Chapter 35
Upper Limb Lymphedema

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Introduction

Within the realm of lymphatic disease treatment, there are many therapeutic interventions available, as highlighted in previous chapters. Treatment of lymphedema (LE) is very challenging. Therapeutic options in LE include conservative and operative modalities and should be individualized with regard to the circumstances of the patient and the lymphedema by a multidisciplinary approach. These circumstances include age, comorbidities, prognosis of (malignant) disease, psychosocial aspects, and physical potential. The goals for conservative treatment are to eliminate edema by reducing interstitial fluid accumulation and to stimulate lymphatic propulsion by compression.

Traditionally, many modalities are performed in combination. The contribution of each individual treatment modality to the outcome is, therefore, still under discussion. In this chapter we will focus on the timing of treatment, the combination of various modalities of treatment, and the phases of intervention.

Many terms are used to describe lymphatic treatments: complex decongestive therapy/treatment, complex physical therapy, or complex decongestive physiotherapy. These terms are confusing because it cannot be seen that the lymphatics are involved, "physiotherapy" is a terms used too generally, and the word "complex" is unclear.

Therefore, in 1998, the term *decongestive lymphatic therapy (DLT*) was advocated to achieve uniformity of nomenclature and foster communication among the health care professionals who administer therapy for lymphedema. DLT comprises a number of interrelated treatment modalities that are most efficacious when utilized in an interdependent fashion, as mentioned in Chap. 9.

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Table 35.1 Useful lymphedema interventions

Therapeutic option	Initial treatment phase	Maintenance phase
Manual lymph drainage	X	
Bandaging	X	
Garments/hosiery		X
Pneumatic compression	X	X
Physiotherapy	X	
Decongestive lymphatic therapy	X	
Exercise	X	X
Weight control	X	X
Skin care	X	X
Awareness	X	X
Self-management		X
Reconstructive surgery	X	
Reductive surgery	X	

Treatment of lymphedema consists of two phases: the initial treatment phase and the maintenance phase. The first phase gradually merges into the maintenance phase. The goal of treatment is to reduce lymphedema during the treatment phase and make the patient independent from the professional health care worker. This provides the patient with as much knowledge as possible and with self-management skills to maintain the result with a good quality of life. The patient plays an active role in the maintenance of the therapeutic result. The role of the therapist during the second phase is more hands-off, monitoring and guiding the patient. The various therapeutic options are listed in Table 35.1.

Physical treatment of lymphedema should not be considered as a single therapeutic modality, but as a continuum that begins with informing and educating the patient, advocating awareness and self-management, objective early diagnostics by volumetry, and, at the end of the spectrum, individual specialized lymphedema treatments. A multidisciplinary approach, as suggested in many guidelines,[1-3] is mandatory to the success of the treatment of upper limb lymphedema.

Lymphedema of the Arm

Lymphedema of the arm is, in most cases, due to treatment of breast cancer. Many factors influence the development of breast cancer–related lymphedema (BCRL), including obesity,[4] hypertension,[5] infection, type of cancer treatment,[6] and individual impaired lymphatic drainage.[7]

Lymphedema frequently develops slowly, often with pre-clinical symptoms and signs, such as heaviness, transient swelling, and slight volume changes compared with preoperative values. Early detection is essential for a treatment program during the initial stages of lymphedema.

The practical issues in the approach to lymphedema in general, and to physical therapy in particular, are centered upon the organization and availability of care for the patient. In cancer-related lymphedema, and especially in BCRL, a protocolized approach is useful because lymphatic awareness can be integrated into the cancer protocol. This gives the opportunity to start primary and secondary prevention programs on lymphedema from the outset. Much work has to be done to achieve this ambition.

Considerations in Manual Lymph Drainage

Only a few studies have been performed to study the additional effects of manual lymph drainage (MLD) over compression therapy in LE. Two controlled studies showed that compression therapy with or without additional MLD was equally effective for BCRL. Andersen et al.[8] performed a randomized controlled study in BCRL comparing MLD and compression ($n=20$) with a control group that was treated with only compression therapy ($n=20$). After 2 weeks, the control group actually had a greater percentage reduction in absolute edema (60%) compared with the MLD group (48%). Both groups experienced an equal reduction in the symptoms of heaviness and tightness, but the control group also had a reduction in reported discomfort. The reduction in absolute edema (66%) was maintained for 12 months' follow up (pooled data). Johansson et al.[9] studied the effect of short-stretch bandages with or without MLD in 38 female patients. Both groups showed significant improvement in volume reduction (−11% after 3 weeks) and fewer complaints.

A comparison of studies on MLD and compression therapy alone by Korpon et al.[10] found no difference in volume change.

In a systematic review, Kligman et al.[11] studied 10 randomized controlled trials of treatment for BCRL. In all of these studies, the authors could not go farther than stating that there was "some suggestion" that compression and MLD "may improve" LE. The effectiveness of the use of life-long compression garments was more obvious.

In daily practice, MLD is used in several therapeutic schemes, especially when it is combined with various forms of compression therapy, such as short-stretch multilayer bandaging applied after each MLD session.[12] Although MLD has been used widely for many decades and is assumed by many to be a panacea for the treatment of LE, there is currently no indisputable published evidence for its effectiveness or its mode of action in improving lymphatic drainage.

Controlled, comparative studies are currently not available for the effectiveness of each separate modality in the treatment of LE.

Moseley et al.[13] conducted an extensive review of the literature in 2006 for common non-operative treatment modalities for LE and concluded that despite the identified benefits, there was still a need for large-scale, clinical trials in this area. A combination of MLD with compression therapy improved the results. In most studies reviewed by Moseley et al. there was a mix of lymphedema types, mainly

BCRL, and specific outcome parameters were often not defined. Specific studies on primary lymphedema are not available.

In 2007, Hamner and Fleming[14] retrospectively studied 135 patients with BCRL who were receiving DLT. After 8 weeks, the volume reduction was about 18%. A surprisingly positive effect on pain was found: 76 patients experienced pain before treatment, and 56 were free of pain after treatment (76% reduction). It was concluded that LE continues to be a problem for patients with breast cancer. A program of lymphedema therapy can reduce the volume of edema and, in particular, reduce pain in this population. Badger et al.[15] compared the effects of treatment for 18 days with short stretch bandaging, followed by compression hosiery with those of compression hosiery alone for leg and arm lymphedema. They showed that initial compression therapy with subsequent use of hosiery was twice as effective as hosiery alone.

Measurement of the undergarment pressure was performed in some studies.[16,17] A major limitation of these studies is the discrepancy between the undergarment pressure claimed by the manufacturer and the actual interface pressure due to the large variety of types of garments and inter-individual variation in measuring garments. Vignes et al.[18] studied 682 patients treated for BCRL for four years in the maintenance phase. Treatment failure was associated with younger age and higher weight and body mass index. Treatment with diurnal garments and nocturnal bandaging decreased the risk of treatment failure significantly (hazard ratio, 0.53 [0.34–0.82], $p = 0.004$), whereas the addition of MLD did not.

General Considerations for Compression

The pressure delivered by compression is different in the legs than in the arms. It is important to note that the hydrostatic pressure that must be overcome by external compression is much higher in the legs than in the arms. In a standing position, the venous pressure in the distal leg is equal to the weight of the blood column between the heart and the measuring point, which is about 80–100 mmHg. The high intravenous pressure in the upright body position always increases the lymphatic load by promoting increased fluid extravasation. High external pressure is necessary in order to counteract this extravasation. The venous pressure in the arm is much lower than that in the leg because of the lower weight of the blood column between the heart and the hand. Thus, less external compression will be needed to reduce extravasation into the tissue and to promote reabsorption of tissue fluid. The arm volume reduction from bandaging is probably due not only to a pressure-dependent shift in Starling's equilibrium, but also to stimulation of lymphatic drainage. Besides veno-dynamic issues, lympho-dynamic issues should also be considered. In healthy arms, the distance from the arm to the thoracic duct is short, and the intra-lymphatic pressure varies with the intra-thoracic pressure. Lymphatic drainage is stimulated with relatively low or even negative intra-lymphatic pressure. In BCRL, lymphatic drainage is deficient because of damage to the major lymph collectors and lymph nodes by surgery and/or radiation, leading to lymphatic congestion.[19]

Compression Therapy in the Arms

Although inelastic, multi-layer, multi-component compression bandages allow immediate reduction of volume in lymphedematous arms and is a mandatory part of treatment, studies to measure the interface pressure in arm LE has rarely been performed before. The deciding parameter of the interface pressure, which is the dosage of compression therapy, has been measured only in patients with chronic venous insufficiency[20] and there is a positive relation between pressure and volume reduction. In arm lymphedema, for example, the compression pressure required to obtain the highest volume reduction per unit of time is unknown.

Damstra and Partsch[21] showed that low sub-bandage pressures between 20 and 30 mmHg are effective and better tolerated than high-pressure bandages by the patient with arm lymphedema. In future, more research will be required to understand the therapeutic effect of types of compression therapy and materials in arm lymphedema.

Recently, published studies have shown the importance of compression therapy after circumferential suction-assisted lipectomy (the Brorson method)[22,23] in order to achieve a 100% volume reduction in end-stage arm lymphedema. The method consists of an operative intervention to remove the complete suprafascial component of the lymphedematous arm, which consists mainly of fat.[24] Postoperatively, compression therapy is provided by short stretch bandaging and garments, which should be worn lifelong, the same as in the conservative treatment of arm lymphedema. All garments are custom-fitted and flat knitted. Long-term results are highly favorable, with sustained complete volume reduction of the pre-operative volume excess, for up to 13 years of follow-up. In this procedure, manual lymph drainage is not necessary to maintain the result.

In lymphedema, intermittent pneumatic compression has been used for decades. Megens and Harris[25] reviewed the literature on physical therapy treatment of BCRL. Most studies were inappropriately designed and often lacked proper comparisons. They concluded that compression therapy should be performed with multi-chamber devices in combination with other therapeutic options, such as MLD and compression. Monotherapy with intermittent pneumatic compression was discouraged.

Bandaging and hosiery can provide compression. In general, hosiery is measured when the maintenance phase is reached. In this phase there is no further volume reduction despite proper LE treatment. The terms hosiery, garments, and sleeves are often used interchangeably and include gloves, gauntlets, Bermudas, and compression devices for toes. For LE, garments should always be custom-fitted and flat-knitted with a high static stiffness and should be measured routinely during long-term follow-up.[26]

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