Lymphoedema is a progressive, chronic oedema condition that can have significant effects on psychosocial health, physical health and quality of life issues for patients. Proper diagnosis, early detection and appropriate treatment can effectively slow the progression of Lymphoedema. Wound care specialists should be aware of this condition and address not just the wound but the proximal and distal oedematous areas contributing to delayed wound healing. Even if diagnosed properly, health care practitioners and patients are often unaware that specialised treatment is available to treat Lymphoedema in South Africa and may not know where to seek help. This is one reason why the Lymphoedema Association of South Africa (LAOSA) was formed in 2012 by a group of Lymphoedema Specialists, each with over 135 hours of International Certified Training.

**What is Lymphoedema?**

Lymphoedema is an abnormal accumulation of protein-rich fluid in the interstitium which causes chronic inflammation, skin integrity changes and fibrosis. It occurs most frequently in the extremities; however, it may also affect the head, neck, breast, abdomen and/or genitals and occurs when there is impaired lymphatic function or an inadequacy in transporting lymph fluid through the body. The lymphatic system is a ‘waste removal system’ or drainage system parallel to the venous system. The initial lymph capillaries transports away all cell waste products in the interstitial spaces that cannot be re-absorbed into the venous capillaries and returns it to the circulatory system via the ductus thoracicus. The lymph system does not have a pump for moving lymph fluid similar to the heart pump that moves blood in the circulatory system. Lymph fluid propulsion through the lymph vessels is dependent on effective muscle contraction, diaphragmatic breathing and peristalsis. Since Lymphoedema is a progressive condition it is imperative that it is diagnosed promptly in the earliest possible stage because early diagnosis leads to more effective care and cost effective treatment options. Lymphoedema can and usually does co-exist with other medical and swelling conditions and can be a challenge to recognise and diagnose correctly.

**Classification of Lymphoedema:**

- **Primary**- This is associated with congenital or developmental abnormalities of the lymphatic system. It may occur in newborns (Congenital Lymphoedema), in adolescents (Lymphoedema Praecox), or in people over the age of 35 years (Lymphoedema Tarda). This occurs less often in men. It is usually unilateral and tends to be worse during warm weather, before menstrual periods, and after prolonged dependency.

- **Secondary**- This is caused by an event or result of surgery, radiation, infection, trauma or burn that occurs when lymph nodes or vessels are removed or damaged and can no longer effectively filter waste products. Filariasis is another form of Lymphoedema caused by a parasitic worm infection transmitted by a mosquito bite typical in developing countries.

**Incidence of Lymphoedema**

It is unknown how many people in Africa live with Lymphoedema. The unavailability of statistics is merely part of a worldwide dilemma.
In 2014, the World Health Organisation (WHO) estimated that worldwide 1-2% of the population suffers from chronic Lymphoedema; 120 million people worldwide suffer from Lymphatic Filariasis and nearly 1.4 billion people in 73 countries are estimated to be at risk of infection; 90 million caused by parasites. An estimated 25 million men suffer with genital disease and over 15 million women are afflicted in the leg; 20 million caused by breast cancer; 2–3 million people with primary Lymphoedema. The incidence of breast cancer related Lymphoedema ranges between 6% and 83%. The economic burden of chronic wounds and associated Lymphoedema are better documented in the developed world and appear to be high. In South Africa, with the growing epidemic of non-communicable diseases, emerging infections, longer life expectancies and slow improvements of socioeconomic conditions, it is likely that the prevalence and impact of chronic wounds and Lymphoedema will increase. To put things in perspective, South Africa’s population is currently 53 million people; conservatively at 1-2% prevalence in the population, estimates could be as high as 530,000-1.06 million people with some form of Lymphoedema.

**Stages of Lymphoedema (International Society of Lymphology)**

**Stage 0 - Latent Stage**
- No visible changes
- Oedema is not evident despite an impaired lymphatic drainage system
- System is still effectively compensating

**Stage 1 - Reversible Lymphoedema**
- Early accumulation of protein-rich oedema fluid with visible oedema
- Pitting oedema noted
- Oedema reduced by elevation of the limb but returns when limb is reverted to normal position

*Prompt treatment can control the condition and may prevent it from becoming more severe.

**Stage 2 - Spontaneously Irreversible Lymphoedema**
- Increase of protein-rich oedema fluid and noted tissue changes in the skin
- Increase risk of fibrosis, infections, and skin problems

*Pressure against limb produces only a slight indentation-pitting or no indentation at all
*Elevation of limb will not completely reduce oedema

*Can improve with intensive treatment.

**Stage 3 - Lymphostatic Elephantiasis**
- Increased protein-rich oedema fluid causing skin tissue to become extremely oedematous and fibrotic
- Normal skin elasticity is lost and skin may hang in folds
- Skin may change colour
- Papillomas-small solid benign tumors that project above the surrounding tissue may develop
- Hyperkeratosis-increase in the thickness and hardening of the dermal layer of the skin may develop
• Limited mobility
• Pressure does not produce pitting
• Infections become more common-cellulitis
• Increased risks of breaks in skin-fungal infections and open wounds may form within the fold of skin

*Requires intensive treatment.

Lymphoedema Assessment

Treatment of Lymphoedema is based on correct diagnosis which would require an evaluation by a qualified practitioner. Lymphoedema has distinguishing features that can be seen over time. The patient history and physical exam should include location and amount of oedema, Stemmer sign, assessment of skin integrity, age of onset and course of progression of swelling, factors associated with onset-injury, infection, surgery, cancer treatment-surgery with lymph node excision (how many nodes removed), radiation and measurement of both limb volumes by tape measure and/or water displacement.

Other diagnostic tests for Lymphoedema may include the following:
• History
• Physical exam
• Measure of Volume- tape measurement, water displacement or perometry
• Soft tissue imaging-MRI, CT, US
• Lymph vessel and lymph node imaging-Lymphoscintigraphy, NIR-ICG (near infra-red florescence imaging with idocyanine green)
• Changes in electrical conductance-Bioimpedance Spectroscopy (BIS)
• Changes in biomechanical properties-tissue dielectric constant and tonometry
• Genetic testing-for primary Lymphoedema a karyotype test and testing for a gene called VEGFR-3, FOXC2 or SOX 18
• Blood tests –there is no blood test for Lymphoedema-rule out other chronic oedemas such as hypothyroidism(myxoedema) or low-protein hypoproteinaemia (liver or renal disease)
• Other vascular imaging-Echocardiogram, computed tomography venograms or arteriograms, venous ultrasound and arterial ultrasound with ankle brachial index (ABI). Patients with reduced ABPI of 0.5-0.8 should not receive sustained compression exceeding 25mmHg. Patients with ABPI <0.5 should not receive compression. If arterial involvement is suspected, referral to a vascular specialist should be made.

Lymphoedema Treatment

The gold standard of care according to the International Lymphoedema Framework, is Combined Decongestive Therapy (CDT). This is the most effective and accepted method of treatment and includes the following:

Phase I CDT: (Reducing oedema)
• Manual Lymph Drainage (MLD)
• Multi-layer short stretch compression bandaging
• Remedial Lymphatic and Diaphragmatic breathing exercises
• Skin care

Each CDT treatment usually lasts between 45-90 minutes in duration, 5 days per week for 2 to 8 weeks, reducing frequency once a plateau of oedema has been achieved. The frequency and duration can be modified and individualised to produce the greatest reduction and improvement in the shortest period of time. Diuretics are often prescribed for oedema, but are however ineffective for removal of protein-rich interstitial fluid from the tissues. Excessive diuretic use can lead to dehydration and electrolyte imbalance. Diuretics may be medically indicated in patients with Lymphoedema 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 and patients with Lymphoedema should not stop any medications before discussing with their physician first.

Phase II CDT: (Maintenance of oedema reduction)

Upon completion of Phase I, a person is set up with a home program that includes the following:
• Self-manual lymph drainage home maintenance program
• Self-bandaging or specialised foam with velcro compression device (night time compression)
• Daily lymphatic exercise program may include aqua lymphatic exercises
• Skin care-moisturise and skin inspection daily
• Use of day time gradient compression garments (20-30 mmHg-30-40mmHg or higher compression as indicated/prescribed to keep oedema reduced)
• Maintaining healthy, low sodium, low preservatives diet and maintaining ideal BMI

Conclusion

The past decade has seen a dramatic increase in the international understanding and awareness of Lymphoedema with a growing evidence base that supports the efficacy and cost effectiveness of utilising CDT with wound healing. In chronic non-healing wound
situations, therapeutic options are limited and expensive. When completed properly and safely by a qualified Lymphoedema specialist, CDT is a powerful and valuable adjunct, and surgeons and wound practitioners should be aware of its capability and potential to assist them in their wound resolution. Results from large numbers of case studies demonstrates combining treatments offers significant benefits, including fewer complications, better functional outcomes, reduced hospital visits, improved wound healing time, fewer infections after treatment, improved appearance of the limb, improved skin integrity, limb volume reductions of 50-70% or more and most importantly improved quality of life.

Selected References


Suzi Davey, OTR/L, CLT-LANA is a Lymphoedema Therapy Specialist and Consultant since 1998 now living in Hillcrest, South Africa. She is the founder of Healing Hands of Lymphatics LLC in Florida USA. She can be reached at lymphatictouch@yahoo.com. For more information about LAOSA or to find a certified Lymphoedema Therapist visit www.laosa.co.za