
The Lymphedema Center and Multidisciplinary Management

6

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Key Points

- Lymphedema is a complex problem, and few physicians focus on the disease.
- Patients with lymphedema often feel isolated because they have difficulty finding a provider who understands their condition and can treat them.
- Plastic surgeons and compression experts are necessary components of a Lymphedema Center.
- More than one specialist is often needed to treat a patient with lymphedema.
- Individuals with lymphedema are best managed in a multidisciplinary Lymphedema Center.

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Introduction

Lymphedema is a complicated medical condition that is poorly understood. Few physicians care for patients with lymphedema and the condition is associated with many myths. Patients often are misdiagnosed, undergo unnecessary tests, and are managed incorrectly. For example, 25 % of individuals referred to our Lymphedema Center with “lymphedema” do not have the disease [1, 2]. One-third of patients sent to our Lymphedema Center previously had undergone tests that were non-diagnostic for lymphedema; 8 % had been treated erroneously with a diuretic [2].

“Lymphedema” often is used as a generic term to describe any condition that causes extremity overgrowth; incorrect diagnosis can lead to incorrect management [1, 3]. We believe patients with lymphedema are best managed in an interdisciplinary Lymphedema Center. Because the disease does not fit easily into one medical specialty, individuals usually require multiple providers. In this chapter we present our patient care model and current treatment algorithm.

Lymphedema Center

Patients with lymphedema typically are medical nomads, being referred from provider to provider until they find someone who can manage their

condition. One-third of patients treated in our Lymphedema Center reside outside of our referral area [2]. Our clinic meets monthly and is directed by plastic surgeons. New patient visits are scheduled for 1 h and they are initially seen by the plastic surgeon. Next, they undergo standardized photography as well as limb measurements by a physician's assistant or nurse. Individuals then are seen by a compression garment specialist and fitted for garments. In Boston patients also are measured for a pneumatic compression device. Individuals who are being considered for operative intervention undergo water displacement to most accurately determine the volume of their diseased extremity (Fig. 6.1).

All patients undergo lymphoscintigraphy to determine their lymphatic function. Patients who reside outside of our region have their lymphoscintigram performed at our institution the day before their scheduled appointment. We have a standard lymphoscintigraphy protocol (images at 45 min, 2 h, 4 h). The use of a consistent protocol enables us to compare patients as well as follow the same individual longitudinally. We have found that when patients undergo lymphoscintigraphy at

outside institutions the test often is not performed properly or is difficult to interpret (e.g., only a 2 h image is taken). We prefer to obtain a lymphoscintigram for most patients because it definitively diagnoses the condition. In addition, the test gives a subjective measure of the severity of lymphedema in patients who likely have the condition. For example, an individual who has transit to the inguinal nodes by 2 h and no dermal backflow would be considered to have "mild" lymphatic dysfunction, whereas a patient with no tracer in the inguinal nodes by 4 h and dermal backflow would be labeled as "severe" dysfunction. Patients with worse lymphoscintigraphic findings are counseled that their prognosis may be less favorable and that they should be vigilant with conservative interventions.

All individuals evaluated in our Lymphedema Center are entered into a database to facilitate research. Using this strategy, we have been able to publish demographic data on patients referred to a Lymphedema Center as well as characterize the disease *obesity-induced lymphedema* [1–6]. Collecting patients in regional referral centers allows retrospective and prospective clinical



Fig. 6.1 Measurement of limb volumes using water displacement

research studies to be performed. In addition, insights into the condition are more easily formed when patients are concentrated among specific providers. Our database also has allowed us to help patients contact other individuals with lymphedema who reside in their area. Often individuals obtain psychological benefit from speaking or meeting with others who share their disease (particularly in the pediatric population).

Interdisciplinary Management

Although many types of specialists care for patients with lymphedema, plastic surgeons have been particularly involved in patient management because they are most capable of providing surgical intervention. Currently performed physiologic procedures (i.e., lymphatic–venous anastomosis, lymph vessel transplantation, lymph node transfer) require the microsurgical expertise of plastic surgeons. Excisional procedures that are carried out (i.e., liposuction, staged skin/subcutaneous excision, Charles procedure) also require plastic surgical training.

A Lymphedema Center should be staffed by a plastic surgeon capable of providing operative intervention if necessary (Table 6.1). Compression experts must participate as well (e.g., rehabilitation medicine physician, physical therapist, occupational therapist, physician assistant, and/or nurse). Our clinic in Boston is staffed by a compression garment specialist who measures, fits,

and troubleshoots garments during the patient's appointment. In addition, we have a pneumatic compression expert who teaches pneumatic compression to patients during their visit.

Our multidisciplinary clinic in Boston also involves the following specialties who do not attend the clinic: nuclear medicine, radiology, bariatric surgery, and infectious disease. We have a close collaboration with our nuclear medicine department with whom we have developed our lymphoscintigraphy protocol. An interventional radiologist with expertise in the field of vascular anomalies is available to review imaging (e.g., MRI, CT, ultrasound) to diagnose patients who have normal lymphoscintigraphy. Patients with obesity-induced lymphedema, or at risk for the disease, are referred to our bariatric surgical collaborator. Individuals with >3 infections a year, or who are at risk for having filariasis, are referred to our infectious disease colleagues.

Current Treatment Algorithm

Almost all patients referred to our Lymphedema Center undergo lymphoscintigraphy to determine whether they have the disease and, if so, the severity of the condition (Fig. 6.2). Patients are prescribed compression garments as well as pneumatic compression. Management of patients diagnosed with lymphedema begins with patient education. Individuals are counseled about activities of daily living that may help or worsen their condition (e.g., exercise the affected extremity and avoid trauma). Often we answer questions regarding myths about lymphedema that can significantly improve the patient's quality of life (e.g., the individual can eat any type of food, take a warm bath, exercise (Nordic pole walking or running)).

After counseling a patient with lymphedema about his/her condition, we initiate conservative compression strategies. We advocate that custom-fitted compression garments should be worn continuously. We also recommend pneumatic compression for 2 h each day if possible. The authors do not routinely use lymphatic

Table 6.1 Multidisciplinary lymphedema center specialists

Plastic surgeon
Compression garment expert (rehabilitation medicine, physical therapy, physician assistant, nurse)
Pneumatic compression expert
Nuclear medicine physician
Radiologist
Bariatric surgeon
Infectious disease physician
Specialists to rule out other causes of extremity edema (orthopedic surgeon, vascular surgeon, internist, pediatrician, rheumatologist)

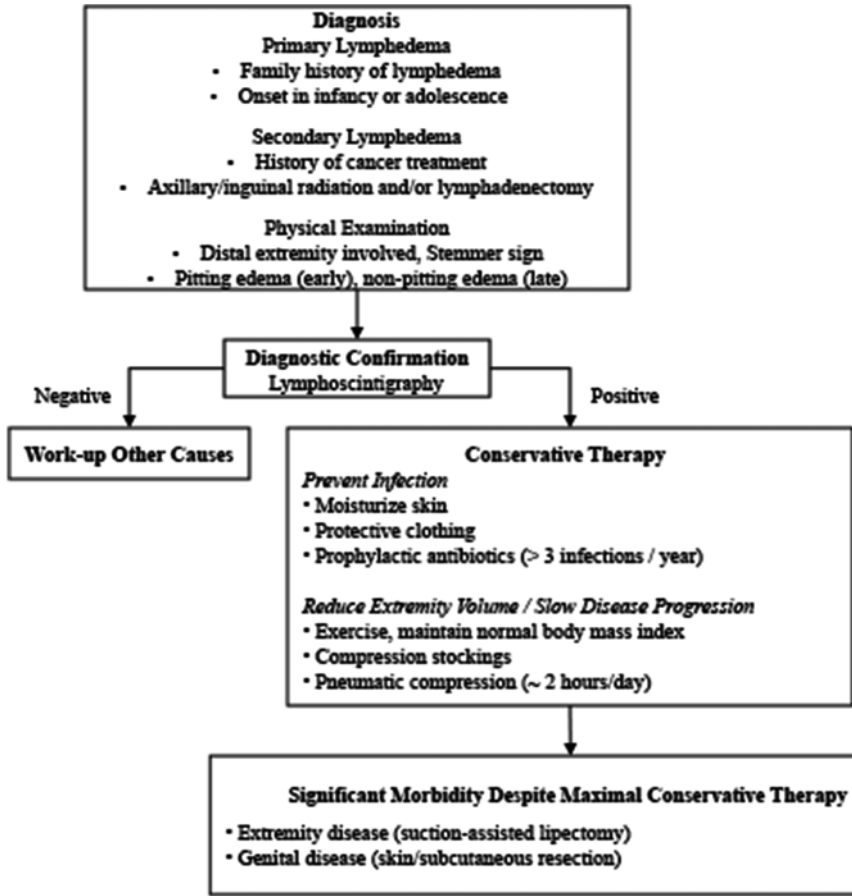


Fig. 6.2 Current lymphedema management algorithm used by the authors

massage or combination compressive regimens (e.g., complex decongestive therapy), although these modalities can be effective. Compared to complex decongestive regimens, compression garments and pneumatic compression are easier for the patient and can be done at their convenience in their home.

Approximately 95 % of our patients are able to be managed conservatively without operative intervention. Indications for surgical treatment include failure of conservative measures and repeated infection, difficulty using the extremity, and/or significant psychosocial morbidity. The authors favor excisional procedures over physiologic operations because we believe the results are most predictable and patients with severe

disease are candidates. Our first-line operation for lymphedema is suction-assisted lipectomy (liposuction). Individuals with genital disease or very severe extremity lymphedema are managed by staged skin/subcutaneous excision.

Patients referred to our center that have a normal lymphoscintigram and are thought to have a condition other than lymphedema undergo further evaluation. Individuals with venous insufficiency are referred to a vascular surgeon. Typically, if we are unsure of the cause of the patient's swelling, we will obtain an MRI. MRI will illustrate whether or not the patient has a vascular malformation (e.g., Klippel-Trénaunay syndrome, venous malformation, lymphatic malformation) or an underlying orthopedic process (e.g., synovitis,

ligament sprain, occult fracture). Individuals with suspected systemic fluid overload (e.g., cardiac, renal, hepatic pathology) are referred to their primary care physician. Occasionally, patients with a possible rheumatological process are sent to a rheumatologist.

Conclusions

Lymphedema is a poorly understood condition and patients often are misdiagnosed, managed incorrectly, and feel isolated. Individuals with this disease should be treated by providers focused on their condition. Because a single specialist can rarely manage a patient with lymphedema, individuals are treated in a multidisciplinary center where they can be diagnosed, educated, and managed both conservatively and operatively. Concentrating patients in regional centers also improves their condition by facilitating research.

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