

LYMPHOEDEMA DIAGNOSIS AND MANAGEMENT

Lymphoedema is a chronic progressive condition that is characterised by swelling of the limb, head, neck, breast and genitalia. It can result in significant physical and psychological morbidity, but the symptoms can be greatly ameliorated with prompt and appropriate management.

Debra Doherty is Senior Lecturer, Centre for Research and Implementation of Clinical Practice, Thames Valley University



Figure 1. Squared toes with forefoot swelling.

Lymphoedema is a chronic progressive condition that is generally characterised by the swelling (oedema) of one or more limbs, and may include the corresponding section (quadrant) of the trunk. Swelling can also affect the head and neck, breast and the genitalia. It occurs as a result of an imbalance between lymph formation and lymph absorption. Fluid and its contents (e.g. subcutaneous fat, protein) accumulate in the tissue spaces because of functional inadequacy of the lymphatic system (International Society of Lymphology, 2003).

There is currently no cure for lymphoedema, but it may be greatly ameliorated with

appropriate management. Many patients are unaware of treatments or where to obtain them and, if ignored, the condition can gradually deteriorate and become difficult to manage. Lymphoedema can result in significant physical and psychological morbidity. The increase in limb size can

interfere with physical functioning and affect body image (Weiss and Spray, 2002). Additionally, pain and discomfort are physical symptoms that are frequently associated with the condition (Franks et al, 2006).

Lymphoedema can be assigned to two categories: primary or secondary. Primary lymphoedema is thought to be the result of a primary abnormality of the lymph-conducting system, but the nature and cause is unknown (Browse et al, 2003). Secondary lymphoedema is an acquired disorder caused by damage to the lymphatic system, resulting in functional deficiency (International Society of Lymphology, 1995) (Tables 1 and 2).

Table 1

Primary lymphoedema

A condition that may be isolated or inherited, and which results from abnormalities or malformation of the lymphatic vessels and/or nodes

Clinical presentation can occur at any age (Browse et al, 2003)

Table 2**Causes of secondary lymphoedema**

Trauma and tissue damage
 Infection
 Malignant disease
 Venous disease
 Inflammation
 Immobility and limb dependency
 Factitious
 (Browse et al, 2003)

The most common form of lymphoedema worldwide — secondary lymphoedema due to filarial infection transmitted by the mosquito — occurs mainly in tropical regions. Filariasis-induced lymphoedema affects as many as 120 million people in 80 countries, with 40 million suffering disability and disfigurement as a result (World Health Organization, 2000). Migrating mosquito populations mean that the UK may be affected by lymphoedema filariasis.

There is a perception that lymphoedema is an uncommon problem in developed countries, and is mainly confined to people who have developed it following treatment for cancer. However, it has been reported that, at birth, approximately one in 6000 people will develop primary lymphoedema and the overall prevalence of lymphoedema/chronic oedema has been estimated as 1.33/1000 in the UK (Dale, 1985; Moffatt et al, 2003).

Primary lymphoedema is usually considered after all possible causes of secondary lymphoedema have been excluded. Where lymphoedema is thought to be primary in origin, a specialist referral

may be necessary for further investigation.

Assessment

In most cases the diagnosis of lymphoedema is determined from clinical history and physical examination (International Society of Lymphology, 2003). However, the wider psychological and social issues require a more holistic approach. The doctor has an important role to play in the assessment process as early diagnosis and treatment is critical in order to prevent worsening of the condition.

Basic assessment should include an evaluation of any tumour burden in the lymph nodes, the patency of the venous system, evidence of soft tissue infection, the frequency of any infections, and identification of any venous insufficiency, and should exclude deep vein thrombosis (DVT) (Caban, 2002). Routine clinical

investigations are required to exclude other causes of oedema, or identify factors that may have contributed to the development of lymphoedema. Additional diagnostic investigations may be necessary to ascertain the cause of the oedema and ensure appropriate treatment (Table 3).

Early identification of the condition and initiation of treatment leads to better therapeutic outcomes (Brennan and Miller, 1998). Subjective symptoms, with or without overt swelling, frequently exist in the early stages of lymphoedema. For example, Rockson et al (1998) highlighted the importance of symptoms such as tension of the arm, leg or chest, heaviness, discomfort and the experience of hardness in the identification of early lymphoedema. Objective limb measurements are also used to identify swelling,

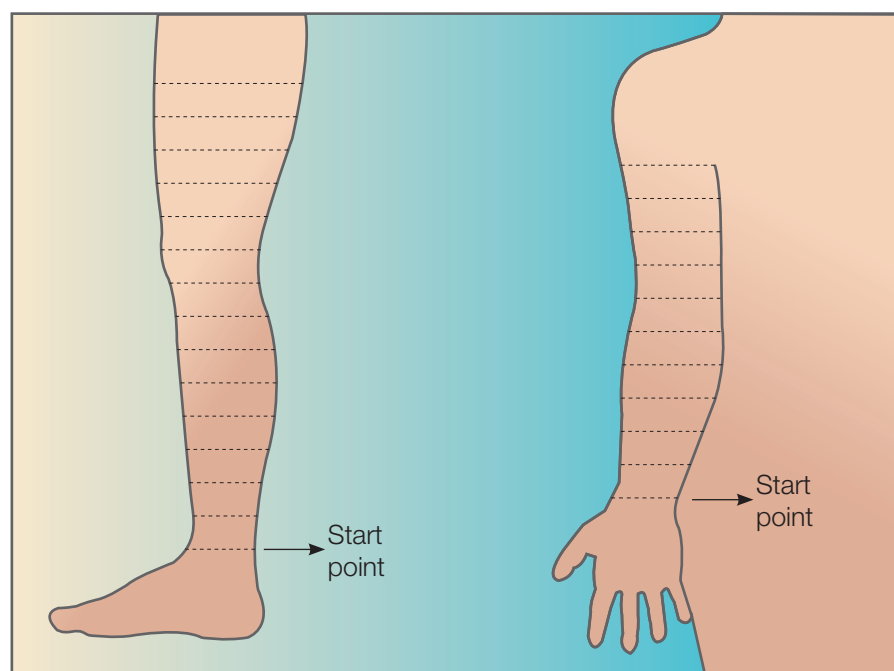


Figure 2. Measuring limb circumference. Using a ruler and tape, measurements are taken at 4cm intervals along the limb from a predetermined, fixed starting point.

Table 3**Useful diagnostic tests**

Lymphoscintigraphy: a lymphatic imaging test used mainly to identify the cause of swelling, or to rule out lymphoedema for those who may just have lipoedema

Computed tomography scan

Magnetic resonance imaging scan

such as measuring the limb's circumference (*Figure 2*).

Using a ruler and tape, measurements are taken at 4cm intervals along the limb from a predetermined, fixed starting point.

Both limbs are always measured at the same time and a determination of lymphoedema is made on the measured differences. It is important to identify the dominant limb as measurements can naturally differ between both limbs.

In one study, Kissin et al (1986) found that in a normal control group there was a difference of up to 2cm in arm measurements that was not considered to be caused by lymphoedema. In the case of volumetric measurements, the percentage difference in volume between limbs in the normal population can be as high as 8–9% (Sitzia, 1997). Lymphoedema is considered if the volume in the swollen limb is at least 10% above that of the contralateral (other) normal limb volume. This is a useful indicator with unilateral limb swelling only. However, where both limbs are swollen, comparisons cannot be made and, in such cases,

both limbs are measured and volume is monitored over time. Other methods of evaluation include changes in skin folds, tissue texture, shape distortion, psychosocial morbidity and impaired general or limb function (Sitzia et al, 1997).

The management of lymphoedema

The management of lymphoedema requires a multi-modal approach and may involve several members of the multidisciplinary team. Essential components of care include skin care and risk-minimisation strategies, the application of external support/compression, a programme of exercise and movement, psychosocial support and patient education. Manual lymphatic drainage may also be an integral component of care for some groups of lymphoedema patients.

Skin care and prevention strategies

The awareness and identification of lymphoedema is an important issue for those at risk of, or who have developed, the condition. There appear to be several factors that can predispose an individual to a higher risk of developing this disease or which can predict the progression (*Table 4*). Skin problems are common in patients with lymphoedema as the skin undergoes significant changes (*Figures 1,3,4*). Excessive subcutaneous swelling can distort shape and lead to deep skin folds. Tissue changes occur as a result of chronic inflammation, causing the deposition of fibrin and collagen. These fibrotic changes contribute to thickening and a firm tissue consistency (Foldi et

al, 2003). This thickened, less compliant skin can compromise lymph flow even further and increase the tendency to infection.

Cellulitis is a complication commonly associated with lymphoedema. This is because impairment of the lymphatic system leads to a disturbance in the movement of white blood cells (lymphocyte trafficking), and can result in reduced defence mechanisms within the lymphatic drainage area. Damage to the skin through household or garden injuries, insect bites, sunburn and injections have to be avoided as the compromised immune response increases the tendency for infection.

A systematic review by Badger et al (2004) highlighted the importance of good foot care as a preventive measure, as fungal infection between the toes is a precipitating cause of

Table 4**Risk factors for lymphoedema**

Breast or gynaecological cancer surgery, radiation therapy or node dissection

Lymph node removal, trauma to lymph nodes/vessels

Advanced cancer

Poor nutritional status and obesity

Recurrent infection in the same site

Family history of chronic oedema

Immobility and prolonged limb dependency

Chronic skin disorders and inflammation

Chronic venous insufficiency, particularly post-thrombotic syndrome

Concurrent illness, e.g. kidney or cardiac disease or phlebitis (inflammation of the walls of a vein)

Varicose vein stripping

Vein harvesting for femoropopliteal bypass graft



Figure 3. Deepened skin folds and resolving cellulitis.



Figure 4. Hyperkeratosis. Overproliferation of the keratin layer of the skin looks brown/grey and there is a distorted limb shape.

cellulitis. Patients at risk and those with swelling should be given information on how to prevent skin damage, treat minor abrasions, and seek medical help in the event of an infection.

Skin care regimens teach patients with lymphoedema and their carers how to support the acidity and hydration of the skin. Maintenance of good skin integrity is important in avoiding infection, which can

cause further swelling because of the inflammatory response. The skin's barrier function is restored by emollients, which soothe the epidermis (Table 6).

Compression

The management of lymphoedema can necessitate different types and levels of intervention. Traditionally, the treatment of lymphoedema is described as consisting of two distinct phases, an intensive treatment period (phase one) and a long-term treatment/management plan (phase two). Following assessment, patients are entered into the most appropriate phase of treatment. In practice, however, these two phases may not be distinct from one another. Treatment usually needs to be tailored to meet the increasing complexity of patient needs. Compression can be applied using bandaging or hosiery.

Intensive therapy (phase one) involves decongesting the reduced lymphatic pathways to reduce the size of the limb, encouraging the development of collateral drainage routes and stimulating the function of remaining patent routes to control swelling in the long term (Mortimer, 1995). It involves treatment that is applied by a trained practitioner over a specified period, usually 2–4 weeks. The frequency and duration of intensive treatment can vary, depending on the increasingly complex needs of patients being managed in the community. During the intensive phase of treatment, compression is usually applied in the form of multilayer lymphoedema bandaging (Table 7).

Multilayer compression systems used in the management of lymphoedema comprise several layers of inelastic bandaging. Inelastic bandages do not have any elasticity, the stretch is caused by the weave only, thus it forms an inelastic covering to the limb. This resists changes in the limb or calf muscle and shape.

On movement of the limb, the lymphatic vessels pump when they are compressed against the inelastic bandage layers by the calf muscle. This generates a high-working pressure on muscle contraction, and a low-resting pressure on muscle relaxation to allow the lymph vessels to fill and facilitate lymph flow (Partsch, 1991).

Multilayer lymphoedema bandaging is indicated for ulceration, fragile or damaged skin, lymphorrhoea, thickened skin changes, pronounced skin folds, distorted limb shape, lymphangiectasia and papillomaosis, or where the limb is too large to fit hosiery. Bandaging is contraindicated where the ankle brachial pressure

Table 6

General principles of skin care

Avoid soap as it tends to cause dry skin

Avoid scented, perfumed products, as they can precipitate sensitivity reactions

Observe, check and monitor skin for cuts, abrasions or insect bites. Careful inspection is important if sensory neuropathy is present

Ensure skin folds, if present, are clean and dry

Moisturisation creates a barrier that prevents further water loss and protects the skin from bacteria and irritants

Table 7**Components of intensive treatment**

Skin care
 Multilayer lymphoedema bandaging
 Exercise and movement
 Patient support and education
 Manual lymphatic drainage (for some groups of patients)

Table 8**Guidelines on exercise**

Maintain normal functioning/activity level
 Gradual introduction of active exercises based on ability and disease status
 Warn patients to warm up and cool down before and after exercise
 Exercise needs to be modified to meet specific needs
 Consider accompanying neurological or joint problems
 Compression should be worn during exercise to maximise its effects
 Flexibility exercises are useful to maintain range of movement
 Where a degree of paralysis exists, the affected limb can be moved passively
 Swimming, aqua aerobics, walking, cycling and low-impact aerobics are recommended activities
 The Lymphoedema Support Network videos are useful for specific exercises

index is <0.6 , in the presence of: acute cellulitis, as the bandages will be too painful to tolerate; acute DVT; uncontrolled heart failure; lymphatic obstruction and extreme sensory neuropathy. *Figure 5* shows how multilayer lymphoedema bandaging can be used to treat lower limb lymphoedema.

Exercise and movement

Exercise and movement are common rehabilitative

interventions and are an important component in the treatment and management of lymphoedema. Breathing, exercise and limb elevation aimed at improving venous functioning also benefit the lymphatic system, as peripheral lymphatic failure is frequently complicated by venous failure (Ryan, 2003). At present, there is little evidence to indicate what is the most effective exercise regime in the management of lymphoedema. The type, intensity, frequency and under what conditions exercise may be safely used to reduce swelling has not been fully defined. Functional capacity should be assessed before patients embark on an exercise programme.

Inactivity will not prevent the onset of lymphoedema, particularly after breast cancer surgery (Johansson et al, 2002). Most patients will benefit from exercise and should be encouraged to wear a compression garment to provide extra support and help the muscles function more effectively. Combinations of stretching and aerobic exercise can be beneficial in controlling lymphoedema (Miller, 1998). *Table 8* provides exercise guidelines.

In some cases, physiotherapy intervention may be required. This is usually indicated for problems related to mobility, if there is no improvement in three months following a standard exercise regimen devised by a specialised practitioner. Physiotherapy may improve joint movement, movement-related pain following breast cancer surgery and joint function.

Long-term management

Effective long-term management of lymphoedema (phase two) necessitates a considerable degree of patient involvement. The management of lymphoedema relies on patients and carers playing an active role. Enhancing the patient's role in the self-management of chronic diseases and reducing dependence on health professionals can improve the patient's knowledge, health-related behaviour, and psychosocial and health status (Lorig et al, 1987; Gibson et al, 2000).

The long-term management of lymphoedema focuses on enhancing the functioning of the lymphatic system, limiting further deterioration of swelling and gaining long-term control of the condition (Mortimer and O'Donnell, 2003) (*Table 9*).

Hosiery is generally used in the long-term management of lymphoedema. Garments aim to work using the same principles that apply to compression bandages. They should ideally facilitate a high-working



Figure 5. Multilayer lymphoedema applied to the leg.

pressure and a low-resting pressure to allow lymphatic refill. As with bandages, this requires garments to be as inelastic as possible in order to achieve this objective. However, some elasticity is essential to permit movement and facilitate application of the garments. Indications for the use of compression hosiery include intact and resilient skin, regular limb shape with no shape distortion and swelling <20% excess volume. Contraindications are similar to those for bandaging but additionally include extensive ulceration, lymphorrhoea or other weeping skin conditions, fragile or damaged skin, pronounced skin folds and limbs too large to fit into hosiery (Moffatt et al, 2005). Custom-made garments are usually required to address the problem of a larger limb size. There are several factors that need to be considered when choosing hosiery (Table 10).

Williams and Williams (1999) highlight the importance of limb shape when choosing hosiery and the fact that poor

limb shapes are not suitable for ready-to-wear hosiery. Ready-made garments can be less precise in fit, and are more likely to roll at the top. Custom-made garments have a number of advantages: they do not roll, curl, twist or tighten and, therefore, fit the patient more effectively.

Accurate measurement is important to achieve a correctly fitting garment. Generally, measurements should be taken when the area is largely free of oedema, i.e. immediately after removal of compression bandages or in the morning before any swelling can develop. It is important not to tighten the tape so that it makes an indentation in the skin. Precise measurements are required for custom-made garments. Most manufacturers provide measuring forms to assist the fitter (Table 11).

After the initial prescription, the practitioner should fit the garment to ensure that there are no problems. He or she should demonstrate and observe the patient's application technique



Figure 6. Example of a poorly-fitting garment and how to overcome poor fit. The practitioner must always check the garment following prescription to check that it fits properly.

Table 9

Focus of long-term management of lymphoedema

Daily skin care to improve skin hydration and reduce the risk of acute inflammatory episodes
 Compression to provide support for muscles and encourage lymphatic and venous flow
 Exercise, including breathing exercises and movement, to enhance lymph and venous flow
 Patient support and psychological care from professionals and other patients
 Patient education enabling people to know when to seek advice and how to access help quickly

Table 10

Factors to consider when choosing hosiery

General health and evaluation of peripheral limb circulation
 Patient dexterity — ability to apply and remove hosiery or access to a carer who can do this for the patient
 Patient preferences and comfort
 Patient mobility and limb function
 Severity of oedema
 Shape and size of the limb
 Extent of fibrosis/fat deposition
 Previous history
 Age
 Lifestyle
 Coverage — to prevent oedema distal or proximal to the garment
 Level of compression required
 Material
 Skin sensitivity
 (Moffatt et al, 2005)

and garment removal, and correct if necessary. All wrinkles should be removed when applied. This can be assisted by wearing application gloves with raised tread on the palm and fingers to smooth the hosiery by stroking gently upwards.

Patients should be advised against folding hosiery over at

Table 11

Items that need to be included in a prescription for compression garments

Quantity of stockings/tights or sleeves
 Level of compression
 Name of manufacturer and order code
 Knitted texture (flat or circular)
 Length of garment
 Attachment/fixation
 Measurements (for custom-made garments)
 Colour
 Any comments or explanations

the top, as tight bands at the top of the stocking may lead to pressure damage. Patients should be instructed to wash their garments frequently and according to manufacturers' instructions to protect durability. The hosiery also needs to be protected from direct contact with emollients as these can damage them. Garments should be replaced every 3–6 months, or when they begin to lose elasticity (*Figure 6*).

Clear verbal and written instructions must be given with regard to fitting errors, such as pressure points, skin discoloration or constrictions discovered after first wearing. Where difficulties occur, the garment should be immediately removed and the practitioner informed via given contact details.

Community staff can play an important role in the long-term support of patients with lymphoedema. They should be adequately trained and competent to identify and provide appropriate care. Their role in the long-term phase of

therapy will involve:

- ▶▶ Observation and monitoring control of swelling
- ▶▶ The use of multilayer bandaging in a small percentage of patients where this may be required
- ▶▶ Renewal of hosiery
- ▶▶ Providing positive reinforcement and motivation to facilitate the implementation of self-management regimes
- ▶▶ Appropriate referral to the lymphoedema specialist team.

Manual lymphatic drainage

Lymphoedema results from a reduction in the transport capacity of the lymphatic system. The lymph system in the affected area is unable to respond to an increase in lymphatic load. Massage therapy can increase the amount of lymphatic load and may have a negative effect on lymphoedema. Traditional massage techniques can apply too much pressure to the superficial lymphatics, and cause focal damage to anchoring filaments and the endothelial lining of lymph vessels (Eliska and Eliska, 1995).

Manual lymphatic drainage (MLD) is a gentle form of massage that is an integral component of lymphoedema treatment for some groups of patients. If correctly applied, MLD increases the activity of lymph vessels, exerts little pressure on the skin and does not cause an increase in local arterial blood flow (Foldi, 1995). MLD aims to evacuate extracellular fluid from the interstitial space.

Where there is lymphoedema of the head, neck, body and breast,

compression may not be well tolerated. Therefore, MLD may be the only realistic option and can be effective in reducing and controlling swelling long term when used in an early intervention programme (Piso et al, 2001).

MLD is generally considered to be too labour-intensive and time-consuming to be widely available to all patients with lymphoedema. Consequently, MLD is rationed and its good effects are mostly unavailable outside specialist centres.

Conclusion

Lymphoedema is an under-recognised problem in the UK, but there has been a recent focus on raising awareness of the condition. Greater access to quality care and early intervention can reduce its progression considerably and improve management. A multi-modal and multi-professional approach is often required to manage lymphoedema and its complications effectively.

Glossary

Lipoedema: excessive subcutaneous deposition of fat in the lower legs between the ankle and waist. It can occur to a lesser extent in the upper limbs from wrist to shoulder

Lymphoerrhoea: leaking of lymphatic fluid through the surface of the skin. It typically presents as beads of fluid trickling down the affected limb or swollen region

Papillomatosis: dilated skin lymphatics surrounded by thickened tissue, which is seen as cobblestone-like projections on the surface of the skin

As patients are managing the condition on a long-term basis, community healthcare professionals require appropriate education and skills to provide care jointly with specialist lymphoedema practitioners, and support where required. Finally, organisational commitment is required to provide appropriate services to meet the needs of patients in the community. **WE**

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Key Points

- » There is currently no cure for lymphoedema but it may be greatly ameliorated with appropriate management.
- » Lymphoedema can result in significant physical and psychological morbidity.
- » Pain and discomfort are physical symptoms that are frequently associated with the condition.
- » The management of lymphoedema requires a multi-modal approach and may involve several members of the multidisciplinary team.