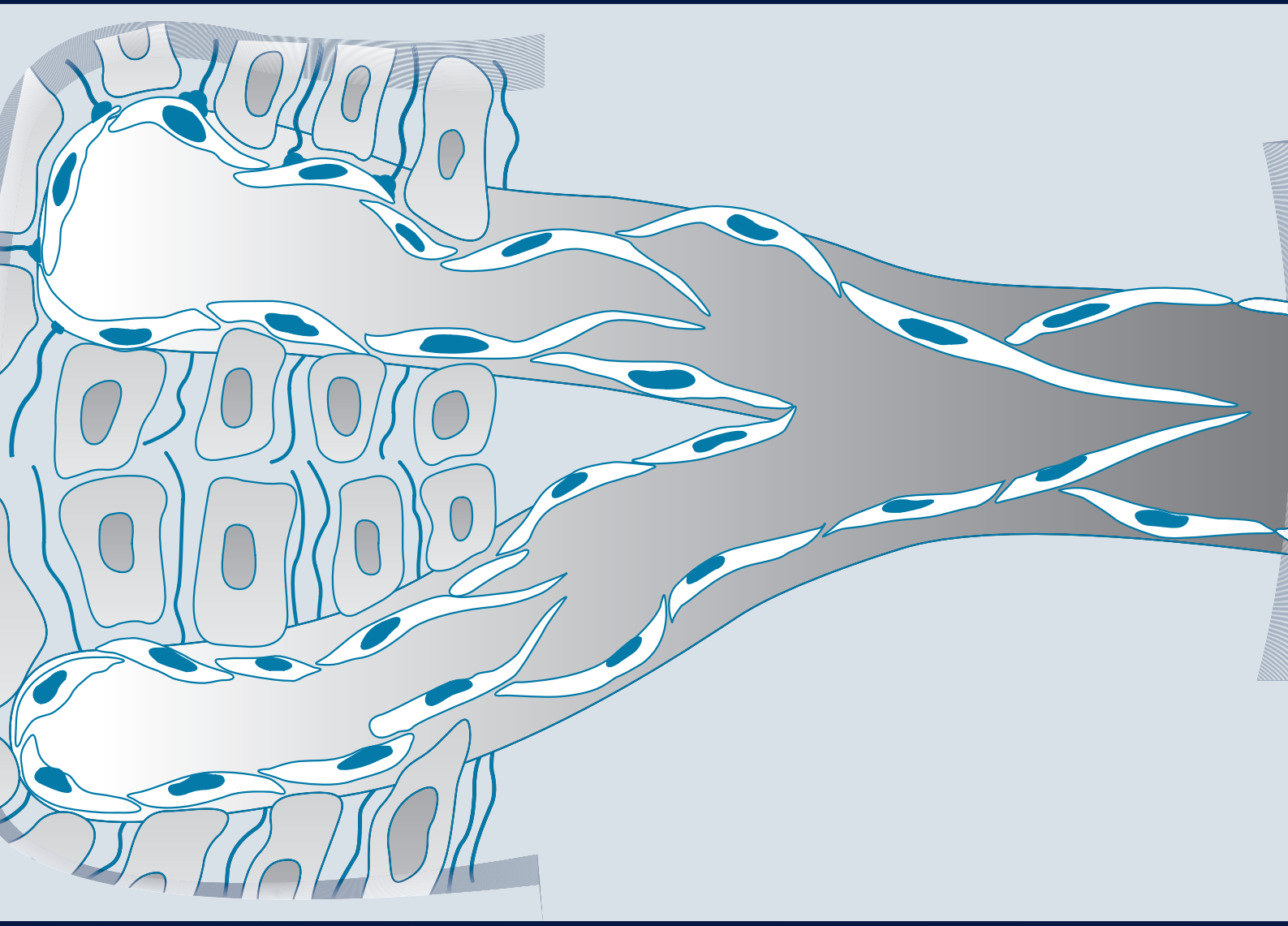
INTERNATIONAL CONSENSUS

BEST PRACTICE FOR THE MANAGEMENT OF LYMPHOEDEMA



LYMPHOEDEMA FRAMEWORK
AN INTERNATIONAL PERSPECTIVE

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THE LYMPHOEDEMA FRAMEWORK
The Lymphoedema Framework is a UK based research partnership launched in 2002 that aims to raise the profile of lymphoedema and improve standards of care through the involvement of specialist practitioners, clinicians, patient groups, healthcare organisations, and the wound care and compression garment industry.

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BEST PRACTICE FOR THE MANAGEMENT OF LYMPHOEDEMA

INTERNATIONAL CONSENSUS

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Introduction

Lymphoedema is a progressive chronic condition that affects a significant number of people and can have deleterious effects on patients' physical and psychosocial health. Even though it may be greatly ameliorated by appropriate management, many patients receive inadequate treatment, are unaware that treatment is available or do not know where to seek help. Several recent systematic reviews have highlighted the distinct lack of evidence for the optimal management of lymphoedema[1-3]. By presenting a model for best practice in lymphoedema in adults, this document aims to raise the profile of the condition and improve the care that patients receive.

ABOUT THIS DOCUMENT

The guidance provided here was derived from a UK national consensus on standards of practice for people who are at risk of or who have lymphoedema (Box 1). The consensus process (Box 2) was launched in 2002 and was driven by the Lymphoedema Framework with input from national patient support groups, patients with lymphoedema, national professional lymphoedema groups, clinical experts and industry (Appendix 1). Production of this document included review by an international panel of experts and endorsement by key national lymphoedema organisations.

The recommendations resulting from the consensus approach are included where relevant. Each recommendation has been classified according to the UK NHS Health Technology Assessment model for guideline development (Box 3)[4].

This document will be reviewed and updated after five years. Key references have been included; a complete list of the references used in the preparation of the text can be found at: www.lf.cricp.org.

BOX 2 Consensus process

The recommendations made in this document are the result of a highly rigorous, systematic process based on an explicit methodology of consultation and consensus[4] that involved:

- developing multidisciplinary, nationally representative groups
- identifying and critically appraising the best available information
- linking recommendations to supporting evidence.

BOX 3 Classification of recommendations[4]

- **A** Clear research evidence
- **B** Limited supporting research evidence
- **C** Experienced common sense judgement

ABBREVIATIONS

ABPI: ankle-brachial pressure index
IPC: intermittent pneumatic compression
MLD: manual lymphatic drainage
MLLB: multi-layer inelastic lymphoedema bandaging
SLD: simple lymphatic drainage (self massage)
TBPI: toe-brachial pressure index

BOX 1 Standards of practice for lymphoedema services, adapted from[5]

Standard 1: Identification of people at risk of or with lymphoedema
Systems to identify people at risk of or with lymphoedema, regardless of cause, will be implemented and monitored to ensure that patients receive high quality education and lifelong care.

Standard 2: Empowerment of people at risk of or with lymphoedema
Individual plans of care that foster self-management will be developed in partnership with patients at risk of or with lymphoedema (involving relatives and carers where appropriate), in an agreed format and language.

Standard 3: Provision of lymphoedema services that deliver high quality clinical care that is subject to continuous improvement and integrates community, hospital and hospice based services
All people at risk of or with lymphoedema will have access to trained healthcare professionals, including lymphoedema specialists, who will work to agreed standards for comprehensive ongoing assessment, planning, education, advice, treatment and monitoring. Care will be of a high standard and subject to continuous quality improvement.

Standard 4: Provision of high quality clinical care for people with cellulitis/erysipelas
Agreed protocols for the rapid and effective treatment of cellulitis/erysipelas, including prevention of recurrent episodes, will be implemented and monitored by healthcare professionals who have completed recognised training in this subject.

Standard 5: Provision of compression garments for people with lymphoedema
Agreed protocols for assessment for and the provision of compression garments for people with lymphoedema, or where warranted, those at risk of lymphoedema, will be implemented and monitored.

Standard 6: Provision of multi-agency health and social care
Following comprehensive assessment, any patient at risk of or with lymphoedema who requires multiagency support will have access to and receive care appropriate to their needs from health and social services.

WHAT IS LYMPHOEDEMA?

Lymphoedema may manifest as swelling of one or more limbs and may include the corresponding quadrant of the trunk. Swelling may also affect other areas, eg the head and neck, breast or genitalia. Lymphoedema is the result of accumulation of fluid and other elements (eg protein) in the tissue spaces due to an imbalance between interstitial fluid production and transport (usually low output failure)[6]. It arises from congenital malformation of the lymphatic system, or damage to lymphatic vessels and/or lymph nodes.

In patients with chronic lymphoedema, large amounts of subcutaneous adipose tissue may form. Although incompletely understood, this adipocyte proliferation may explain why conservative treatment may not completely reduce the swelling and return the affected area to its usual dimensions.

Lymphoedema may produce significant physical and psychological morbidity. Increased limb size can interfere with mobility and affect body image[7-10]. Pain and discomfort are frequent symptoms, and increased susceptibility to acute cellulitis/erysipelas can result in frequent hospitalisation and long-term dependency on antibiotics[11,12].

Lymphoedema is a chronic condition that is not curable at present, but may be alleviated by appropriate management; if ignored, it can progress and become difficult to manage.

At birth, about one person in 6000 will develop primary lymphoedema; the overall prevalence of lymphoedema/chronic oedema has been estimated as 0.13-2%[13-15]. In developed countries, the main cause of lymphoedema is widely assumed to be treatment for cancer. Indeed, prevalences of 12-60% have been reported in breast cancer patients[16-19] and of 28-47% in patients treated for gynaecological cancer[20,21]. However, it appears that about a quarter to a half of affected patients suffer from other forms of lymphoedema, eg primary lymphoedema and lymphoedema associated with poor venous function, trauma, limb dependency or cardiac disease[14,22].

LYMPHATIC FILARIASIS

Lymphatic filariasis is a parasitic infection transmitted by mosquitoes. In endemic areas, infection usually occurs in childhood. The parasites damage the lymphatic system, eventually causing lymphoedema. Although lymphatic filariasis is a major cause of lymphoedema worldwide, detailed information on its management in endemic areas is outside the scope of this document. For more information on the condition, see:

- Dreyer G, Addiss D, Dreyer P, Norões J. *Basic Lymphoedema Management.* Hollis, USA: Hollis Publishing Company, 2002.
- Dreyer G, Norões J, Figueredo-Silva J, Piessens WF. Pathogenesis of lymphatic disease in Bancroftian filariasis: a clinical perspective. *Parasitol Today* 2000; 16(12): 544-48.
- Vaqas B, Ryan TJ. Lymphoedema: pathophysiology and management in resource-poor settings - relevance for lymphatic filariasis control programmes. *Filaria J* 2003; 2(1): 4.
- Global Alliance to Eliminate Lymphatic Filariasis www.filariasis.org.

Identifying the patient at risk

C People at risk of lymphoedema should be identified early during routine assessment, monitored and taught self care.

Effective identification of patients at risk of lymphoedema relies on awareness of the causes of lymphoedema and associated risk factors, implementation of preventive strategies, and self monitoring. Patients, carers and healthcare professionals should be aware that there may be a considerable delay of several years from a causative event to the appearance of lymphoedema.

RISK FACTORS FOR LYMPHOEDEMA

The true risk factor profile for lymphoedema is not known. There may be many factors that predispose an individual to developing lymphoedema or that predict the progression, severity and outcome of the condition (Box 4). Further epidemiology is required to identify these factors, and research is needed to establish how risk factors themselves can be modified to reduce the likelihood or severity of consequent lymphoedema.

BOX 4 Risk factors for lymphoedema

Upper limb/trunk lymphoedema

- Surgery with axillary lymph node dissection, particularly if extensive breast or lymph node surgery
- Scar formation, fibrosis and radiodermatitis from postoperative axillary radiotherapy
- Radiotherapy to the breast, or to the axillary, internal mammary or subclavicular lymph nodes
- Drain/wound complications or infection
- **Cording (axillary web syndrome)**
- **Seroma** formation
- Advanced cancer
- Obesity
- Congenital predisposition
- Trauma in an 'at risk' arm (venepuncture, blood pressure measurement, injection)
- Chronic skin disorders and inflammation
- Hypertension
- Taxane chemotherapy
- Insertion of pacemaker
- Arteriovenous shunt for dialysis
- Living in or visiting a lymphatic filariasis endemic area

Lower limb lymphoedema

- Surgery with inguinal lymph node dissection
- Postoperative pelvic radiotherapy
- Recurrent soft tissue infection at the same site
- Obesity
- Varicose vein stripping and vein harvesting
- Genetic predisposition/family history of chronic oedema
- Advanced cancer
- Intrapelvic or intra-abdominal tumours that involve or directly compress lymphatic vessels
- Orthopaedic surgery
- Poor nutritional status
- Thrombophlebitis and chronic venous insufficiency, particularly post-thrombotic syndrome
- Any unresolved asymmetrical oedema
- Chronic skin disorders and inflammation
- Concurrent illnesses such as phlebitis, hyperthyroidism, kidney or cardiac disease
- Immobilisation and prolonged limb dependency
- Living in or visiting a lymphatic filariasis endemic area

Cording (axillary web syndrome): the appearance of tender, painful cord-like structures below the skin; may be due to inflammation or thrombosis of lymph vessels
Seroma: an accumulation of fluid at or near a surgical wound

Lymphadenitis: inflammation of the lymph nodes, which become swollen, tender and painful

TABLE 1 Classification of causes of secondary lymphoedema, adapted from[23]

Classification	Example(s)
Trauma and tissue damage	■ lymph node excision ■ radiotherapy ■ burns ■ varicose 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

Classification of lymphoedema

Lymphoedema is classified as primary or secondary depending on aetiology[23]. Primary lymphoedema is thought to be the result of a congenital abnormality of the lymph conducting system. Secondary or acquired lymphoedema (Table 1) results from damage to the lymphatic vessels and/or lymph nodes, or from functional deficiency. It may also be the result of high output failure of the lymphatic circulation, eg in chronic oedema due to venous insufficiency or post-thrombotic syndrome, when the function of the overloaded lymphatic system eventually deteriorates.

REDUCING RISK

The diverse aetiology of lymphoedema means that patients at risk of lymphoedema will be encountered in a wide variety of primary and secondary/tertiary care settings, eg cancer services, vascular surgery units, wound care/tissue viability services, dermatology services, plastic surgery units and services where patients receive symptom management for advanced cancer. To guarantee that patients at risk are recognised and their risk of lymphoedema is minimised, each setting should ensure that staff are aware of the potential risk factors for lymphoedema, the appropriate actions to take and relevant referral pathways (Figure 1). The setting should also offer structured patient education that follows an established methodology[24].

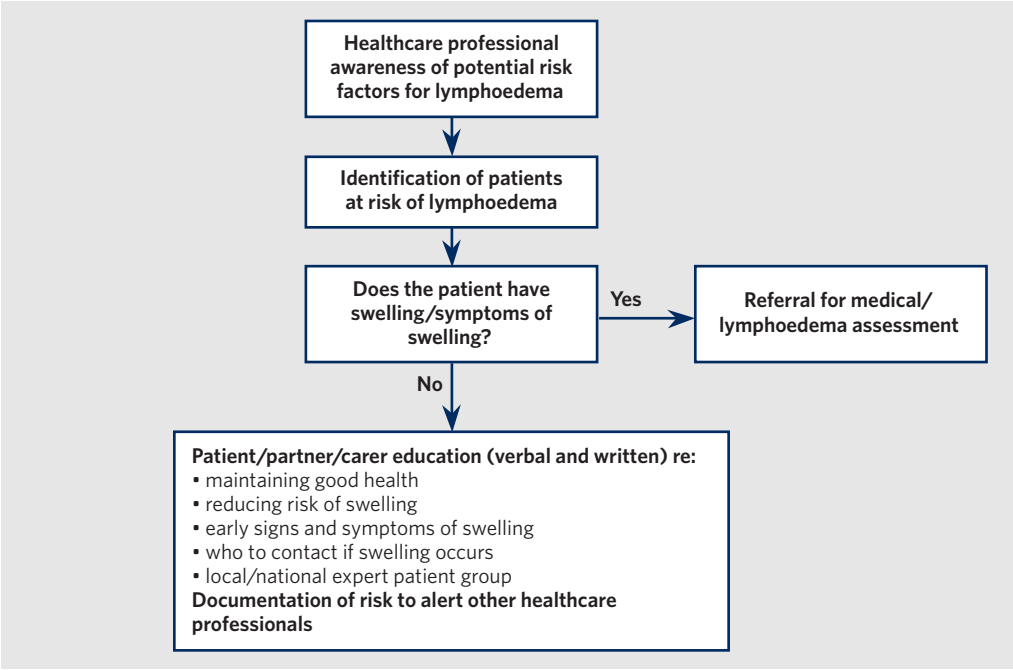


Figure 1 Management of patients at risk of lymphoedema

C Patients and carers should be offered information about lymphoedema and its management.

Patients at risk of developing lymphoedema and their partners/carers need to know what lymphoedema is, why the patient is at risk, how to maintain good health, how to minimise the risk of developing lymphoedema (Box 5), early symptoms and signs (Box 6), and who to contact if swelling develops.

A number of organisations disseminate information about lymphoedema (Box 7). Individual settings could use this information to devise patient education programmes, information leaflets and resources.

BOX 5 Common sense approach to minimising the risk of developing lymphoedema

- Take good care of skin and nails
- Maintain optimal body weight
- Eat a balanced diet
- Avoid injury to area at risk
- Avoid tight underwear, clothing, watches and jewellery
- Avoid exposure to extreme cold or heat
- Use high factor sunscreen and insect repellent
- Use mosquito nets in lymphatic filariasis endemic areas
- Wear prophylactic compression garments, if prescribed
- Undertake exercise/movement and limb elevation
- Wear comfortable, supportive shoes

NB While robust evidence is lacking that these actions reduce the risk of lymphoedema, they reflect a common sense approach. These actions may also help patients with existing lymphoedema to reduce the risk of deterioration.

BOX 6 Early signs and symptoms of lymphoedema

- Clothing or jewellery, eg sleeve, shoe or ring, becoming tighter
- Feeling of heaviness, tightness, fullness or stiffness
- Aching
- Observable swelling

BOX 7 Examples of organisations that supply information for patients

- British Lymphology Society www.lymphoedema.org/bls
- Dutch Lymphoedema Network www.lymfoedeem.nl
- Lymphoedema Association of Australia www.lymphoedema.org.au
- Lymphoedema Support Network (UK) www.lymphoedema.org/lsn
- Lymphovenous Canada www.lymphovenous-canada.ca
- National Lymphedema Network (USA) www.lymphnet.org

Assessment

An accurate assessment is essential for the appropriate treatment of lymphoedema.

Effective assessment of a patient at risk of or with possible lymphoedema will be comprehensive, structured and ongoing. Here, assessment has been divided into medical assessment and lymphoedema assessment, but the two may run in parallel within the same healthcare setting.

MEDICAL ASSESSMENT

The medical assessment is used to diagnose lymphoedema and to identify or exclude other causes of swelling (Box 8). In a primary care setting, this assessment is usually carried out by the general practitioner. If the patient presents to secondary/tertiary care, assessment may be by a medical specialist.

Most cases of lymphoedema are diagnosed on the basis of the medical history and physical examination. The choice of investigations used to elucidate the cause of the swelling (Box 9) will depend on the history, presentation and examination of the patient.

Specialist investigations

In secondary/tertiary settings, specialist investigations may be conducted including:

- ultrasound[25] – to assess tissue characteristics, eg for skin thickening and tissue fibrosis
- colour Doppler ultrasound[26] – to exclude deep vein thrombosis and evaluate venous abnormalities
- lymphoscintigraphy[27] (Figure 2) – to identify lymphatic insufficiency in patients where the cause of the swelling is unclear, to differentiate lipoedema and lymphoedema (Table 2, page 9), and to evaluate potential candidates for surgery

Chyluria: milky coloured urine due to reflux of chyle (the fat-bearing lymph that normally drains from the intestine to the thoracic duct) into the lymphatics of the urinary system

BOX 8 Differential diagnosis of lymphoedema

Unilateral limb swelling:
- acute deep vein thrombosis
- post-thrombotic syndrome
- arthritis
- Baker's cyst
- presence/recurrence of carcinoma*

Symmetrical swelling:
- congestive heart failure
- chronic venous insufficiency
- dependency or stasis oedema
- renal dysfunction
- hepatic dysfunction
- hypoproteinaemia
- hypothyroidism/myxoedema
- drug induced (eg calcium channel blockers, steroids, non-steroidal anti-inflammatories)
- lipoedema

NB These conditions may co-exist with or cause lymphoedema.
*Presence or recurrence of carcinoma requires direct referral to the appropriate oncology service.

BOX 9 Screening investigations

Blood tests:
- full blood count (FBC)
- urea and electrolytes (U&Es)
- thyroid function tests (TFTs)
- liver function tests (LFTs)
- plasma total protein and albumin
- fasting glucose
- erythrocyte sedimentation rate (ESR)/C-reactive protein (CRP)
- B-natriuretic peptide

Urine dipstick testing, including observation for **chyluria**
Ultrasound
Chest X-ray

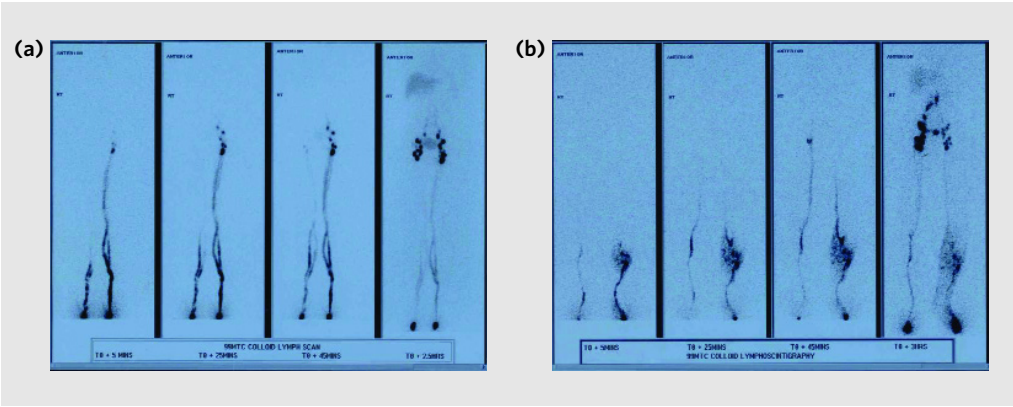


FIGURE 2 Lymphoscintigraphy
Radiolabelled colloid or protein is injected into the first web space of each foot or hand, and is tracked as it moves along the lymphatics by a gamma camera. (a) Normal lower limb images with fast lymph drainage in left leg because of associated venous disease. (b) Normal right leg with disturbances to lymph drainage in left leg from past cellulitis/erysipelas.

- micro-lymphangiography using fluorescein labelled human albumin[28] – to assess dermal lymph capillaries
- indirect lymphography using water soluble contrast media[29] – to opacify initial lymphatics and peripheral lymph-collectors and to differentiate lipoedema and lymphoedema
- CT/MRI scan[30] – to detect thickening of the skin and the characteristic honeycomb pattern produced by lymphoedema, to detect lymphatic obstruction by a tumour at the root of a limb or in the pelvis or abdomen, and to differentiate lipoedema and lymphoedema
- bioimpedance[31] – to detect oedema and monitor the outcome of treatment
- filarial antigen card test – to detect infection with *Wuchereria bancrofti* by testing for antibodies to the parasite in a person who has visited or is living in a lymphatic filariasis endemic area.

Primary lymphoedema is usually diagnosed after exclusion of secondary lymphoedema. Genetic screening and counselling may be required if there is a suspected familial link. Three gene mutations have been linked with primary lymphoedema:

- FOXC2 – **lymphoedema-distichiasis syndrome**
- VEGFR-3 – **Milroy's disease**
- SOX18 – **hypotrichosis-lymphoedema-telangiectasia syndrome.**

LYMPHOEDEMA ASSESSMENT

A lymphoedema assessment should be performed at the time of diagnosis and repeated periodically throughout treatment. The findings of the assessment should be recorded systematically (Box 10, page 8) and form the baseline from which management is planned, further referral made and progress monitored. Specialist computer programs can assist in standardising assessment (eg LymCalc; details can be found at: www.colibri.demon.co.uk).

Lymphoedema assessment is usually carried out by a practitioner who has undergone training at specialist level.

Lymphoedema staging

Several staging systems for lymphoedema have been devised, including the International Society of Lymphology system (Box 11). None has achieved international agreement and each has its limitations.

BOX 11 International Society of Lymphology (ISL) lymphoedema staging[6]

ISL stage 0
A subclinical state where swelling is not evident despite impaired lymph transport. This stage may exist for months or years before oedema becomes evident

ISL stage I
This represents early onset of the condition where there is accumulation of tissue fluid that subsides with limb elevation. The oedema may be pitting at this stage

ISL stage II
Limb elevation alone rarely reduces swelling and pitting is manifest

ISL late stage II
There may or may not be pitting as tissue fibrosis is more evident

ISL stage III
The tissue is hard (fibrotic) and pitting is absent. Skin changes such as thickening, hyperpigmentation, increased skin folds, fat deposits and warty overgrowths develop

Lymphoedema-distichiasis syndrome: a form of primary lymphoedema with onset at or after puberty in which the patient has accessory eyelashes along the posterior border of the eyelids. Has a clear family history
Milroy's disease: a form of primary lymphoedema that is present at birth, only affects the lower limbs and has a clear family history
Hypotrichosis-lymphoedema-telangiectasia syndrome: a form of primary lymphoedema associated with sparse or absent hair and telangiectasia (localised collections of distended blood capillary vessels observed in the skin as red spots)

BOX 10 Lymphoedema assessment proforma

Lymphoedema Assessment Form **Assessor:** **Date:**

Name: Male/female DOB: Tel:
Address:

Patient number: Next of kin:
Referred by: Primary care physician:

Diagnosis
Primary/secondary lymphoedema/lipoedema
Onset of oedema (age/symptoms):
Investigations:
Current symptoms:
Current/previous cellulitis:
Current treatment for lymphoedema:
Past treatment for lymphoedema:

Past medical history

Surgery:
Cancer status:
Axillary clearance/ sentinel node biopsy:
No. nodes removed:
No. nodes +ve:
Radiotherapy:
Chemotherapy:
Hormonal therapy:
Venous/arterial disease:
Neurological disease:
Family history:

Current medication

Allergies:

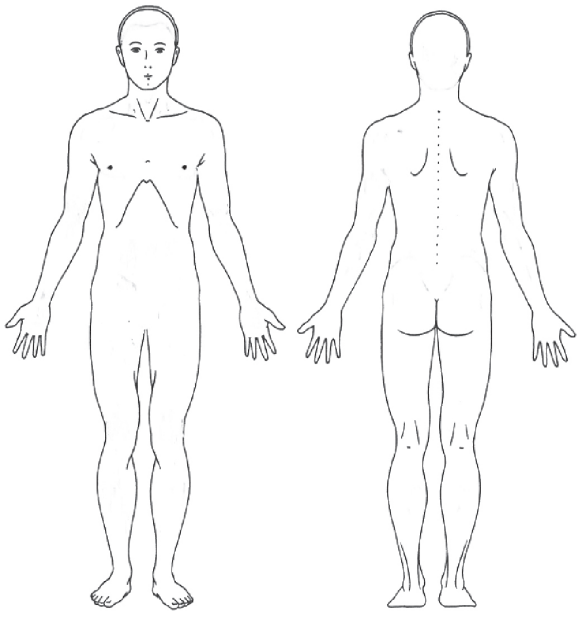
Psychosocial/functional status
Emotional state:
Social support:
Employment:
Mobility:
Activities of daily living:

Nutritional assessment
Weight (kg):
Height (m):
BMI:
Waist circumference (cm):

Current location of swelling

√ Swelling Pitting Tissue thickening

Dominant side: upper limb R/L; lower limb R/L
Skin condition:
Tissues in swollen area are predominantly: soft/firm
Swelling is predominantly: pitting/nonpitting
Sensory changes:



Stemmer sign: Hand: R +/- L +/-
Foot: R +/- L+/-

ABPI/TBPI: R leg
L leg

Pain assessment
Present?
Site/character/pain score:
Current treatment:

Limb circumference measurements

	Upper limb		Lower limb	
	R	L	R	L
Hand/foot circumference (cm)				
Starting point (cm)				
Above elbow/knee (cm)				
Below elbow/knee (cm)				
Total limb volume (ml)				
Distal volume (ml)				
Proximal volume (ml)				
Distal:proximal ratio				
Excess total limb volume (ml and %)				
Excess distal limb volume (ml and %)				
Excess proximal limb volume (ml and %)				

TABLE 2 Differentiating lymphoedema and lipoedema

	Lymphoedema	Lipoedema*
Signs and symptoms	Can involve the legs, arms, trunk, genitalia or head and neck Swelling of limbs affects hands and feet Affects either sex Stemmer sign may be positive; usually not painful on pinching	Usually causes symmetrical bilateral swelling of the lower limbs; can occur in arms Swelling stops at ankles and wrists Pain and bruising are prominent features Affects mainly women In pure lipoedema, Stemmer sign is negative; often painful on pinching
Aetiology	Results from inadequate lymphatic drainage May be congenital or result from insults to the lymphatic system Not usually associated with hormonal imbalances	Unknown; results in excessive subcutaneous fat deposition Appears to be oestrogen requiring and starts at time of hormonal change eg pregnancy, puberty Family history of lipoedema often positive
Lymphoscintigraphy	Identifies disordered lymphatics	Often indicates normal lymphatic functioning
MRI scan	Honeycomb pattern in the subcutis and thickened skin	Subcutaneous fat, but no fluid

*Lipoedema can progress to develop an oedematous component - lipolymphoedema.

BOX 12 Severity of unilateral limb lymphoedema[6]

Mild:
<20% excess limb volume

Moderate:
20-40% excess limb volume

Severe:
>40% excess limb volume

Classification of severity

One method of establishing the severity of unilateral limb lymphoedema is based on the difference in the limb volume of the affected and unaffected limbs (Box 12).

There is currently no formal system for the classification of the severity of bilateral limb swelling or lymphoedema of the head and neck, genitalia or trunk.

The severity of lymphoedema can also be based on the physical and psychosocial impact of the condition. Factors to consider include:

- tissue swelling – mild, moderate or severe; pitting or nonpitting
- skin condition – thickened, warty, bumpy, blistered, lymphorrhoeic, broken or ulcerated
- subcutaneous tissue changes – fatty/rubbery, nonpitting or hard
- shape change – normal or distorted
- frequency of cellulitis/erysipelas
- associated complications of internal organs, eg pleural fluid, **chylous ascites**
- movement and function – impairment of limb or general function
- psychosocial morbidity.

A more detailed and comprehensive classification applicable to primary and secondary lymphoedema remains to be formulated.

Chylous ascites: the accumulation of chyle (fat-bearing lymph) in the abdominal cavity
Lymphadenopathy: enlargement of the lymph nodes

TESTING FOR PITTING

Pitting indicates the presence of excess interstitial fluid, ie tissue oedema. Pitting is usually tested for by pressing firmly, but without hurting the patient, on the area to be examined with a finger or thumb for a count of at least 10 seconds. If an indentation remains when the examiner ceases pressing, pitting is present. The depth of the indentation reflects the severity of the oedema. In a research setting, the pitting test may be defined in terms of the pressure applied and the length of application, and measurement of the depth of any resulting indentation.

Assessment of swelling

The duration, location and extent of the swelling and any pitting should be recorded, along with the location of any **lymphadenopathy**, the quality of the skin and subcutaneous tissue, and the degree of shape distortion. Limb circumference and volume should be measured.

Limb volume measurement

Limb volume measurement is one of the methods used to determine the severity of the lymphoedema, the appropriate management, and the effectiveness of treatment. Typically, limb volume is measured on diagnosis, after two weeks of intensive therapy with multi-layer inelastic lymphoedema bandaging (MLLB), and at follow-up assessment.

In unilateral limb swelling, both the affected and unaffected limbs are measured. The difference in limb volume is expressed in millilitres (ml) or as a percentage.

Oedema is considered present if the volume of the swollen limb is more than 10% greater than that of the contralateral unaffected limb. The dominant limb should be noted: in unaffected patients, the dominant limb can have a circumference up to 2cm greater and a volume as much as 8-9% higher than the nondominant limb[32,33].

In bilateral limb oedema, the volume of both limbs can be measured and used to track treatment progress.

There is no effective method for measuring oedema affecting the head and neck, breast, trunk or genitalia. Digital photography is recommended as an appropriate means to subjectively record and monitor facial and genital lymphoedema[34].

Water displacement method

The water displacement method (also known as water plethysmography) is considered the 'gold standard' for calculating limb volume and is the only reliable method available for the measurement of oedematous hands and feet[35]. It uses the principle that an object will displace its own volume of water. However, practicalities, such as hygiene issues and accessing this method, limit its use.

Circumferential limb measurements

Calculation of volume from circumferential measurements is the most widely used method. It is easily accessible and its reliability can be improved if a standard protocol is followed.

Circumferential measurements of limbs (Figure 3) are put into a specialist computer program or calculator for determination of individual limb volume and excess limb volume. Some practitioners have set up standard spreadsheet programs to calculate volume.

A simplified method for patients with palliative care needs is shown in Figure 4 (page 12). These measurements are not used to calculate limb volume, but to track sequential changes in circumference.

Perometry

Perometry uses infrared light beams to measure the outline of the limb. From these measurements, limb volume (but not hand or foot volume) can be calculated quickly, accurately and reproducibly[36]. Although the use of perometry is becoming more widespread, the cost of the machine limits it to specialist centres.

Bioimpedance

Bioimpedance measures tissue resistance to an electrical current to determine extracellular fluid volume. The technique is not yet established in routine practice. However, it may prove useful in demonstrating early lymphoedema, identifying lipoedema, and in monitoring the outcome of treatment[31]. The technique is currently of limited use in bilateral swelling.

Limitations of excess limb volume

Calculation of excess limb volume is of limited use in bilateral lymphoedema. In such cases measurements can be used to track sequential changes in limb circumference to indicate treatment progress. In patients with extensive **hyperkeratosis**, **elephantiasis** or tissue thickening it should be recognised that a proportion of the excess volume will be due to factors other than fluid accumulation.

Hyperkeratosis: thickening of the outer layer of the skin
Elephantiasis: severe lymphoedema characterised by severe swelling, hard thickened tissue, deep skin folds and skin changes such as hyperkeratosis and warty growths

Figure 3 Method for obtaining measurements for calculating limb volume
A pretensioned tape measure should be used. No tension should be applied to the tape during measuring.

Upper limbs

- Ask the patient to sit with the arm supported on a table with the hand palm down
- On the ulnar aspect of the arm* measure with a ruler and record the distance from the nail bed of the little finger to 2cm above the ulnar styloid (wrist)†. Mark this point on the patient. This determines the starting point
- Mark the same point on the contralateral arm
- Lie a ruler along the ulnar aspect of the arm and mark the limb at 4cm intervals from the starting point to 2cm below the axilla
- With the limb in a relaxed position, measure the circumference at each mark, placing the top edge of the tape measure just below the mark
- Note measurements above the elbow in the correct section of the paper or electronic recording form
- Repeat the process on the other limb. Ensure there are the same number of measurements for both arms
- Document the position the patient was in when measurements were taken

Lower limbs

- Ask the patient to stand or sit with both feet firmly on the ground
- On the medial aspect of the leg* measure with a ruler and record the distance from the floor to 2cm above the middle of the medial malleolus†. Mark this point on the patient. This determines the starting point
- Mark the same point on the contralateral leg
- Seat patient on a chair with bottom as close to the edge as possible, or seat on a couch with the leg straight
- Lie a ruler along the medial aspect of the leg and mark the limb at 4cm intervals from the starting point to 2cm below the popliteal fossa for swelling below the knee
- If swelling extends above the knee, ask the patient to stand or to lie on a couch. Continue the marks at 4cm intervals above the knee to 2cm below the gluteal crease
- With the limb in a relaxed position, measure the circumference at each mark, placing the top edge of the tape measure just below the mark
- Note measurements above the knee in the correct section of the paper or electronic recording form
- Repeat the process on the other limb. Ensure there are the same number of measurements for both legs
- Document the position the patient was in when measurements were taken

*If only one limb is affected, start with the unaffected side.
†If the ulnar styloid or medial malleolus cannot be located, alternative fixed anatomical points can be used to determine the starting point, eg olecranon process or anterior iliac spine. The distance from the fixed anatomical point to the starting point should be recorded to ensure consistency when measurements are repeated subsequently.

MEASURING LIMBS FOR VOLUME OR COMPRESSION GARMENTS

Figures 3 and 4 illustrate methods for measuring limbs to assess limb volume and swelling. These methods differ from the techniques used to measure for compression garments, which are shown on pages 41 and 42.

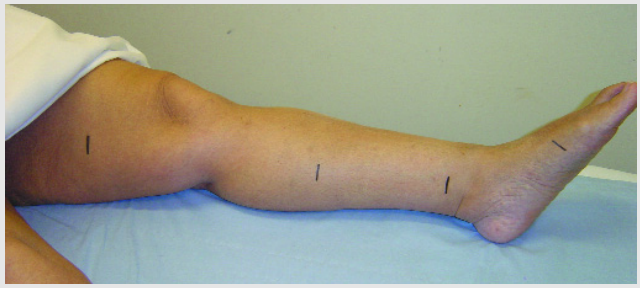
FIGURE 4 Simplified measuring method for patients with palliative care needs
A pretensioned tape measure should be used. No tension should be applied to the tape during measuring.

Upper limbs
With the limb in a supported position and the arm straight, measure circumference:
- around the dorsum of the hand (if oedematous)
- around the wrist
- 10cm below the olecranon process
- 10cm above the olecranon process

Repeat with the other arm

Lower limbs
With the limb in a relaxed position, measure circumference:
- of the foot (if oedematous)
- 2cm above the medial malleolus
- 10cm above the superior pole of the patella
- 10cm below the inferior pole of the patella

Repeat with the other leg



Lymphangiectasia: dilatation of lymph vessels; may appear as blister-like protuberances on the skin
Lymphorrhoea: leakage of lymph from the skin surface
Papillomatosis: the development of warty growths on the skin consisting of dilated lymphatics and fibrous tissue
Lipodermatosclerosis: thickening and hardening of the subcutaneous tissues of the lower leg with brown discolouration of the skin; associated with chronic venous insufficiency; in severe cases lymphatics become damaged

Assessment of skin condition

The general condition of the patient's skin and that of the affected area should be assessed for:
- dryness
- pigmentation
- fragility
- redness/pallor/cyanosis
- warmth/coolness
- dermatitis
- cellulitis/erysipelas
- fungal infection
- hyperkeratosis
- **lymphangiectasia**
- **lymphorrhoea**
- **papillomatosis**
- scars, wounds and ulcers
- **lipodermatosclerosis**
- orange peel skin (peau d'orange)
- deepened skin folds
- Stemmer sign (Figure 5).

Examples of some of the skin changes seen in lymphoedema can be found on pages 24-27, along with the indications for referral of patients to dermatology or other specialist services.

Vascular assessment

The arterial vascular status of the legs of all patients with lower limb lymphoedema should be assessed. The presence of peripheral arterial occlusive disease may contraindicate compression therapy or necessitate a reduction in the level of compression used.

Ankle-brachial pressure index (ABPI) provides an objective measure of the patency of the large arteries supplying blood to the foot. It is calculated from the ratio of the highest ankle systolic pressure for each limb to the highest systolic pressure in the arm. There are limitations to the test particularly in the presence of lymphoedema. Tissue thickening, hyperkeratosis or oedema may make it difficult to detect blood flow using the standard 8MHz probe. The use of a 4MHz probe and a larger size blood pressure cuff may overcome these problems[37].

An ABPI of 1.0-1.3 is normal; an ABPI of <0.8 indicates a degree of lower limb arterial occlusive disease that precludes the use of high compression. Inability to obliterate the pulse signal during measurement or an ABPI>1.3 also indicates vascular disease.

Measurement of toe-brachial pressure index (TBPI) may be useful when obtaining an ABPI is not possible or too painful[38]. Alternatives for assessing vascular status include pulse oximetry and pulse oscillography of the limbs, but may be subject to false-positive ischaemic results in the presence of oedema.

If there is any doubt about the patient's peripheral arterial status, a vascular opinion should be sought.

Use of these vascular assessment methods requires appropriate training in measurement technique and interpretation of results.

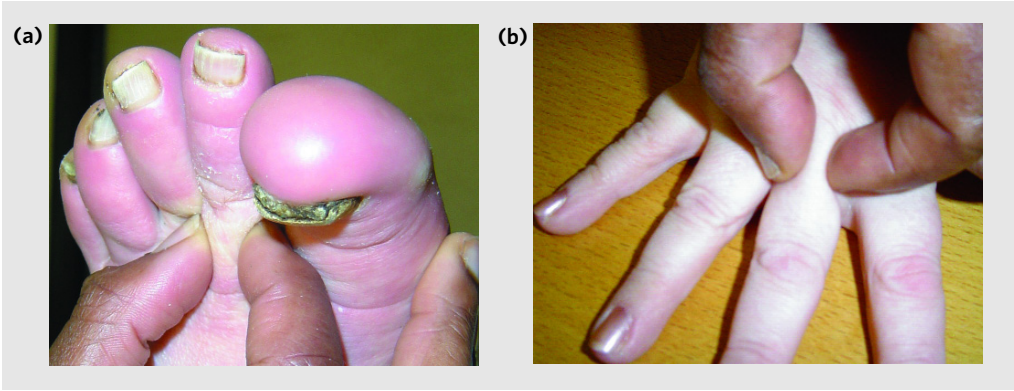


FIGURE 5 Stemmer sign
In a healthy person, a fold of skin can be pinched and lifted up at the base of the (a) second toe or (b) middle finger. The Stemmer sign is present and indicative of lymphoedema when a skin fold cannot be raised.

NB A negative sign may occur in proximal descending lymphoedema and does not exclude lymphoedema.

Pain assessment

Pain has been reported to affect 50% of patients with lymphoedema, with most taking regular analgesia[14]. Pain may be caused by:

- inflammation
- tissue distension
- infection
- ischaemia
- lipoedema
- nerve entrapment or neuropathy
- complex regional pain syndrome
- factitious swelling
- radiation-induced fibrosis
- cancer recurrence/progression
- taxane chemotherapy
- degenerative joint disease.

Effective assessment of pain requires noting the cause, nature, frequency, timing, site, severity and impact of the pain. Effective management strategies are dependent on the understanding that there are layers of pain in lymphoedema, eg:

- procedural pain – resulting from the treatment of lymphoedema
- incident pain – breakthrough pain caused by day to day activities
- background pain – intermittent or continuous pain at rest.

Any of these can be influenced by environmental factors or psychosocial factors that affect patient experience and ability to communicate pain[39].

Local pain management clinics and palliative care teams can provide help in the consistent and regular use of formal pain assessment tools and the management of pain.

Nutritional assessment

B **Patients with lymphoedema should be encouraged to maintain a healthy body weight.**

Nutritional assessment has two components: determining obesity and assessing the patient's diet.

As yet, the role of diet in lymphoedema is not established. However, lymphoedema is associated with obesity and obesity is a risk factor for the development of lymphoedema after treatment for breast cancer[40,41]. The frequent co-existence of obesity and lymphoedema suggests that obesity may contribute to the development of lymphoedema, possibly by reducing mobility[42].

Body mass index (BMI), calculated from the patient's weight and height, may be used to determine obesity. Overweight patients should be encouraged to reduce their BMI to <25; patients with BMI ≥30 should be offered dietary treatment or advice[43].

Waist measurement and waist-to-hip ratio provide an indication of total body fat and are simple methods for the assessment of obesity. A waist-to-hip ratio of >0.80 for women and >0.90 for men is associated with increased health risk[44]. A reduction in waist circumference, indicating decreased central body fat, with no overall weight change may result in a significant reduction in health risk (Table 3).

TABLE 3 The relationship between waist circumference and health risk[45,46]

	Waist circumference	Health risk
Women	80-87cm (32-34in)	Increased
	88cm (35in) or greater	Substantially increased
Men	94-101cm (37-39in)	Increased
	102cm (40in) or greater	Substantially increased

Psychosocial assessment

C **Patients with lymphoedema should receive psychological screening to identify those who require help to cope with the condition and those who require specialist psychological intervention.**

Lymphoedema can result in functional impairment, reduced self esteem, distorted body image, depression, anxiety, and problems with sexual, family and social relationships[7,10,47]. Psychosocial assessment will highlight areas that require referral for specialist intervention and factors that may have an impact on management and concordance with treatment.

Psychological evaluation should include asking the patient how their swelling makes them feel about themselves alongside assessment for:

- depression – eg low mood, loss of interest, low energy, changes in weight, appetite or sleep patterns, poor concentration, feelings of guilt or worthlessness, suicidal thoughts (Box 13)
- anxiety – eg apprehension, panic attacks, irritability, poor sleeping, situation avoidance, poor concentration
- cognitive impairment – may contribute to lack of motivation and inability to be independent
- lack of motivation
- ability to cope
- understanding of disease and concordance with treatment.

BOX 13 Screening for depression[48]

NICE recommends that screening for depression should include the use of at least two questions concerning mood and interest, eg:

- During the last month, have you often been bothered by feeling down, depressed or hopeless?
- During the last month, have you often been bothered by having little interest or pleasure in doing things?

BOX 14 Functional assessment of limbs affected by lymphoedema

Arm:

- range of joint movement
- ability to use fastenings, eg buttons, bra fastenings
- ability to put on or remove underwear/compression garments or bandaging
- hand grip and pincer movement
- effect of lymphoedema on activities of daily living
- use of any aids

Leg:

- range of joint movement
- ability to get up from sitting or lying
- ability to walk; gait analysis
- ability to lift individual legs
- posture when sitting and standing
- ability to put on and take off footwear/compression garments or bandaging
- suitability of footwear
- effect of lymphoedema on activities of daily living
- use of any aids

Social factors that should be assessed include:

- accommodation – accessibility, general living standards, heating/cooling
- support – involvement of carers, effect of lymphoedema on personal relationships, social isolation
- employment – ability to work, effect of work on lymphoedema
- education – ability to attend educational establishment and study
- financial status – benefit entitlement, medical insurance
- recreational activities, exercise, sport.

Mobility and functional assessment

Assessment of a patient's mobility and functional status (Box 14) will contribute to the formulation of a management plan and determine whether referral for further assessment is necessary. Functional assessment of lymphoedema affecting the head, neck, trunk or genitalia should be undertaken by a lymphoedema specialist.

The World Health Organization has produced a standardised, cross-cultural, non-disease specific tool for functional assessment – the WHO Disability Assessment Scale, available at: www.who.int/icidh/whodas.

Patients with functional, joint or mobility problems should be referred as appropriate for physiotherapy and/or occupational therapy assessment.

Treatment decisions

B **Patients with lymphoedema should receive a coordinated package of care and information appropriate to their needs.**

B **Patients and carers should have early active involvement in the management of lymphoedema.**

The best practice management of lymphoedema has a holistic, multidisciplinary approach that includes:

- exercise/movement – to enhance lymphatic and venous flow
- swelling reduction and maintenance – to reduce limb size/volume and improve subcutaneous tissue consistency through compression and/or massage, and to maintain improvements
- skin care – to optimise the condition of the skin, treat any complications caused by lymphoedema and minimise the risk of cellulitis/erysipelas
- risk reduction – to avoid factors that may exacerbate lymphoedema
- pain and psychosocial management.

Swelling reduction is achieved through a combination of compression (eg MLLB and/or compression garments) and exercise/movement with or without lymphatic massage (manual lymphatic drainage – MLD, simple lymphatic drainage – SLD or intermittent pneumatic compression – IPC).

The precise form of management programme required will be determined by the site, stage, severity and complexity of the lymphoedema, and the patient's psychosocial situation (Figure 6). Patients may require referral to a lymphoedema service (Box 15), or for assessment of co-existing medical, functional or psychosocial problems. Successful management of lymphoedema relies on patients and carers playing an active role.

BOX 15 Indications for referral to a lymphoedema service

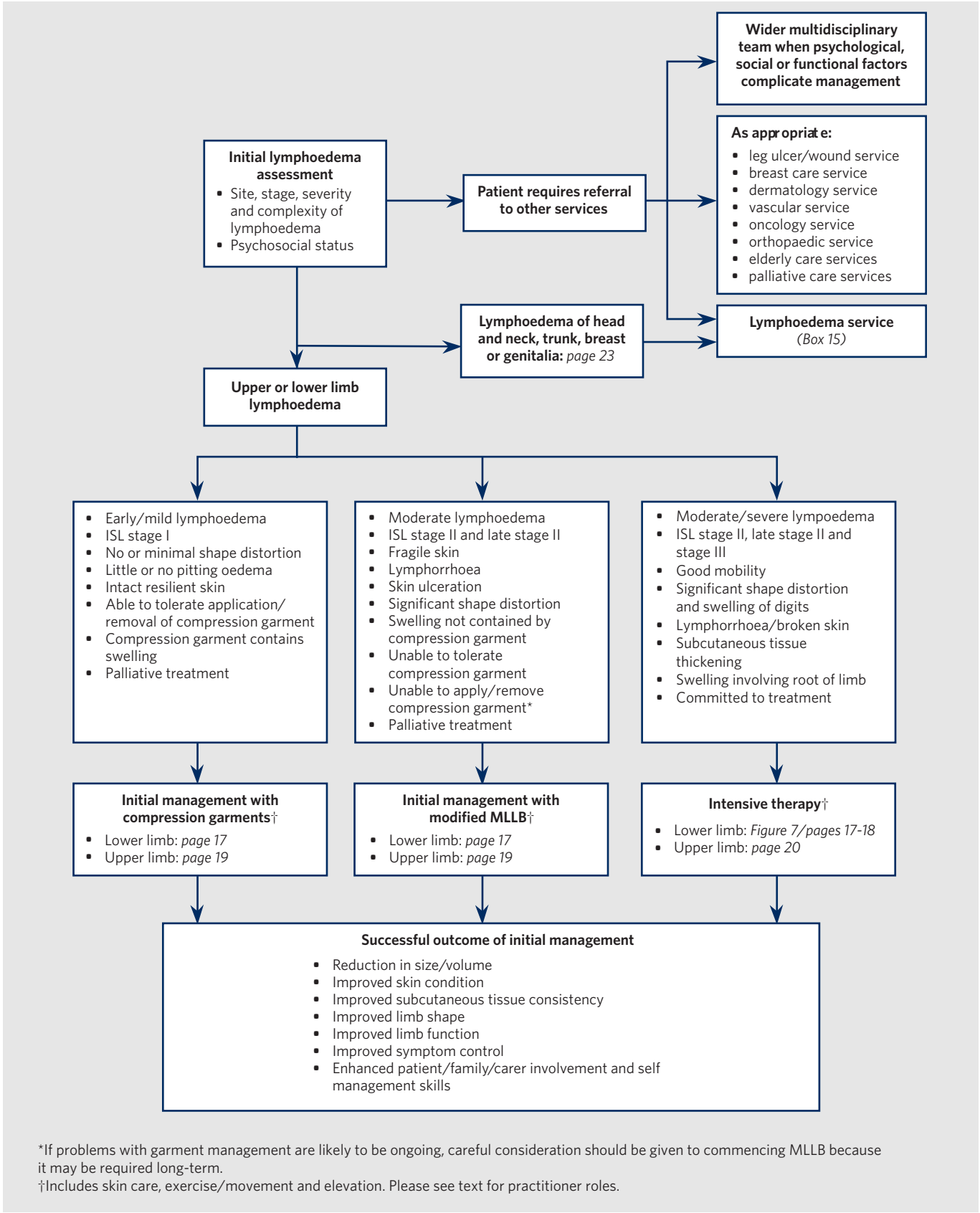
Special groups:
- swelling of unknown origin
- midline lymphoedema (head, neck, trunk, breast genitalia)
- children with chronic oedema
- primary lymphoedema
- lymphoedema in family members

Factors complicating management:
- concomitant arterial disease
- concomitant diabetes mellitus
- concomitant venous insufficiency with ulceration
- long-term complications due to surgery or radiotherapy
- severe papillomatosis, hyperkeratosis or other chronic skin condition
- severe foot distortion/ bulbous toes
- sudden increase in pain or swelling of lymphoedematous site
- chylous reflux, eg chyluria, **chyle**-filled lymphangiectasia
- neuropathy
- functional, social or psychological factors
- obesity

Management difficulties:
- compression garment fitting problems
- failure to respond after three months' standard treatment
- wound that deteriorates or is unresponsive after three months' treatment
- recurrent cellulitis/erysipelas

Chyle: the milk-coloured, fat-bearing lymph that usually drains from the intestine into the thoracic duct

FIGURE 6 Initial management of lymphoedema
This algorithm guides the practitioner in choosing the appropriate form of management for the patient, and indicates where in this document to find further information.



*If problems with garment management are likely to be ongoing, careful consideration should be given to commencing MLLB because it may be required long-term.
†Includes skin care, exercise/movement and elevation. Please see text for practitioner roles.

LOWER LIMB LYMPHOEDEMA – INITIAL MANAGEMENT

Initial management of lower limb lymphoedema will involve psychosocial support, education, skin care, exercise/movement, elevation and management of any pain or discomfort (Figure 6). The patient's initial management may also include:

- compression hosiery
- modified MLLB
- intensive therapy.

Compression hosiery

Patients with mild lower limb lymphoedema (ISL stage I), minor pitting, no significant tissue changes, no or minimal shape distortion, or palliative needs may be suitable for initial management with compression hosiery. The pressure used should be guided by the patient's vascular status and their ability to tolerate compression and manage the garment (pages 39-45). Skin care, exercise/movement, elevation and SLD should be taught alongside self monitoring and proper application, removal and care of hosiery. Patients' application/removal technique should be monitored and assessed.

Patients should be reviewed four to six weeks after initial fitting, and then after three to six months if response is satisfactory. The patient should be reviewed at each garment renewal, ie approximately every three to six months.

The practitioner will be appropriately trained.

Modified MLLB

Patients with ISL stage II or late stage II lower limb lymphoedema may be candidates for initial treatment with modified MLLB, outside an intensive therapy regimen. Modified MLLB may also be useful in controlling symptoms in patients with cancer-related lymphoedema and frail patients who have complex medical problems (page 34). Management should include skin care, exercise/movement, elevation, SLD and psychosocial support.

The practitioner will be appropriately trained.

Intensive therapy

Intensive therapy reduces swelling by decongesting impaired lymphatic pathways, reducing lymphatic load, encouraging the development of collateral drainage routes, and stimulating the function of remaining patent routes.

Intensive therapy is used in patients with ISL stage II, late stage II and stage III lower limb lymphoedema. Intensive therapy regimens use a combination of skin care, MLLB, exercise/movement and elevation. The regimen may include MLD or MLD with IPC.

The frequency of treatment, degree of compression and type of bandaging used should be adapted according to the patient's physical and psychosocial needs, and to the presence of venous ulceration and arterial or venous insufficiency (Figure 7, page 19).

Intensive therapy programmes are likely to be undertaken for a period of two to four weeks, although a maximal effect may be achieved more quickly in some patients. During this time treatment should be evaluated continuously and appropriate alterations made according to patient need and the effectiveness of the selected regimen. Appropriate training is required for all practitioners who deliver intensive therapy programmes[49].

Standard intensive therapy (>45mmHg)

This involves skin care, exercise/movement, elevation, MLD, and MLLB with inelastic bandages (sub-bandage pressure >45mmHg) undertaken daily.

Patients undergoing standard intensive therapy must be carefully selected and be willing and able to commit physically and emotionally to daily intensive therapy, including participation in exercise programmes.

The practitioner will be appropriately trained at specialist level.

INTENSIVE THERAPY

The combination of skin care, exercise, MLD and MLLB is often known as decongestive lymphatic therapy (DLT) or complete decongestive therapy (CDT). The term intensive therapy has been used in this document to denote a holistic approach that includes education, psychosocial support and pain management, and that may also include SLD and IPC.

MODIFYING MLLB

Where necessary, MLLB may be modified and individualised, according to patient need and resources available, by altering the:

- compression produced
- frequency of bandage change
- materials used.

SAFETY ISSUES

Lower limb peripheral arterial occlusive disease
Patients with lower limb lymphoedema with reduced ABPI of 0.5-0.8 should not receive sustained compression exceeding 25mmHg[50]. Patients with ABPI <0.5 should not receive compression. If arterial involvement is suspected, referral to a vascular specialist should be made before introducing compression.

Cellulitis/erysipelas
During periods of acute infection, the level of compression should be reduced or removed if too painful, medical supervision may be required, and any form of lymphatic massage should be discontinued. The usual type and level of compression should be recommenced when the acute phase of the infection has resolved and the patient is able to tolerate it again. Patients who wear compression garments can use one of lower compression if available, or receive modified bandaging until garments can be tolerated.

Modified intensive therapy with high pressure (>45mmHg)
This involves skin care, exercise/movement, elevation, MLD/SLD and MLLB with inelastic bandages undertaken three times weekly.

Suitable patients are able to tolerate high levels of compression, but are unable to commit to standard intensive therapy for physical, social, psychological or economic reasons. This may include those who are elderly, obese or have poor mobility.

The practitioner will be appropriately trained, and have access to physiotherapy assessment and to a practitioner with specialist training.

Modified intensive therapy with reduced pressure (15-25mmHg)
This involves skin care, exercise/movement, elevation, SLD, MLLB +/– IPC undertaken three times weekly.

Patients are selected for this treatment when high levels of compression are either unsafe or difficult to tolerate. This includes those with:

- moderate concurrent lower limb peripheral arterial occlusive disease (ABPI 0.5-0.8)[50]. NB Patients with ABPI <0.5 should not receive sustained compression therapy, but may benefit from special forms of IPC
- a neurological deficit that will make sensing complications difficult
- lipoedema/lipolymphoedema – lower levels of compression may be easier to tolerate
- cancer requiring palliative treatment
- co-morbidities requiring less aggressive reduction in swelling.

The practitioner will be appropriately trained, and have access to physiotherapy assessment and to a practitioner with specialist training.

Intensive therapy for lymphovenous disease (35-45mmHg or 15-25mmHg)
This involves skin care, exercise/movement, elevation, and MLLB +/– IPC undertaken either daily or three times weekly. Treatment frequency will be determined by the severity of the oedema, skin condition and rate of swelling reduction.

Suitable patients include those who have had deep vein thrombosis or those who have post-thrombotic syndrome, who may be at risk of developing or have existing leg ulceration. A recent review concluded that immediate ambulation with appropriate compression does not significantly increase the incidence of pulmonary embolism, produces a faster reduction of pain and swelling, and reduces the severity of post-thrombotic syndrome[51]. MLLB may need to be modified in the presence of venous ulceration, peripheral arterial occlusive disease or immobility (Appendix 2). IPC may be particularly useful for the many patients with venous ulceration who have poor mobility and are unable to elevate their legs[52-54].

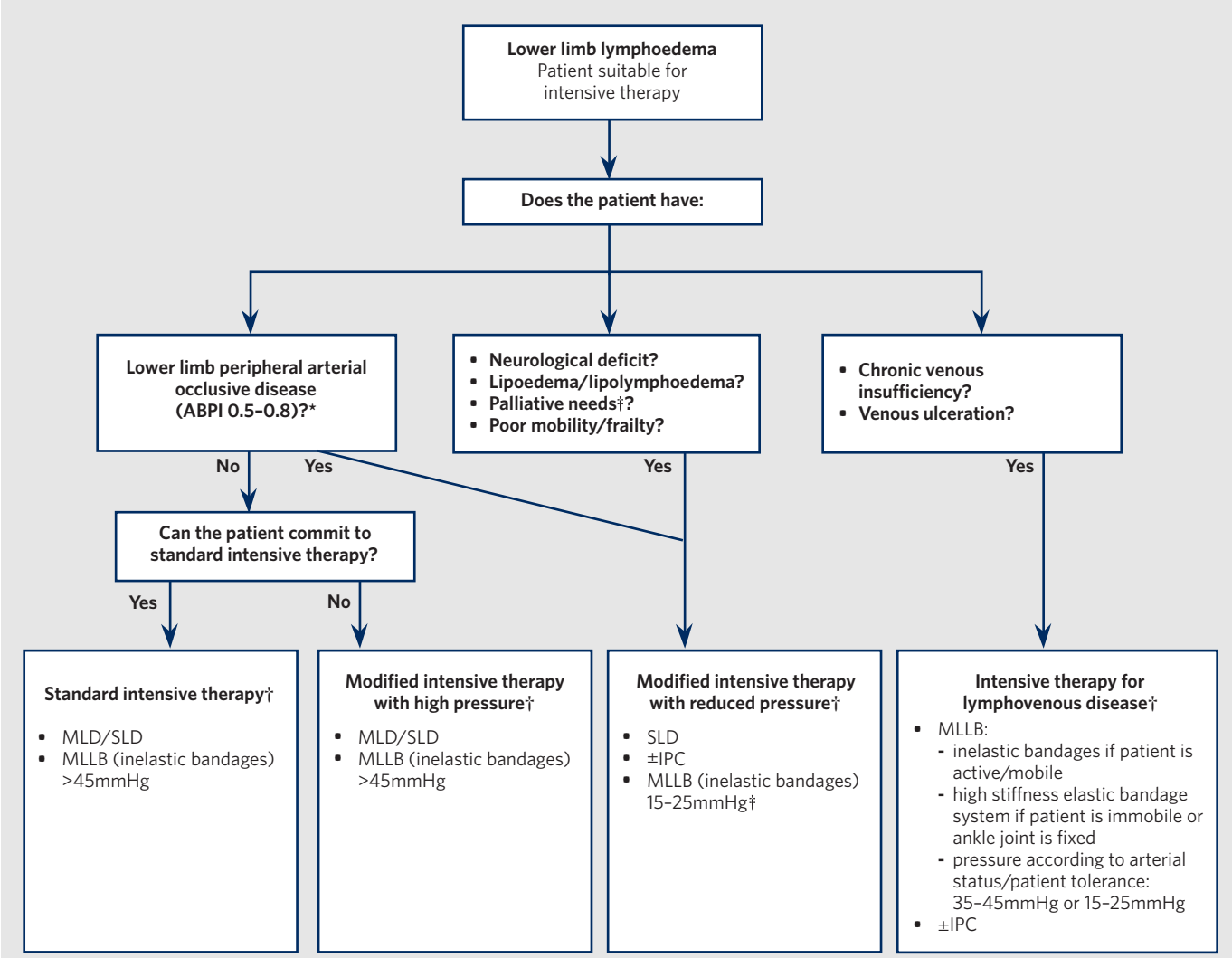
NB In severe cases with significant limb distortion, oedema and tissue thickening, fitter patients may benefit from a period of standard intensive therapy.

The practitioner will be appropriately trained at specialist level.

SUB-BANDAGE PRESSURE

- The pressures given here are sub-bandage pressures measured at the ankle in the supine position.
- The optimal sub-bandage ankle pressures for the MLLB systems used in lymphoedema have yet to be determined.
- The recommendations here relate to the sub-bandage ankle pressures recommended for venous disease[55].

FIGURE 7 Intensive therapy options for patients with lower limb lymphoedema



*Patients with ABPI <0.5 should not receive compression therapy and should be referred to a vascular specialist.
†Includes skin care, exercise/movement and elevation.
‡In the palliative situation, bandages may be used to support the limb and would apply very little compression.

UPPER LIMB LYMPHOEDEMA – INITIAL MANAGEMENT

As for the lower limb, initial management for upper limb lymphoedema will involve psychosocial support, education, skin care, exercise/movement, elevation and management of any pain or discomfort (Figure 6, page 16). The patient's initial management may also include:

- compression garments
- modified MLLB
- intensive therapy.

Compression garments

Compression garments can be used as initial management in patients who have mild upper limb lymphoedema (ISL stage I) with minimal subcutaneous tissue changes and shape distortion. Where there is considerable soft pitting oedema, MLLB (inelastic bandaging) will be required to reduce and stabilise the swelling prior to the application of compression garments.

In general, the level of compression used to treat lymphoedema of the upper limb is lower than that required for lower limb lymphoedema. Lower pressure compression garments also have a role to play in managing symptoms in a palliative context.

Management of patients treated initially with compression garments will include education about risk reduction and self management, skin care, exercise/movement, elevation, SLD and psychosocial support.

The practitioner will be appropriately trained.

Modified MLLB

Initial management of upper limb lymphoedema with MLLB will usually be part of an intensive therapy regimen (see below). Selected patients with ISL stage II or late stage II upper limb lymphoedema who are unable to wear compression garments may better tolerate adapted forms of MLLB. The initial and longer term management of patients with palliative care needs may also involve modified MLLB (page 34).

The practitioner will be appropriately trained.

Intensive therapy

Intensive therapy of upper limb lymphoedema involves the use of MLLB to reduce oedema and improve, where required, limb shape, subcutaneous tissue consistency and skin condition. In the intensive phase of treatment, daily bandaging is undertaken for two to four weeks, and all aspects of standard intensive therapy are implemented, ie skin care, exercise/movement, elevation and MLD.

In the palliative situation, where modified MLLB is used, it may be possible to reduce the frequency of bandaging after at least an initial week of daily treatment.

The practitioner will be appropriately trained, and have access to physiotherapy assessment and to a practitioner with specialist training.

TRANSITION MANAGEMENT – UPPER AND LOWER LIMB LYMPHOEDEMA

Following intensive therapy, some patients may benefit from a one to three month period of transition management before progressing to long-term therapy. The transition period may be helpful to:

- maximise the effects of intensive therapy and stabilise fluctuations in swelling to an individually acceptable level
- prevent rebound swelling on transfer to compression hosiery
- evaluate long-term maintenance strategies
- support and facilitate self management
- reduce practitioner input.

An algorithm has been developed to guide practitioners in deciding which patients require transition therapy (Figure 8).

Transition management requires a practitioner who has received appropriate training at specialist level, and may be shared with community staff.

Compression choices

Success and concordance demand that an individualised compression regimen is developed that is comfortable and acceptable to the patient. Treatment may include a combination of compression garments and MLLB, with or without MLD or IPC (Figure 9).

Promotion of self management

An important aim of the transition phase is promotion of self management and long-term control. Patients should, wherever possible, be actively engaged in all stages of their treatment. Patient involvement during the transition phase, with education, training and supervision, can include:

- skin care
- exercise/movement, elevation and weight reduction
- use of an inelastic adjustable compression device
- SLD
- compression garments +/- MLLB
- self monitoring for complications
- treatment adjustment according to fluctuations.

A trained and competent health or social carer or a relative can support any or all of these activities.

FIGURE 8 Transition management – upper and lower limb lymphoedema

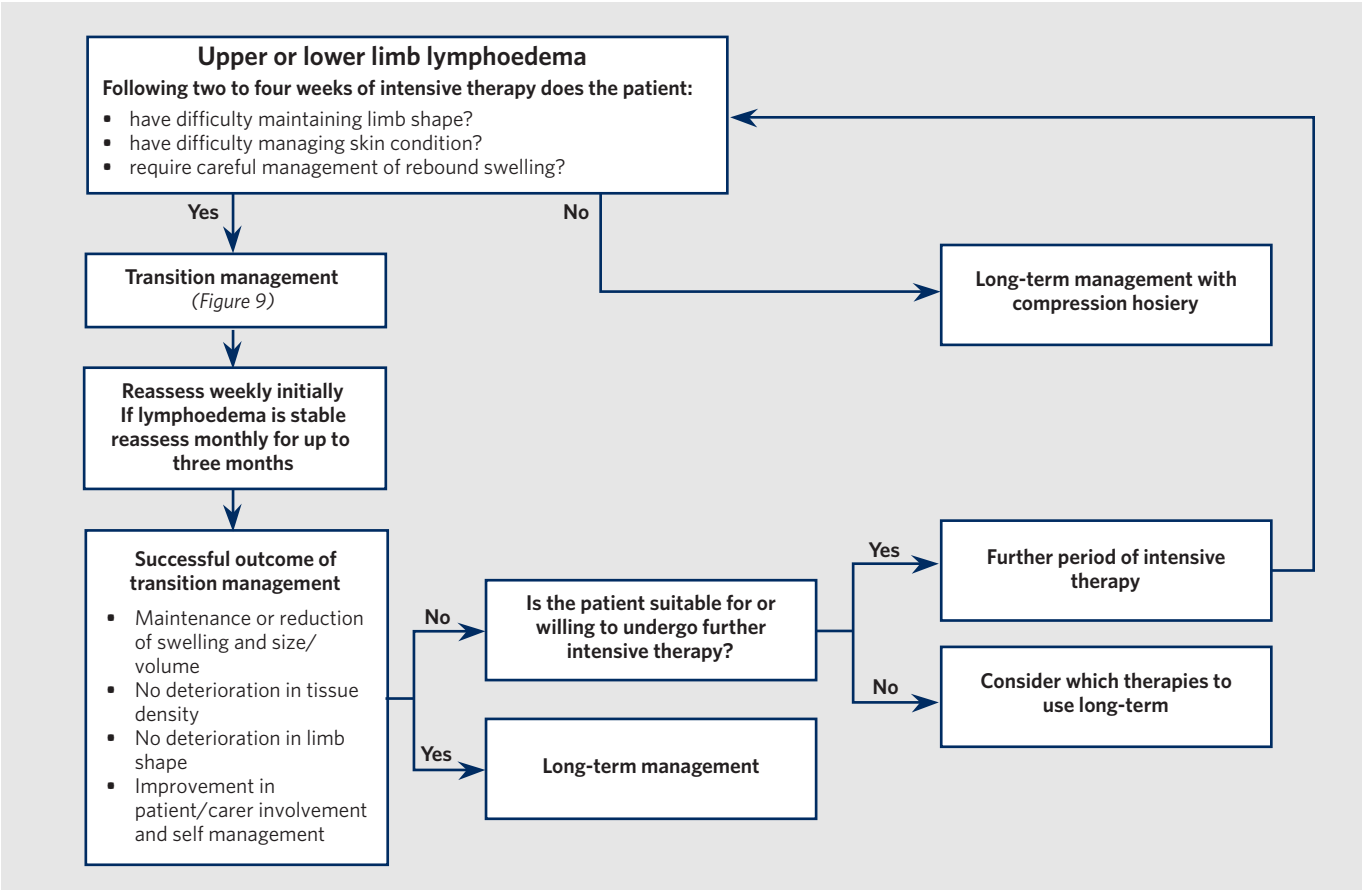
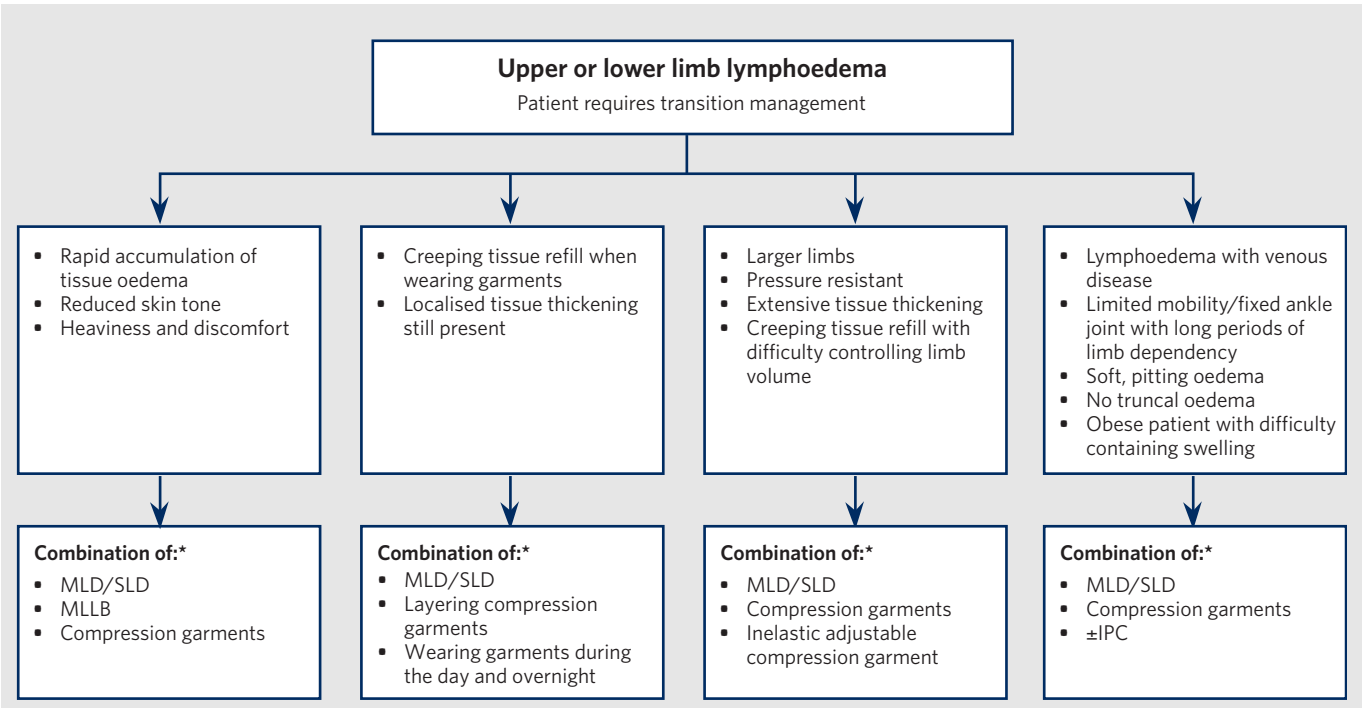


FIGURE 9 Compression choices in transition management for upper or lower limb lymphoedema



*Includes skin care, exercise/movement and elevation.

COMPRESSION GARMENTS

Most patients with lymphoedema who require long-term management will use compression garments.

LONG-TERM MANAGEMENT – UPPER LIMB AND LOWER LIMB LYMPHOEDEMA

The long-term management of lymphoedema focuses on enhancing the function of the lymphatics, limiting further deterioration of swelling, and gaining long-term control of the condition. Success relies on self management by patients and carers, with appropriate and effective education, training, and medical and psychosocial support. It involves:

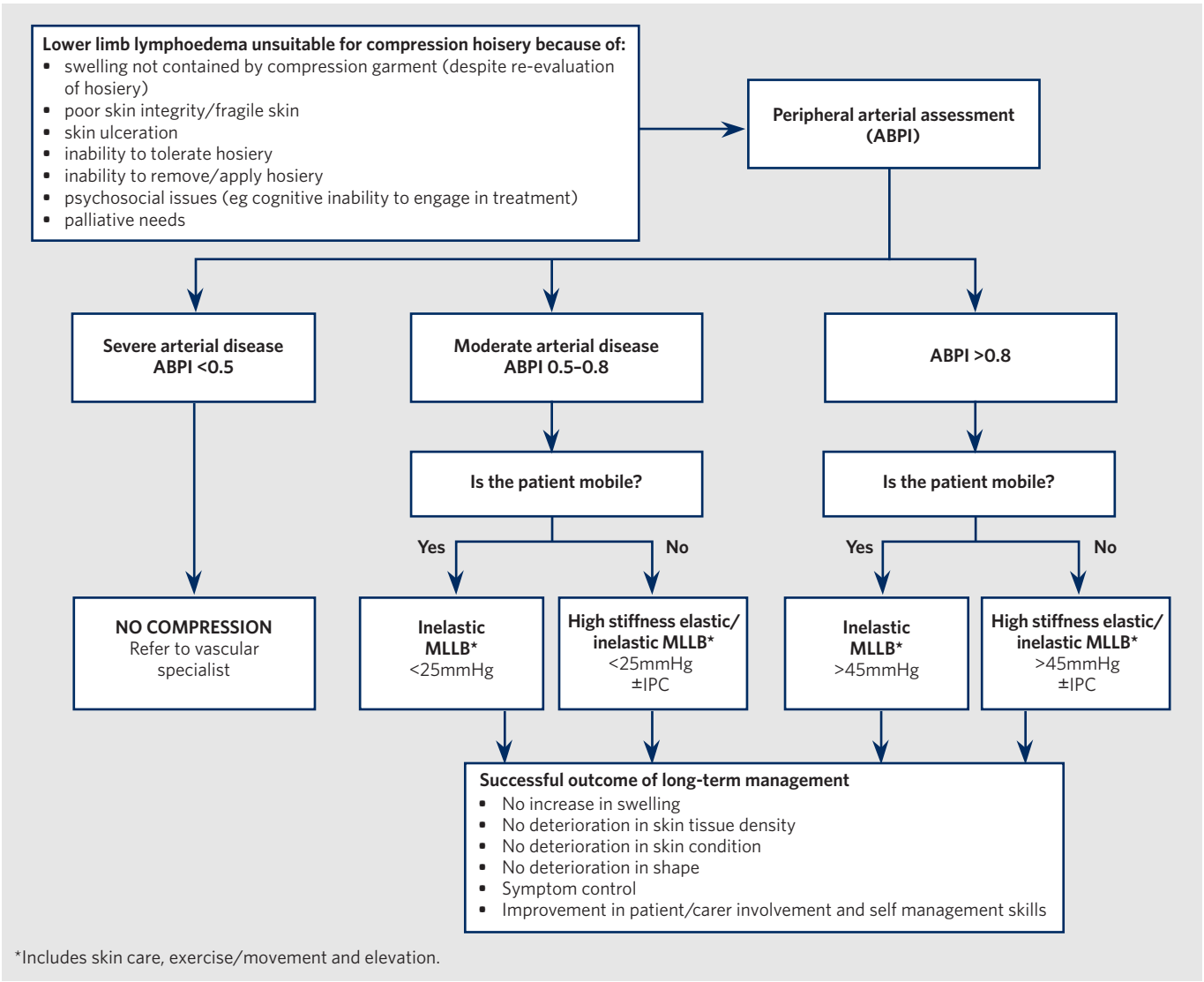
- daily skin care
- exercise/movement
- compression – compression garments, bandaging or an inelastic adjustable compression device
- limb elevation
- SLD performed by the patient or a trained carer/relative
- self monitoring.

Long-term management of lymphoedema usually involves compression garments. However, for some patients the most appropriate form of compression in the long-term will be bandaging (Figure 10) or a combination of compression garments and bandaging.

Occasionally, patients with upper limb lymphoedema who have developed expertise in managing their condition will be able to manage their lymphoedema mainly through exercise, using compression garments when needed.

Long-term management requires that the practitioner has appropriate training, and access to a practitioner with specialist training.

FIGURE 10 Long-term management of lower limb lymphoedema with MLLB



*Includes skin care, exercise/movement and elevation.

MANAGEMENT OF MIDLINE LYMPHOEDEMA

The management of midline lymphoedema (Box 16), ie lymphoedema of the head and neck, trunk, breast or genitalia, can be particularly challenging, especially because of the lack of standardised objective measurement methods to evaluate treatment effects and to facilitate measurement for appropriate compression garments.

Practitioners treating midline lymphoedema will be trained at specialist level. Management will require collaboration with the patient and a multidisciplinary team. In some circumstances, care may be managed jointly with community staff.

Truncal lymphoedema

Lymphoedema can affect the chest, back, abdomen, buttocks, breast or genitalia in isolation or in combination with limb oedema. Lymphoedema of the trunk is often secondary to a tumour compressing the lymphatics or to trauma and tissue damage from cancer treatment. Consequently, particular attention should be paid to determining the presence or recurrence of cancer during initial assessment.

The management strategies described for breast and genital lymphoedema can be combined, where necessary, with those for the management of limb lymphoedema[56].

Breast lymphoedema

There is little consensus on the best approach to the management of breast lymphoedema. However, prevention, early diagnosis and supportive care have much to offer. MLD and SLD form an important part of treatment. Medium compression may be applied using suitable bras (including sports bras), Lycra foundation garments or custom made garments. Tissue thickening may be softened by using customised foam pads. The anatomy of the area may make bandaging difficult.

Genital lymphoedema

Genital lymphoedema can be highly incapacitating and extremely difficult to manage. Careful monitoring for signs of infection and scrupulous skin care are crucial. MLD and SLD are important treatment components.

When genital lymphoedema and lower limb lymphoedema co-exist, treatment of the lower limb swelling may exacerbate the genital oedema. In this situation, clearance of the core lymphatics through MLD is particularly important.

Women usually require custom made compression garments with anatomically contoured stasis pads to treat thickened and swollen areas. In men, MLLB may be used and self-bandaging taught. Depending on the degree of swelling, supportive close fitting shorts containing Lycra (eg cycle shorts) may be a useful alternative to ready to wear or custom made scrotal supports or compression garments. In either sex, surgical management may sometimes be necessary.

Lymphoedema of head and neck

Lymphoedema of the head and neck is often a complication of cancer or secondary to tissue damage in this area. MLD and SLD are key elements of treatment. Low pressure compression may be applied using bandaging or custom made garments. Low density foam pads can be used to apply localised pressure. Compression should never be applied to the neck area. Surgical management of eyelid lymphoedema may be considered.

Box 16 Principles of management for midline lymphoedema

The individually tailored management plan for patients with lymphoedema of the head and neck, trunk, breast or genitalia, is likely to include:

- daily skin care
- exercise/movement
- massage – MLD and/or SLD
- compression – bandaging, compression garments and individualised foam pads
- self monitoring

NB Compression may not be well tolerated in midline lymphoedema and MLD may be the only realistic option.

Skin care and cellulitis/erysipelas

FIGURE 11 Intact skin

FIGURE 12 Rough and scaly dry skin

B **Good skin care regimens should be implemented by patients and carers in the management of lymphoedema.**

Skin problems are common in patients with lymphoedema. Swelling may produce deep skin folds where fungal and bacterial infections can develop. Chronic inflammation causes deposition of fibrin and collagen, contributing to skin thickening and firm tissue consistency. Reduced tissue compliance may further compromise lymph flow and increase the tendency to infection. **Maintenance of skin integrity and careful management of skin problems in patients with lymphoedema are important to minimise the risk of infection.**

The general principles of skin care (Box 17) aim to preserve skin barrier function through washing and the use of emollients. Ordinary soaps, which usually contain detergents and no glycerin, should be avoided because they tend to dry the skin. Natural or pH neutral soap can be used. The perfumes and preservatives in scented products may be irritant or allergenic. In high concentrations, mineral and petrolatum based products may exacerbate dry skin conditions by occluding skin pores and preventing natural oils from surfacing.

Emollients re-establish the skin's protective lipid layer, preventing further water loss and protecting the skin from bacteria and irritants. Emollients can be bath oils, soap substitutes or moisturisers (lotions, creams and ointments). In general, ointments, which contain little or no water, are better skin hydrators than creams, which are better than lotions.

The best method of emollient application is unknown. Some practitioners recommend applying them using strokes in the direction of hair growth (ie towards the feet when applying to the legs) to prevent blockage of hair follicles and folliculitis. Others recommend applying emollients by stroking towards the trunk to encourage lymph drainage.

Emollients may damage the elastic component of compression garments, and it is preferable to avoid application immediately prior to donning.

Box 17 General principles of skin care

- Wash daily, whenever possible, using pH neutral soap, natural soap or a soap substitute, and dry thoroughly
- Ensure skin folds, if present, are clean and dry
- Monitor affected and unaffected skin for cuts, abrasions or insect bites, paying particular attention to any areas affected by sensory neuropathy
- Apply emollients
- Avoid scented products
- Particularly in hot climates, vegetable-based products are preferable to those containing petrolatum or mineral oils

SKIN CARE REGIMENS

Following are descriptions of skin care regimens for skin conditions that can occur in patients with lymphoedema. These conditions may occur simultaneously and require combinations of regimens. The general principles of skin care apply to all conditions (Box 17).

Intact skin

The condition of intact skin (Figure 11) should be optimised by applying emollient at night.

Dry skin

Dry skin may vary from slightly dry or flaky to rough and scaly (Figure 12). Patients may complain of itching.

Emollients should be applied twice daily (including after washing) to aid rehydration. If the heels are deeply cracked, emollients and hydrocolloid dressings may help and the patient should be referred according to local dermatology guidelines.

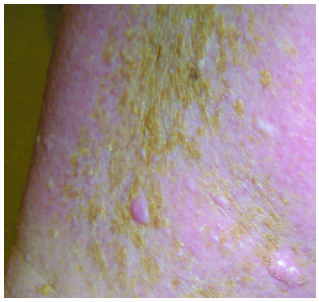


FIGURE 13 Hyperkeratosis

FIGURE 14 Folliculitis

FIGURE 15 Fungal infection on the sole of the foot

FIGURE 16 Lymphangiectasia

Hyperkeratosis

Hyperkeratosis (Figure 13) is caused by overproliferation of the keratin layer and produces scaly brown or grey patches.

Emollients with a low water content are recommended. MLLB reduces the underlying lymphoedema and improves skin condition. If the condition has not improved within two weeks, the patient should be referred according to local dermatology guidelines.

Folliculitis

Folliculitis (Figure 14) is due to inflammation of the hair follicles. It causes a red rash with pimples or pustules, and is most commonly seen on hairy limbs. The cause is usually *Staphylococcus aureus*, and it may precede cellulitis/erysipelas. Swabs should be taken for culture if there is any exudate or an open wound.

An antiseptic wash/lotion, eg one containing chlorhexidine and benzalkonium, should be used after washing. Emollient should be applied without being rubbed in. If there is no response after one month, the patient should be referred according to local dermatology guidelines.

Fungal infection

Fungal infection (Figure 15) occurs in skin creases and on skin surfaces that touch. It causes moist, whitish scaling and itching, and is particularly common between the toes. It can precede the development of cellulitis/erysipelas. Skin scrapings and, if nails are affected, nail clippings should be sent for mycological examination.

Treatment is with terbinafine 1% cream for up to six weeks alongside meticulous skin care. In some countries, Whitfield ointment is used as an alternative. Any sign of bacterial infection should be treated promptly (pages 27-29). Nail infection requires treatment with an oral antifungal agent under medical supervision. The patient should be referred to a dermatologist if there is no response after six weeks' treatment.

Lymphangiectasia

Lymphangiectasia (Figure 16 – also known as lymphangiomata) are soft fluid-filled projections caused by dilatation of lymphatic vessels. Treatment is compression with MLLB. If there is no response to initial compression, or the lymphangiectasia are very large, contain chyle or cause lymphorrhoea, the patient should be referred immediately to a lymphoedema practitioner with training at specialist level.

Papillomatosis

Papillomatosis (Figures 17 and 18) produces firm raised projections on the skin due to dilatation of lymphatic vessels and fibrosis, and may be accompanied by hyperkeratosis.

The condition may be reversible with adequate compression. If the condition does not improve after one month, the patient should be referred to a lymphoedema practitioner with training at specialist level.

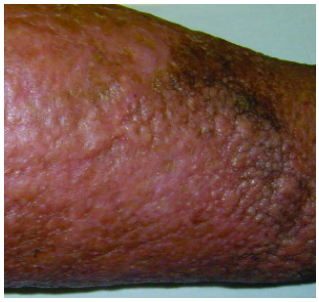


FIGURE 17 Papillomatosis

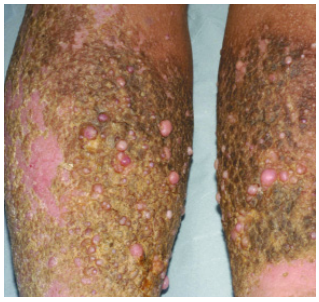


FIGURE 18 Severe papillomatosis

FIGURE 19 Lymphorrhoea and resulting maceration

FIGURE 20 Ulceration

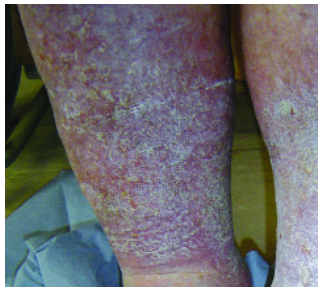


FIGURE 21 Venous eczema

FIGURE 22 Contact dermatitis

Lymphorrhoea

Lymphorrhoea (Figure 19) occurs when lymph leaks from the skin surface. The patient may require medical review to determine the underlying cause, eg worsening congestive heart failure.

The surrounding skin should be protected with emollient, and nonadherent absorbent dressings should be applied to the weeping skin. MLLB will reduce the underlying lymphoedema, but needs to be changed frequently to avoid maceration of the skin. Frequency of change will be determined by factors such as strikethrough and the rate of swelling reduction. In the palliative situation, light bandaging may be more appropriate. If the condition does not improve with two weeks of treatment, the patient should be referred to a lymphoedema practitioner with training at specialist level.

Ulceration

It is important to establish the underlying cause of the ulcer because it determines treatment and whether compression is appropriate (Figure 20). If venous and/or arterial disease is present, the internationally agreed leg ulcer management algorithm should be followed (Appendix 2). The ulcer will require an appropriate dressing and the surrounding skin will need to be treated according to its condition. Exercise/movement and optimal nutrition should be encouraged and long periods of limb dependency minimised. The patient should be referred to the appropriate specialist service if the ulcer is unresponsive after six to eight weeks, there is rapid deterioration or a drop in ABPI.

Venous eczema

Venous eczema (also known as varicose eczema or stasis dermatitis) usually occurs on the lower legs (Figure 21), particularly around the ankles, and is associated with varicose veins. The skin becomes pigmented, inflamed, scaly and itchy.

Treatment is with topical corticosteroids in ointment form as recommended in local guidelines, eg a potent corticosteroid such as betamethasone valerate 0.1% with clioquinol 3% for seven days followed by a mildly potent corticosteroid such as clobetasone butyrate 0.05% or betamethasone valerate 0.025%. A non-sensitising, low water content emollient should be applied during steroid treatment. If ABPI is <0.5, the patient should be referred to a vascular surgeon. The patient should be referred according to local dermatology guidelines if the condition persists.

Contact dermatitis

Contact dermatitis (Figure 22) is the result of an allergic or irritant reaction. It usually starts at the site of contact with the causative material, but may spread. The skin becomes red, itchy and scaly, and may weep or crust.

Acute episodes are treated with a potent topical corticosteroid in ointment form, eg betamethasone valerate 0.1% once or twice daily. For dermatitis unresponsive to less potent corticosteroids, treatment is with a very potent topical corticosteroid such as clobetasol propionate 0.05% once or twice daily. Treatment should continue for three to four weeks, during which time the strength of the steroid and amount applied are gradually reduced. The patient should be referred according to local dermatology guidelines if the condition does not improve.

Lymphangiosarcoma

In the most severe cases of lymphoedema, lymphangiosarcoma, a rare form of lymphatic cancer (Stewart-Treves syndrome) can develop (Figure 23). It mainly occurs in patients who have been treated for breast cancer with mastectomy and/or radiotherapy. The sarcoma first appears as a reddish or purplish discolouration or as a bruised area that does not change colour. It progresses to an ulcer with crusting, and eventually to extensive necrosis of the skin and subcutaneous tissue. It can metastasise widely. Patients with suspected lymphangiosarcoma require urgent referral to an oncologist.

CELLULITIS/ERYSIPELAS

Patients with lymphoedema are at increased risk of acute cellulitis/erysipelas, an infection of the skin and subcutaneous tissues. The cause of most episodes is believed to be Group A β-haemolytic streptococci. It may also be caused by staphylococci or other bacteria.

Good skin care reduces the likelihood of cellulitis/erysipelas, and consequently the need for antibiotics.

NOTE: CELLULITIS TERMINOLOGY

Cellulitis may also be known as:

- erysipelas
- acute inflammatory episode
- lymphangitis
- dermohypodermal infection
- lymphoedema-related acute dermatitis
- dermatolymphangioadenitis (DLA)

Symptoms are variable. Episodes may come on over minutes, grumble over several weeks or be preceded by systemic upset. Symptoms include pain, swelling, warmth, redness, **lymphangitis**, lymphadenitis and sometimes blistering of the affected part (Figure 24). More severe cases have a greater degree of systemic upset, eg chills, rigor, high fever, headache and vomiting. In rare cases, these symptoms may be indicative of necrotising fasciitis. The focus of the infection may be tinea pedis (athlete's foot), venous eczema, ulceration, ingrowing toe nails, scratches from plants or pets, or insect bites. Box 18 (page 28) outlines the principles involved in the management of acute cellulitis/erysipelas at home or in hospital.

Summary of guidelines for the management of cellulitis/erysipelas in lymphoedema[57]

The guidelines summarised here describe the indications for hospital admission and antibiotic therapy for acute and recurrent cellulitis/erysipelas in patients with lymphoedema.

Prompt treatment of cellulitis/erysipelas is essential to prevent further damage that can predispose to recurrent attacks.

Criteria for hospital admission

The patient should be admitted to hospital if they show:

- signs of septicaemia (hypotension, tachycardia, severe pyrexia, confusion or vomiting)
- continuing or deteriorating systemic signs, with or without deteriorating local signs, after 48 hours of oral antibiotics
- unresolving or deteriorating local signs, with or without systemic signs, despite trials of first and second line oral antibiotics.

It is essential that patients with cellulitis/erysipelas, who are managed at home, are monitored closely, ideally by the general practitioner.

FIGURE 23 Lymphangiosarcoma
Lymphangiosarcoma developing in long standing breast cancer related lymphoedema.

FIGURE 24 Cellulitis/erysipelas

Lymphangitis: inflammation of lymph vessels

BOX 18 Principles of home- or hospital-based management of acute cellulitis/erysipelas

Exclude:
- other infections, eg those with a systemic component
- venous eczema, contact dermatitis, intertrigo, microtrauma and fungal infection
- acute deep vein thrombosis
- thrombophlebitis
- acute lipodermatosclerosis
- lymphangiosarcoma (Stewart-Treves syndrome)

Swab any exudate or likely source of infection, eg cuts or breaks in the skin

Before commencing antibiotics establish:
- extent and severity of the rash - mark and date the edge of the erythema
- presence and location of any swollen and painful regional lymph nodes
- degree of systemic upset
- erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) and white cell count

Commence antibiotics as soon as possible (Table 4), taking into account swab results and bacterial sensitivities when appropriate

During bed rest, elevate the limb, administer appropriate analgesia (eg paracetamol or NSAID), and increase fluid intake

Avoid SLD and MLD

If tolerated, continue compression at a reduced level or switch from compression garments to reduced pressure MLLB

Avoid long periods without compression

Recommence usual compression and levels of activity once pain and inflammation are sufficiently reduced for the patient to tolerate

Educate patient/carer - symptoms, when to seek medical attention, risk factors, antibiotics 'in case', prophylaxis if indicated

TABLE 4 Antibiotics for cellulitis/erysipelas in lymphoedema[57]

Situation	First-line antibiotics*	If allergic to penicillin*	Second-line antibiotics*	Comments*
Home care Acute cellulitis/erysipelas	Amoxicillin 500mg eight hourly +/- flucloxacillin 500mg six hourly†	Clindamycin 300mg six hourly	Clindamycin 300mg six hourly If fails to resolve, convert to iv regimen as for hospital admission	Treat for at least 14 days or until signs of inflammation have resolved
Hospital admission Acute cellulitis/erysipelas + septicaemia	Amoxicillin iv 2g eight hourly (or benzylpenicillin iv 1200-2400mg six hourly) plus gentamycin iv 5mg/kg daily	Clindamycin iv 1.2g six hourly	Clindamycin iv 1.2g six hourly (if poor or no response by 48 hours)	Switch to amoxicillin 500mg eight hourly when: ■ temperature down for 48 hours ■ inflammation much resolved ■ CRP <30mg/L
Prophylaxis to prevent recurrent cellulitis/erysipelas (≥two attacks per year)	Phenoxymethylpenicillin 500mg once daily (1g once daily if weight >75kg)	Erythromycin 250mg once daily	Clindamycin 150mg once daily or clarithromycin 250mg once daily	After one year, halve dose of penicillin to 250mg once daily (500mg once daily if weight >75kg)
Emergency supply of antibiotics, 'in case of need' (when away from home)	Amoxicillin 500mg eight hourly	Clindamycin 300mg six hourly	If fails to resolve, or constitutional symptoms develop, convert to iv regimen as for hospital admission	
History of animal bite	Co-amoxiclav 625mg six hourly	Ciprofloxacin 500mg twelve hourly	Consult microbiologist	Causes may be *Pasteurella multocida, Eikinella corrodens* or *Capnocytophaga canimorsus*

NB Local guidelines may determine which antibiotics may be used.
*Dosages are for oral treatment unless stated otherwise; iv = intravenously.
†Add if infection with *Staphylococcus aureus* is suspected, eg if folliculitis, pus formation, and/or crusted dermatitis are present.

Antibiotic regimens
Antibiotic regimens for cellulitis/erysipelas in lymphoedema vary according to the clinical situation (Table 4). Antibiotics should be continued for at least 14 days after an acute episode has responded clinically to treatment. It may take one to two months of antibiotic treatment to achieve complete resolution.

Antibiotics 'in case'
The risk of further attacks of cellulitis/erysipelas in lymphoedema is high. It is recommended that patients who have had an attack of cellulitis/erysipelas carry a two week supply of oral antibiotics, particularly when away from home for any length of time, eg on holiday. Patients should be advised to start antibiotics immediately when familiar symptoms of cellulitis/erysipelas arise and to seek a medical opinion as soon as possible.

Recurrent cellulitis/erysipelas
Antibiotic prophylaxis should be offered to patients who have two or more attacks of cellulitis/erysipelas per year (Table 4). After two years of successful prophylaxis the antibiotics can be discontinued. However, if cellulitis/erysipelas recurs, lifelong antibiotic prophylaxis is required.

The risk of recurrent cellulitis/erysipelas can be reduced by controlling swelling, and by treating interdigital scaling, fungal infections, folliculitis, dermatitis, open wounds (including leg ulcers) and weeping lymphangiectasia.

Lymphatic massage

Lymphatic massage – manual lymphatic drainage (MLD) and simple lymphatic drainage (SLD) – aims to reduce swelling by encouraging lymph flow.
The efficacy of MLD and SLD remains to be proven, but there is no doubt that they are of immense value in providing psychological and symptomatic benefits.

MANUAL LYMPHATIC DRAINAGE

C MLD and compression can reduce and control lymphoedema of the head, neck and body.

Manual lymphatic drainage (MLD) is a gentle massage technique that is recognised as a key component of decongestive therapy. MLD aims to encourage fluid away from congested areas by increasing activity of normal lymphatics and bypassing ineffective or obliterated lymph vessels. Although there is a wealth of clinical opinion advocating the benefits of MLD, there are little research data to conclusively support its use[2,58-60]. The most appropriate techniques, optimal frequency and indications for MLD, as well as the benefits of treatment, all remain to be clarified.

MLD remains a specialist skill that needs regular practice in order to maintain competence. Deep, heavy-handed massage should be avoided because it may damage tissues and exacerbate oedema by increasing capillary filtration.

Indications
MLD may be indicated as part of intensive therapy, transition management, long-term management or palliative care (Box 19). MLD on its own is not sufficient treatment for lymphoedema; it should be combined with compression therapy to support and maintain its effects. However, where compression is difficult or is not well tolerated, eg in lymphoedema of the head, neck, trunk, breast and genitalia, MLD may be the only realistic option.

BOX 19 Indications for MLD and SLD

- Swelling at the root of a limb
- Trunk and midline oedema (eg chest, breast, back, abdomen, genitalia, head and neck)
- Provision of comfort and pain relief when other physical therapies are no longer appropriate
- Adjunctive treatment to pain management

An important contraindication to MLD and SLD is acute cellulitis/erysipelas (Box 20). In advanced cancer, MLD/SLD can be used with medical practitioner and patient consent, but should not be used over the sites of known primary tumours or metastases.

Technique

A number of different techniques exist for MLD. However, there is little evidence to demonstrate which is the most effective and for what clinical indications. Essentially, MLD is a gentle massage technique that follows the lymphatic pathways. The different methods have several aspects in common:

- performed for up to an hour daily
- usually performed with the patient in the lying position, unless for lymphoedema of the head and neck
- starts with deep diaphragmatic breathing
- treats the unaffected lymph nodes and region of the body first
- moves proximally to distally to drain the affected areas
- movements are slow and rhythmical
- uses gentle pressure – if the pressure is too hard it stimulates blood flow, the skin becomes red, and more fluid is encouraged to move into the tissues
- ends with deep diaphragmatic breathing.

MLD may be conducted daily (or sometimes twice daily) or three times weekly. A course of therapy may last three or more weeks, and may be repeated at intervals of three months to one year[61]. However, the ideal frequency and length of course for MLD remains to be defined.

MLD is conducted by practitioners with training at specialist level.

SIMPLE LYMPHATIC DRAINAGE

Simple lymphatic drainage (SLD) is a simplified self-administered version of MLD that patients and carers can learn and apply themselves[62,63]. Ideally, all patients should be taught SLD, unless contraindicated (Box 20). While there may be benefits, some patients find it difficult to learn, memorise and effectively incorporate this treatment into a daily regimen. Patients who have MLD may find it easier to learn SLD.

Technique

In common with MLD, there is little robust evidence to support the use or effect of SLD. There is no definitive technique for SLD, but it is similar to MLD and is conducted for 10-20 minutes daily.

For SLD to be effective, the healthcare professional must ensure that:

- the patient/carer is motivated
- the patient/carer is sufficiently dextrous to perform SLD
- time is allocated for initial teaching
- teaching is progressive and enables the patient or carer to become skilled
- written instruction is given and technique is observed
- competence in the procedure and the patient's ability to cope with treatment are checked regularly.

SLD is conducted and taught by practitioners with appropriate training.

BOX 20 Contraindications to MLD and SLD

General contraindications

- Acute cellulitis/erysipelas
- Renal failure
- Unstable hypertension
- Severe cardiac insufficiency
- Hepatic cirrhosis with abdominal fluid (ascites)
- Superior vena cava obstruction
- Untreated tuberculosis or malaria

Local contraindications*

- Untreated thyroid dysfunction
- Primary tumours
- Metastases

Caution required: cardiac insufficiency.
*MLD and SLD should not be performed at these sites.

Intermittent pneumatic compression

Although there is considerable international debate over its effectiveness in lymphoedema, intermittent pneumatic compression (IPC) is widely used. It may form part of an intensive therapy regimen or long-term management in selected patients, and may be used with caution in the palliative situation.

WHAT IS IPC?

IPC consists of an electrical air compression pump attached to an inflatable plastic garment that is placed over the affected limb. The garment is inflated and deflated cyclically for a set period, usually about 30-120 minutes. The pressure produced by the garment can be varied. Garments may be single chambered, or contain multiple chambers (usually three, five or 10) that are inflated sequentially to provide a peristaltic massaging effect along the length of the limb towards its root.

The question of whether single or multichambered devices are more effective remains open. However, multichambered devices are used most frequently and randomised controlled trials have shown them to produce a faster effect[64,65].

IPC is thought to reduce oedema by decreasing capillary filtration, and therefore lymph formation, rather than by accelerating lymph return.

IPC is particularly effective in nonobstructive oedemas, eg those due to immobility, venous incompetence, lymphovenous stasis or hypoproteinaemia. In obstructive lymphoedema, ie lymphoedema resulting from lymphatic vessel/node damage or lymph node resection, SLD or MLD is recommended before IPC to stimulate lymphatic flow[66].

It is important that compression therapy with garments or bandaging is continued after IPC to prevent rapid rebound swelling. Contraindications to IPC are listed in Box 21.

BOX 21 Contraindications to IPC

- Untreated nonpitting chronic lymphoedema
- Known or suspected deep vein thrombosis
- Pulmonary embolism
- Thrombophlebitis
- Acute inflammation of the skin, eg cellulitis/erysipelas
- Uncontrolled/severe cardiac failure
- Pulmonary oedema
- Ischaemic vascular disease
- Active metastatic disease affecting oedematous region
- Oedema at the root of the affected limb or truncal oedema
- Severe peripheral neuropathy

Caution required: peripheral neuropathy, pain or numbness in the limb, undiagnosed, untreated or infected wounds, fragile skin, grafts, skin conditions that may be aggravated by IPC, extreme limb deformity (may impede correct use of IPC).

GUIDELINES FOR USE

Consensus on the pressures suitable for IPC in lymphoedema is lacking.

Careful surveillance is required to ensure that the correct technique and pressures are applied. Pressures should be adjusted according to patient tolerance and response to treatment. In general:

- pressures of 30-60mmHg are advised
- lower pressures are advised in palliative care, eg 20-30mmHg
- duration and frequency of 30 minutes to two hours daily are recommended[66-68].

IPC may exacerbate or cause congestion or a ring of fibrosis at the noncompressed root of a treated limb if the lymphatics in the root of the limb have not been cleared. IPC of the lower limbs may precipitate genital oedema[69].

IPC is not recommended if there is oedema at the root of the limb or in the adjacent trunk.

IPC should be prescribed and performed by practitioners who have received appropriate training at specialist level.

Multi-layer inelastic lymphoedema bandaging

BOX 22 Indications for MLLB

Lymphoedema with:
- fragile, damaged or ulcerated skin
- distorted limb shape
- limb too large to fit compression garments
- areas of tissue thickening
- lymphorrhoea
- lymphangiectasia
- pronounced skin folds

Cautionary notes: Patients with significant skin sacs/lobes or extensive tissue thickening should be referred to a lymphoedema practitioner with training at specialist level. If there is swelling at the root of the limb or adjacent to the trunk, MLD should be performed in conjunction with MLLB.

B **Multi-layer systems followed by compression garments are more effective than single layer compression garments when used in the initial phase of lymphoedema treatment[70].**

Multi-layer lymphoedema bandaging (MLLB) is a key element of intensive therapy regimens. For some patients it may also form part of their transition, long-term or palliative management.

MLLB uses inelastic bandages that have low extensibility and that produce high working pressures and lower resting pressures (Figure 25), ie they create peak pressures that produce a massaging effect and stimulate lymph flow. In certain situations (page 34), elastic bandages may be used instead. Elastic bandages produce sustained compression with smaller variations during movement.

USES FOR MLLB

As well as reducing oedema, MLLB:
- restores shape to the limb/affected area
- reduces skin changes such as hyperkeratosis and papillomatosis
- supports overstretched inelastic skin
- eliminates lymphorrhoea
- softens subcutaneous tissues.

MLLB is indicated when skin changes are marked or limb distortion and skin folds preclude compression garments (Box 22).

Contraindications to MLLB include severe peripheral arterial occlusive disease (Box 23).

MLLB SYSTEMS

The purpose and characteristics of the usual components of MLLB in their order of use are described in Table 5.

MLLB regimens can be adapted to individual patient's needs by varying the:
- pressure produced by the bandages
- frequency of bandage change
- bandage bulk
- type of bandage, eg using elastic bandages instead of inelastic bandages.

BOX 23 Contraindications to MLLB

- Severe arterial insufficiency (ABPI <0.5), although modified MLLB with reduced pressures can be used under close supervision
- Uncontrolled heart failure
- Severe peripheral neuropathy

Caution required: cellulitis/erysipelas (MLLB can be continued, if tolerated, at reduced pressure), diabetes mellitus, paralysis, sensory deficit, controlled congestive heart failure (application of MLLB to one limb at a time may be advisable).

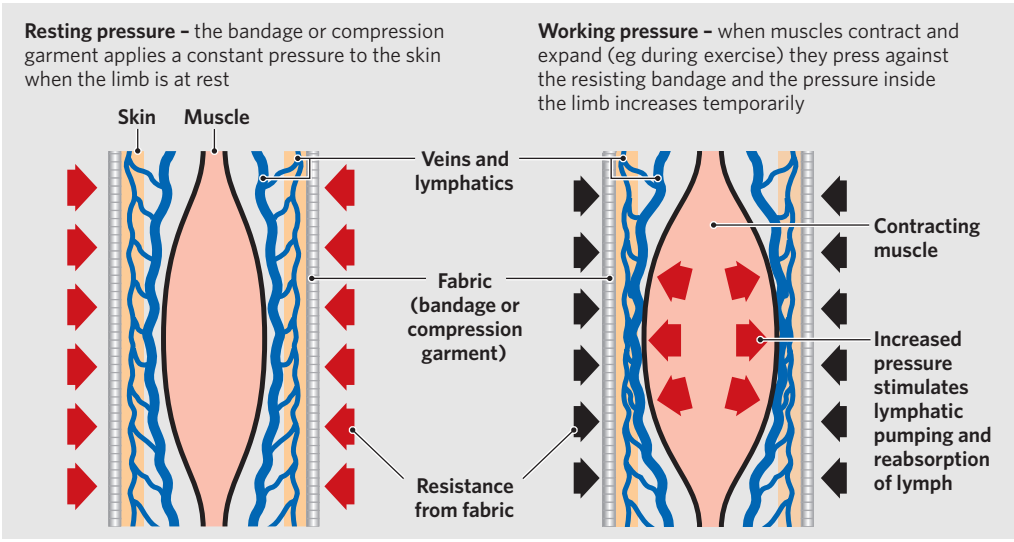


FIGURE 25 Resting and working pressures

TABLE 5 Components of MLLB (in order of use)

Component	Purpose	Characteristics	Notes
1. Skin care	To optimise skin health and treat any skin conditions, eg hyperkeratosis or ulceration	According to need	As a minimum, emollient should be applied to the skin before bandaging
2. Finger or toe bandaging (if indicated)	To prevent or reduce swelling of the fingers To reduce swelling of the toes	Conforming bandage	Bandaging should not impede function of digits
3. Tubular bandage	To provide a protective, absorbent layer between the skin and other bandages	A light cotton or cotton-viscose bandage applied to the whole area to be bandaged Does not contribute significantly to compression	Should be long enough to be folded back over the padding layer at either end to prevent fraying or chafing
4. Soft synthetic wool ('sub-compression wadding bandage') or foam roll or sheet	To protect the skin and subcutaneous tissues, to normalise shape*, to protect bony prominences and to equalise the distribution of pressure produced by other bandage layers	Soft synthetic wool or polyurethane foam is available in different widths and thicknesses, and as bandages or sheets Polyester undercast padding is available in sheets of various widths Higher densities of foam are used with greater degrees of shape distortion or tissue thickening	Extra padding may be required on vulnerable pressure points such as the Achilles' tendon, dorsum of the foot, tibialis anterior tendon, the malleoli, the popliteal fossa and the elbow
5. Dense foam	Applied locally to soften hard areas of tissue thickening* or areas particularly vulnerable to oedema, eg the malleoli	Polyurethane high density foam is available in sheets or pads of different thicknesses that can be cut to shape	Applied over soft synthetic wool or under foam Edges should be bevelled to prevent rubbing
6. Inelastic bandages	To provide compression	Constructed of crimped cotton yarns Available as nonadhesive, cohesive or adhesive Most types are available in 4cm, 6cm, 8cm, 10cm and 12cm widths	Several layers are used Cohesive and adhesive bandages can help to prevent slippage and are used to prolong the time the bandage is worn
7. Tape	To secure ends of bandages		The tape appropriate to the bandage being secured should be used

* Foam chip bags contain low density foam pieces in a tubular bandage and can be used to bulk out areas such as the palm of the hand or over areas of tissue thickening.

Achieving the desired pressure

The pressure produced by a compression bandage can be predicted according to Laplace's Law (Box 24). This law shows that sub-bandage pressure will:

- rise with increasing bandage tension and number of bandage layers
- decrease with increasing limb circumference and bandage width.

In practice, therefore, Laplace's Law shows that for a larger limb requiring high levels of compression, the desired pressure may be achieved by increasing the number of bandage layers applied and increasing the tension used during application.

BOX 24 Laplace's Law[71]

$$P = \frac{T \times N \times 4630}{C \times W}$$

P = sub-bandage pressure (mmHg)
T = bandage tension (kilograms force – kgf)
N = number of layers
C = limb circumference (cm)
W = bandage width (cm)

Frequency of MLLB system change

As yet, there is no empirical evidence to indicate how frequency of bandage change affects speed of oedema reduction or final outcome. Clinical experience recommends that MLLB systems should be changed daily

for the first seven days. This will minimise bandage slippage and ensure that sub-bandage pressure is maintained as swelling reduces. According to therapy regimen and wound/skincare requirements, it may then be possible to reduce the frequency of change to two to three times per week. Continence issues may also influence the frequency of change.

Commencement of bandaging and the timing of bandage change may need to be co-ordinated with any orthotic or podiatric needs of the patient.

Use of elastic bandaging

In some situations, the inelastic bandages used in MLLB may be replaced with a multi-layer elastic bandage regimen. The stiffness produced by the combination of layers and the inclusion of a cohesive elastic bandage produces high working pressures. However, the resting pressure is higher than with inelastic systems.

The sustained resting pressure produced by high stiffness elastic bandage systems may be useful when:

- the patient is immobile
- the ankle joint is fixed, ie the calf muscle pump cannot be used
- the patient has venous ulceration and lymphatic disease
- the patient has proven venous disease
- large volume loss is expected, ie to increase time worn.

Modifications for long-term or palliative use

MLLB can be modified to apply reduced pressure for long-term, palliative or night time use. In most cases, the bandages are applied using a spiral technique only. Materials include:

- cotton tubular bandage
- soft synthetic wool or foam padding
- cohesive or adhesive inelastic bandages – using fewer layers.

Self/carer bandaging

For selected patients, self bandaging or bandaging by a carer may be appropriate. The patient or carer needs good dexterity, a clear understanding of the technique involved, and to demonstrate proficiency in application. The bandaging technique would be modified as described for long-term management.

Self/carer bandaging may be helpful to patients with:

- pressure resistant lymphoedema
- obesity/larger limbs
- experience of treatment
- a desire to be actively engaged in their management
- refill not controlled by hosiery alone.

Patients may also choose self/carer bandaging to enhance comfort or for use at night when they wear a compression garment during the day.

ALLERGY AND MLLB

Where possible, tubular bandages with high cotton content should be used to avoid exposing the patient to potential allergens. Direct contact between skin and foams should be avoided.

BANDAGE CARE

Some components of the MLLB system can be washed and dried according to the manufacturer's instructions and reused. Over time, inelastic bandages will progressively lose their extensibility, which will increase their stiffness. Heavily soiled materials should be discarded. Cohesive and adhesive bandages should be discarded after use.

PRINCIPLES OF MLLB

Practical bandaging skills are important for the effective use of MLLB (Boxes 25 and 26).

Practitioners will be appropriately trained. The use of tailored foam pads requires training at specialist level.

Clear guidance is given for MLLB of the leg in Figures 26-33 and Box 27 (pages 35-37) and for MLLB of the arm in Figures 34-38 and Box 28 (pages 37-38).

BOX 25 Avoiding bandage slippage[72]

- Use foam to pad (more likely to stay in place than soft wool underpadding)
- Place narrow strips of foam between the inelastic bandage layers at the thigh to act as a brake
- Apply a cohesive or adhesive bandage in ≥ one layer, and particularly as the final layer
- Use ordinary noncompressive pantyhose over the bandage or suspenders attached to the proximal end of the bandages. This avoids changing the pressure gradient over the leg

BOX 26 Principles of MLLB

- Protect the affected area using tubular bandage and soft synthetic wool or foam underpadding
- Start bandaging distally and move proximally
- Guide bandages close to the limb using the entire hand to ensure good fit and to prevent creasing
- Always apply additional padding to the popliteal fossa and the inside of the elbow
- Apply inelastic bandages at full extension (lock-out point), except when applied to fingers and toes
- If elastic bandages are used, they are usually applied at 50% extension and with 50% overlap
- Use several layers of inelastic bandages to achieve the desired pressure
- Minimise creases at joints by bandaging the limb in a slightly flexed position and using figure of eight turns at the joint
- Extend partial limb bandaging beyond the area of swelling and ideally incorporate the knee or elbow joint to prevent proximal displacement of fluid into the joint
- Figure of eight bandaging increases the number of layers of bandage applied and results in higher sub-bandage pressures than spiral bandaging. Its use over the whole limb may be appropriate to reduce slippage or for inverted champagne bottle shaped legs, when high sub-bandage pressures are required
- Assess security of bandages and fixation, range of movement, circulation, sensation and level of comfort after application. Ask the patient to report bandage slippage and any change in digit sensation or colour
- The patient should be encouraged to contribute to the development of an individualised bandage system that fulfils their needs

MLLB OF THE LEG

FIGURE 26 Application of tubular bandage to lower leg
Apply a cotton tubular bandage next to the skin. The tubular bandage can be applied after toe bandaging, if indicated. If applied before toe bandaging, the tubular bandage should be folded back temporarily to allow access to the toes.

Box 27 Recommended materials for MLLB of the leg

- Cotton tubular bandage
- Toe bandages (if indicated) – 4cm conforming bandage
- Soft synthetic wool or soft foam roll (10cm or 20cm) or sheet
- Inelastic bandages – one 8cm, three to four 10cm for lower leg, and four to six 12cm for thigh

(a)

(b)

(c)

(d)

FIGURE 27 Bandaging the toes and foot
Toes should be bandaged if swollen. If not bandaged, the toes should be monitored and bandaged if they become swollen.
(a) Anchor the 4cm conforming bandage with one complete circle at the base of the toes.
(b) Take the bandage to the distal end of the big toe.
(c) Bandaging should be distal to proximal starting from the base of each toenail with a turn around the base of the toes before starting the next toe.
(d) Keep slight tension on the bandage. Avoid making creases on the underside of the toes. The little toe can be bandaged on its own, with the adjacent toe, or left unbandaged. On completion check that the bandage does not slip off, and check the toes for cyanosis and sense of touch.

FIGURE 28 Application of underpadding to lower leg
Apply soft synthetic wool padding to protect and reshape the limb. Soft foam underpadding can also be used.

(a)

(b)

(c)

(d)

FIGURE 29 Spiral bandaging of foot and lower leg with inelastic bandage
(a) Anchor an 8cm inelastic bandage with a turn around the base of the toes.
(b) Bandage the foot using spiral technique. Use figure of eight technique around the ankle. Continue up the leg using spiral technique with any remaining bandage.
(c) Bandage the lower part of the leg using a 10cm inelastic bandage and spiral technique, and continue up the limb.
(d) The end of the tubular bandage can be folded back and concealed under the next layer of bandage.

(a)

(b)

(c)

(d)

(e)

(f)

FIGURE 30 Spiral bandaging of the thigh with inelastic bandage
(a) If swelling occurs above the knee, the thigh should be bandaged. Ensure the cotton tubular bandage is long enough to cover the thigh.
(b) After bandaging the lower leg, allow the patient to stand with the knee slightly bent. Apply soft synthetic wool padding to the knee and thigh.
(c) At the popliteal fossa, double or triple the padding or apply a foam insert.
(d) Ask the patient to shift their weight to the leg to be bandaged, providing support if necessary, so that the thigh can be bandaged with the muscle contracted. Use a 10cm or 12cm inelastic bandage and apply a loose turn to anchor the bandage below the knee.
(e) After anchoring the bandage obliquely across the popliteal fossa, make a circular turn once around the distal aspect of the thigh. Then continue down to the starting point of the bandage, wrapping the flexed knee with figure of eight turns. Then wrap through the popliteal fossa over the patella using spiral technique.
(f) Continue the bandage up the thigh to the groin using spiral bandaging technique. The next layer is applied in the same way, but in the opposite direction.

Addressing specific problems

FIGURE 31 Padding skin folds
Deep skin folds can occur on the toes. Forefoot swelling may also be present. Skin folds must be padded. Bevel edged foam strips can be used. This is an area of treatment that is initiated and monitored by practitioners with training at specialist level

FIGURE 32 Forefoot swelling
Foam padding can be applied to the forefoot and fastened with a toe bandage to increase local pressure. This care is initiated and monitored by practitioners with training at specialist level, as it requires accurate use of appropriately cut foam.

FIGURE 33 Padding for retromalleolar oedema
Foam padding can aid oedema reduction around the malleoli.

MLLB OF THE ARM

BOX 28 Recommended materials for MLLB of the arm

- Cotton tubular bandage
- Finger bandages – 4cm conforming bandage
- Soft synthetic wool or soft foam roll (10cm)
- Inelastic bandages – one 6cm, one 8cm, and two to three 10cm

FIGURE 34 Application of tubular bandage
Apply a cotton tubular bandage, first cutting a hole for the thumb.

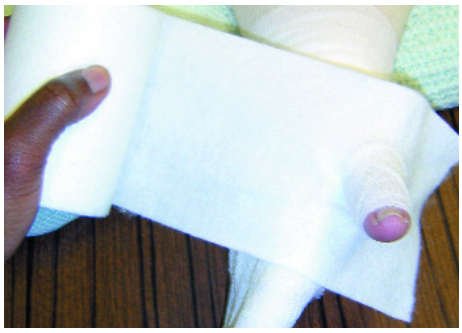
FIGURE 35 Finger and hand bandaging
(a) Begin with the palm of the hand facing down. Make one loose complete turn with the 4cm conforming bandage around the wrist to anchor it.
(b) Ask the patient to spread their fingers and thumb. Then begin to bandage the hand. Wrap each finger individually.
(c) Bring the bandage over the back of the hand to the fingertips without tension. Bandaging should be distal to proximal, leaving the fingertips uncovered. Make circular turns around each finger. Maintain light tension on the bandage.
(d) On completion check that the bandage does not slip off, and check digits for cyanosis and sense of touch.

(a)

(b)

(c)

(d)


(a)

(b)

(c)

(d)

FIGURE 36 Application of foam underpadding
(a) Start the soft synthetic wool or soft foam underpadding at the hand. Cut a hole for the thumb and anchor around the wrist.
(b) Apply extra padding to the palm of the hand by fanning padding back and forth over the palm to keep it in a natural open position. This helps to provide opposing pressure on the dorsum of the hand when the inelastic bandage is applied. Then proceed up the arm using spiral technique.
(c) Apply double or triple padding or a thin foam sheet to the inside of the elbow to protect it from the inelastic bandage.
(d) If a second padding bandage is required to cover the arm, overlap its beginning with the end of the first bandage.

(a)

(b)

(c)

(d)

FIGURE 37 Spiral bandaging of the arm with the inelastic bandage
(a) Begin with a 6cm inelastic bandage applied loosely at the wrist with one turn to anchor. For patients with small hands, a 4cm bandage may be used instead. Wrap the hand with the fingers spread. Use moderate tension on the bandage. Cover all of the hand including the knuckles and palm of the hand at the base of the thumb to mid palm.
(b) Use spiral technique to bandage the forearm with any remaining material. Overlap the second inelastic bandage (8cm or 10cm) with the end if the first. Bandage the forearm with the muscles tightened by asking the patient to make a fist. This is to prevent excess pressure increase in this part of the arm during active movement that might worsen venous and lymphatic return.
(c) Use figure of eight turns to bandage the elbow while it is slightly flexed. This further protects the inner elbow.
(d) Start the final inelastic bandage (10cm) at the wrist. Apply it using spiral technique in a reverse direction to cover the whole arm up to the armpit. This helps to maintain an optimal pressure gradient from the distal to proximal part of the arm.

FIGURE 38 Padding for dorsal and palmar oedema
Additional pressure can be applied to palmar and dorsal oedema by inserting foam padding that has been cut to shape and bevelled.

Compression garments

Correctly fitted compression garments should be prescribed appropriately for patients with lymphoedema.

The main use of compression garments is in the long-term management of lymphoedema, usually following a period of intensive therapy. Compression garments are also used for prophylaxis or as part of initial treatment. They may provide the only form of compression used, or form part of a regimen that includes other types of compression. Some patients wear garments during waking hours only, for exercise only, or up to 24 hours per day.

A wide variety of factors must be taken into account when determining whether a patient is suitable for compression garments[73] (Boxes 29 and 30).

BOX 29 Criteria indicating patient suitability for compression garments

- Good dexterity
- Intact, resilient skin
- No or minimal shape distortion
- Absent or minimal pitting oedema
- Swelling that can be contained by compression garments
- Concordant and motivated
- Ability to tolerate and manage hosiery (+/- carer support)
- Ability to monitor skin condition and engage in prevention strategies
- Symptom-based management/palliative needs

BOX 30 Contraindications to compression garments

- Arterial insufficiency - ABPI <0.5 in the lower limb
- Acute cardiac failure
- Extreme shape distortion
- Very deep skin folds
- Lymphorrhoea, or other weeping skin condition
- Extensive ulceration
- Severe peripheral neuropathy

Caution required: cellulitis/erysipelas (if tolerated, patients can continue garment use or switch to reduced pressure MLLB), sensory deficit, paralysis, fragile or damaged skin.

COMPRESSION GARMENT CONSTRUCTION

Compression garments can be categorised according to method of fabric manufacture[74]:

- Circular knit garments – the material is continuously knitted on a cylinder and has no seam, and is used mainly to make ready to wear garments. Garments are shaped by varying stitch height and yarn tension (Figure 39). Circular knit garments may be thinner and more cosmetically acceptable than flat knit garments.
- Flat knit garments – the material is firmer and thicker than that of circular knit garments. Garments are knitted as a flat piece that is shaped by adding or removing needles (Figure 40). The flat piece is then joined by a seam to form the garment. Most custom made garments are made from flat knit material.

COMPRESSION GARMENT STANDARDS

National standards for compression garments are usually prerequisites for reimbursement and cover parameters such as testing methods, yarn specification, compression gradient and durability. Existing standards do not cover compression garments other than hosiery, eg they do not cover arm sleeves, and differences in class pressure ranges and testing equipment make comparisons between standards difficult (Table 6)[74]. Furthermore, practitioners should be aware

TABLE 6 Comparison of hosiery classification in the British, French and German compression hosiery standards[74] The mmHg ranges refer to the pressures applied at B (ankle circumference at smallest girth) by the compression hosiery. NB There are no national standards for compression hosiery in the USA; the compression classification used most widely there is: Class 1 20-30mmHg; Class 2 30-40mmHg; Class 3 40-50mmHg and Class 4 50-60mmHg. Hosiery is also available in the USA in a 15-20mmHg pressure range.

	British standard BS 6612:1985	French standard ASQUAL	German standard RAL-GZ 387:1987
Testing method	HATRA	IFTH	HOSY
Class I	14–17mmHg	10–15mmHg	18–21mmHg
Class II	18–24mmHg	15–20mmHg	23–32mmHg
Class III	25–35mmHg	20–36mmHg	34–46mmHg
Class IV	Not reported	>36mmHg	>49mmHg

FIGURE 39 Circular knit knee high garment
Shaped by varying yarn tension and stitch height during knitting.

FIGURE 40 Flat knit knee high garment before sewing
Shaped by adding and removing needles during knitting.

that some manufacturers' compression class pressure ranges for lower limb hosiery may be different from the compression class ranges used for upper limb garments. To assist comparison, therefore, garment packaging and studies involving compression garments should state the pressure ranges within the classes quoted and the testing method used to determine the pressures.

LIMB SHAPE AND GARMENT CHOICE

Limb shape plays an important role in choosing compression garments. Ready to wear compression garments are suitable where there is no or minimal limb distortion, but can be more difficult to fit precisely and, if circular knit, may roll at the top. Custom made garments can be made to accommodate a wide range of anatomical distortion. Flat knit garments do not roll, curl, twist or tourniquet, can achieve a better fit, and can be made with zippers to aid application.

FITTING COMPRESSION GARMENTS

C Compression garments for patients with lymphoedema should be fitted by appropriately trained practitioners.

Prescription of compression garments should only be undertaken after full assessment of the patient, and should take into account factors such as the stage and severity of the lymphoedema, the patient's comfort, preferences, lifestyle, psychosocial status, concurrent disease, and ability to apply and remove garments. Patients with skin problems such as dermatitis or psoriasis and those with known allergies to substances like elastane benefit from the use of cotton rich garments.

Patients should be measured for garments when swelling has been minimised, pitting oedema is absent or minimal, any shape distortion optimised and the area stabilised (Box 31).

Accurate measurement is important to achieve correct fit of ready to wear and custom made garments. Measurements required will usually include circumferential measurements at several given sites and longitudinal measurements between specified points (Figures 41 and 42). The prescription should also specify style, knitted texture and any fixation or attachment (Box 32).

Measurement for ready to wear or custom made compression garments requires that the practitioner has appropriate training, and access to a practitioner with training at specialist level.

NOTE: FIGURES 41-42

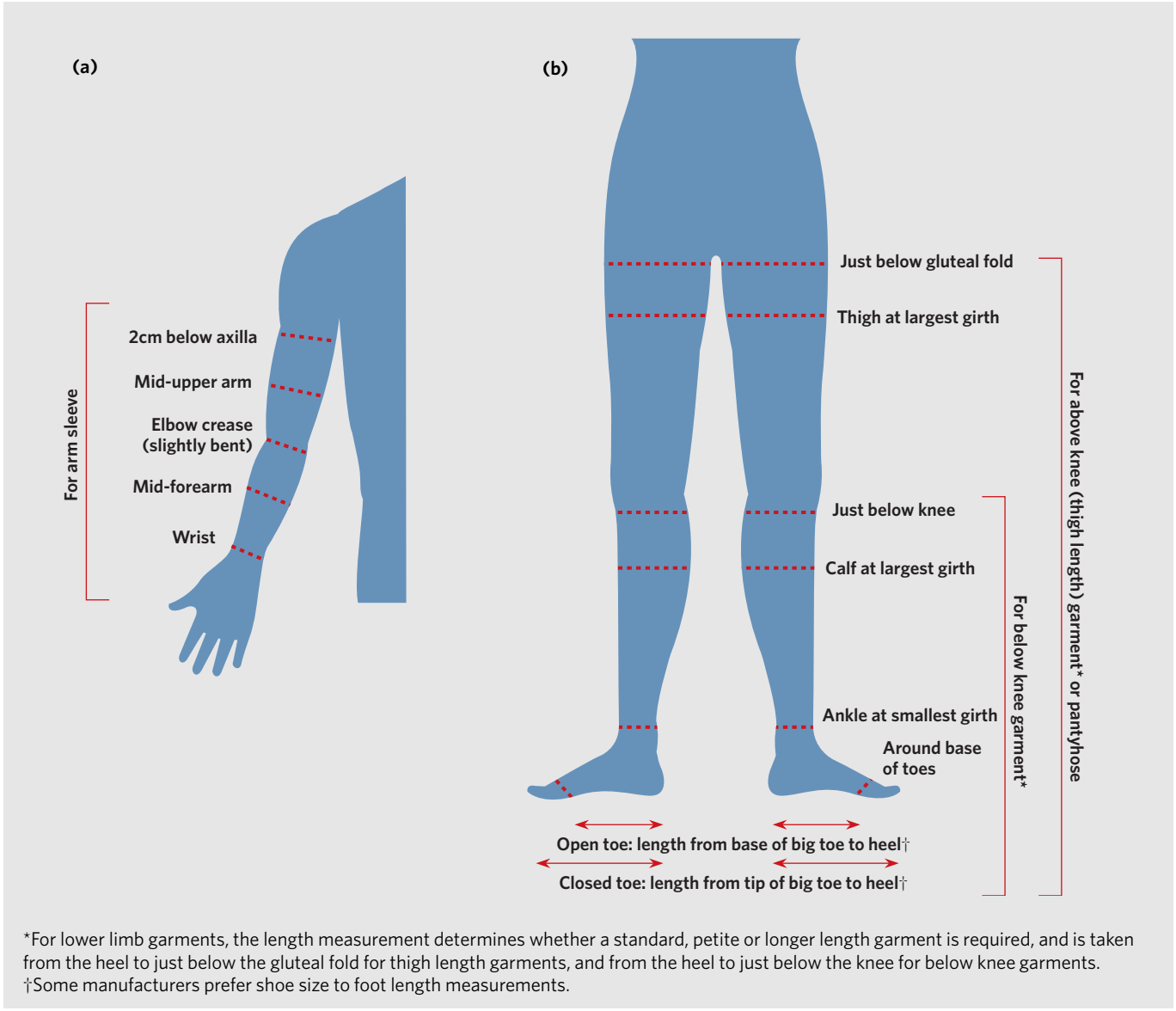
These figures provide a guide to measuring for compression hosiery. Careful attention should be paid to the specific measuring instructions of the manufacturers from which garments are ordered.

BOX 31 Tips for compression garment measurement

- Measure when the area is largely free of pitting oedema, ie immediately after removal of compression bandages, or in the morning before swelling can develop
- The measuring tape should be pulled firmly, but not so tightly that it indents the skin
- Measure with the patient in the recommended position
- Continue bandaging until the patient has received the prescribed garments

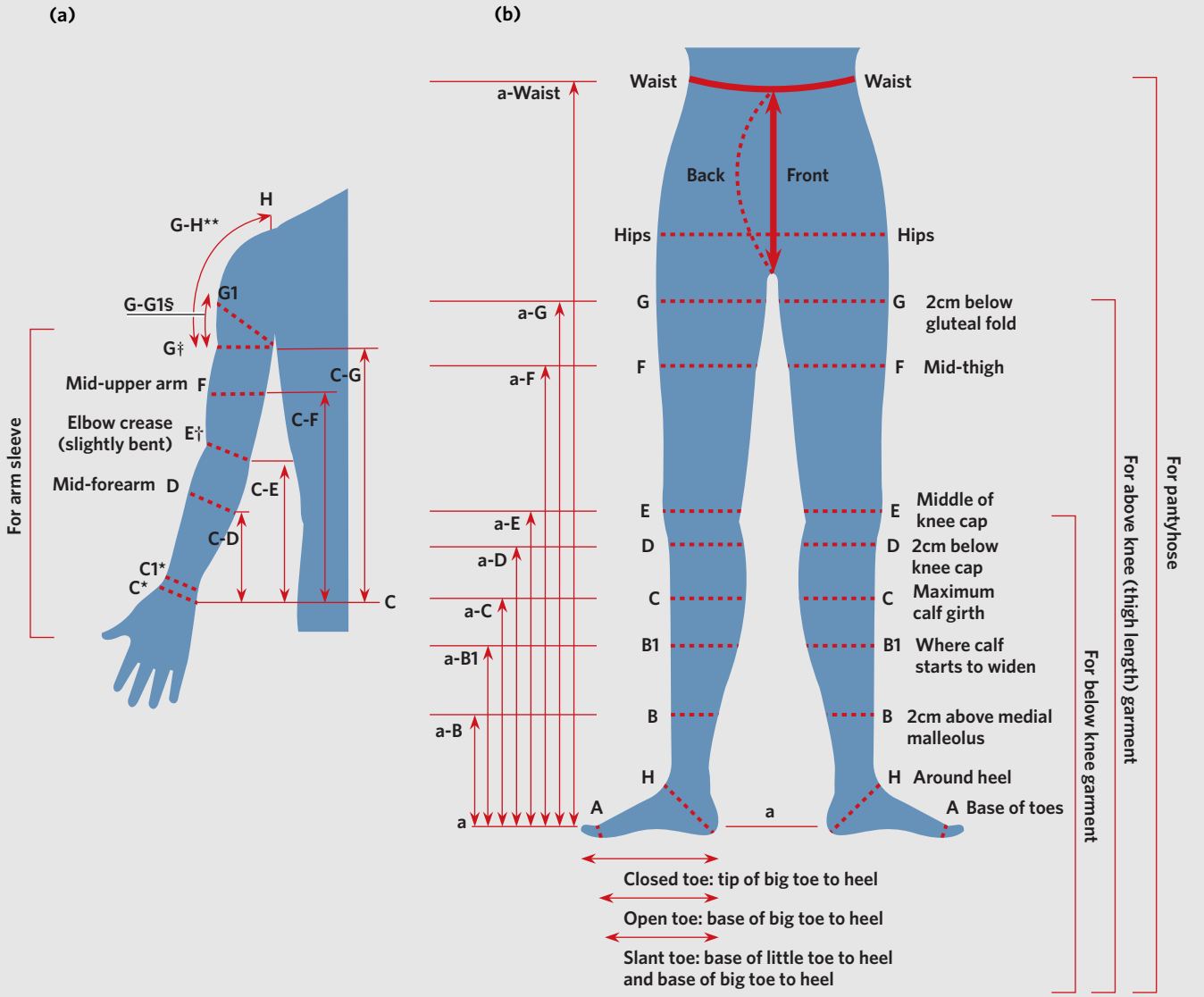
BOX 32 Components of a compression garment prescription

- Quantity of garments (at least two – one for wearing, one for washing)
- Manufacturer, style and garment code
- Level of compression required
- Knitted texture, ie circular knit or flat knit
- Length
- Fixation and attachment, if needed, eg silicone top, waist attachment
- For ready to wear garments, state size
- For custom made garments, provide measurements required by the manufacturer
- Sex of the patient
- Colour



*For lower limb garments, the length measurement determines whether a standard, petite or longer length garment is required, and is taken from the heel to just below the gluteal fold for thigh length garments, and from the heel to just below the knee for below knee garments.
†Some manufacturers prefer shoe size to foot length measurements.

FIGURE 41 Measurements for ready to wear compression garments for limbs
Circumferential measurements are taken at the levels indicated.
(a) Upper limb
Measurements may be taken while the patient is sitting comfortably with the arm supported. The length measurement is taken along the inside of the arm from the wrist to 2cm below the axilla to determine whether a standard or longer length garment is required.
(b) Lower limb
According to patient mobility and the circumstances in which the measuring is taking place, measurements may be taken while the patient is standing, lying or sitting. In ideal conditions, measurements from the foot to the knee may be taken while the patient is lying on a couch, and measurements above the knee while the patient is standing. A measuring board should be used if available.



* To find C, ask the patient to flex the wrist. Use the level of the second crease from the hand to measure circumference C. C1 is about 3cm proximal to C.
† Measure circumference E at the elbow crease with the elbow slightly bent. Measure again 1-2cm proximal to E. If this circumference is larger than the E measurement, record this as E.
‡ To measure circumference G, ask the patient to place a piece of paper in the axilla to show where they would like the garment to finish while putting the arm at their side. Fold the paper around the arm and mark the level of G at the top edge of the paper. When measuring circumference G do not apply any tension to the tape.
§ Measure length G-G1 for bias top.
**Measure length G-H for shoulder attachment.

FIGURE 42 Measurements for custom made compression garments for limbs
Circumferential and longitudinal measurements are taken as indicated for the style of garment required.
(a) Upper limb
Measurements may be taken while the patient is sitting comfortably with the arm supported. Length measurements are taken along the inside of the arm.
(b) Lower limb
According to patient mobility and the circumstances in which the measuring is taking place, measurements may be taken while the patient is standing, lying or sitting. In ideal conditions, measurements from the foot to the knee may be taken while the patient is lying on a couch, and measurements above the knee while the patient is standing. A measuring board should be used.

Checking fit

A trained practitioner should check that a newly prescribed garment is as ordered, fits properly and fully covers the area requiring treatment. Initial fitting should include a demonstration of how to put on and remove the garment, and observation and assessment of the patient's/carer's application and removal techniques. Clear verbal and written instructions should be given on errors of fit that may be discovered after first wearing, and on how to care for the garment (Box 33).

BOX 33 Application and care of compression garments

Patients and carers should be advised that:

- All folds and wrinkles should be removed. This can be assisted by wearing household rubber gloves whilst smoothing the garment
- Emollients may damage compression garments. Ensure emollient is absorbed before donning garment or use products approved by the garment manufacturer. A cotton liner can be used if emollient is applied just before donning, the skin is at risk of trauma, or there is dermatitis
- Compression garments should not be worn with the top folded down
- Garments combining an armsleeve and gauntlet should not be worn with the handpiece folded back
- Any distortion in limb shape, skin redness/damage/discolouration, or peripheral swelling may indicate garment unsuitability
- The garment should be removed immediately if problems occur and the patient should contact their practitioner
- Garments should be washed frequently according to the manufacturer's instructions (performance may be impaired by infrequent washing)

At follow up visits, the practitioner should check that the patient is concordant with garment wear, that the garment has not been altered, and that swelling is not occurring proximal or distal to the garment.

Avoiding problems

Garment slippage can be overcome in a number of ways (Box 34). A variety of aids is available for easing application of compression garments (Box 35). Aids also exist to assist with garment removal.

Oily or greasy emollients can damage compression garments and make garment application difficult. A cotton underlayer can be used to assist application and minimise damage.

Garment replacement

Garments should be replaced every three to six months, or when they begin to lose elasticity. Young or very active patients may require more frequent garment replacement.

ALLERGY AND COMPRESSION GARMENTS

Patients may develop an allergy to compression garments. Allergens include fabric dye, latex and nylon. If an allergy is suspected:

- treat contact dermatitis appropriately
- use garments without latex
- use garments with high cotton content, or that have double covered yarns to limit skin contact with elastic components
- consider the use of a cotton tubular bandage underlayer (which must be unwrinkled during wear) or a garment with an inbuilt lining.

COMPRESSION GARMENTS FOR LIMBS

The following recommendations for compression garments for the lower limb (Figure 43 and Table 7) and for the upper limb (Table 8) have been developed by the British Lymphology Society compression garments group and the Lymphoedema Framework working groups.

Patients with severe shape distortion may find flat knit garments more appropriate. However, the finer finish of circular knit hosiery may make it more cosmetically acceptable.

If the patient in unable to tolerate the therapeutically indicated level of compression, lower pressure garments may be necessary to encourage concordance.

Tolerability of high levels of compression may be enhanced by layering garments.

BOX 34 Avoiding compression garment slippage

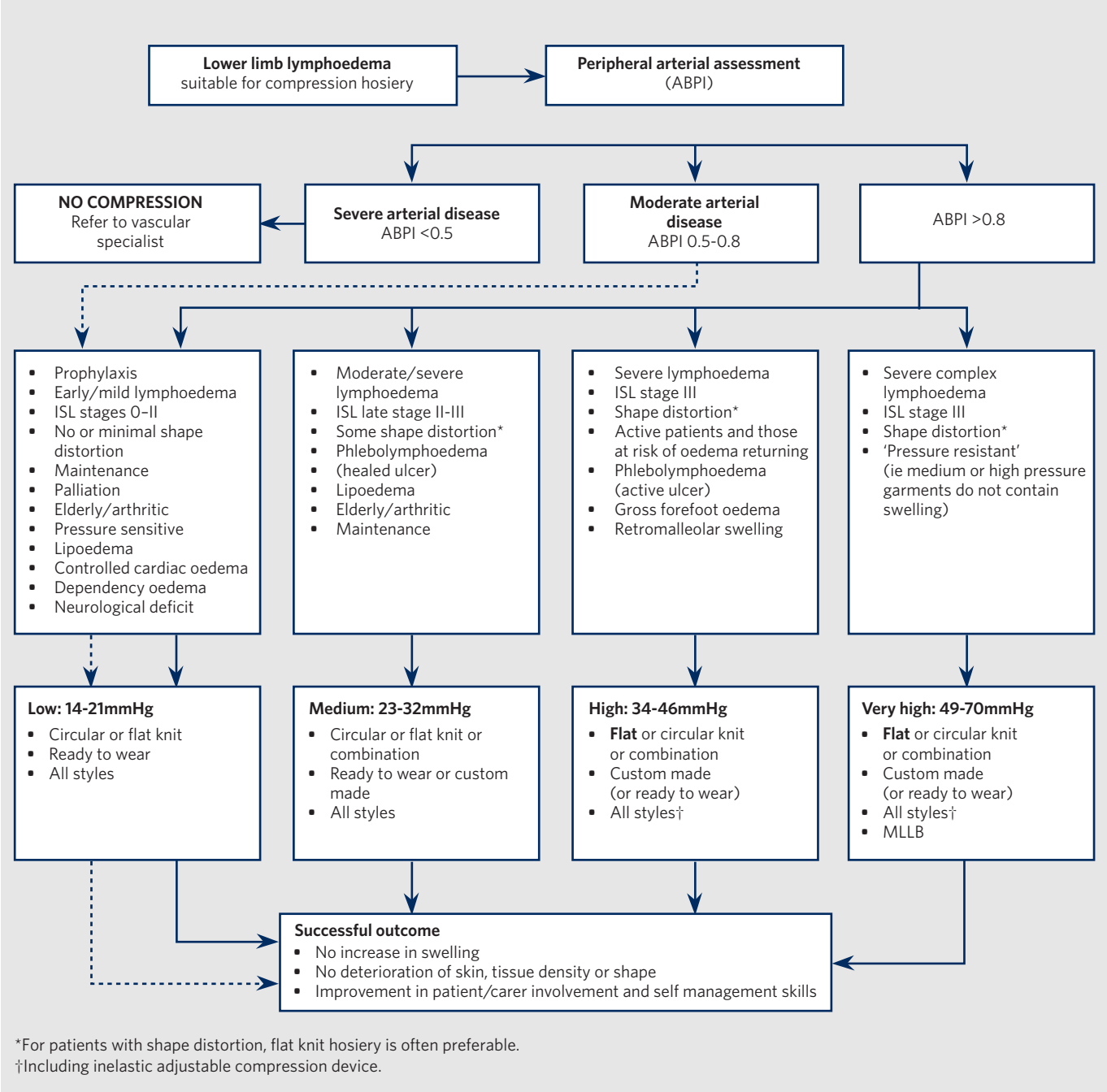
Ensure garment fits correctly
Ensure style is appropriate
Consider:

- skin glue or surgical adhesive tape
- silicone coated band at top edges
- fixation mechanism - eg waist fastening/half panty/full panty/shoulder cap/bra attachment and strap

BOX 35 Application aids

- Garment application gloves
- Glide on applicator
- Silk slippers
- Anti-slip mat
- Metal applicator frames

FIGURE 43 Compression garments for lower limb lymphoedema/lymphovenous oedema, adapted from[73]



*For patients with shape distortion, flat knit hosiery is often preferable.
†Including inelastic adjustable compression device.

Layering compression garments

The practice of layering compression garments has been described in the management of lymphoedema[12], but there is little evidence of its efficacy. Two layers of garment produce a higher pressure on the limb and are stiffer than one garment. The second layer is likely to add about 70% of the pressure it would when applied alone[75].

Patients may find that wearing an additional garment layer can help to manage exacerbations of their condition. Furthermore, patients who have difficulty applying a single higher compression garment may be able to manage to apply two layers of a lower compression garment. When layering two garments, it is recommended that a flat knit garment is used next to the skin and that the outer layer is a circular knit garment.

TABLE 7 Compression garment recommendations for specific problems in lower limb lymphoedema

Problem	Recommendations/notes
Swollen toes	Where toe caps are difficult to manage, closed toe garments may be helpful
Forefoot swelling	No risk of toe swelling – use open toe garments; flat knit is preferable Toe swelling – use open toe garment and toe caps, unless toe caps are impractical, when a closed toe garment may suffice Lymphoedema of the foot only – inelastic adjustable foot wrap may be useful
Forefoot bulge	Custom made flat knit garments may be required to produce sufficient pressure An individually shaped foam pad can apply additional pressure Inelastic adjustable footwrap may be useful Check that footwear is well-fitting and supportive
Retromalleolar swelling	Foam, crescent shaped stasis pads can be used to focus pressure
Fat/arthritic knees	Low classification pantyhose under a calf stocking may be useful for shape distortion of the knee and thigh If using circular knit, use an extra wide calf range
Thickened tissue just below patella	Below knee garments can exacerbate the problem; ideally use full leg garments Pressure can be focused by using a crescent shaped ribbed or foam chip stasis pad over thickened area If a below knee compression garment is necessary, a stasis pad can be used with an orthopaedic elasticated knee support
Inverted champagne bottle legs	Limb shape should be corrected with MLLB Flat knit appears to be more effective than circular knit May need higher pressure levels May need custom made garments If using two garment layers, use a combination of flat knit and circular knit
Lymphoedema extends to groin	Flat knit custom made garments, eg one- or two-legged closed gusset panty, should be used A foam chip pad angled into the groin under the compression garment may be used to focus pressure Close fitting shorts with Lycra (eg cycle shorts) are convenient for some patients
Obesity	May need custom made garments; flat knit may be easier to apply Garments designed to accommodate pregnancy may be useful Severe distortion of the lower limb or patient preference may restrict treatment to the lower part of the leg Using separate overlapping garments for above and below the knee may make application easier

SAFETY ISSUES

Lower limb peripheral arterial occlusive disease

The lower limb peripheral arterial status of patients with lower limb lymphoedema should be assessed prior to compression. Patients with ABPI <0.5 should not receive compression and should be referred to a vascular specialist.

Risk reduction

Patients should be advised to wear compression garments when performing high risk, repetitive activities. Although there is no robust evidence that long sitting while travelling, eg by aeroplane, increases or precipitates lymphoedema, patients should exercise caution and wear a compression garment if they are at risk of or have lymphoedema.

TABLE 8 Compression garments for upper limb lymphoedema

Indications	Compression garment classification	Recommendations	Notes
Prophylaxis Mild lymphoedema ■ ISL stage I-II ■ No shape distortion ■ Maintenance ■ Palliation	LOW 14-18mmHg	Circular or flat knit Ready to wear*	Application aids may be required by less dextrous and elderly patients If patient will be travelling by aeroplane, it is advisable to prescribe a handpiece as well as a sleeve
Moderate lymphoedema ■ ISL late stage II-III ■ Some shape distortion	MEDIUM 20-25mmHg	Circular or flat knit Ready to wear or custom made*	Garments can be made that incorporate pads to treat areas of thickened tissue Silk inserts can be used at the inner elbow if irritation and trauma occur
Severe lymphoedema ■ ISL stage III ■ Major shape distortion	HIGH 25-30mmHg	Circular or flat knit Custom made*	Such high pressure is required only in exceptional cases

*All upper limb styles including gloves and gauntlets and inelastic adjustable compression devices.

COMPRESSION GARMENTS FOR MIDLINE LYMPHOEDEMA

Compression garments can be used to treat lymphoedema of the head and neck, breast, trunk or genitalia. These garments may be custom made or ready to wear. Garments for the torso are usually classified as providing medium compression (25-30mmHg), while lower pressures are used on the head. However, there is no recognised agreement on the appropriate level of compression for these patients.

Leotard or bodice style garments may be useful for patients with truncal oedema and flat knit construction is preferable. Patients with breast lymphoedema may require a ready to wear or custom made bra. For patients with scrotal swelling, scrotal supports can be used. Anatomically contoured foam padding inserted into compression pantyhose or shorts can be used in female genital lymphoedema. Groin swelling is often accompanied by tissue thickening, and may occur in combination with lower limb lymphoedema; one- or two-legged closed gusset pantyhose angled across the groin with foam chip stasis pads may be helpful.

OTHER COMPRESSION DEVICES

Inelastic adjustable compression devices are available for the treatment of lymphoedema. The compression the device applies can be adjusted by altering how tightly the straps used to fix the garment in place are pulled. They can be used to contain swelling in patients with moderate or severe lymphoedema of the upper or lower limb and the torso, and are useful self management tools.

Exercise/movement and elevation

Exercise/movement are common rehabilitative interventions used to reduce oedema. At present, there is little evidence to indicate which types, intensities and frequencies of exercise may be safely used in the management of lymphoedema.

EFFECTS OF EXERCISE/MOVEMENT

Exercise improves muscular strength, cardiovascular function, psychological wellbeing and functional capacity. Gentle resistance exercise stimulates muscle pumps and increases lymph flow; aerobic exercise increases intra-abdominal pressure, which facilitates pumping of the thoracic duct[76].

TAILORED EXERCISE/MOVEMENT PROGRAMMES

Combinations of flexibility, resistance and aerobic exercise may be beneficial in controlling lymphoedema[77-79], and should be tailored to the individual patient (Box 36). Physiotherapy referral is required for patients who have difficulty with mobility, joint function or joint movement.

ELEVATION

Elevation of the affected limb, ideally to just above the level of the heart, is often advised to reduce swelling. It is thought that elevation acts by maximising venous drainage and by decreasing capillary pressure and lymph production.

BOX 36 General guidelines on exercise

- Patients should be encouraged to maintain normal functioning, mobility and activity
- Exercise/movement should be tailored to the patient's needs, ability and disease status
- Patients should be encouraged to include appropriate warming up and cooling down phases as part of exercise to avoid exacerbation of swelling
- Compression should be worn during exercise
- Expert patients can help to demonstrate, teach and monitor exercise, and provide information on access to local exercise programmes

Types of exercise:

- start with low to moderate intensity exercise
- paralysed limbs can be moved passively
- walking, swimming, cycling and low impact aerobics are recommended
- heavy lifting and repetitive motion should be avoided
- flexibility exercises maintain range of movement

Anecdotal evidence suggests that limb elevation when the patient is sitting or in bed may be a useful adjunct to active treatment, but should not be allowed to impede function or activity. Patients should be encouraged not to sleep in a chair and to go to bed at night to avoid the development of 'arm chair' legs or exacerbation of lower limb lymphoedema.

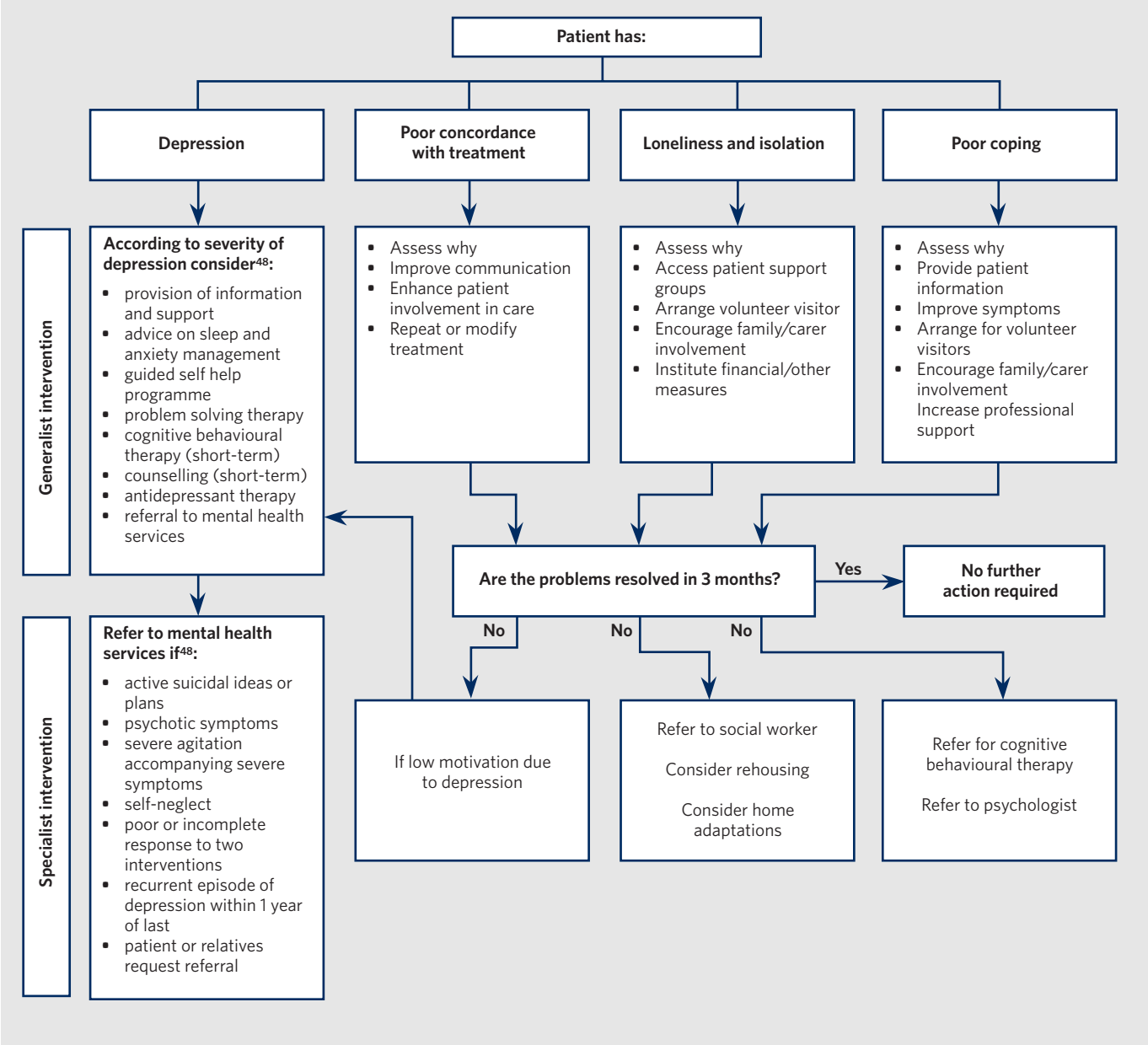
Psychosocial support

Psychosocial support is an important element of the holistic treatment of lymphoedema: it has the potential to have considerable influence on outcome by enhancing concordance, encouraging self-management and maximising quality of life.

Intervention involves planning and implementing psychosocial care strategies that help patients and their family/carers to take a positive role in the management of their lymphoedema and to achieve as good a quality of life as possible (Figure 44).

If psychosocial problems are not resolved within three months, the patient should be referred for specialist intervention.

FIGURE 44 Algorithm to address psychosocial problems



Palliative care

The needs of patients with lymphoedema who are otherwise ill with advanced disease and who require palliative care can be complex. This document can provide only an indication of supportive measures and treatments that may be helpful.

Lymphoedema can produce distressing and debilitating symptoms that affect lifestyle and function. Patients with advanced disease may not be able to tolerate a full programme of assessment and treatment, but require a palliative approach in which assessment techniques are modified and individual treatments are selected to ease specific symptoms (Box 37 and Table 9).

BOX 37 Guide to selection of treatment in advanced disease

- Ascertain type and cause of oedema, and contributory factors
- Identify levels of symptoms such as pain
- Establish significance of the swelling to the patient and consider patient circumstances and perspective
- Establish realistic goals
- Consider response to treatment

TABLE 9 Management of lymphoedema in patients with palliative care needs

Problem	Intervention
Unable to tolerate full assessment procedures	■ Use modified monitoring and limb volume measurement techniques
Fragile or dry skin	■ Maintain skin integrity – refer to skin management guidance
Discomfort in a swollen limb	■ Reduced compression MLLB with modification to materials used ■ Low pressure compression garments
Swollen limb due to dependency or inactivity, or mainly venous oedema of lower limbs with no truncal oedema	■ Good skin care and guidance on limb positioning ■ Gentle passive or active exercises ■ Reduced compression MLLB ■ Low pressure compression garment ■ IPC ■ Refer to physiotherapist
Severe limb or digit swelling	■ Good skin care ■ Reduced compression MLLB with modification to materials used
Swelling of scrotum and/or penis	■ Close-fitting shorts with Lycra to provide scrotal support ■ Custom made garments and scrotal support for use by ambulant patients ■ Scrotal bandaging ■ Teach SLD
Swelling of female genitalia	■ Lycra shorts with 1cm thick anatomically contoured foam pads ■ Flat knit custom made shorts with foam pads ■ Compression tights with localised padding ■ Teach SLD
Truncal oedema	■ MLD by practitioner with training at specialist level ■ Teach SLD ■ Supportive garments, eg bodice or bra for comfort
Lymphorrhoea	■ Good skin care and guidance on limb positioning ■ Modified reduced compression MLLB
Loss of independence and restricted mobility	■ Teach self care measures ■ Teach SLD ■ Refer to occupational therapist or physiotherapist as required ■ Appropriate psychological intervention ■ Low classification compression garments or shaped tubigrip if compression garments not tolerated

Surgery

BOX 38 Potential indications for surgery in lymphoedema[81-84,92-94]

- Severe deformity or marked disability due to swelling
- Removal of redundant tissue after successful conservative therapy
- Proximal lymphatic obstruction with patent distal lymphatics
- **Lymphocutaneous fistulae** and **megalymphatics**
- Eyelid and external genital lymphoedema
- Lack of response to compression therapy
- Recurrent cellulitis/erysipelas
- Intractable pain
- Lymphangiosarcoma

Lymphocutaneous fistulae: abnormal connections between the lymphatic system and the surface of the skin; may leak large quantities of lymph
Megalymphatics: large, dilated incompetent lymph vessels that allow lymphatic reflux

Surgical treatment of lymphoedema can be divided into three main categories:

- surgical reduction
- procedures that bypass lymphatic obstructions
- liposuction.

Patients for surgery need to be selected carefully (Box 38) and counselled to ensure realistic expectations of likely outcome. Maintenance of any improvement gained requires long-term postsurgical compression therapy.

SURGICAL REDUCTION

Surgical reduction (sometimes also known as debulking surgery) aims to remove excess subcutaneous tissue and skin, and may be useful in the symptomatic treatment of severe lymphoedema. However, the postsurgical morbidity of reduction operations may be considerable[80,81]. In some cases, surgical reduction may be considered for lymphoedema of the eyelid or genitalia.

BYPASS OPERATIONS

Bypass operations aim to restore lymphatic function through lymphovenous anastomoses and lymphatic or venous vessel grafting, or lymph node transplantation[82]. Anastomosis of lymph vessels to the venous system may be attempted in patients with proximal lymphatic obstruction and patent distal lymphatics, and produces better results at earlier stages of lymphostatic disease[83-85]. Lymphatic grafting and lymph node transplantation require microsurgical techniques, and show promising results in carefully selected patients[86,87].

LIPOSUCTION

In patients with chronic lymphoedema, adipocyte proliferation (which may be related to an inflammatory process) may mean that conservative treatment or microsurgery do not completely resolve limb enlargement[88].

Liposuction has been performed on patients with long-standing breast cancer related lymphoedema. It removes excess fat tissue and is considered only if the limb has not responded to standard conservative therapy. Liposuction does not correct inadequate lymph drainage and is not indicated when pitting is present. Where concordance with compression garments after treatment is high, results have been maintained[89,90]. Liposuction has also been used for primary and secondary leg lymphoedema with promising results[91].

Other treatments

A variety of other treatment modalities may be used to treat lymphoedema; many require further evaluation (Box 39). National use of these treatments is variable.

DRUG TREATMENT

Two main groups of drug have been used in the treatment of lymphoedema: benzopyrones and diuretics.

Benzopyrones

Benzopyrones are based on a variety of naturally occurring substances. Examples include flavonoids, oxerutins, escins, coumarin, and ruscogen combined with hesperidin.

There is little evidence to support the use of these drugs in lymphoedema[1,95]. There is some data, however, that flavonoids may stabilise swelling by reducing microvascular filtration[96].

Oxerutins have been licensed in some countries, usually for use in chronic venous insufficiency, but there are insufficient data to draw conclusions about their efficacy in lymphoedema. The same conclusion has been reached about flavonoids. Coumarin has been most widely trialled, but the most recent study reported no significant effect[95] and the drug has been withdrawn in Australia because of liver toxicity.

Diuretics

Diuretics encourage the excretion of salt and water, and by reducing blood volume might be expected to reduce capillary filtration and lymph formation. There is no evidence that diuretics encourage lymph drainage.

A diuretic is likely to be prescribed on a pragmatic basis for anyone with oedema almost irrespective of cause. However, higher doses of thiazides or loop diuretics (eg furosemide or bumetanide) can reduce body potassium levels with long-term use and may cause muscle weakness, promote oedema formation and affect the heart.

Diuretics are not recommended for use in the treatment of lymphoedema. Occasionally, short courses may be of benefit in chronic oedema of mixed aetiology, and in older patients in whom enhanced lymphatic drainage as a result of lymphoedema therapy precipitates cardiac failure.

BREATHING EXERCISES

Breathing exercises are recommended by some clinicians as a preliminary manoeuvre that may help to clear the central lymphatics prior to interventions that promote lymph drainage from the peripheries[97]. However, other clinicians question the physiological basis of breathing exercises as there are no experimental data in humans to confirm that variations in intrathoracic pressure due to breathing assist central lymphatic drainage into the venous system.

Although a recent human study demonstrated that a combination of exercise and deep breathing significantly reduced the volume of lymphoedematous limbs[79], evidence is lacking of the effect of breathing exercises in isolation. Nonetheless, breathing exercises are not harmful, are inexpensive, and may be proven beneficial in some groups of patients with lymphoedema.

LYMPHOEDEMA TAPING

Lymphoedema taping is an emerging form of treatment for lymphoedema. It involves the application of narrow strips of elastic tape to the affected area, and can be used in combination with compression garments or bandaging. It is thought to improve muscle function and lymph flow[98] and may have a role to play in the treatment of midline and peripheral swelling. However, evidence is lacking of its efficacy in lymphoedema.

HYPERBARIC OXYGEN

Hyperbaric oxygen therapy is known to promote healing in bone that has become ischaemic following radiotherapy. In patients with upper limb lymphoedema following radiotherapy, two small studies have indicated that hyperbaric oxygen may improve lymph flow and reduce limb volume in the short-term[99,100]. Further research is required to establish whether benefits can be demonstrated in randomised trials and in the long-term.

LASER THERAPY

Low level laser therapy has shown potential for the treatment of lymphoedema, particularly of the upper limb, where it has reduced limb volume and tissue hardness[101]. Further research is required to establish the benefits of treatment and the optimal regimen.

BOX 39 Other treatments

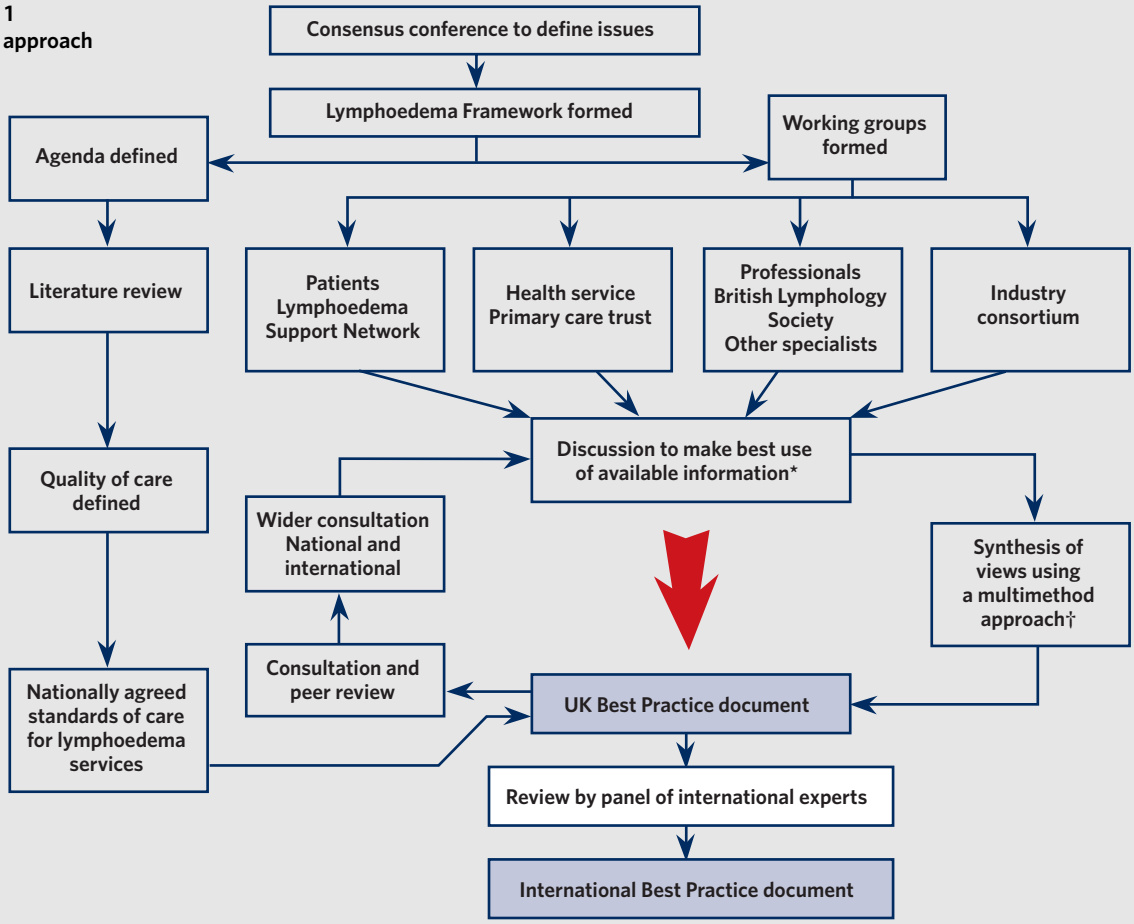
Other treatments that have been used for the treatment of lymphoedema, mainly in breast cancer patients, that all require further evaluation, include:

- cryotherapy
- transcutaneous electrical nerve stimulation (TENS)
- pulsed magnetic fields, vibration and hyperthermia
- thermal therapy
- ultrasound
- complementary medicine

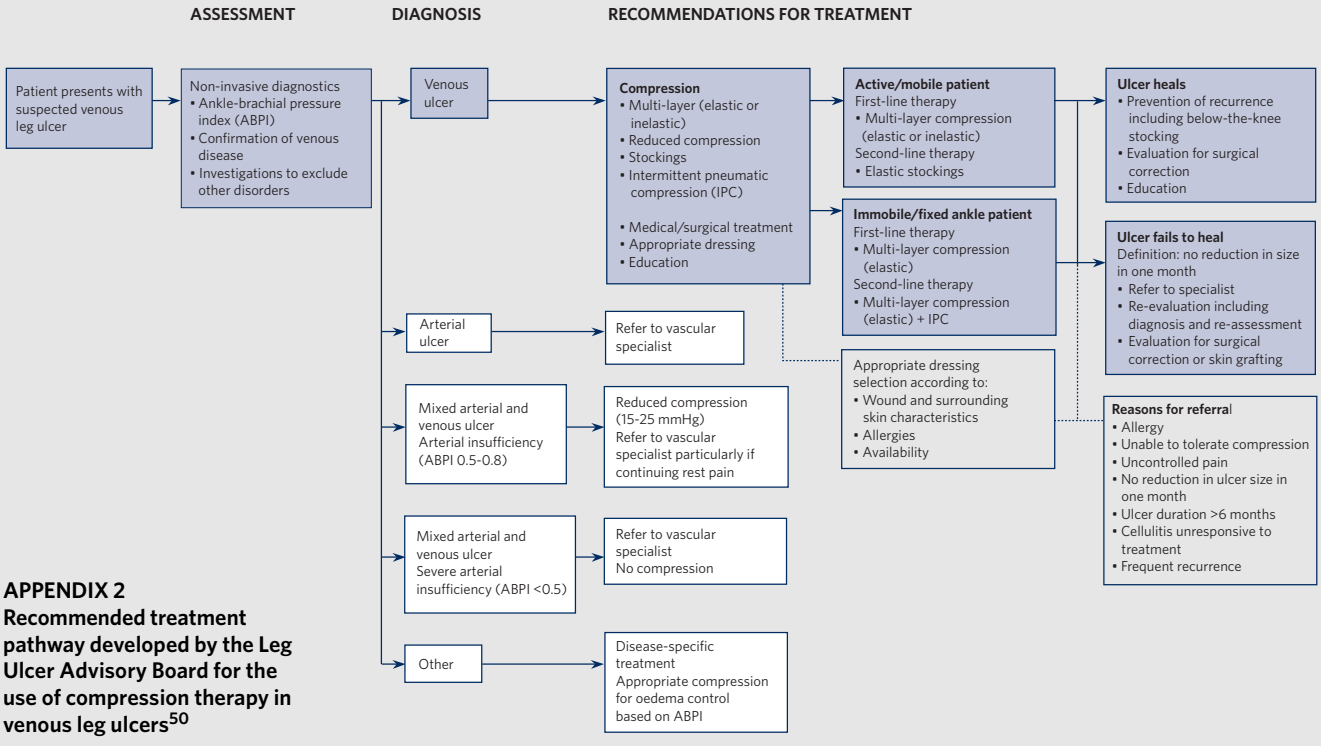
RECOMMENDED READING

- Badger C, Preston N, Seers K, Mortimer P. Benzo-pyrones for reducing and controlling lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 2: CD003140.
- Badger C, Preston N, Seers K, Mortimer P. Physical therapies for reducing and controlling lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 4: CD003141.
- Badger C, Seers K, Preston N, Mortimer P. Antibiotics/anti-inflammatories for reducing acute inflammatory episodes in lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 2: CD003143.
- Browse N, Burnand K, Mortimer P. *Diseases of the Lymphatics*. London: Arnold, 2003.
- European Wound Management Association (EWMA). Focus Document: *Lymphoedema bandaging in practice*. London: MEP Ltd, 2005.
- European Wound Management Association (EWMA). Position Document: *Understanding compression therapy*. London: MEP Ltd, 2003.
- Földi M, Földi E, Kubik S (eds). *Textbook of Lymphology for Physicians and Lymphedema Therapists*. San Francisco: Urban and Fischer, 2003.
- Lymphoedema Framework. *Template for Practice: compression hosiery in lymphoedema*. London: MEP Ltd, 2006.
- Olszewski WL. *Lymph Stasis: pathophysiology, diagnosis and treatment.* Boca Raton: CRC Press, 1991.

APPENDIX 1
Consensus approach



*Information used: published data, systematic reviews, national and European guidelines
†Multimethod approach: face to face discussion, structured interaction, formal group feedback, mailed questionnaires



APPENDIX 2
Recommended treatment pathway developed by the Leg Ulcer Advisory Board for the use of compression therapy in venous leg ulcers[50]

References

1. Badger C, Preston N, Seers K, Mortimer P. Benzo-pyrones for reducing and controlling lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 2: CD003140.
2. Badger C, Preston N, Seers K, Mortimer P. Physical therapies for reducing and controlling lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 4: CD003141.
3. Badger C, Seers K, Preston N, Mortimer P. Antibiotics/anti-inflammatories for reducing acute inflammatory episodes in lymphoedema of the limbs. *Cochrane Database Syst Rev* 2004; 2: CD003143.
4. Murphy MK, Black NA, Lamping DL, et al. Consensus development methods, and their use in clinical guideline development. *Health Technol Assess* 1998; 2(3): i-iv, 1-88.
5. Standards of Practice for Lymphoedema Services. *Lymphoedema Framework Journal* 2003; 1: 10-18.
6. International Society of Lymphology. The diagnosis and treatment of peripheral lymphedema. Consensus document of the International Society of Lymphology. *Lymphology* 2003; 36(2): 84-91.
7. Tobin MB, Lacey HJ, Meyer L, Mortimer PS. The psychological morbidity of breast cancer-related arm swelling. *Cancer* 1993; 72(11): 3248-52.
8. De Godoy JMP, Braile DM, de Fatima Godoy M, Longo O Jr. Quality of life and peripheral lymphedema. *Lymphology* 2002; 35(2): 44-50.
9. Williams AF, Moffatt CJ, Franks PJ. A phenomenological study of the lived experiences of people with lymphoedema. *Int J Palliat Nurs* 2004; 10(6): 279-86.
10. McWayne J, Heiney SP. Psychologic and social sequelae of secondary lymphedema: a review. *Cancer* 2005; 104(3): 457-66.
11. Carroll D, Rose K. Treatment leads to significant improvement. Effect of conservative treatment on pain in lymphoedema. *Prof Nurse* 1992; 8(1): 32-36.
12. Mortimer PS. Managing lymphedema. *Clin Exp Dermatol* 1995; 20(2): 98-106.
13. Dale RF. The inheritance of primary lymphoedema. *J Med Genet* 1985; 22(4): 274-78.
14. Moffatt CJ, Franks PJ, Doherty D, et al. Lymphoedema: an underestimated health problem. *QJM* 2003; 96: 731-38.
15. Rabe E, Pannier-Fischer F, Bromen K, et al. Bonner Venenstudie der Deutschen Gesellschaft für Phlebologie. *Phlebologie* 2003; 32: 1-14.
16. Mortimer P, Bates D, Brassington H, et al. The prevalence of arm oedema following treatment for breast cancer. *QJM* 1996; 89: 377-80.
17. Schrenk P, Reiger R, Shamiyeh A, Wayand W. Morbidity following sentinel lymph node biopsy versus axillary lymph node dissection fro patients with breast carcinoma. *Cancer* 2000; 88(3): 608-14.
18. Meric F, Buchholz TA, Mirza NQ, et al. Long-term complications associated with breast-conservation surgery and radiotherapy. *Ann Surg Oncol* 2002; 9(6): 543-49.
19. Ozaslan C, Kuru B. Lymphoedema after treatment of breast cancer. *Am J Surg* 2004; 187(1): 69-72.
20. Hong JH, Tsai CS, Lai CH, et al. Postoperative low pelvic irradiation for stage I-IIA cervical cancer patients with risk factors other than pelvic lymph node metastasis. *Int J Radiat Oncol Biol Phys* 2002; 53(5): 1284-90.
21. Ryan M, Stainton MC, Slaytor EK, et al. Aetiology and prevalence of lower limb lymphoedema following treatment for gynaecological cancer. *Aust N Z J Obstet Gynaecol* 2003; 43(2): 148-51.
22. Hardy D, Taylor J. An audit of non-cancer-related lymphoedema in a hospice setting. *Int J Palliat Nurs* 1999; 5: 18, 20-27.
23. Browse N, Burnand K, Mortimer P. *Diseases of the Lymphatics*. London: Arnold, 2003.
24. Department of Health and Diabetes UK. *Structured Patient Education in Diabetes*. Report from the Patient Education Working Group. London: Department of Health, 2005. Available at: www.dh.gov.uk/publications (accessed July 2006).
25. Mellor RH, Bush NL, Stanton AW, et al. Dual-frequency ultrasound examination of skin and subcutis thickness in breast cancer-related lymphedema. *Breast J* 2004; 10(6): 496-503.
26. Wheatley DC, Wastie ML, Whitaker SC, et al. Lymphoscintigraphy and colour Doppler sonography in the assessment of leg oedema of unknown cause. *Br J Radiol* 1996; 69: 1117-24.
27. Burnand KG, McGuinness CL, Lagattolla NRF, et al. Value of isotope lymphography in the diagnosis of lymphoedema of the leg. *Br J Surg* 2002; 89: 74-78.
28. Bollinger A, Jäger K, Sgier F, Seglias J. Fluorescence microlymhography. *Circulation* 1981; 64(6): 1195-200.
29. Partsch H, Stöberl Ch, Urbanek A, Wenzel-Hora BI. Clinical use of indirect lymphography in different forms of the leg edema. *Lymphology* 1988; 21(3): 152-60.
30. Liu NF, Wang CG. The role of magnetic resonance imaging in the diagnosis of peripheral lymphatic disorders. *Lymphology* 1998; 31(3): 119-27.
31. Hayes S, Cornish B, Newman B. Comparison of methods to diagnose lymphoedema amongst breast cancer survivors: 6-month follow-up. *Breast Cancer Res Treat* 2005; 89(3): 221-26.
32. Kissin MW, Querci della Rovere G, Easton D, Westbury G. Risk of lymphoedema following the treatment of breast cancer. *Br J Surg* 1986; 73(7): 580-84.
33. Sitzia J, Stanton AW, Badger C. A review of outcome indicators in the treatment of chronic limb oedema. *Clin Rehabil* 1997; 11(3): 181-91.
34. Kwaliteitsinstituut voor de Gezondheidszorg CBO. *Richtlijn Lymfoedeem*. Netherlands: van Zuiden Communications BV, 2002. Available at: www.lymfoedeem.nl (accessed July 2006).
35. Bernas M, Witte M, Witte C, et al. Limb volume measurements in lymphedema: issues and standards. *Lymphology* 1996; 29 (Suppl): 199-202.
36. Stanton AW, Northfield JW, Holroyd B, et al. Validation of an optoelectronic limb volumeter (Perometer). *Lymphology* 1997; 30(2): 77-97.
37. Vowden K, Vowden P. Managing leg ulcers: a review of clinical guidelines. *Nurs Times* 2000; 96(14 Suppl): 19-20.
38. Bonham PA. Steps for determining toe brachial pressure index. *Adv Skin Wound Care* 2004; 17(1): 44-45.
39. World Union of Wound Healing Societies (WUWHS). Principles of best practice: *Minimising pain at wound dressing-related procedures. A consensus document*. London: MEP Ltd, 2004.
40. Werner RS, McCormick B, Petreck J, et al. Arm edema in conservatively managed breast cancer: obesity is a major predictive factor. *Radiology* 1991; 180(1): 177-84.
41. Goffman TE, Laronga C, Wilson L, Elkins D. Lymphedema of the arm and breast in irradiated breast cancer patients: risks in an era of dramatically changing axillary surgery. *Breast J* 2004; 10(5): 405-11.
42. Ryan TJ. Risk factors for the swollen ankle and their management at low cost: not forgetting lymphedema. *Int J Low Extrem Wounds* 2002; 1(3): 202-8.
43. National Obesity Forum. Guidelines on management of adult obesity and overweight in primary care, 2004. Available at: www.nationalobesityforum.org.uk (accessed May 2006).
44. National Institutes of Health. *Clinical guidelines on the identification, evaluation, and treatment of overweight and obesity in adults.* NIH Publication no. 98-4083. Maryland: NIH, 1998.
45. Report of a joint WHO/FAO expert consultation. *Diet, Nutrition and the Prevention of Chronic Diseases*. WHO Technical Report Series 916. Geneva: World Health Organization, 2003.
46. International Diabetes Federation. The IDF consensus worldwide definition of the metabolic syndrome. Brussels, Belgium: International Diabetes Federation; 2005. Available at: http://www.idf.org/webdata/docs/MetSyndrome_FINAL.pdf (accessed July 2006).
47. Passik SD, McDonald MV. Psychosocial aspects of upper extremity lymphoedema in women treated for breast carcinoma. *Cancer* 1998; 83 (12 Suppl American): 2817-20.
48. National Institute of Clinical Excellence (NICE). *Depression: management of depression in primary and secondary care*. London: NICE, 2004. Available at: www.nice.org.uk (accessed July 2006).
49. Morgan PA, Moody M, Franks PJ, et al. Assessing community nurses' level of knowledge of lymphoedema. *Br J Nurs* 2005; 14(1): 8-13.
50. Marston W, Vowden K. Compression therapy: a guide to safe practice. In: European Wound Management Association (EWMA). Position Document: *Understanding compression therapy*. London: MEP Ltd, 2003; 11-17.
51. Partsch H. Ambulation and compression after deep vein

REFERENCES

thrombosis: dispelling myths. *Semin Vasc Surg* 2005; 18: 148-52.
52. Franks PJ, Moffatt CJ, Connolly M, et al. Factors associated with healing leg ulceration with high compression. *Age Ageing* 1995; 24(5): 407-10.
53. Vowden K. The use of intermittent pneumatic compression in venous ulceration. *Br J Nurs* 2001; 10(8): 491-509.
54. Alpagut U, Dayioglu E. Importance and advantages of intermittent external pneumatic compression therapy in venous stasis ulceration. *Angiology* 2005; 56(1): 19-23.
55. Partsch H. Understanding the pathophysiological effects of compression. In: European Wound Management Association (EWMA). Position Document: *Understanding compression therapy*. London: MEP Ltd, 2003; 2-4.
56. Gültig O. Lymphoedema bandaging for the head, breast and genitalia. In: European Wound Management Association (EWMA). Focus Document: *Lymphoedema bandaging in practice*. London: MEP Ltd, 2005; 15-17.
57. British Lymphology Society and Lymphoedema Support Network. Guidelines on the management of cellulitis in lymphoedema. Available at: http://www.lymphoedema.org/bls/CellulitisGuidelines.pdf (accessed July 2006).
58. Williams AF, Vadgama A, Franks PJ, Mortimer PS. A randomized controlled crossover study of manual lymphatic drainage therapy in women with breast cancer related lymphoedema. *Eur J Cancer Care* 2002; 11(4): 254-61.
59. Woods M. The experience of manual lymph drainage as an aspect of treatment for lymphoedema. *Int J Palliat Nurs* 2003; 9(8): 336-42.
60. McNeely ML, Magee DJ, Lees AW, et al. The addition of manual lymph drainage to compression therapy for breast cancer related lymphedema: a randomized controlled trial. *Breast Cancer Res Treat* 2004; 86(2): 95-106.
61. British Lymphology Society. *Chronic Oedema Population and Needs*. Sevenoaks, Kent: BLS, 1999.
62. De Godoy JMP, Batigalia F, Godoy M de F. Preliminary evaluation of a new, more simplified physiotherapy technique for lymphatic drainage. *Lymphology* 2002; 35(2): 91-93.
63. Bellhouse S. Simple lymphatic drainage. In: Twycross R, Jenns K, Todd J (eds). *Lymphoedema*. Oxford: Radcliffe Medical Press, 2000: 217-35.
64. Bergan JJ, Sparks S, Angle N. A comparison of compression pumps in the treatment of lymphedema. *J Vasc Surg* 1998; 32: 455-62.
65. Pohjola RT, Pekanmäki K, Kolari PJ. Intermittent pneumatic compression of lymphoedema. *Eur J Lymphol* 1995; 5: 87-90.
66. Bray T, Barrett J. Pneumatic compression therapy. In: Twycross R, Jenns K, Todd J (eds). *Lymphoedema*. Oxford: Radcliffe Medical Press, 2000; 236-43.
67. Johansson K, Lie E, Ekdahl C, Lindfeldt J. A randomized study comparing manual lymph drainage with sequential pneumatic compression for treatment of postoperative arm lymphedema. *Lymphology* 1998; 31(2): 56-64.
68. Szuba A, Achalu R, Rockson SG. Decongestive lymphatic therapy for patients with breast carcinoma-associated lymphedema. A randomised, prospective study of a role of adjunctive pneumatic compression. *Cancer* 2002; 95(11): 2260-67.
69. Boris M, Weindorf S, Lasinski BB. The risk of genital edema after external pump compression for lower limb lymphedema. *Lymphology* 1998; 31(1); 15-20.
70. Badger CM, Peacock JL, Mortimer PS. A randomized, controlled, parallel-group clinical trial comparing multilayer bandaging followed by hosiery versus hosiery alone in the treatment of patients with lymphedema of the limb. *Cancer* 2000; 88(12): 2832-37.
71. Thomas S. The use of the Laplace equation in the calculation of sub-bandage pressure. Available at: www.worldwidewounds.com/2003/june/Thomas/Laplace-Bandages.html (accessed July 2006).
72. Williams AF, Keller M. Practical guidance on lymphoedema bandaging of the upper and lower limbs. In: European Wound Management Association (EWMA). Focus Document: *Lymphoedema bandaging in practice*. London: MEP Ltd, 2005; 10-14.
73. Doherty D, Morgan P, Moffatt C. Role of hosiery in lower limb lymphoedema. In: Lymphoedema Framework. *Template for Practice: compression hosiery in lymphoedema*. London: MEP Ltd, 2006: 10-21.
74. Clark M, Krimmel G. Lymphoedema and the construction and classification of compression hosiery. In: Lymphoedema Framework. *Template for Practice: compression hosiery in lymphoedema.* London: MEP Ltd, 2006: 2-4.
75. Dale JJ, Ruckley CV, Gibson B, et al. Multi-layer compression: comparison of four different four-layer bandage systems applied to the leg. *Eur J Vasc Endovasc Surg* 2004; 27(1): 94-99.
76. Cohen SR, Payne DK, Tunkel RS. Lymphedema: strategies for management. *Cancer* 2001; 92(4 Suppl): 980-87.
77. Miller LT. Exercise in the management of breast cancer-related lymphoedema. *Innovations in Breast Cancer Care* 1998; 3(4): 101-6.
78. Johansson K, Tibe K, Weibull A, Newton RC. Low intensity resistance exercise for breast cancer patients with arm lymphedema with or without compression sleeve. *Lymphology* 2005; 38(4): 167-80.
79. Moseley AL, Piller NB, Carati CJ. The effect of gentle arm exercise and deep breathing on secondary arm lymphedema. *Lymphology* 2005; 38(3): 136-45.
80. Puleo CA, Luh M. Management of extremity lymphedema. *Cancer Control* 1995; 2(5): 424-28.
81. Gloviczki P. Principles of surgical treatment of chronic lymphoedema. *Int Angiol* 1999; 18(1); 42-46.
82. Campisi C, Boccardo F, Zilli A, et al. The use of vein grafts in the treatment of peripheral lymphedemas: long-term results. *Microsurgery* 2001; 21(4): 143-47.
83. Carrell T, Burnand K. Surgery and lymphoedema. In: Twycross R, Jenns K, Todd J (eds). *Lymphoedema*. Oxford: Radcliffe Medical Press, 2000: 285-92.
84. Campisi C, Davini D, Bellini C, et al. Lymphatic microsurgery for the treatment of lymphedema. *Microsurgery* 2006; 26(1): 65-69.
85. Campisi C, Boccardo F. Microsurgical techniques for lymphedema treatment: derivative lymphatic-venous microsurgery. *World J Surg* 2004; 28(6): 609-13.
86. Weiss M, Baumeister RG, Hahn K. Dynamic lymph flow imaging in patients with oedema of the lower limb for evaluation of the functional outcome after autologous lymph vessel transplantation: an 8-year follow-up study. *Eur J Nucl Med Mol Imaging* 2003; 30(2): 202-6.
87. Becker C, Assouad J, Riquet M, Hidden G. Post mastectomy lymphedema: long-term results following microsurgical lymph node transplantation. *Ann Surg* 2006; 243(3): 313-315.
88. Cluzan R, Ryan T, Brorson H. Adipose tissue and lymphedema – what is proved? *Lymphology* 2006; 39(Suppl): [in press].
89. Brorson H. Liposuction in arm lymphedema treatment. *Scand J Surg* 2003: 92: 287-295.
90. Brorson H, Ohlin K, Olsson G, Svensson B. Long term cosmetic and functional results following liposuction for arm lymphedema: An eleven year study. *Lymphology* 2006; 39 (Suppl): [in press].
91. Brorson H, Ohlin K, Olsson G, Svensson B. Liposuction of leg lymphedema: Preliminary 2 year results. *Lymphology* 2006; 39 (Suppl): [in press].
92. Mortimer P. Swollen lower limb-2: lymphoedema. *BMJ* 2000; 320(7248): 1527-29.
93. Porter W, Dinneen M, Bunker C. Chronic penile lymphedema: a report of 6 cases. *Arch Dermatol* 2001; 137(8): 1108-10.
94. Campisi C, Bellini C, Eretta C, et al. Diagnosis and management of primary chylous ascites. *J Vasc Surg* 2006; 43(6): 1244-48.
95. Loprinzi CL, Kugler JW, Sloan JA, et al. Lack of effect of coumarin in women with lymphedema after treatment for breast cancer. *N Engl J Med* 1999; 340(5): 346-50.
96. Roztocil K, Prerovsky I, Oliva I. The effect of hydroxyethylrutosides on capillary filtration rate in the lower limb of man. *Eur J Clin Pharmacol* 1977; 11(6): 435-38.
97. Vaqas B, Ryan TJ. Lymphoedema: pathophysiology and management in resource-poor settings – relevance for lymphatic filariasis control programmes. *Filaria J* 2003; 2(1): 4.
98. Shim JY, Lee HR, Lee DC. The use of elastic adhesive tape to promote lymphatic flow in the rabbit hind leg. *Yonsei Med J* 2003; 44(6): 1045-52.
99. Gothard L, Stanton A, MacLaren J, et al. Non-randomised phase II trial of hyperbaric oxygen therapy in patients with chronic arm lymphoedema and tissue fibrosis after radiotherapy for early breast cancer. *Radiother Oncol* 2004; 70(3): 217-24.
100. Teas J, Cunningham JE, Cone L, et al. Can hyperbaric oxygen therapy reduce breast cancer treatment-related lymphedema? A pilot study. *J Womens Health* 2004; 13(9): 1008-18.
101. Carati CJ, Anderson SN, Gannon BJ, Piller NB. Treatment of postmastectomy lymphedema with low-level laser therapy: a double blind, placebo-controlled trial. *Cancer* 2003; 98(6): 1114-22.