

## CHAPTER 20 LYMPHEDEMA DIAGNOSIS AND THERAPY

**Original authors: Audrea Cheville, Cindy Felty, Gail L. Gamble,  
Peter Gloviczki, Thom W. Rooke, David Strick  
Abstracted by Raghu Motaganahalli**

### Introduction

**Lymphedema** is a condition in which an excessive amount of **lymph** (liquid with some solid parts) collects in one place in the body, causing **edema** (swelling) and later if not properly treated, skin problems. **Lymphedema** occurs most frequently in the arms and legs. **Lymphedema** is most often a chronic condition and rarely cured, but it can generally be controlled by carefully following a treatment program.

### Lymphatic System

The **lymphatic system** plays a very important part in keeping your body healthy. It works as part of your body's cleaning system, by collecting and removing **lymph** fluid (which contains proteins, fatty acids, waste from cell feeding and bacteria that enter the body) from the body. **Lymph** fluid is taken up in **lymphatic vessels** and passes through **lymph nodes** where harmful substances are destroyed and cleaned by special infection-fighting cells that are present in our body (by the **immune system**). After **lymph nodes** have filtered the **lymph** going through them, the **lymph** fluid returns to join the **blood** pumping through your **veins**. When this protein-rich fluid is not removed because of damage to the **lymphatic vessels** themselves or the **lymph nodes**, the result is **lymphedema** or swelling of the leg or arm. If the **lymph vessels** do not develop normally, or if the **lymph nodes** or **lymph vessels** have been removed or damaged (such as with radiation from cancer treatment, or from repeated infections in the leg or arm), **lymph** fluid may collect (stay) in the soft tissues of the affected body part causing **lymphedema**.

### Types of Lymphedema

There are 2 types of lymphedema: **primary lymphedema** and **secondary lymphedema**. **Primary Lymphedema** occurs when the development of **lymph vessels** and/or **lymph nodes** is abnormal. This condition may be present at birth, but may only show up as **edema** at the time of puberty or even later in life. **Primary lymphedema** is much more common in women and occurs most often in the legs. A significant number of patients with **primary lymphedema** (perhaps as many as 10%) are thought to have chronic **reflux** (back flow of lymph fluid in the **lymph vessels**). This includes patients with congenital **lymphatic hyperplasia**, which in its most extreme form is sometimes known as "**megalympathic**" disease. There are 3 types of **primary lymphedema**. **Congenital lymphedema** presents at birth and accounts for about 10% of all cases. **Lymphedema praecox** shows up between the ages of 2 and 25 years old and represents about 80% of

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all **primary lymphedema**. Finally, **lymphedema tarda** is generally noted first after the age of 35 and accounts for 10% of these cases.

**Secondary Lymphedema** has 2 causes. The first is obstruction or blockage of **lymph** fluid movement in the **lymph vessels** or **nodes**. The cause can be injury, infection, blockage from tumor invasion, radiation therapy, or by surgical removal or damage of the **lymph nodes** or **lymph vessels**. Surgical damage of the **lymph system** may be the most common cause in the United States. Scarring of the **lymph vessels** or **nodes** is the final result of most causes. The most common cause of **lymphedema** world wide is **filarial disease**, caused by infecting worms. These are parasites (worm type invaders) that get into the patient and damage the **lymph vessels** as well as causing other problems. The world health organization estimates that the number of people affected worldwide by this problem may approach 100 million. A less common cause of **lymphedema** is overproduction of interstitial / **lymphatic fluid**, the second cause of **secondary lymphedema**. High body temperature, injury or inflammation (infection) are thought to produce **lymphedema** by this mechanism. Most of these conditions resolve in time and so does the **lymphedema**. Chronic overload occurs when the **lymphatic system** is repeatedly damaged over time due to recurrent infections, chronic **vein** problems, or obesity. These types of overload to the **lymphatic system** can cause **lymphedema** to progress more quickly.

### **Symptoms of Lymphedema**

There are four **stages of lymphedema**. In the **first stage**, the body can make up for any decreased movement of **lymph fluid** so no physical changes are seen (**latency stage**). Stage 2 is called “**reversible lymphedema**” and even though swelling is seen no actual damage has occurred to the body and so the swelling can be removed by medical means. Stage 3 (called “**spontaneously irreversible lymphedema**”) means that actual damage (scarring or fibrosis) has occurred to the body and the only way to change the situation is by surgery. The final stage is called “**lymphostatic elephantias**”, or elephant like appearance, with severe thickening and hardening of the tissues to the point of disfigurement. What the doctor and patient sees with **lymphedema** is swelling in the leg, arm or other body part, limited movement, feeling of heaviness, tightness or aching, recurrent infection, and hardening and/or thickening of the skin. **Lymphedema** does not cause focal pain (pain in a localized, specific area). You should contact your doctor if you are having pain, weakness, or numbness in the affected body part.

### **Diagnosis of Lymphedema**

The **diagnosis of lymphedema** is based on a history of progressive and non-painful swelling especially into the foot or hand if the limbs are affected. The physical findings are swelling in the leg, arm or other body part, limited movement, feeling of heaviness, tightness or aching, recurrent infection, and hardening and/or thickening of the skin. An **ultrasound study** uses sound waves to bounce off underlying structures to make sure that the **edema** that is present is not as a result of a new **blood clot** or other damage to the **vein**. **Magnetic resonance imaging (MRI)** is a study using magnets to see into the body

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and can help to prove that the type of tissue involved is in the fatty tissue under the skin and is not a **vein** related problem. **Lymphoscintigraphy** is the most common test use to show that the **lymphatic system** is the problem. The study involves giving an injection of a small amount of **technetium 99m-labeled colloid** (a substance that can be seen by this type of study) into the space between the toes and takes pictures of the leg or arm or other affected part of the body as the colloid is taken into then **lymph system**. This test is very sensitive for diagnosing **lymphedema**. The **lymph system** is followed for up to 24 hours or until the medication is taken up in the liver. The test shows if there are areas where the medication is not taken up indicating obstruction or damage to the **lymph vessel** system. **Lymphangiography** is not used in most cases since it can cause **lymphedema** but in certain cases it is the only test that can actually show the anatomy of the **lymph vessels**. It is usually done on patients with **chylous reflux** or abdominal or **thoracic chylous fistulas** or connections in the abdominal or chest cavity that makes **lymph fluid** come in contact with parts of the body that it usually does not connect with. If actually surgery on the **lymphatic system** is planned (microsurgery), a **lymphangiogram** may be required.

### **Medical Treatment of Lymphedema**

Early treatment is key in achieving long-term control of **lymphedema**. The medical treatment of **lymphedema** involves two phases: **reduction therapy** which focuses on actively decreasing the swelling and maintenance therapy which is used to maintain or keep the reduced size gained in the reduction phase. **Reduction therapy** involves aggressive wrapping of the limb with **compression bandages** sometime of a stiff material or sometimes with elastic material or a combination of both. **Short stretch or low stretch (not elastic) wraps** are most often used since the **compression** is more constant especially when the patient walks. **Short stretch wraps** have the added benefit of being able to mold most closely to the shape of the affected body part so all of the swelling can be reduced. **Multilayered techniques** are the most popular used. In addition, specially created padding using cut pieces of foam can be applied to specific areas that might have more thickened skin to help remove the protein rich tissue that has accumulated. The idea is to push the fluid out of the affected part by applying the wraps during the entire day with rewrapping whenever the **compression** seems less tight. By measuring the size of the limb frequently during this phase of treatment, the limb can be decreased to its near normal size when compared to the other limb (leg or arm). By compressing the limb the fluid is forced back up into the trunk where it can be reabsorbed by the body. The wraps should only be taken off just before bathing or just before a **massage therapy** session. Exercise is important during **compression** treatment to help get the fluid out of the affected part of the body. A therapist will give the patient special instructions on best exercise. Skin care is also an important part of the care of patients with **lymphedema**. Pressure and friction from the wraps and the appearance of dry skin as the swelling decreases may contribute to problems. The patient must pay close attention to dry or cracked heels, toes or fingers and if prone to athletes foot, the use of an anti-fungal powder or cream to skin creases such as between toes, and skin folds daily or more often may help to decrease infections. A daily moisturizer to prevent dry skin may also help. If you notice a scratch or break in the skin of the affected limb, ask your doctor if an antibiotic ointment applied to the skin might be helpful.

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In addition, there are many **lymphedema** treatment programs available in the US. The therapists are specially trained to use a special type of **massage** to decrease lymphedema swelling. This treatment requires certification after special training. This technique called **manual lymph drainage (MLD)** is a type of specialized **massage** which is different from **deep congestive massage** for reducing **lymphedema**. The technique encourages the natural flow of **lymph** fluid out of the affected limb by first clearing out the **lymph vessels** further up the limb before encouraging the **lymph** fluid at the most distal part of the limb. The therapist uses a specific set of hand movements and sequences to gently move the fluid away to more healthy **lymph vessels**. Stimulating (rubbing) the **lymph vessels** allows them to function more normally and filter and remove the excess fluid. Areas of skin may be especially thick with (protein) fibrinous tissue. The **MLD** technique can soften these areas to become more like normal skin. You or a family member may be taught to use this technique at home although it can be challenging to do it on yourself. **MLD** is not used as a therapy alone. It is most often combined with other therapies including **compression wrapping**, exercise, garments, and skin care.

Before **MLD** became popular, **compression pumping** was one of the main treatments for **lymphedema**. **Pumps** are available that can produce external pressures high enough to push fluid from a limb (arm or leg) and yet gentle enough not to cause harm to tissue. The use of the **compression pumps** is controversial by those who believe that **MLD** is the best way to gently encourage the fluid to move out the limb. There is concern that the **pump** pressures are too high and may be traumatic to the tissue. There are newer **pumps** that may actually mimic the motions of **MLD** and are programmed to do so. In addition, the ability to manipulate every phase of the **pumping** sessions by adjusting the pressures and timing sequences make **compression pumping** attractive to those with busy lifestyles. The **pumping** devices are compact enough to carry and use during travel allowing patients with **lymphedema** to have a lifestyle not dependent upon a therapist or family member to help handle their daily routine of **edema** reduction. Patient's experiencing this type of **pumping** session which lasts about 1 hour per limb report that the motion is quite tolerable and evidence has suggested that it is very effective. Your therapist will help you make decisions about which is the best approach for you.

The **maintenance phase** of therapy begins after the affected limb is reduced as much as can be expected (determined by daily measurements of the limb by the therapist). The goal of **maintenance therapy** is to prevent the limb from swelling again. **Compression** garments play a critical role in preventing swelling from happening again. The patient is given a prescription for a **compressions stocking**. It is important that the stocking fit the size of your limb with all the **edema** out since fluid will again appear if the stocking is too big. An experienced fitter should measure your limb. You should not stop wearing your **compression wrapping** until the stocking is available. The stocking should fit snug but should not be painful. You should put the stocking on as soon as you awake from sleep and preferably before you get out bed in the morning and should only take it off when you are ready to go to bed. **Compression stockings** can have pressures of 20-30 mmHg (for arm **lymphedema**) and 30-40 mmHg or even up to a custom made stocking providing 50 mmHG of pressure for lower extremity **lymphedema**. The pressure is

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determined by the severity of the **lymphedema** and the previous success with lower pressure **compression stockings**. The stocking should be replaced every 4-6 months because they wear out. The patient should have 2 garments at the same time, so that one can be worn while the other is being washed. Always put the garment on using rubber or similar gloves to avoid putting a hole into the garment. The garment loses its **compression** once a hole gets in it. If 10-15 pounds or more are lost during the **maintenance phase**, a re-measured stocking is likely needed since the stocking will not be fitting well. If the affected limb begins to get larger despite your wearing the compression garment you should contact your therapist. Re-measurement is needed every year to make sure that the fit is still correct. Sometimes there may be additional thickened skin and fluid that could be reduced out of the limb in the year following a reduction phase. This may happen as the body remodels itself. There might be consideration given to another reduction phase of treatment if this does occur.

In addition to this standard program, you doctor may have you use **compression wrapping** at night to help reduce swelling even further. It is very important to continue exercising for best decrease in swelling and to improve the general quality of life. Walking, swimming, and bicycling are all good exercises to strengthen your muscles, help you to maintain a healthy weight and also help to keep **edema** from returning the affected limb. You should always wear your **compression** garment or wrap when you exercise. Elevation of the affected limb or body part allows gravity to help in draining the fluid especially at the end of the day. Raising the affected limb higher than the level of the heart is the recommended height. Some patients need **compression pumping** or **MLD** periodically or at night to keep the limb size down. Skin care is as important in this phase of treatment as during the reduction phase. Additional hints to improving the chances of having the best results are to avoid extreme temperatures such as getting into hot tubs or saunas. These can allow dilation of **blood vessels** and fluid to come back into the leg. Having an antibiotic available to be used at the first sign of infection may help to decrease recurrent problems with skin and fatty tissue infections. There are not proven effective drugs to treat lymphedema.

### **Surgical Treatment for Lymphedema**

A variety of surgical techniques have been proposed to treat patients with **lymphedema** especially when it has become too difficult to manage by medical therapy alone. The type of **lymphedema** may determine if a surgical treatment is possible for your condition. **Excisional operations** remove (cut out) excess tissue to help decrease the size of the limb. The most extensive **excisional operation** is the **Charles's procedure** which removes all the skin and subcutaneous tissue from the knee down to the ankle. This procedure may be done when there have been multiple infections in the skin and fatty tissue under the skin, skin sloughing or the patient is having difficulty in performing normal daily functions. Reducing the bulk of skin is usually helpful but this invasive procedure can have complications such as poor wound healing, prolonged hospitalization, long scars, changes in feeling, and swelling in the foot and ankle. A less extensive form of surgery to reduce **lymphedema** tissue is **liposuction**. **Lymphedema** tissue in time turns into fat which can not be helped by **compression** so **liposuction** does this for the

body. In rare cases and when there are nicely open **lymph vessels** above and below an area of damage, **lymphatic reconstruction** or direct reconnections of the **lymph vessels** to each other or a vein is possible. These are called **lymphovenous anastomosis** or **lymphatic grafting** and use microsurgery. It is noted that this procedure is most useful when done early in the course of **lymphedema**, before subcutaneous fibrosis occurs and **lymphatic vessel** sclerosis or hardening begins and the **lymphatic vessels** still have the ability to contract. It has been done in patients with surgical damage to **lymph vessels** (such as after breast cancer surgery), **chylus reflux**, and in people affected with **filariasis** who often have very large **lymph vessels**. It is very delicate work working with very small vessels so it is done only in very select cases and the results are not well known.

## Commonly asked questions

### What is lymphedema?

**Lymphedema** is swelling in the leg, arm or other parts of the body due to the body's inability to effectively remove **lymph** fluid (which contains proteins, fatty acids, waste from cell feeding and bacteria that enter the body).

### What can cause lymphedema?

**Lymphedema** can happen because the formation of the **lymph vessels** during maturation (as a preborn baby) was less than adequate (called **primary lymphedema**) or damage (injury) has occurred to the normally formed **lymph channels** and **lymph nodes** during surgery, from infection, from radiation treatment or other causes. It can also occur from too much **lymph** fluid being made such as can happen when **venous disease** is present. Because the **lymphedema** caused by damage or overproduction is the result of something other than a problem with how the **lymph vessels** matured it is called **secondary lymphedema**.

### What can I do if I have lymphedema?

Your doctor can make sure that you actually have **lymphedema**. If you do and there is no infectious or other acute medical problem that must be dealt with, then external **compression** treatments are begun to decrease the swelling and to keep it down. This may involve external **massage**, **compression pumping**, **compression wraps** and eventually **compression stockings**. How well you do will be determined by how faithfully you use the treatments prescribed.

### Is surgery for lymphedema a good option?

Surgery for **lymphedema** is really a last option for only a select group of patients. Medical treatment is the main stay of treatment and surgery is only considered if this fails. An in-depth discussion of this option with a vascular surgeon would be the best way to find out if you are a candidate for surgery.

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