KK Women's and Children's Hospital
SingHealth

Lymphatic Malformations

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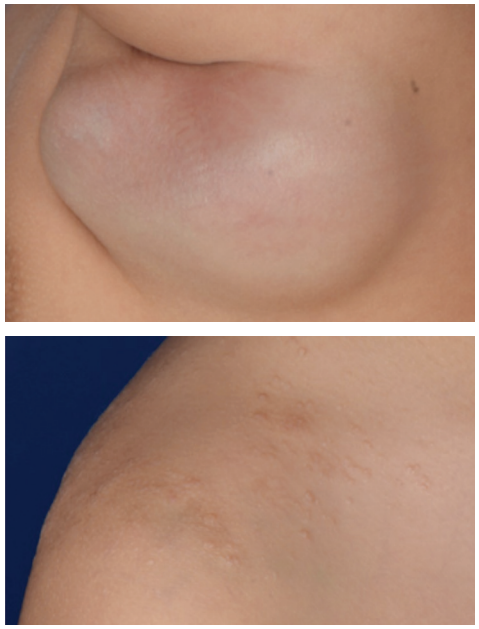
What are lymphatic malformations?

Lymphatic malformations (LM) are abnormally developed clusters of lymphatic vessels that may or may not communicate with normal lymphatic vessels.

They are fluid-filled spaces containing lymph fluid and can consist of large spaces (macrocystic LM), multiple tiny spaces (microcystic) or a combination of both (mixed).

When a combination of LM and venous malformation (VM) are found in the same lesion, these are called veno-lymphatic malformations.



Lymphatic malformation

What do lymphatic malformations look like?

LM can occur anywhere in the body and are present at birth, although they may not become apparent until later in life.

LM appear as skin-coloured soft swellings, sometimes with tiny bubbles or vesicles over the skin. There may be a bluish hue when there is bleeding into the lesion from an adjacent vein. They usually do not feel warmer than surrounding skin.

LM usually grow in proportion with the child's growth. They may become more obvious after episodes of local trauma, or hormonal changes occurring at puberty or during pregnancy.

LM can become larger and warmer after vaccinations or infections (e.g. flu, bacteria) as a result of increased lymph fluid flow due to activation of the body's immune system.

LM are not known to become cancerous.

What problems can lymphatic malformations cause?

Depending on their location, LM may cause pain/numbness, swelling, restriction of movement, breathing difficulty or cosmetic concerns. LM can occasionally become infected causing fever, pain and swelling.

Occasionally they can leak fluid, sometimes blood stained, especially if they are located superficially.

Rarely, LM may be part of a vascular malformation syndrome e.g. Klippel-Trenaunay syndrome (KTS) where the affected limb can become larger or longer than the normal limb and there may be abnormal large "anomalous" vessels that may clot easily.

How are lymphatic malformations diagnosed?

LM are diagnosed with a combination of clinical history, physical examination, imaging including ultrasound, magnetic resonance imaging (MRI), and rarely, tissue biopsies.

Ultrasound is a useful non-invasive, painless test that can be performed either in the clinic or at the diagnostic imaging center. It involves using a probe placed on the skin over the site of the suspected LM. Depending on the size of the lesion, this may take 10 to 30 minutes and will require some cooperation from the child. Ultrasound may be used on its own or with other investigations to confirm the diagnosis of LM, as well as to assess the suitability for treatment by injection sclerotherapy or surgical excision.

MRI provides more detail than ultrasound and can objectively demonstrate the size and extent of the lesion. There is no radiation involved. However, the child needs to stay still for about 45 - 60 minutes, sometimes longer. As the movement will affect the image quality, general anaesthesia (GA) is required for infants and younger children who are unable to cooperate. GA is administered by a team of paediatric anaesthetists.

Tissue biopsies are rarely required for the diagnosis of LM. If required, this involves cutting the skin and removing a small piece of tissue to look under the microscope. This procedure may be done under local anaesthetic if the child is cooperative. Otherwise, it can also be performed under sedation in the ward or under GA in the operating theatre.

How are lymphatic malformations treated?

Treatment for LM may be indicated for associated functional problems or to improve the appearance.

Treatment options for LM include conservative management, oral medications, sclerotherapy, lasers, surgery or a combination of these.

If there are no symptoms or the symptoms are mild, treatment may not be necessary. However, the child will be reviewed regularly.

Conservative management includes pain relief with anti-inflammatory medications, compression garment (if the lesion is in a limb) and alteration of lifestyle accordingly.

LM that are infected may require oral or intravenous antibiotics for treatment. Removal of the infected fluid through the insertion of a tube may be necessary if swelling progresses rapidly.

Definitive treatment for LM is guided by various factors including the size, extent of the lesion, the number of "spaces" within the lesion, the amount of solid tissue component, and proximity to vital structures like nerves, blood vessels or airway.

Sclerotherapy is a type of treatment that involves the injection of a special chemical into the LM to incite swelling initially but ultimately shrink the lesion.

Depending on the site, size and complexity, certain lesions are also suitable for surgical removal.

Some LM that are not easily treated by sclerotherapy or surgery may be treated with an oral medication called sirolimus.

Lasers may be a useful "add-on" treatment for LM that have a superficial component.

The various treatment options will be discussed with the patient and caregiver at the Multidisciplinary Vascular Anomalies Clinic.

Useful telephone number

Central Appointments 6294-4050

KK Women's and Children's Hospital
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KK Women's and Children's Hospital
100 Bukit Timah Road
Singapore 229899
Tel: 6-CALL KKH (6-2255 554)
Fax: 6293-7933
Website: www.kkh.com.sg
www.facebook.com/kkh.sg

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