Diseases of lymphatic system

Lymphatic system

- Lymphatics commences from tissue spaces and empty into veins
- Lymph nodes
- Epitheliolymphoid tissues – walls of GIT , thymus spleen
- Function – return of protein rich fluid into circulation
  - Includes albumin, globulin, coagulation factors, growth factors , cholesterol, fat soluble vitamins etc

Development

- Four cystic spaces .. Neck and groin
- Cisterns
- Lower limb abdomen – through cisterna chyli – thoracic duct - left jugular vein
- Head and neck to right
- Lymphatics follow vein
- No lymphatics in brain and bone cortex

- Lymphatic system includes lymphatic channels, lymphoid organs and circulating elements
- Starlings law plays a role
- Lymph flows against a pressure gradient
  - Helped by muscle contractions
  - Contactions and relaxations of lymphangions
  - Valves

Diseases

- Acute lymphangitis
  - Staph/streptococcus
  - Lymphadenitis/abscess
  - Fever pain etc
  - Red streak
  - Rest, elevation, IV antibiotics
  - Incision and drainage
  - Recurrences --- acute inflammatory episode

- Chronic lymphangitis
  - Repeated attacks of acute lymphangitis
  - May be a cause for lymphoedema

Neoplasms

- Benign neoplasms
  - From birth
  - Neck, axilla, shoulder, groin
  - Localised clustures of dilated lymph sacs not connected to lymphatic system – lymphangiomas
  - Three types – capillary/simple, cavernous, cystic hygroma

Simple / capillary lymphangioma

- lymphangioma circumscriptum
- Dilated dermal lymphatics that blister on to skin
- Contains clear fluid or blood stained
- Inner thigh, shoulder or axilla
- Lymphangiography diagnostic – separate from main lymphatic trunk
- Treatment excision – after confirming the dignosis

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CAVERNOUS (DIFFUSE)LYMPHANGIOMA

- Bigger swelling
- Face, mouth, lips, tongue
- Intramuscular sometimes
- Very difficult to remove
- recurrence

Cystic hygroma

- Most common
- 75% neck, 20% axilla, 5% in mediastinum, pelvis, groin, retroperitoneum sometimes combinations
- Children from birth
- Painless, infection –produce pain
- Complete surgical excision
- Sclerotherapy ?
- Recurrence

Malignant neoplasm

- Lymphangiosarcoma
  - Rare
  - Long standing lymphoedema
  - Post mastectomy
  - Upper limb common
  - Bluish/purple discoloration- nodule- ulcer – diffuse
  - Treatment ineffective – fatal

- Lymphoedema
  - defined as abnormal limb swelling caused by the accumulation of increased amounts of high protein ISF secondary to defective lymphatic drainage in the presence of (near) normal net capillary filtration.

- 1 in 6000 births
- Physical , emotional , psychological

Summary box 58.1

Symptoms frequently experienced by patients with lymphoedema

- Swelling, clothing or jewellery becoming tighter
- Constant dull ache, even severe pain
- Burning and bursting sensations
- General tiredness and debility
- Sensitivity to heat
- 'Pins and needles'
- Cramp
- Skin problems, including flakiness, weeping, excoriation and breakdown
- Immobility, leading to obesity and muscle wasting
- Backache and joint problems
- Athlete's foot
- Acute infective episodes

- Unilateral lymphoedema
  - Mild - < 20% limb volume
  - Moderate – 20 – 40%
  - Severe - > 40%
- Pathophysiology of edema
  - Capillary filtration abnormality
  - Lymphahtic abnormality (true lymphoedema)
  - Both

Classification

- Primary
  - Cause unknown
  - Congenital lymphatic aplasia/hypoplasia
- Secondary
  - Definite cause

Table 58.1 Aetiological classification of lymphoedema.

Primary lymphoedema	Congenital (onset <2 years old): sporadic; familial (Nonne–Milroy's disease)
	Praecox (onset 2–35 years old): sporadic; familial (Letessier–Meige's disease)
	Tarda (onset after 35 years old)
Secondary lymphoedema	Parasitic infection (filariasis)
	Fungal infection (tinea pedis)
	Exposure to foreign body material (silica particles)
	Primary lymphatic malignancy
	Metastatic spread to lymph nodes
	Radiotherapy to lymph nodes
	Surgical excision of lymph nodes
	Trauma (particularly degloving injuries)
	Superficial thrombophlebitis
	Deep venous thrombosis

Table 58.2 Clinical classification of lymphoedema.

Grade (Brunner)	Clinical features
Subclinical (latent)	There is excess interstitial fluid and histological abnormalities in lymphatics and lymph nodes, but no clinically apparent lymphoedema
I	Oedema pits on pressure and swelling largely or completely disappears on elevation and bed rest
II	Oedema does not pit and does not significantly reduce upon elevation, positive Stemmer's sign
III	Oedema is associated with irreversible skin changes, i.e. fibrosis, papillae

Risk factors

Upper limb/trunk lymphoedema	Lower limb lymphoedema
Surgery with axillary lymph node dissection, particularly if extensive breast or lymph node surgery	Surgery with inguinal lymph node dissection
Scar formation, fibrosis and radiodermatitis from postoperative axillary radiotherapy	Postoperative pelvic radiotherapy
Radiotherapy to the breast or to the axillary, internal mammary or subclavicular lymph nodes	Recurrent soft tissue infection at the same site
Drain/wound complications or infection	Obesity
Cording (axillary web syndrome)	Varicose vein stripping and vein harvesting
Seroma formation	Genetic predisposition/family history of chronic oedema
Advanced cancer	Advanced cancer
Obesity	Intrapelvic or intra-abdominal tumours that involve or directly compress lymphatic vessels
Congenital predisposition	Orthopaedic surgery
Trauma in an 'at-risk' arm (venepuncture, blood pressure measurement, injection)	Poor nutritional status
Chronic skin disorders and inflammation	Thrombophlebitis and chronic venous insufficiency, particularly post-thrombotic syndrome
Hypertension	Any unresolved asymmetrical oedema
Taxane chemotherapy	Chronic skin disorders and inflammation
Insertion of pacemaker	Concurrent illnesses such as phlebitis, hyperthyroidism, kidney or cardiac disease
Arteriovenous shunt for dialysis	Immobilisation and prolonged limb dependency
Air travel	Air travel
Living in or visiting an area for endemic lymphatic filariasis	Living in or visiting an area for endemic lymphatic filariasis

Summary box 58.3

Malignancies associated with lymphoedema

- Lymphangiosarcoma (Stewart–Treves' syndrome)
- Kaposi's sarcoma (HIV)
- Squamous cell carcinoma
- Liposarcoma
- Malignant melanoma
- Malignant fibrous histiocytoma
- Basal cell carcinoma
- Lymphoma

Primary lymphoedema

- Familial or sporadic
  - Familial – milroy's disease and Meige's disease
- Age at onset
  - Congenita – 10% by birth
  - Praecox – 75% adolescence
  - Tarda – beyond 35 yrs – 15%
- There is some developmental anomaly
- Unilateral in 50%
- Family history 1/5th cases

- Lymphangiographically three types
  - Aplasia – 15% , congenital type
  - Hypoplasia – solitary lymph channel- 75%
  - Varicose/dilated lymph trunks in 12%- associated AV malformations
- Prognosis bad for aplasia and varicose type

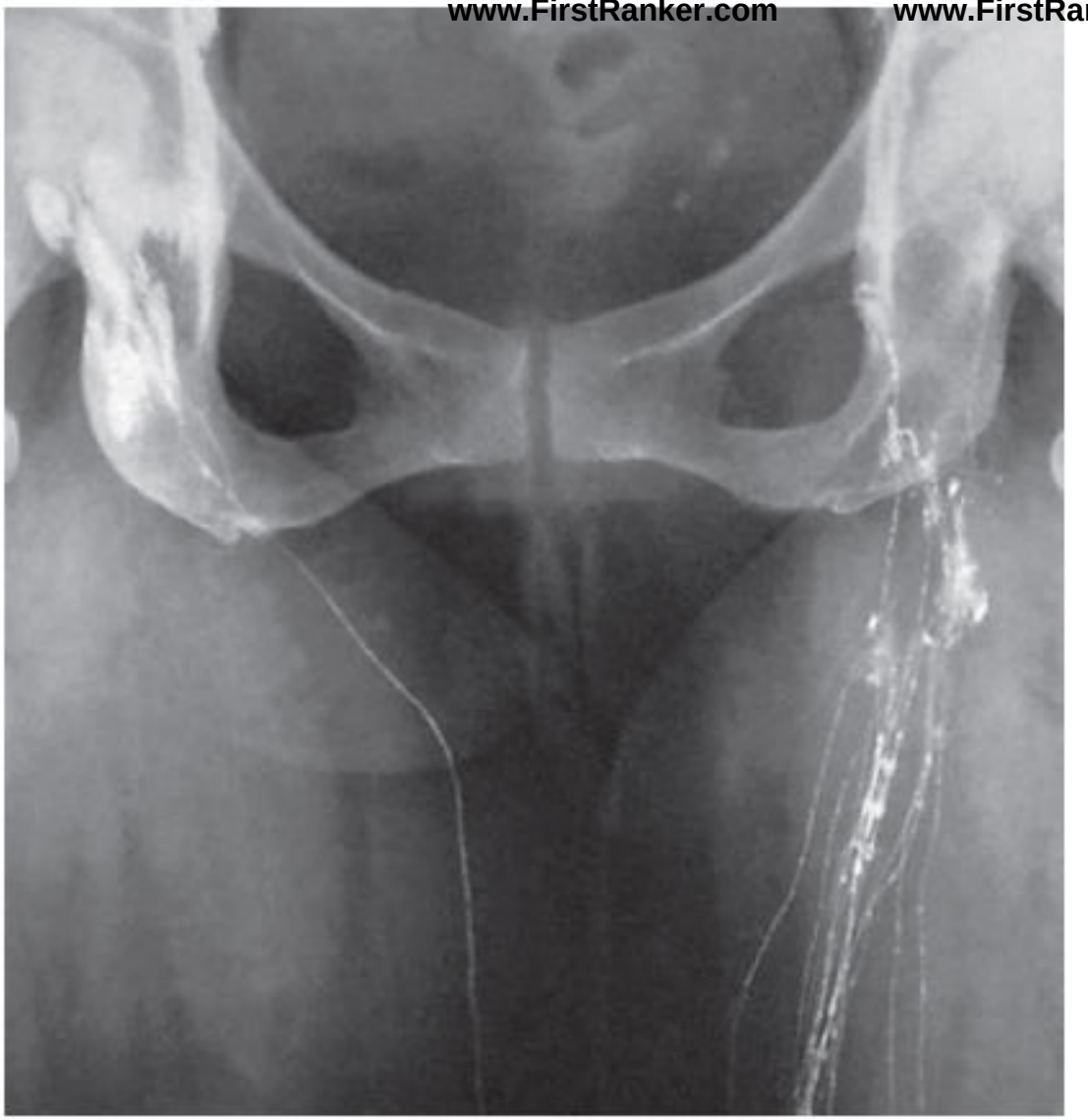


Figure 58.5 This patient presented with congenital lymphoedema of the right leg. The lymphangiogram shows lymphatic hypoplasia.

Secondary lymphoedema

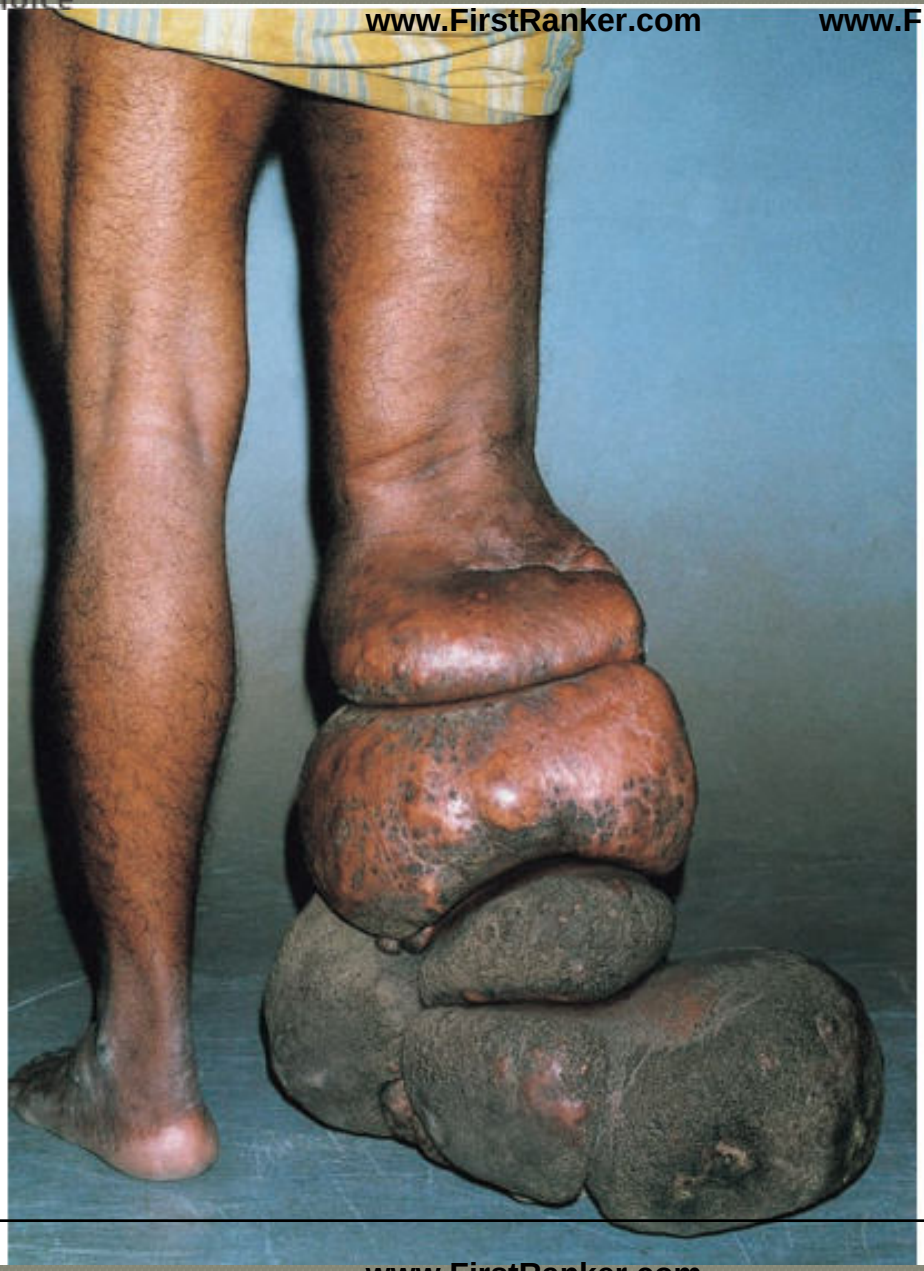
- Most common
- Infection/inflammation/trauma/tumor
- Filariasis commonest cause
- Certain conditions can mimic lymphodema like – factitious, immobility, lipoedema

Summary box 58.5

Features of lipoedema that help differentiate it from lymphoedema

- Occurs almost exclusively in women
- Onset nearly always coincides with puberty
- Nearly always bilateral and symmetrical
- Involvement of trunk
- The feet are not involved, leading to an inverse shouldering effect at the malleoli
- No pitting
- No response to elevation or compression
- No skin changes of lymphoedema (negative Stemmer's sign)
- MRI shows subcutanteous fat but no fluid accumulation

Trauma and tissue damage	Lymph node excision
	Radiotherapy
	Burns
	Variscose vein surgery/harvesting
	Large/circumferential wounds
	Scarring
Malignant disease	Lymph node metastases
	Infiltrative carcinoma
	Lymphoma
	Pressure from large tumours
Venous disease	Chronic venous insufficiency
	Venous ulceration
	Post-thrombotic syndrome
	Intravenous drug use
Infection	Cellulitis/erysipelas
	Lymphadenitis
	Tuberculosis
	Filariasis
Inflammation	Rheumatoid arthritis
	Dermatitis
	Psoriasis
	Sarcoidosis
	Dermatosis with epidermal involvement
Endocrine disease	Pretibial myxoedema
Immobility and dependency	Dependency oedema
	Paralysis
Factitious	Self-harm



Investigations

- Routine
- Lymphangiography
  - Direct
  - Indirect
- Isotope lymphoscintigraphy
- CT scan – Mass lesions
- MRI – LN
- USS – Venous system /DVT

	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					

e 58.9 Lymphangiographic patterns of primary lymphoedema.

Management

- Initial evaluation

Initial evaluation of the patient with lymphoedema

- History (age of onset, location, progression, exacerbating and relieving features)
- Past medical history including cancer history
- Family history
- Obesity (diet, height and weight, body mass index)
- Complications (venous, arterial, skin, joint, neurological, malignant)
- Assessment of physical, emotional and psychosocial symptoms
- Social circumstances (mobility, housing, education, work)
- Special needs (footwear, clothing, compression garments, pneumatic devices, mobility aids)
- Previous and current treatment
- Pain control
- Compliance with therapy and ability to self-care

- Relief of pain
- Control of swelling
  - Bedrest, elevation, compression, bandaging, exercise
  - Manual lymphatic drainage (MLD)
  - Multilayer lymphoedema bandaging (MLLB)
- Skin care

Skin care

- Protect hands when washing up or gardening; wear a thimble when sewing
- Never walk barefoot and wear protective footwear outside
- Use an electric razor to depilate
- Never let the skin become macerated
- Treat cuts and grazes promptly (wash, dry, apply antiseptic and a plaster)
- Use insect repellent sprays and treat bites promptly with antiseptics and antihistamines
- Seek medical attention as soon as the limb becomes hot, painful or more swollen
- Do not allow blood to be taken from, or injections to be given into, an affected arm (and avoid blood pressure measurement)
- Protect the affected skin from sun (shade, high-factor sun block)
- Consider taking antibiotics if going on holiday

Effects of MLLB

- Reduces oedema
- Restores shape to the affected area
- Reduces skin changes (hyperkeratosis, papillomatosis)
- Eliminates lymphorrhoea
- Supports inelastic skin
- Softens subcutaneous tissues

- Treat acute infective episodes
- Exercises
- Drugs
  - Flavanoids / benzpyrones
  - Diuretics ?
- Surgery

Surgeries

- Rare
- Minimal benefit
- Three types
- Bypass procedures
  - Gille's – Omental/skin bridge
  - Kinmonth – ileal mucosal patch
  - Neibulowitz – LN to vein
  - Lymphaticovenular Anastomosis (LVA)

- Liposuction
- Limb reduction procedures
  - Sistrunk – wedge excision and suturing
  - Homans
  - Thompson's
  - Charle's

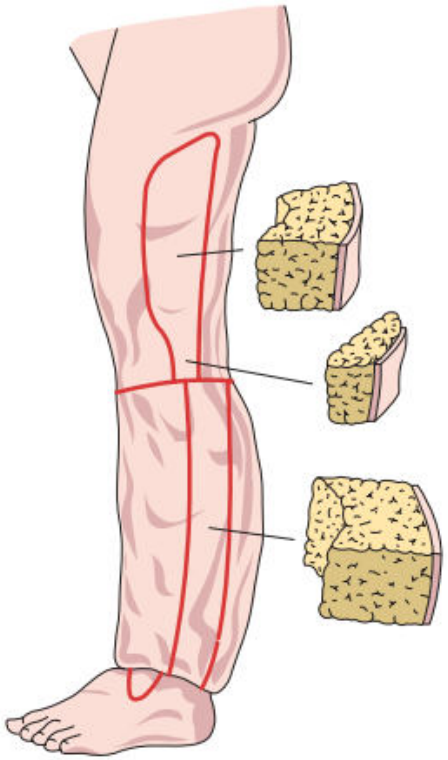


Figure 58.11 Homans' procedure involves raising skin flaps to allow the excision of a wedge of skin and a larger volume of subcutaneous tissue down to the deep fascia. Surgery to the medial and lateral aspects of the leg must be separated by at least six months to avoid skin flap necrosis.

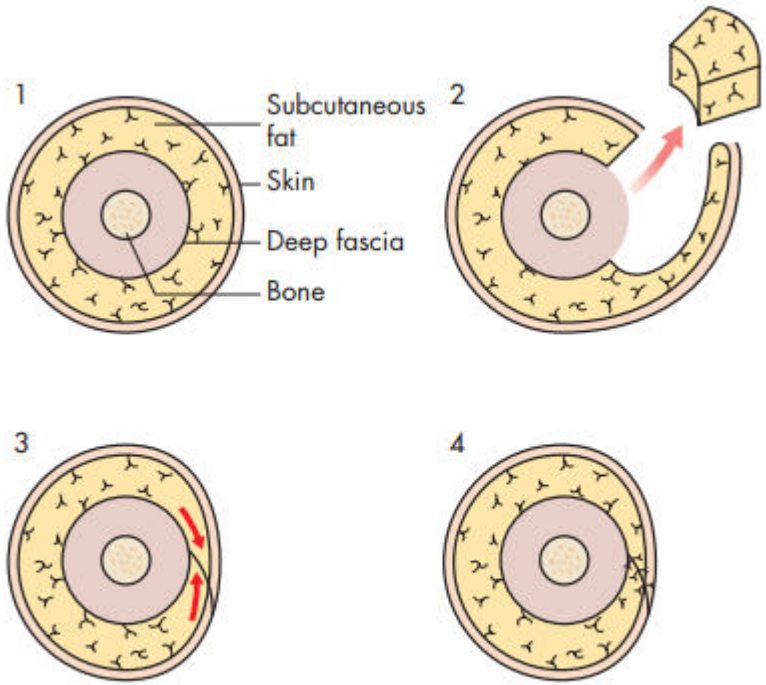


Figure 58.12 A cross-sectional representation of Thompson's reduction operation; red arrows illustrate the buried dermal flap sutured to deep fascia.

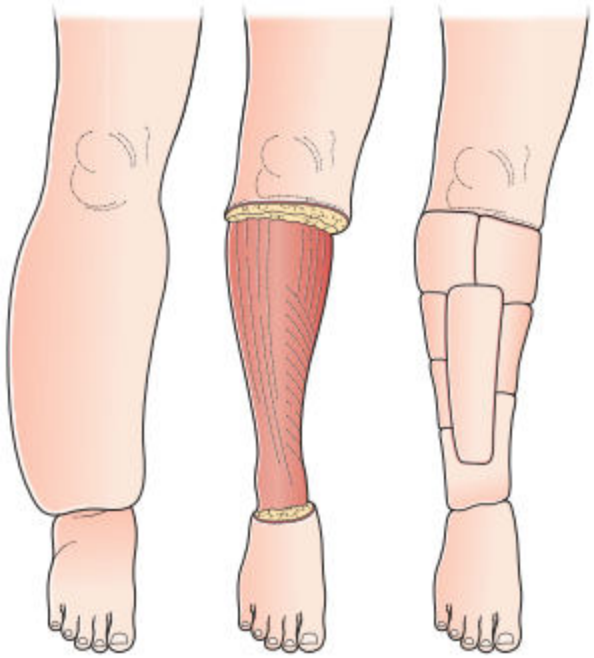


Figure 58.13 The Charles procedure involves circumferential excision of lymphoedematous tissue down to and including the deep fascia followed by split-skin grafting. This procedure gives a very poor cosmetic result but does allow the surgeon to remove very large amounts of tissue and is particularly useful in patients with severe skin changes.