

Lymphedema: a condition impacting millions

In my hometown, there was a schoolteacher who had one arm that was three or four times larger than the other. She always wore sleeveless outfits, even in the winter. If she wore a jacket, the sleeve for the affected arm was enlarged and looked out of place. I was always curious as to what had happened to cause this problem. When I was older I learned that this lady had an operation for breast cancer and the swelling was a result of the surgery. I remember hearing that this school teacher was lucky to be alive, despite the fact she had been disfigured. All of this tragic had happened to someone who was one of the best dressed ladies in our community. In nursing school, I learned the official name for the condition, lymphedema, but it was not a condition that was widely addressed or emphasized.

In 1998, I was diagnosed with breast cancer myself, and the term lymphedema took on new meaning and greater importance. After my mastectomies I visioned myself as the schoolteacher I was so curious about thirty years before. You can imagine the fear when after a few weeks I noticed the swelling in my fingers, my bracelets were fitting tighter and there was this collection of fluid under my right arm pit and upper back that could not be drained away. I remember being told that lymphedema was a possibility because I had so many lymph nodes removed and if swelling happens, I would need to wear a compression sleeve to keep it under control. That was the extent of my education and preparation, for what would be a life long condition after my mastectomies.

As nurses we have a responsibility to help clients adapt and adjust to the changes that illness, and the consequence of illness, have on their lives. In the area of lymphedema, there is much that nurses need to learn to be the effective caregivers and advocates for clients with this condition. I am fortunate to live in a community that is progressive in most areas of health care and when I sought help in dealing with my lymphedema I was able to find a program in the area that specialized in the treatment of the condition using a process of lymph massage along with compression garments and other strategies. As a result of my treatment, my right hand and arm appear normal and I have just minor swelling in my upper shoulder and back, more importantly my clothes fit.

I shared my story with you so that you could have a sense of the importance of this issue in the lives of many of the clients that we serve. The remainder of this article is written with one of the therapist who has cared for me over the past three years. It is hoped that you will use the material presented here to spark your interest in the topic of lymphedema and become informed so that you can be an effective resource in your community on the condition.

What is Lymphedema?

Lymphedema, which is also spelled *lymphoedema*, is abnormal swelling of the tissues, due to the presence of excess protein-rich fluid known as lymph. This swelling occurs when the lymphatic system malfunctions or is damaged and lymphatic fluid cannot drain as quickly as it is produced. It affects an estimated 100 million men, women, and children around the world, including at least 3 million Americans. The story that is told in the beginning of this article is about lymphedema of the arm as a result of treatment for breast cancer. Breast cancer is only one of the many causes of lymphedema. Therefore, almost every nurse will come across lymphedema in her practice. Lymphedema most commonly occurs in the extremities (arms or legs); however, it can also affect the trunk, breast, abdomen, neck, head, and or genitals.

The fluid that causes the swelling of lymphedema is protein-rich and this makes the tissues easily susceptible to infections. Each infection damages lymphatic structures and places the area more at risk for developing lymphedema. Once the lymphatic systems malfunctions or lymphatic structures are damaged and lymphedema has developed it can be treated but not cured. Early treatments can usually effectively control the swelling. Without treatment, the lymphedema symptoms become progressively more serious. Left untreated, lymphedema is much more serious than the inconvenience of a swollen limb. It is a progressive condition in which frequent infections cause serious pain and may require hospitalization. Skin changes, like benign papillomas may develop. Eventually, lymphedema can lead to the loss of mobility and

total disability. In rare occasions malignant skin cancer develops, like lymph-angiosarcoma (Stewart-Treves-Syndrome.)

The Categories of Lymphedema

Lymphedema is classified as being either primary (genetic) or secondary (acquired.)

Primary Lymphedema

Primary Lymphedema, usually, is caused by the malformation of lymphatic vessels and affects approximately one out of every 6,000 individuals. Typically, the characteristic swelling begins at the foot and progresses upward toward the body. Primary lymphedema is congenital and can have its onset at different moments in life. The phase of life in which primary lymphedema develops is described in the following paragraphs.

Millroy's Disease is lymphedema that is present at birth or develops within the first two years of life. Lymphedema that is present at this early age is not always called Millroy's Disease. Lymphedema can also be present at birth as part of the genetic condition Turner's syndrome.

Lymphedema Praecox, also known as *Meige Disease*, is lymphedema that appears in puberty. Approximately 75% of all primary lymphedema cases develop during this period. Lymphedema Praecox affects four times as many females as males and the swelling usually occurs in only one leg.

Lymphedema Tardum, which develops around the age of 35, affects both males and females; however, it occurs most frequently in females. Onset of this condition is sudden, often with no apparent cause, and the swelling may affect one or both of the legs.

Secondary Lymphedema

The lymphedema that is referred to in Lorna Harris' personal story is known as secondary lymphedema. This is the more common form of lymphedema and is an acquired disorder caused by damage to the lymphatic system. As part of the treatment of cancer, lymph nodes in the armpit or groin are removed. With cancer surgery the limb adjacent to the surgery area is most frequently affected; however, lymphedema can develop in more parts of the body, including the head and neck, trunk (chest and abdomen), or the genitalia. In contrast to primary lymphedema, this swelling can begin anywhere in the limb, depending which nodes are removed, how heavy radiation was and how severe scarring had become.

Risk Factors for Developing Secondary Lymphedema

Not only, treatments for breast cancer makes an individual at risk of developing secondary lymphedema, there are more damaging actions that can cause trauma to the lymphatic system. When any of the following potential causes are part of the medical history, the person can develop secondary lymphedema:

Cancer treatments that include a biopsy, the removal of lymph nodes, radiation therapy, chemotherapy, or a combination of these treatments that result in the destruction of lymphatic structures.

Burns, radiation, or injuries that destroy large areas of the skin or damage the lymphatic capillaries located in, and just under, the skin.

Scars due to injuries or surgery can block the normal flow of lymph.

Excess weight places additional stress on the lymphatic system.

Conditions affecting the circulatory system, such as Congestive Heart Failure or Chronic Venous Insufficiency, place added stress on the lymphatic system.

Conditions that limit muscle movement, such as paralysis or multiple sclerosis, prevent the normal muscle activity that aid in the flow of lymph.

The Treatment of Lymphedema

Treatment of lymphedema consists of a myriad of different techniques and areas and is called Combined Decongestive Therapy (CDT.) The goal of CDT is to improve drainage through the still existing healthy lymphatic vessels and so decongest the tissue and decrease swelling. The initial treatment period is an intensive time in which the individual gets daily or twice daily therapy sessions after which the involved

limb is bandaged to keep the swelling down, so the limb cannot fill up with lymph fluid in between treatments. The period of intensive treatment last till the lymphedema has come to a point that no more improvement can be expected. Usually, this state can be reached after ten or more days. When there is a status quo of the lymphedema, the person is measured for therapeutic compression garments. Exercises are important to help the lymphatic system in moving the lymph fluids. In short are here the four steps of CDT:

- Skin care
- Manual Lymph Drainage
- Compression Therapy and
- Exercises

Skin Care

Normally a thin protective layer, which is known as the *acid mantle*, is maintained on the surface of skin by the flow from the pores of oil from the sebaceous gland and water from sweat glands. This film is mildly acidic with a pH ranging from about 4 to 5.5. (A pH of 7 is neutral. A pH lower than 7 is acidic and a pH higher than 7 is alkaline.) Maintaining this acid mantle is very important because it inhibits the growth of harmful bacteria and fungi on the skin. Unfortunately, it can easily be disrupted for many reasons including:

- The swelling and fragility of the skin due to lymphedema.
- Washing the skin with harsh soaps that remove this protective coating.
- Ongoing exposure to the sun, wind, cold and harsh weather elements.

Therefore, maintaining the acid mantle is an important part of treating and managing lymphedema.

When starting the treatment session, skin inspection is performed to determine if there is an area that needs special attention. The areas in between the fingers and toes and around the nail bed are prone for getting infected by fungi. Daily checks of these areas, especially when peripheral neuropathy exists in concomitance with lymphedema, needs to be performed. Also, check for unnoticed cuts or insect bites. The skin needs to be washed and cleaned with a perfume free, anti-bacterial soap. It is important to dry the skin thoroughly. Wet edematous skin is known for getting infected easily. After receiving Manual Lymph Drainage and before compression is applied, antibacterial ointment is applied on detected cuts or insect bites and the skin needs to be creamed with a pH-neutral lotion or crème (for example; Eucerin®) to keep the skin moist and healthy. Compression garments and bandages that are worn to prevent the arm from swelling also dry out the skin. If those lotions or crèmes are not applied to the skin, the skin can crack and is an easy 'port of entry' for bacteria that can cause erysipelas and cellulites.

Manual Lymph Drainage

Manual Lymph Drainage (MLD) is a pumping, circular massage stroke that stretches the lymphatic vessels by means of a light stretch during the circular movement of the stroke. Lymph vessels start in the hand and feet and follow the veins up into the body. The smooth muscles in the vessels react to the stretch of the MLD-stroke by way of a contraction and they help move up the lymph fluid. Valves prevent the lymph fluid from draining in a reverse way. Behind the chest bone, lymph passes the Thoracic Duct after which it enters into the venous system.

By applying MLD to the neck first, the lymphatic system in this area is opened and ready to receive lymph from the congested area. Once the neck area is cleared, the therapist will massage the non-involved side. Then the involved side is treated. If there is only mild edema it is sufficient to use superficial MLD-strokes. When the edema is more severe with pockets of hardened tissue, also called fibrosis, deeper edema techniques can be used to drain deeper in the tissues and to break up those pockets of fibrosis. Especially, in edema after cancer treatments fibrosis easily develops after heavy radiation and it can block proper drainage. Also, non-treated primary lymphedema of the legs can get those tissues changes.

Compression Therapy

There are several ways of using compression to treat and manage lymphedema. Compression helps decrease the limb, help drain the tissues, and prevent an increase in size. In the initial treatment phase short-stretch bandages are donned to adjust to the changing shape of the limb easily and to provide equal pressure all around the limb. Often, multi layers of different compression materials, like foam and quilted pats, help in softening fibrosis and shaping the limb. Once the lymphedema is stable, and no more improvement is to be expected, the limb is measured to fit for compression garments. For daytime use a compression sleeve is worn and for the night a quilted compression device. When the client is not able to

apply bandages him or herself, nighttime devices can be applied during the day for easy donning and doffing (= to place or remove an article of clothing on a person.)

Exercises

Exercises play an important role in bringing down the size of a limb and in maintaining the reached level. One aspect of exercising is that it improves lymphatic drainage in the impaired area itself. Another aspect is that it has a positive impact on cardio-vascular function, improves overall lymphatic function, and enhances overall health and well-being. Usually, exercises are performed wearing bandages or compression garment.

Exercises in the impaired area focuses on improving lymph drainage and maintaining Range of Motion (ROM) of the involved joints. The exercises need to be performed in a gentle and slow manner to accommodate the slow pace of the lymphatic flow. Deep abdominal breathing, engaging the diaphragm, is key to helping fluid flow and improves lymph drainage out of the trunk up into the chest area. The compression garment or short-stretch bandages that are worn while performing the exercises, act as a counter force to the lymphatic system and aids in better lymph drainage out of the impaired area.

Cardio-vascular exercises, or endurance exercises, increase heart rate and breathing for an extended period of time. Activities like walking, swimming, or bicycling are an important part of any exercise regimen because they:

- Improve the circulation of both blood and lymph,
- Aid in the flow of fluid away from affected areas, and
- Combat fatigue.

Endurance exercises can all start on a very easy level but individuals can be trained to a more strenuous level with a high cardio-vascular output that moves a lot of lymph fluid. The level of physical condition one had before one developed lymphedema is in general the level that one can work their way up to with these exercises.

When One is at Risk for Lymphedema

As a nurse it is important to know what a healthy life style is in regard to conditions that place a person at risk for developing lymphedema. The next list shows *what not to do* and *what to do* to avoid developing lymphedema:

- When traveling by air, wear a prescription compression garment on the “at risk arm” during all flights.
- Never allow a blood pressure measurement, injection, blood draw, or IV placement in the at risk limb.
- Body weight should be within a normal range.
- Exercise and eat well to maintain good health.
- Sunburn and other injury on the “at risk for lymphedema” area should be avoided.
- A cut in the skin needs to be cleansed thoroughly and an antibiotic cream needs to be applied.

Early Warning Signs of Lymphedema

Clients might complain of minor discomforts that not directly give the impression that their lymphatic system is compromised and that lymphedema is developing. Refer for CDT-treatment if your client complains of the following symptoms:

- A feeling of heaviness in the affected area.
- Discomfort, such as a “pins and needles” sensation, in the affected area.
- A sensation of tightness of the skin surrounding the affected tissues.
- Swelling, no matter how slight.
- Changes such as a ring that no longer fits, a watch that is too tight, or a shoe that is suddenly too small.
- Aching in the adjacent shoulder or hip.
- Decreased mobility in the affected joints (wrist and fingers or ankle and toes).

- An infection within the “at risk” area. Any infection requires immediate medical care!
- Pitting edema. To test for pitting edema, press a finger against the swollen tissue. If this pressure creates an indentation that gradually goes away, pitting edema is present.

If a client complains about one of these discomforts, please listen and respond by referring the client to the appropriate health care provider promptly! Once lymphedema begins, it will not go away by itself and without proper treatment, the lymphedema symptoms become progressively more serious. It will lead to increased swelling, which can be painful, disfiguring and can limit mobility and it will lead to progressive fibrosis, which will impair lymph drainage and increases the risk of infection.

Who treats Lymphedema?

In 2000, the National Lymphedema Network (NLN), which is an American organization that represents the interests and concerns of lymphedema patients and therapists, felt the need to create national standards to qualify as a lymphedema therapist. The Lymphology Association of North America (LANA) was founded with the mission of establishing training standards for lymphedema therapists and creating a national program to certify qualified therapists. Only those, who are already licensed professionals in related medical fields, are allowed to train to become lymphedema therapists. This includes physical therapists, physical therapy assistants, occupational therapists, certified occupational therapy assistants, registered nurses, physicians, and massage therapists who have completed 500 massage-school hours and/or have National Therapeutic Massage and Bodywork Certification.

LANA standards for schools offering CDT-training is at least 135 hours of specialized lymphedema education, of which 1/3 consists of theoretical instruction in the anatomy and physiology of the lymphatic system and 2/3 consists of hands-on mentored training. One year after graduating from a CDT-program, the healthcare professional can sit for the LANA-exam. LANA’s website <http://www.clt-lana.org> lists those therapists that have passed the exam and earned the right to call themselves CLT-LANA (Certified Lymphedema Therapist –LANA.) LANA certification is a relatively new and voluntary program. There are many well-qualified lymphedema therapists who have met the LANA training requirements but have not elected to take part in the exam. Those healthcare professionals usually call themselves CLT (Certified Lymphedema Therapist.)

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