Putting the squeeze on lymphedema

When a portion of the lymphatic circulation develops abnormally or is injured by trauma, surgery, or radiation therapy, fluid balance is thrown off kilter and the patient can experience severe swelling in an extremity. In this article, we’ll help you learn about the different types and stages of lymphedema and the treatment options for this difficult condition. We’ll also offer you advice on prevention strategies and patient teaching.

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MOST OF US are familiar with lymphedema following breast cancer therapy, especially when axillary dissection and axillary radiation therapy are used. Slightly more than a quarter of patients who receive treatment for breast cancer are affected by lymphedema, and it can cause substantial functional and psychological impairment.

Of course, cancer treatment isn’t the only cause of lymphedema. In this article, I’ll help you understand the types of lymphedema, its causes, possible complications, and preventive and treatment strategies.

Let’s start off with the causes and a description of the various types of lymphedema.

A blockage in the flow
Lymphedema is the result of excess proteins, fluid, inflammation, and fibrosis in the lymphatic system. It’s divided into two types: primary and secondary, based on the underlying cause.

Primary lymphedema affects anywhere from 1 to 2 million individuals in the United States. It’s twice as common in women as in
men, and it affects the lower extremities three times more often than the upper extremities. It’s bilateral in two-thirds of cases. Primary lymphedema isn’t a progressive disease. Congenital lymphedema, lymphedema praecox, and lymphedema tarda are forms of primary disease distinguished by the age of onset.

Congenital lymphedema is typically characterized by absence or abnormality of the lymphatic tissue; it’s clinically evident at birth. Milroy disease, a kind of congenital lymphedema, accounts for about 2% of all primary lymphedema cases. Lymphedema praecox, which expresses itself between birth and 35 years of age, is the most common type of inherited primary lymphedema, accounting for 65% to 80% of all cases of primary lymphedema. Lymphedema tarda, also known as Meige disease, has a late onset; it usually doesn’t appear until after age 35. It’s the rarest form of primary lymphedema.

Secondary lymphedema, the kind most of us are familiar with, is an acquired condition. It’s usually caused by an obstruction or trauma to the lymphatic system that interferes with lymphatic flow. In the United States, it’s most commonly caused by injury to or removal of regional lymph nodes during surgery or radiation therapy,
infection, or tumor growth. Secondary lymphedema affects between 2 and 3 million individuals in the United States. Worldwide, the most common cause of secondary lymphedema is filariasis, an infestation of the lymph nodes by the parasite Wuchereria bancrofti; it’s this organism that gives rise to elephantiasis.

No way out
The lymphatic system is made up of lymph vessels, tissue, and organs. Lymphocytes, a specialized type of white blood cell, circulate throughout the lymphatic system to help the body fight off disease and infection. Lymph vessels collect a fluid that’s made up of protein, water, fats, and wastes from the cells of the body and then carry the fluid to the lymph nodes. Lymph nodes filter out potentially harmful waste and foreign materials. Lymph vessel walls are thinner than those of venous and arterial blood vessels, allowing larger proteins to permeate the vessel walls.

If excessive amounts of protein and fluid overload the lymphatic system, they begin to accumulate in the interstitial spaces. Lymphedema is distinct from other forms of edema in that material trapped in the interstitial spaces has a higher concentration of protein.

The protein and fluid in the interstitial spaces prompt an inflammatory response. Fibroblasts migrate to the area and deposit collagen. This changes the initial pitting edema into the brawny (orange), nonpitting edema characteristic of lymphedema. Pitting refers to the indentation that persists when a finger is pressed into an edematous area. See It’s the pits for the International Society of Lymphology’s staging system.

We’ll look next at the clinical presentation of secondary lymphedema.

Keep it up
Secondary lymphedema can occur acutely, or it may have a more insidious, gradual onset. Four patterns of acute lymphedema are recognized:

I. Mild acute lymphedema lasts for only a few days. It usually occurs following surgery to remove a lymph node or after an injury to lymphatic vessels that occurs during surgery. This form of acute lymphedema may be recognized by a warm, erythematous, and nonpainful limb associated with the lymphatic chain near the area.

II. The second type of acute lymphedema occurs 6 to 8 weeks after surgery or radiation to the affected area. It’s noted by a warm to hot, erythematous, very tender limb.

The trouble really starts when the lymph nodes stop working. The International Society of Lymphology has set the following staging system:

- **Stage 0**—Subclinical condition where swelling isn’t evident despite impaired lymph transport. It may exist for months or years before edema occurs.
- **Stage I**—Pitting may occur and is reversible. It may take up to a few hours of rest and elevation to reverse.
- **Stage II**—Pitting occurs, and the edema isn’t appreciably reduced with elevation of the affected limb. In late Stage II, the tissue hardens and becomes fibrotic, and pitting no longer occurs.
- **Stage III**—This stage is also referred to as elephantiasis. Pitting is absent. Skin changes like acanthosis, fat deposits, and warty overgrowths may develop. Fluid may ooze from the skin due to high pressure in the lymphatic and venous vessels. It most commonly occurs in the legs and results from long-standing, inadequately treated or untreated lymphedema.
To treat it, the affected limb is elevated and anti-inflammatory medications are given to reduce inflammation of the lymphatic vessels and/or veins.

- The third type of acute lymphedema affects the superficial lymphatic vessels and nodes following an insect bite or other minor injury in the area. The affected area’s signs and symptoms are very similar to the second type of acute lymphedema in that the area is painful, warm to hot, and erythematous. Edema may also be apparent. The affected limb is elevated, and because the risk of infection is increased, antibiotic prophylaxis is given.

- The fourth and most common type of acute lymphedema develops 18 to 24 months following cancer surgery, although in some cases, it may take years to develop. Pain is the hallmark symptom of the fourth type of acute lymphedema. It’s typically felt in the neck, shoulders, back, and hips.

  This type of acute lymphedema is more complicated to treat because the painful affected areas tend not to be in an extremity, making elevation tricky. Pain medications and/or anti-inflammatory agents may help.

  Acute lymphedema generally resolves within 6 months. Factors that could cause acute lymphedema to become chronic include a surgical drain leaking protein into the surgical site, inflammation, paralysis of the affected limb, loss of lymphatic function in the area, and/or blockage of a vein by a blood clot or inflammation.

  In chronic lymphedema, the lymphatic flow can’t meet the demands made on it. Many of the problems that cause acute lymphedema also contribute to chronic lymphedema. Unlike acute lymphedema, however, chronic lymphedema isn’t reversible. Treatment is palliative, not curative. Pain, heat, edema, and erythema are also characteristic of chronic lymphedema. Elevation of the affected extremity doesn’t help edema in chronic lymphedema, and the skin eventually becomes hardened or fibrotic.

**did you know?**

The following conditions can mimic signs and symptoms of lymphedema:
- allergic disorders
- heart failure
- hepatic cirrhosis
- hereditary angioedema
- hypoproteinemia
- idiopathic cyclic edema
- kidney failure
- lipedema
- postphlebitic syndrome
- total body excess of free water
- venous disease.

Let’s move on next to diagnosis and treatment of secondary lymphedema.

**Nailing the diagnosis**

Unlike patients with arterial or venous disease, most patients with lymphedema don’t report symptoms other than those related to the weight and size of the limb. Oozing fluids may cause pruritus.

Your patient assessment must include a history and physical exam. Make sure you ask about past surgery, postoperative complications, radiation treatment, and how long it was between the time of surgery or radiation and the onset of lymphedema. You should assess the quality and behavior of the edema: Does it change with position? Has it progressed over time? Find out if there’s a history of trauma or infection, and be sure you get the whole scoop on current medications.

No specific tests are used to diagnose lymphedema. Diagnosis is generally made on clinical presentation and history. Edema usually isn’t detectable clinically until the interstitial volume is about 30% above normal.

Occasionally, imaging studies may be helpful to assess lymphatic flow or to identi-
The most commonly used noninvasive techniques are lymphoscintigraphy, computed tomography scans, and Doppler ultrasonography. In lymphoscintigraphy, a water-based radionuclide is injected into the lymphatic tissues, allowing the dynamics of the flow to be traced, any reversal of flow to be identified, and the severity of the obstruction to be determined.

If there are any lingering doubts over a diagnosis of lymphedema, a chemical evaluation of the protein content of the edema fluid should put them to rest. A tissue fluid analysis with a protein content between 1 and 5.5 g/dl usually indicates lymphedema; 0.1 to 0.9 g/dl is more indicative of venous or cardiac edema.

Better sooner than later
The main focus of treatment for secondary lymphedema lies in prevention. Lifestyle plays an important role, and patients should be encouraged to consume a healthy diet, maintain a normal body weight, and participate in regular exercise. Patients with lymphedema need to take particular care not to do anything that could lead to infection. Even a minor scratch to the skin near the area may become infected. The National Lymph-

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### Educating patients about lymphedema

<table>
<thead>
<tr>
<th>General measures</th>
<th>Prevent infection</th>
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<tbody>
<tr>
<td>• Elevate the affected arm or leg above your heart as much as possible.</td>
<td>• Keep the affected arm or leg clean and dry; apply moisturizer often.</td>
</tr>
<tr>
<td>• Avoid having blood drawn or injections given in the affected arm, and avoid injections into the hip on the side of the affected leg.</td>
<td>• Use gloves and wear shoes when gardening.</td>
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<tr>
<td>• Don’t have your blood pressure measured on the affected arm.</td>
<td>• Cut nails straight across, and don’t cut your cuticles. Do not bite your nails or fingers. See a podiatrist or primary care provider if a nail infection or an ingrown nail occurs.</td>
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<tr>
<td>• If your legs are affected, don’t sit for more than 30 minutes before getting up for a break. Also, don’t cross your legs when sitting.</td>
<td>• Use an electric razor for shaving.</td>
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<td>• Avoid tight clothing and jewelry.</td>
<td>• Use an antibacterial ointment on cuts and scrapes.</td>
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<tr>
<td>• Don’t hold or wear a purse on the affected arm.</td>
<td>• Use gauze, not tape bandages, to cover cuts or scrapes.</td>
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<td>• Don’t use ice packs or heating pads on the arm or leg that has lymphedema.</td>
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<tr>
<td>• Don’t use the affected limb to test the temperature of water. Feelings of touch, temperature, and pain may be reduced in the affected leg or arm.</td>
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<tr>
<td>• Follow the diet and exercise plan developed by your health care team.</td>
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<td>• Keep your medical appointments.</td>
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<td></td>
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<tr>
<td>Prevent inflammation</td>
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<td>• Avoid injury to the affected arm or leg.</td>
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<tr>
<td>• Avoid physical exercise or activities that might lead to cuts or bruises.</td>
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<tr>
<td>• Always wear socks and shoes, especially outdoors.</td>
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<tr>
<td>• Use extreme care and use protective devices on hands when using needles, knives, scissors, or other sharp objects if you have an affected arm.</td>
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<tr>
<td>• Avoid getting sunburned; use sunscreen with an SPF of 30 or greater.</td>
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<tr>
<td>When to call your primary care provider</td>
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<tr>
<td>• You have signs and symptoms of infection like redness, pain, heat, swelling, and fever.</td>
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<tr>
<td>• You notice a rash.</td>
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<tr>
<td>• Your affected limb suddenly gets larger.</td>
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edema Network, a nonprofit organization, offers an excellent guideline for risk reduction at [http://www.lymphnet.org/riskReduction/riskReduction.htm](http://www.lymphnet.org/riskReduction/riskReduction.htm). See also Educating patients about lymphedema for more information.

The good news is that tissue loss or ulceration related to lymphatic obstruction with subsequent lymphedema is unusual. Tissue loss occurs because of a concurrent secondary pathology. As already noted, patients with lymphatic obstruction are prone to infections in the affected extremity caused by bacteria or, more commonly, fungus. As part of your assessment, you should examine intertriginous folds for ulcers and areas of infection. Explain to your patients that this is important to do at home too. It may be helpful to show them how to use a mirror to visually examine those hard-to-see nooks and crannies.

To be effective, lymphedema treatment must improve lymph flow and drainage. The best outcomes are obtained when therapy begins before irreversible sclerotic changes occur in the interstitium. Strict adherence to therapy is essential and lifelong.

Let’s take a closer look at the types of lymphedema treatments.

Complete decongestive therapy (CDT) has four stages:

- **Manual lymph drainage (MLD)** is a gentle manual treatment administered by a trained therapist. MLD reroutes lymph flow around the blocked area into more centrally situated, healthy lymph vessels. From there, the flow can drain into the venous system. Special massage techniques help to break down fibrotic tissue to restore flow.

- **Compression bandaging**, slightly elastic cotton bandages are wrapped around the affected extremity to increase tissue pressure, helping to push the excess fluid and protein out of the area. Compression bandaging also helps keep more fluid and protein from flowing in and breaks down areas of scarring and fibrosis. Note that compression-bandage therapy for lymphedema requires the application of more pressure than compression-bandage therapy for venous disease: about 30 to 40 mm Hg for venous disease versus about 50 to 60 mm Hg for lymphedema.

- **Remedial exercises** are performed with compression bandages or compression garments in place. Breathing exercises help to increase the volume of lymph fluid carried through the thoracic duct (the body’s largest lymph vessel). Remedial exercises increase lymph vessel activity and improve lymph circulation.

- **Meticulous skin and nail care** reduces bacterial and fungal growth, which decreases the risk of infection.

Contraindications for CDT and MLD include acute infection, cardiac edema, malignant disease, and pulmonary edema from chronic obstructive pulmonary disease.

### Thanks for your support

Numerous local and national support and education organizations are available to assist patients with lymphedema. A good place to start is the National Institutes of Health National Cancer Institute (NCI). The NCI can be reached at [http://www.cancer.gov](http://www.cancer.gov) or at 1-800-4-CANCER (1-800-422-6237). Other resources include:

- Cancer Supportive Care Programs [http://www.cancersupportivecare.com](http://www.cancersupportivecare.com)
- Circle of Hope Lymphedema Foundation, Inc. [http://www.lymphedemacircleofhope.org](http://www.lymphedemacircleofhope.org)
- National Lymphedema Network [http://www.lymphnet.org](http://www.lymphnet.org)
- Y-ME National Breast Cancer Organization [http://www.y-me.org](http://www.y-me.org)
tion, however: Whether a bandage or garment is used, it must be applied correctly; improper application can compress nerves or blood vessels, which can cause complications.

Arterial disease is a contraindication for the use of compression bandages or garments. Caution should also be used in patients with hypertension, paralysis, diabetes, asthma, and heart failure.

Specialized lymphatic drainage pumps, called gradient pneumatic lymphedema pumps or intermittent pneumatic compression pumps, are also available. The extremity or part of the extremity, such as the foot, is wrapped, and the pump applies gradient pressure with more pressure on the distal end than on the proximal end. The pump also provides sequential pressure, a technique like milking that pushes the fluid from the distal end of the extremity toward the trunk and central body cavity.

Usually, a combination of these approaches is used along with lifestyle changes for the best outcome.

**Water pills don’t work**

There’s not much available in the way of effective medical therapy for lymphedema. Diuretics have little or no value. They draw off the excess water in the interstitial spaces, but not the excess protein. As soon as the diuretic is stopped, the concentrated proteins pull more water back into the affected area.

The dietary supplement coumarin, a benzopyrone, isn’t approved in the United States and may produce serious adverse effects, in particular, liver toxicity. Selenium therapy for secondary lymphedema of the arm caused by breast cancer surgery is currently under investigation in a clinical trial. The theory behind the selenium trial is that the damage to the lymphatic system is linked to excessive generation of oxygen radicals; the researchers hypothesize that selenium, an antioxidant that gobbles up oxygen radicals, might decrease the damage to the lymph system.

Also under investigation in a clinical trial is the use of hyperbaric (high-pressure) oxygen therapy for the treatment of secondary lymphedema caused by breast cancer treatment. According to the National Institutes of Health (http://www.clinicaltrials.gov/ct/gui/show/NCT00077090), the researchers think that hyperbaric oxygen may stimulate the growth of new lymphatic channels as well as lead to a reduction in scar tissues surrounding existing lymphatic channels in the armpit.

Occasionally, surgery is used as a palliative treatment to relieve pressure and edema. It’s usually considered the treatment of last resort to relieve otherwise intractable pain.

Complications associated with lymphedema are, in general, associated with an infection in the affected limb. If the pelvic lymphatic system is affected, problems of elimination may occur.

A rare and lethal complication is lymphangiosarcoma, cancer of the lymphatic vessels; it can develop after the patient has had chronic lymphedema for 10 years or more. The incidence is estimated to be 0.45% in patients who survive at least 5 years after radical mastectomy; a lower incidence of 0.07% is seen in patients after simple mastectomy. Death from lymphangiosarcoma generally occurs within 18 months, with the 5-year survival rate at around 10%.

Physical findings of lymphangiosarcoma are bluish-red or purplish bumps on the affected extremity that resemble slightly raised ecchymosis. The tumors are very aggressive and characteristically metastasize early to multiple sites, most often to the lungs. Treatment may involve amputation of the affected extremity or wide local excision of the lesions. Lymphangiosarcoma that arises following mastectomy is called Stewart-Treves syndrome.

Let’s finish up with a discussion of some
ways to keep your patient with lymphedema from getting worse or running into complications.

At home
Stress to all patients with lymphedema the importance of reporting any signs and symptoms of infection in the affected extremity to the primary care provider.

Patients should also be given nutritional guidance to help them maintain adequate protein balance to help fight off infection. In general, the diet should provide 30 to 35 calories/kg/day, including 1.25 to 2 g protein/kg/day. Most patients can benefit from taking a daily multivitamin with minerals.

Lymphedema is disfiguring, sometimes painful, disabling, and often leads to big changes in a patient’s lifestyle. These factors can all lead to depression. Support groups, counseling, and antidepressant medications may be useful for treating emotional disorders associated with a diagnosis of lymphedema (see Thanks for your support).

Keep in mind that early recognition and appropriate treatment can help minimize progression and complications of lymphedema. It’s your responsibility to help patients recognize lymphedema, avail themselves of appropriate interventions, and help them maintain a good quality of life.

Learn more about it


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Putting the squeeze on lymphedema

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