

Lymphedema



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Definition

- *Abnormal limb swelling*
 - *due to accumulation of high protein ISF*
 - *secondary to defective lymphatic drainage*
 - *in the presence of (near) normal net capillary filtration.*
- *Confined to the epifascial space.*

Pathophysiology

1. *Lymphatic aplasia, hypoplasia, dysmotility, obliteration by inflammatory, infective or neoplastic processes, or surgical excision.*
2. *Lymphatic hypertension & distension, with valvular incompetence.*
3. *Accumulation in IFS of fluid, proteins, growth factors & other active peptide moieties, glycosaminoglycans & particulate matter, including bacteria.*

3. *There is increased collagen production, accumulation of inflammatory cells esp. macrophages & lymphocytes, & activation of keratinocytes.*
4. *End result is protein rich edema fluid, increased deposition of ground substance, subdermal fibrosis & dermal thickening & proliferation.*

Classification

Etiological Classification (Allen)

Primary lymphedema

1. *Congenital* → Onset < 2 years
 - *Sporadic*
 - *familial (Milroy's disease).*
2. *Praecox* → Onset 2-35 years
 - *Sporadic*
 - *familial (Meige's disease).*
3. *Tarda* → Onset after 35 years.

Secondary lymphedema

1. *Parasitic infection (filariasis).*
2. *Fungal infection (tinea pedis).*
3. *Exposure to foreign body material (silica particles).*
4. *Primary lymphatic malignancy.*
5. *Metastatic spread to lymph nodes.*
6. *Radiotherapy to lymph nodes.*
7. *Surgical excision of lymph nodes.*
8. *Trauma (particularly degloving injuries).*
9. *Superficial thrombophlebitis.*
10. *Deep venous thrombosis.*

Clinical Classification (Brunner)

1. *Subclinical (latent)*
2. *Grade I*
3. *Grade II*
4. *Grade III*

Lymphangiographic Classification (Browse)

1. *Congenital hyperplasia (10%)*
2. *Distal obliteration (80%)*
3. *Proximal obliteration (10%)*





















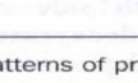
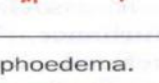
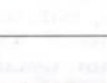
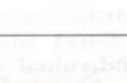

	Normal	Congenital hyperplasia	Distal obliteration (hypo/aplasia)	Proximal obliteration (hypo/aplasia) with distal hyperplasia	Proximal obliteration (hypo/aplasia) with distal obliteration
Thoracic duct					
Nodes					
Para-aortic					
Iliac					
Femoral					

Fig. 17.13 Lymphangiographic patterns of primary lymphoedema.

Clinical features

Symptoms

1. *Constant dull ache, even severe pain or cramp*
2. *Pins & needles, burning & bursting sensations*
3. *Sensitivity to heat*
4. *Skin problems, including dehydration, flakiness, weeping, excoriation & breakdown*
5. *Athlete's foot*
6. *Acute infective episodes*
7. *Backache & joint problems*
8. *Immobility, leading to obesity & muscle wasting*
9. *General tiredness & debility*

Signs

1. *Lymphedema characteristically involves the foot;*
 1. *Contour of the ankle is lost.*
 2. *Buffalo hump forms on the dorsum of the foot.*
 3. *Toes appear square.*
 4. *Stemmer's sign.*
2. *It usually spreads proximally to knee level & less commonly affects the whole leg.*



Fig. 17.2 The foot of a patient with typical lymphoedema.



Fig. 17.3 The lower leg of patient with typical lymphoedema.

3. *In the early stages, it will pit & the swelling is down in morning.*
4. *Later, fibrosis, dermal thickening & hyperkeratosis occur.*
5. *Chronic eczema, dermatophytosis, onychomycosis, fissuring, verrucae & papillae are frequently seen in advanced disease.*

6. *Lymphangiomas* are dilated dermal lymphatics that blister onto the skin surface;

1. If lymphangiomas are < 5 cm across, they are termed *lymphangioma circumscriptum*.
2. If they are more widespread termed *lymphangioma diffusum*.
3. If they form a reticulate pattern of ridges then termed *lymphedema ab igne*.
4. Lymphangiomas frequently weep →
 - Lymphorrhea & chyloorrhea (in skin),
 - protein losing diarrhea (in intestine),
 - chylous ascites (in peritoneal cavity),
 - chylothorax (in pleural cavity), &
 - chyluria (in urinary tract).

Discharge from lymphangiomas suggest lymphangectasia (megalymphatics) & chylous reflux.

7. *Ulceration, non-healing bruises, & raised purple-red nodules should lead to suspicion of malignancy.*

- *Lymphangiosarcoma in post mastectomy lymphedema is termed **Stewart-Treves syndrome**.*

Differential diagnosis

Non vascular or lymphatic:

■ General disease states

1. *Cardiac failure*
2. *Hepatic failure*
3. *Hypoproteinemia due to nephrotic syndrome, malabsorption, protein losing enteropathy*
4. *Hypothyroidism (myxedema)*
5. *Allergic disorders, including angioedema & idiopathic cyclic edema*
6. *Prolonged immobility & lower limb dependency*

■ Local diseases

1. *Ruptured Baker's cyst*
2. *Myositis ossificans*
3. *Bony or soft tissue tumors*
4. *Arthritis*
5. *Hemarthrosis*
6. *Calf muscle hematoma*
7. *Achilles tendon rupture*

■ Others

1. *Retroperitoneal fibrosis*
2. *Gigantism*
3. *Drugs: Corticosteroids, estrogens, progestogens, monoamine oxidase inhibitors, methyldopa, hydralazine, nifedipine*
4. *Trauma*
5. *Obesity*

Venous disorders

1. *Deep venous thrombosis*
2. *Post thrombotic syndrome*
3. *Varicose veins*
4. *Klippel Trenaunay syndrome*
5. *External venous compression*

Arterial disorders

1. *Ischaemia reperfusion*
2. *Arteriovenous malformation*
3. *Aneurysm*

Investigations

■ *Routine tests*

1. *Full blood count.*
2. *Urea, creatinine, & electrolytes.*
3. *Liver function tests.*
4. *Chest radiography.*
5. *Blood smear for microfilariae.*

■ *Diagnostic tests*

1. *Lymphangiography*

1. *Direct*

2. *Indirect*

2. *Isotope lymphoscintigraphy*



Fig. 17.7 This patient presented with congenital lymphoedema of the right leg. The lymphangiogram shows lymphatic hypoplasia.

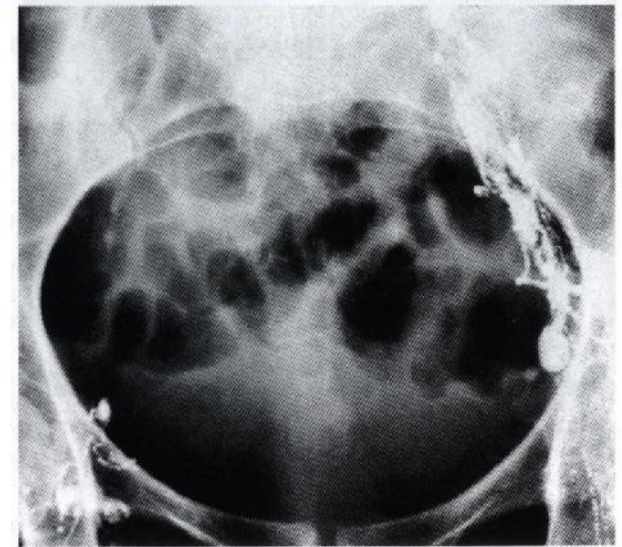


Fig. 17.8 This patient presented with lymphoedema of the right leg. Bipedal lymphangiogram demonstrated normal lymphatics in the right leg up to the inguinal nodes but no progression of contrast above the inguinal ligament; a case of proximal obstruction.

3. *Computerised tomography*

1. *A single axial slice thru the midcalf helps in differentiating;*
 1. *Lymphedema* → *Enlarged subcutaneous compartment, with coarse, reticular honeycomb pattern.*
 2. *Venous edema* → *Increased volume of muscular compartment.*
 3. *Lipoedema* → *Increased subcutaneous fat.*
2. *Also used to exclude pelvic or abdominal mass lesions.*

4. *Magnetic resonance imaging*

Useful in assessment of patients with lymphatic hyperplasia, & also distinguishes venous & lymphatic causes of a swollen limb.

5. *Ultrasound*

Provide useful information about venous function.

6. *Pathological examination*

Fine needle aspiration, needle core biopsy or surgical excision.

Management

Medical Treatment

1. *Relief of pain*

- *non-opioid & opioid analgesics,*
- *corticosteroids,*
- *tricyclic antidepressants,*
- *muscle relaxants,*
- *anti-epileptics,*
- *nerve blocks,*
- *physiotherapy,*
- *adjuvant anti-cancer therapies, &*
- *measures to reduce swelling if possible.*

2. *Control of swelling*

- *Decongestive lymphedema therapy (DLT)*

1. *Intensive phase*

- *It is a short intensive period of therapist led care, & comprises skin care, MLD, MLLB & exercises.*

2. *Maintenance phase*

- *Patient uses a self care regimen..*

Skin care

- *Guidance on skin hygiene &, specifically, the avoidance of acute infective episodes;*
 - *Protect hands when washing up or gardening.*
 - *Never walk barefoot & wear protective footwear outside.*
 - *Use an electric razor to depilate.*
 - *Never let the skin become macerated.*
 - *Treat cuts promptly.*
 - *Use insect repellent sprays & treat bites promptly.*
- *Antifungal prophylactic therapy to prevent athlete's foot.*
- *Antibiotic therapy &, if necessary, hospital admission for acute infective episodes.*
- *Management of lymphorrhea with elevation, compression & sometimes cautery under anesthetic.*

Manual lymphatic drainage (MLD)

- *Aim is to evacuate fluid & protein from the ISF space, & stimulate lymphangion contraction.*
- *Therapist should perform MLD daily; they should also train the patient &/or carer to perform a simpler massage, termed simple lymphatic drainage (SLD).*

Multilayer lymphoedema bandaging (MLLB) & compression garments

- *MLLB exert support thru the production of a semi rigid casing, where the resting pressure is low but changes quite markedly in response to movement & posture.*
- *Elastic garments provide compression, produce a sustained high resting pressure & follow in as limb swelling reduces; however, it does not alter greatly in response to muscular activity & posture.*
- *Pneumatic compression devices*

Exercise

- *Slow, rhythmic, isotonic movements eg swimming & massage will increase venous & lymphatic return.*
- *Vigorous exercise, esp. if it is anerobic & isometric (eg carrying heavy shopping bags or prolonged standing), will tend to exacerbate lymphedema.*
- *When at rest, the lymphedematous limb should be positioned with foot / hand above the level of heart.*

Surgical Treatment

1. *Bypass procedures*

1. *Ileal mucosal patch (Kinmonth).*
2. *Omental pedicle.*
3. *Skin bridge (Gillies).*
4. *Anastomosing lymph nodes to veins (Neibulowitz).*
5. *Direct lymphovenous anastomosis.*

2. *Limb reduction procedures*

1. *Sistrunk's operation*
2. *Homans' operation*
3. *Thompson's operation*
4. *Charles' operation*

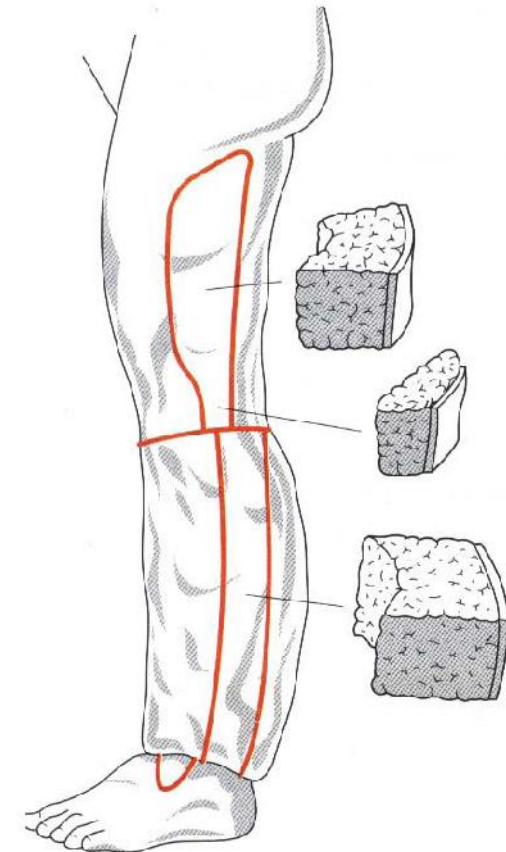


Fig. 17.14 Homan's procedure involves raising skin flaps to allow the excision of a wedge of skin and a larger volume of subcutaneous tissue down to the deep fascia. Surgery to the medial and lateral aspects of the leg must be separated by at least 6 months to avoid skin flap necrosis.

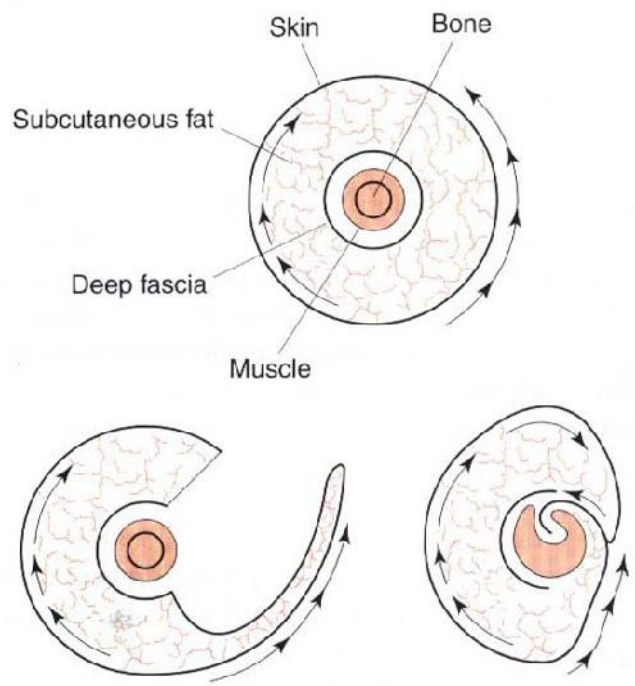


Fig. 17.16 A cross-sectional representation of Thompson's reduction operation; the buried dermal flap.

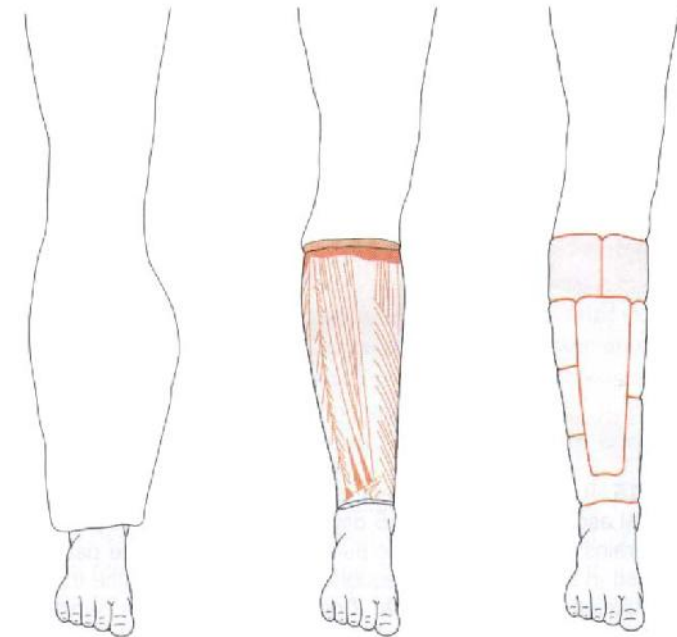
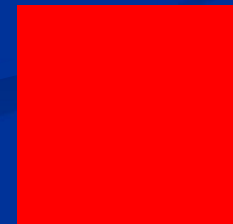


Fig. 17.17 Charles' procedure involves circumferential excision of lymphoedematous tissue down to and including the deep fascia followed by split skin grafting. This procedure gives a very poor cosmetic result but does allow the surgeon to remove very large amounts of tissue and is particularly useful in patients with severe skin changes.



■ **Causes of secondary lymphedema includes all of the followings, except?**

- A. Filariasis
- B. Tinea pedis
- C. Meige's disease
- D. Skeletal radiotherapy
- E. Metastasis to lymph nodes

Answer: D

■ Symptoms & signs of lymphedema includes all of the followings, except ?

- A. Intermittent limb pain
- B. Pins & needles
- C. Sensitivity to heat
- D. Stemmer's sign
- E. Dermal thickening & hyperkeratosis

Answer: A

■ **Useful investigations for lymphedema does not include ?**

- A. Lymphangiography
- B. Isotope lymphoscintigraphy
- C. Magnetic resonance imaging
- D. Doppler ultrasound
- E. CT scan

Answer: D

■ **Decongestive lymphedema therapy consists of all of the followings, except ?**

- A. Skin care
- B. Manual lymphatic drainage
- C. Multilayer lymphedema bandage
- D. Compression garments
- E. Vigorous isometric exercise

Answer: E

■ **Surgical treatment options of lymphedema includes all of the followings, except ?**

- A. Kinmonth's operation
- B. Appendix mucosal patch
- C. Sistrunk's operation
- D. Homan's operation
- E. Skin bridge

Answer: B

Thank you!