

# Symptomatic brain cavernomas

## Information leaflet for people with symptoms due to their brain cavernoma



Helping the Cavernoma Community

Website: [www.cavernoma.org.uk](http://www.cavernoma.org.uk)

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Registered charity number: 1114145

## Who is this leaflet for?

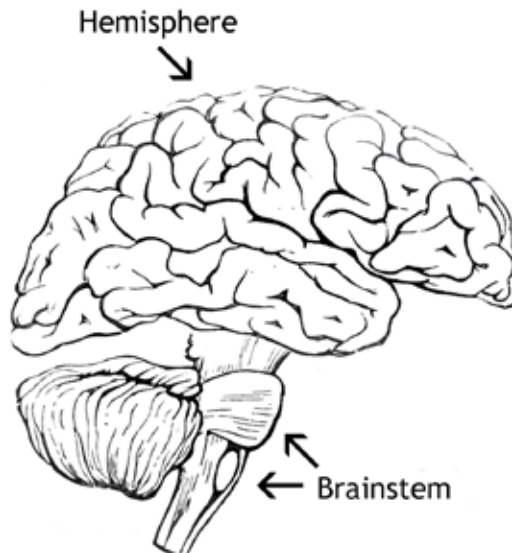
This leaflet is for people affected by a brain cavernoma, which is also known as a 'cavernous angioma', 'cavernous haemangioma', or 'cavernous malformation'. In particular, this information is for people who have symptoms caused by a brain cavernoma.

There are other leaflets in the Cavernoma Alliance UK series:

- one for people who do not have symptoms due to their brain cavernoma; and
- another for people concerned about the genetic causes of brain cavernomas and whether they run in families.

## 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.



Picture of the lobes and hemispheres of the brain, and the brainstem (labelled).



Cavernomas look like raspberries.

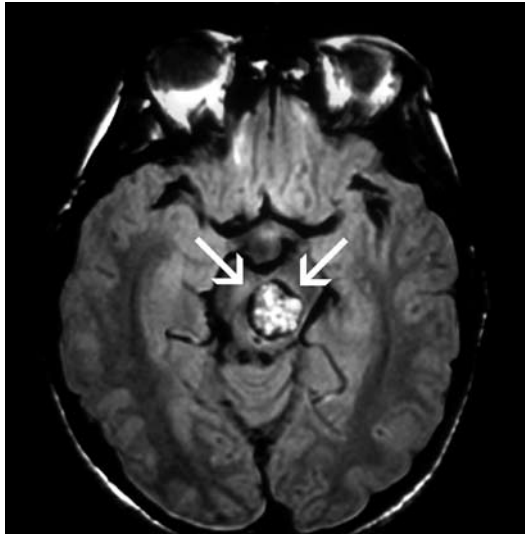
Cavernomas look like raspberries. They are made up of abnormal blood vessels through which blood flows slowly - these are the caverns that give the condition its name. The cells that line these caverns sometimes ooze small amounts of blood into surrounding brain tissue, which sometimes causes symptoms.

Cavernomas can get bigger, but this growth is not cancerous - 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.

## 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



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 arrows on the picture.

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.

## 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 fewer 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 more detail in the '**Information leaflet 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.

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 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 7 to 10 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. So far, most of these studies have only involved people attending neurosurgery units, so they do not represent the entire population. At the moment, most of the information available concerns the risk of cavernomas bleeding in the future. Another problem is that the average follow-up period in these studies has been no more than five years. This limits the amount that anyone can know about what will happen in the long term.

It would be useful to know what the outlook is for the following three groups of people with symptoms as a result of their brain cavernoma.

- People who have had one or more epileptic seizure
- People with a neurological deficit (whose brain scan does not show fresh bleeding from the cavernoma)
- People whose cavernoma has bled

## Risks for people who have had one or more epileptic seizure, or who have a neurological deficit, and whose brain scan did not show bleeding from the cavernoma

Two studies showed that for people who had never had a haemorrhage from their brain cavernoma, bleeding seemed to affect only one in 200 people (0.5%) **each year**.

## Risks for people who have had symptoms caused by their brain cavernoma bleeding

The risk of future bleeding appears to be higher for people who first come to medical attention with bleeding from their cavernoma. However, the risks of having another bleed vary widely. These risks range from roughly one in 25 people (4%) a year, to roughly one in four people (23%) a year.

Some, but not all, studies have found a higher risk of future bleeding from cavernomas in the brainstem. The brainstem is a small, tightly packed structure that all information going between the brain and the spinal cord flows through. There is a higher risk of further symptoms from cavernomas in the brainstem. If this is the case, it will probably be a result of the way the brainstem is made up, because it only takes a very small amount of blood to cause symptoms.

A few studies have suggested that women and people under 40 years of age may have a higher risk of future bleeding. However, other studies have not given the same results, so this may not be correct.

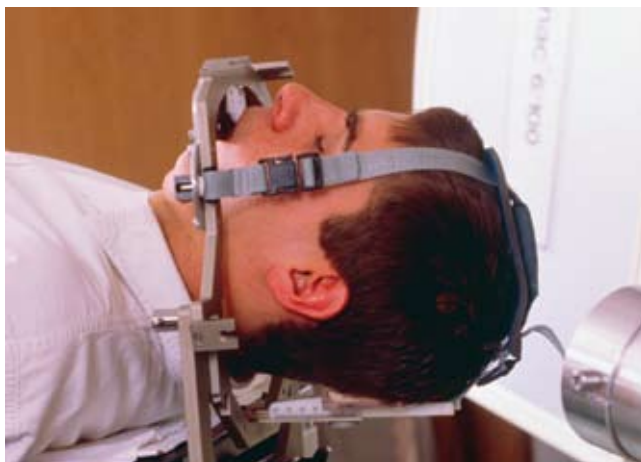
## Treatment

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 being performed in an operating theatre.

**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'.



Head frame used during one type of stereotactic radiotherapy.

**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 above. 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 a lot between patients and in the results of **short-term** studies that have been done. Many researchers think that 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.

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 cavernomas?

As you can tell from reading this leaflet, 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**' leaflet. 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 best way to work out whether treatment is beneficial, or not.

## This leaflet

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

## How Cavernoma Alliance UK can help

We, Cavernoma Alliance UK, were set up by people affected by brain cavernomas to support sufferers and their families. Our website, [www.cavernoma.org.uk](http://www.cavernoma.org.uk), provides information about brain cavernomas, our current members and information about how you can become a member. Along with our parent organisation, Angioma Alliance, based in Virginia, USA, we also provide updates on research.

## Mission statement

- 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 cavernomas and to provide support.
- To increase awareness of brain cavernomas so that those affected by the condition receive understanding and support.

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