Lymphedema: Everything You Need to Know

What is Lymphedema?
Lymphedema is an abnormal accumulation of protein rich fluid that causes chronic inflammation and reactive fibrosis of affected tissues. It can interfere with wound healing and cause significant damage to the skin.

Symptoms of lymphedema include swelling of a body part or parts, skin changes, discomfort, restricted range of motion and pitting or non-pitting edema.

Lymphedema can occur virtually anywhere in the body, but is usually seen in the arms or legs. If left untreated, lymphedema can cause:
✓ a progressive hardening of the affected area
✓ increased size of an extremity
✓ hardening of dermal (skin) tissues
✓ hyperkeratosis
✓ papillomas

• Affecting over 2.5 million Americans, it remains poorly understood in the medical community and can have a profound impact on one’s quality of life.
• The diagnosis of lymphedema can be made by medical history, physical exam, and comparative clinical measurements of the affected and non-affected limb.
• Lymphedema can develop when lymphatic vessels are missing or impaired (primary), or when lymph vessels are damaged or lymph nodes have been surgically removed (secondary).
• Lymphedema is a chronic condition, which means that it can be a lifelong challenge. It can be managed, but it is neither curable nor life-threatening.
• Lymphedema should not be confused with edema resulting from venous insufficiency/poor circulation. However, untreated venous insufficiency can progress into a combined venous/lymphatic disorder which is treated in the same way as lymphedema.

Who’s at Risk?
There are a few factors that can put someone at risk of developing lymphedema:
• Anyone who has had gynecological, melanoma, prostate, or kidney cancer in combination with lymph node dissection and/or radiation.
• Anyone with a history of cellulitis.
• Those with certain preoperative and postoperative factors such as a high body mass index (obesity), tumor location, delayed wound healing, postoperative infection, and postoperative hematoma or seroma.

While at-risk people can incorporate risk-reduction measures into their daily lives, even the most watchful person has no guarantee that lymphedema will not occur.
The Normal Lymphatic System

Consisting of microscopic lymph vessels, nodes and lymphoid tissues, the lymphatic system is part of the circulatory system of veins and arteries. The role of the normal lymphatic system is to remove impurities from the body’s tissues through the circulatory system and to produce cells of the immune system (lymphocytes) that are vital in fighting bacteria and viruses. These important lymphocytes, produced in the lymphatic system, are a crucial part of the immune system.

Lymph vessels are microscopic channels that contain colorless fluid called lymph. The lymph is comprised of white blood cells and waste products from tissue/cells. Lymph fluid passes through nodes, or valves, located in the lymph channels at 1 – 2 cm intervals. As the fluid passes through the nodes, it is purified of harmful bacteria and viruses. The 2 largest concentrations of lymph nodes are in the head and neck region (approximately 170 – 200) and in the deep abdominal region (approximately 400 – 700). Smaller accumulations of nodes are present in the axilla (armpits) and inguinal folds (groin). Not all persons are born with the exact same number of lymph nodes. It is believed that most individuals have approximately 500 – 1500, some as small as a sesame seed and others as large as an olive.

What Causes Lymphedema?

**Primary Lymphedema:** Lymphedema can occur from idiopathic (unknown) causes. This is called primary lymphedema and has been known to affect from one to as many as four limbs and/or parts of the body. Primary lymphedema can be present at birth, or associated with arterial-venous abnormalities (AV malformations) such as hemangioma, Port Wine Stain, Klippel Trenaury, and Parker-Weber Syndrome.

The most common form of primary lymphedema is lymphedema praecox which typically appears during puberty, primarily in girls, and usually affecting one lower extremity. A second form of primary lymphedema is lymphedema tarda which can occur in both male and female adults, typically between ages 25 – 35.

**Secondary Lymphedema:** Secondary lymphedema, or acquired lymphedema, can develop as a result of surgery, radiation, infection, or trauma. The number one cause of lymphedema in the US is treatment for cancer. This includes certain surgeries, radiation, and some chemotherapy. Other non cancer-related surgeries (as well as trauma and infections) which inadvertently or accidently disrupt lymphatic pathways, can result in lymphatic impairment and lead to the eventual onset of lymphedema.

Lymphedema can develop immediately post-operatively, or weeks, months, even years later. It also can develop when chemotherapy is unwisely administered to the already affected area (the side on which surgery was performed) or after repeated aspirations of a seroma (a pocket of fluid which occurs commonly post-operatively) in the axilla, breast, or groin area. This often causes localized infection and, subsequently, lymphedema. For some patients, taking tamoxifen, a medication used for breast cancer prevention, can contribute to the onset of lymphedema.
Radiation Therapy is used in treatment of various cancers and AIDS-related diseases (such as Kaposi-Sarcoma), can damage otherwise healthy lymph nodes and vessels, causing scar tissue to form, interrupting normal flow of lymphatic fluid. Radiation can also cause skin dermatitis or burn similar to sunburn.

**What are the Symptoms of Lymphedema?**

- Lymphedema can develop in any part of the body. Signs or symptoms include:
- Sensation of fullness or heaviness in a limb(s)
- Tightness of skin, decreased flexibility in a joint
- Difficulty fitting into clothing in one specific area
- Tight feeling in ring, watch or bracelet

**If you notice persistent swelling, it is important to seek immediate medical advice, as early diagnosis and management generally improve the outcome.**

**Lymphedema Develops in a Number of Stages, From Mild to Severe**

**Stage 0:** The lymphatic system is becoming impaired but swelling may not be evident. Most patients are asymptomatic but some report a feeling of heaviness in the limb. Stage 0 may exist for months or years before the onset of visible lymphedema.

**Stage I:** (spontaneously reversible): There is an accumulation of protein-rich edema fluid causing swelling. Swelling will reduce (lessen) with elevation. There is a “pitting” or indentation when pressed by fingertips. A limb may feel “fine in the AM” but feel heavy or have swelling by the end of the day.

**Stage II:** (spontaneously irreversible): Pitting becomes progressively more difficult due to an accumulation of connective/fibrotic tissue. At this stage, the edema does not reduce with elevation.

**Stage III:** (lymphostatic elephantiasis): Typically seen in Stage III:
- Greater accumulation of protein-rich fluid, which makes pitting is no longer possible
- Thickening, hardening, or scarring of the tissues (fibrosis and sclerosis)
- Skin changes such as warty overgrowths
- Skin becomes susceptible to deep, poorly healing wounds and is in danger of infection
- Fat deposits or cornmeal appearance of skin (hyperkeratosis)
- Skin may thicken or discolor
- Lymph(fluid) may leak or weep through damaged skin

**How is Lymphedema Treated?**

The recommended treatment plan is an approach based on the Complete Decongestive Therapy (CDT) method which consists of:

A. Manual Lymph Drainage
B. Compression Bandaging
C. Remedial/Lymphedema Specific Exercises
D. Meticulous Skin and Nail Care
E. Instructions in Self-Care
Manual Lymphatic Drainage (MLD) is performed by Occupational or Physical Therapists who are certified in the treatment of lymphedema. It is a gentle manual treatment which improves the activity of the lymph vascular system. In lymphedema, MLD reroutes the lymph flow around blocked areas into more centrally located healthy lymph vessels which drain into venous system.

Compression Bandaging is used in conjunction with manual lymphatic drainage. Short stretch bandages are applied to increase tissue pressure, improve the efficiency of the muscle and joint pumps and prevents re-accumulation of evacuated lymph fluid.

Remedial Exercises are performed with the bandages or support garments in place. These exercises are specifically designed to increase lymph vessel activity which then increases the volume of lymph transported out of the affected limb. Lymph circulation is also improved with these exercises.

Goals of CDT treatment:
- Utilize the remaining lymph vessels and other lymphatic pathways.
- Decongest swollen limbs.
- Eliminate fibrotic (scar) tissue.
- Avoid the re-accumulation of lymph fluid.
- Prevent/eliminate infections.
- Maintain normal or near normal size of limb.

Frequently Asked Questions

Will my Lymphedema Ever Go Away?
It is extremely rare for lymphedema to spontaneously resolve. It is not considered a curable condition. Rather, it is a condition that requires life-long management including meticulous skin care, compression bandaging, use of compression garments and lymphedema specific exercise.

Why Don’t Water Pills Help my Lymphedema?
Lymphatic fluid is comprised of proteins, salts, white blood cells and water and carries cellular debris to the lymph nodes for filtration. Diuretics do not help manage lymphedema.

Will my Insurance Cover Treatment for Lymphedema?
Most insurances will cover skilled therapy sessions for the treatment of lymphedema. Not all insurances cover compression bandages and/or garments needed for treatment. This can best be addressed on a one-to-one basis with your therapist.

Does Lymphedema Cause Pain?
Swelling can stretch tissues, put pressure on nerves or cause scar tissue to form and limit range of motion. The most effective way of reducing pain or discomfort is by treating lymphedema and reducing swelling.

Can I Have Surgery to Cure my Lymphedema?
There are a few types of micro-surgical procedures that are being trialed on a limited number of patients. Unfortunately, success has been limited in most cases and, in some cases, have made the lymphedema worse. Additional research is needed.