

Excerpt from:

Coping with Lymphadema

Written by Joan Swirsky and Diane Sackett Nannery

What is Lymphadema?



This pamphlet will address questions we are often asked by physicians, nurses, therapists, and other healthcare professionals regarding pneumatic compression therapies.

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What is Lymphadema?

Lymphedema is a combination of two words: *lymph* (a natural body fluid) and *edema* (swelling)—literally, it means “swelling caused by lymph.” This is a chronic disorder in which lymph fails to circulate properly and, as a result, accumulates in the tissues of a limb or other part of the body. Its symptoms can include pain, numbness, a loss of mobility, a loss of skin elasticity, hardening of the skin, increased susceptibility to infection, chronic ulceration of the skin, and swelling that can make an arm, leg, or other body part as much as two, three, four, or even more times its normal size. Once the onset of lymphedema takes place, it affects every facet of your life—and it never goes away. If untreated, the damage it causes is irreversible and progressive. In order to understand this condition, it is first necessary to understand how the body’s circulatory systems work, and also to understand the role of the immune system. As you will see, these systems are separate, yet intrinsically related.

THE BODY’S CIRCULATORY SYSTEMS

When we think of circulation, we tend to think of blood and the blood vessels. But the body has another circulatory system that works in concert with the blood system. This is the lymphatic system. The lymphatic system is a mechanism for removing excess blood protein and water from the spaces around the cells and returning them to the blood system.

The Blood System

Blood is composed of red blood cells, which carry oxygen; white blood cells, which fight infection; platelets, which stop bleeding; and plasma, the liquid portion, which is 90 percent water and also contains a number of proteins that are essential to life. Blood is pumped through the blood vessels by the heart. Upon leaving the heart, the blood goes first to the lungs, where it picks up oxygen. It then travels to the cells of the body by means of the arteries and capillaries. Since capillaries are somewhat porous, they allow for oxygen and nutrients to be transferred out of the blood by means of the plasma and to enter the tissues, where they are used to produce the energy needed to carry out all the activities necessary for life. When fluid diffuses out of the capillaries, it joins the, interstitial fluid, which is found in spaces between the body’s cells and tissues. It is the interstitial fluid that bathes all the cells of the body and serves as an exchange medium for the nutrients carried in the blood. From this fluid, oxygen and nutrients enter the cells. In turn, the cells release carbon dioxide and other cellular waste products and eliminate them through the circulatory system. The blood is returned to the

heart by means of the veins and the journey begins all over again. As blood passes through the capillaries in the tissues, there is a continual exchange of its plasma portion (the liquid, and nutrients, including protein molecules, that are needed by the cells for the maintenance of life) and the interstitial fluid. Normally, the amount of fluid that exits the capillaries for the interstitial fluid is roughly equal to the amount of fluid that reenters the capillaries from the interstitial fluid. However, a small percentage of the fluid is left behind. It is this fluid that must rejoin the blood circulation by means of the lymphatic system.

The Lymphatic System

The lymphatic system consists of the lymph, lymphatic vessels, and lymph nodes. It is responsible for returning excess fluid and blood protein from the tissues to the blood circulation, and also plays an important role in protecting the body against illness. Lymph is a clear, colorless fluid composed of water, protein, salts, glucose, and urea, plus white blood cells. Basically, lymph is interstitial fluid that has left the tissue spaces and entered the lymphatic capillaries. Unlike the blood vessels, which are closed (that is, the blood is always contained within the blood vessels), the lymphatic system’s capillaries are open at one end. It is through these openings that the interstitial fluid enters the capillaries to become lymph. At the other end, lymphatic capillaries join with other lymphatic capillaries to form the lymphatic vessels. These vessels act as channels through which lymph is moved through the body, back toward the blood circulation. The lymph is as thick as honey or syrup and it moves rather slowly, its movement powered by the contractions of skeletal muscles and the motion of breathing. An amazingly efficient system of one-way valves within the lymphatic vessels prevents the fluid from flowing backward toward the tissues again. Lymphatic vessels also absorb most of the digested fats that drain from the intestines, and transport these to the bloodstream as well. This intestinal lymphatic fluid is called **chyle** and often appears whitish because of its fat content. As lymph is transported through the body, it passes through structures called lymph nodes, which are located along the lymphatic vessels in various regions of the body. These round or kidney-shaped nodes, roughly the size of lima beans, are found in the head and neck, armpits, groin, abdomen, pelvis, and chest, either deep in the body or near to the surface, where they can be detected through manual examination. There are between 500 and 1,500 lymph nodes in the body. The lymph nodes have two main functions. First, they produce **lymphocytes**, which are white blood cells that combat infection by producing antibodies to fight bacteria and viruses. Second, they filter the lymph, destroying and removing dead cells and waste materials, as well as bacteria, viruses, and other potential disease-causing pathogens. In essence, the nodes catch and annihilate these toxins, preventing them from entering the bloodstream and wreaking havoc there. When the lymph nodes have performed their detoxifying function, the lymphatic fluid is ready to be returned to the blood.

At the end of the lymphatic system, the lymphatic vessels join together to form the two lymphatic ducts. The ducts connect the lymphatic circulation with the

blood circulation; through them, lymphatic fluid passes back into the blood. The left duct, which is the larger of the two, serves as the drainage system for 80 percent of the body, including the left side of the neck and head, the left arm, the trunk of the body, and the legs. This duct begins in the area of the lower spine, collecting the lymph from all the lymphatic vessels of the lower limbs, pelvis, abdomen, and lower chest. The fluid is then transported up to the chest, where it empties into the central blood circulation through a vein at the base of the left side of the neck. The right lymphatic duct collects lymph from the right side of the neck, the right chest area, and the right arm, and empties into a vein in the right side of the neck.

Circulation And The Immune System

As we have seen, the circulatory systems are central to nourishing the tissues, removing wastes, and maintaining the body's critical fluid balance. That is not their only role, however. They also are important components of the immune system. The immune system is that complex mechanism that defends and protects the body against the bacterial and viral invaders that threaten to undermine our health and well-being. To be sure, our bodies have other ways of warding off infections and diseases. Among them are the skin, which covers and protects the entire body; the mucous membranes that line our breathing passages and snare or sweep away minute organisms; our temperature-regulating mechanism, which elevates body temperature to destroy organisms that cannot withstand the heat; and even our sticky earwax, salty tears, and acidic stomach juices, all of which protect against and can destroy or retard the growth of microorganisms. But if these defenses fail to ward off bacterial or viral invasion, the body depends on its second line of defense: blood and lymph. At the first sign of foreign invasion, blood and lymph containing infection-fighting white blood cells rush to the affected tissues in an attempt to localize the infection and prevent its spread. Different types of white blood cells perform different functions. Some physically attack, engulf, and consume invading organisms; others secrete substances that kill specific enemies. Once infection has been contained, the debris resulting from this cellular warfare is cleaned up by means of the lymphatic circulation—it enters the lymphatic capillaries with the interstitial fluid and is filtered out by the lymph nodes. The purified lymphatic fluid is then reintroduced into the blood system.

THE EFFECTS OF LYMPHATIC DAMAGE

If the lymphatic system is damaged, its normal functions are compromised. If the nodes are injured, they cannot filter toxins from the system or produce the lymphocytes that fight infection; if nodes are removed, this leaves the body with fewer nodes to fight disease and also disrupts the pathways by which lymph is normally drained from the tissues; if the vessels are injured, they can no longer transport lymph through the system as they should. Put simply, a damaged lymphatic system works inefficiently, giving rise to symptoms that demand treatment.

Both blood plasma and interstitial fluid contain protein, but plasma normally contains about 7 percent protein, whereas interstitial fluid contains approximately 2 percent. Biochemically, this results in most plasma protein being held in the circulatory system. If some leaks out, it is returned to the blood by the lymphatic circulation. However, if the lymphatic system is damaged, a concentration of protein eventually accumulates in the tissues. This protein further interferes with the flow of tissue fluid into the lymphatic capillaries and, ultimately, the blood. The result is edema, or swelling. How might the lymphatic system be damaged? The filtering mechanism of the lymph nodes or the lymphatic vessels may be injured as a result of surgery, injury, infection, invading cancer cells, a congenital or acquired malformation or obstruction of the veins, or radiation treatment.¹ One common scenario that can lead to damage is cancer surgery. If there is cancer anywhere in the body, its cells may enter the draining lymphatic vessels and be carried to other parts of the body. So for the purpose of determining if cancer is present or has spread, lymph nodes are often removed for microscopic examination to determine whether they are cancerous or noncancerous.

Whatever the cause of the damage, a damaged lymphatic system is unable to tolerate the demand for the drainage of lymph fluid. In short order, large molecules of proteins and lipids (fats) accumulate in the interstitial spaces, surrounding tissues are deprived of their vital oxygen supply, and chronic inflammation and scar tissue develop, which further impedes the flow of lymph. The result: a backup of fluid and swelling in the affected limb lymphedema. The trapped protein becomes stagnant, creating an environment ripe for infection. At the same time, because the lymphatic system is an important component of the immune system, the immune response is often unpaired. The result can be repeated bouts of infectious disease. Some doctors refer to lymphedema as a “plumbing problem.” In fact, this analogy is quite apt. It is as if water were coming into your home through three underground pipes, but one of the pipes—the one with a filter to remove impurities—is damaged and has become clogged with dirt. It is easy to see why this would severely compromise both the purity and the flow of water, as well as resulting in a backup of water. Your lymphatic vessels can be compared to the pipes in this illustration; your lymphatic fluid to the water; and the accumulating protein to the dirt. If the flow of lymph through lymphatic vessels is impeded, both by injury to the vessels and by the resulting accumulation of protein molecules, the backup that ensues is forced into the tissues of the affected arm or leg, creating a seriously swollen limb.

TYPES OF LYMPHEDEMA

There are two general categories of lymphedema, primary and secondary. Primary lymphedema is lymphedema unrelated to any other known condition. Secondary lymphedema is lymphedema acquired as a result of some other disease or trauma. Approximately 1 in 6,000 people in the United States is affected by hereditary primary lymphedema. The symptoms of type I hereditary lymphedema, also known

as Milroy's Disease, are typically pre-sent at birth, and the swelling tends to worsen slowly with advancing age. This type of lymphedema affects more females than males.

In type II hereditary lymphedema, or Meige disease, symptoms usually develop during childhood, adolescence, or early adulthood. This form of the disease generally produces severe swelling in areas below the waist, with the first symptoms usually including reddened skin over areas of swelling. This type of lymphedema affects males and females in equal numbers. Lymphedema tarda is a form of hereditary lymphedema that usually occurs after the age of thirty-five. Its symptoms are similar to those of type II hereditary lymphedema. In the hereditary form of the disease, symptoms of lymphedema develop because the lymphatic vessels are obstructed as a result of malformation of the lymphatic system. In some cases, there may be fewer lymphatic vessels than normal or the vessels may be underdeveloped. In other cases, lymphatic vessels may be unusually large and numerous. Complications of hereditary lymphedema may include lymphangitis (inflammation of lymphatic vessels) and cellulitis (these conditions will be discussed in detail in Chapter 2). Red streaks on the skin may develop, as may a general feeling of ill health, fever, chills, and/or headaches. Some people with this condition develop a persistent accumulation of fluid in the lungs. The most serious long-term complication of all forms of hereditary lymphedema is a slightly increased risk of developing lymphangiosarcoma, a type of cancer, in the affected area.

Most cases of congenital lymphedema are inherited as what geneticists call an autosomal dominant trait. Simply put, if an individual inherits a defective copy of a certain gene, from either the mother or the father, he or she will be affected. This is because the defective gene will "dominate" the corresponding normal gene received from the other parent, resulting in the appearance of the disease. If an affected person has children with an unaffected partner, the likelihood of any individual child inheriting a defective gene, and thus the disorder, is 50 percent, regardless of the sex of the child. It is also possible for congenital lymphedema to occur because of genetic mutation (a spontaneous change in genetic material) early in fetal development. Once such a mutation occurs, it can be passed down to the affected individual's offspring. Secondary lymphedema is lymphedema that comes about as a result of illness or injury that damages the lymphatic system. It is much more common than primary lymphedema. It can occur after surgery with lymph-node removal, after radiation therapy, or after an injury to or infection of the lymphatic system. This is because these factors can result in lymphatic vessels being severed, damaged, or blocked. Sometimes kidney failure or heart problems may lead to lymphedema in one or both legs. In such cases it is likely to remain undiagnosed and be either ignored or treated improperly. Primary and secondary lymphedema are equally threatening, and affect both children and adults.

SYMPTOMS OF LYMPHEDEMA

Lymphedema is a serious condition involving the lymphatic, circulatory, and immune systems, which cannot be seen with the naked eye. What can be seen, as a kind of bold representation of the inner damage that has taken place, are swelling of the limb and changes in the skin. The first symptoms of lymphedema may be subtle, starting when you notice that the ring on your finger is hard to remove, or that your arm appears slightly swollen, or that you seem to have a minor infection from the scratch of a pin. Or they may be quite blatant—a sudden ballooning of an arm or leg or a lack of mobility in the affected limb.

Swelling

Lymphedema often begins with a condition called pitting edema, in which the tissues swell due to an excess of fluid and small indentations form when the affected area is pressed with a finger. In pitting edema, the indentations return to normal if the area is elevated, and no noticeable or lasting changes are apparent. This is considered grade one, or acute, lymphedema. Without careful monitoring and treatment, a person with acute lymphedema is at risk of progressing to grade two lymphedema, usually after three to six months.

In grade two lymphedema, the condition becomes chronic. As it does so—and especially if left untreated—it involves less pitting edema, but significant skin changes set in. The affected limb swells, but the swelling is not reduced by elevation. In addition, the skin hardens as fibrous tissue develops, and the skin does not pit when pressed. This hardened tissue further blocks the fluid's flow, which makes the lymphedema worse. It is at this point—when the thick, coarse skin may become ulcerated and bacteria are able to enter the skin—that a person becomes vulnerable to cellulitis, an infection of the soft tissues of the skin. Grade three lymphedema occurs after repeated attacks of cellulitis. If a lower limb is affected, it can lead to a chronic condition known as lymphostatic elephantiasis, in which the leg becomes hugely enlarged and the skin and underlying tissue become hardened. The term elephantiasis is used to describe the similarity in appearance to an elephant's leg.

Skin Changes and Problems

It is important to understand the role the skin plays in lymphedema not only because it often reflects the severity of the disorder but because it is so visible to the outside world. Your appearance affects how you feel about yourself, and also affects your ability to proceed with a normal work and social life. According to Deborah Samoff, MD, a clinical professor in the Department of Dermatology at New York University Medical School who has treated the skin problems of many people with lymphedema, people suffer as much from the change in the appearance of their skin as they do from the discomfort and potential danger of the other

symptoms of this stubborn condition. It is therefore crucial to treat skin problems before they reach an advanced stage.

When lymphatic vessels are obstructed or destroyed, they are unable to drain away microbes that can colonize in the tissues and penetrate the skin. In short order, this protein-rich, stagnant lymph creates a perfect environment for bacteria to grow and flourish, leading to chronic dermatitis (skin inflammation), hyperkeratosis (patches of roughened skin), and a brawny or leathery appearance. The skin may also become warty or homy or callused. In addition, there may be significant disfigurement, with the limb swelling to several times its normal size. Often, there is weeping of lymphatic fluid through the skin, giving the skin a mossy texture. Chronic weeping, leaking, and oozing leads to a mushy softening of the skin that can give rise to additional bacterial, yeast, and/or fungal infections—a kind of vicious cycle.

For example, a fungal infection such as athlete's foot may develop in the spaces between the toes, causing peeling and scaling and a breakdown of the skin. Once an opening in the skin occurs, a portal for infectious organisms exists. *Staphylococcus* or *Streptococcus* bacteria may enter the system, thrive in the stagnant lymphatic fluid, and cause repeated bouts of infection. As this vicious cycle continues, the individual may experience a loss of normal sensation, a lack of mobility, and a diminishing sense of self-esteem as a result of the limb's unsightly appearance.

Once lymphedema becomes chronic, changes occur in the dermis, which is the deeper layer of skin. In addition to swelling of the lymphatic vessels, there is a thickening of the skin and a development of scarlike formations called fibrosis. Since early diagnosis of both skin and other problems is the key to better overall health, and in some cases longer survival, all patients with chronic lymphedema must undergo regular skin examinations on a lifelong basis.

Very rarely, lymphangiosarcoma, a form of cancer that affects the tissues of the lymphatic system, may develop as a complication of chronic lymphedema. The reason or reasons for this are not well understood, but it may be related to the degeneration of collagen (a protein that is a key component of skin tissues) and fats below the dermis, or to impairment of the immune system in people with lymphedema. Anyone with lymphedema who suddenly develops crops of purple-red patches or bumps on the skin of the involved limb should consult a doctor without hesitation, as these are the primary symptoms of lymphangiosarcoma. If a doctor suspects lymphangiosarcoma, a biopsy should be performed at once.

DIAGNOSING LYMPHEDEMA

Often, people with symptoms of lymphedema hesitate to seek help, either because they think those symptoms will go away, or because they believe the symptoms “go along with” their cancer surgery or accident. However, since diagnosis is the first step toward treatment and effective management, any person who has had a traumatic accident, undergone surgery in which lymph nodes were removed, or been through any kind of radiation treatment must pay close attention to any signs of swelling and seek medical advice immediately.

When you go to your doctor, you may find that he or she knows about as much—or as little—about the condition as you do. There are no standard criteria for diagnosing lymphedema. One doctor may depend on visual examination, another on imaging scans, yet another on arm or leg measurements that chart the course of swelling. Some doctors combine these methods to arrive at a definitive diagnosis. Many people actually end up diagnosing themselves.

However it is done, getting a correct diagnosis is essential if you are to proceed with effective treatment—treatment that will manage the condition before it gets worse.

Measuring

One way to assess lymphedema is to measure the affected limb. Using an ordinary tape measure, your doctor measures the circumference of your limb in several places to monitor the increase or decrease in swelling on an ongoing basis. In an arm, measurements are taken in at least one location on the lower arm and two locations on the upper arm; this may be reversed for a leg. It is important that both the affected limb and its unaffected counterpart be measured.

Having one limb somewhat larger than the other does not necessarily mean there is anything wrong. If you are right-handed, for instance, your right arm may naturally be a little larger than your left. In fact, a healthy dominant arm may even be larger than a swollen nondominant arm. Generally, however, if one limb is two centimeters (about three-quarters of an inch) larger than the other, a doctor may make a diagnosis of lymphedema. Even with such a relatively small degree of swelling, you are likely to experience a feeling of heaviness or fullness in the affected limb. A difference of two and a half centimeters (about one inch) indicates moderate lymphedema.

Another way of measuring the limb is to submerge it in water and measure the volume of displaced liquid. But even though this method is highly accurate, it is rarely used because of its messy nature.

Imaging

In addition to measuring your limbs, your physician may recommend that you see a radiologist, a medical doctor who specializes in the use and interpretation of x-ray tests for diagnosis and treatment. This professional may perform one or more different tests to visualize the condition of the lymphatic system and to determine where any blockages may be. These tests include the color flow Doppler, magnetic resonance imaging (MRI), lymphangiography (LAS), and a computerized tomography (CT) scan.

Color Flow Doppler

Through the use of ultrasound technology, this noninvasive technique creates, an image of the flow of fluids through the vascular system and detects any blockages. To perform this test, the doctor places blood-pressure cuffs on different areas of the limb to measure the highest pressure of the artery. Then the cuffs are attached to a machine that measures the amplitude of the sound at each cuff. The test takes approximately fifteen minutes to perform.

Magnetic Resonance Imaging (MRI)

This test detects abnormalities of the circulatory system, discriminating between the muscle, fat, and fluid, as well as determining the extent of underlying tissue damage. It also shows enlarged lymph nodes, but not tumors or obstructions.

To undergo this test, you lie in a tunnel-shaped electromagnetic machine that is open at both ends. Short bursts of alternating electromagnetic energy—not radiation—emit tiny signals that are used to create cross-sectioned images of the area being observed.

While the process is lengthy—it takes about an hour—and requires you to remain immobile, it causes no discomfort or pain. However, according to a study published in the *Journal of the American Medical Association*, nearly 30 percent of those who undergo this test suffer anxiety-related reactions in the confinement of the machine. Some radiologic centers now have “open” MRI machines for people who tend to experience claustrophobia. The new machine has open ends and sides, allowing you a full range of movement of your limbs and an unobstructed view in four different directions.

Lymphangiography (LAS)

This is an invasive test in which a radioactive solution is injected into the tissues. Images are obtained as the radiotracer travels through the lymphatic system, creating detailed pictures of lymphatic channels and lymph nodes.

A combination of MRI and LAS is considered highly effective in viewing the lymphatic system, since MRI alone cannot detect abnormalities or other problems in the veins or arteries. MRI gives important information about soft tissues, lymph nodes, and blood vessels, and is particularly useful for determining whether vessels are obstructed, while LAS depicts the lymphatic channels and the functioning of the lymphatic system.

Computerized Tomography (CT)

This technology is sometimes used to help diagnose lymphedema. However, it involves doses of radiation and sometimes the injection of a contrast dye into the veins, which has the potential to cause severe allergic reactions. Therefore, many doctors prefer MRI scans. There are also other tests, such as venography and lymphography, that can be used to assess the functioning of the circulatory systems. However, these tests too involve injecting dyes into the veins, and they are associated with a high incidence of complications, including permanent swelling. As a result, these tests are not among the first choices for diagnosing lymphedema.

In spite of the seriousness and widespread incidence of lymphedema, few people have ever heard of it. It has been hidden from view by those who suffer in silence with its symptoms; by a medical system that offers a cornucopia of treatments for all sorts of other ailments but no comprehensive preventive or treatment strategies for lymphedema; and by a scientific community that has few research projects designed to give us a better understanding of how to prevent or treat this lifelong condition.

Thousands of people each year become vulnerable to lymphedema. Many of them might be able to avoid the condition altogether—if they learn what makes them vulnerable and what strategies might be employed to prevent it or, if necessary, to manage the condition once it appears. We will explore these subjects in the chapter that follows.

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