Cavernoma Alliance UK

Helping the Cavernoma Community

Information booklet

Spinal cavernoma

For people with symptoms due to their spinal cavernoma

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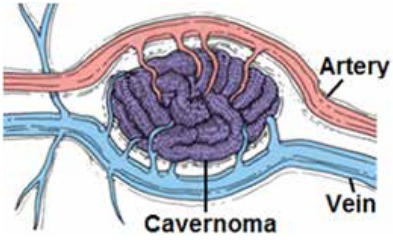
This booklet is for people affected by a spinal cavernoma, which is also known as a ‘cavernous angioma’, ‘cavernous haemangioma’, or ‘cavernous malformation’.

In particular, this booklet is for people who have symptoms caused by a spinal cavernoma.

There are other booklets in the Cavernoma Alliance UK series. They are for:

- people who have symptoms due to their brain cavernoma;
- people who do not have symptoms from their brain cavernoma;
- people concerned about genetic causes of brain cavernoma and whether cavernomas run in families; and
- people interested in pregnancy and birth who are affected by cavernoma.

What are cavernomas?



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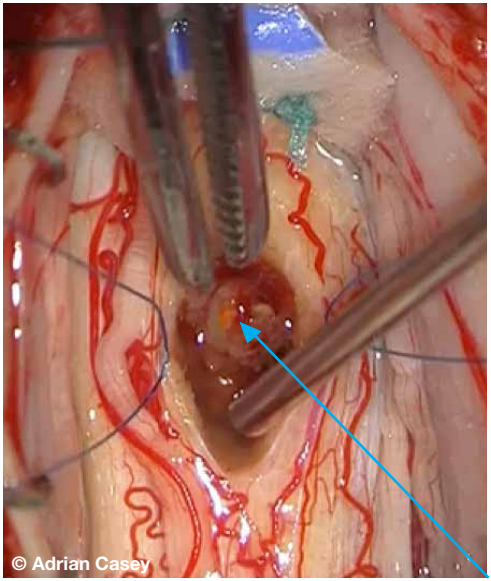
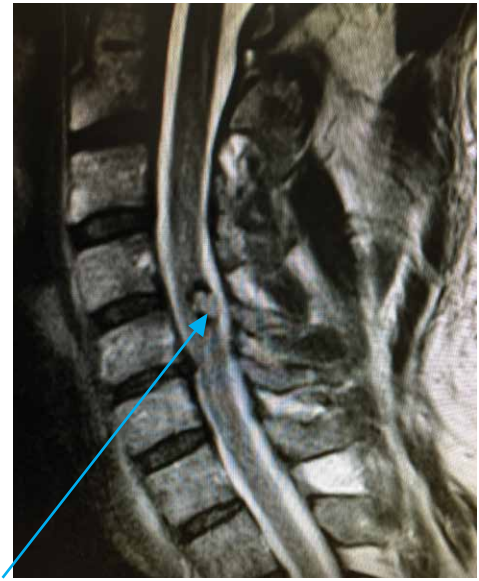
Cavernomas are clusters of abnormal blood vessels through which blood flows slowly – these are the caverns that give the condition its name. They are found in the brain, brainstem, spinal cord and, rarely, in other areas of the body. The cells that line these caverns sometimes ooze small amounts of blood into surrounding brain tissue, which sometimes causes symptoms. In the spinal cord, a cavernoma is usually the size of a pea (1 centimetre). On a scan, a cavernoma looks like a piece of popcorn and under the microscope it looks like a blueberry.





Cavernoma

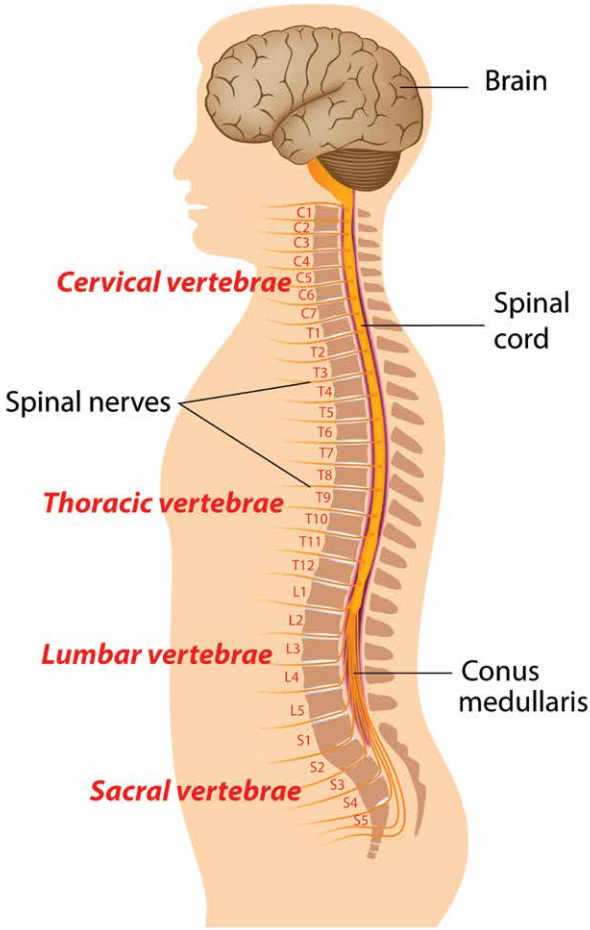
Spinal cavernomas

Spinal cavernomas are cavernomas within the spinal cord.

The spinal cord is the main bundle of nerves that connects the brain to the rest of the body. The nerves are very tightly packed, so even small cavernomas can cause symptoms. This is one of the reasons spinal cavernomas tend to cause more problems than brain cavernomas.

Spinal cavernomas may be intramedullary (inside the spinal cord) or outside the spinal cord but in the spinal bony canal (extramedullary, or intradural). About 85% of spinal cord cavernomas are inside the spinal cord. Spinal cavernomas are usually discovered when people are in their thirties or forties.

Cavernomas in the spinal cord are not as common as cavernomas in the brain. However, if you have a spinal cavernoma, there is about a 1:6 to 1:4 (16% to 25%) chance you will have a brain cavernoma too.

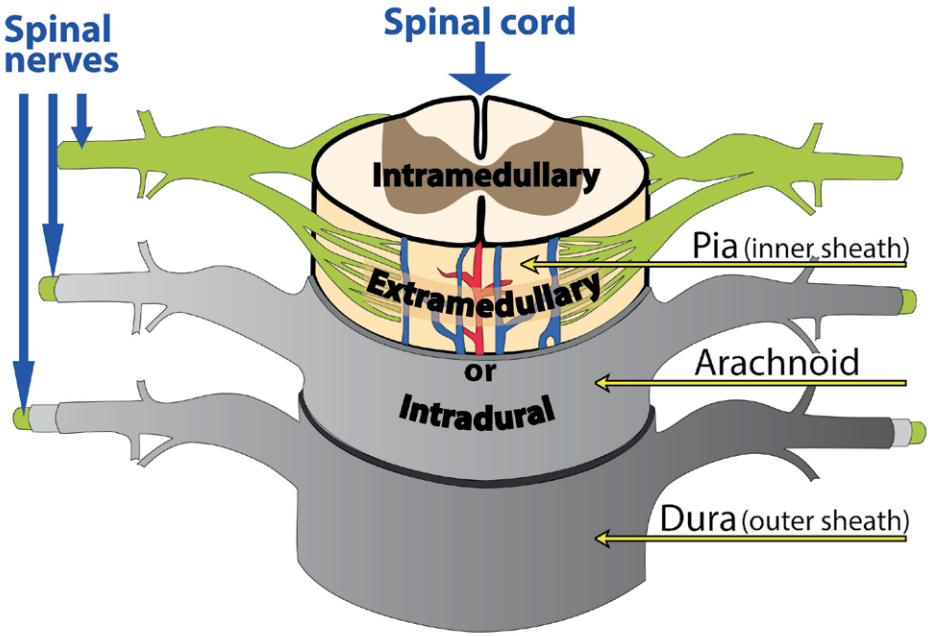


What we know about intramedullary spinal cavernomas

Because it is relatively rare to have a cavernoma inside the spine, no large studies have taken place. The information that we have is from small studies of about 20 or 30 patients.

Here are some basic facts about spinal cavernomas.

- The average age people first visit their doctor with symptoms of a spinal cavernoma is 39, though cavernomas can affect people as young as two or as old as 80.
- Equal numbers of men and women are affected by spinal cavernomas.
- The upper part of the spine (cervical spine) is involved in about 40% of cases and the thoracic in about 55%. The remaining 5% involves the conus medullaris and lumbar spine (see the diagram on page 4).



The spinal cord is surrounded by three sheaths, of which the outer is known as the 'dura'. The space inside the inner sheath (or 'pia'), is known as 'intramedullary', and the space between the inner and outer sheaths is 'extramedullary' or 'intradural'.

What causes cavernomas?

Cavernomas may be hereditary or spontaneous. (Spontaneous cavernomas are also referred to as 'sporadic'.) They are caused by a change in the cell walls of the blood vessels (capillaries), which results in blood leaking and the cavernoma developing.

Hereditary (genetic) causes

We explain the genetics of cavernomas in more detail in our information booklet for people concerned about genetic causes of brain cavernomas. Briefly, three genes are known to be responsible for the development of cavernomas – CCM1, CCM2 and CCM3. Mutations (changes in the structure) of these three genes are the cause of about a quarter of cases of cavernomas. A parent with one of these genes has a 50% chance of passing the gene to their children. These numbers apply to all cavernomas – we do not know if they are different for spinal cavernomas.

Radiation treatment

Radiation to the spine, usually given for childhood cancers, leukaemia or lymphoma, may result in a cavernoma.

What are the symptoms of a spinal cavernoma?

Symptoms are caused by injury to the nerves and are related to what the spinal cord normally does. They include the following.

- Weakness in the hands or legs
  Our muscles are controlled by nerves in the spinal cord. If the nerves stop working properly as a result of a haemorrhage higher up in the cervical neck area, both the hands and the legs are more likely to be involved. Lower down, in the thoracic area of the spinal cord, only the legs are likely to be affected.
- Sensory problems – pins and needles, numbness, or problems with balance.
- Pain – this is a sign that the nerves are not working properly.
- Bladder and bowel problems
  To be able to control our bowel and bladder we need to be aware of the sensation that we want to 'go' and we need to have muscle control. As discussed above, spinal cavernomas can affect muscle and sensation control.

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 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 cavernomas were sometimes missed, or mistaken for symptoms of other neurological conditions, such as multiple sclerosis. Even now, some people with a 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 treatment is available for cavernomas?

Microsurgery

In microsurgery, surgeons use modern techniques to completely remove the cavernoma, while keeping the injury to the rest of the spinal cord to a minimum. The aim of surgery is to reduce or remove the chances of the cavernoma bleeding again. This is best achieved by completely removing it. Surgery needs to take place in highly specialized centres by surgeons who have significant experience. This treatment has benefits but, even at the hands of a very experienced surgeon, it also carries risks.

Conservative treatment

This is supportive treatment that does not treat the cause (the cavernoma) but aims to achieve the best chance of neurological recovery and reduce the side effects of nerve injury. There are various forms of conservative treatment, such as rehabilitation programmes consisting of physical therapy and exercises, and targeted psychological support including coping strategies. This treatment has none of the risks of surgery, but does have other risks (the chance of another bleed and further injury).

What are the risks of each treatment?

Microsurgery

When choosing a treatment, you need to weigh the risks against the benefits.

The risks of surgery include the following.

- Risk from general anaesthetic. The younger and healthier someone is, the lower this risk becomes.
- Risks from general surgery. These are the risks encountered in most surgical procedures. They include the risk of bleeding and infection and, in spinal operations, the risk of spinal fluid leaking.
- Risks of microsurgery. The main risk is further injury to the spinal cord (the very thing the surgeon is trying to protect). This can result in your balance becoming worse, and pain or weakness which, in the worst cases, can lead to paralysis and bowel and bladder incontinence.

About 1 in 4 patients will notice that their symptoms get worse immediately after surgery. However, by six weeks to three months:

- 50% of patients will be better than before the surgery (their symptoms and nerve function will have improved);
- 40% will be the same as before surgery (but with the cavernoma re moved); and
- 10% will have a worse nerve function compared with before the surgery.

Everyone is different, and the risks vary depending on where the cavernoma is and the surgeon's experience.

Conservative treatment

The risks of conservative treatment depend on the natural history of a spinal cavernoma. The natural history of a disease is what happens to someone who has a specific health problem over a long period, which doctors do not intervene in.

The risk of bleeding from a cavernoma within the spinal cord varies between 1.4% to 6.8% a year. 2% a year is probably more accurate, but we cannot be certain and every individual is different.

Common questions about spinal cavernomas

- Will the cavernoma start to bleed (or bleed again)? If so, how often?
- What will be the consequences of having a cavernoma? How will my symptoms change over the years?
- Can one cavernoma be more benign or have a more favourable course than another, and if so how will we know?
- Who should be treated and when? For example, research is still ongoing into whether it is best to remove a cavernoma.

The answers to these questions are based on observations of patients who were diagnosed with a cavernoma and did not have surgery. (More patients had surgery than those who didn't.)

What we know

People with spinal cavernomas follow two patterns. Less than half (45%) of patients will have one or more episodes of acute deterioration. A bit more than half (55%) will find that their nerve function slowly but steadily gets worse.

The patient's age, the location of the cavernoma in the spinal cord, how deep it is, or what clinical course a patient follows do not appear to affect the outcome.

On the other hand, if you choose to have surgery, having your cavernoma completely removed has a better outcome than if it is only partly removed.

This booklet

This booklet was written by Mr Adrian Casey from the National Hospital for Neurology and Neurosurgery, Queen Square, London, compiled by David White and Ian Stuart, and reviewed by Professor Rustam Al-Shahi Salman.

How Cavernoma Alliance UK can help

Our organisation 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 and spinal 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 brain or spinal cavernomas, 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 and spinal cavernomas so that those affected by the condition receive understanding and support.

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