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Lymphatic Malformations (LMs)

A lymphatic malformation (LM) is a misshaped lymph vessel that traps lymph fluid. The trapped fluid forms a pocket, or cyst, which may gradually or suddenly grow. An enlarged LM may put pressure on nearby organs.

Your doctor may use Ultrasound or Body MRI to diagnose and evaluate your condition. Not all LMs require treatment. If it is not causing a problem, you and your doctor may decide to watch it over time. If you do need



treatment, options include antibiotics, sclerotherapy, surgery, laser therapy, and compression therapy.

What is a lymphatic malformation?

The lymphatic system is part of the body's immune system. It helps fight infection and other disease and keeps the fluid balance in the body. A network of small tube-like vessels throughout the body collect extra watery fluid (called lymph) and carry it to lymph nodes and eventually back into blood circulation.

A misshaped lymph vessel that traps lymph fluid is called a lymphatic malformation (LM). This trapped lymph fluid forms a pocket, or cyst, which may gradually or suddenly grow. Blood from nearby vessels may also enter a cyst. This may cause the area around the LM to swell. An enlarged LM may be pressure on nearby organs.

LMs can appear anywhere in the body. They are most common on the neck, face, and armpit. They may also appear inside the body, in organs or bones.

LMs are congenital, which means the malformation is present at the time of birth.

An LM may be found in a fetus during pregnancy. Others may be found later in childhood, when it appears as a fluid-filled lump.

There are two LM types. Macrocystic LMs may include one or more cysts one centimeter wide or larger. They may be anywhere within the body but are most often found directly under the skin on the neck or chest. They are usually soft to the touch. They may grow larger due to infection or bleeding inside the cyst. Microcystic LMs are a group of smaller cysts that may feel solid. These may appear anywhere in the

body.

The two types of LMs may appear together.

LMs are prone to infection. Nearby blood vessels may also bleed into the malformation, causing it to swell. If an LM becomes enlarged or painful, you should see your physician.

LMs are not hereditary. They are not associated with any other medical conditions. However, the condition is more common in children with:

- Down syndrome
- Turner syndrome
- an overgrowth syndrome (genetic disorders that cause an unusual increase in the size of the body or a body part)
- Noonan syndrome.

Symptoms

If you have an LM, your symptoms will depend on the size of the malformation and where it is located. Symptoms may include:

- A soft, smooth, and discolored lump on the skin. It may be found anywhere on the body including the neck, head, mouth, tongue, eye, chest, stomach, arms, legs, scrotum, and penis.
- A lump or mass that gets larger quickly.
- A lump that shows signs of infection, including redness, warmth, pain, swelling and drainage (rarely).
- Chronic small bumps, blisters, or bloody crusts on the surface of the skin that may rupture and ooze blood or clear lymph fluid.
- An LM in the head, neck or tongue region may make it hard to breathe or swallow.
- LMs in an arm or leg may cause swelling and pain throughout the entire limb.

How is a lymphatic malformation diagnosed and evaluated?

Before a baby is born, an LM may be found during an obstetrical ultrasound (US), which provides pictures of a fetus within a woman's uterus.

An LM on the skin may be found during a physical examination. Imaging is needed to confirm the malformation. Imaging is also used to diagnose an LM that is inside the body.

You may have one or both of these imaging tests:

• Ultrasound: Ultrasound imaging uses sound waves to produce pictures of the inside of the body.

For more information about ultrasound performed on children, visit the Pediatric Abdominal Ultrasoundpage.

• Body magnetic resonance imaging: MRI uses a powerful magnetic field, radiofrequency pulses and a computer to produce detailed pictures of internal body structures. MRI does not use radiation (x-rays).

For more information about magnetic resonance imaging performed on children, visit the Pediatric MRI page.

How is a lymphatic malformation treated?

Not all LMs require treatment. If the LM is not causing problems, you and your doctor may decide to watch it over time. Treating an LM will depend on:

- its location in the body.
- whether it is macrocystic, microcystic, or mixed.
- whether nearby tissues, blood vessels and/or organs are affected.
- the patient's age (for children), medical history and overall health.

Treatment options include:

- Antibiotic medications to treat an infected LM.
- Wearing a tight-fitting garment on the affected body part (if possible) to prevent pain or to prevent the malformation from getting larger (called compression therapy).
- Draining the cysts. For this procedure, a drainage tube is placed through a small skin opening into the cyst(s) using image guidance.
- Sclerotherapy. Your doctor will inject a solution directly into the malformed lymph vessels. This causes them to shrink and collapse. The doctor begins by draining the cyst(s) using image guidance. Sclerotherapy is most often used for macrocystic LMs. Some microcystic LMs also respond to sclerotherapy. Several sclerotherapy sessions may be required.
- Surgery. Your doctor may remove some LMs using surgery. LMs that affect too large an area, such as an entire arm, may not be suitable for surgery.
- Laser therapy. Doctors use a strong beam of light to destroy the LM. Laser therapy is used for LMs of the skin or mouth. Several treatments spaced over several months may be needed. This therapy may be used in combination with other treatments.
- Medical therapy. For extensive or complicated LMs, your doctor may prescribe skin ointments, oral medication, or intravenous medication. Your doctor may also use medication to shrink an LM or to reduce the risk of an LM coming back after surgery or sclerotherapy.

Doctors are investigating other ways to treat LMs, including the use of cold (cryotherapy) or heat (radiofrequency ablation).

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