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POSITION STATEMENT OF THE NATIONAL LYMPHEDEMA NETWORK

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TOPIC: TREATMENT

Introduction

Lymphedema (LE) is a chronic condition characterized by the abnormal accumulation of interstitial fluid due to insufficiency of the lymphatic system. Lymphatic dysfunction may be related to primary malformation of the lymphatic system, or to secondary causes. The leading cause of LE in the United States today is cancer and its treatment.

The progression of LE is characterized by swelling, as well as changes of the skin and subcutaneous tissue.¹ Changes typically manifest as roughness, dryness, and hardening of the skin.² Limbs may become grossly enlarged and distorted in contour with exaggerated skin creases, folds, and lobules.³ Progressive LE may be complicated by medical morbidity including recurrent tissue infections and non-healing wounds. Functional, psychological, and social morbidity can occur as well. LE has no cure but can be successfully managed following timely diagnosis with appropriate treatment. Diagnosis may require evaluation by a physician with expertise in LE and, when indicated, diagnostic testing.

Treatment of LE: Complete Decongestive Therapy (CDT)

CDT is comprised of an initial reductive phase (Phase I) followed by an ongoing, individualized maintenance phase (Phase II).⁴

The primary goals of CDT are to:

1. decrease edema
2. increase lymph drainage from the congested areas⁵
3. reduce subdermal fibrosis
4. improve the skin condition
5. enhance patient's functional status⁶
6. enable the patient to adhere to an independent self-care program

Components of CDT

Manual lymph drainage (MLD), multi-layer, shortstretch compression bandaging, remedial exercise, skin care, education in LE self-management, and elastic compression garments comprise CDT.⁷

Frequency and Duration of CDT

Optimally,^{7,8} CDT is performed daily until the reduction of fluid volume has plateaued, often after 3 to 8 weeks.

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 Training Position Paper.) Additional specialty training may be required for therapists treating facial, truncal, and genital LE, or LE in the context of advanced systemic illness.

Manual Lymph Drainage

Manual lymph drainage is a specialized manual (hands-on) technique which stimulates superficial lymphatic vessels. MLD may direct lymphatic flow out of congested areas and into functional lymph node basins.

Compression Bandaging

Multiple layers of short stretch bandages are applied to the lymphedematous area(s). Short stretch bandages have limited extensibility under tension (50%), in contrast to Ace^R bandages (300%). To achieve an effective compression gradient, bandages must be strategically applied with low to moderate tension using more layers in the distal, relative to the proximal, portions of the affected territory(ies). Pressure within the short stretch bandages is low when the patient is

inactive, “resting pressure”. Muscle contractions increase interstitial pressure, “working pressure”, as muscles expand within the limited volume of the semi-rigid bandages. Interstitial cycling between low resting and high working pressures creates an internal pump that encourages movement of congested lymph along the distal to proximal gradient created by bandaging. The non-elastic⁴ bandage sheath also counters refilling of fluid and reduces tissue fibrosis which further reduces volume.

LE Exercises (Remedial Exercise)

LE exercises are beneficial for all patients. Although activity and exercise may temporarily increase fluid load, appropriate LE exercises may enable the person with LE to resume exercise and activity while minimizing the risk of exacerbation of the swelling.¹⁰ Compression garments or compression bandages must be utilized during exercise to counterbalance the excessive formation and stasis of interstitial fluid. (See NLN Exercise Position Paper for exercise guidelines.)

Skin and Nail Care

Meticulous hygiene is recommended to decrease dermal colonization with fungus and bacteria. Low pH moisturizers should be applied to limit dermal desiccation and microbial growth.¹¹ Because of impaired local immunity in a lymphedematous limb, breaks in the skin may allow entry of bacteria and result in serious infections. (See NLN Position Paper on Risk Reduction.)

Compression Garments

Following maximal volume reduction with Phase I CDT, patients should be fitted with a compression garment. Properly fitted garments are essential for long-term control of LE volume.¹² Garment style and compression strength should be prescribed to enhance patient compliance and volume control. Garments should be washed regularly to maximize the garment’s longevity and effectiveness. Garments must be replaced at regular intervals.

Patient Education

LE is a life-long condition. Patient education in self-management techniques is therefore a critical dimension of effective treatment. All LE patients should be taught LE risk reduction, self- manual lymph drainage, the importance of skin care, the signs and symptoms of cellulitis, the proper fit and care of garments, the importance of weight control, and an individualized LE exercise program. Emphasis on specific LE self-care elements should be adjusted on a case-by-case basis.

Modifications and Individualization of CDT

CDT programs should be individualized based on the presence of concomitant medical conditions. Patients with wounds, musculoskeletal problems, adhesive scars, or post-radiation fibrosis causing limited mobility of the involved area or areas adjacent to the swelling, may require adjunctive therapeutic interventions in addition to CDT.¹³⁻¹⁵ Alternative nonelastic compression devices are often helpful adjuncts to simplify nighttime compression. These devices may enhance Phase II CDT effectiveness in persons who are unable or unwilling to apply traditional short-stretch compression bandages. In selected cases, they may be useful during Phase I treatment in combination with short-stretch bandaging.

Pressotherapy (Intermittent Pneumatic Compression, “Compression Pump”)

Pressotherapy is not a component of conventional CDT. Pressotherapy may be used as an adjunct to CDT.¹⁶ Pressotherapy involves insertion of the lymphedematous extremity into a multi-cell inflatable appliance, which is attached to an air compression pump. Sequential inflation and deflation of the cells creates a distal to proximal compression wave that moves the water component of the lymph and interstitial fluid out of the affected territory. There is a two-phase pump that creates a proximal to distal gradient (preparation phase) and a distal to proximal gradient (drainage phase) to simulate MLD.

Pressotherapy can decrease capillary filtration, thereby decreasing lymph formation. Pressotherapy does not accelerate lymph return¹⁷ and does not enhance the removal of the excess protein component of lymphatic fluid. Potential complications of pressotherapy include displacement of the edema to the proximal limb, adjacent trunk and/or genitalia. A fibrosclerotic ring may develop above the proximal end of the pump appliance, further obstructing lymphatic flow. The use of a pump should be supervised by a trained therapist or healthcare provider experienced in lymphedema management. Pump pressures generally range from 30-60 mmHg

Surgical Treatment of LE

The surgical treatment of LE has been advocated in a few specific circumstances to reduce the weight of the affected limb, to help minimize the frequency of inflammatory attacks, to improve cosmesis, and to potentially reduce the risk of secondary angiosarcoma.¹⁸⁻²¹ However, the surgical treatment of LE is not curative. The potential benefits of surgery need to be weighed against the risks, which are related to the extent of the procedure, the individual medical needs of the patient, and the expertise of the surgical team. There are two basic types of surgical procedures used for the treatment of LE: 1) excisional operations, including debulking^{21, 22} and liposuction,²³ and 2) lymphatic reconstruction.²⁴⁻²⁸ Most reports of the outcomes from surgical treatment for LE are drawn from the experience of a single institution or group of surgeons and focus on small numbers of patients with insufficient objective outcome data and long-term follow-up. In general, surgical treatment is associated with significant risks and does not eliminate the need for compression garments.²⁹⁻³¹

Pharmaceutical Approaches

LE should not be exclusively treated with drugs or dietary supplements. Diuretics may be harmful.³² Dehydration of soft tissues with diuretic therapy may increase interstitial protein concentration and secondarily potentiate inflammation and tissue fibrosis. Diuretics may be indicated in LE associated with systemic conditions (e.g. anasarca), but this must be medically assessed on a case-by-case basis. Benzopyrones, such as Coumarin and Hydroxyethylrutin and flavonoid derivatives such as Diosmin have been tested in research studies.³³⁻³⁷ The United States and Australia abandoned the use of oral Coumarin due to liver toxicity and inconsistent efficacy.³⁶ Trials with hydroxyethylrutin demonstrate improved skin softening. Daflon 500 (a combination of Diosmin and Hesperidin) is used widely in the equatorial regions of the world to treat LE. The benefit appears to result from the reduction of microvascular permeability^{38,39} and increased thoracic duct pumping.⁴⁰ However, these mechanisms have only been verified in animal models to date.

Natural supplements

Studies indicate potential benefit from American horse chestnut. The effects may be mediated by reducing venous capillary permeability.^{41,42} There are also studies demonstrating mixed benefits of selenium in radiation-associated LE.⁴¹⁻⁴³ Bromelain is a natural diuretic found in fresh pineapple.⁴⁴⁻⁴⁷ It appears to have an anti-inflammatory and diuretic effect that may have a beneficial impact.^{48,49} Further investigation of these and other supplements is necessary.

Conclusion

LE is a chronic condition characterized by the abnormal accumulation of interstitial fluid due to insufficiency of the lymphatic system, either as a primary or a secondary disorder. Diagnosis of LE may require evaluation by a physician with training and expertise in LE. Sometimes special diagnostic testing is needed. CDT is the current international standard of care for managing LE. The efficacy of CDT is supported by large case series demonstrating a tight temporal association between the initiation of Phase I CDT and limb volume reductions of 50-70%, enhanced cosmesis, and reduced incidence of cellulitis.^{1,3,7,50-}

⁵² Chronic or gradually progressive lymphedema for intervals up to 30 years or more prior to the initiation of CDT consistently respond to Phase I therapy. Patient adherence during Phase II CDT preserves the volume reduction.^{7,52} The literature establishes an emphatic causal relationship between CDT and lasting LE volume reduction.

At present it is unclear to what degree deviation from the intensive and integrated approach that is the hallmark of CDT, will compromise LE treatment outcomes. It is recognized that such deviations may be imposed by provider, patient, and third party payer constraints. Means do not exist to accurately identify which LE patients will be most adversely affected by changes in the intensity or components of CDT. A variety of alterations have been formally proposed, however, their impact remains undetermined.¹⁷ It is

therefore recommended that CDT alterations be instituted on a case by case basis under the supervision of an experienced LE therapist or lymphologist with the goals of preserving volume reduction, preventing medical comorbidity, and enhancing patient adherence and comfort.

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