The Standard of Care for Lymphedema: Current Concepts and Physiological Considerations

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Abstract

The standard of care for lymphedema encompasses risk reduction, early detection, Phase-I Complete Decongestive Therapy in a clinical setting, and Phase-II Self Management at home. This review discusses these lymphedema treatment components, including relevant physiological aspects that form the basis of treatment and an understanding of which are critical to successful outcomes. Phase-I therapy includes manual lymphatic drainage (MLD), short-stretch compression bandaging, decongestive exercise, skin care, and sometimes intermittent pneumatic compression (IPC). Home management includes many of the same elements, with adaptations for home treatment. Significant to the success of Phase-I and Phase-II care is effective bandaging and lymphatic drainage treatment via manual (MLD) or automated (IPC) processes. Truncal clearance plays a key role in facilitating lymphatic drainage from affected limbs by achieving effective pressure gradients, reducing lymphatic network resistance, and stimulating lymphatic contractility. The transition from Phase-I to Phase-2 presents challenges to patients and clinicians. Poor patient compliance with time-consuming technique dependent-home care regimens contribute to losing gains achieved in the clinic. Ineffective treatment results in complications, including increased pain, reduced range of motion and mobility, increased risk of infection, and other physical and psychological sequelae. The use of an advanced programmable IPC device for lymphedema treatment may support successful home management by addressing some of the home treatment barriers. Device selection requires careful consideration of the mode of action and device functionality. An advanced programmable device that provides truncal and proximal clearance and low applied pressure represents the current choice for such in-home devices.

Introduction

In discussing the broad concept of the standard of care for lymphedema, it is useful to delineate four major focus areas; risk reduction, early detection, Phase I Complete Decongestive Physiotherapy in a clinical setting, and Phase II Self-Management at home. Drawing on relevant aspects of anatomy and physiology of the lymphatic system, this review discusses the current treatment approaches to lymphedema, with particular attention to patients who have undergone treatment for breast cancer.

Risk Reduction

The surgeon, caregiver, and at-risk patient can take several steps to minimize the risk of developing lymphedema. First and foremost, the patient should be informed and educated as early as possible. Since it is the surgeons who likely have the earliest contact with the patient, it falls on them to alert the patient to the risk of lymphedema complications, and to provide precautionary dos and don’ts, either directly or by pointing patients to reliable sources.

Risk reduction strategies and practices are broadly aimed at (1) minimizing injury and chances of infection of at-risk limbs; (2) reducing activities and behaviors that are linked to triggering or exacerbating lymphedema; (3) promoting informed self-monitoring of changes that are suggestive of early-onset lymphedema; and (4) facilitating immediate medical evaluation if lymphedema is suspected. Symptoms such as limb heaviness, weakness or aching, skin tightness or firmness, pain or numbness, or impaired mobility or range of motion may indicate a latent or pre-clinical stage of lymphedema. Since latency may extend for months or even years before it progresses to significant swelling, continued patient vigilance is important. Standardized questionnaires codifying these and other symptoms are available in the literature. Because increased weight (BMI) is associated with a greater risk of developing lymphedema, weight management may also be indicated. Good educational information may be found in the literature and at various web sites, including risk reduction guidelines from the National Lymphedema Network (NLN) and the American Cancer Society.
A recent study reported that breast cancer patients who were educated about the risks for developing lymphedema developed fewer symptoms and practiced more risk reduction behaviors (such as avoiding blood pressure readings, blood draws, and injections on the affected limb), than those who did not receive educational information. Although not always the case, it is highly recommended that the standard of care include an initial patient consultation with a professional skilled in lymphedema therapy and education, either prior to surgery or soon after, in order to improve patient outcomes.

Early Detection

It is well known that lymphedema, left untreated, will progressively become worse. The earlier lymphedema is detected and properly treated, the better will be the outcome. Early detection and treatment can lead to near normalization of a swollen limb or an edematous trunk, and a greater chance of minimizing or avoiding significant complications. Complications include fibrosis, infection, pain, range-of-motion limitations, negative body image, and an array of activity-limiting sequelae. An important component of early detection is the patient’s own recognition of symptoms and to seek therapy promptly. Too often, symptoms are either unrecognized or disregarded, resulting in initial therapy being sought at a more advanced stage when the ability to “arrest and reduce” the developing lymphedema is limited. Although patient education and informed awareness can improve this scenario, even under favorable conditions, self-referral may be unnecessarily delayed.

Thus, there have been efforts to help practitioners identify latent or incipient lymphedema using more quantitative methods, including the detection of early tissue changes by electrical impedance, changes in limb volume via circumferential measurement, or changes in tissue dielectric constant values measured anywhere on the body surface. Although promising, these approaches are limited by the absence of accepted standard criteria for preclinical lymphedema. Additionally, the patient must be evaluated at the lymphedema clinic, preferably prior to surgery, and at follow-up intervals after surgery if preclinical lymphedema is to be readily detected. Again, this emphasizes the importance of early presurgical consultation and evaluation that is beginning to be recognized and implemented as standard of care.

Phase I: In-Clinic Decongestive Therapy

Once lymphedema is detected, patients are usually referred to a lymphedema clinic staffed by certified, trained lymphedema therapists. This intensive, short-term therapy is known as Complete Decongestive Physiotherapy (CDP) or Complex Decongestive Therapy, CDT. The goals of CDP are to: (1) arrest disease progression; (2) reduce lymphedema volume; (3) prevent infections; (4) restore mobility and range of motion; and (5) train patients for life-long, self-management of the condition at home.

The standard of care of Phase I CDP therapy includes manual lymphatic drainage (MLD), short-stretch compression bandaging (SSCB), decongestive exercises, skin care, and sometimes the use of intermittent pneumatic compression (IPC). The details of the various CDP components and their utility to achieve initial reductions in lymphedema are well documented by earlier literature and more recent reviews. The MLD component is used to move lymph fluid out of affected lymphedematous regions to more centrally located healthy regions, thereby reducing edema volume. Exercise constructively influences lymph flow through body movement and transient muscle contraction, particularly when combined with SSCB. The use of SSCB also serves to sustain the reduced volume until the next MLD treatment, in part by decreasing the transcapillary ultrafiltration rate. The short-stretch nature of the specific bandages used for lymphedema treatment is significant. Short-stretch bandage material has few elastic fibers and will stretch less than a material with many elastic fibers (long-stretch). During transient muscle contraction, the reduced expansion of the bandage creates resistance resulting in high sub-bandage dynamic or “working pressure.” This is illustrated in Figure 1 when a patient squeezes a rubber ball. Such dynamic pressures are transmitted inward, making fibrotic tissue softer while simultaneously facilitating fluid movement by its effects on interstitial space contents and lymphatic vessel activation. With short-stretch bandages, large functional dynamic pressures can be achieved with low resting pressures, thereby helping to prevent circulatory compromise, especially important in patients with vascular deficiencies.

Because MLD is central to the Phase I therapeutic processes, and self-MLD and/or IPC is used in Phase II, the following discussion will consider the physiological basis for successful application of these therapies in upper extremity lymphedema.

The lymphatic system consists of an enormous number of lymphatic vessels, punctuated with lymph glands (nodes). Movement of lymph fluid through these vessels is directed toward specific entry points into the venous system. For drainage of the upper extremity under normal healthy conditions (Fig. 2A), axillary lymph nodes (LN) receive most upper extremity lymph fluid via superficial and deep lymphatic channels, and the ipsilateral trunk quadrant via multiple pathways. Lymph also drains from the arm to and through the supra- and infraclavicular nodes, bypassing the axillary nodes. It is collected by lymphatic vessels that join major trunks to empty into the venous system via junctions within the venous angle, as schematized in Figure 2.

If nodes are removed, or lymphatic channels damaged by radiation (e.g., as part of breast cancer treatment), then the ability of upper extremity lymphatic channels and all truncal tributaries to move lymph out of their respective territories is reduced (Fig. 2B). This reduction depends on the number of lymph nodes impacted, their prior share of the overall drainage load, and other effects associated with surgery and radiation. Subsequent effects depend on the extent of lymphatic channels with direct connections to the venous system, the presence of anastomotic lymph collectors connecting the affected quadrant to the contralateral axillary and supraclavicular nodes, and on differences of upper extremity lymph-flow features that affect subsequent arm lymphedema patterns. If the reduction in lymph drainage capacity is large enough, lymphedema develops.

Successful MLD requires brief application of mild, variable, directional pressures followed by an immediate release or resting phase. The effectiveness of both MLD and IPC depends on stimulating lymphatic flow in a way that facilitates lymph movement through alternative pathways, so that lymph is moved from lymphedematous regions circumventing damaged normal pathways. Key to effective therapy is the...
recognition of the underlying physiological processes, anatomical features, and physical principles governing the lymph movement. Collectively, these underscore the importance of treating the truncal lymphatics for effective lymphedema therapy.

Lymph movement within and then out of lymphatic territories is determined by: (1) normal contractile activity of lymphatic vessels; (2) the associated dynamic pressures that are developed; and (3) the resistance to lymph flow attributable to the overall network of lymphatic vessels and nodes.
Dynamic pressures, in turn, depend on distension of smooth muscle within lymph vessel walls as influenced by their lymph volume or load. An optimal therapeutic approach facilitates lymph flow into and out of the cleared areas, ultimately emptying into the venous circulation.

Figure 3A shows three physical factors that affect the process of normal lymph transport from a limb to and through lymph nodes, lymph collectors, and conduits terminating at junctions with the venous system: (1) dynamic propulsive pressures developed by contracting lymphatic vessels located within the limb (PL); (2) resistance to lymph flow between limb and venous junction (RL); and (3) pressures within the network of lymphatic vessels linking nodes to the venous system (PLV). These factors determine lymph flow (QL) from the limb as described by the following functional relationship:

\[ Q_L \sim \frac{(P_L - P_{LV})}{R_L} \]

Thus, lymph flow is proportional to the pressure gradient between intralymphatic vessels and lymph–venous conduits, divided by the lymphatic network resistance that lies between them.

With lymphedema, lymph movement through normal channels and out of lymphedematous regions is reduced due to increased RL (node removal and injury to lymphatic vessels and tissues) and reduced PL within lymph vessels in the lymphedematous regions. Promotion of lymph flow out of lymphedematous areas, partly by bypassing higher resistance pathways (as is the strategy with MLD and some advanced IPC devices), now depends on the pressure gradient (PL – P_L) in which P_T conceptually represents the truncal tissue pressure (Fig. 3B). This underscores the importance and basis for preparatory truncal clearance in which truncal pressure and volumes are reduced prior to attempting to drain lymphedematous regions. Truncal clearance, therefore, facilitates lymph flow through remaining ‘normal’ pathways by reducing PLV and PT, thereby optimizing the gradient and facilitating treatment-related lymph movement through alternate pathways.

Based on the foregoing, standard-of-care therapy, whether manual or automated, should include a thorough truncal clearance phase and proceed in clearing proximal to distal as demonstrated schematically in Figure 4. This approach is needed to minimize volume and pressure within lymphatic territories that will subsequently collect lymph fluid drained from lymphedematous tissue regions. Thus, the prepared normal truncal region serves as an optimized pathway for the adjacent affected quadrant, and the prepared affected truncal region serves as an optimized pathway for draining the lymphedematous arm. A similar proximal clearance requirement and analysis applies to lower extremity lymphedema.

This discussion of the physiological basis for Phase I care elaborates upon clinical concepts that have been described in textbooks and used in clinical practice for many years. Phase I care concludes when the therapist determines that the patient has attained the maximum benefit to be achieved by the intensive treatment. (i.e., a plateau has been reached) The optimal outcome is a near normalization of the affected limb in terms of limb volume, tissue health, pain reduction, and improved range of motion and mobility. During the final stages of Phase I, the therapist will focus on educating and training the patient in the techniques necessary to achieve effective long term management in the home.

**Phase II: Self-Management at Home**

In Phase II, the patient assumes responsibility for maintaining or improving the clinical results achieved in Phase I, and managing the condition long-term at home by reducing the risks of exacerbations and associated complications. Self management in the home includes the following: (1) self-MLD;
Poor patient compliance in performing home self-care is a major factor contributing to the loss of gains so diligently achieved during Phase I. Such losses (and in fact reversals) may trigger complications, including fibrosis, inflammation, cellulitis, diminished range of motion, and pain. Lack of compliance, especially with respect to self-MLD and bandaging, may be attributable to a number of factors. The affected areas may be difficult to reach because of impaired mobility, or because of their location (e.g., the backside). The location of the affected area also complicates application of compression bandaging, which can be more difficult to apply to the trunk than to the extremities. Further, self-care is emotionally challenging because of the daily time required. Self-MLD is physically demanding and requires the patient’s complete attention. The affected areas may also be difficult to reach because of impaired mobility, or because of their location (e.g., the backside). The location of the affected area also complicates application of compression bandaging, which can be more difficult to apply to the trunk than to the extremities. Further, self-care is emotionally challenging because of the daily time required. Self-MLD is physically demanding and requires the patient’s complete attention. These demands are becoming more difficult to meet in light of the longevity of cancer survivors and their declining physical prowess. To be effective, self-MLD must also be done consistently and correctly, which for many patients is a significant challenge. This self-care regimen may also lead to repetitive stress injuries of the already compromised limb or unaffected limb.

Together these factors can impede compliance, particularly as the patients may be coping with cancer and its aftermath. As a result, many patients who initially make diligent efforts to adhere to the in-home treatment regimen experience exacerbations of the lymphedema or related infections, and require further rounds of in-clinic treatment, including CDP. While the therapist’s specialized attention is often psychologically supportive to the patient, the costs associated with cycling through multiple sessions of professionally administered CDP continue to mount. Some third-party payers do not cover repeated CDP sessions, creating a financial burden for the patient. The use of advanced, programmable IPC devices provides an effective treatment approach that can ease the physical and emotional burdens of self-care.

**FIG. 4.** Schematized depiction of effective truncal-limb treatment. Normal flow pathways are reduced or absent. Truncal clearance reduces $P_T$ and $F_{LV}$ in ipsilateral and contralateral quadrants and augments normal lymphatic vessel pumping actions prior to attempting limb drainage therapy. Treatment-related lymph flow is thus optimized. (See text for further discussion.) The numbers show the sequential order in which applied therapeutic decongestion or clearance is most appropriate and effective. The main emphasis is on preparing the truncal regions prior to treating the affected limb. The physiological basis for this approach is discussed in detail within the text.

**Truncal clearance**

As noted, the capacity to provide automated initial truncal or proximal clearance is essential. Without it, therapy focused solely on the affected limb is inefficient and less effective, and also may result in injury. In a recent survey of 50 certified lymphedema therapists conducted by the author in February 2009 (unpublished, data available on request), their greatest concerns about using or recommending IPC was that it would cause truncal or genital edema or produce fibrotic cuffs. Each of these is a concern with simple ‘older generation’ pumps that do not provide truncal clearance.

**Magnitude and duration of applied pressure**

The literature describes the possibility of lymphatic vessel injury when sustained high pressures are applied to the skin. High sustained pressure (squeeze and hold approach) with
IPC use was also a prime concern of the surveyed therapists. The amount and duration of pressure is not only a safety consideration but is also important to enhance patient tolerance and to address the pain associated with lymphedema. Significant to any discussion about applied pressure is an understanding of the impact that both duration and surface area play. Stated succinctly, applying pressure for a longer duration to a larger surface area will result in a greater pressure time gradient than applying that same pressure to a smaller surface area for a brief period of time (Fig. 5). Older generation, simple IPC systems, inflate sequentially and hold inflation for a period of time, utilizing a squeeze and hold approach (long duration, large surface area) that may result in higher pressures being applied than therapeutically necessary or appropriate.28 Further, with some older-generation simple IPC pumps, device settings do not accurately reflect applied pressures.31 Therefore, the clinicians’ ability to prescribe appropriate therapy is impeded not only by limited device functionality (nonprogrammable) but also the possible inaccuracies in calibration. Some newer advanced IPC systems employ a short duration pressure profile (work and release) which delivers lower applied pressures.28

In summary, significant barriers exist to successful long-term home management for the lymphedema patient. Home management programs are demanding and technique dependent. Patient adherence to such regimens are challenging. One alternative to support patients’ in home efforts may be the use of advanced programmable IPC devices.

**Risks and Complications of Ineffective Therapy**

Lymphedema is a chronic condition that, without effective treatment, progressively grows worse.7 Its impact, whether in upper or lower extremities, is multidimensional and may include chronic pain, severe mobility limitations, impairment in function, predisposition to serious inflammatory episodes and infections (cellulitis, erysipelas), and compromised immune function.28 Psychological and quality of life impacts are also significant and include loss of self-esteem, depression, impaired body image, and social withdrawal.29

Progressive lymphedema induces physiological changes that can inhibit normal bodily functions. Interstitial accumulation of protein, debris, and cellular fragments trigger a variety of negative processes. Proteins act to pull more fluid into the interstitium, thereby further increasing tissue edema. As time passes, accumulated proteins, together with connective tissue changes, cause tissue fibrosis. Macrophages that normally clean up interstitial debris are inhibited by fibrosis, and the interstitium suffers other progressive derangements. In addition, accumulated protein and other substances act as stimuli for chronic inflammation accompanied by increased capillary blood pressure and flow due to vasodilation of small blood vessels supplying the area. This increased pressure causes more fluid accumulation and tissue warming that, combined with the bacterial growth stimuli of protein, creates an infection-ripe environment, setting the stage for a vicious cycle of worsening symptoms and progressive complications. These considerations emphasize the importance of continuity in treatment from acute care to home management as being critical to avoid worsening of the swelling and associated complications.

In addition to the well-recognized complications of progressive lymphedema, failure to treat the truncal lymphatics and the affected extremity is associated with significant risks to the patient. These include development of a fibrous band at the proximal limb that further impedes lymphatic drainage out of the limb, and development of new areas of edema in the truncal quadrant adjacent to the affected limb that include

![FIG. 5. Comparison of pressure-time integrals associated with different IPC devices. Data are mean and SD of measurements on 10 subjects. At all forearm sites, the Lympha Press® device (Tel-Aviv, Israel) resulted in pressure-time integrals that were significantly greater (**p < 0.001) than for the Flexitouch® device (Minneapolis, Minneosta). (Data from Ref. 28).](image-url)
swelling of the chest, axilla, back, abdomen, and genitalia. These conditions present new treatment challenges, as well as further debilitation for the patient.

An additional aspect of truncal treatment should be mentioned. Lymphedema is seldom confined to just the limb that is obviously swollen. In many cases, the lymphedema extends into the adjacent body quadrants. Truncal edema presents its own set of complications. Lack of effective treatment of the truncal region may result in infection, pain in the back, chest, shoulder, or breast, and worsening neuropathic pain of the fingers. Pain may be localized or may radiate across the chest, trunk, or back. In addition, significant body changes may lead to emotional distress and altered body image. Patients may also report discomfort with movement and difficulty performing activities of daily living. These additional facts further emphasize the vital need for the treatment of adjacent body regions (truncal lymphatics), in addition to the obviously affected extremity.

Lymphedema complications, whether in the limb or trunk, require careful monitoring to minimize symptom progression and their growing impact on a patient’s ability to participate successfully in daily activities. Effective life-long treatment of the affected limb and trunk is necessary to reduce the risk of these complications.

Summary

The standard of care for lymphedema embraces a broad spectrum of considerations that include: (1) risk reduction; (2) early, potentially preclinical detection; (3) early and intensive initiation of therapy (Phase I); and (4) effective and accepted in-home self-care (Phase II). Risk reduction can be enhanced by early education of patients, including when possible, presurgical and follow-up evaluations. Accepted phase I therapy includes MLD, SSCB, decongestive exercise, and skin care, in some cases supplemented with IPC use. Phase II, life-long in-home self-management, includes self-MLD and/or IPC, use of compression garments SSCB or an alternative compression system, exercise, and skin care. Significant phase II compliance issues that may negate phase I gains can be mitigated by the adjunctive use of physiologically appropriate IPC devices. IPC selection for lymphedema treatment requires careful consideration of the mode of action and device functionality. Simple nonprogrammable devices are no longer viewed as clinically appropriate for lymphedema care because they do not address the specific physiological needs of lymphedema patients, resulting in increased risk associated with treatment pressure, pattern, and the lack of truncal clearance. Such risks cannot be overcome because of the simple nature of these nonprogrammable devices. Advanced programmable devices that provide truncal clearance, lower applied pressure, and optimum adjustability are physiologically sound and represent the standard of care approach. Risks of ineffective treatment are significant, and clinicians are challenged to develop care plans that can be effectively implemented by the patient to optimize long-term outcomes.

Disclosure

Dr. Mayrovitz is a scientific advisor to Tactile Systems Inc., the manufacturer of the Flexitouch® system.

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