

There are 2 types of lymphatic malformation -  
Lymphoedema and Cystic Lymphatic Malformation

### **Lymphoedema**

Lymphoedema is a build up of lymphatic fluid in the tissues. In children, it is usually due to absent or reduced lymphatic vessels in the affected area. Lymphoedema may be obvious at birth or be noticed for the first time in childhood.

Please see the LYMPHOEDEMA fact sheet for more information.

### **What is a Cystic Lymphatic Malformation?**

A Cystic Lymphatic Malformation is a sponge-like collection of cysts either in the skin or beneath the skin. The cysts contain clear fluid called Lymph. The cysts may be large (macro) cysts, small (micro) cysts or a combination of both. If a Cystic Lymphatic Malformation occurs in the skin, it results in fluid filled blisters or lumps that look like warts (but aren't). If it is in the deeper tissues, it looks like a swelling beneath normal looking skin. It may cause enlargement of a normal body part such as the ear, lip, tongue, finger, or a limb.

Old names for Cystic Lymphatic Malformation include "Cystic Hygroma" or "Lymphangioma". Other less commonly used and outdated names include lymphangioma circumscriptum, lymphangioma simplex, verrucous haemangioma and angiokeratoma circumscriptum.

Although Cystic Lymphatic Malformations are always present at birth, they may not be noticed until lymph fluid slowly builds up in them, or suddenly accumulates due to an infection or some bleeding into the cysts.

Sometimes Lymphatic Malformations occur in association with abnormalities of other blood vessels – capillaries, veins, and/or arteries.

### **Why does my child have a Lymphatic malformation?**

The Lymphatic system transports clear, protein rich fluid from the intestine and from body fluids into the blood circulation. The Lymph vessels start in the tissues or intestines and join into the veins at various sites but particularly in the neck, armpits and groins.

Lymphatic Malformations occur when there is an error in the formation of lymph vessels or where the lymph vessels don't connect properly into the venous system. They can occur anywhere in the body but are more common in the neck, groins and armpits.

The exact cause of Lymphatic Malformations is unknown. They are not due to any foods, medications, infections or anything the mother may or may not have done during pregnancy. They are rarely associated with a genetic abnormality

### **Is the Lymphatic Malformation Dangerous?**

Most Lymphatic Malformations are not immediately dangerous and time can be taken to evaluate them and plan treatment.

Lymphatic Malformations may become infected or bleeding may occur into them causing sudden swelling and pressure on nearby structures. If the Lymphatic Malformation is in the neck region, it is important to find out whether it is close to the windpipe (airway) because swelling in this area can cause feeding and breathing problems.

Those in the abdomen or chest may cause problems with absorption of nutrients or a build up of fluid. If the Lymphatic malformation is in the skin, leakage of fluid commonly occurs. This can lead to infection.

### **What tests are needed?**

There may be more to the Lymphatic Malformation than what can be seen on the surface. Ultrasound with Doppler, CT scan or MRI may be recommended by your doctor, to confirm the diagnosis, to show the extent, and to see if there are any capillary, venous or arterial abnormalities associated with it.

### **Can Lymphatic Malformations be treated?**

It is crucial to have the right diagnosis and to assess the extent of the Lymphatic Malformation prior to planning treatment.

It is uncommon to be able to completely eradicate a Lymphatic Malformation but much can be done to reduce their size and to reduce the chance of complications

Treatment depends on the size and location of the vascular malformation and may include sclerotherapy, surgery or a combination of both.

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Please consult with your doctor or health professional to make sure this information is correct for your child.

This document was reviewed on 9 July 2010

A multi-disciplinary team of vascular anomaly specialists including dermatologists, interventional radiologists and surgeons will help to ensure that the best care is tailored to the needs of your child. The team will discuss with you the recommended treatment/s, the possible outcomes and possible risks. You should feel free to ask any questions.

### **Sclerotherapy**

Sclerotherapy involves the injection of an irritant into the cysts. The procedure is usually carried out under general anaesthetic in the X-ray department or the operating theatre. Under ultrasound or radiological guidance, the fluid is drained from the cyst/s. They are then injected with an agent designed to cause irritation and obliteration of the cyst/s. If possible, the area is then kept compressed for some days after the treatment to minimize the re-collection of fluid in the area. Usually, a number of treatments are required. The Vascular Anomalies Team will discuss with you the recommended sclerosing agent.

Sclerotherapy is successful to a degree in most Lymphatic Malformations but results are best in the treatment of large (macro) cysts. There is a small risk of infection, and damage to nearby structures or to the skin. Even after a good result, the cyst may re fill with fluid over time. Treatment can be repeated.

### **Surgery**

Some small and well defined Lymphatic Malformations can be excised. In others, sclerotherapy may be done first to shrink the malformation and make surgery more likely to be successful. Sometimes it is a safer option to 'debulk' the malformation, rather than entirely remove it. For lesions that surround vital structures (e.g. the airway, major blood vessels or nerves) surgery carries higher risks and these areas may be best treated with sclerotherapy or left alone.

An operation will always result in a scar. There is also a risk of bleeding and infection. Even though the malformation may appear to be completely removed, the fluid may re accumulate over time.

### **Surface Treatments**

Lymphatic Malformations in the skin or on other surfaces (e.g. tongue) may be improved by CO2 Laser treatment.

### **What other support is available?**

It is often difficult for parents to deal with their own response to their child's condition, in addition to the response of other people. The Vascular Anomalies Team social worker is available to discuss this and other issues with you, and to help ensure that any questions of the specialist you have, are answered. Practical assistance to do with your hospital visits is also available.

Much internet based information is readily accessible. If you read anything on-line or else where that you would like clarified, please do not hesitate to bring it to the attention of your doctor.

[www.vascularanomalies.org/NOVAAroundtheWorld.htm](http://www.vascularanomalies.org/NOVAAroundtheWorld.htm)

### **Remember**

Every Lymphatic Malformation is different. Most will require some investigations to help confirm the diagnosis and to plan management. Treatment is aimed at improving symptoms and the appearance, and at reducing the risk of complications

Please direct inquiries to:

Vascular Birthmarks Service  
Dept Paediatric Surgery  
Sydney Children's Hospital  
High Street, Randwick 2031  
Tel 02 9382 1776  
Email: [SCHVBC@sesiahs.health.nsw.gov.au](mailto:SCHVBC@sesiahs.health.nsw.gov.au)

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