

What is the lymphatic system?

The lymphatic system is a network of vessels or channels throughout the body. Lymph vessels carry clear fluid (known as 'lymph') from all the tissues of the body, back to the blood stream. Lymph is mainly made up of protein and water. Lymph can also carry any foreign matter that enters the tissues, such as bacteria.

Lymphatic vessels start in almost every area of the body (eg organs, muscles, fat, and skin). Initially the vessels are tiny. They gradually join into bigger channels before emptying the lymph into the blood stream. Lymph is pumped into and along lymphatic vessels by movement of adjacent muscles as well as by contractions in the walls of the larger lymph vessels. Back-flow is prevented by valves inside the vessels. On its way along the lymph vessels, the lymph is filtered through lymph nodes ('glands'). These remove any bacteria and start the fight against infection.

What is Lymphoedema?

Lymphoedema (lymphedema) is a build-up of lymph in the tissues. The fluid and protein enters the tissues in the normal way but the lymphatics (because they are too small or absent or do not work properly) are unable to remove the fluid at a fast enough rate. The fluid causes swelling. If lymphoedema is left untreated, the protein rich fluid damages the tissues, interfering with oxygen delivery to cells and increasing the risk of infection (such as fungal and bacterial infections) and can result in poor healing. Eventually, the swollen tissues become scarred and the swelling may be difficult to reverse. Usually the legs are involved in lymphoedema, but any body part may be involved.

Why does lymphoedema occur?

Lymphoedema may be 'primary' or 'secondary'

Primary lymphoedema is either due to

- an absence or reduction in the number of lymph vessels in the affected area, or
- malformation or malfunction of the lymph vessels that are present.

Secondary lymphoedema occurs when the lymph vessels have been damaged by disease, injury or surgery.

Children with lymphoedema usually have primary lymphoedema. It may appear before or at birth (congenital), during childhood or adolescence, or not until adult life.

Are there any associated problems?

Primary lymphoedema usually occurs in isolation. Very rarely, genetic abnormalities are associated. There are some rare syndromes associated with primary lymphoedema. Sometimes there are abnormal veins and capillaries in the affected area.

Will any tests or investigations help?

Most of the time, the diagnosis of primary lymphoedema in babies and children can be made without the need for tests. Whether the cause is an absence of lymph vessels, or abnormality in lymph vessels, the treatment is the same. Sometimes further tests are advised. Blood tests looking for genetic and protein abnormalities may be recommended. MRI scanning will help determine if there are associated vein and capillary abnormalities. Lymphoscintigraphy can help assess lymphatic function. Your doctor will discuss any recommended tests with you.

What treatment is available for lymphoedema?

Because the lymph vessels are not able to do their job of removing fluid from the tissues, the treatment is aimed at providing alternative ways for this to happen. If the tissue swelling is kept to a minimum, then the risk of complications and long term problems will be markedly reduced.

The recommended treatment is called complex physical therapy which is an active non surgical treatment of lymphoedema. The main components of this are manual lymphatic drainage, compression therapy, skin care and exercise.

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

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Lymphoedema

The manual lymphatic drainage is a specialised massage technique that should be performed by a therapist with specific training.

Compression therapy involves multilayered bandaging, and custom made compression garments for the specific area of lymphoedema.

Meticulous skin care: Skin hygiene is extremely important. Shower or bathe daily and apply moisturiser regularly to prevent drying and cracking. Any sign of infection should be treated immediately with antibiotics.

Protect skin from cuts, scratches, insect bites and sunburn.

Avoid needles for injection or taking of blood from affected limb.

Treat tinea (fungal infections such as athlete's foot or "ringworm" or nail infection) immediately.

The exercise program is designed by the child's therapist and is age and ability dependent. Keep weight in normal range. Lymphoedema can worsen in hot conditions so it is important to avoid overheating.

This specialized complex physical therapy treatment is available at Sydney Children's Hospitals Randwick and Westmead. The physiotherapist will be able to explain, start and maintain treatment and teach parents/carers how to undertake some aspects of the therapy. Often it is possible to arrange some support closer to home for ongoing care but regular review at the Children's Hospital will usually be required.

Will my child be able to lead a normal life?

Children should be encouraged to pursue normal activities, as exercise is very beneficial and can assist with maintaining normal weight and fitness. Children can exercise and play sport while wearing the compression garments and this can be monitored by their therapist.

What about the long term?

Lymphoedema is a condition that does not go away. It will require active therapy through out life.

At present there is no cure but when managed well and consistently, the amount of fluid in the tissues can be kept to a minimum and the secondary effects prevented or reduced.

Constant vigilance is needed to reduce the risk of injuries and infection.

Skin care

If redness or increased weeping occurs from the affected area, medical advice is needed. Any sign of infection should be treated immediately with antibiotics.

Skin hygiene is extremely important as the skin is the main barrier to infection. Use moisturiser regularly to prevent drying and cracking.

Keep weight in normal range.

Compression garments should be worn to maintain the limb/limbs and prevent increases in swelling.

Compression garments **MUST** be worn for air travel.

Please direct inquiries to
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