Lower Limb Swelling &Lymphoedema

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Swelling

Swelling is the enlargement of a body part or organ beyond it normal size and usually causing a distortion of the shape and structure of the affected area.

Fluid

Gas

Mass

The swollen leg

Swelling is a sign for many conditions affect the human body .

These conditions could be vascular or non vascular .

It could be unilateral or bilateral .

Non vascular or lymphatic

General disease states :

- Cardiac failure from any cause .
- Liver failure .
- Hypoproteinaemia due to nephrotic syndrome, malabsorption, protein-losing enteropathy
- Hypothyroidism (myxoedema)
- Allergic disorders, including angioedema and idiopathic cyclic oedema
- Prolonged immobility and lower limb dependency

Non vascular or lymphatic

Local disease processes :

- Ruptured Baker's cyst
- Myositis ossificans
- Bony or soft-tissue tumours
- Arthritis
- Haemarthrosis
- Calf muscle haematoma
- Achilles tendon rupture
- Cellulitis
- Athlete's foot

Non vascular or lymphatic

- *Retroperitoneal fibrosis May lead to arterial, venous and lymphatic abnormalities*
- *Gigantism: Rare All tissues are uniformly enlarged*
- *Drugs : Corticosteroids, oestrogens, progestogens Monoamine oxidase inhibitors, phenylbutazone, methyldopa, hydralazine, nifedipine*
- *Trauma : Painful swelling due to reflex sympathetic dystrophy*
- *Obesity : Lipodystrophy Lipoidosis*

Venous

- Deep venous thrombosis
-
- Phlebitis
- Post-thrombotic syndrome
- Varicose veins
- Klippel–Trenaunay’s syndrome
- External venous compression

Arterial and lymphatic

- Aneurysm
- Inflammation in the lymph nodes
- Lymphedema

Common Causes of Leg Edema

Table 1

Unilateral		Bilateral	
Acute (<72 hours)	Chronic	Acute (<72 hours)	Chronic
Deep vein thrombosis	Venous insufficiency		Venous insufficiency
			Pulmonary hypertension
			Heart failure
			Idiopathic edema
			Lymphedema
			Drugs
			Premenstrual edema
			Pregnancy
			Obesity

Less Common Causes of Leg Edema

Table 2

Unilateral		Bilateral	
Acute (<72 hours)	Chronic	Acute (<72 hours)	Chronic
Ruptured Baker's cyst	Secondary lymphedema (tumor, radiation, surgery, bacterial infection)	Bilateral deep vein thrombosis	Renal disease (nephrotic syndrome, glomerulonephritis)
Ruptured medial head of gastrocnemius	Pelvic tumor or lymphoma causing external pressure on veins	Acute worsening of systemic cause (heart failure, renal disease)	Liver disease
Compartment syndrome	Reflex sympathetic dystrophy		Secondary lymphedema (secondary to tumor, radiation, bacterial infection, filariasis)
			Pelvic tumor or lymphoma causing external pressure
			Dependent edema
			Diuretic-induced edema
			Dependent edema
			Preeclampsia
			Lipidema[8]
			Anemia

Rare Causes of Leg Edema

Table 3

Unilateral		Bilateral	
Acute (<72 hours)	Chronic	Acute (<72 hours)	Chronic
	Primary lymphedema (congenital lymphedema, lymphedema praecox, lymphedema tarda)		Primary lymphedema (congenital lymphedema, lymphedema praecox, lymphedema tarda)
	Congenital venous malformations		Protein losing enteropathy, malnutrition, malabsorption
	May-Thurner syndrome (iliac-vein compression syndrome)[51]		Restrictive pericarditis
			Restrictive cardiomyopathy
			Beri Beri
			Myxedema

Idiopathic edema

A pitting edema of unknown cause that occurs primarily in pre-menopausal women who do not have evidence of heart, liver, or kidney disease.

In this condition, the fluid retention at first may be seen primarily pre-menstrually , which is why it sometimes is called "*cyclical*" edema. However, it can become a more constant and severe problem. Obesity and depression can be associated with this syndrome, and diuretic abuse is common

Spironolactone is considered the drug of choice for idiopathic edema , Avoiding environmental heat, low salt diet, avoiding excessive fluid intake, and weight loss for obese patients.

Venous insufficiency

Chronic pitting edema, often associated with brown hemosiderin skin deposits on the lower legs. The skin changes can progress to dermatitis and ulceration, which usually occur over the medial maleoli.

Other common findings include varicose veins and obesity. Most patients are asymptomatic but a sensation of aching or heaviness can occur.

Diagnosis **: is usually made clinically but can be confirmed with a Doppler study. Although CVI is thought to result from previous DVT , only one third of patients will give that history**

Treatment **: compression stockings**

History

Key elements of the history include :

What is the *duration* of the edema (acute [<72 hours] vs. chronic)? If the onset is acute, DVT should be strongly considered.

Is the *edema* painful ? DVT and reflex sympathetic dystrophy are usually painful. CVI can cause low-grade aching. Lymphedema is usually painless.

What *drugs* are being taken? Calcium channel blockers, prednisone, and anti-inflammatory drugs are common causes of leg edema

Is there a history of *systemic disease* (heart, liver, or kidney disease)?

Antihypertensive drugs
Calcium channel blockers
Beta blockers
Clonidine
Hydralazine
Minoxidil
Methyldopa
Hormones
Corticosteroids
Estrogen
Progesterone
Testosterone
Other
Nonsteroidal anti-inflammatory drugs
Pioglitazone, Rosiglitazone
Monoamine oxidase inhibitors

Physical Examination

Body mass index. Obesity

Distribution of edema

Tenderness

Pitting

Varicose veins

Kaposi-Stemmer sign : *(lymphedema)*

Skin changes

Signs of systemic disease: findings of heart failure (especially jugular venous distension and lung crackles) and liver disease (ascites, spider hemangiomas, and jaundice) may be helpful in detecting a systemic cause.

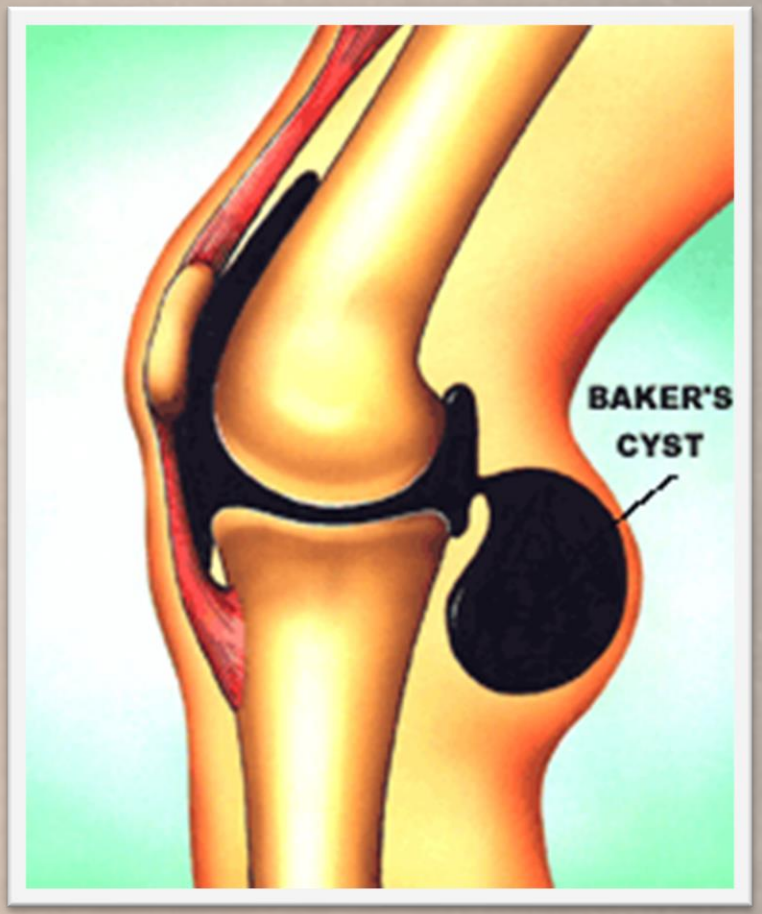
***Kaposi-Stemmer sign*: inability to pinch a fold of skin at base of second toe because of thickened skin indicates lymphoedema**

cellulitis

Varicose veins

Clinical presentation :

- **Local pain and edema**
- **Local inflammation**
- **Local hemorrhage into the surrounding tissue**
- **Dilated superficial veins**


BAKER'S
CYST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LYMPHOEDEMA

Definition

Lymphedema may be 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.

At birth, 1 in 6000 persons will develop lymphoedema

LYMPHOEDEMA

Classification

Two main types of lymphoedema are recognised:

1 *primary lymphoedema*, in which the cause is unknown (or at least uncertain and unproven); it is thought to be caused by congenital lymphatic dysplasia

2 *secondary or acquired lymphoedema*, in which there is a clear underlying cause.

LYMPHOEDEMA

Aetiological Classification

- Primary lymphoedema
- Congenital -onset <2 yrs
  Sporadic
  Familial (Milroy's disease
- Praecox(onset -2-35yrs)
  Sporadic
  Familial(Meige's dise)
- Tarda – onset – after 35 yrs old)

- Secondary lymphoedema
  Parasitic – Filariasis
  Fungus - Tinea pedids
  Exposure to F B particles silica particles Primary lymphatic malignancy
  Lymph nodes metastasis
  Radiotherapy to L N
  Surgical excision of L N
  Trauma –Degloving injury
  Supf. Thrombophlebitis
  Deep venous thrombosis

primary lymphoedema

Three types of primary lymphedema are distinguished by age of onset.

- ***Congenital lymphedema*** **is present at birth or occurs early in infancy.It accounts for fewer than 10% of primary lymphedema cases.Lymphedema that is both congenital and hereditary is known as** ***Milroy's disease*****.**
- ***Lymphedema praecox*** **occurs at any time from puberty until the end of the third decade. Most cases of primary lymphedema are of this type.It is three times more common in women than in men.**
- ***Lymphedema tarda*** **occurs after age 30.**

secondary lymphoedema

Secondary lymphedema is due to obstruction from a variety of causes, including infection, parasites, mechanical injury (including surgery), postphlebitic syndrome, and neoplasms.

***In developed countries*, the most common causes are obstruction by malignancies, postsurgical lymphedema (e.g., after mastectomy), and lymphatic destruction from therapeutic radiation.**

***In less well-developed countries*, parasitic obstruction (elephantiasis) is a common cause. Wuchereria bancrofti is the most common offending parasite.**

Table 55.6 Classification of causes of secondary lymphoedema

Classification	Example(s)
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

Malignancies associated with lymphoedema

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

Table 55.2 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
III	Oedema is associated with irreversible skin changes, i.e. fibrosis, papillae

Filariasis

- This is the most common cause of lymphoedema worldwide, affecting up to 100 million individuals.
- It is particularly prevalent in Africa, India and South America where 5–10% of the population may be affected.
- The viviparous nematode *Wuchereria bancrofti*, whose only host is man, is responsible for 90% of cases and is spread by the mosquito.
- The disease is associated with poor sanitation.
- The parasite enters lymphatics from the blood and lodges in lymph nodes, where it causes fibrosis and obstruction

Summary box 55.4

Features of filariasis

Acute

- Fever
- Headache
- Malaise
- Inguinal and axillary lymphadenitis
- Lymphangitis
- Cellulitis, abscess formation and ulceration
- Funiculo-epididymo-orchitis

Chronic

- Lymphoedema of legs (arm, breast)
- Hydrocele
- Abdominal lymphatic varices
  - Chyluria
  - Lymphuria

elephantiasis

Symptoms

- Constant dull ache or severe pain
- Burning and bursting sensations in limb
- General tiredness and debility
- Sensitivity to heat, cramps, pins and needles
- Skin problems- dehydration, flakiness, weeping excoriation and breakdown
- Immobility of patient leading to obesity and muscle weakness
- Backache and joint problems
- Athlete's foot and acute infective episodes

Conditions mimicking lymphoedema

- Factitious lymphoedema- Caused by application of a tourniquet (a start and sharp cut off is seen on examination) or hysterical disuse of limb in pts with psychological or psychiatric problems
- Immobility lymphoedema- Generalised or localised immobility of any cause leads to chronic limb swellling e.g.elderly person who spends all day or night sitting in a chair (arm chair legs), the hemiplegic stroke patient or young patient with multiple sclerosis
- Lipoedema- Seen only in women as B/L symmetrical enlargement of legs and sometimes lower half of the body due to abnormal deposition of fat . It may or may not be associated with obesity .It can coexist with other causes of limb swelling.

Complications

- Slow wound healing
- Infection- Cellulitis – Lymphangitis
  - Lymphadenitis
- Skin Ulcers ,thickening of skin ,follicles forms
- Malignancy - Lymphangiosarcoma
  (Stewart Treves Synderome)
  - Retiform haemangioendothelioma
  (low grade angiosarcoma)

Investigation of Lymphoedema

- Are investigations necessary ?

  It is usually possible to diagnose and manage lymphoedema purely on the basis of history and examination when swelling is mild and there are no complicating features In pts with severe , atypical and multifactorial swelling investigations may help to confirm the diagnosis ,management and prognostic information

Investigations

- **Routine Tests** - TLC,DLC RFT, LFT, Thyroid function tests, total plasma proeins,albumen , fasting blood sugar, urine exam for chyluria , blood smear for microfilariae, X ray chest and ultrasound
- **Lymphangiography**- Direct method involves injection of contrast medium into peripheral lymphatic channel followed by radiographic visualisation of the lymphatic vessels and nodes . It is the gold standard for showing abnormalities of large lymphatics and lymph nodes. It can be technically difficult , is unpleasant for the pt , may cause further injury to lymphatics. As a routine it has become obsolete. Indirect lymphangiography involves intradermal inj. Of water soluble non ionic contrast into a web space it is taken up by lymphatics and is followed radiographically . It will show distal lymphatics but not proximal lymphatics and lymph nodes.

- **Isotope lymphoscintigraphy** - This has become primary diagnostic technique in case of unccertainty. Radioacctve technitium labelled protein or colloid particles are injected into interdigital web space and taken up by lymphatics .Serial films are taken by a gamma camera.It provides a qualitative measure of lymphatic function. Quantitative function is performed using a dynamic (exercise) component and provides information on lymphatic transport

Investigations

- **Computed tomography** - A single axial CT slice through the midcalf is a useful test for lymphoedema (coarse, non enhancing reticular honeycomb pattern in an enlarged subcutaneous compartment), Venous oedema (increased volume of the muscular compartment and lipoedema (increased sub cutaneous fat). CT will diagnose a pelvic or abdominal mass lesion
- **MRI** – It can provide clear images of lymphatic channels and lymph nodes and is useful in assessment of lymphatic hyperplasis,MRI can differentiate between venous and lymphatic causes of a swollen limb and can detect tumours causing lymphatic obstruction
- **Ultrasound**- It can provide information about venous function like DVT and venous abnormalities
- **Pathological examination-** If malignancy is suspected FNAC , neddle cone biopsy or surgical excision from lymph nodes is useful . Skin biopsy will confirm the diagnosis of lymphangiosarcoma

Management of Lymphoedema

- Overview - History (age of onset, location, progresssion,aggravating and relieving factors)
- Past & Family medical history (H/O cancer)
- Obesity (Diet, Ht and wt, BMI)
- Complications –venous,arterial,skin,joint
- Relief of Pain *Control of swelling
- Skin care * Exercise
- Manual lymphatic drainage (MLD)
- Multilayer bandaging & compressive garments
- Drugs *Surgery

Management

- Skin care to treat infections and optimise condition of the skin
- Manual Lymphatic Drainage (MLD) to enhance lymph flow.This precedes bandaging and directs lymph fluid to functional territories and helps to form collateral pathways
- Application of multi layer compression bandaging
- Exercise to increase lymphatic & venous flow-Massage and swimming are beneficial.Avoid vigrous exercises
- A compressive garment is used to preserve the reduction acieved by treatment and prevent progression of lymphoedema
- Educat ion and Psychosocial support

Management

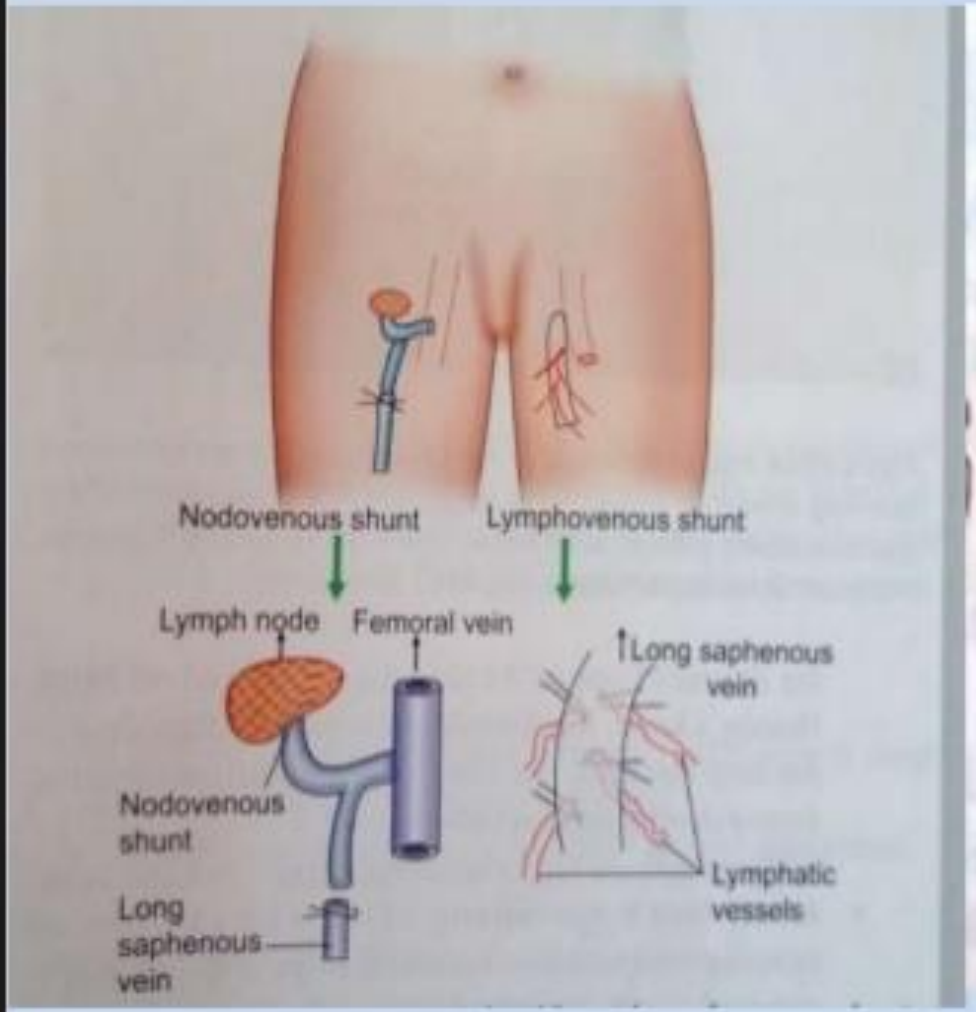
*__Compressive Therapy__ - Use of compression garments is very important in treatment of lymphoedema. They apply medically proper pressure to the swollen region to reduce pooling of fluid. This preserve the limb size and shape , supports and improves lymphatic circulation. These garments are available in a variety of styles,sizes, colours and grades of compression (Class I- IV).More swelling needs stronger support.They come as Pre –sized (ready to wear) or custom made as a circular knit or flat knit design. They are worn during day and removed at night

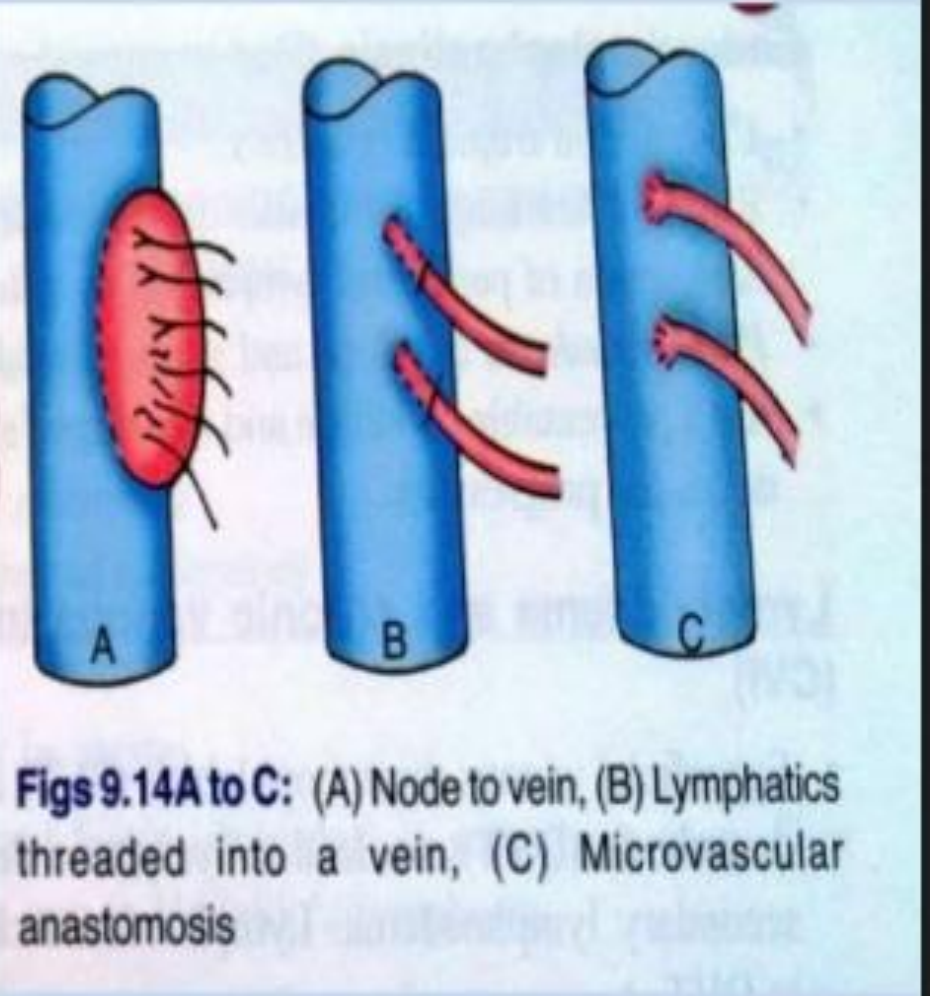
Surgery

- **Bypass Procedures-** In patients with proximal ilioinguinal obstruction and normal distal lymphatic channels , lymphatics bypass may help
  * Omental Pedicle * Skin bridge
  * Ileal mucosal patch
  * Lymph node to vein anastomosis
  * Direct lymphaticovenularanastomosis

  Technically these are demanding operations and there is no conclusive proof that they produce better results than best medical management .

By Pass operations





Figs 9.14A to C: (A) Node to vein, (B) Lymphatics threaded into a vein, (C) Microvascular anastomosis

Surgery

- Lymphatic liposuction –

  Using vibrating canulae excessive fluid, fat and thickened tissue is removed from the limb having advanced lymoedema. It has been shown to be an effetive method to reduce the size and stiffness of the affeted limb. It is less traumatic and a simple method comparing with standard excisional operations

Low level Laser therapy- Started in 2008 This has been used in post mastectomy upper arm lymphoedema and hav e shown good results in decreasing size of the involved upper limb

Surgery

Excisional Operations

- They are indicated when a limb is so swollen that it interferes with mobility and livelihood. They are not ' cosmetic' as they do not create a normally shaped leg and cause significant scarring. Various operations are-
- **Sistrunk -** A wedge of skin and subcutaneous tissue is excised and wound is primarily closed Mainly done to reduce girth of the thigh.
- **Homans** – Skin flaps are elevated then sub cutaneous tissue is excised beneath flaps , these are trimed in size and closed primarily.Good operation for calf . Main complication is skin flap necrosis.There should be 6 months gap between operations on medial and lateral sides of limb .Can be used for upper limb if there is no venous obstruction or malignancy

Surgery

- **Thompson operation** - One skin flap is sutured to the deep fascia and buried under second skin flap (buried dermal flap).Cosmetic results are not better than Homans operation and formation of pilonidal sinus is common.
- **Charles operation** – It was intially designed for filariasis and involved excision of all the skin and subcutaneous tissues down to deep fascia and covering with splitt skin grafts. This gives very unsatisfactory cosmetci result and graft failure is common.Advantage is that it reduces the size of massively swollen limb very much

Thomson operation

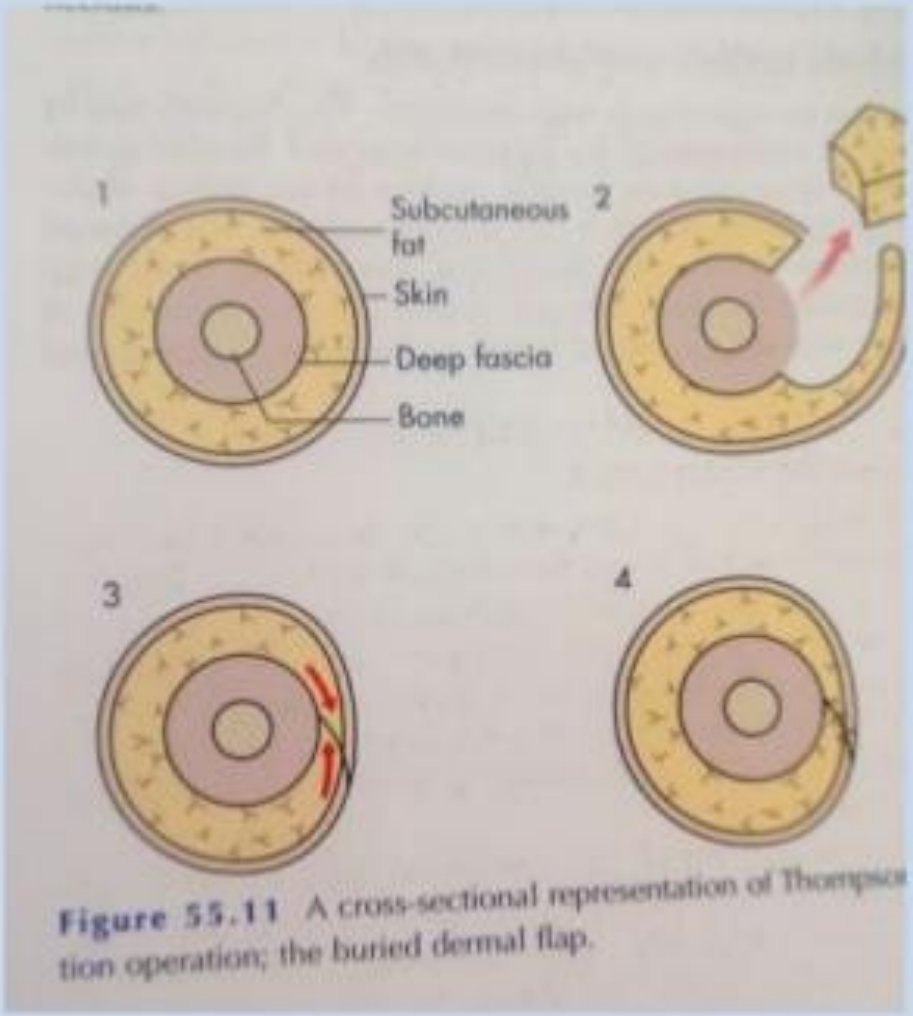
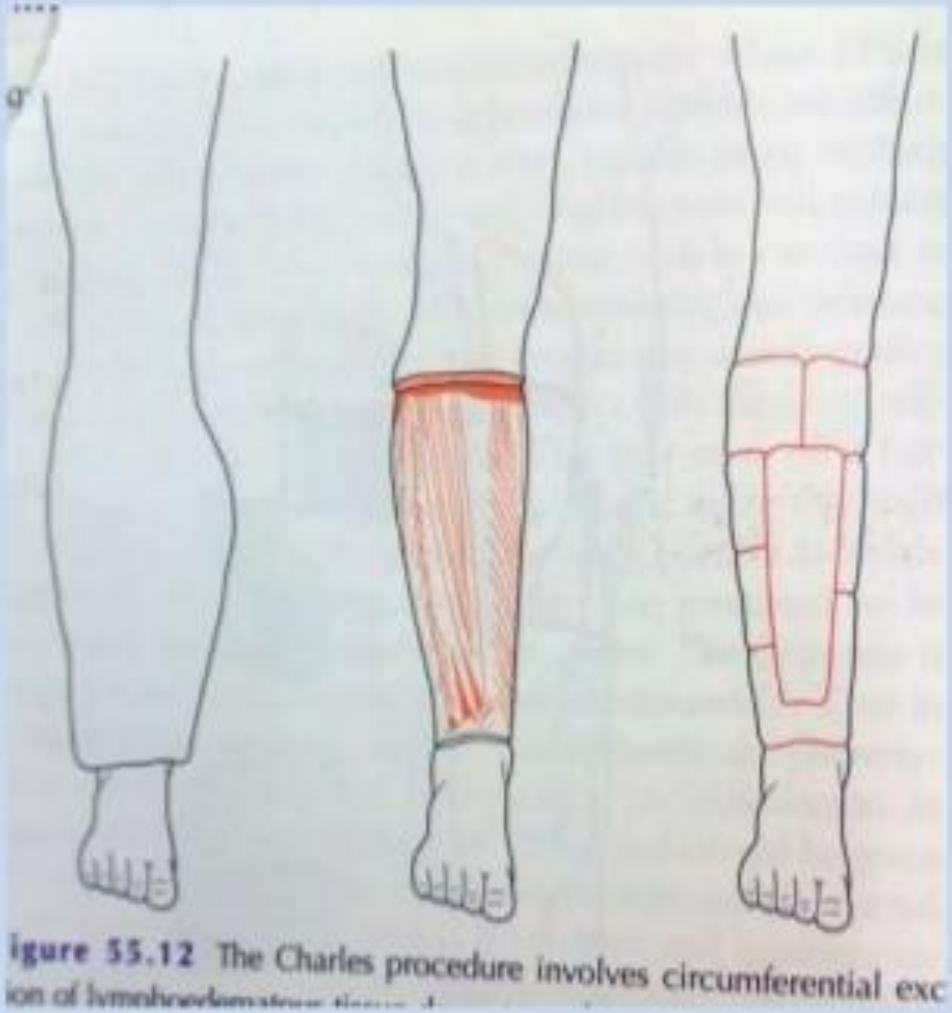


Figure 55.11 A cross-sectional representation of Thompson tion operation; the buried dermal flap.

Charles operation



igure 55.12 The Charles procedure involves circumferential exc

Thank you