

# Lymphedema



## Diagnostic workup and management

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### Learning objectives

After completing this learning activity, participants should be able to describe the diagnostic criteria and appropriate workup required to make the diagnosis of lymphedema; describe the range of treatment modalities available for lymphedema; and identify patient-centered concerns, including pain and activities of daily living.

### Disclosures

#### Editors

The editors involved with this CME activity and all content validation/peer reviewers of the journal-based CME activity have reported no relevant financial relationships with commercial interest(s).

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Lymphedema is a localized form of tissue swelling resulting from excessive retention of lymphatic fluid in the interstitial compartment. It is caused by impaired lymphatic drainage. Lymphedema is a chronic progressive disease with serious physical and psychosocial implications. It can be challenging to diagnose, especially in obese patients and in those with coexisting venous disease. We performed PubMed and Google Scholar searches of the English-language literature (1966-2017) using the terms lymphedema, lymphedema management, and lymphatic complications. Relevant publications were manually reviewed for additional resources. There are currently no standard guidelines for the diagnosis of lymphedema. There is no cure yet for lymphedema, and the objective for management is to limit disease progression and prevent complications. (J Am Acad Dermatol 2017;77:995-1006.)

**Key words:** cellulitis; compression; lymphedema; management; skin care.

## DIAGNOSIS

### Key points

- **Lymphedema is a clinical diagnosis; other diagnostic tools can be used to rule out other causes of extremity swelling**
- **The Kaposi–Stemmer sign is pathognomonic of chronic lymphedema**
- **Lymphoscintigraphy is the criterion standard imaging tool to confirm diagnosis**
- **Unless surgical intervention is considered, imaging is not necessary**

### Abbreviations used:

APCD:	advanced pneumatic compression device
BCRL:	breast cancer-related lymphedema
CDT:	combined decongestive therapy
DEC:	diethylcarbamazine
MLD:	manual lymphatic drainage
PCD:	pneumatic compression device
STS:	Stewart–Treves syndrome

Lymphedema is primarily a clinical diagnosis, and thorough history-taking and physical examination

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are invaluable. Advanced lymphedema can be diagnosed clinically. However, in early stages of the disease, concomitant conditions, such as obesity, lipodystrophy, and venous insufficiency, can make the diagnosis challenging; therefore, additional workup may be necessary. There are many diagnostic tools to evaluate lymphatic function, and the decision to use any of them must be based on the patient's situation. Early diagnosis of lymphedema is important because it significantly increases the success of therapy.<sup>1</sup>

### History and physical examination

History should include age of onset, family history, course of the condition, history of penetrating trauma, infection, cancer, heart failure, hypothyroidism, hypoalbuminemia, sepsis, venous or lymphatic obstruction, inguinal/axillary radiation, lymphadenectomy, and travel to areas endemic for filariasis. Lower extremity lymphedema usually manifests as swelling in the dorsal surface of the foot with a characteristic blunt "squared-off" appearance of the digits in the involved extremity (Fig 1). Lymphedema in the legs often produces preferential swelling of the distal extremity and then progresses proximally. Pitted or dimpled texture of the skin (peau d'orange) and the Kaposi–Stemmer sign (an inability to pinch the fold of skin on the dorsal aspect of the base of the second toe) are characteristics of chronic lymphedema. In advanced stages, the skin over the affected area becomes hyperkeratotic and develops verrucous cobblestone-like papules, plaques, and nodules with underlying woody fibrosis.

### Clinical staging of lymphedema

Based on clinical findings in a physical examination, the International Society of Lymphology developed a staging system that divides lymphedema into 4 stages (Table I).

### Imaging studies

In most cases, imaging is not necessary for diagnosis, but it can be used for confirmation of diagnosis and to assess the extent of involvement in order to determine the most appropriate therapeutic intervention.

**Lymphoscintigraphy.** Lymphoscintigraphy is the criterion standard imaging tool to confirm the diagnosis of lymphedema. It is the most commonly used study for the evaluation of lymphatic function. By visualizing the lymphatic network, lymphoscintigraphy allows for detection of lymphatic abnormalities.<sup>2,3</sup> The reported sensitivity and specificity for detecting lymphedema is approximately 73% and 100%, respectively.<sup>4,5</sup> A



**Fig 1.** Characteristic squared off appearance of digits in a patient with lymphedema of the leg.

**Table I.** Clinical staging of lymphedema

Stage	Features
0 (Subclinical)	Swelling is not evident. Heaviness and discomfort
1 (Spontaneously reversible)	Swelling relived by limb elevation; includes pitting edema
2 (Spontaneously irreversible)	Swelling not improved by limb elevation; may include or not include pitting edema
3 (Lymphostatic elephantiasis)	Swelling not improved by elevation, skin hardening, nonpitting edema, verrucous changes, and recurrent soft tissue infections

water-soluble technetium-labeled large molecule (colloid or human serum albumin) is injected intradermally into the interdigital space of the affected limb. The lymphatic system takes up the radiolabeled colloid macromolecule, and the lymph movement is then monitored with a gamma camera. Major lymphatic trunks and lymph nodes can be visualized. Typical abnormalities visualized in lymphedema include delayed transport of tracer to the regional lymph nodes, absent or delayed visualization of lymph nodes, formation of collateral lymphatic vessels, and dermal backflow. Despite its clear advantages, lymphoscintigraphy is time-consuming and technically challenging. It involves exposure to radiation and is not available at every facility.

**Magnetic resonance imaging.** Magnetic resonance imaging (MRI) is neither sensitive nor specific for lymphedema. However, it can be useful for the evaluation of other causes of limb swelling, such as obstructive tissue or untreated cancer.<sup>6,7</sup> MRI can be used to assess the presence and severity of edema and differentiate fluid from fat deposition. The characteristic absence of edema within the muscular compartment helps distinguish lymphedema from other forms of edema. MRI is superior to computed tomography because it can detect water and the

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