LYMPHOEDEMA

External manifestation of lymphatic insufficiency in which progressive, chronic swelling causes distortion in size/shape of limbs and reduction in mobility/function
Low output: impaired capacity of lymphatics to drain microvascular filtrate
High output: excessive filtrate exceeds drainage capacity of normal lymphovascular system (cirrhosis/nephrotic/CVI/CCF)

AETIOLOGY OF LOW OUTPUT LYMPHOEDEMA

1. Primary (intrinsic): aplasia/hypoplasia/hyperplasia/dysmotility of developing lymphatics
(i)**CONGENITAL** (soon after/at birth): (i)Familial (Milroy's disease: autosomal; VEGFR-3) (ii)Non-familial
(ii)**PRAECOX** (<35yrs): (i)Familial (lymphoedema-distichiasis syndrome: FOXC2) (ii)Non-familial
(iii)**TARDA** (>35yrs)

2. Secondary (extrinsic): lymphatic obliteration 80%/obstruction 10% /valvular incompetence 10%
(i)**MALIGNANCY:** commonest in Europe; starts in thighs
(ii)I**NFECTION:** parasitic (*Wucherichia bancrofti* filariasis commonest worldwide)/bacterial (BHS, S. aureus, TB)
(iii)**SURGERY**: ALND/groin dissection/arterial or venous surgery
(iv)**RADIOTHERAPY**
(v)**VENOUS DISEASE** (inc DVT/DVI)
(vi)**TRAUMA** (degloving)

BROWSE'S CLASSIFICATION (lymphangiographic findings)
OBLITERATIVE (80%): progressive destruction of distal lymphatics (women, bilateral)
PROXIMAL OBSTRUCTION (10%): proximal occlusion in abdo/pelvic nodes (unilateral)
LYMPHATIC VALVULAR INCOMPETENCE (10%: incomplete valve development → diltation and hyperplasia (bilateral)

PATHOGENESIS
Progressive lymphatic HTN → impaired contractility/valve incompetence
Accumlate protein rich fluid in ISF
Collagen synthesis/keratinocyte activation → fibrosis of skin

CLINICAL FEATURES

HISTORY
Swelling: slowly progressive/?reduction with elevation/commence distally and progresses proximally (not Ca)
Pain/heaviness
Lymphangitis/Cellulitis/Erysipelas

EXAMINATION
I: swelling (uniform); skin (peau d'orange, hyperkeratosis, thickens with loss of elasticity (Stemmer's sign), vesicles)
look for scars of surgery/DXT; fungal infection; cellulitis/erysipelas
Pa: pitting early, non-pitting later; Stemmer's sign; nodes
Pe:
A:

ISL STAGING:
STAGE 0: (subclinical) no swelling despite impaired flow
STAGE 1: (early) fluid accumulates and reduces with elevation; pitting
STAGE 2 (late) elevation fails to reduce swelling; non-pitting due to fibrosis
STAGE 3: Lymphostatic elephantiasis (no pitting; trophic skin changes ie hyperkeratosis warts, fat deposits, fibrosis, fissuring, verrucae, eczema acanthosis)

INVESTIGATION
(1) uncertain diagnosis (2) assess causes (3) surgery planned
DUPLEX US
LYMPHANGIOSCINTIGRAPHY: isotope flow gives overall limb assessment; 92% sensitive, 100% specific
INTERSTITIAL MR LYMPHANGIOGRAPHY: dilated lymphatics, collateral with dermal backflow and obstruction
CT: dilated lymphatics/assess causes/monitor treatment response by CSA
MRI: circumferential subcutaneous oedema, dermal thickening, honeycomb fibrosis between muscles and dermis
CONTRAST LYMPHANGIOGRAPHY: if considering microvascular reconstruction
FLUORESCENCE MICROLYMPHANGIOGRPAHY: diagnosis and subtyping:
Milroys: aplasia   Lipoedema: lymphatic aneurysms

Bloods: renal/liver function, albumin, TFTs   CXR: cardiomegaly
Urine dip   ECHO: CCF

<u>MANAGEMENT</u>

1. Reduce swelling 2. Reduce infection 3. Improve function
Conservative treatment successful early (pitting phase) || Surgery for severe or resistant cases

<u>GENERAL</u>

Elevation/exercise/weight loss/skin care and hygiene   (ISL: no specific diet, no fluid restriction)s

<u>CONSERVATIVE</u>

MANUAL LYMPHATIC DRAINAGE/MASSAGE: squeeze prox and work prox to distal; not beneficial alone
GRADUATED ELASTIC COMPRESSION: 50mmHg+; multilayer bandages improve limb shape before fitting stockings
IPC (INTERMITTENT PNEUMATIC COMPRESSION
THERMAL TREATMENT
COMPLEX DECONGESTIVE PHYSIOTHERAPY

<u>SURGERY</u>

SEVERE AND RESISTANT CASES

1. DEBULKING OPERATIONS
(i)Excise skin/subcutaneous tissue; for advanced fibrotic lymphoedema
(Homan's= ellipse excise w/ primary closure)(Charles= all skin and subcut tissue down to fascia)
(Sistrunk's= wedge excision)(Thompson=buried dermal flap)
(ii) Lipectomy/lipsuction in non-fibrotic lymphoedema

2. BYPASS PROCEDURES= regional lymphatic obstruction
Lymphatic-lymphatic bypass/lymphatico-venous shunts

AVOID skin/muscle/omentum transposition and enteromesenteric bridging, no supportive evidence