The Vascular Education Foundation was established to increase awareness of the dangers and severity of vascular disease through the education of both the community and healthcare professionals.

The VEU™

The Vascular & Endovascular Update

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LEG SWELLING: LYMPHEDEMA…A FRUSTRATING DISEASE

Lymphedema can be a frustrating disease, not only for the patient, but for physicians as well.

A 40-year-old woman presents with bilateral leg swelling that has progressively worsened for the past 2 years. She states that, initially, the swelling was intermittent, but recently has become progressively worse causing her legs to feel heavy and tired. She was in the ED a few months ago because the legs became red and warm and she was treated for cellulitis with antibiotics for the third time in two years. On exam, the left leg is noted to be larger than the right with thickened skin along the shin. Now she has developed an open wound on the left shin which is weeping clear fluid. The patient does not have congestive heart failure nor does she have renal dysfunction. She has tried ACE bandages, over-the-counter stockings and local wound care, but without any improvement. She is frustrated and, as her treating physician, you may feel that there is not much more that can be offered to treat her leg swelling. Lymphedema can be a frustrating disease, not only for the patient, but for physicians as well.

The Lymphatic System

Lymphedema is often dismissed as generalized edema. However, an estimated 5 to 10 million people are affected by it. While vascular surgeons mainly diagnose and treat diseases affecting either the arteries or the veins, when it comes to leg swelling, a vascular surgeon will evaluate the lymphatic system as well.

The lymphatic system is comprised of a complex network of lymph channels that are microscopic "capillary"-like vessels that absorb and transport fluid from tissue to lymph nodes *(figure 1)*. This lymph fluid contains white blood cells, triglycerides, cellular debris, protein and bacteria. The lymph nodes, another organ of the lymphatic system, filters and processes the lymph fluid to maintain homeostatic balance within the tissues of the body. Any pathology or process that affects the ability of the lymph channels or lymph nodes to transport or process the lymph fluid can result in a backup of fluid within the part of the body it serves. This accumulation of fluid leads to the skin and tissue changes that are seen in lymphedema.

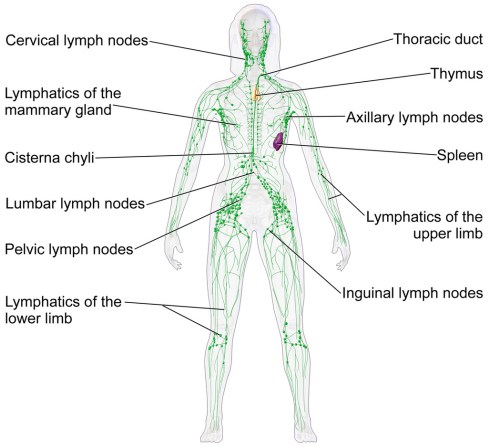

Figure 1. The lymphatic system demonstrating lymphatic channels of the body.

Causes of Lymphedema

There are two classifications of lymphedema: primary and secondary. Primary lymphedema, in general, results from an inherent problem within the lymphatic system. Due to congenital underdevelopment and subsequent malfunction of the lymphatic system, patients develop swelling of the limbs; most commonly, the left leg. If the diagnosis is made between birth and age 35, the disease is known as *lymphedema praecox*, or Meige disease. If the symptoms arise after the age of 35, the disease is

termed *lymphedema tarda*. Both of these forms are congenital and hereditary. A third type of hereditary lymphedema is Milroy disease which often occurs at birth and is associated with other abnormalities such as hydroceles and papillomas. These disorders are inherited in an autosomal dominant pattern.

Secondary lymphedema refers to lymphedema caused by an external, non-congenital factor. Most commonly in the United States, secondary lymphedema is caused by cancer. The cancer cells invade the lymph nodes and disrupt their ability to process and transport the lymphatic fluid. Often, surgery or radiation therapy can lead to secondary lymphedema because of scarring of the lymphatic channels. Other causes of secondary lymphedema include trauma and obesity.

Symptoms and Signs

Despite the cause of lymphedema, the symptoms and signs are similar and patients have typical complaints. Most often, patients will complain of swelling of a limb or limbs that may worsen throughout the day. Typically, the swelling improves only slightly, if at all, when the limb is elevated. The swollen limb will be described as feeling tired, heavy and achy. Patients may complain that they have difficulty wearing clothing such as pants, socks or shoes. They may report difficulty with ambulation or decreased range of motion of the affected limb. There may be a history of frequent infections such as cellulitis or erysipelas. Exam findings will not only help to make the diagnosis of lymphedema, but will also reflect the stage of the disease *(figure 2)*. In stage 1, or mild lymphedema, some swelling is noted with mild pitting. Stemmer's sign may be positive (inability to pinch skin overlying the base of the second toe of the swollen limb). However, the skin is otherwise healthy appearing. In stage 2, or moderate lymphedema, pitting edema is seen along with thickened, often times discolored, skin. At this stage, fibrosis of the tissue has begun. Stage 3 is severe lymphedema, also known as elephantiasis, and is characterized by severe enlargement of the limb with thickened, hardened, scaly skin, evidence of trophic skin changes (acanthosis, fat deposits and hyperkeratosis) and the development of skin crevasses with resultant fungal infections or weeping wounds. Of course, there can be variations in presentation and overlapping of symptoms from stage to stage depending on the individual patient.

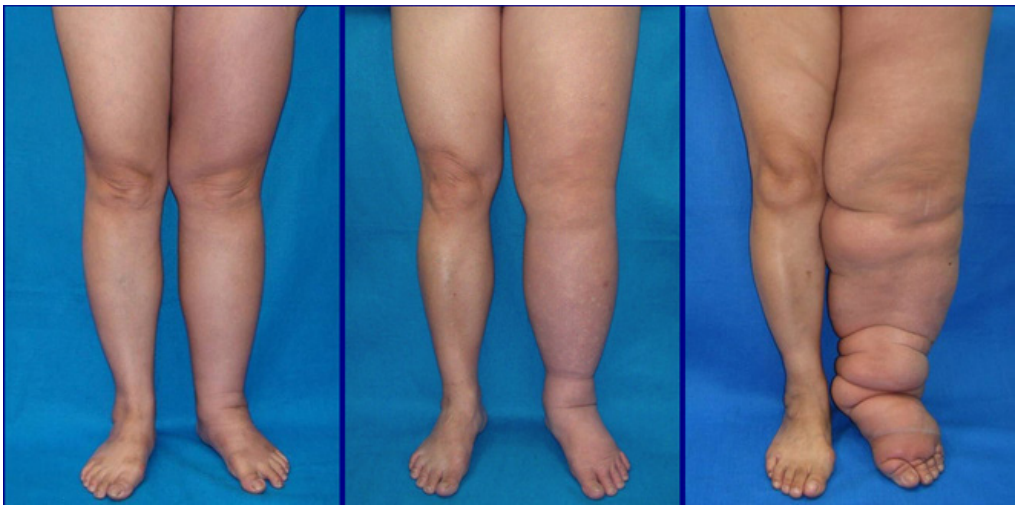

Figure 2. Stages of lymphedema. Changes occur within the affected limb ranging from mild to moderate swelling (Stage 1) to diffuse swelling with thickened, scaly skin (Stage 3, also known as elephantiasis).

Diagnosis of Lymphedema

Diagnostic imaging is not usually done for lymphedema. A tracer scan called a lymphscintongram can be performed which provides a picture of the lymphatics with areas of disruption or pooling of lymph fluid. However, lymphscintography is mostly done at selected centers which, in most cases, will not change treatment plans for a patient with clinically suspected lymphedema. And although 90% of the time, lymphedema can be diagnosed by history and physical alone, it should be thought of as a diagnosis of exclusion. Other sources of generalized edema should be ruled-out prior to conferring the diagnosis of lymphedema to a patient who has leg swelling. Diagnostic testing to check cardiac function and renal function should be done if the clinical picture points towards a systemic cause of the swelling. Certainly, elderly patients who are at risk for cancer should be screened appropriately if there is clinical concern or if the patient is found to have hard, fixed, non-tender lymph nodes. Obvious causes for a patient's swelling that are teased out from history-taking such as trauma or concern for deep vein thrombosis should be pursued with appropriate imaging

Figure 3. A patient with bilateral lymphedema is undergoing compressive therapy with stockings and bandage wrapping to help control swelling.

modalities.

Most importantly, however, is to rule-out chronic venous insufficiency and other venous disorders that may mimic or exacerbate lymphedema. Up to 30% of patients will have an underlying venous problem and many researchers postulate that much of lymphedema is caused by a malfunctioning venous system. Therefore, all patients with leg swelling should undergo venous ultrasound looking for reflux within the deep and saphenous venous systems. Moreover, other venous disorders such as pelvic venous stenosis and May-Thurner syndrome, both of which occur in the pelvic veins, may need to be evaluated. Luckily, as compared to lymphedema which has no reliable surgical cure, these venous pathologies can often be treated very successfully with minimally invasive procedures and alleviate symptoms from lymphedema.

Lymphedema Treatment

There is no cure for lymphedema. However, there are many methods to help control the symptoms of lymphedema and to restore patients to a better quality of life. Treatment consists of aggressive swelling control and wound management during the treatment phase. Once all wounds have healed and the swelling is minimized, then patients will need to continue therapy for swelling control during the maintenance phase. Initially, aggressive treatment modalities may include daily wrapping or medicated wrapping (Unna boot) with bandages of the affected limb in order to heal any wounds that may be present *(figure 3)*. Elevation and compression therapy will be recommended for treatment and maintenance of swelling control. Compression garments are typically of prescription strength (20-30 mmHg or higher). Some patients may prefer physical therapy or lymphedema massage techniques that can be done either at a lymphedema clinic or at home. Most patients will end up needing pneumatic compression devices, or lymphedema pumps, to assist with long term maintenance of leg swelling *(figure 4)*. These pumps are fitted for each patient and are provided for home use. Ultimately, patients will be able to use the pumps for 30-45 minutes a day to minimize the lymph fluid in the legs and then wear compression stockings as they go about their day. These pumps help to minimize leg symptoms, increase mobility and restore healthy skin.

Leg swelling is a common complaint amongst patients. Although as clinicians, it may be sufficient to rule-out CHF exacerbation or DVT as a cause, patients still have real concerns about what is going on with their legs. However, this does not have to lead to a frustrating situation for the patient or for the physician. Your vascular surgeon partner, dedicated to treating the various pathologies affecting the legs, including lymphedema, can offer solutions…and hope *(figure 5)*.

Talk to your vascular surgeon partner to ensure that your patient is getting the best treatment for their unique physiology, medical history and condition.

When you have questions, contact vascular surgeon Dr. Sanjeev Pradhan at 708-821-7794

Figure 4. Patient is using a lymphedema pump at home to maintain a healthy and functional extremity once affected by extreme swelling.

Figure 5. Before and after photographs of a patient suffering for years with left leg lymphedema. Utilizing multiple treatment modalities, the patient was able to restore her leg to a healthier and more functional state.

About the author, Dr. Sanjeev Pradhan

With Vascular Specialists in Tinley Park, Illinois, ***Dr. Sanjeev Pradhan*** is board-certified in both vascular and general surgery. He earned Vascular and Endovascular Fellowships at Yale University School of Medicine.

Dr. Pradhan designed a unique hybrid vascular surgery suite which integrates advanced imaging techniques with a traditional operating room setting, and was the first surgeon in the Chicago area to use fenestrated stents to treat abdominal aortic aneurysms.

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