Chronic Edema/Lymphedema: The Hidden Epidemic

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Day 2 of the conference contains a fascinating block of presentations, under the heading International/WAWLC Sessions. These sessions have been organized by members of the World Alliance for Wound and Lymphedema Care and will provide attendees with insight about wound care in other countries and how lessons learned there can be applied to Canada. Lymphedema will be in the spotlight. To help readers and conference attendees understand more about this prominent but poorly understood condition, the following article provides an excellent primer on chronic edema/lymphedema.

hronic edema/ lymphedema has been described as the hidden epidemic. Yet despite significant morbidity it remains under-recognized, underfunded and under-treated. According to Stout et al., the main reasons for this is lack of public awareness, insufficient education and knowledge among health-care providers and failure of insurers to recognize the extent of the problem.¹ In a Canadian study carried out by Hodgson et al., many patients with lymphedema, especially those with non-cancer-related lymphedema and children with lymphedema, had limited or no access to treatment.² This comprom-

ised access stems from a lack of hospital-based services, an insufficient number of private clinics and the cost of treatment. Patients in rural/suburban areas may not have access to trained therapists or physicians who can properly diagnose and treat chronic edema/lymphedema.

Extent of the Problem

Primary lymphedema occurs in one in 6000 at birth. The most common cause of secondary lymphedema worldwide is lymphatic filariasis, a parasitic infection transmitted by a mosquito vector. It is estimated to impact more than 120 million individuals worldwide.³ About 40 million individuals are disfigured and incapacitated by the disease. Lymphatic filariasis is uncommon in the Western hemisphere, being endemic in only two locations: Léogâne, Haiti, and Recife, Brazil. It is estimated that three to five million people in the United States are affected by secondary lymphedema.

There are no Canadian statistics, but by extrapolation from the U.S. study it is estimated that at least 300,000 people in Canada may be affected by lymphedema. Likewise, there are few Canadian statistics documenting the numbers of people with secondary lymphedema; however it is suggested that the incidence of breast cancer-related lymphedema ranges from 6% to 70%, but lymphedema may be a common and under-reported morbidity.⁴ Up to 70% of men with prostate cancer may have lymphedema. Similarly, morbid obesity is rapidly increasing in North America, and 80% of these individuals are thought to suffer with an element of lymphedema.³

Definition of Chronic Edema/Lymphedema

Lymphedema is defined as an abnormal swelling of a limb and/or the related quadrant of the trunk due to the accumulation of protein-rich fluid in the tissue spaces of the skin.⁵ Clinically lymphedema may be defined as chronic edema lasting more than three months that is minimally responsive to overnight leg elevation or diuretics and is accompanied by skin changes such as thickened skin, hyperkeratosis and papillomatosis.⁶ Lymphedema may be primary or secondary. Primary lymphedema is related to the congenital absence or malformation of lymphatics and may appear clinically at birth or later in life. There is growing recognition that primary lymphedema is related to specific genetic abnormalities. Secondary lymphedema results from damage to lymphatics. Common causes of secondary lymphedema include chronic venous insufficiency,



obesity, recurrent infections, surgery—particularly surgeries involving damage or removal of lymph nodes or lymph vessels trauma, burns and radiation treatments. Medical conditions such as congestive heart failure, renal failure and liver failure may lead to chronic edema, which, if persistent, ultimately will lead to lymphatic dysfunction.

Pathophysiology of Chronic Edema/ Lymphedema

Lymphatic fluid (also known as lymph) primarily consists of water and protein filtrate. Lymph contains large quantities of macromolecules that are not absorbed into the arteriovenous capillary

> bed but are transported by the lymphatic system entering the venous system in the thorax. The lymphatic system also transports fat (chyle) and waste

products of metabolism.

Lymphatic failure is best discussed in the context of overall tissue fluid dynamics. The cells are nested in an extracellular matrix and are bathed in a constant flow of tissue fluid that nourishes and supports the cells as well as carries away the products of metabolism. Fluid moves under the influence of the push of hydrostatic pressure within the capillaries and in the extracellular compartment and the pull of osmotic force where fluid moves across a semi-permeable membrane from areas of low concentration of dissolved proteins to a region of higher concentration. Newer research has shown that in the extremities there is no net reabsorption of fluid into the capillary bed and that 100% of excess tissue fluid is handled by the lymphatics.⁷ A normal lymphatic system may fail because the capillary filtrate exceeds the ability of the lymphatic system to handle the fluid load.

Conference Tip:

Consider attending at least one

session that is completely outside your area of expertise or interest.

Gaining insight and information from

new people with different approaches

can be enlightening and inspirational!

There are three methods by which the lymphatic system may fail:

- 1. Dynamic insufficiency (or high-output failure)
- 2. Mechanical insufficiency (also known as low-output failure)
- 3. A combination of the two

In high-output failure the lymphatic system becomes overwhelmed, the tissue spaces become saturated with proteinacious fluid and swelling of the affected area occurs. If left untreated, the presence of macromolecules, such as growth factors, proteases and pro-inflammatory molecules may lead to chronic inflammation, infection and hardening of the skin.⁸ Additionally, the accumulation of cellular debris and blocking of the lymphatic vessels impede transportation of macrophages and lymphocytes—thus limbs affected by lymphedema are more prone to infections. Examples of high output failure include hepatic cirrhosis (ascites), nephrotic syndrome (anasarca) and venous insufficiency of the leg.

Low-output failure is characterized by decreased lymphatic transport. It may be related to congenital absence or malformation of the lymphatics, tissue damage from trauma or surgical treatment. It also my result from obstruction of the lymphatics by tumour, morbid obesity or scar tissue from thermal or chemical burns. Recurrent infection or other inflammatory conditions will ultimately lead to damage of the lymphatics. Lymph transport occurs within the lymphatic system through intrinsic contraction of the lymphangion, pulsations of adjacent arteries aided by calf-muscle-pump function. Thus chronic dependency of the limb and poor calfmuscle-pump action may also lead to low-output failure.

In situations where high-output transport failure is long-standing, functional deterioration of the current lymphatics system is inevitable and results in a reduction of overall transport capacity, leading to a combination of both high output and low output failure.

The Costs

Early intervention and management of chronic edema are key factors in reducing the risks to overall patient health, recovery, lifestyle and work. Unaddressed, these risks may lead to expensive hospitalization/IV treatment. These are avoidable costs, which will only multiply due to the escalating number of patients with obesity, complex medical conditions and cancer.

A prevalence study done in the United Kingdom by Christine Moffat in 2003 revealed that 823 patients in 619,000 had chronic edema.⁹ Of these:

- 27% were admitted to hospital for antibiotic treatment at an increased burden on the health-care system.
- 32% received some form of compression bandaging.
- 29% had an infection in the

12 months prior to the study.

- The mean length of stay was 12 days at a mean cost of £2300 (\$4200).
- 80% had taken time off work.
- 8% had to give up work.

The study estimated that for every £1 spent on lymphedema treatments £100 in hospital admission costs were saved.

In 2011 the challenges of chronic edema were compared between the UK and Canada. It was noted that both had a lack of public awareness, poor professional knowledge, inadequate information, delayed diagnosis and inappropriate treatment, poor understanding of treatment options, a lack of evidence-based guidance and difficulties ensuring concordance with treatment.¹⁰ The geographical issues patients in Canada face are considerable, and many patients deliver their own care. There is frustration with the lack of funding for lymphedema from both government and insurance companies.

Best Practice

The most comprehensive document on best practice is the document from the International Lymphedema Framework.¹¹ When combined with the recent Position Statement on Compression Therapy (www.lympho.org) it provides guidance to the practising clinician. The management of chronic edema/lymphedema has several components, which may include:

- Compression therapy
- Meticulous skin care
- Manual lymphatic drainage (MLD) done by professionals or simple lymphatic drainage done by patients or informal caregivers after instruction by a professional
- Exercise
- Intermittent pneumatic compression therapy
- Compression garments for maintenance

Of these, the most critical is compression therapy. Compression therapy works to enhance both venous and lymphatic drainage from the extremity. Short stretch, more rigid bandage systems have proven to be more effective than elastic systems in promoting mobilization of tissue fluid through:

- Reduction of the cross-section of the veins
- Improved valvular function, which reduces reflux and

decreases hydrostatic pressure in the veins

- Improved venous flow velocity by enhancement of calfmuscle pumping activity
- Increased tissue hydrostatic pressure leading to decreased net filtration

This has several beneficial effects on the lymphatic system:

- Reduction of venous congestion decreases net filtration and fluid load on the lymphatic capillaries.
- Increased interstitial tissue fluid hydrostatic pressure increases tension on the anchoring filaments, which will increase the ingress of lymph fluid and macromolecules into the lymphatic capillaries.
- Enhanced muscle-pump activity improves lymph propulsion though the lymph vessels.
- Down regulation of inflammatory cytokines leads to breakdown of fibrosclerotic tissue.

Summary

Lymphedema is a hidden epidemic. It is under-recognized and undertreated worldwide. It is a chronic condition and is best managed using chronic disease models that focus on self-management with professional support for assessment, treatment recommendations and monitoring. The cornerstone of treatment is appropriate compression therapy to manage edema. It requires a partnership between health-care systems, health professionals, persons living with lymphedema and their families or caregivers. 🥙

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