

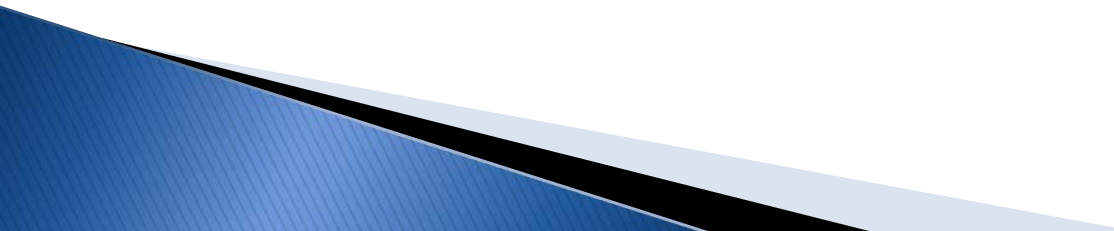
Lymphedema

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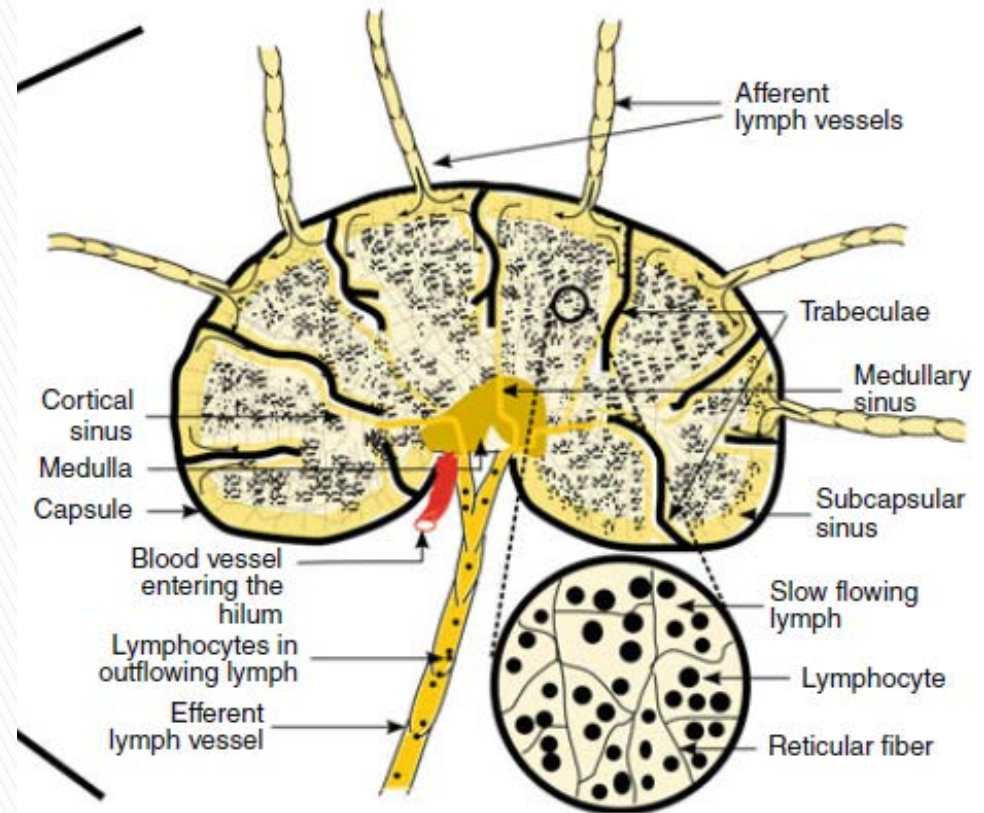
ACP Board Review, March 2014

Agenda

- ▶ Pathophysiology
 - ▶ Classification schemes
 - ▶ Symptoms and signs
 - ▶ Differential diagnosis
 - ▶ The CEAP class C3 CVD leg
 - ▶ Lipedema
 - ▶ Diagnostic testing
 - ▶ Management
 - ▶ Complications
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Pathophysiology

- ▶ Lymphatic system collects & drains interstitial fluid that escapes capillary circulation
- ▶ Drains into veins
- ▶ Obstruction
- ▶ Reflux
- ▶ Overproduction of lymph fluid

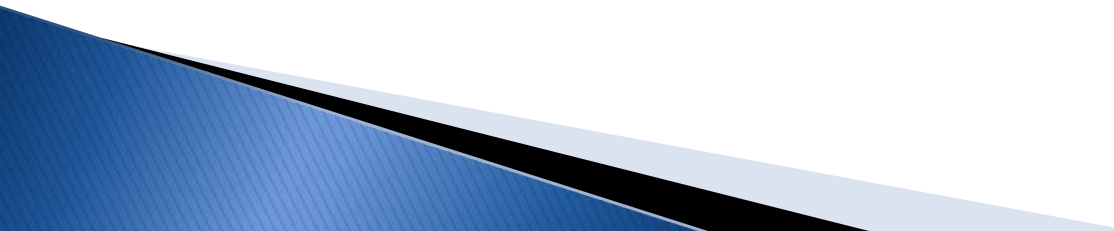


Laredo & Lee, Lymphedema, in Mowatt-Larssen et al. (eds), *Phlebology, Vein Surgery & Ultrasonography*, 2014.

Primary lymphedema

- ▶ 10% of cases
- ▶ Congenital, < 1 year – aplastic, bilateral
- ▶ Praecox, 1–35 years – hypoplastic, unilateral foot & calf, females 10:1
- ▶ Tarda, > 35 years

Secondary lymphedema

- ▶ 90% of cases
 - ▶ Obesity
 - ▶ CVI
 - ▶ Filariasis
 - ▶ Recurrent infection
 - ▶ Cancer, e.g. lymphatic, ovarian, prostate
 - ▶ Radiation therapy
 - ▶ Surgery or trauma
- 

Natural history

Table 23.1 Stages of lymphedema

Latency	Risk for lymphedema present. No clinical change evident
Stage I	Pitting, reduces overnight with simple measures (elevation). No fibrosis
Stage II	No longer pitting, no full reduction with elevation, evident fibrosis
Stage III	Nonreversible, hardened fibrosis and sclerosis of cutaneous and subcutaneous tissues

Laredo & Lee, Lymphedema, in Mowatt-Larssen et al. (eds), *Phlebology, Vein Surgery & Ultrasonography*, 2014.

DDx – unilateral swelling

- ▶ Deep vein thrombosis or obstruction
- ▶ Iliac vein obstruction
- ▶ Chronic venous insufficiency
- ▶ Lymphedema
- ▶ Baker's cyst
- ▶ Cellulitis
- ▶ Orthopedic injury

DDx – bilateral swelling

- ▶ Bilateral CVI
- ▶ Bilateral DVT
- ▶ Caval obstruction
- ▶ Lymphedema
- ▶ Lipedema
- ▶ Obesity
- ▶ Right CHF
- ▶ Pulmonary hypertension
- ▶ Liver insufficiency
- ▶ Kidney insufficiency
- ▶ Hypothyroidism
- ▶ Medications – CCB, corticosteroids, hormones, NSAIDs

Lymphedema signs

Buffalo hump,
courtesy S Dean



Stemmer's sign,
courtesy Laredo & Lee

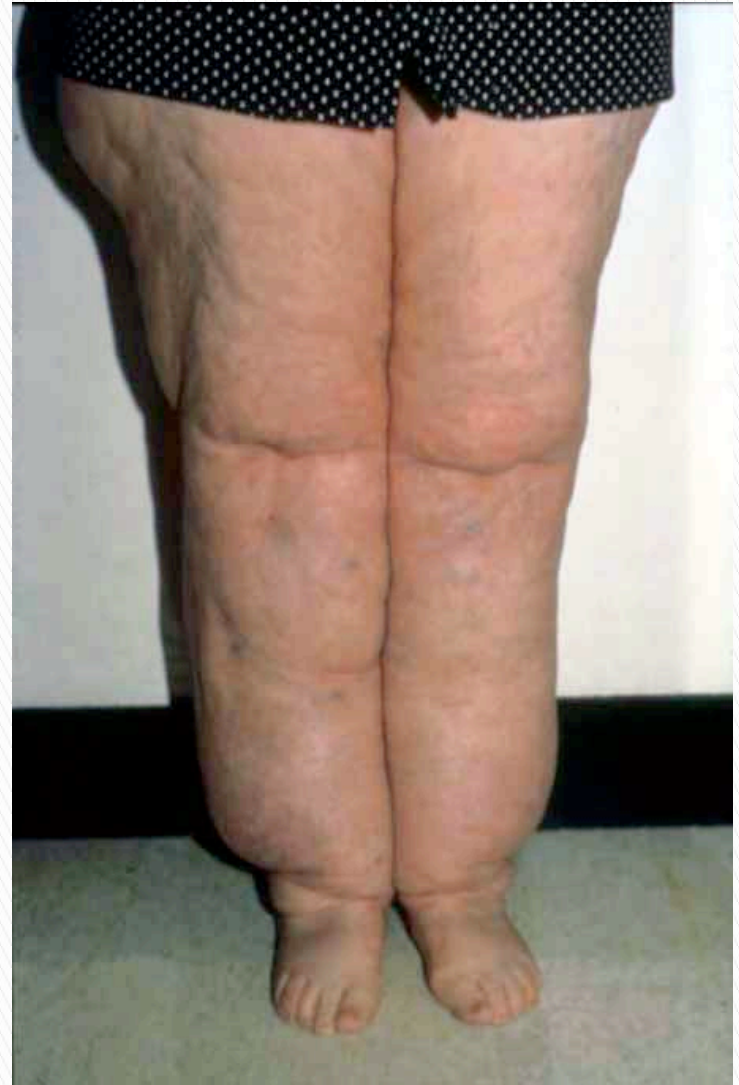


CEAP class C3 vein disease

- ▶ Starts at ankle, can progress up calf and thigh
- ▶ Saphenous ablation corrects only mild ankle swelling
- ▶ Rule out deep vein thrombosis or chronic obstruction
- ▶ Consider ilio caval vein obstruction
- ▶ Consider popliteal or axial deep vein reflux
- ▶ *Phlebolymphe*dema - 20–30% of patients with CVI have associated lymphatic dysfunction

Lipedema

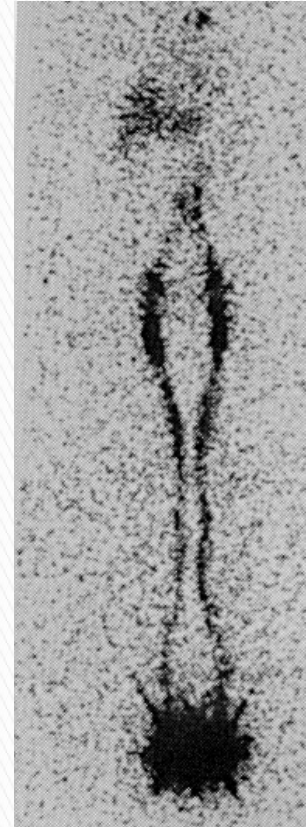
- ▶ Cause – subcutaneous fat deposition
- ▶ Painful fat syndrome
- ▶ Female
- ▶ Edema buttocks to ankles but spares feet
- ▶ Onset puberty to 20s
- ▶ Tx – exercise, weight control
- ▶ Can consider liposuction



Courtesy S Dean

Diagnostic testing

- ▶ Lymphedema is usually a clinical diagnosis
- ▶ Lymphoscintigraphy is best test to confirm
- ▶ Radiolabeled contrast injected between toes
- ▶ Delayed transport or backflow of tracer, asymmetric pattern



Courtesy S Dean

Treatment

- ▶ Compression stocking, 30–40 mm Hg
- ▶ Skin hygiene
- ▶ Consider physical therapy (lymphedema specialist) – manual lymphatic drainage
- ▶ Consider pneumatic compression

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Table 23.2 Physical treatments for lymphedema

Treatment	Effect
Exercise	Dynamic muscle contractions encourage movement of lymph along tissue planes and noncontractile, initial lymph vessels (passive drainage) and increased contractility of collecting lymph vessels (active drainage)
Compression garments	Opposes capillary filtration Acts as a counterforce to muscle contractions generating greater interstitial pressure changes
Manual lymphatic drainage	Form of massage therapy that stimulates lymph flow in more proximal, normally draining lymphatics to “siphon” lymph from congested areas
Compression bandaging	Used as an intensive treatment in combination with exercise to reduce large, misshapen lower limbs and permit subsequent maintenance treatment with compression stockings
Pneumatic compression	Softens and reduces limb volume but can forcibly displace fluid into the trunk and genitalia. Compression garments must be worn after treatment
Elevation	Does not stimulate lymph drainage, but lowers venous pressure and therefore capillary filtration, allowing lymph drainage to catch up

Complications

- ▶ Cellulitis - *Streptococcus, Staphylococcus*
- ▶ Lymphosarcoma - malignant degeneration

What can we do?

- ▶ Rule out venous sources – DVT, deep vein reflux, ilio caval
- ▶ Suspect lymphedema & lipedema
- ▶ Help PCP with workup
- ▶ Skin hygiene
- ▶ Obesity education
- ▶ Compression stockings
- ▶ Consider physical therapy (Lymphedema Clinic) and pneumatic compression
- ▶ Centers of excellence – Stanford, Ohio State, George Washington

Conclusions

- ▶ Rule out venous sources of swelling – deep vein thrombosis, ilio caval obstruction, deep vein reflux – and Baker’s cyst
- ▶ Suspect lymphedema with toe → foot → calf swelling & signs
- ▶ Consider lipedema for painful bilateral swelling with ankle cutoff in females

