

Lymphatic Malformations



What is a lymphatic malformation?

Lymphatic malformations are sponge-like collections of abnormal vessels filled with clear fluid. The lymphatic system keeps fluids in balance and helps fight infections, along with the immune system. It collects extra fluid from the tissues and transports it through a series of small vessels back into the bloodstream.

With a lymphatic malformation (LM), the transfer of this fluid through these vessels is slowed or stopped. The extra fluid collects and widens (dilates) the vessels. This causes swelling of the affected area and sometimes enlargement of soft tissues and bones.

Where are LMs found?

They are most often seen in the neck and armpit (axilla), but they can be found in any area of the body. In some areas the dilated vessels tend to be small (microcystic), while in other areas, they tend to be large (macrocytic). Both microcystic and macrocystic malformations may cause enlargement of any part of the body (for example, the lip, cheek, ear, tongue, limb, finger or toe). LMs may be found just under the skin (superficial) or deep. They can be in one place (localized) or widespread (diffuse). When LMs are widespread in bone and soft tissue, the condition may be called lymphangiomatosis.

What causes a LM?

We do not know the exact cause of LMs. They are thought to be caused by errors in the formation of lymph vessels while a baby is developing in the womb. The cause is not related to any known drug or medicine that may have been taken during pregnancy or to any environmental exposures during pregnancy.

We think some LMs are related to a change (mutation) in a gene called PIK3CA. This change is mosaic. It means only cells in the malformation are affected. The change is not seen in other cells elsewhere in the body.

How does a LM grow?

LMs can increase in size or grow as your child grows. They may swell quickly and be painful if your child is sick. When your child is feeling better, the swelling usually goes down slowly. Occasionally, LMs decrease on their own without any treatment.

How is a LM diagnosed?

Doctors will use your child's medical history and a physical exam to diagnose LMs. Your child may also need an MRI (magnetic resonance imaging), US (ultrasound), or CT scan (computed tomography). This is to confirm the diagnosis or to find out the size and how widespread the lesion is.

Is a LM harmful?

A LM can be prone to infection since it is connected to the immune system. LMs can also cause these problems:

- Fluid leakage from the skin. This can cause cellulitis, an infection of tissue in and beneath the skin.
- If cellulitis keeps coming back, it can cause pain, changes in how the skin and body looks, and may lead to serious infection. If this happens, your child will need aggressive antibiotic therapy. They may also be treated with steroid therapy. Steroid therapy can help to decrease inflammation in the malformation.
- Bleeding into the LM which may cause pain and swelling.

There may be other problems depending on the area of the body that is affected by the LM. LMs in the:

- Trachea (windpipe), tongue or chest can make it hard to breathe.
- Gastrointestinal tract (stomach and intestines) can cause protein loss.
- Chest (thorax) can cause chylothorax, which is a leakage of lymphatic fluid into the chest cavity. It can also cause heart and lung problems.
- Stomach (abdomen) can cause chylous ascites, which is a leakage of lymphatic fluid into the stomach.

- Bone can cause bony destruction and pain. When a lymphatic malformation affects nearby bone, it is called Gorham-Stout Disease.

LMs may cause nearby tissues, like skin, muscle, fat or bone to overgrow.

How can we prevent an infection?

Good oral hygiene can help prevent an infection in a LM in the head or neck area. Your child should brush their teeth 2 to 3 times a day and floss once a day. Your child's teeth should be cleaned by a dental hygienist every 6 months. A mouth guard may be recommended to help reduce trauma to the tongue or cheek while sleeping.

Being up-to-date with vaccines can also prevent infection.

How is a LM treated?

Which therapy is offered for your child depends on the location and size of the LM. The methods most often used to treat a LM include:

Surgery

Some LMs, especially those that are localized, can be removed during surgery. Surgery may be used along with some of the other treatment options listed below. Surgery may not be possible if the LM is widespread and surrounds important body parts and organs.

Surgery cannot be done without some scarring. Years after the surgery, lymphatic vesicles may return at the site of a scar. Also, surgery may cause damage to body parts affected by the LM.

Sclerotherapy

During this procedure pronounced SCLAIR-oh-THAIR-uh-pee, a doctor removes the fluid, sometimes with a drain, from the LM then injects a solution to reduce its size by creating an inflammatory response. For macrocystic LMs, sclerotherapy can cause total or near-total shrinkage. This treatment has limited effect on microcystic LMs.

Sclerotherapy may cause superficial or deep tissue scarring. Also, in some cases, the LM may not shrink or it may come back.

Laser therapy

Surgical laser therapy may be used depending on the type of LM. It is often used along with other treatment options. It requires multiple treatments that are spaced over several months. It is done under general anesthesia (a medicine given to make your child sleep without pain during the treatment).

This may cause some tissue scarring and/or changes in the color of the skin. These can sometimes be improved by additional cosmetic procedures.

Medicine

LMs are sometimes managed with medicines. Some medicines are used to treat symptoms, like pain or bleeding, that are related to the LM. Other medicines are used to treat LMs directly. Your healthcare team will watch

To Learn More

- Vascular Anomalies Clinic 206-987-2105
- Ask your child's nurse or doctor
- www.seattlechildrens.org

your child closely while they are taking these medicines. Your doctor will discuss the benefits and risks of each medicine.

When do I call the doctor?

Call the clinic if your child has a rapid increase in the size of their LM or the warning signs of an infection, including fever, swelling, local pain, redness and bleeding.

Free Interpreter Services

- In the hospital, ask your child's nurse.
- From outside the hospital, call the toll-free Family Interpreting Line 1-866-583-1527. Tell the interpreter the name or extension you need.

Adapted with permission from Cincinnati Children's Hospital.

Seattle Children's offers interpreter services for Deaf, hard of hearing or non-English speaking patients, family members and legal representatives free of charge. Seattle Children's will make this information available in alternate formats upon request. Call the Family Resource Center at 206-987-2201.

This handout has been reviewed by clinical staff at Seattle Children's. However, your child's needs are unique. Before you act or rely upon this information, please talk with your child's healthcare provider.

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