

Lymphatic malformations and their treatment

Information for parents & carers



What is a vascular malformation?

Vascular malformations or vascular anomalies are general terms used to describe an abnormal cluster of vessels. Malformations are often present at birth and tend to grow slowly in proportion with the body. However at certain times, when the body's hormone levels increase, such as in puberty and pregnancy, they can grow more quickly. They can come in a number of forms.

Venous malformations

These often have a slow flow of blood running through them.

Arterio-venous malformations

These often have a higher flow of blood running through them.

Lymphatic malformations

These contain a clear fluid rather than blood; lymphatic malformations tend to be very slow flow.

Sometimes there can be a combination of the above. In this information booklet we will be looking at **lymphatic malformations**.

Are they harmful?

Lymphatic malformations are benign. They are not a tumour or cancer, and whilst some people have extensive problems affecting several areas of their body, they cannot spread to other areas of the body.

Patients who have lymphatic malformations may experience the following symptoms:

- **Swelling:** This is often a soft lump that can appear on any part of the body; the swelling can increase and decrease at different times.
- **Pain:** This can be mild to severe depending on the size and location of the malformation
- **Skin discolouration.**
- **Increased swelling** and pain can occur in association with any viral infection.
- **Rarely ulceration.**

How do we treat lymphatic malformations?

We will go through several steps to allow us to make the best decision regarding your child's care

Diagnosis:

Once we have received a referral from the team looking after your child we will, in the first instance, need to confirm the type of malformation they have and its extent as this will help us identify the correct course of treatment. They will need to undergo a number of investigations; these may include an ultrasound scan and/or an MRI, sometimes we may request a biopsy.

MDT (Multidisciplinary Team)

We will often discuss thier case with other professionals involved in treating patients with vascular malformations to make sure we are taking the best course of action in each case.

Treatment options

Once we have the results from their tests we will invite you and your child to attend an outpatient appointment at which point the interventional radiologists will discuss their treatment options with you. We may also discuss their case further with colleagues from other disciplines. After discussion we will normally offer the following options.

No treatment

If the malformation is causing your child no real problems in terms of living a normal active life, it may be reasonable to leave the malformation alone. If you decide, after discussion, to take this option then we will normally discharge your child from our care. This does not mean they cannot be re-referred if their symptoms increase.

Conservative treatment

We may offer conservative treatment in the form of compression garments. Some patients find relief from their symptoms by the use of a fitted grade two compression garment. We will refer your child to your local Orthotics Department to be accurately measured and fitted. We will often invite you back to clinic after three to six months to see if the garments are helping your child's symptoms.

Interventional treatment

If your child's malformation is causing them significant problems and it is agreed that interventional treatment would be the best option we will place them on our waiting list for treatment. It is important you understand that the treatments we offer are for symptom control, we cannot cure malformations at present.

Surgical Treatment

If your child's malformation is causing them significant problems and it is agreed, after MDT discussion, that it would be best treated by surgery then we will arrange for your child to be referred to the appropriate service. We will discuss this further with you and your child in clinic.

Symptom diary

In the interim between referral and treatment we may get you and your child to log the severity and location of pain and swelling, as well as any other problems they may have related to their malformation. If you are given a diary please remember to bring it with you to show to the doctor on their treatment date or when they attend clinic.

What does the treatment involve?

We would normally admit children to a ward when we treat lymphatic malformations. This is due to the fact that most children may find the procedure difficult to tolerate if they were awake and therefore benefit from a general anaesthetic. However as children get older we find they become able to tolerate uncomplicated procedures under a local anaesthetic and would therefore be offered this as an alternative route to general anaesthetic.

When we are ready to carry out your child's procedure we will call the ward and let them know. On arrival into Radiology Theatres your child will be clerked in by one of our interventional radiology team, we will need to put your child in a hospital gown; we will also insert a cannula (plastic tube) into their arm to administer drugs and fluids if needed. Finally the consultant interventional radiologist performing the

procedure will confirm your consent for your child to undergo the procedure and answer any further questions you or your child may have.

When your child is ready we will take them into theatre to have their procedure, they may take a toy in with them or any comforter and one of their parents or guardians can come into the room with them whilst they are settled and given their general anaesthetic.

Sclerotherapy

This is the usual course of treatment for lymphatic malformations; the aim of this treatment is to cause the malformation to shrink in size and reduce symptoms. Sclerotherapy involves injecting a liquid agent or sclerosant into the malformation by placing needles directly into it.

The needles are placed into the malformation under ultrasound guidance. Percutaneous sclerotherapy normally takes between thirty minutes to an hour to complete. After the treatment children will normally be recovered in our recovery area and then go back to their ward. They may have a compression dressing on the treated area; this can normally be removed after 24 to 48 hours. The area that has been treated may feel lumpy or harder to the touch this is normal and should settle down after a couple of weeks.

Sclerotherapy is a safe and effective procedure, but as with any medical procedure complications can arise.

Increased swelling and pain will normally occur post procedure. The swelling and pain should generally settle down after a few weeks, during this time simple pain killers such as paracetamol or ibuprofen can be used to control

the pain. We will contact you to make sure the pain and swelling post procedure is not excessive. If your child does not experience some swelling and pain it may be an indication that the treatment may not have worked.

If the malformation is close to, or just under the skin, there is a small risk of skin blistering and ulceration. Most ulcers will heal of their own accord or may require simple bandaging, this depends on their location. However, rarely a skin graft may be required to treat damaged areas of skin.

Occasionally, nerve damage can occur if the malformation is close to a nerve. This may be in the form of a feeling of numbness, tingling or pins and needles and is usually temporary. However, rarely the nerve may be heavily bruised causing longer term symptoms or the damage could be permanent.

There is always a small risk, as with any procedure involving needle insertion, of bleeding and infection. Risks are minimised by using image guidance to aim the needle into the correct area and by using a sterile technique.

Some sclerosants can cause discolouration of the skin, particularly if injected into areas of the malformation that are close to the skin surface. This will normally fade with time, but rarely can leave some permanent discolouration.

Sclerotherapy with Picibanil (OK432)

This is the sclerosant commonly used in the treatment of lymphatic malformations. Picibanil (or OK432) is a penicillin-killed bacterium. We inject this drug in a similar manner to other sclerosant agents. Needles are placed into the malformation under ultrasound guidance and fluid is aspirated

to confirm if the malformation is lymphatic in nature. This is followed by injection of OK432 directly into the malformation.

Injection of OK432 is a safe and effective procedure, but is associated with similar side effects and complications to other sclerotherapy agents as outlined above. It can, however, be associated with more prolonged inflammation and swelling.

If your child has or has had an allergic reaction to penicillin they may not be able to have treatment with OK432; we may need to arrange an allergy test before we can use it to treat them.

They will probably feel like they have mild flu-like symptoms for a week or so. This is completely normal and is a result of the treatment. However if these symptoms become excessive with resultant temperature and fever you should contact us or their GP.

Sclerotherapy with Doxycycline

Doxycycline is an antibiotic that has been used in the treatment of lymphatic malformations. We will inject this drug in a similar way to other sclerosant agents; we may also inject it via a larger tube inserted directly into the area of abnormality. This will be discussed with you at clinic.

Doxycycline is not advised to be used on patients with liver or renal problems. Doxycycline can cause darkening of the colour of your skin, nails, eyes, teeth, gums, or scars.

Doxycycline may cause your child's skin to be more sensitive to sunlight than it is normally. Exposure to sunlight, even for brief periods of time, may cause a skin rash, itching, redness or other discoloration of the skin, or severe sunburn. It is advisable therefore that your child:

- Stays out of direct sunlight, especially between the hours of 10:00am and 3:00pm, if possible for five days post procedure.
- Wear protective clothing, including a hat. Also, wear sunglasses that protect against UVA and UVB rays.
- If they go outside, use a high quality sunblock lotion. They should continue to use this for three weeks after.
- Apply a sunblock product that has a sun protection factor (SPF) number of at least 30. Some patients may require a product with a higher SPF number, especially if they have a fair complexion. Make sure you use a quality sunblock covering both UVA and UVB. If you have any questions about this, check with your doctor.
- Apply a sunblock lipstick that has an SPF of at least 15 to protect your lips.
- Do not use a sun lamp or tanning bed or booth.

Sclerotherapy with Bleomycin

Bleomycin is a medicine that has been used for many years in the treatment of other illnesses, we may suggest this treatment if your child has a lymphatic malformation in a region we would be keen to avoid any swelling, such as around the eye for example or if previous therapies have failed. We have a separate information leaflet for this type of sclerotherapy.

Other risks

Radiation

Sometimes we use X-ray to visualise the blood vessels during the procedure. Radiation doses for the procedure are generally accepted to be low, though occasionally higher doses are required if the procedure is long or demanding.

Compartment syndrome

Sometimes if the malformation is located in a certain area of the body swelling post procedure may cause a condition called compartment syndrome. This is caused when swelling occurs in an area where there is not much room for the swelling to take place without impacting on other structures in the body, the forearm for example. We will often arrange for your child to take a small dose of steroids before and after their treatment to combat this. This will depend on their age.

Follow up

Before your child commences treatment we will ask you how is best for us to follow them up.

We have three methods

- **By telephone:** We will take the best contact number for yourself.
- **By email:** We will take the best contact email for yourself.
- **By virtual clinic:** We can talk to you via a private and secure video link. Again we will need your email to arrange a convenient time to talk.

We will contact you one week and then eight weeks after your child's procedure to see how they are recovering.

At one week we will ask a number of questions which will enable us to establish how they have been after the treatment and make sure your child is not having any problems post procedure or suffering from any complications.

At eight weeks we will discuss if their symptoms have improved or not. We will use this information to determine the next best course of action for your child; this is normally either further treatment or a clinic appointment.

If you have any questions or concerns in the meantime please contact:

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Please note, if your child is feeling unwell or you are unable to contact us regarding any urgent concerns or complications please attend your local GP Surgery, local walk in centre or your local Accident and Emergency Department if appropriate.



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