



Helping the Cavernoma Community

Information booklet

Symptomatic brain cavernomas

For people with
symptoms due
to their brain
cavernoma

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LOTTERY FUNDED

Registered Charity Number: 1114145



This booklet is for people affected by a brain cavernoma, which is also known as a ‘cavernous angioma’, ‘cavernous haemangioma’, or ‘cavernous malformation’.

In particular, this booklet is for:

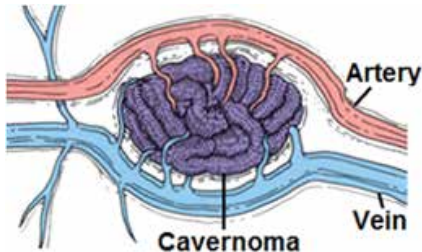
- people who have symptoms caused by a brain cavernoma.

There are other booklets in the Cavernoma Alliance UK series:

- one for people who have symptoms due to their brain cavernoma; and
- another for people who do not have symptoms from their brain cavernoma.

What are brain cavernomas?

Cavernomas are clusters of abnormal blood vessels. Cavernomas are found in the brain, brainstem, spinal cord and, rarely, in other areas of the body.



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Cavernomas look like raspberries, and are about the same size.

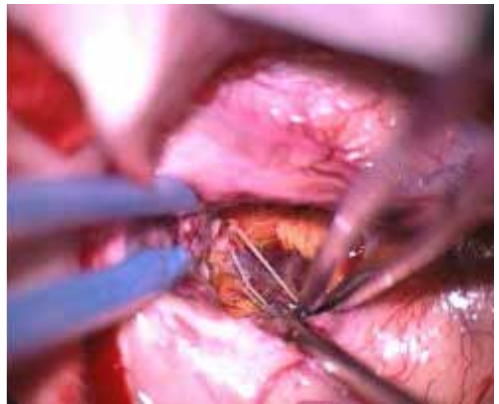
Cavernomas are made up of abnormal blood vessels through which blood flows slowly – these are the caverns that give the condition its name. The cells that line these caverns sometimes ooze small amounts of blood into surrounding brain tissue, which sometimes causes symptoms.

Cavernomas can get bigger, but this growth is not cancerous and they do not spread to other areas of the body. Most people with the condition only have one cavernoma. Some people have more than one cavernoma, and these people sometimes develop new cavernomas over time. Cavernomas can also be diagnosed by looking at brain tissue under the microscope.

View of a cavernoma under the operating microscope



The cavernoma is indicated by the arrow.



The brain after the removal of the cavernoma.

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How are cavernomas diagnosed?

Cavernomas have been known about since the middle of the 19th century, when they were seen by pathologists examining brains under the microscope.

Magnetic resonance imaging (MRI) is the most accurate test and provides pictures of brain cavernomas. Computed tomography (CT) scans and angiograms cannot be relied upon to show up cavernomas. Doctors only began to see how common cavernomas were when the MRI test was first developed in the 1980s. Before the MRI test was available, symptoms of brain cavernomas were sometimes missed, or mistaken for symptoms of other neurological conditions, such as multiple sclerosis. Even now, some people with a brain cavernoma wonder if they might have been diagnosed earlier if they had had an MRI test to investigate neurological symptoms that they had in the past.



Magnetic resonance imaging (MRI) of the brain (above) showing the typical 'raspberry' or 'popcorn' appearance of a cavernoma in an area called the 'midbrain', which is shown by the arrow on the picture.



A patient about to enter MRI scanner, with radiologist and radiographer (on left of picture).

What causes brain cavernomas?

In most cases, it's impossible to know what caused a brain cavernoma, but there are two recognised causes.

Radiation treatment

Radiation is used to treat a variety of medical conditions. During radiation treatment, a patient's brain may be exposed to radiation. Some of these people are later found to have a brain cavernoma. If these people have never had an MRI scan in the past, it is impossible to be sure whether the radiation had anything to do with the brain cavernoma developing. Even for patients who have had an MRI scan in the past, the age of the machine and the types of pictures taken during the scan may explain why a cavernoma was never found. However, for a few people who have had an MRI scan before receiving radiation treatment, and another MRI scan later on that identifies a brain cavernoma, it is possible that the radiation may have caused the cavernoma.

Genes

In less than half of the people affected by brain cavernomas, there is likely to be a genetic cause. Some people have more than one cavernoma, or may develop more than one cavernoma over time. In these cases there may be a genetic cause. These issues are discussed in further detail in the **'Information booklet for people concerned about genetic causes of brain cavernomas'**.

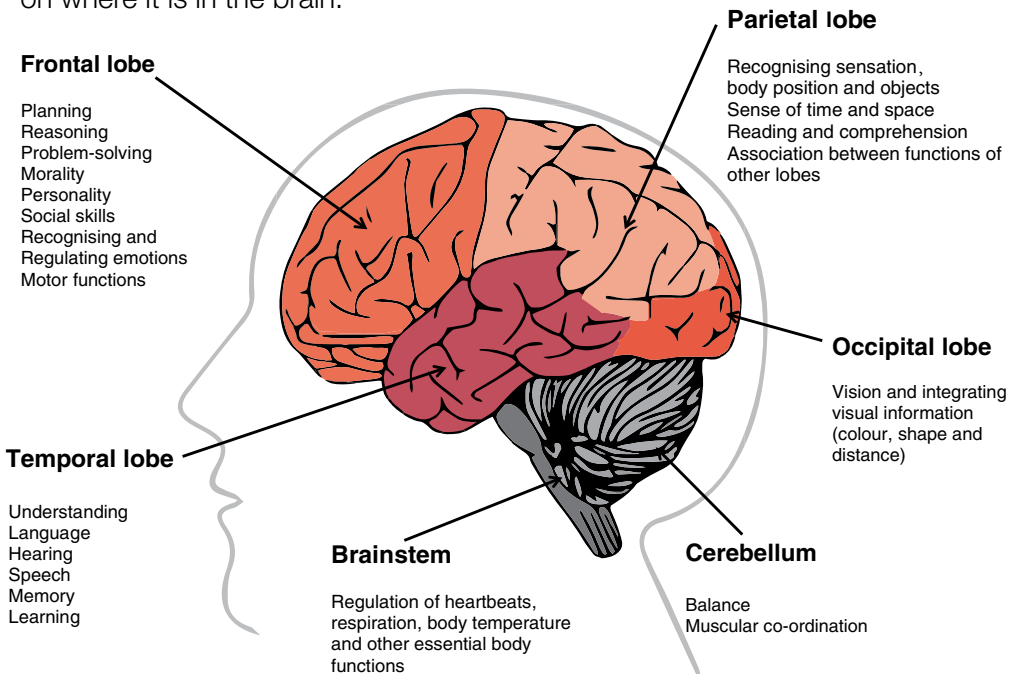
How common are brain cavernomas?

Brain cavernomas are quite common. Studies into the results of MRI scans on people who had no symptoms have revealed that approximately one person in every 600 has an **asymptomatic** brain cavernoma. This means that in the United Kingdom there are roughly 90,000 people with an **asymptomatic** brain cavernoma – enough to fill Wembley Stadium.

People who experience symptoms from a brain cavernoma are much rarer. A modern study based on the entire population of Scotland has found that, each year, one person out of every 400,000 is diagnosed with a **symptomatic** brain cavernoma for the first time in their life.

Symptoms

Many people who are diagnosed with a brain cavernoma have no symptoms at all, or they do have symptoms but they are not thought to be related to the cavernoma. This leaflet is for people whose cavernoma is thought to have caused symptoms. The symptoms a cavernoma might cause depend on where it is in the brain.



Drawing of the lobes and hemispheres of the brain

Cavernomas near the surface of the ‘hemispheres’ or ‘lobes’ of the brain can cause **epileptic seizures**.

Cavernomas can cause neurological deficits in any area of the brain. The most common symptoms of **neurological deficit** are dizziness, numbness, weakness, disturbed vision, speech difficulty, problems swallowing, and unsteadiness. The type and combination of neurological deficits caused by a cavernoma depend on where exactly in the brain the cavernoma is.

These neurological deficits are sometimes caused by bleeding from the cavernoma, known as **‘haemorrhage’**. If new symptoms occur because of a haemorrhage and last more than a day, this is called a **‘stroke’**. If a cavernoma bleeds, usually only a small amount of blood oozes from it (half a teaspoonful on average), so cavernoma bleeds rarely cause death or severe disability. In fact, bleeding from a cavernoma sometimes causes no symptoms at all. Brain scans can find any new bleeding, but this can only be done accurately by a CT scan within seven days after the symptoms began, or by an MRI scan within two months after the symptoms began.

Some people affected by cavernomas also experience problems with memory, attention, concentration, energy levels, and mood. These symptoms can have a variety of causes, not necessarily the cavernoma. In the study based on the Scottish population, half (50%) of the people had no symptoms when they were first diagnosed with their brain cavernoma. The symptoms affecting the others were as follows.

- 15% of people had one or more epileptic seizure
- 8% of people had a brain haemorrhage
- 27% of people had neurological deficits, without a haemorrhage showing up on the pictures of their brain

What is the risk of developing symptoms?

A few research studies provide information about the outlook (sometimes called ‘prognosis’) for people who have symptoms due to a brain cavernoma. The most precise estimates of the risk of stroke due to haemorrhage come from a combined analysis of seven studies involving 1,620 people with brain cavernomas. These studies were able to describe the risk of haemorrhage over five years following diagnosis, which is described overleaf.

For people with a cavernoma that has caused one or more epileptic seizure, the risk of stroke due to haemorrhage is the same as for people with incidental cavernoma. Please see the information booklet about incidental cavernomas for these risks. For people with a cavernoma that has caused one epileptic seizure, the risk of going on to have a second seizure over five years is approximately 94%.

For people with a brainstem cavernoma that had already caused a stroke due to haemorrhage in the past, the risk of having another haemorrhage over five years is approximately 30.8%. In other words, 1 in approximately 3 people with a **brainstem cavernoma** that had already caused a stroke due to haemorrhage will experience another stroke due to haemorrhage during the first five years after the diagnosis of their cavernoma. Put another way, 2 out of these 3 people will not have another stroke due to haemorrhage.

For people with a cavernoma that is not in the brainstem and had already caused a stroke due to haemorrhage in the past, the risk of haemorrhage over five years is approximately 18.4%. In other words 1 in approximately 5 people with a **cavernoma that is not in the brainstem** that had already caused a stroke due to haemorrhage will experience another stroke due to haemorrhage during the first five years after the diagnosis of their cavernoma. Put another way, 4 out of these 5 people will not have another stroke due to haemorrhage.

At the moment, we do not have reliable evidence to show that your age or sex affects the risk of bleeding from a cavernoma. Nor do we have reliable evidence to show that having more than one cavernoma makes the risk of bleeding higher. We do not know whether lifestyle influences the risk of bleeding from cavernomas. It seems that the risks of haemorrhage are similar for a person who has had an epileptic seizure due to their cavernoma and a person who has had no symptoms at all.

Because there is a shortage of information about what happens to people more than five years after diagnosis, we are not sure whether these risks of haemorrhage remain the same, or change, over the rest of someone's lifetime.

Treatment

Medication is available to treat seizures caused by cavernomas. Other symptoms, such as headaches, can also be treated with medication. The main ways used to try to stop cavernomas bleeding are **neurosurgery** and **stereotactic radiotherapy**.

Neurosurgery involves an operation under general anaesthetic. In this operation, the skull is penetrated (called a ‘craniotomy’) and the cavernoma is removed. The method used and the risks involved depend on where the cavernoma is in the brain. Surgery for cavernomas has been made safer using the operating microscope (used in treatment called ‘microsurgery’). Brain scanning during the operation allows surgeons to reach cavernomas with as little disruption to the normal brain tissue as possible. This procedure is sometimes known as ‘image-guided surgical navigation’ or ‘computer-assisted or frameless stereotaxy’.

Stereotactic radiotherapy involves beams of radiation being targeted at a brain cavernoma from many different points around the head.

The radiotherapy beams are targeted very accurately, so it is important that the head does not move while a patient is being treated. Various techniques are used to keep the head still, such as the head frame shown in the picture. When radiotherapy is given as a one-off treatment, it is sometimes called ‘radiosurgery’. Unlike brain surgery, ‘radiosurgery’ does not involve a general anaesthetic or an operation. Stereotactic radiotherapy is usually carried out to treat cavernomas in regions where brain surgery would be too dangerous. Whether or not stereotactic radiotherapy helps is uncertain.

It can be difficult to decide whether or not to have treatment for a cavernoma that has caused symptoms. One thing to think about is the risk of leaving the cavernoma alone. As mentioned above, these risks vary between patients in the short-term studies that have been done. The risks of bleeding from a cavernoma may settle down in the long term. The other thing to consider is the risk of treatment itself, which varies depending on where the cavernoma is and the type of treatment used. Before deciding whether to have treatment, you should talk to your doctor about your cavernoma.



Neurosurgery being performed in an operating theatre.
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Fitting the frame for stereotactic radiotherapy.
Image courtesy of Elekta.

Some people ask whether they can have their cavernoma **'monitored'** by going to clinic appointments and having repeated brain scans. This can be useful when there is any uncertainty about the diagnosis of a cavernoma. However, even when diagnosis is certain, no features on a brain scan (such as signs that a cavernoma has grown) are known to show that the risk of bleeding would be higher. For most people, clear information from a doctor, and a clear discussion about whether or not to treat the cavernoma, is probably enough.

What is still not known about brain cavernomas?

As you can tell from reading this booklet, there are still many important questions about cavernomas which we do not yet have answers to. For this reason, you may be asked to take part in research studies.

- We know of three genes that can cause certain types of cavernomas. If you want more information about these, please read the **'Genetics of brain cavernomas'** booklet. However, we still don't know how cavernomas are formed, which is why researchers may want to study the genetic code (DNA) in a blood sample, or brain tissue taken during neurosurgery.
- We know very little about the long-term outlook for people with brain cavernomas, or about what influences the risk of future bleeding. A few long-term studies are ongoing.
- For some people and some cavernomas, there is uncertainty about whether to treat them and, if so, whether to treat them using neurosurgery or stereotactic radiotherapy.
- There may be clinical trials of these treatments in the future, and these trials are likely to be randomised (in other words, whether a patient in the trial gets treatment or no treatment, or one treatment or another, would be chosen at random by a computer). Such randomised controlled trials are the fairest test of whether treatment is beneficial.

This booklet

This booklet was written by Rustam Al-Shahi Salman, and reviewed by Neil Kitchen and members of Cavernoma Alliance UK.

How Cavernoma Alliance UK can help

Cavernoma Alliance UK was set up by people affected by brain cavernomas to support those with the condition and their families. Our website, www.cavernoma.org.uk, provides information about brain cavernomas, current members, and information about how you can become a member, membership is free. Along with our affiliated organisation, Angioma Alliance, and associated groups throughout the world, we also provide updates on research.

Our aims

- To make sure that every person with a brain cavernoma, and their family, has access to clear information about the illness.
- To provide information, through our website, about others affected by brain and spinal cavernomas.
- To increase awareness of brain cavernomas so that those affected by the condition receive understanding and support.

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Acknowledgements

We are very grateful to
The Alice Ellen Cooper Dean Charitable Foundation
for their financial support in producing
this information booklet.



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