National Lymphedema Network

Position Statement of the National Lymphedema Network

The Diagnosis and Treatment of Lymphedema

By the NLN Medical Advisory Committee; February2011

Introduction

Lymphedema is caused by an abnormality of the lymphatic system leading to excessive build up of tissue fluid that forms lymph, known as interstitial fluid. Stagnant lymph fluid contains protein and cell debris that causes swelling of affected tissues. Lymph is responsible for transporting essential immune chemicals and cells. Left untreated, lymphedema leads to chronic inflammation, infection and hardening of the skin that, in turn, results in further lymph vessel damage and distortion of the shape of affected body parts. [1-4,199,201]

Interstitial fluid can build up in any area of the body that has inadequate lymph drainage and cause lymphedema. Lymphedema is a condition that develops slowly and once present is usually progressive.[143,192] People can be born with abnormalities in the lymphatic system. This type of lymphedema is known as Primary Lymphedema. Depending on how severe the condition is, swelling can be present at birth or may develop later in life.[198] Most lymphedema in the United States is Secondary Lymphedema. This type of lymphedema occurs from damage to the lymphatic system, commonly from cancer and its treatment but also from trauma to the skin such as from burns or infections. [5,189] Lymphedema after breast cancer has been studied the most, but lymphedema can occur as a result of other cancers, including melanoma, gynecologic cancer, head and neck cancer and sarcoma.[76-78, 185-187] The overall risk of lymphedema for all cancers is reported to be 15.5%.[186] The risk of developing lymphedema does not diminish over time but is a lifelong risk. [6,143] Progressive lymphedema is complicated by recurrent infections, non-healing wounds, discomfort or pain, difficulty with daily tasks, emotional and social distress.[7-9]

Effective treatment for lymphedema is available. Early diagnosis is important since treatment is most effective when lymphedema is diagnosed at the earliest stage.[188,193,194] Every patient with lymphedema should have access to established effective treatment for this condition. Lymphedema has no cure but can be successfully managed when properly diagnosed and treated.

Diagnosis of Lymphedema

Since lymphedema is progressive and early diagnosis leads to more effective treatment, the diagnosis of lymphedema at the earliest possible stage is very important. Treatment of lymphedema is based on correct diagnosis. Many conditions that cause swelling (edema) are not lymphedema. True lymphedema is swelling caused by abnormality in the lymphatic system. Lymphedema can also co-exist with other medical and swelling conditions. Correct diagnosis of lymphedema may require evaluation by a physician or other health-care provider with expertise in lymphedema who can, when needed, perform specialized diagnostic testing.[10,198] Diagnostic tests for lymphedema come under the following categories:

- History and physical examination
- Soft tissue imaging
- Lymph vessel and lymph node imaging
- Measures of volume
- Changes in electrical conductance
- Changes in biomechanical properties
- Genetic testing
- Other vascular imaging
- Blood tests for other conditions that can look like

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History and Physical Examination

A history and physical examination by a health-care provider who has experience with diagnosis and treatment of lymphedema is important for all patients with chronic swelling.[193,194,198] Primary and Secondary lymphedemas have characteristic features that can be seen over time. The history should include age of onset, location(s) of swelling, pain and other symptoms, medications that can cause swelling, the course of progression of the swelling, and factors associated with swelling onset such as cancer, injury, or infection. A family history is important to the diagnosis of inherited forms of lymphedema. The physical examination includes an assessment of the vascular system (lymphatics, veins and arteries), skin and soft tissues in the swollen body part(s), palpation of lymph nodes, and looking for changes in body systems associated with various forms of inherited lymphedemas.[181,182,202] Diagnostic tests and imaging must be paired with the information from the history and physical examination to make a correct diagnosis. For trunk, breast, genital, head and neck lymphedema, the history and physical examination is the currently accepted method of diagnosis.[76,78]

Soft Tissue Imaging

Magnetic resonance imaging (MRI), computed tomography (CT) and some types of ultrasound (US) are able to detect the presence of extra fluid in the tissues.[159,160,170] Fluid that is outside of cells (extracellular) and also outside of vessels (extravascular) is called tissue fluid or interstitial fluid. Lymphedema is one type of interstitial fluid build up that occurs when fluid is not being removed effectively by the lymph vessels. MRI, CT and US can show the presence of increased interstitial fluid but cannot tell the cause. These imaging techniques have to be put together with history, physical examination and sometimes other imaging tests.[194] Other conditions such as heart failure or low proteins in the blood from liver disease or malnutrition can cause fluid to build up in the tissues. MRI, US and CT scans may be required to determine the cause of lymphedema, especially if there is a concern that the lymphedema might be the result of an untreated cancer.

Lymph Vessel Imaging

Lymphoscintigraphy is a nuclear medicine study used for imaging lymph vessels and lymph nodes.[195] Radio-labeled particles of protein are injected just under the skin of the area of the body to be imaged. Usually technetium labeled sulphur colloid is used. Lymphoscintigraphy is accurate for detecting abnormalities of the lymphatic system in the extremities regardless of the cause.[163,165,189] It demonstrates slow or absent lymph flow and areas of reflux (backflow). Lymphoscintigraphy can reveal abnormalities of lymph uptake in lymph nodes with some forms of lymphedema.[168] Lymphoscintigraphy can predict response to treatment.[167] Lymphoscintigraphy shows the main, larger lymph vessels and nodes. It shows the basic architecture of the peripheral lymphatic system. It does not show the deep transport lymph vessels carrying lymph from the nodes back to the blood circulation. Lymphoscintigraphy identifies lymphatic abnormalities at a late stage, after lymphedema has occurred. The type of lymphoscintigraphy done for the diagnosis of lymphedema is not available at all radiology departments. Most radiology departments, however, can do a form of lymphoscintigraphy used to identify the sentinel lymph node for cancers such as breast and melanoma. These studies for the sentinel lymph node are different from the lymphoscintigraphy studies done for diagnosis of lymphedema. Before undergoing a lymphoscintigraphy study the patient should inquire if the radiologist performing and reading the study has a large amount of experience with lymphoscintigraphy studies for the diagnosis of lymphedema. Lymphoscintigraphy, in combination with other vascular studies, can differentiate venous edema from lymphedema.[190,196] Lymphoscintigraphy may not be necessary in some forms of secondary lymphedema where the diagnosis is clear from the history and physical examination or other imaging. In order to diagnose primary lymphedema, however, a lymphoscintigraphy must be done. Especially in children, a detailed study must be done that includes all potential areas of involvement and the contralateral normal limb or body part for comparison.[166,169] These studies must be done by a radiologist familiar with primary lymphedema and genetic forms of edema. In children being evaluated for lymphedema, other vascular and imaging studies are necessary because primary lymphedema can occur in combination with many vascular abnormalities and other organ defects. The specific

tests needed should be determined by a specialist in lymphedema.

A new technique for imaging lymph vessels is Near Infra-Red Florescence Imaging (NIR) using a substance known as indocyanine green (ICG).[82,171-175] The ICG is injected into the skin and immediately imaged with a dynamic (real time) infrared florescence camera. With NIR-ICG, even very small lymphatic vessels can be seen. The study is dynamic which means that the actual function of the lymphatic vessels can be analyzed. Diseased lymphatics that do not contract (or pulse) normally can be seen with NIR-ICG. ICG is a green dye that has been used safely in other areas of the body such as the liver and eyes. It can be used in very small amounts to image the lymphatics. NIR-ICG can diagnose lymphedema and find abnormalities at an early stage, possibly before swelling is obvious. Although this technique shows promise for the diagnosis of lymphedema, it is currently available at very few centers, most of which are involved in research.

Measures of Volume

Measures of limb (arm and leg) volume have been the standard way of detecting lymphedema for years and have been shown to be accurate when properly done.[156,157,161,170] Enlargement of the limb (increase in volume) is the end result of fluid building up in the tissues. Therefore, volume measurements are used to quantify the presence and severity of lymphedema and follow the response to treatment. Volume is measured by 3 main methods: tape measurements, perometry, and water displacement.[170] Tape measurements are taken at defined intervals, using geometric formulas to calculate the total volume. This technique can be accurate if it is done in precisely the same way each time, and is most accurate when the same person takes the measurements each time. Perometry uses an infra-red optical electronic scanner and computer to calculate the volume of the body part. Perometry is accurate if the body part is positioned exactly the same way each time and the machine has been calibrated for accuracy. Perometry has been used for a decade in research on lymphedema and has been accurate when compared to the long used 'gold standard' of water displacement.[149] Perometry has been demonstrated to detect as little as a 3% change in limb volume in breast cancer survivors followed over time.[207] Water displacement, the bench 'gold standard' for assessing volume, is rarely used these days due to its inconvenience. The body part to be measured is immersed in a large cylinder and the water that is pushed out (displaced) is measured. All of these volume methods are effective and accurate when done properly.[170] They are most accurate on arms and legs. Measures of volume cannot differentiate lymphedema from other types of edema and do not determine when temporary post-operative arm edema becomes chronic lymphedema. Although tape measurements have been developed for head and neck,[78] they are not true volume measurements, nor have they been standardized. They are best used for following the effects of treatment rather than making a diagnosis.

Electrical Conductance Testing (BIS)

Bioimpedance Spectroscopy (BIS) is a method for measuring water content in tissues. It has been used for many years to assess the total water content of the body and body composition for fitness and weight loss purposes. BIS is now available to measure interstitial fluid as a component of assessment leading to the diagnosis of lymphedema.[144,148,149] BIS has been shown to provide reliable data to be used in the diagnosis of breast cancer-related lymphedema.[150] BIS can detect early changes associated with lymphedema.[162,164] BIS is done by passing a small, painless, electrical current through the limb and measuring the resistance to current (impedance). The machine uses certain electrical current frequencies to determine if more fluid exists as compared to the contralateral limb. It does this by comparing the difference in resistance to electricity passed through interstitial fluid compared to intracellular fluid. BIS currently is done on the whole limb since the resistance to current flow for standard technique is calculated to the length of the body part. The higher the water content in the interstitial tissue, the lower the resistance (impedance). BIS may show promise for detecting smaller areas of localized lymphedema, but this application has not been subjected to adequate study to recommend it.[206] BIS is not as accurate in advanced, fibrotic edema. As in measures of volume, BIS cannot differentiate lymphedema from other types of edema and does not determine when temporary post-operative arm edema becomes chronic lymphedema.[145,170,191]

Changes in Biomechanical Properties of Tissues

Lymphedema causes the affected skin and subcutaneous tissues to become inflamed and hardened (fibrotic).[199] Lymphedema is graded clinically, not just by increased size or volume, but also by the progressive change in the skin texture as it becomes denser and harder.[10,158] Currently, these skin changes are documented by physical examination of tissue texture, pitting, enlarged skin folds and other dermatologic conditions such as wounds or papillomas (benign growths on the skin in areas of lymphedema). Methods available for measuring skin texture and resistance quantitatively are: tissue dielectric constant[151-153] and tonometry.[148,152,154,155] The tissue dielectric constant is a measure of tissue water content. The test is performed with a device that passes an electrical current of a specific frequency to one location of the skin and measures the reflected wave that returns. The reflected wave form indicates the amount of water present in the tissue. Tonometry uses a device that measures the amount of force required to indent a tissue which gives a specific measurement value to the degree of firmness or fibrosis. There are some technical difficulties to the use of these tools and a number of environmental factors and operator differences can give variable values. These measures of the biomechanical properties of tissues are important for research. Hopefully continued development will lead to better tools for clinical use so that diagnostic methods will include quantitative methods of skin and subcutaneous tissue changes associated with progressive lymphedema.

Genetic Testing

For patients who have been diagnosed with primary lymphedema, genetic counseling and genetic testing may be appropriate.[176-178,184,202] All young children diagnosed with primary lymphedema should have a karyotype test performed. The karyotype determines the presence of chromosome abnormalities such as Turner's syndrome that can be associated with lymphedema. Other types of primary lymphedema involve specific genes. For example, Milroy's disease has a specific defect of the FLT4 gene that is responsible for producing a protein called vascular endothelial growth factor receptor 3 (VEGFR-3). The FOXC2 and SOX18 genes are also associated with lymphedema. Most forms of inherited lymphedema are not detected on gene or chromosome tests. Children diagnosed with primary lymphedema should be referred to a Medical Geneticist or a Genetic Counselor to determine which tests are indicated for that child's condition. With late onset of primary lymphedemas, genetic testing is of limited benefit, but Genetic Counseling may be offered on a case-by-case basis.

Other Vascular Imaging

Some forms of edema are caused by diseases or abnormalities in the cardiovascular system (heart, arteries veins). For children, and some adults, diagnosed with primary lymphedema, it is important to evaluate for other vascular abnormalities.[179,180] Conditions such as congestive heart failure, vein clots known as deep venous thrombosis (DVT), damaged vein valves known as venous insufficiency, and some arterial conditions can lead to swelling or exist concurrent with lymphedema. With secondary lymphedema from cancer, obstruction of a vein can contribute to the severity of edema.[196] Imaging studies of the heart, veins or arteries may be needed to get a complete and accurate diagnosis of the cause and proper treatment for edema.[190] The most common cardiovascular studies ordered for the evaluation of complex edemas are: echocardiogram, venous ultrasound and arterial ultrasound with ankle brachial index (ABI). Ultrasound studies of veins looking for a clot can be done lying down. To accurately diagnose venous insufficiency (incompetent valves) the ultrasound must be done standing or on a tilt table that can be tipped into a standing position (for patients who cannot stand for the test). If there is a concern for abnormalities of blood vessels in the chest, abdomen or pelvis, more advanced imaging, such as computed tomography venograms or arteriograms, may be recommended.

Other Diagnostic Tests

There is no blood test for lymphedema. Other medical conditions such as hypothyroidism (myxedema) or low protein (hypoproteinemia) can cause edema and need to be done in a complete evaluation of swelling. Standard plain x-rays may be ordered for some inherited lymphedemas to evaluate for orthopedic conditions.[182,183]

Treatment of Lymphedema: Complete Decongestive Therapy (CDT)

Complete Decongestive therapy is also called Combined,

Complex or Comprehensive Decongestive Therapy. All refer to the same method known as CDT. CDT is the main treatment for lymphedema. Experts who treat lymphedema consider CDT the "gold standard" of treatment.[11,12] CDT has been shown to be safe and effective.[13-19,197] CDT consists of an initial reductive phase (Phase I) followed by a maintenance phase (Phase II).[11, 20-23] In Phase I, the main goals are reducing the size of the affected part and improving the skin. After Phase I, the person with lymphedema needs to continue into Phase II, an ongoing, individualized self-management phase to make sure the gains of Phase I are maintained long term.[24]

Effects of CDT are to:

1. decrease swelling[25,26]
2. increase lymph drainage from the congested areas[27,28]
3. reduce skin fibrosis and improve the skin condition[1]
4. enhance patient's functional status[29]
5. relieve discomfort and improve quality of life[8,25,26,30-34]
6. reduce the risk of cellulitis and Stewart-Treves-Syndrome, a rare form of angiosarcoma[35-45,197]

Components of CDT

1. manual lymph drainage (MLD)
2. multi-layer, short-stretch compression bandaging
3. lymphatic exercise
4. skin care
5. education in lymphedema self-management, and elastic compression garments[22,46]

Frequency and Duration of Phase I (Reductive) CDT

Optimally, CDT is performed daily (5 days/week) until the reduction of fluid volume has reached a plateau, which can take 3 to 8 weeks.[22,47] Some patients may have good results from CDT with modifications of the frequency and duration of treatment.[49] CDT frequency and duration should be individualized to produce the greatest reduction of swelling and improvement of skin condition in the shortest period of time.

Maintenance (Phase II) CDT

At the completion of Phase I CDT, the person with lymphedema is set up on a self-management program that includes self-lymph drainage (sometimes called Simple Lymphatic Drainage), home lymphatic exercises, a skin care regimen, and compression garments or bandages that the individual learns to apply. Some individuals may require additional measures at home to maintain the gains achieved in Phase I. These measures may include garments with Velcro, specialized foam construction garments, and pneumatic compression devices.[136] Phase II maintenance must be monitored and changed periodically, just as treatment for any other chronic medical condition. Compression garments must be replaced every 4-6 months to be effective. Specialized equipment requires maintenance and replacement according to manufacturers' guidelines. Phase II CDT and periodic medical monitoring are essential to the long-term success of lymphedema treatment.[16,22-24]

Therapist Training

Therapists providing CDT should have completed at least 135 hours of training as recommended by the Lymphology Association of North America® (LANA®). (See NLN Position Paper: Training of Lymphedema Therapists.[205]) Additional specialty training may be required for therapists treating facial, truncal, and genital lymphedema, or lymphedema in people with complex illnesses or disabilities.

Manual Lymph Drainage (MLD)

Manual lymph drainage is an essential part of CDT. It is a specialized manual (hands-on) technique that appears to work by two mechanisms. It stimulates superficial lymphatic vessels to remove excess interstitial fluid and it moves it through subepidermal (under the skin) fluid channels that form when lymphatics are damaged. [47,201] Some people refer to MLD as massage, but it is different from the usual types of muscle or myofascial massage commonly known to the public. MLD is a light, skin technique learned by certified lymphedema therapists designed to improve fluid removal from congested areas where the lymphatics are not working properly and into lymph vessels and lymph nodes that are functioning.[48]

Compression Bandaging

Compression bandaging refers to a specific technique[54] utilizing multiple layers of several materials to create safe and effective gradient compression. The necessary components of

compression bandaging are:

1. Tubular bandage lining
2. Digit bandages
3. Polyester, cotton, or foam under-cast padding
4. Multiple layers of short-stretch bandages with 50% overlap and 50% stretch to cover the entire limb

In some patients, it is also necessary to utilize polyurethane foam in various densities and configurations within the bandaging system. These materials are applied according to standard technique to body parts with lymphedema. Short-stretch bandages have limited stretchability when pulled. They can stretch 40-60% from resting length, compared to long-stretch bandages such as Ace® bandages that stretch to greater than 140% of resting length. To achieve an effective compression gradient, short-stretch bandages must be strategically applied with low-to-moderate tension using more layers at the ends of the extremities than higher up.[50-55] Pressure within the short-stretch bandages is low when the patient is not moving ("resting pressure"). Muscle contractions increase interstitial fluid pressure to assist the fluid to move out of congested areas ("working pressure"), as muscles expand within the limited space of the short-stretch bandages.[56] The cycling between low-resting and high-working pressures in the interstitial fluid areas under the bandages creates an internal pump-like action. This action encourages movement of congested interstitial fluid into the vascular circulation. The short-stretch bandages also prevent refilling of the fluid into the tissues. Another property of short-stretch bandages is to reduce the tissue hardening (fibrosis).[1] Compression Bandaging is always a part of Phase I CDT. Some individuals with more severe forms of lymphedema may need to use home compression bandaging longer term as part of Phase II. Some locations of the body, such as the head and neck, are not amenable to standard short-stretch bandaging so other compression techniques have to be used.[204]

Exercise (including lymphatic "Remedial Exercise")

With lymphedema, specific exercise is beneficial for all patients.[203] Although heavy activity may temporarily increase fluid load, appropriate exercise enables the person with lymphedema to resume activity while minimizing the risk of exacerbation of swelling.[29,57-60,139] For people who have lymphedema, compression garments or compression bandages must be worn during exercise (except in aqua therapy) to counterbalance the build up of interstitial fluid.[58,61] (See NLN Position Paper: Exercise for Lymphedema Patients.[205]) Since exercise has been shown to have major positive effects during and after cancer treatment, safe exercise must be a goal for all cancer-related lymphedema.[60,62] For other forms of lymphedema, exercise also has positive effects. People with or at-risk for lymphedema are encouraged to work with a lymphedema specialist to incorporate an individualized exercise program into lymphedema management.

Skin and Nail Care

Meticulous hygiene is recommended to decrease the amount of fungus and bacteria on the skin. Low pH moisturizers should be applied to keep skin from drying and cracking.[63] Cracks and dry areas of the skin are entry points for bacteria and fungus, which can result in infections and wounds.[64,65] Skin infections are known as cellulitis (or erysipelas). Cellulitis is a serious infection of the skin that requires antibiotic treatment in people with lymphedema.[36-38] (See NLN Position Paper: Lymphedema Risk Reduction Practices.[205])

Compression Garments

Following achievement of maximal volume reduction with Phase I CDT, patients should be fitted with a compression garment. The patient should receive two garments at a time for each affected body part: one to wear and one to wash and dry. Having two garments insures that the patient does not wear a dirty or wet garment which promotes bacterial or fungal infection. Garments may be sleeves, stockings, bras, compression shorts, face or neck compression wear, etc. The type of garment depends upon the body part with lymphedema. Properly-fitted garments are essential for long-term control of lymphedema.[4,66] Garment style and compression strength should be prescribed according to the patient's ability to manage the garment and maintain the best volume control and skin health.[67] Ready-made garments come in a variety of sizes and can be fitted to many individuals. Custom garments are made specifically for the individual who cannot fit a ready-made garment. They are more expensive than ready-made garments. Custom garments may be required for patients with irregularly-shaped limb(s) or body parts, wounds, lack of sensation or

difficulty with hand dexterity. Custom garments are often a necessity for growing children. Custom garments allow for options such as special linings to reduce the risk of skin breakdown and fastening devices which can help the patient put on and remove the garment. Garments should be washed daily so the garment lasts as long as possible and does not lose its compression strength. Manufacturer instructions must be followed for washing and drying to prolong the life of the garment. Most daily garments must be replaced every 4-6 months to maintain compression strength. Replace compression garments for children when growth necessitates, which is usually multiple times per year for babies and younger children.

In addition to the day garments used in Phase II, some patients with more severe forms of lymphedema will need night garments or advanced day garments to maintain the reductions obtained in Phase I. There are a variety of options for advanced and night garments that may be required for control of lymphedema, such as Velcro closure garments and specialized foam compression garments.[68,136,137]

Patient Education

Since lymphedema is a life-long condition, patient education in self-management is very important.[69] To reduce the risk of developing lymphedema or having lymphedema worsen, all patients with lymphedema or at-risk for lymphedema should be instructed in essential self care. The important areas of education include risk-reduction practices, self-lymph drainage, skin care, signs and symptoms of infection, proper fit and care of garments, and the importance of good nutrition, exercise and weight control.

Weight Loss

Lymphedema risk increases with obesity, so weight loss should be a part of lymphedema treatment in overweight individuals, as well as maintenance of optimal weight in normal-weight individuals. [45,70-74] In one study, weight loss alone was shown to reduce arm volume in the lymphedema arm more than the uninvolved arm of obese women with post-mastectomy lymphedema.[75]

Modifications and Individualization of CDT

CDT programs should be individualized based on the presence of other medical conditions or patient abilities. Patients with wounds, scars, or musculoskeletal conditions; palliative care patients; or patients with post-radiation fibrosis may require adaptations of CDT. If there is limited mobility of the body part with or near the swelling, the patient may require other therapies, such as scar massage or myofascial therapy, in addition to CDT, to have a benefit from CDT.[49,64,65,68]

Decongestive Therapy for Head & Neck Lymphedema

Lymphedema can be a complication of treatment for head and neck cancer. Manifestations of lymphedema in patients with head and neck cancer are both internal (difficulty swallowing, vocal cord swelling) and external (swelling of the face, jaw and neck). Modifications of CDT have been shown to be beneficial, especially manual lymphatic drainage and modified garments. [76-78]

Intermittent Pneumatic Compression Therapy (IPC)

IPC, also known as compression pump therapy, can be useful in some patients as an adjunct to Phase I CDT[79- 88] or a necessary component of a successful home program (Phase II CDT).[89-91]

Single-chamber pumps, used in the past, are not used for lymphedema now. Single chamber pumps can cause fluid to move in both directions, meaning fluid can build up in the already-swollen area. Also, the pressure in single-chamber pumps does not stimulate lymphatic flow as sequential pumps do.[92] Acceptable pumps should have appliances (pump garments) with multiple chambers and have a sequential pressure delivery with the chambers compressing in a specific pattern determined individually for the patient's diagnosis and pattern of lymphedema.[93] Since lymphedema is a condition involving a quadrant of the body (upper or lower trunk, chest, abdomen), and not just the limb with the swelling, many patients who require IPC will need a pump that treats the trunk of the body and not just the limb with the swelling.

Recommended pump pressures generally range from 30-60 mmHg, although lower or higher pressures may be indicated.[200] The pressure displayed on the pump may not accurately reflect what is delivered to the skin surface. One study demonstrated considerable differences in skin/device interface pressure patterns and magnitude which may have an impact on therapeutic outcomes.[94] This is a significant

concern because superficial structures may be harmed if the pressures applied in therapy are too high.[95] In general, lower pressures are considered to be safer, but the pressure has to be individualized to the patient's diagnosis and skin condition.[95,200] The length of each treatment is usually one hour. IPC is not a "stand-alone" treatment. It is utilized along with standard CDT to maintain control of lymphedema at home.[90,91] (Phase II). To maintain edema control, a compression garment, or short-stretch bandages, should be worn between pump treatments and also when IPC therapy is discontinued.[136]

Patients being considered for IPC therapy must be evaluated by a physician or health-care provider with expertise in lymphedema. It is important to insure safe selection of the proper device and appropriateness of IPC. The prescription must include the intensity of pressure and pattern of pressure needed, taking into consideration several aspects of the patient's situation including determination of need for programmable pressure to treat fibrotic areas,200,201 address treatment of ulcers, and adjust for patient's level of pain and skin sensitivity. If trunk, chest or genital swelling is present, the physician must determine whether a pump that provides appliances to treat those areas is necessary or if the patient can manage the trunk swelling through self-MLD or garments. If a pump with only extremity attachments is used, close monitoring should be instituted to detect an increase in edema or fibrotic (hard) tissue above the device sleeve, called a fibroscelerotic ring.96 If this occurs, consideration should be given to using a device that treats the trunk in addition to the extremities. Additionally, the physician or health-care provider must evaluate the impact of various other medical conditions that are usually considered contraindications for pneumatic compression therapy, including acute infection, severe arterial vascular disease, acute superficial or deep vein phlebitis (inflammation or clot), recurrent cancer in the affected area, or uncompensated congestive heart failure.

Surgical Treatment of LE

Surgery for lymphedema is not curative, but it has been used in specific circumstances for control of a severe condition. Circumstances where surgery may be considered are: reducing the weight of the affected limb, minimizing the frequency of inflammatory attacks, improving cosmetic appearance, or fitting the limb into garments. As with all surgical procedures, the risks and benefits must be weighed against the individual needs of the patient, and the expertise of the surgical team. Surgery is usually only considered when adequate trials of all usual methods of treatment have failed.[97,98] There are several types of surgical procedures available that have been used for lymphedema: (a) excisional operations, including debulking and liposuction, (b) tissue transfers, and (c) microsurgical lymphatic reconstruction. There are very few surgeons who perform these procedures. It is extremely important that patients with lymphedema are treated by surgeons experienced in the care of lymphedema and who work with certified lymphedema providers for the patient's on-going care after surgery. Surgery for lymphedema must be done in conjunction with CDT.[147]

Debulking

Debulking surgery removes the hard connective tissue and any large folds of fatty tissue associated with the lymphedema-affected body part.[99-101] The potential risks of this surgery include prolonged hospitalization, poor wound healing, nerve damage or loss, significant scarring, destruction of the remaining lymphatic vessels in that body part, loss of limb function, return of swelling, poor cosmetic results, and decrease in quality of life. Post-operatively, compression garments are still necessary for the maintenance of the limb and must be worn life-long due to the lymphatic scarring from these surgeries and lymphatic insufficiency.

Liposuction

Liposuction involves the circumferential removal of fatty tissue deposits in the body part affected by long-standing lymphedema. It is generally performed under general anesthesia and involves the creation of many small incisions. Tubular suction devices are inserted into the incisions by the surgeon to break up, liquefy, and suction out the fat.[102-104] Liposuction for lymphedema is similar, but not exactly the same, as cosmetic liposuction. Tight bandaging is necessary to stop the bleeding after liposuction for lymphedema. Life-long compression garments are generally needed to prevent lymphedema from coming back due to the scarring of lymph vessels that can occur from the procedure. The risks of liposuction include bleeding, infection, skin loss, abnormal sensation (such as numbness, tingling, "pins and needles" feeling), and lymphedema returning.

Tissue Transfers

Tissue transfers (grafts) have been attempted to bring lymph vessels into a congested area to remove excess interstitial fluid. There are few studies of the long-term effectiveness of tissue transfers for lymphedema. Published articles are either outdated, done on animals, or describe lymph vessel function in breast reconstruction flaps.[104,105]

Microsurgical Lymphatic Reconstruction

Microsurgical and supramicrosurgical (much smaller vessels) techniques have been developed to move lymph vessels to congested areas to try to improve lymphatic drainage. Surgeries involve connecting lymph vessels and veins, lymph nodes and veins, or lymph vessels to lymph vessels. Reductions in limb volume have been reported and a number of preliminary studies have been done, but there are no long-term studies of the effectiveness of these techniques.[106-115]

Summary on Surgical Treatments

In general, surgical treatment is associated with significant risks, may result in reduced swelling for an unknown time, and is done by very few surgeons with experience in lymphedema. Surgical management of lymphedema should always be done in conjunction with CDT and does not stop the need for compression garments and Phase II maintenance. Since CDT, and other adjunctive therapies such as advanced garments and IPC, can usually produce good management in compliant patients, surgery is rarely a necessary consideration.[138]

Pharmaceutical Approaches

Lymphedema should not be exclusively treated with drugs or dietary supplements. Diuretics are ineffective for removal of interstitial fluid from the tissues. Excess diuretic use can lead to dehydration, electrolyte imbalance, and tissue damage. However, diuretics may be medically indicated in patients with lymphedema who have other medical conditions such as high blood pressure and heart disease. Therefore, diuretic use must be assessed on a case-by-case basis. Individuals with lymphedema should not stop diuretics before checking with their physician or health care provider.

Some drugs such as Coumarin (not coumadin) and Diosmin have been tried for lymphedema. They have not been found to be effective and have adverse side effects.[116-123]

Natural Supplements

There is limited evidence from rigorously-designed studies on the use of natural supplements for lymphedema. Studies have indicated American horse chestnut may help venous edema but not lymphedema.[124] Selenium has been reported to improve lymphedema in head and neck cancer.[125,126] Bromelain, a substance found in pineapple, has anti-inflammatory, anticoagulant, enzymatic, and diuretic effects. Some have wondered if there might be a benefit for bromelain use with lymphedema, but it has not been studied for use specifically for lymphedema.[127-132]

Due to potential interactions with prescription drugs and other negative side effects, patients should check with their physician or health-care provider before taking any natural supplement.

Complementary, Integrative and Alternative Treatments

A number of promising treatments have been reported, but they have not yet been subjected to sufficient rigorous research to recommend as the standard of care. These treatments include cold laser, electrical stimulation, vibratory therapy, oscillation therapy, endermologie and aqualymphatic therapy.133-142,146 All of these techniques are done in combination with components of CDT. Acupuncture has shown benefit for some symptoms of cancer and cancer treatment, including fatigue, hot flashes, muscular or joint pain, neuropathy and nausea. There are no rigorous studies on using acupuncture for treating lymphedema or using acupuncture on lymphedema extremities (see NLN Position Paper on Risk Reduction.[205]). Rebounder trampolines have been advocated by some for treating lymphedema, but there are no published studies on this treatment. Rebounding is good exercise, it but is not known to be superior to other forms of aerobic exercise in individuals with lymphedema.

Summary on Treatment and Diagnosis of Lymphedema

Treatment of lymphedema should be undertaken only after a thorough diagnostic evaluation has been done according to accepted guidelines by qualified practitioners. CDT is the current international standard of care for managing lymphedema. CDT has been shown to be effective in large numbers of case studies demonstrating limb volume reductions of 50-70% or more, improved appearance of the

limb, reduced symptoms, improved quality of life, and fewer infections after treatment. Even people with progressive lymphedema for 30 years or more before starting CDT have been shown to respond. Patient adherence during Phase II CDT is critical for preserving volume reduction. It is recommended that CDT adaptations or other lymphedema treatments be used on a case-by-case basis under the supervision of a health-care provider (physician, nurse, physician assistant, therapist) with demonstrated expertise in lymphedema management. IPC is a demonstrated effective adjunct to CDT. All interventions for lymphedema must have the goals of inducing and maintaining volume reduction, preventing medical complications, improving skin condition, reducing infection, enhancing patient adherence, and improving comfort and quality of life.

This document has been written and reviewed by members of the 2010-2011 NLN Medical Advisory Committee (MAC). MAC members have disclosed no relevant financial arrangements or affiliations.

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