

Arteriovenous malformations (AVM)

A close-up portrait of a young girl with long, dark brown hair and bangs. She is smiling warmly at the camera. Her skin has a slightly reddish tint, particularly around her eyes and on her cheeks, which is characteristic of an arteriovenous malformation (AVM).

Information for families

Great Ormond Street Hospital
for Children NHS Foundation Trust

This information sheet from Great Ormond Street Hospital (GOSH) provides information about arteriovenous malformations (AVM) and how they can be treated. It also explains what to expect when you come to GOSH for diagnosis and treatment.

What is an arteriovenous malformation (AVM)?

An arteriovenous malformation (AVM) is a specific term used to describe a tangle of blood vessels with abnormal connections between arteries and veins.

High pressure arteries containing fast flowing blood are directly connected to low pressure veins which normally only contain slow flowing blood. This means that blood from the arteries drains directly into the veins without stopping to supply the normal tissues in that part of the body with essential substances like oxygen and nutrition; over time this can lead to the normal tissues becoming painful or fragile. It also means that the AVM gets progressively larger over time as the amount of blood flowing through it increases and it can cause problems due to its size.

Finally, it may also mean that the heart has to work harder to keep up with the extra blood flow.

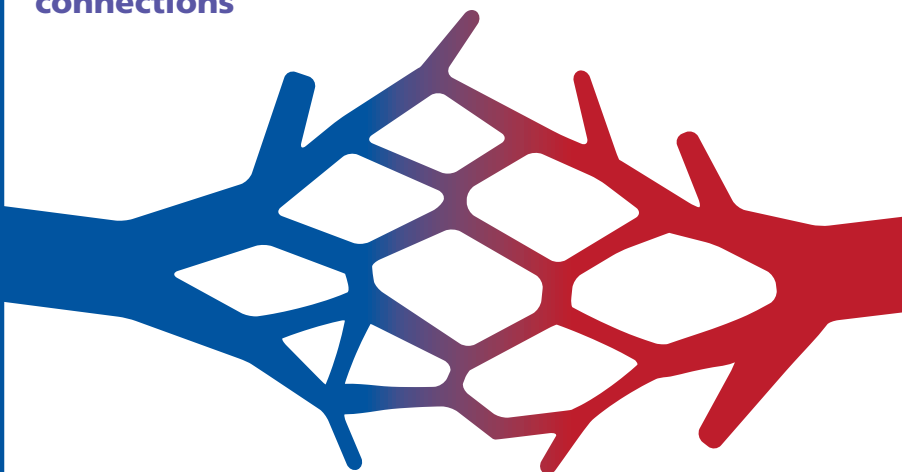
One of our specialist doctors describes an AVM as a ring road that bypasses the high street of a town. Traffic (or blood) will use the bypass rather than the high street which suffers as a result.

Many AVMs, however, are small and do not cause any significant problems during a person's life.

An AVM usually has many arteries supplying it and several draining veins. The centre of the AVM is often called the nidus.

AVMs may occur anywhere in the body, including near the surface of the skin. AVMs in the brain and spinal cord have different symptoms and treatment so are explained in another information sheet.

**Normal
connections**



**Arteriovenous
malformation**



What causes an AVM?

AVMs are almost always congenital (present at birth), although they are often not very obvious at birth. It is thought that they arise from an error in blood vessel formation very early in pregnancy. In most cases we do not know exactly what causes this error. In a small proportion of cases, AVMs arise as part of an inherited condition. There is no evidence to suggest that AVMs are the result of something you did or did not do during pregnancy.

How common are AVMs?

The incidence of all types of AVM taken together is thought to be approximately 1.4 in every 100,000.

What are the signs and symptoms of an AVM?

An AVM involving the skin may cause blue, purple or red discolouration. They often cause swelling and sometimes the blood vessels are visible close to the skin surface. The area usually feels warm compared to the surrounding skin, due to the high rate of blood flow through it. A pulse or buzz may be felt. The skin over the AVM may be delicate and more likely to break down or develop an ulcer, which can be painful. Over time, the increased blood flow may affect heart function.

Unlike haemangiomas, AVMs do not shrink. They tend to increase slowly in size during childhood and adolescence, and may become more prominent during pregnancy.

How is an AVM diagnosed?

As an AVM can be confused with other birthmarks, the diagnosis is best confirmed by a specialist centre. The doctors there will carry out a physical examination and take details of the child's medical history and usually this is enough to make a diagnosis, although ultrasound scans and/or Magnetic Resonance Imaging (MRI) scans may be carried out to confirm it. Other imaging techniques, such as catheter angiograms, may be suggested to better define the AVM and plan treatment.

What day to day care do AVMs need?

Generally, AVMs do not need any special care on a daily basis.

If the blood vessels in an AVM are near the surface of the skin, they can bleed if they are knocked or scratched.

If the AVM starts to bleed, apply pressure over it with a clean handkerchief, cloth or tissue for at least five minutes. If blood soaks through the handkerchief, cloth or tissue, put another one on top and keep up the pressure. Do not take it off to have a look as this could start the bleeding again. If the bleeding continues, even after pressing down on the AVM for five minutes, go to your nearest NHS Walk-In Centre or Accident and Emergency department. The team at GOSH can usually be contacted for advice if needed.

Occasionally, the skin over an AVM can form an open sore or ulcer, which is painful. Ulcers can become infected, so a visit to your doctor is important, as infected ulcerated areas may need treatment with antibiotics.

How is an AVM treated?

Whether and how an AVM is treated depends on its location and what problems it is causing. Some AVMs are small and do not cause any significant problems during a person's life, so can be safely left untreated.

If an AVM is troublesome because of its size, location or other symptoms, there are several treatment options available. Some small AVMs can be removed surgically but the majority of AVMs are too complex to treat in this way and are treated with embolisation instead. This is a minimally invasive procedure usually performed by an interventional radiologist. It aims to slow down or block off blood flow through the AVM by closing some or all of the abnormal blood vessels from the inside. Various substances can be used to block the blood vessels, including medical glue, medical putty, tiny metal coils or plastic beads.

The substance used depends on the area being embolised, the speed of the blood flow in that area and the size of the blood vessels. Sometimes two or more substances are used together to get the best result. Embolisation of high flow AVMs is a very specialised procedure and is only performed in specialist centres. Planning such a procedure is often complex and the options and risks will be discussed in detail before any decision is made. Embolisation is a good way of controlling the symptoms of an AVM but may not offer a complete cure.

If the blood flow through the AVM is successfully reduced, surgery may be suggested to improve the cosmetic appearances afterwards or to remove what is left. Any surgery has a risk of bleeding and infection and some scarring will result as well. In some cases, it may be preferable to embolise the AVM and not carry out any further surgery afterwards.

What happens next?

If some of the AVM remains after treatment, blood can start to flow again through the AVM. This will be monitored over time, by seeing your child in clinic and sometimes performing repeat scans. Repeat embolisation procedures may be required if the blood flow through the AVM becomes troublesome again.

Conclusion

Vascular malformation research is an area of medicine that is continually advancing; studies have already given us improved options for treating AVMs and continue to tell us more about how and why they develop. We hope that this leaflet has been helpful in learning more about your child's AVM; if you have any comments about it, please contact the Vascular Anomalies Unit.

Further information and support

At GOSH, contact the **Vascular Anomalies Unit** on 020 7829 8668.

The **Butterfly AVM Charity** can offer advice and support to anyone affected by an AVM.

Telephone them on 07811 400 633 or visit their website at www.butterflyavmcharity.org.uk

The **Birthmark Support Group** can also offer advice and support.

Telephone them on 0845 045 4700 or visit their website at www.birthmarksupportgroup.org.uk

Nikki's story

I go to GOSH for lots of ops to get better. I find it boring having to go to the hospital for check-ups all the time as it's in Central London. I also have to have regular blood tests because I'm on a special medicine. People are always staring at me and ask questions about my eye and they think that I may have fallen down and hurt myself. This makes me sad. My Daddy says to me that it's what's on the inside that matters not the outside. As AVMs are rare I have not met many children that have one.

On the bright side I get to make lots of friends and have met some lovely doctors and nurses that are really kind and help me although I just wish I was a normal little girl. I also like the arts and crafts at GOSH and when I stay in hospital I go to the school. My Mummy and Daddy support me all the way on my journey and look after me and I love them lots :)



George's story (Nikki's dad)

Every day is both special and unpredictable when you have a child with an AVM. Special because you cherish the good days when your child is relatively symptom free and unpredictable because you appreciate that due to the nature of Nikki's illness we can quickly move from parent to carer mode! I find myself looking through the eyes of Nikki as people cannot stop staring and all I wish for is her acceptance into society that is often cruel and based more often than not on the way we look.

Once the trauma of finding out that Nikki had a very rare illness subsided, I quickly became aware of the do's and don'ts making sure that whoever she was with both in and out of school was informed about her condition and what to do if

she had a bleed and who to contact. Also due to her AVM Nikki cannot over exert herself too much so this too has to be monitored, although looking at her running up and down in the house and dancing you would never know!

It is our job as parents to teach Nikki how to deal with the general public in a positive way and to take care of her emotional side as well as physical health, although children are far more resilient than we give them credit for. The future for Nikki I hope is positive.

There is certainly a lot of research ongoing with regard to pharmacological approaches to AVMs, and given the pretty rapid development of doctors understanding of the underlying vascular biology, I'm confident that in a few years from now, we will have far superior active agents and treatments. It is my goal as a parent to build awareness of AVMs and ensure that Nikki has as normal a life as she can.

Notes

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