

Vascular malformations and their management

This leaflet tells you about the condition known as vascular malformation, including venous malformation, arteriovenous malformation and lymphatic malformation. It also explains the various treatment options for these conditions, including percutaneous sclerotherapy which is a common treatment.

This information is not meant to replace informed discussion between you and your doctor but can act as a starting point for such a discussion. Further sources of information are suggested at the end of the leaflet. Your consultant will be happy to discuss any questions you might have.

As with any medical procedure, you should always understand the benefits and risks of any planned treatment before you sign a consent form.

What is a vascular malformation?

Vascular malformation is a ‘catch all’ phrase for a lesion or abnormality that arises during the development of blood vessels or lymphatic channels even before you have been born. This means these lesions are present before birth but normally become evident either in childhood or adulthood. Vascular malformations consist of abnormal channels which have enlarged to develop fluid-filled sacs containing either blood (with blood circulating in the lesion) or lymph, a fluid which surrounds all the cells of the body and helps fight off infection.

We can therefore classify vascular malformations according to the main type of vessel or channel affected and what is flowing in them. These are most commonly veins (“slow flow” venous malformations), arteries and veins (“high flow” arteriovenous malformations) and lymph channels (lymphatic malformations). While veins and arteries carry blood, lymph channels carry lymph. Your doctor will tell you the type affecting you. Overall, the incidence of vascular malformations is around 1.5% in the population with the commonest type being venous malformations.

Are they harmful?

Vascular malformations are benign. This means you may carry the malformation without suffering any adverse effects. With the exception of very rare childhood syndromes they are not inherited or passed on to children. They are not a tumour or cancer. They cannot spread to other areas of the body, although some people can have very extensive problems or several areas are affected. While vascular malformations can occur in any organ in the body, small vascular malformations occurring deeper in the body usually simply go unnoticed. This leaflet refers to those malformations that occur close to the skin surface and are most commonly noticed as a soft lump.

What symptoms do vascular malformations cause?

This can depend on what type of malformation they are but in general they appear as a soft lump or bump which often persists and can affect any part of the body. They can cause mild discomfort or occasionally more severe pain, especially if they are in an area under constant movement or pressure, or if they become inflamed or infected. For similar reasons vascular malformations can enlarge rapidly and they may also vary in size depending how active you are. They can also become larger during puberty or pregnancy, or if clots develop within them. In children the lesions most commonly enlarge as the child grows. Overlying skin can discolour, especially if they have bled under the skin surface in the past. Very rarely the skin can break down to form an ulcer.

How do we diagnose vascular malformations?

You have probably already undergone a complete assessment to determine the nature of your lesion. Your doctor will have assessed your history of vascular malformations, performed an examination and requested specific tests. These may include an ultrasound and MRI study to assess the nature and extent of the lesion. Sometimes, an X-ray can also be helpful. In rare instances, an additional biopsy might be performed to confirm a vascular malformation.

How are vascular malformations normally treated?

If treatment is necessary, you will normally be referred to the Interventional Radiology Department where you can discuss your condition and treatment options with an interventional radiologist. They work closely with other doctors such as plastic and vascular surgeons. Normally a joint decision is made on the most appropriate treatment.

Most vascular malformations do not require active treatment once the diagnosis has been made and the patient is reassured of its benign nature. If lesions are relatively small, cause only minimal discomfort or symptoms, do not involve sensitive structures (for example joints) and do not limit daily activities, then it is entirely reasonable to leave things alone. This does not mean treatment cannot be considered in future. Some patients find the additional use of supportive clothing or a grade 2 compression stocking over the lesion useful to prevent discomfort or enlargement when active, or simply to prevent it being knocked during the day. Compression stockings should be worn during the day but taken off at night.

If treatment is considered necessary then one of the following options may be considered.

Surgery: When lesions are focal (affecting a discrete area) surgical removal can be considered. Lesions which respond poorly to percutaneous sclerotherapy (see below) could also be considered for surgery. The risks of surgery depend on many factors, including the site and nature of the lesion, but it will normally involve a general anaesthetic (you will be asleep) and it will result in a scar although this may be small and will normally fade considerably with time. It is rarely possible to guarantee the entire lesion has been removed and symptoms can therefore re-occur. The surgeon will explain the precise details of treatment and help you weigh up the risks and benefits.

Percutaneous sclerotherapy: Percutaneous sclerotherapy is a minimally invasive method of treating vascular malformations. Percutaneous means ‘through the skin’ and sclerotherapy refers to injection of a ‘sclerosant’ through the skin into the sacs which form the lesion. A sclerosant is an irritant which is designed to irritate the lining of the abnormal sac-like channels of the lesion. This produces inflammation and encourages the walls of the sacs to stick together and therefore obliterate the channels.

The procedure is performed by a consultant interventional radiologist normally in the Radiology Department angiography suite. Patients can be treated as a day case using local anaesthetic applied as a cream over the skin and sometimes injected under the skin surface (to numb the affected area). Sedative drugs can also be injected through a vein in the arm. This keeps patients relaxed and less aware but still awake or rousable. Children usually require a general anaesthetic.

Sclerotherapy should not be considered as a one off treatment but is more likely to involve a course of treatment to be repeated at intervals until a satisfactory control of symptoms has been achieved.

Frequently asked questions

What happens before the procedure?

Treatment is rarely urgent and should be performed at a time convenient for you when work and other commitments allow you a 1 to 2 week recovery period. Once you have met the doctor, discussed to your satisfaction the treatment plan and agreed to proceed, you will be given an appointment to attend the hospital, normally as a day case. We may carry out some blood tests before you attend.

What happens on the day of the procedure?

Most patients are admitted on the day of the procedure having been asked to starve for 6 hours prior to the procedure. You may have clear fluids (water) up to 2 hours before the procedure. The nurse will make sure you are in a hospital gown, check your blood pressure and confirm your details. A cannula (plastic tube) will be inserted into your arm for the administration of drugs and fluids, if required. You will be seen by your consultant before the procedure. Your consent for the procedure will be confirmed and you will again have an opportunity to ask any questions.

What happens during the procedure?

Percutaneous sclerotherapy normally takes between 30 minutes to one hour. It starts with cleaning the affected area and covering with sterile drapes. Pain relief and anaesthetic are provided as described above. You will be constantly monitored to ensure you are comfortable. The doctor then inserts tiny needles (one or two at a time) in to the sacs of the lesion using an ultrasound machine to make sure they are placed in exactly the correct spot. X-ray contrast (dye) is then injected and watched on a screen using a special X-ray machine (fluoroscope). This confirms the needle is in a safe place for injection. Finally the sclerosant is injected under X-ray guidance. The procedure is repeated in different areas of the lesion. Once completed, all

needles are removed, small plasters applied and the area then covered with bandaging to compress the lesion and encourage the small sacs to close.

Occasionally, some malformations require more complex treatment requiring direct access to the blood vessel by placing a tube in the artery or vein in the groin or arm. The doctor will make this clear when explaining the procedure.

What happens after the procedure?

Post-procedure care is tailored to the individual but may include the following:

- You are transferred to the recovery area, provided with further pain relief as required and discharged after 4 to 6 hours of monitoring.
- The compression dressing should be left on for 48 hours and removed at a wound check which will be performed in the X-ray department. This is then replaced with a compression stocking to be worn during the day for about 2 weeks.
- Depending on the site of the lesion you may require a sling to elevate the arm or crutches and remain non-weight bearing for 48 hours.
- Generally you should aim to return to work within 1 or 2 weeks of treatment and clinic follow-up will be arranged for 6 weeks' time and further treatment arranged if required.

What are the benefits of the procedure?

Sclerotherapy is an effective treatment in the majority of patients. It also has some benefits over surgery. Minimal access mean minimal trauma with shorter recovery times. There is no surgical scar. The treatment can also be offered for lesions that are unsuitable for surgery; those that are spread out within the soft tissues and are not easily surgically excised, and lesions near to sensitive structures which could be damaged in surgery.

What are the risks of percutaneous sclerotherapy?

There are risks associated with any procedure and these should be balanced against the potential benefits. This will be explained to you in detail before you consent for the procedure. Percutaneous sclerotherapy can have serious complications but this is rare and steps are always taken to minimise the risks.

- **Pain:** Sometimes the procedure can still be uncomfortable despite the pain relief provided. Because sclerosant naturally produces inflammation there is always swelling and discomfort after the procedure which will last a few days and is normally well controlled with pain killing tablets like paracetamol and anti-inflammatories like ibuprofen. Raising the affected area also helps.
- **Damage to surrounding structures:** Leakage of sclerosant in to surrounding tissues can cause damage. Nerve damage can lead to pain, weakness or numbness, but this is normally temporary and rarely long-standing or permanent. Muscle scarring which could limit movement and require treatment such as physiotherapy. Damage to overlying skin can include staining or discolouration (which normally fades), scarring or sometimes loss of skin that might require skin grafting. Sclerosant can also leak into normal blood vessels where there is a risk of causing clots and deep vein thrombosis.

- **Bleeding and infection:** There is always a small risk of this with any procedure involving needle insertion. Risks are minimised by using image guidance to aim the needle and by using a sterile technique, sometimes with additional antibiotic cover to further reduce the risk of infection.
- **Failure or recurrence:** Some patients respond poorly to sclerosant injection and a further procedure using more sclerosant might be considered. In such cases surgery may also be considered instead. A lesion can very rarely be treated entirely and there is always a risk that symptoms could return in future and further treatment may be necessary.

How successful is percutaneous sclerotherapy?

It is difficult to accurately predict how successful treatment will be for any individual. Studies looking at the outcomes from treatments suggest that, across the range of different types of vascular malformations, around 65-90% of people benefit from initial partial or complete relief of symptoms from a course of sclerotherapy treatment. The success of a procedure depends a lot on the anatomy of a lesion, its blood supply or drainage of a lesion. Your doctor will explain if there are any particular features of the lesion that make success more or less likely.

NICE (The National Institute for Health and Care Excellence) has not evaluated percutaneous sclerotherapy for vascular malformations in adults because they regard it as an “Established procedure - Procedures do not fall within the Programme's remit if they are considered standard clinical practice with an efficacy and safety profile that is sufficiently well known.” (NICE, March 2009).

Data collection

This leaflet is part of our commitment to provide the best possible care we can to you. We collect information on patients' conditions, the procedures they receive and the outcomes to ensure our practices are safe and to monitor how well the treatment works. This information is confidential and will be anonymised if submitted to an external electronic database. A doctor looking after you can fully explain why we collect this information and what details are recorded.

Sources of further information:

- British Society of Interventional radiology, www.bsir.org see patient section for an overview of the different types of malformations
- Great Ormond Street Hospital for Children, www.gosh.nhs.uk see medical conditions section and search for “venous malformation” or lymphatic malformation” or “arteriovenous malformation”. While focused on children (when vascular malformations are most commonly diagnosed) the information is still relevant to anyone wishing to gain further knowledge on the condition.

And finally....

Some of your questions should have been answered by this leaflet but remember this is only a starting point for discussion about your treatment with the doctors looking after you.

Make sure that you are satisfied that you have received enough information about the procedure before you sign the consent form.

Contacting us

Radiology Department, Tel 0118 322 7991.

Radiology Day Case Unit, Tel 0118 322 8368 (Monday-Friday 8.30am-5pm).

Interventional Radiology Depart, Tel 0118 322 7961

To find out more about our Trust visit www.royalberkshire.nhs.uk

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