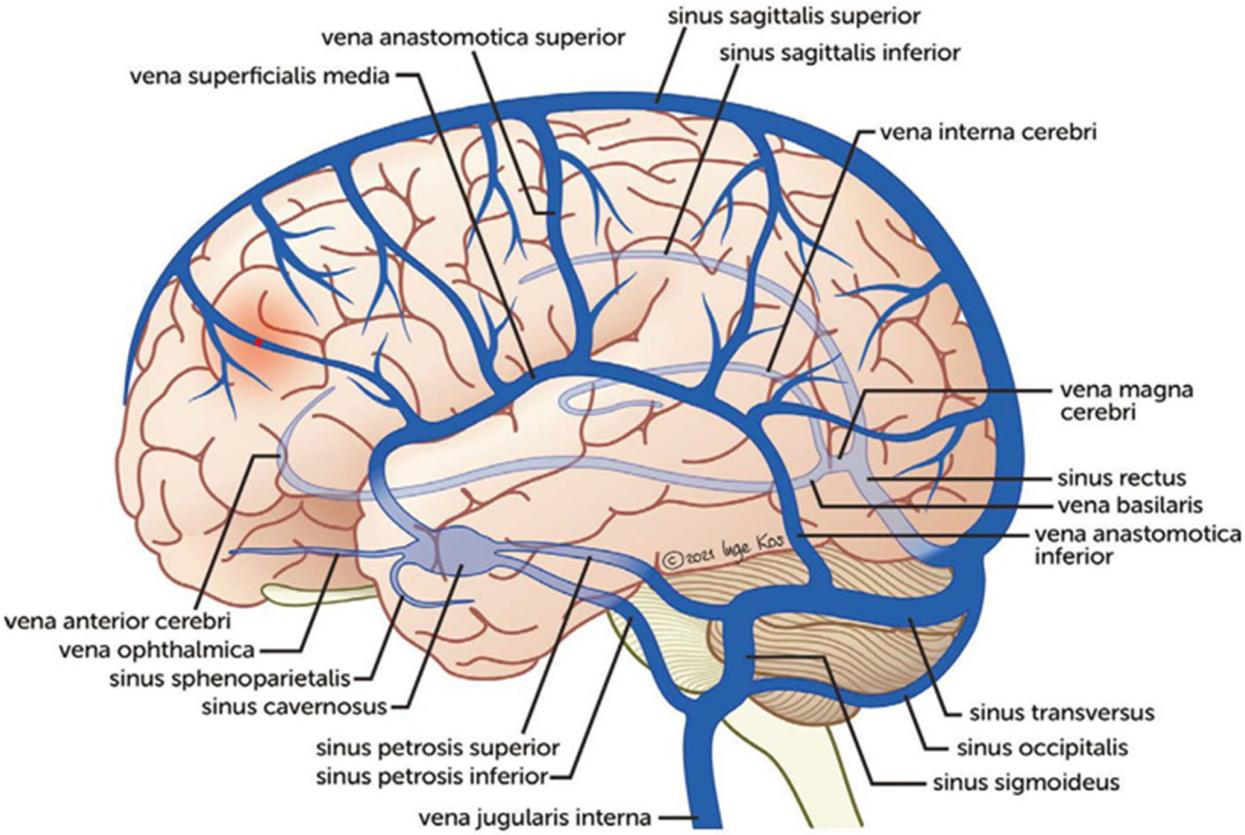
In order for your body to function properly, the blood must keep flowing. If you have a wound, the blood in that spot should clot quickly. Otherwise you will lose too much blood. The coagulation system ensures that the blood clots at the right time in the right place. A scab is formed in this way. It is a kind of natural plaster. Platelets and clotting proteins in the blood, together with the blood vessel wall, ensure blood clotting. If you have a wound, the clot should not continue to grow indefinitely. And the clot also has to be broken off in time. Various systems in the body take care of this. They also ensure that your blood remains thin enough to flow properly.

What is thrombosis?

If your blood clots when there is no wound, or if it continues to clot when the wound is already closed, blood clots form on the inside of the blood vessels when they shouldn't. When a vein becomes blocked by a blood clot, we call it venous thrombosis. Venous means 'of a vein'. Thrombosis of a vein in the leg, deep vein thrombosis, is most common. Venous thrombosis can also develop in other places in the body such as the arm, pelvis or brain.

What is cerebral venous sinus thrombosis?

Cerebral venous sinus thrombosis is thrombosis in a vein of the brain. It is a brain disorder that mainly affects people between the ages of 20 and 50. Cerebral venous sinus thrombosis also occurs in children (particularly shortly after birth). Three quarters of adults with cerebral venous sinus thrombosis are female. It is a rare condition. Cerebral venous sinus thrombosis is the result of a blood clot in the veins of the brain, which prevents the blood from draining properly. The drainage of the cerebrospinal fluid through the veins is often also disturbed. In half of the people, the brain is damaged, for example because the disturbed drainage of the blood leads to an accumulation of fluid (edema) in part of the brain. We call this a venous stroke. Small blood vessels in the brain can also rupture, resulting in a brain hemorrhage. The severity of cerebral venous sinus thrombosis varies greatly from person to person.



Which complaints can cerebral venous sinus thrombosis cause?

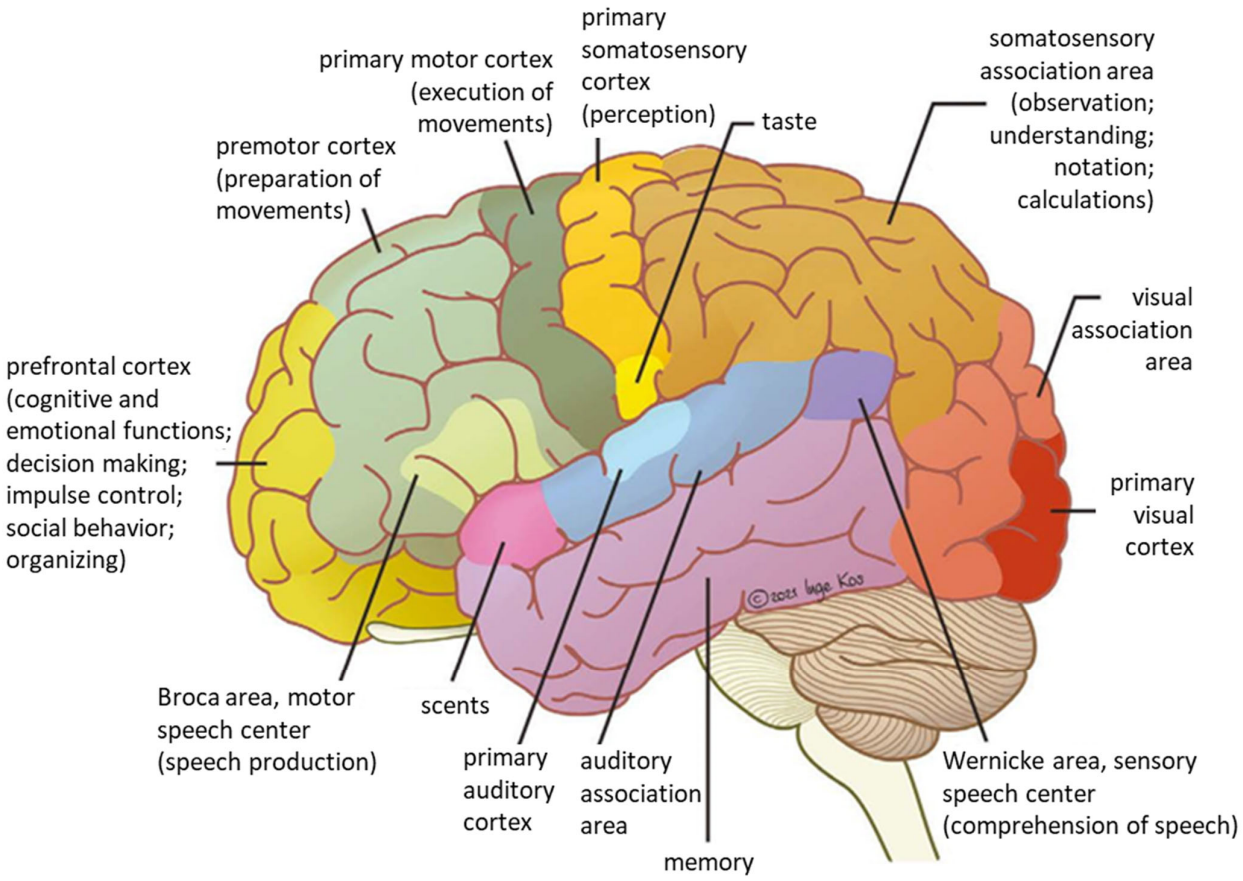
Severe headache, blurred vision, nausea, and throwing up

Severe headache is the most common complaint in cerebral venous sinus thrombosis. Nine out of 10 people with cerebral venous sinus thrombosis have this severe headache. Sometimes people develop a blurred vision. Nausea and vomiting are also common. These complaints are the result of increased pressure in the head as a result of the disturbed drainage of blood and cerebrospinal fluid.

Neurological deficits

In addition, 4 out of 10 people suffer from neurological deficits, such as paralysis of one side of the body or face (e.g. drooping corner of the mouth), slurred speech, a disturbance of vision, and sometimes a lowered consciousness. Neurological deficits occur when part of the brain tissue cannot function properly, for example due to a venous stroke or bleeding.

The location and size of the affected area determine the consequences. A problem in the left half of the brain, for example, can cause a speech disorder (aphasia), paralysis and/or sensory problems on the right side of the body. Problems in the right hemisphere cause problems in the left side of the body. However, the division into the left and right hemispheres of the brain is not always strict.



Epileptic seizures

Cerebral venous sinus thrombosis can also provoke an epileptic seizure. Epileptic seizures are caused by a sudden disruption of the electrical impulse transmission between certain brain cells. Seizures occur in 4 out of 10 people during the first weeks of cerebral venous sinus thrombosis.

How does cerebral venous sinus thrombosis occur?

The causes of thrombosis are very diverse. It can be caused by:

- changes in your blood composition
- obstructions in your blood flow
- damage to the blood vessel wall

Below you can read more about the causes of the change in the composition of the blood. The composition of your blood is not always the same, this can change due to, for example:

Sickness

If you are sick, the composition of your blood may change. For example, your blood may contain many infectious proteins or waste products. Sometimes the blood clots more easily and the risk of thrombosis increases. This is especially the case if there is an inflammation, such as an infection in the region of the head or neck. But cancer or hormonal disorders can also change the composition of the blood.

Medication

Some medicines can also make your blood clot more easily, for example when treating cancer with chemotherapy or hormone therapy.

Female hormones

The female sex hormone estrogen increases the risk of thrombosis due to an altered blood composition. Estrogen is present in your body in a higher concentration during pregnancy, IVF treatments, hormone treatments around the menopause, and when using the contraceptive pill. Age plays a role here: the older you are when you use the contraceptive pill, the greater the risk of thrombosis. The risk increases especially if you are over 40. If you are overweight and use the contraceptive pill, you also have a higher risk of thrombosis.

Hereditary factors

Due to hereditary factors, your blood can be composed in such a way that you have a greater chance of thrombosis. Common hereditary defects are a deficiency of antithrombin, protein C and protein S. These proteins are produced in your liver and inhibit blood clotting. If you have a deficiency in one of these proteins, the risk of thrombosis is 5 to 10 times higher than average. Other common hereditary defects are factor V Leiden mutation and prothrombin mutation. With these abnormalities, the effect of certain coagulation proteins is increased, with the risk of thrombosis being 3 to 5 times greater than average.

Genetic testing

Currently, most people with thrombosis are not routinely checked for possible hereditary

causes. For the treatment of the thrombosis, it usually does not matter whether there is a hereditary cause. Only if a doctor suspects that the outcome of a genetic test does influence the treatment, your doctor can decide to conduct further research into hereditary causes of thrombosis. For example, if thrombosis is remarkably common in your family. If you have a hereditary factor, this can sometimes mean that anticoagulants are prescribed as a precaution in high-risk situations for thrombosis. If you know that you have a hereditary risk of thrombosis, it is possible that it will be more difficult to get some types of insurance policies, for example with a life insurance policy.

How is cerebral venous sinus thrombosis diagnosed?

Your doctor suspects that you have cerebral venous sinus thrombosis based on your symptoms. To further investigate this, you will receive a CT or MRI scan of the brain with contrast of the blood vessels.

Brain CT scan

The CT scan works with X-rays. Just before the scan, you will be given contrast fluid through an injection in your arm. The contrast medium makes the flowing blood in the blood vessels easier to see on the scan. During the scan, the examination table on which you lie will slide through the CT scanner. A series of pictures of your brain and blood vessels will be taken. The pictures show contrast in the blood vessels. If there is a clot, there is a recess in the blood flow at the site of the clot.

Brain MRI scan

The MRI scan of the brain works with magnetic fields and radio waves. No X-rays are used. Sometimes contrast fluid is administered through an IV, but not always. With this scan, the doctor gets information about your blood vessels as well as detailed information about your brain tissue.

How is cerebral venous sinus thrombosis treated?

Blood thinners

If your doctor has diagnosed you with cerebral venous sinus thrombosis, you will usually be given anticoagulants (blood thinners) right away. The three most commonly prescribed anticoagulants are listed below:

Heparin

Heparin is either given intravenously by means of a pump or injected under the skin. The latter form of heparin is called low-molecular-weight heparins, abbreviated to LMWHs. These LMWHs have a direct anticoagulant effect. Your nurse or doctor will teach you how to use LMWHs yourself.

Coumarin derivates

This anticoagulant medicine is given to you through a pill. In most cases, you will be given a drug derived from coumarin, such as warfarin, acenocoumarol or phenprocoumon. Coumarin is a so-called vitamin K inhibitor. Vitamin K is involved in the clotting of the blood: with less

vitamin K the blood clots less quickly. It takes a few days for a coumarin to work, so in the beginning you will be given the pills at the same time as heparin. If you take a coumarin, you will be checked by a thrombosis service. An employee of this service uses a blood test to determine the INR value of your blood. The INR value is a measure of the time it takes for your blood to clot. Things like food with a lot of vitamin K, alcohol and illness affect the INR value. This blood test is used to determine how many pills you should take per day. This can differ per day. You will always receive a new schedule, so that you can see every day whether you should take one or more tablets. With a short instruction from the thrombosis service, you can often also learn how to test the INR value of your blood at home, so that you do not have to go to the thrombosis service every time.

DOAC
The abbreviation DOAC stands for direct oral anticoagulants. You receive these anticoagulants in tablet or capsule form. These are the drugs: dabigatran, rivaroxaban, edoxaban and apixaban. With these drugs, the blood does not have to be checked by the thrombosis service as with coumarins. DOACs are usually prescribed for deep vein thrombosis and pulmonary embolism. Recent research shows that these drugs can also be used in most people with cerebral venous sinus thrombosis.

How long should you take the anticoagulant drugs?
Your doctor will determine how long you should take the anticoagulant medicines. In cerebral venous sinus thrombosis without apparent cause, this is usually 3 to 12 months. In some cases, you have to take anticoagulant medicines for life, for example if you have previously had a thrombosis.

Epilepsy drugs

If you have had one or more epileptic seizures, you will be given medication for this. Most often, epileptic seizures occur only in the initial stages of cerebral venous sinus thrombosis. The duration of treatment with epilepsy drugs varies and is on average 3 to 6 months. If seizures continue to occur after the acute phase, you will need to take these medicines for a long time and sometimes for life.

Rehabilitation

If there are neurological deficits, you will usually get a referral for a rehabilitation program in a rehabilitation center. The following specialists are involved in this:

- rehabilitation doctor
- physiotherapist
- occupational therapist
- speech therapist
- (neuro)psychologist
- social worker
- occupational health expert

The rehabilitation is tailored to your own situation and therefore looks different for everyone.

What are the consequences of cerebral venous sinus thrombosis?

The first period after a cerebral venous sinus thrombosis is a violent and uncertain phase. Fortunately, the prognosis has improved in recent years due to better recognition of the disease, improved imaging techniques, and more effective treatments. Nevertheless, cerebral venous sinus thrombosis is still a serious brain disorder of which 5-10% of people die from.

The long-term consequences differ per person and depend, among other things, on whether and how much damage the brain tissue has suffered, for example as a result of a venous stroke or bleeding. Damage to brain tissue can manifest in physical problems, disturbances in thinking, and/or changes in emotions or behavior. The most important recovery occurs in the first half year after cerebral venous sinus thrombosis. In the long term, more than 3 in 5 people recover without symptoms. More than 1 in 5 people can perform all the usual work and activities again, but less and/or more slowly than before. About 8 in 100 people experience limitations in daily functioning, but are independent. Finally, about 6 in 100 people are not (or no longer) independent after a cerebral venous sinus thrombosis.

The following complaints are common after a cerebral venous sinus thrombosis:

- Headache

Headache is the most common complaint after cerebral venous sinus thrombosis. The headaches can get worse by fatigue or stress. Sleeping, resting or taking a break provides relief. The headache usually goes away on its own after a few days or weeks. If the headache does not diminish or gets worse, it is advised to see your doctor. If a headache remains chronically present, it is usually less severe than the headache in the initial phase of the disease.

- Epilepsy

Usually epileptic seizures occur only in the initial phase of cerebral venous sinus thrombosis, but about 1 in 10 people continue to have seizures after cerebral venous sinus thrombosis. Further on in this folder you will find sources of information about epilepsy and some lifestyle advices.

- Neurological deficits

In most people, neurological deficits only occur in the initial phase of cerebral venous sinus thrombosis. Recovery from these deficits varies greatly from person to person and depends on the location, size, and type of damage to the brain tissue. If there are persistent neurological deficits, you will usually follow a rehabilitation program in a rehabilitation center.

- Clumsiness

Perhaps you are temporarily less handy than usual, you are unsteady on your legs, bump into the furniture, or let things fall out of your hands. If this is the case, take it easy. Your brain is the control center for your entire body. It needs to process the information coming in through your eyes, ears, and other senses and it needs to send the right signals to the right muscles to make sure you can do something. Give yourself more time to do things.

- Trouble seeing

You may have difficulty focusing your gaze and/or you may be hypersensitive for bright light. This usually passes quickly. Wearing sunglasses can help with sensitivity to bright light, including indoors. If you suffer from blurred vision or impaired vision, it is important that you contact your specialist immediately.

- Fatigue

In the beginning, even the smallest effort can be very tiring. You notice that you need more sleep than usual. If necessary, go to bed during the first few weeks when you feel tired, even if it is in the middle of the day. If the fatigue symptoms persist, do not hesitate to take rest breaks during the day. Consult an occupational therapist for your daily schedule and energy consumption.

- Concentration problems

No one can concentrate when they're tired, so it is not strange if you have trouble with your ability to concentrate. Maybe you like to read the newspaper, but you can't concentrate enough. In that case, it is helpful to read in small chunks with breaks in between. This also applies to all other activities that require concentration. Issues that require your full attention are better handled when you have the most energy.

- Slowness

You may find that, for example, you have trouble following conversations and/or follow instructions. It may take longer to complete a task. Ask others to repeat what they have said or ask questions of your own. Give yourself the extra time you need to complete tasks. Avoid situations where you will be pressured to get things done quickly.

- Memory problems

Remembering appointments and tasks may be less easy than before. Use a calendar and/or a planning board to keep track of your appointments. For important appointments, it helps to record the conversation and listen to it at a later time. Please mention that you will be recording the conversation to the person you are talking to. In addition, it is advisable to bring someone with you to important appointments.

- Irritability

You may be more easily irritated by situations that did not bother you before. This can be difficult for you and those around you. The brain is the control center of your emotions and regulates them. If this center is affected, you have less control over your emotions. There are several ways to deal with this. Leaving the room or walking away from an irritating situation is a good solution for some people. Another option to counteract the irritated feeling is to seek distraction by, for example, walking or cycling (if the fatigue allows it). The irritability is worse when you are tired, so make sure to rest and do relaxation exercises if necessary.

- Sensitivity to sound

If you don't want to see something, you just close your eyes. It is much more difficult to close your ears. Normally, your brain prevents you from noticing all kinds of disturbing environmental sounds. It is possible that your brain (temporarily) does not filter out these

sounds well. You then notice that sounds disturb you. Explain this to family and friends and ask them to not make unnecessary noises. Sound-canceling headphones or earplugs can help to muffle ambient noise.

Tip: The consequences of cerebral venous sinus thrombosis are not always visible. It is sometimes difficult for others to understand what is bothering you. Therefore, give this folder to your employer, teachers, family and friends.

Return to work

The best time to return to work depends on how you feel and the type of work you do. Consult with your company doctor to discuss this.
Depending on your situation, a neuropsychological examination may be necessary to gain insight into any cognitive impairments. Such an examination is possible at a rehabilitation center. You need a referral from your GP for this.

Check-ups

As long as you are receiving anticoagulants, you will remain under the supervision of a specialist and possibly the thrombosis service. If you are monitored by a thrombosis service, the coagulation values of your blood will be checked regularly and the dosage of the anticoagulant medication will be determined. You also go to your specialist for a check-up. How often you need to go depends on your complaints. Sometimes a referral to a rehabilitation doctor is necessary.

Risk of recurrence

Once you have had a cerebral venous sinus thrombosis, you have an increased risk of having another thrombosis in the future. About a third of these recurrences are cerebral venous sinus thrombosis. Of all recurrences, the majority occur within the first year after the cerebral venous sinus thrombosis. Do you get the same symptoms as with your previous cerebral venous sinus thrombosis? Call your doctor immediately.

Living with cerebral venous sinus thrombosis

Below we list some tips for you:

Medication

- Always take your anticoagulant medicines according to the instructions of your thrombosis service or doctor and do not miss the monitoring appointments with the thrombosis service or doctor.
- Anticoagulant drugs inhibit the clotting of the blood. This makes you more likely to bruise or bleed. You are less able to staunch wounds. Contact the thrombosis service or your doctor if you experience unexplained bruising or unusual bleeding such as a nosebleed, a wound that continues to bleed, heavy or unexpected menstruation, blood in the urine or stool, coughing or vomiting blood or something that looks like coffee grounds.
- If you hit your head resulting in a bruise or wound, it is advisable to contact the thrombosis service or your doctor.

- Avoid injury-prone sports and contact sports, such as football, rugby, hockey, martial arts, skiing and horseback riding. This way you limit the risk of bleeding.
- Are you sick? Flu or diarrhea can interfere with how your anticoagulant medicines work. Therefore, always contact the thrombosis service or your GP if you are ill.
- Are you undergoing surgery or do you need to have a tooth extracted? In that case, tell your specialist or dentist that you have (had) a thrombosis and that you use(d) anticoagulant medicines.
- Do you have holiday plans and are you currently under the control of the thrombosis service? Inform them about your holiday. They can then adjust the dosing period of the anticoagulant medicines and provide you with an international statement stating that you are being treated for thrombosis.
- If you wish to have children, please contact your doctor. Certain anticoagulants and medicines for epilepsy can be harmful to your unborn child.
- Request a medicine passport from your pharmacy.

Other advice

- If you smoke, stop smoking.
- Live healthy and get enough exercise, eat healthy and varied. You can consult a lifestyle coach to help you with this.
- When your symptoms have disappeared, you can resume your daily activities. It differs for everyone when that is. Listen carefully to your body.
- Women who use contraceptive pills are advised to stop these at the same time as when the anticoagulant medicines are stopped. It is not necessary to stop immediately when the cerebral venous sinus thrombosis is diagnosed. You can safely take the contraceptive pills during the period that you are taking anticoagulants, without an increased risk of thrombosis. This may be desirable to reduce menstrual blood loss with concomitant use of anticoagulant drugs. Alternative forms of contraception are, for example, an IUD.
- Women who wish to have children are advised to contact their general practitioner prior to their pregnancy for a referral to the outpatient clinic of vascular medicine or gynaecology. Women with a history of cerebral venous sinus thrombosis who become pregnant are usually treated with anticoagulant drugs (low molecular weight heparin) to prevent thrombosis during and the first weeks after pregnancy.

Advice for epilepsy

- Take your epilepsy medicines on time.
- Do not go swimming or bathing alone, but have someone with you who could lift your head above water in the event of an epileptic seizure.
- Live with order and regularity: an irregular day and night rhythm or lack of sleep can trigger an epileptic seizure.
- After an epileptic seizure, you are not allowed to drive a car or motorcycle for a certain period of time. Ask your doctor which period of time applies to you.

Frequently asked questions

Can you drive after a cerebral venous sinus thrombosis?

After a cerebral venous sinus thrombosis you are sometimes (temporarily) not allowed to drive a car or motorcycle. There are several reasons for a driving ban. Therefore, ask your doctor which situation applies to you.

Can you drive after an epileptic seizure?

After an epileptic seizure, different rules apply with regard to driving a car and motorcycle. It is important, among other things, how many epileptic seizures you have had. Ask your doctor about the length of the driving ban in your specific situation.

Can you fly after a cerebral venous sinus thrombosis?

If you wish to fly within two weeks of the diagnosis of cerebral venous sinus thrombosis, please contact the relevant airline. For a flight longer than 4 hours, we advise you to use anticoagulant drugs (low-molecular-weight heparin in a low dose or a DOAC) on the day of departure, and 24 hours afterwards if necessary. This is to prevent a thrombosis. **Please note:** this advice does not apply if you are already taking anticoagulant medicines on a daily basis.

Websites
https://cerebralvenousthrombosis.com/

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Hartstichting

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