

# Best Practice Guidelines

## The management of lipoedema

2017

Diagnosis and assessment

Lipoedema management

Life style support and  
self care

Compression therapy

Non-surgical and surgical  
interventions

## **BEST PRACTICE GUIDELINES: THE MANAGEMENT OF LIPOEDEMA**

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# Wounds UK

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# Developing best practice guidelines for the management of lipoedema

People with lipoedema in the UK face significant challenges. Many are not recognised by healthcare professionals as having the condition or are misdiagnosed. Awareness of lipoedema among medical practitioners is poor, and little clinical research is focused on the condition. To date, no good quality guidelines for the management of the disease have been published, resulting in inconsistent and frequently inappropriate care for people with lipoedema.

Even when lipoedema is diagnosed correctly, accessing appropriate care within the NHS may be difficult because of poor understanding of treatment and referral routes, and geographical variations in clinic availability, funding and capacity.

Lipoedema is a chronic, incurable disease that can have a severe impact on quality of life, and physical and psychosocial wellbeing. Some patients are so seriously affected that they lead very restricted lives, sometimes to the extent of being unable to leave their homes. The complexity of the issues faced by patients with lipoedema necessitates interprofessional, multidisciplinary care with an emphasis on supporting self management and working in partnership with the person to identify realistic goals and to manage expectations.

These best practice guidelines on lipoedema were inspired by a group of clinicians who first started discussing the need for clear guidance in 2015. The discussions culminated in a meeting in September 2016 that had the specific aim of developing guidelines on management that improve the lives and outcomes of people with lipoedema. The meeting was ground breaking: not only did it bring together key opinion leaders and experts involved in the treatment of lipoedema from all around the UK, but, significantly, it also included people with lipoedema representing UK third sector organisations.

The meeting participants recognised a general paucity of clinical evidence relating to the management of lipoedema. The conclusions of the meeting formed the basis for this document, which draws, where possible, on relevant literature. Where evidence is lacking, expert opinion has been used to inform the guidelines and make recommendations. The content was subject to review by the Expert Working Group and additional reviewers before being finalised.

This document will be of interest to anyone involved in delivering support and clinical services to people with lipoedema, including general practitioners, lymphoedema therapists, community nurses, plastic surgeons, dietitians, commissioners, third-sector organisations and more.

There is still a considerable amount to learn about lipoedema. Undoubtedly, the next few years will bring rapid advances in understanding of the pathophysiology of lipoedema and the most effective ways of managing the condition. As a result, the Group recognises that this document is likely to need to be reviewed within three years.

The Group hopes that the document will be useful to people with lipoedema, and the wide range of professionals who have contact with them. This document is an early step towards achieving tangible benefits for patients, enhancing recognition and diagnosis of the condition by professionals and the public, improving access to best practice management, and providing scope for future development of lipoedema services in the UK.

Anne Williams and  
Denise Hardy  
Co-Chairs

## GUIDE TO USING THIS DOCUMENT

Each section of the document helps healthcare practitioners to provide appropriate support and effective treatment and care for patients with lipoedema. The key points for each section summarise the information most relevant to clinical practice

# SECTION 1: EPIDEMIOLOGY AND PATHOPHYSIOLOGY OF LIPOEDEMA

Lipoedema was first described in 1940 and is a chronic incurable condition involving a pathological build-up of adipose tissue (Allen & Hines, 1940). It typically affects the thighs, buttocks and lower legs, and sometimes the arms, and may, although not always, cause considerable tissue enlargement, swelling and pain. It may significantly impair mobility, ability to perform activities of daily living, and psychosocial wellbeing. Current conservative management involves encouraging self-care, managing symptoms, improving functioning and mobility, providing psychosocial support, and preventing deterioration in physical and mental health and wellbeing.

Lipoedema is predominantly a chronic adipose tissue disorder (the word lipoedema means ‘fat swelling’), with clinically apparent oedema due to fluid accumulation in the tissues occurring as a secondary feature in some individuals (Todd, 2010; Herbst, 2012a; Reich-Schupke et al, 2013; Herbst et al, 2015). Although most commonly called lipoedema, the condition has a variety of other names (Box 1).

## Prevalence

Lipoedema almost exclusively affects women, but a few cases have been reported in men (Chen et al, 2004; Langendoen et al, 2009). Relatively little epidemiological research has been carried out on lipoedema and so it is unclear exactly how many people are affected and to what extent. The research so far has produced widely varying figures. In the UK, the minimum prevalence of lipoedema has been estimated to be 1 in 72,000 (Child et al, 2010). However, the authors noted that this is likely to be an underestimate (Child et al, 2010). In Germany, the prevalence of lipoedema has been estimated to be 11% in women and post-pubertal girls (Földi et al, 2006; Szél et al, 2014).

Further research is needed to establish clearly the proportion of the population affected by lipoedema. It is likely to be more common than the limited evidence available

suggests: cases may be ‘hidden’ because of their mild nature or because the person is reluctant to contact health services. Other cases may be unrecognised or misdiagnosed by health services. Common misdiagnoses include obesity or lymphoedema (Box 2) (Goodliffe et al, 2013), although both conditions may co-exist with lipoedema.

## Cause

The precise mechanisms responsible for the development of lipoedema are unknown, but it is likely that multiple factors are involved (Okhovat & Alavi, 2014).

Lipoedema often first presents during puberty, although oral contraceptive use, pregnancy and the menopause also appear to be triggers. These observations suggest that hormonal change may be involved in initiating the characteristic build-up of adipose tissue (Fonder et al, 2007; Bano et al, 2010; Godoy et al, 2012). Onset of the disease after periods of significant weight gain have also been reported (personal communication, K Gordon).

There is also evidence of a genetic predisposition to lipoedema. A family history of the condition has been found in 15%–64% of patients (Harwood et al, 1996; Child et al, 2010; Schmeller & Meier-Vollrath, 2007). The genetic variants involved have not been identified fully, but research suggests that autosomal dominance with male sparing is the most likely mode of inheritance (Child et al, 2010). Investigations into the genetics of lipoedema are ongoing, and include researching whether men may act as carriers for the associated genetic factor(s).

### Box 1. Synonyms for lipoedema (Schmeller & Meier-Vollrath, 2007; Langendoen et al, 2009; Herbst 2012a; Cornely, 2014)

- Adiposalgia
- Adiopoalgia
- Lipalgia
- Lipedema (American spelling)
- Lipohyperplasia dolorosa
- Lipohypertrophy dolorosa
- Lipomatosis dolorosa of the legs
- Painful column legs
- Painful fat syndrome
- Riding breeches syndrome
- Stovepipe legs.

### Box 2. Lymphoedema and lipoedema (Harwood et al, 1996; Lymphoedema Framework, 2006; Goodliffe et al, 2013)

Patients with lipoedema may be misdiagnosed as having lymphoedema. Lymphoedema results from malfunction of the lymphatic system, whereas lipoedema is thought to primarily be a disorder of adipose tissue (a lipodystrophy). Confusingly, however, patients with lipoedema may develop lymphatic dysfunction. This combination of lipoedema and secondary lymphoedema is sometimes referred to as lipolymphoedema.

### Enlargement of fat tissue

The characteristic increase in subcutaneous fat tissue seen in lipoedema may be due to adipocyte hypertrophy (increase in size but not necessarily number of fat cells) and/or hyperplasia (increase in number of fat cells) (Suga et al, 2009; Schneble et al, 2016) (Figure 1). In addition, there is evidence of an increase in the rate of adipocyte death, possibly due to hypoxia induced by excessive tissue enlargement, and infiltration of fat tissue by scavenger inflammatory cells (macrophages) (Suga et al, 2009).

By inducing growth of new fragile capillaries in the fat tissue, it has been suggested that hypoxia may contribute to the easy bruising often reported by patients with lipoedema (Fife et al, 2010). Other tissue changes that may occur include reduced elasticity of the skin and connective tissue (fascia) (Jagtman et al, 1984; Herbst, 2012a).

The cause of the pain and hypersensitivity often mentioned by patients with lipoedema is unclear, but may relate to compression of nerve fibres by enlarged fat deposits, inflammation and/or central sensitisation (a process which involves changes in the brain and spinal cord that are associated with the development of chronic pain) (Langendoen et al, 2009; Peled & Kappos, 2016).

### Development of oedema

In many patients, lipoedema is accompanied by the formation of fluid oedema. It has been suggested that the oedema may result from overloading of an essentially normal lymphatic system (see Appendix 1, page 32 for information on the lymphatic system). Although, changes in the structure and function of the lymphatic system have been observed in some patients, much research is needed to discover whether these changes are a common feature of lipoedema and whether they relate to the pathophysiology of the condition (Amann-Vesti et al, 2001; Bilancini et al, 1995).

Increased interstitial fluid formation due to capillary fragility and possible mechanical obstruction of small lymphatic vessels by adipose tissue enlargement, combined with reduced skin and connective tissue elasticity, reduced mobility due to pain or

joint problems, may act to decrease the effectiveness of the venous and lymphatic systems (Harwood et al, 1996; Cornely, 2006; Langendoen et al, 2009). As a result, the rate of interstitial fluid accumulation may exceed the rate of clearance, and oedema may occur.

In patients with lipoedema who also have chronic venous insufficiency (CVI) the tendency for interstitial fluid accumulation may be compounded.

Age-related changes that cause the lymphatic vessels to harden (lymphangiosclerosis) and become less effective at removing fluid may also contribute to the development of lipolymphoedema (Cornely, 2006).

Some women with lipoedema report premenstrual fluid retention that can have a considerable cyclical impact on the size and shape of lipoedematous areas.

### Key points

1. Lipoedema is underdiagnosed and almost exclusively affects women
2. Although lipoedema is often misdiagnosed as simply being obesity, lipoedema and obesity can co-exist
3. Hormonal and genetic factors are likely to contribute to the adipose tissue enlargement characteristic of lipoedema
4. Patients with lipoedema may develop secondary lymphoedema (lipolymphoedema), which may be compounded if chronic venous insufficiency is also present.

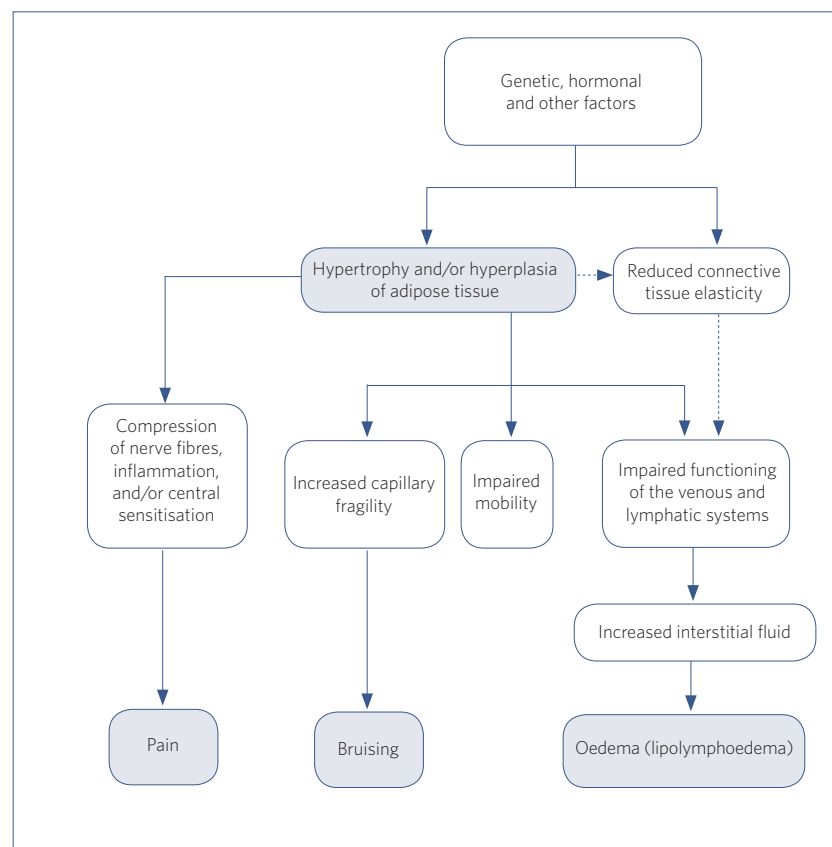


Figure 1: Possible pathophysiology of lipoedema

## SECTION 2 : DIAGNOSIS AND ASSESSMENT

Lipoedema is often not recognised in primary care, and awareness and understanding of the condition among medical professionals is limited (Goodliffe et al, 2013; Evans, 2013).

A diagnosis of lipoedema is made on clinical grounds that are based on the history and examination of the patient. Currently, there are no known blood or urine biomarkers, nor are there any specific diagnostic tests, for lipoedema (Herbst, 2012a).

In the absence of definitive diagnostic tests, clinicians need to have a clear understanding of the unique characteristics of lipoedema and how they differ from other apparently similar conditions such as lymphoedema and obesity (Fife et al, 2010) (see pages 8–9).

Diagnosis of lipoedema may be delayed due to poor recognition of the condition by health professionals. Making an accurate diagnosis may be challenging, particularly in the early stages or when a patient has co-existing obesity.

The course of lipoedema over time is not fully understood, but is highly variable and unpredictable. The condition may progress relentlessly in some patients, and yet in others the only symptom is a relatively minor increase in subcutaneous fat that remains stable for many years (Langendoen et al, 2009; Dutch Guidelines, 2014).

### History and symptoms

Typically, a patient with lipoedema is female and reports onset at puberty or at another time of hormonal change. Only a handful of male cases have been reported in the literature: all were thought to have developed lipoedema secondary to hormonal disturbances, with reduced testosterone levels being a common factor (Child et al, 2010).

The development of tissue enlargement is often insidious (Todd, 2016). It is usually bilateral and symmetrical, and most commonly affects the legs, thighs, hips and/or buttocks, with sparing of the feet. Diagnosis of lipoedema may be difficult in

the early stages or in mild forms as the symptoms and signs may be subtle. The characteristics of lipoedema become more obvious as the disease progresses and in more severe forms (Table 4, page 11).

Although the lower limbs and buttocks are the most commonly affected areas, it is suggested that lipoedema may occur in any part of the body (Herbst et al, 2015) and there is a great deal of variation between individuals in areas affected. In one study, about 30% of patients with affected lower limbs also had affected arms (Fife et al, 2010). However, anecdotal reports suggest the proportion of patients with affected lower and upper limbs is much higher, particularly in established lipoedema (stage 2 onwards). In about 3% of cases of lipoedema, the arms alone are affected, usually with sparing of the hands (Fife et al, 2010).

In patients with lower limb lipoedema, the lower body will often be disproportionately large: individuals may require clothes for their lower body that are several sizes larger than those needed for their upper body (Fife et al, 2010).

The adipose tissue enlargement may be accompanied by bruising without apparent cause or due to minor trauma only. Many patients with lipoedema also often mention pain and extreme sensitivity/tenderness to touch and pressure in the affected areas. They also report that the affected areas are cooler than unaffected areas. (The skin over obese tissue may also feel cooler because of the insulating effect of fat.)

Patients with lipolymphoedema may mention that standing for long periods, hot environments or weather, and aeroplane journeys may exacerbate pain, swelling and feelings of heaviness in the limbs, probably due to fluid accumulation in the tissues.

Mobility may be restricted due to pain, mechanical hindrance, and/or hip and knee joint problems, particularly in patients with severe lipoedema. There are anecdotal reports of a possible association between lipoedema and hypermobility (Willams & MacEwan, 2016; Lontok et al,

Lipoedema UK and the Royal College of General Practitioners (RCGP) have partnered to develop an online course called *Lipoedema — An Adipose Tissue Disorder*. The Royal College of Nursing (RCN) has endorsed the course, which takes about 30 minutes to complete and can be accessed at: [www.elearning.rcgp.org.uk/lipoedema](http://www.elearning.rcgp.org.uk/lipoedema)

### Box 3. Areas for discussion with a patient suspected of having lipoedema

- Age at onset and association with potential hormonal triggers, e.g. puberty, oral contraceptive use, pregnancy, weight gain
- Areas of the body affected, and whether and how the degree and extent of enlargement or swelling have changed over time
- Effect of dieting, calorie restriction and physical activity/exercising on weight and limb size
- Presence and severity of pain, discomfort or hypersensitivity to touch
- Presence, extent and triggers (if any) of bruising
- Presence of knee or hip pain, and related mobility issues
- Differences in skin texture and temperature between affected and unaffected areas
- Effect of rest or leg elevation on leg size and pain/discomfort in patients with lower limb enlargement
- Effect of prolonged standing, heat or hot weather on swelling and pain/discomfort
- Clothing sizes for upper and lower body
- Impact on:
  - Daily living
  - Mobility (e.g. need for aids such as walking stick or wheelchair)
  - Personal relationships
  - Work
  - Emotional state
- Family history
- Previous investigations and management (including surgery such as liposuction)
- Other medical and surgical history (e.g. comorbidities, regular medication, allergies, previous episodes of cellulitis and previous surgery)
- Reasons for presenting now, understanding of disease, and expectations of treatment outcomes.

2017). Muscle weakness may also play a part: a study in women with lipoedema and women with obesity found that those with lipoedema had statistically significantly lower leg muscle strength (Smeenge, 2013). Some people become so restricted that they are housebound or unable to care for themselves.

In addition, patients with lipoedema may report family history of relatives with similar tissue enlargement. They often mention repeated attempts to lose weight through calorie-restricted diets and exercising that have little or no impact on lipoedema-affected areas and result in weight loss from unaffected areas only (Fife et al, 2010).

Box 3 lists areas for discussion during history taking in a patient suspected of having lipoedema. It is important to recognise that the patient may be presenting for the first time or may have had investigations and management elsewhere previously. Also, in some cases, the patient may have encountered dismissive or negative responses during their contact with health services. Ascertaining the patient's reasons for presenting and their hopes for treatment and outcomes will form a good basis for a partnership approach to management.

### Examination

As lipoedema is a clinical diagnosis, examination is particularly important, and individuals appreciate time taken by

clinicians to examine them. In addition to characteristic signs such as braceleting at the ankles, reduced skin temperature and altered tissue texture may be present and require palpation to detect (Table 1, page 8). Clinicians should check for Stemmer's sign (Box 4), which can assist in differentiating lipoedema from lymphoedema, and for pitting oedema (Box 5, page 8), which if present may indicate lipolymphoedema.

### Differential diagnosis

Part of the reason that lipoedema may be underdiagnosed is that it may be mistaken for other conditions that cause sub-cutaneous tissue enlargement/swelling or fat deposition. The two most frequent misdiagnoses are generalised obesity (particularly in young, otherwise well patients) and lymphoedema (Table 2, page 9).

Medical causes of bilateral symmetrical lower limb swelling are listed in Box 6 (page 10). Infrequent causes of unusual fat deposition include Dercum's disease, polycystic ovary disease, Cushing's syndrome, growth hormone deficiency and lipodystrophies that cause lipohypertrophy (e.g. analbuminaemia) (Box 7, page 11).

### Investigations

Currently, there are no diagnostic tests for lipoedema and the main purpose of investigations is to exclude other diagnoses or to inform lipoedema management strategies.

### Box 4. Stemmer's sign (Lymphoedema Framework, 2006)

Stemmer's sign is negative or not present when a fold of skin can be pinched and lifted up at the base of the second toe or at the base of the middle finger.

A positive sign (a) in a patient with lipoedema, when a fold of skin cannot be lifted, indicates secondary lymphoedema.

Stemmer's sign is usually negative (b) in patients with 'pure' lipoedema.

**Table 1. Characteristic signs of lipoedema that may be found during clinical examination**

| Sign                                                                                                                              | Description                                                                                                                                                                                                                                                                                                                                        |                                                                                       |
|-----------------------------------------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------|
| Subcutaneous tissue enlargement                                                                                                   | <ul style="list-style-type: none"> <li>Usually bilateral and symmetrical without involvement of the hands and feet (at least initially)</li> <li>However, the pattern of areas affected and overall shape may vary between patients</li> </ul>                                                                                                     |    |
| Cuffing or braceleting at the ankles/wrists                                                                                       | <ul style="list-style-type: none"> <li>The tissue enlargement stops abruptly at the ankles or wrists so that there is a 'step' before the feet or hands which are usually unaffected</li> <li>May also be called 'inverse shouldering'</li> </ul>                                                                                                  |    |
| Loss of the concave spaces either side of the Achilles tendon                                                                     | <ul style="list-style-type: none"> <li>Occurs in lower limb lipoedema</li> <li>The concave areas posterior to the malleoli (retromalleolar sulci) and either side of the Achilles tendon are filled in</li> </ul>                                                                                                                                  |                                                                                       |
| Bruising                                                                                                                          | <ul style="list-style-type: none"> <li>Bruising may occur anywhere in areas affected by lipoedema, often with no known cause</li> </ul>                                                                                                                                                                                                            |                                                                                       |
| Altered skin appearance, temperature and texture                                                                                  | <ul style="list-style-type: none"> <li>The skin of affected areas may feel softer and cooler than unaffected areas</li> <li>The skin may have the texture of orange peel or have larger dimples</li> </ul>                                                                                                                                         |  |
| Abnormal gait and limited mobility                                                                                                | <ul style="list-style-type: none"> <li>May be due to bulk of the legs and/or fat pads on the medial aspect of the knees</li> <li>May include: <ul style="list-style-type: none"> <li>- Reduced or poor heel to toe strike during walking</li> <li>- Flat feet</li> <li>- Genu valgum (knock knees)</li> </ul> </li> <li>Muscle weakness</li> </ul> |                                                                                       |
| Stemmer's sign negative (Box 4, page 7)                                                                                           | <ul style="list-style-type: none"> <li>Usually negative</li> <li>A positive Stemmer sign represents failure to pinch a fold of skin at the base of the second toe, and is pathognomonic of lymphoedema</li> </ul>                                                                                                                                  |                                                                                       |
| Pitting oedema (Box 5) in patients with lipoedema and secondary lymphoedema (lipolymphoedema) and/or chronic venous insufficiency | <ul style="list-style-type: none"> <li>Usually absent in the early stages of the disease</li> <li>Patients with lipoedema may find testing for pitting oedema particularly uncomfortable</li> <li>Pitting indicates the presence of excess interstitial fluid and may be present in patients with lipolymphoedema</li> </ul>                       |                                                                                       |

*Pictures supplied courtesy of BSN Medical*

## Box 5. Pitting oedema (Lymphoedema Framework, 2006)

Pitting oedema is a sign of excess interstitial fluid. It can be detected by applying a thumb or finger to tissues with pressure that is sustained for at least 10 seconds. Oedema is present when a dimple or pit remains in the tissues when the pressure is removed. The depth of the pit produced may indicate the severity of the oedema. Repetition of the test across the area suspected of involvement can help to determine the extent of the oedema. N.B. Elucidation of this sign may cause discomfort or pain and should be performed gently.

**Table 2. Differentiating lipoedema from lymphoedema and obesity (Forner-Cordero et al, 2009; Langendoen et al, 2009; Fife et al, 2010; Child et al, 2010; Fetzer & Wise, 2015)**

| Characteristic                    | Lipoedema                                                                                                                         | Lymphoedema                                                                                     | Obesity                                                  |
|-----------------------------------|-----------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------|----------------------------------------------------------|
| Gender                            | • Almost exclusively female                                                                                                       | • Male or female                                                                                | • Male or female                                         |
| Age at onset                      | • Usually 10–30 years                                                                                                             | • Childhood (mainly primary); adult (primary or secondary)                                      | • Childhood onwards                                      |
| Family history                    | • Common                                                                                                                          | • Only for primary lymphoedema                                                                  | • Very common                                            |
| Areas affected                    | • Bilateral<br>• Usually symmetrical<br>• Most frequently affects legs, hips and buttocks; may affect arms<br>• Feet/hands spared | • May be unilateral or bilateral depending on cause                                             | • All parts of the body<br>• Usually symmetrical         |
| Effect of dieting on condition    | • Weight loss will be disproportionately less from lipoedema sites                                                                | • Proportionate loss from trunk and affected limbs                                              | • Weight reduction with uniform loss of subcutaneous fat |
| Effect of limb elevation          | • Absent or minimal                                                                                                               | • Initially effective in reducing swelling; may become less effective as the disease progresses | • None                                                   |
| Pitting oedema (Box 5, page 8)    | • Absent or minor in the early stages of the disease                                                                              | • Usually present but pitting may cease as the disease progresses and tissues fibrose           | • No                                                     |
| Bruises easily                    | • Yes                                                                                                                             | • Not usually                                                                                   | • No                                                     |
| Pain/discomfort in affected areas | • Often<br>• Hypersensitivity to touch in affected areas                                                                          | • May be uncomfortable<br>• No hypersensitivity to touch                                        | • No                                                     |
| Tenderness of affected areas      | • Often                                                                                                                           | • Unusual                                                                                       | • No                                                     |
| Skin consistency                  | • Normal or softer/looser                                                                                                         | • Thickened and firmer                                                                          | • Normal                                                 |
| History of cellulitis             | • Unusual (unless lipolymphoedema is present)                                                                                     | • Often                                                                                         | • Unusual                                                |
| Stemmer's sign (Box 4, page 7)    | • Usually negative (unless secondary lymphoedema is present)                                                                      | • Usually positive                                                                              | • Usually negative                                       |

### Laboratory tests

Routine screening blood tests useful in excluding or identifying other or concomitant conditions, especially if weight gain and lethargy are present, may include urea and electrolytes (U&Es), full blood count (FBC), thyroid function tests (TFTs), liver function tests (LFTs), plasma proteins (including albumin), brain natriuretic peptide (BNP – a test for congestive heart failure) and glucose (Forner-Cordero et al, 2012; NVDV, 2014).

Even though hormonal factors are thought to contribute to the development of lipoedema, there is no evidence that endocrinological tests will detect any abnormalities (NVDV, 2014). Similarly, blood tests to measure the levels of inflammatory markers, such as C-reactive protein (CRP) or erythrocyte sedimentation rate (ESR) are unlikely to provide abnormal results.

### Imaging investigations

Imaging investigations such as ultrasound scans, magnetic resonance imaging (MRI) scans and computed tomography (CT) scans are usually not necessary to diagnose lipoedema, but may have a role if there is diagnostic uncertainty.

Lymphoscintigraphy, a method of imaging the lymphatic system that involves injection of radioactive tracers into the skin, should detect lymphoedema (Keeley, 2006).

Ultrasound measurement of dermal thickness may help to differentiate lymphoedema and lipoedema (Naouri et al, 2010).

Venous duplex ultrasound scanning may be indicated if chronic venous insufficiency is suspected (Wounds UK, 2016).

## Hand-held devices

There is increasing interest in the potential role of hand-held devices that measure the electrical properties of skin and superficial subcutaneous tissues as a way of differentiating lipoedema and lymphoedema. The reading obtained (the tissue dielectric constant) is a measure of the amount of water in the tissues. Higher readings indicate higher water content. Although patients with lymphoedema have been found to have higher readings than patients with lipoedema, further research is needed to determine the role of this technology in diagnosis and management (Birkballe et al, 2014).

Another device under development examines the effect of a small area of suction over affected tissues. The suction is maintained for 30 seconds and an associated smartphone app videos the skin being tested (Levin-Epstein, 2016).

In patients with lipoedema, the suction is thought to produce characteristic skin changes that do not occur in patients without the disease. A pilot trial is underway (Levin-Epstein, 2016).

## Classification and staging

Lipoedema has been classified according to:

- Distribution of the adipose tissue enlargement
- The shape of the enlargement (Table 3).

However, these classifications are of limited clinical use because neither indicates severity or disease progression, and neither guides treatment.

The first system devised to describe the severity and progression of lipoedema comprised three stages. More recent versions include a fourth stage to account for the development of lipolymphoedema (Table 4, page 11). However, as oedema can arise at any stage of lipoedema (Fife et al, 2010), inclusion of this fourth stage is potentially confusing.

The staging system in Table 4 may indicate a patient's position in the progression of lipoedema. However, it does not take account of the severity of symptoms, e.g. pain and impact on lifestyle, neither of which is necessarily related to the degree of tissue enlargement.

### Box 6. Other causes of bilateral lower limb chronic oedema (Ely et al, 2006; Traves et al, 2013)

- Chronic venous insufficiency (CVI)
- Congestive cardiac failure
- Dependency or stasis oedema
- Obesity
- Hepatic or renal dysfunction
- Hypoproteinaemia
- Hypothyroidism
- Pregnancy and premenstrual oedema
- Drug-induced swelling, e.g. calcium channel blockers, steroids, non-steroidal anti-inflammatories.

*N.B. These conditions will usually cause pitting oedema, and may co-exist with lipoedema*

**Table 3. Classifications of lipoedema (Meier-Vollrath & Schmeller, 2004; Földi & Földi, 2006; Langendoen et al, 2009; Herbst, 2012a)**

| According to the anatomical areas affected                                                   |                                                                                                   |
|----------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------|
| Type                                                                                         | Anatomical areas affected                                                                         |
| Type I    | Pelvis, buttocks and hips (saddle bag phenomenon)                                                 |
| Type II   | Buttocks to knees, with formation of folds of fat around the inner side of the knees              |
| Type III  | Buttocks to ankles                                                                                |
| Type IV   | Arms                                                                                              |
| Type V    | Lower leg                                                                                         |
| According to the shape of the tissue enlargement                                             |                                                                                                   |
| Type                                                                                         | Description                                                                                       |
| Columnar                                                                                     | Enlargement of the lower limbs which become column-shaped or cylindrical                          |
| Lobar                                                                                        | Presence of large bulges or lobes of fat overlying enlarged lower extremities, hips or upper arms |
| <i>Columnar lipoedema is much more common than lobar lipoedema</i>                           |                                                                                                   |

*Pictures supplied courtesy of BSN Medical*

Table 4. Lipoedema staging (Schmeller &amp; Meier-Vollrath, 2007; Herbst, 2012a; NVDV, 2014)

| Stage |                                                                                    | Description                                                                                                                                                                                                                                                |
|-------|------------------------------------------------------------------------------------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| 1     |   | <ul style="list-style-type: none"> <li>• Skin appears smooth</li> <li>• On palpation, the thickened subcutaneous tissue contains small nodules</li> </ul>                                                                                                  |
| 2     |   | <ul style="list-style-type: none"> <li>• Skin has an irregular texture that resembles the skin of an orange ('peau d'orange') or a mattress</li> <li>• Subcutaneous nodules occur that vary from the size of walnut to that of an apple in size</li> </ul> |
| 3     |   | <ul style="list-style-type: none"> <li>• The indurations are larger and more prominent than in Stage 2</li> <li>• Deformed lobular fat deposits form, especially around thighs and knees, and may cause considerable distortion of limb profile</li> </ul> |
| 4     |  | <ul style="list-style-type: none"> <li>• Lipoedema with lymphoedema (lipolymphoedema)</li> </ul>                                                                                                                                                           |

Pictures supplied courtesy of BSN Medical

### Future developments

Some studies of the impact of liposuction (see pages 29–31) on patients with lipoedema have used assessments of symptoms and functioning to monitor outcomes (Schmeller et al, 2012; Baumgartner et al, 2016).

Questionnaires were used to grade spontaneous pain, pain upon pressure, oedema, bruising, restricted movement, cosmetic impairment and reduction of quality of life on a five-point scale. Scores for individual items as well as a total score were compared pre- and post-operatively.

The Expert Working Group suggested that a similar approach that considers symptoms and functioning could be developed to indicate non-surgical treatment needs and response in patients with lipoedema. The Group also suggested that the terminology 'mild', 'moderate' or 'severe' is more intuitive than the use of stages, and that each grade could include scope for indicating whether

secondary lymphoedema is present (i.e. whether lipolymphoedema is present).

Such a system would need to be defined fully and formally validated, but could be based on a scoring system for each of the following items:

- Degree of limb enlargement
- Level of pain
- Presence and extent of bruising
- Presence and extent of lymphoedema
- Alterations in gait
- Restrictions to mobility
- Restrictions to performing activities of daily living
- Impact on quality of life.

The scoring system would need to be clear and simple. Total scores could be used to indicate whether the patient falls into the mild, moderate or severe grade. In addition, the system could be used for monitoring, e.g. changes in total or individual item scores could be used to assess the effectiveness of management approaches.

**Box 7. Other diseases that may have unusual patterns of fat deposition** (Sam, 2007; Florenza et al, 2011; Herbst, 2012a; Kandamany & Munnoch, 2013; Melmed, 2013; Nieman, 2015)

- **Dercum's disease** — individuals have painful fatty nodules often accompanied by a wide range of other symptoms including fatigue; may be on the 'lipoedema spectrum'
- **Multiple symmetrical lipomatosis (Madelung's disease)** — painless symmetrical tumour-like accumulations of fat in the subcutaneous tissues
- **Polycystic ovary disease** — a hormonal disorder with increased production of androgen hormones often accompanied by generalised obesity
- **Cushing's syndrome** — due to excess cortisol production; obesity is one of a wide range of symptoms and may be accompanied by a characteristic dorsal fat pad
- **Growth hormone deficiency** — causes may include pituitary disease or trauma; the accompanying obesity is often centralised
- **Lipodystrophies that cause lipohypertrophy (e.g. analbuminaemia)** — rare; may be congenital or acquired.

## Assessment

Assessment of a patient with lipoedema should be holistic and aim to define the patient's current disease severity, to indicate suitability for management options and to signal need for referral (Figure 2). In practice, diagnosis and assessment are often conducted concurrently and elements of the two processes often overlap.

### *Degree and extent of adipose tissue enlargement*

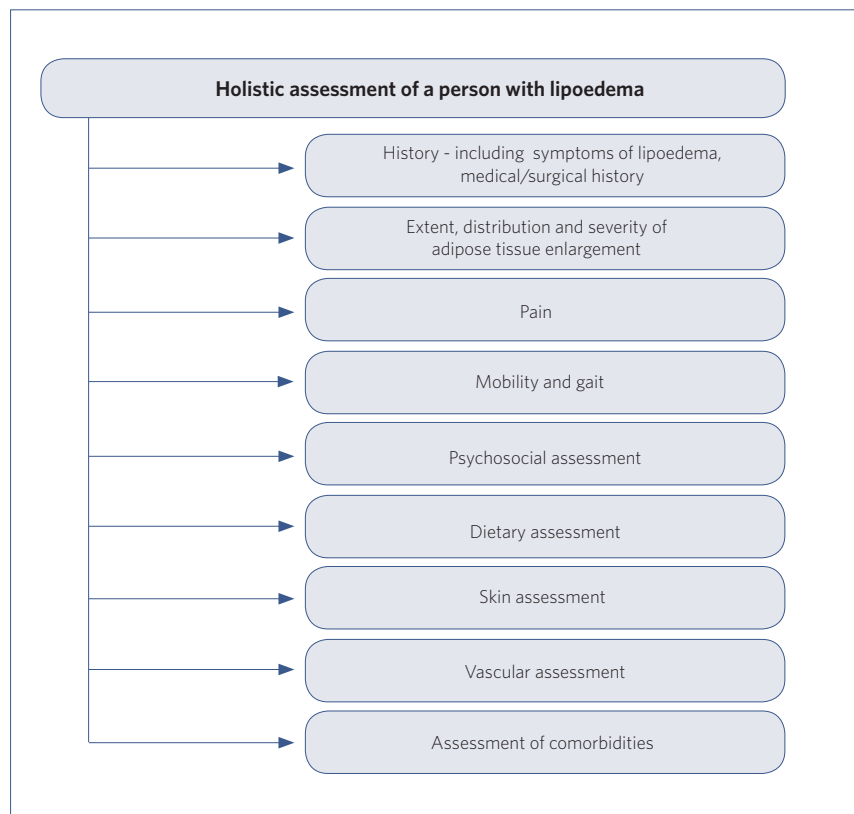
Measurement of the degree and extent of adipose tissue enlargement in lipoedema is not straightforward and is not used for diagnosis. However, sequential measurements may be useful for assessment and monitoring purposes. A wide range of types of measurement may be employed, from bodyweight to limb volume measurement (Table 5, page 13).

In general, simple methods are likely to be the most useful and the easiest to use consistently. Clinicians may find that they tailor the measurement method used to the needs of individuals. Documentation of the details of the measurement method used is important to ensure that future measurements are performed consistently and that changes detected are not artefacts of differences in measurement location or technique.

For some patients, tracking measurements is highly motivating. However, the distortion and flaccidity of the tissues in patients with lipoedema may make measurement impractical. In such situations, serial photographs may be useful.

Body mass index (BMI) is a measure of the ratio between weight and height. It is used widely to define and diagnose obesity and to monitor efforts to lose weight. In lipoedema, however, BMI is likely to be high even when the person is not obese and is therefore of limited value (Reich-Schupke et al, 2013).

It should be noted that measurement for fitting compression garments is a separate process from measuring for monitoring purposes. Where available, clinicians should follow the measuring requirements for compression garments as stipulated by the manufacturer (see pages 23–27).



### *Pain*

Pain is a common and often distressing feature of lipoedema that can impact significantly on daily life. The pain may take several forms, including aching, heaviness, tenderness or pain on touch. The cause of the pain is unclear, but may be related to compression of nerves and/or inflammation (Lontok et al, 2017).

Pain may also be related to joint problems, especially of the knees and hips, arising from increased tissue laxity that may cause joint misalignment or hypermobility, or from degenerative changes (Hodson & Eaton, 2013).

Assessment should aim to determine the cause, nature, frequency, site, severity and impact of the pain. Rating scales can be used to ask patients to quantify their pain at the initial and ongoing assessments. Rating scales include:

- Numerical rating scale — e.g. individuals are asked to rate their pain on a scale from 0 to 10, where 0 is no pain and 10 is the worst pain imaginable
- Visual analogue scale (VAS) — e.g. individuals are asked to mark or indicate the level of pain on a 10cm line where 0cm is no pain and 10cm is the worst pain imaginable (Dansie & Turk, 2013).

*Figure 2: Holistic assessment of a patient with lipoedema*

**Table 5. Measurement for assessment and monitoring in lipoedema (de Koning et al, 2007; Langendoen et al, 2009; Lopes et al, 2016; Madden & Smith, 2016)**

|                       |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                   |
|-----------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Weight                | <ul style="list-style-type: none"> <li>The simplest method of monitoring change in body size</li> <li>Not a specific measure of body areas affected by lipoedema</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                       |
| Waist                 | <ul style="list-style-type: none"> <li>Waist measurement provides information about the distribution of body fat</li> <li>Increased waist circumference can be used to indicate whether a person is overweight or obese, and is associated with increased risk for metabolic syndrome (<math>\geq 80</math> cm and <math>\geq 94</math> cm for Caucasian women and men, respectively)</li> <li>Not a specific measure of a body area usually affected by lipoedema, but may be useful in helping to avoid obesity and to monitor efforts to lose non-lipoedematous fat</li> </ul> |
| Waist to hip ratio    | <ul style="list-style-type: none"> <li>A higher waist-to-hip ratio (waist circumference <math>\div</math> hip circumference; using same units) is associated with increased risk for metabolic syndrome and cardiovascular disease (<math>\geq 0.85</math> for women and <math>\geq 0.90</math> for men)</li> <li>In lower limb lipoedema waist to hip ratio may be unreliable because of disproportionate adipose tissue enlargement over the buttocks and upper thighs. A changing ratio may be due to a reduction in waist size or an increase in hip size</li> </ul>          |
| Circumferential       | <ul style="list-style-type: none"> <li>For example, in lower limb lipoedema: at ankle, calf, knee, thigh</li> <li>A simple method, but requires consistent use of measurement location for meaningful monitoring over time</li> </ul>                                                                                                                                                                                                                                                                                                                                             |
| Limb volume           | <ul style="list-style-type: none"> <li>Limb volume measurement is a complicated process</li> <li>Methods include water displacement and the use of computer programs that calculate volume from circumferential limb measurements taken at 4cm intervals with a spring-tension tape</li> </ul>                                                                                                                                                                                                                                                                                    |
| Body mass index (BMI) | <ul style="list-style-type: none"> <li>A ratio that is calculated by dividing weight by height squared (weight (kg) <math>\div</math> height<sup>2</sup> (m<sup>2</sup>))</li> <li>Widely used to diagnose obesity (BMI <math>\geq 30</math>) and monitor weight change</li> <li>Of limited value in patients with lipoedema</li> </ul>                                                                                                                                                                                                                                           |

### *Mobility and gait*

Patients with lipoedema should be asked about mobility and observed when walking so that gait and footwear can be assessed. Shape distortion and fat pads at the inner knee area may alter gait, which in turn may cause other problems in the legs, knees, hips and back. Lipoedema may hinder mobility because of tissue bulk, pain or hip and knee problems.

Muscle strength may also be reduced: a study of quadriceps strength found that patients with lipoedema had significantly lower strength than people with obesity (Smeenge, 2013).

Asking whether aids are needed for walking and in what circumstances may highlight issues that may otherwise have gone unmentioned. Patients with lipoedema may also have flat feet or genu valgum (knock knees) and require podiatric biomechanical assessment. Restricted ankle mobility (e.g. poor ankle dorsiflexion) and reduced heel to toe movement with reduced heel strike may induce a laboured or plodding gait. This may

contribute to oedema if present by reducing the effectiveness of the foot and calf muscle pump on venous return.

### *Psychosocial assessment*

Patients with lipoedema may suffer considerable psychosocial distress and have significantly reduced quality of life (Box 8 and Box 9, page 14). The initial relief of finding out what is wrong when a diagnosis is received is often followed by feelings of frustration and despair when the patient realises that treatment may not improve symptoms as much as they had hoped.

The social stigma attached to increased body size and physical restrictions, coupled with shame and embarrassment can damage self-esteem, lead to difficulties with personal relationships and work, and cause mental health issues including anxiety and depression (Hodson and Eaton, 2013; Kirby, 2016; Fetzer & Fetzer, 2016).

Practical difficulties, such as those due to reduced mobility and difficulties in finding clothes that fit, along with fear of

### **Box 8. Lipoedema UK's Big Survey 2014 key findings on quality of life (Fetzer & Fetzer, 2016)**

The 250 respondents to Lipoedema UK's Big Survey 2014 reported that lipoedema had a considerable impact on their lives:

- 95% reported difficulty in buying clothes
- 87% reported that lipoedema had a negative effect on quality of life
- 86% reported low self esteem
- 60% reported restricted social life
- 60% reported feelings of hopelessness
- 51% reported that lipoedema had an impact on ability to carry out their chosen career
- 50% reported restricted sex life
- 47% reported feelings of self blame
- 45% reported eating disorders
- 39% felt that lipoedema had restricted their career choices.

## Box 9. Quality of life assessment in lipoedema

Formal quality of life assessment is usually reserved for research purposes or for health economic evaluations undertaken for regulatory purposes. General tools available include the Short-Form (36) Health Survey (SF-36) (Lins & Carvalho, 2016). Currently, there is no quality of life assessment tool for people with lipoedema, although a tool has been developed for people with lymphoedema (LYMQOL) (Keeley et al, 2010). A Patient Benefit Index, a scoring system that evaluates the benefit of treatment from the individual's perspective, has been developed for people with lymphoedema and lipoedema (Blome et al, 2014).

### The illness narrative

An illness narrative (Hyden, 1997) is derived from an individual's explanation of their struggle with a chronic or disabling illness. It is their story of living with the condition.

For decades, the medical profession was sceptical about the veracity of a person's description of their illness. Yet the words of the individual are likely to provide the most accurate account of what it is like to live with a condition. Through such narratives the complexity of the illness experience can be seen. As Hyden (1997) stated: "One of our most powerful forms for expressing suffering and experiences related to suffering is the narrative. Patients' narratives give voice to suffering in a way that lies outside the domain of the biomedical voice."

Gathering information using the illness narrative enables clinicians to gain a more complete understanding of how the condition is impacting each individual person and therefore how to best meet their needs, in particular, how to better address their psychosocial needs.

discrimination or not fitting into seats in public spaces, may discourage a patient from leaving their home, resulting in social avoidance, withdrawal and isolation. These issues may be compounded by lack of understanding and fear expressed by family, friends and colleagues. Patients with lipoedema have also reported receiving verbal abuse from members of the public (Kirby, 2016).

Patients with lipoedema should be asked about their home situation (e.g. accessibility, general living standards, household members, involvement of carers), activities of daily living, social interactions, recreational/physical activities and exercise. Psychological assessment should include evaluation of mood for signs of depression or anxiety, ability to cope, energy levels and sleep quality.

Assessment should also include gaining an understanding of the patient's insight into the condition and their personal goals and expectations of the components and outcomes of treatment.

### Dietary assessment

Many patients with lipoedema have tried repeatedly and often unsuccessfully over many years to reduce the size of the affected areas through dieting and physical activity or exercise. These efforts may

have produced weight loss from non-lipoedematous areas, but may also have resulted in disordered eating behaviours, including anorexia nervosa, binge eating and bulimia (Fife et al, 2010; Forner-Cordero et al, 2012; Williams & MacEwan, 2016; Todd, 2016; Fetzer & Fetzer, 2016). However, up to half of patients with lipoedema may also be overweight or obese (Langendoen et al, 2009; Fife et al, 2010).

Dietary assessment should be approached sensitively and include:

- Current diet, eating habits, and fluid and alcohol consumption
  - Previous attempts to lose weight and the effects of these
  - The patient's:
    - Beliefs about eating, weight gain and physical activity
    - Willingness to change
    - Understanding of the role of diet in the management of lipoedema
- (NICE CG189, 2014).

### Skin assessment

Skin should be assessed for general condition and the effectiveness of personal care. The skin of patients with lipoedema is soft and easily damaged and some patients develop ulceration. It is particularly important to examine any skin folds as these may develop friction or moisture-related skin damage, and fungal or bacterial infections.

### *Vascular assessment*

Compression therapy is an important element of the management of lipoedema. Patients with lipoedema should undergo vascular assessment according to local protocol. Significant arterial disease is a contraindication to compression therapy (Wounds UK, 2015).

The vascular assessment should include consideration of signs, symptoms and risk factors for arterial disease. Doppler ultrasound to determine ankle-brachial-pressure index (ABPI) is a method often used for vascular assessment. However, tissue enlargement may make it difficult to get an accurate ABPI in patients with lipoedema.

Furthermore, inflation of a cuff around the limb may be very painful for patients with lipoedema.

### *Comorbidities*

Comorbidities should be identified and management optimised to minimise impact on patients with lipoedema. Patients with lipoedema have self-reported the presence of several conditions: fibromyalgia, gluten allergy (coeliac disease), hypothyroidism, polycystic ovary syndrome, vitamin D deficiency and arthritis (Herbst et al, 2015; Smidt, 2015; Williams & MacEwan, 2016). However, evidence of direct links between lipoedema and many of these conditions is currently very limited.

More information on the role of ABPI in determining suitability for compression therapy can be found in the *Wounds UK Best Practice Statement on Compression Hosiery*, which is available at: [www.wounds-uk.com](http://www.wounds-uk.com)

### **Key points**

1. The diagnosis of lipoedema is made on clinical grounds: there are no diagnostic tests for the condition
2. Lipoedema is a condition that is distinct from lymphoedema
3. Lipoedema may have a significant impact on a patient's physical and mental health and wellbeing
4. Patients with lipoedema generally report a history of bilateral symmetrical limb enlargement, with sparing of the hands and feet, which is not responsive to dieting. They may also report pain, sensitivity to touch and easy bruising, and a family history of similar tissue enlargement and shape disproportion
5. Affected areas of the body may be softer and cooler, with a texture that is dimpled or resembles a mattress
6. The presence of pitting oedema in affected areas indicates lipolymphoedema
7. Routine blood tests may be useful to exclude or identify other conditions
8. Imaging investigations are not used routinely
9. Further work is required to develop a classification/staging system for lipoedema that takes into account disease progression along with symptoms such as pain or restrictions to mobility
10. Holistic assessment should include the degree and extent of adipose tissue enlargement, presence and level of pain, mobility and gait, psychosocial assessment, dietary assessment, skin assessment, vascular assessment and assessment of any comorbidities
11. Psychosocial assessment is particularly important in people with lipoedema because of the long-term nature of the disease and the importance of self-management.

## SECTION 3 : PRINCIPLES OF MANAGEMENT

Lipoedema is a long-term condition that has wide-ranging impacts on the health and psychosocial wellbeing of patients. Consequently, an interprofessional or multidisciplinary approach to management is often required. However, there is currently inconsistency and inequity across the UK in referral patterns and care for patients with lipoedema.

Patients recognised as possibly having lipoedema in a primary care setting may be referred to a lymphoedema service, where available, for investigation, diagnosis, management and co-ordination of care. However, there is variation throughout the UK in provision of lymphoedema services, and some services do not have sufficient capacity to manage patients with lipoedema. Where there is no provision of lymphoedema services, a referral to vascular or plastic surgery services may be appropriate.

Even so, the Expert Working Group concluded that lymphoedema services are the most appropriate setting for the management of patients with lipoedema, not least because of the expertise held within these services in differentiating the two conditions and in the use of compression therapy. The Group considers that improved recognition of the disease and appropriate referral patterns are reliant on enhancing awareness and recognition of the disease in primary care settings, and in the wider provision of lipoedema/lymphoedema services.

Third sector organisations, such as Lipoedema UK ([www.lipoedema.co.uk](http://www.lipoedema.co.uk)) and Talk Lipoedema ([www.talklipoedema.org](http://www.talklipoedema.org)), provide help with self-management and are important sources of peer support.

### Principles of lipoedema management

The management of lipoedema requires a holistic approach (Figure 3) that includes:

- Facilitating and enhancing the patient's ability to self-care and cope with the physical and psychosocial impact of the condition
- Managing symptoms
- Optimising health and preventing disease progression.

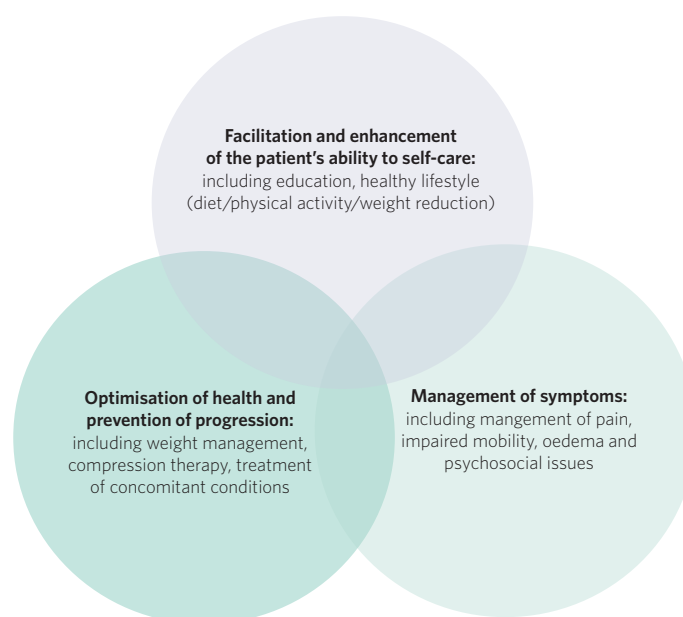


Figure 3: Principles of lipoedema management

In keeping with the NHS goal for personalised care for people with long-term conditions, clinicians should take a collaborative approach to the management of a patient with lipoedema, providing individualised care plans according to need and person-centred treatment goals (NHS Outcomes Framework; Coulter et al, 2013; WHO, 2004; Woods & Burns, 2009; Welsh Assembly Government, 2007).

The main components of lipoedema management are:

- Psychosocial support, management of expectations and education, including family planning, pregnancy advice and genetic counselling
- Healthy eating and weight management
- Physical activity and improving mobility
- Skin care and protection
- Compression therapy
- Management of pain.

Each element needs to be tailored according to the severity of symptoms, degree and complexity of tissue enlargement, whether there has been progression to lipolymphoedema, and the psychosocial status of the patient.

Patients with lipoedema may be well informed about their condition and possible

management routes following internet searching and participation in social media. However, the advice and information found may not be necessarily grounded in evidence. Individuals may be susceptible to misinformation and may need help in understanding what is best practice and most likely to be of benefit based on current evidence, and what is not yet clear or may be detrimental. Such discussions require a sympathetic, non-judgemental approach to avoid discouraging or offending individuals in their efforts to improve their condition.

Discussions should also bear in mind that individuals are often very vulnerable and sensitive after a long journey to diagnosis, which may have included disheartening and upsetting comments from healthcare professionals seen previously.

Support and encouragement alongside working in partnership with the patient and their carer(s) with careful management of expectations, including sensitive discussions about the life-long nature of the condition, should underpin the best practice management of lipoedema.

Clinicians specialising in the management of lipoedema have a key role in providing education and support around a healthy lifestyle, and in implementing and managing compression therapy. Potential roles for other members of the multidisciplinary team are listed in Table 6. It should be noted that referral may not always be available within the NHS; where available, individual services may have specific restrictions and criteria for referral. Private referrals may be possible for patients with sufficient financial resources. Primary care and community-based services have an important role in supporting and enabling self-care and ensuring referral when appropriate (Todd, 2016).

### Benefits of lipoedema management

Lipoedema is a long-term condition that is not curable. However, management of lipoedema according to best practice has the potential to produce benefits including:

- Reduction in pain
- Improved limb shape
- Avoidance of impairment or improvement in mobility
- Management or avoidance of obesity

**Table 6. Involvement of the multidisciplinary team in the management of lipoedema**

| Indication                                                                                                                                                                                                                                                                                  | Clinician/service                                                                                 |
|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|---------------------------------------------------------------------------------------------------|
| <ul style="list-style-type: none"> <li>• Tissue enlargement ± oedema</li> <li>• Pain, aching, sensitivity to touch</li> </ul>                                                                                                                                                               | → Lipoedema/lymphoedema specialist clinician                                                      |
| <ul style="list-style-type: none"> <li>• Abnormal gait</li> <li>• Muscle weakness</li> <li>• Joint pain</li> </ul>                                                                                                                                                                          | → Physiotherapist                                                                                 |
| <ul style="list-style-type: none"> <li>• Mobility problems</li> <li>• Difficulty with day-to-day activities</li> </ul>                                                                                                                                                                      | → Occupational therapist                                                                          |
| <ul style="list-style-type: none"> <li>• Advice and education about weight management, healthy eating, disordered eating, nutritional supplements, diabetes</li> </ul>                                                                                                                      | → Dietitian                                                                                       |
| <ul style="list-style-type: none"> <li>• Flat feet</li> <li>• Abnormal gait</li> </ul>                                                                                                                                                                                                      | → Podiatrist                                                                                      |
| <ul style="list-style-type: none"> <li>• Unmanageable/chronic pain</li> </ul>                                                                                                                                                                                                               | → Pain clinic                                                                                     |
| <ul style="list-style-type: none"> <li>• Concomitant conditions</li> </ul>                                                                                                                                                                                                                  | → Appropriate specialist service (e.g. vascular service, diabetic clinic, psychological services) |
| <ul style="list-style-type: none"> <li>• In carefully selected patients, after non-surgical approaches have been implemented: <ul style="list-style-type: none"> <li>• Severe tissue enlargement causing mobility impairment</li> <li>• Management of severe obesity</li> </ul> </li> </ul> | → Plastic surgeon<br>→ Bariatric surgeon                                                          |

### Patient pathway

Appendix 2, page 33, summarises the patient pathway through assessment and management

- Reduced likelihood of progression to lipolymphoedema
- Where present, reduced severity of lipolymphoedema and reduced risk of complications such as cellulitis
- Minimisation of secondary joint problems, such as knee and hip osteoarthritis
- Minimisation of impact on ability to perform daily activities, including work
- Enhanced ability to self-care
- Improved psychosocial wellbeing.

These effects are also likely to result in wider benefits to the healthcare system including an overall reduction in healthcare utilisation due to lipoedema and for obesity-related conditions such as diabetes.

Although there is currently no evidence that early treatment improves prognosis in lipoedema, the Expert Working Group considers that early diagnosis, intervention and initiation of self-care would produce the greatest health and economic benefits. As yet, no formal health economic analyses have been done on the impact of lipoedema management.

### Key points

1. A multidisciplinary approach to the management of lipoedema is necessary
2. Management aims to manage symptoms, to facilitate and enhance the patient's ability to self-care and optimise health and to prevent disease progression
3. The main components of lipoedema management are: psychosocial support and education, healthy eating, weight management, physical activity, skin care, compression therapy and management of pain.

## SECTION 4: PSYCHOSOCIAL SUPPORT AND SELF-CARE

Patients with lipoedema may experience similar psychological and social challenges to those faced by people with other long-term conditions. Recently published research on the psychosocial impact of lipoedema found that psychological flexibility and social connectedness were associated with higher quality of life irrespective of severity of symptoms (Dudek et al, 2016).

Psychological flexibility was defined as willingness to contact difficult, unpleasant thoughts, feelings and sensations in order to engage in a valued activity; social connectedness was defined according to a scale that measured feelings of belonging and connection with friends and society.

In addition, research into a range of other chronic diseases has concluded that:

- Psychological adjustment has a major impact on chronic disease outcomes and can be aided by encouraging people to stay as active as possible, to express their emotions and to engage in self-care
- People who have a healthy diet, engage in physical activity/exercise or employ other aspects of self-management have fewer symptoms, better functional capability and fewer complications than those who do not
- Mental health issues, such as depression, anxiety or psychological distress, can compromise the ability to self-manage which in turn may lead to feelings of helplessness and hopelessness (de Ridder et al, 2008; Dekker & de Groot, 2016).

Recognition of the importance of and need for emotional and psychological support that encourages self-efficacy (Box 10) is crucial to the successful management of lipoedema. In providing person-centred, empathetic care that is realistic but positive in managing expectations, clinicians play a key role in supporting and empowering patients to adjust to their new circumstances, engage with self-care and seek additional help when needed.

Support can be provided in several ways including discussions during clinic or health centre visits, and the provision of written materials or links to websites, and information on support organisations (Box 11, page 19) and education programmes. Peer-led groups may have a particular role in reducing feelings of isolation. Potential barriers to self-care (Box 12, page 19) should be identified and addressed where possible.

Many patients with lipoedema adjust well, and may even be able to identify positive outcomes of their diagnosis, such as a greater appreciation for life and reassessed life priorities. However, some patients may benefit from interventions such as group/individual counselling, cognitive behavioural therapy (CBT) (Box 13, page 19) or mindfulness (Mantzios & Wilson, 2015). Unfortunately, access to these interventions within the NHS is variable.

Mental health issues such as depression or anxiety may need treatment according to National Institute of Health and Care Excellence (NICE) guidance (NICE CG91, 2009; NICE CG113, 2011). Furthermore, clinicians should be alert to hidden mental health issues in carers (Turner & Kelly, 2000).

Patients with lipoedema who have or are planning to have children may have concerns and anxieties about genetic and hereditary aspects of lipoedema. These concerns may impact their ability to accept the condition and the treatment strategies that may be used. Clinic appointments can provide an opportunity to discuss these worries and to put in place referrals for genetic counselling or to a lymphoedema clinic as appropriate for the patient or other family members.

### Box 10. Self-efficacy (Frei et al, 2009; Lorig et al, 2001; Adams et al, 2010; Hoffman, 2013)

Self-efficacy (the extent or strength of a person's belief in their own ability to produce specific goals) influences health outcomes and may also affect level of healthcare utilisation. Although self-efficacy has not been measured in patients with lipoedema, research in other chronic diseases indicates that high self-efficacy is an important mediator in altering health behaviours. In addition, improvements in self-efficacy generated through self-management or educational programmes have been associated with reductions in healthcare utilisation and symptom burden.

### Box 11. UK lipoedema support organisations

- Lipoedema UK\* – [www.lipoedema.co.uk](http://www.lipoedema.co.uk)
- Talk Lipoedema – [www.talklipoedema.org](http://www.talklipoedema.org)
- British Lymphology Society (BLS) – [www.thebls.com](http://www.thebls.com)
- Lymphoedema Support Network – [www.lymphoedema.org](http://www.lymphoedema.org)

N.B. A number of overseas and international organisations also exist; their websites can be accessed via internet searching

*\*Lipoedema UK has produced an advocacy pack, which is available on request, that contains information on accessing services*

### Box 12. Potential barriers to self-care in patients with lipoedema

- **Previously ineffective care and long duration** of the disease that has entrenched a standpoint that nothing can be done to improve the situation
- **Lack of knowledge and skills** to support treatments and behaviour changes, e.g. lack of understanding of the role of compression therapy in improving symptoms and of the role of physical activity and healthy eating in improving symptoms and overall health, inability to apply compression garments correctly and safely
- **Severity of the condition**, e.g. increased bulk, the presence of pain, reduced mobility or concomitant conditions, may hinder physical activity or ability to self-apply compression garments
- **Poor relationship with healthcare professionals** may result in a lack of trust with unwillingness to take healthcare advice or to seek help when needed
- **Low self-esteem, self-efficacy or social/emotional support** may hinder ability to take responsibility and implement self-care
- **Financial restrictions**, e.g. difficulty paying for transport to appointments may prevent opportunities for education and encouragement
- **Inaccurate or misleading information**, e.g. from social media sites or the internet may cause confusion or reinforce misconceptions.

### Box 13. Cognitive behavioural therapy (CBT) in lipoedema

- CBT is a type of psychotherapy that can help people by changing the way that they think and behave
- CBT is recommended by the National Institute for Health and Care Excellence (NICE) for patients who have a mental health condition, an eating disorder, body dysmorphic disorder, or a chronic physical health problem with depression (NICE CG31, 2005; NICE CG9, 2004; NICE CG91, 2009; NICE CG113, 2011)
- In patients with lipoedema, CBT has the potential to help with a range of issues, including encouraging realistic but optimistic attitudes, treating depression, encouraging self-management and improving functioning (de Ridder et al, 2008; Deter, 2012; Fetzer, 2016).

### Key points

1. In common with other chronic conditions, psychosocial support underpins the management of lipoedema and is important in encouraging self-management and realistic expectations
2. Clinicians need to identify and help patients to deal with potential barriers to self-care
3. Mental health issues may affect carers as well as patients with lipoedema.

## SECTION 5: HEALTHY EATING AND WEIGHT MANAGEMENT

Nutrition plays an important role in the management of lipoedema, not just in weight management but also in engaging the patient, providing a sense of control over the evolution of lipoedema, and reducing the risk of obesity-related conditions such as diabetes and joint degeneration (Todd, 2010; Todd, 2016).

All patients with lipoedema who are not overweight or obese should be encouraged to eat healthily to avoid weight gain through accumulation of non-lipoedema fat. Avoidance of weight gain will prevent deleterious effects on general health and possibly slow progression of the condition (Todd, 2010).

Patients with lipoedema are likely to have tried a variety of diets and may have a complex relationship with food. In Lipoedema UK's Big Survey 2014, 98% of participants reported trying to lose weight: 82% had lost some weight, but this was lost evenly across the body in only 5% (Fetzer & Fetzer, 2016).

Failure to lose weight and feeling unattractive may lead to a cycle of emotional or comfort eating and further weight gain. Denial that obesity is present, the use of the diagnosis of lipoedema as an explanation for weight gain due to overeating, and fixed ideas about diet and 'good'/'bad' foods may complicate attempts to discuss diet and weight loss.

Anecdotally, weight loss programmes have little or no effect on the amount of tissue enlargement in lipoedema (Todd, 2010; Fife et al, 2010). Any weight loss that does occur is likely to be disproportionately lower in lipoedema-affected areas than in unaffected areas. However, a significant proportion of patients with lipoedema also have obesity (Langendoen et al, 2009). These individuals should be encouraged to reduce the amount of non-lipoedema fat tissue through diet and physical activity. Benefits include improved general health with a likely reduction in risk for cardiovascular disease and diabetes, less strain on joints and muscles with potential benefits for mobility, and a probable reduction in the risk of developing lipolymphoedema (Fonder et al, 2007; Langendoen et al, 2009; Todd, 2010).

Key to effective weight management is support and advice enabling patients to find a nutritional plan that suits their food preferences and lifestyle. Despite the plethora of dietary advice available on the internet and other media, there is no clinical evidence to support the use of a particular dietary plan in lipoedema.

Some patients advocate the Harvie and Howell diet which involves a restricted calorie intake for two days each week and eating a Mediterranean-style diet on the other days (Harvie & Howell, 2014). Anecdotally, this regimen appears to result in easier, longer-lasting weight loss (Fetzer & Wise, 2015; Todd, 2016).

The Rare Adipose Disorders (RAD) diet has also been advocated for patients with lipoedema (Todd, 2016). The basis of this diet is reduced consumption of pasteurised dairy products, animal fats, simple sugars, carbohydrates, salt and artificial preservatives, flavours and sweeteners (Herbst, 2012b). However, as with other diets, evidence of effectiveness in patients with lipoedema is awaited.

Where weight loss is advisable, patients with lipoedema are likely to benefit from referral to a dietetic-led service. A stepwise approach to goal setting is important to enable success and prevent discouragement. A non-prescriptive, sympathetic approach that supports healthy eating and maintains a dialogue about food and diet is very important.

Some patients with lipoedema find attending proprietary weight loss programmes that involve weekly meetings with weigh-ins and discussions motivating. When discussing healthy eating, the focus should be on encouraging sustainable healthy, balanced changes in eating habits.

Referral according to local protocol for eating disorders may be necessary if there is evidence of a problem, such as anorexia, bulimia or binge eating (NICE CG9, 2004).

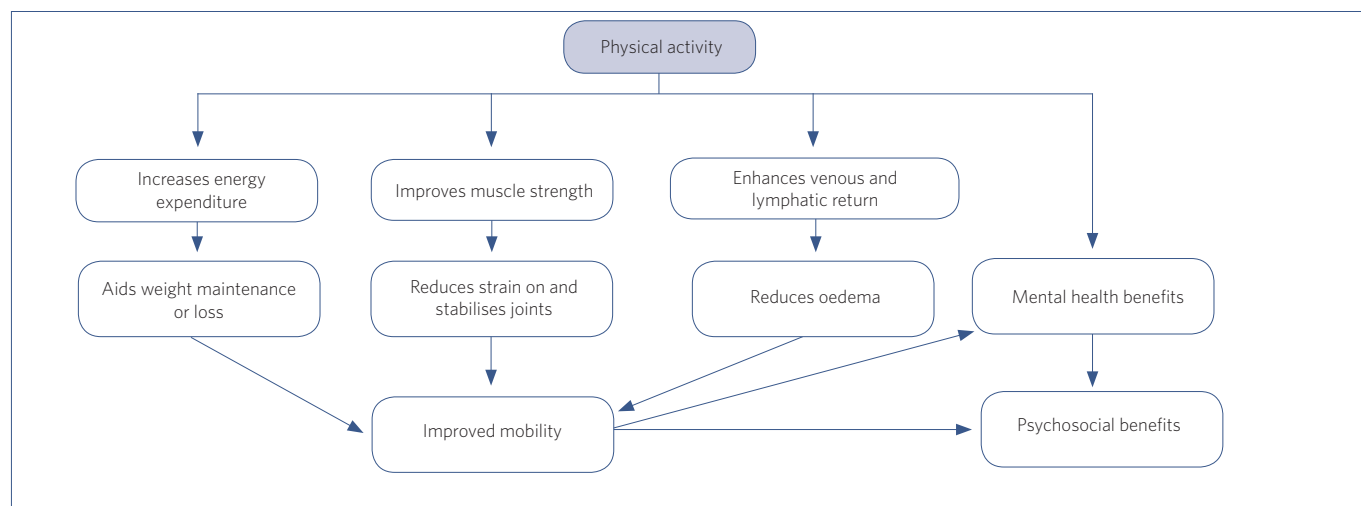


Figure 4: Potential benefits of physical activity in patients with lipoedema

### Dietary supplements

Some healthcare practitioners recommend dietary supplements for people with lipoedema. Currently, there is no robust clinical evidence supporting the use of dietary supplements and further work is needed to identify which may be of benefit and in what ways. Discussions around supplements need to be approached with sensitivity to prevent alienation and loss of trust.

### Physical activity and improving mobility

Increased physical fitness and activity in the general population has been demonstrated to have numerous health benefits, including reduced mortality, reduced rates of obesity, diabetes, cardiovascular disease and cancer, and improved mental health and quality of life (Salmon, 2001; Penedo & Dahn, 2005; Bishop-Bailey, 2013).

Current UK guidelines recommend that adults aged 19 to 64 years should be active daily and each week undertake at least 2.5 hours of moderate intensity activity. They also recommend that adults should undertake physical activity to improve muscle strength at least two days each week (Davies et al, 2011). In addition, the guidelines strongly recommend minimising sedentary behaviour, e.g. by reducing the amount of time spent watching television or using a computer, taking regular breaks at work, and walking for part of journeys

undertaken by bus or car (Davies et al, 2011). Physical activity in patients with lipoedema can also have a number of benefits including weight maintenance or loss and improved mobility (Figure 4) (Fetzer & Wise, 2015). The psychological benefits of physical activity may help to combat negative feelings associated with lipoedema.

Patients with lipoedema who have severely impaired mobility, abnormal gait and/or pain/joint problems should seek the advice of a physiotherapist on suitable types of activity.

Patients with lipoedema embarking on increased physical activity should start to increase activity levels slowly, aiming for some form of physical activity every day. The apparently small gains made will increase confidence.

High intensity exercise or activities that cause or aggravate pain or bruising should be avoided (Fetzer & Wise, 2015). Low intensity activities include walking, water-based exercises, yoga, pilates and the use of resistance bands. However, patients already involved in a high intensity form of exercise should not be discouraged from participating, but may need to consider how to minimise the risks of joint strain and bruising.

Exercising in water, e.g. water aerobics, may be particularly beneficial in patients with lipoedema as the support provided by the water reduces strain on and aids the range of

### Key points

1. Although attempts to lose weight may not have an impact on enlarged adipose tissue, preventing or reducing obesity in other parts of the body through healthy eating and physical activity will help to prevent deterioration in general health
2. There is no clinical evidence supporting the use of a particular diet. Patients should be encouraged to find a healthy, balanced diet that suits their needs and lifestyle
3. Patients with lipoedema should be encouraged to be physically active and undertake activities that suit their needs and lifestyle, while taking into account that some patients may have individual limitations.

motion of joints, reduces pain, and improves venous and lymphatic return (Fetzer & Wise, 2015). Over 75% of people with lipoedema who answered the question about exercising in water in Lipoedema UK's Big Survey 2014 found it to be helpful (Fetzer & Fetzer, 2016).

Exercise in water can take the form of swimming or formal exercise classes (aqua-exercise/aerobics). Patients who cannot swim need not be deterred: simply walking in water is good exercise and the pressure exerted by the water on the tissues is beneficial.

### Overcoming barriers to exercise and physical activity

The term 'exercise' may be worrying and imply high intensity exercise in a gym, particularly for people who are mainly sedentary, have severe lipoedema and/or

restricted mobility. Use of the term 'physical activity' and reassurance that increasing activity does not necessarily need to involve exercise classes may be more successful in encouraging increased levels of movement. Suggestions could include home-based exercise, chair exercises, walking, using the stairs rather than a lift, or parking further from the supermarket door.

Feeling self-conscious and embarrassed, along with difficulties finding sports garments and swimwear that fit, can be major barriers to exercising in public or participating in exercise classes for some people with lipoedema. Solutions may include enrolling in single sex classes, wearing a sarong between the changing rooms and pool, and asking a friend or relative to attend, especially when starting a new class.

## SKIN CARE AND PROTECTION

# SECTION 6: SKIN CARE AND PROTECTION

Patients with lipoedema should be encouraged to pay particular attention to gently drying any skin folds after washing, and to applying appropriate emollients on a daily basis, particularly when the skin is dry (Williams & MacEwan, 2016). Folds in the skin may be prone to irritation and the development of fungal infections that require treatment with antifungal agents (Langendoen et al, 2009).

Where feasible, patients may prefer to avoid procedures such as taking routine blood samples, injections and blood pressure in lipoedema-affected areas (Todd, 2016).

Patients with lipolymphoedema are at increased risk of cellulitis and should be advised to protect themselves from insect bites, burns, scratches and other skin injuries in the affected areas. Cellulitis is

a spreading bacterial infection of the skin and subcutaneous tissues (Al-Niami & Cox, 2009). Local signs include warmth, swelling, erythema, pain and lymphangitis (inflammation of the lymphatic vessels that may be seen as red streaks), and are often accompanied by raised body temperature and feeling unwell. Skin necrosis and abscess formation can occur (Morris, 2008). Prompt treatment with antibiotics is required: severe cases may require intravenous antibiotics (Al-Niami & Cox, 2009).

More information about the treatment of cellulitis in lymphoedema, which would be relevant to those with lipolymphoedema, can be found in the recently updated guidelines from the British Lymphology Society and Lymphoedema Support Network (BLS/LSN, 2016).

### Key points

1. Skin folds may be prone to fungal infections and should be washed and dried with care
2. Patients with lipolymphoedema are at increased risk of cellulitis.

## SECTION 7: COMPRESSION THERAPY

As the names suggests, compression therapy is designed to exert pressure on body tissues. In lipoedema, the use of compression therapy has three main purposes:

- To reduce discomfort, aching and pain by supporting the tissues
- To support tissues and streamline uneven, distorted limb shape and so by reducing mechanical impairment to movement, improve mobility
- To reduce oedema in lipolymphoedema by reducing interstitial fluid formation and encouraging venous and lymphatic return (Reich-Schupke et al, 2013; NVDV, 2014; Fetzter & Fetzter, 2015).

Compression therapy will not reverse the adipose tissue enlargement of lipoedema (Fetzter, 2016). Therefore, unless there is oedema present, compression therapy will not produce a reduction in limb size. Apparent decreases in limb size may occur while wearing compression therapy, however, due to the streamlining effect of compression therapy in limbs that are considerably distorted in shape by tissue lobes or pads.

Compression therapy may also prevent lipoedema worsening and decrease the risk of progression to lipolymphoedema, although evidence for such effects is not currently available (Fonder et al, 2007; Langendoen et al, 2009; Todd, 2010). By aiding mobility, patients with lipoedema who wear compression therapy may be able to be more active (Reich-Schupke et al, 2013).

Over time, consistent use of compression therapy may reshape limbs to a degree; in particular it may help to reduce ankle cuffing (Reich-Schupke et al, 2013; Hodson & Eaton, 2013). Once lipolymphoedema has developed, compression therapy becomes especially important (Todd, 2010).

### Assessment and contraindications

Before the selection of compression therapy, vascular assessment according to local protocol is essential to determine whether arterial compromise is present and to what extent. Compression therapy is contraindicated in patients with severe peripheral arterial disease,

severe peripheral neuropathy and uncontrolled heart failure (Lymphoedema Framework, 2006; Wounds UK, 2015).

Careful assessment of a patient with lipoedema is required to determine which type(s) of compression therapy are indicated. Assessment should include the severity and extent of the lipoedema, including the presence of skin folds or fat lobes, whether oedema is also present, the presence of pain, the goals of treatment and the patient's ability to tolerate and self-manage compression garments. In practice, availability on prescription is a major influencing factor in the type of compression therapy selected.

Clinicians will need to explain the rationale for the use of compression therapy and the need for daily wear and long-term use to maximise concordance.

### Types of compression therapy

There are several types of compression therapy (Table 7, page 24). The type used most commonly for patients with lipoedema is the compression garment (called compression hosiery when used on lower limbs). In patients with lipolymphoedema, multi-layer inelastic bandaging may be used initially to reduce the oedema (Lymphoedema Framework, 2006).

Compression garments can be bought ready-to-wear ('off-the-shelf') or can be custom-made to an individual's requirements. The fabric used in compression garments may be:

- Circular knit — garments are produced by knitting on a round knitting cylinder to produce a shaped fabric tube that does not have a seam; they tend to be thinner than flat knit garments, but more likely to cut in to soft skin or around lobes
- Flat knit — garments are usually produced by knitting a flat shaped piece of fabric that is then stitched together with a longitudinal seam; some garments have seam-free sections; they tend to be thicker and firmer than circular knit garments and more suitable when there is uneven or distorted limb shape (Clark & Krimmel, 2006).

### Compression or containment?

For patients who find compression therapy difficult to tolerate and who have 'pure' lipoedema (i.e. do not have lipolipoedema), the concept of containment may be helpful. The word containment may communicate better the intended purpose of the compression garments in these patients, i.e. supporting the tissues to improve the shape, contour and also possibly the function of the affected areas, while not implying that the volume or condition of the affected area will be improved.

**Table 7. Compression types and roles in the management of lipoedema (Lymphoedema Framework, 2006; Hodson & Eaton, 2013; Reich-Schupke et al, 2013; Wounds UK, 2015; Williams and MacEwan, 2016; Todd, 2016; Williams, 2016; Fetzer, 2016)**

| Type                                                                         | Description                                                                                                                                                                                                                                                                                                                                                                                  | Notes and role in the management of lipoedema                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                  |
|------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| Compression garments                                                         | <ul style="list-style-type: none"> <li>Available ready-to-wear or custom-made</li> <li>Available in a range of styles, e.g. hosiery (below knee, thigh length, leggings, tights, capri-style) with or without feet, and arm sleeves</li> <li>Often available in a limited colour selection only (beige or black), although some companies provide a wider range of colours</li> </ul>        | <ul style="list-style-type: none"> <li>Custom-made flat knit garments may allow more accurate fitting and accommodate uneven or distorted limb shape better than circular knit garments</li> <li>In patients who have skin folds, fat lobes and soft tissues, ready-to-wear circular knit garments may be prone to cutting into tissues and may cause distal oedema; custom-made flat knit garments may be more appropriate</li> <li>If the feet are not involved, there is no oedema and no risk factors for oedema, footless garments can be prescribed</li> <li>Garments may be available with hook and loop (VELCRO®) or zip fastenings</li> <li>Applicators may be required to assist with donning and doffing</li> </ul> |
| Adjustable compression wraps                                                 | <ul style="list-style-type: none"> <li>Sections of inelastic fabric joined together that wrap around the limb and are secured by straps with hook and loop fixings</li> <li>More rigid than compression garments</li> <li>Available in a variety of styles for the upper and lower limbs, including below knee/ elbow +/- foot/hand; full-length limb length +/- hand/foot; thigh</li> </ul> | <ul style="list-style-type: none"> <li>Designed to allow easy application and removal by the patient or carer; often easier to use than compression garments</li> <li>Mainly used to reduce oedema</li> <li>More rigid than compression garments and so less likely to cut in</li> <li>Level of compression for different sections can be adjusted easily</li> <li>More durable than compression garments</li> <li>May be used post-operatively following liposuction</li> </ul>                                                                                                                                                                                                                                               |
| Compression bandaging (N.B. In the USA, bandages are sometimes called wraps) | <ul style="list-style-type: none"> <li>Multi-layer inelastic bandaging systems usually comprise tubular bandage, a padding layer and inelastic bandages</li> </ul>                                                                                                                                                                                                                           | <ul style="list-style-type: none"> <li>Used to reduce oedema, particularly in severely distorted and painful limbs</li> <li>Generally not used for pure lipoedema</li> <li>Can be applied to legs or arms</li> <li>May need to be reapplied daily especially during initial use as the oedema reduces</li> <li>Bandaging of toes or fingers may also be required if affected by oedema</li> <li>Often need to be applied by a clinician; self-application is not easy, but can be taught</li> </ul>                                                                                                                                                                                                                            |
| Intermittent pneumatic compression (IPC)                                     | <ul style="list-style-type: none"> <li>An inflatable plastic garment with one or more chambers that are inflated and deflated cyclically by an electrical air pump; sessions last 30–120 minutes</li> </ul>                                                                                                                                                                                  | <ul style="list-style-type: none"> <li>Main indication is reduction of oedema, which is achieved through the peristaltic massaging effect produced by the inflation/deflation cycles</li> <li>May help to reduce pain, even in the absence of oedema</li> </ul>                                                                                                                                                                                                                                                                                                                                                                                                                                                                |

## Compression therapy selection

Selection of compression therapy (Figure 5, page 25 and Table 7) for patients with lipoedema needs to take account of a wide range of factors including:

- Location, extent and severity of:
  - Tissue enlargement
  - Shape distortion
  - Deep skin folds and fat lobes
- Presence and degree of accompanying:
  - Pain or tenderness
  - Secondary oedema (lipolymphoedema)
- Lifestyle, mobility and preference
- Access to and availability of compression type, and to the expertise needed to apply/fit and use the compression safely and optimally (particularly important for made to measure garments)
- Availability on prescription/cost.

## Informed choice and shared decision making

A patient's willingness to wear and ability to tolerate compression therapy is key to ensuring concordance. Engaging the person in the decision-making process, ensuring that they understand why compression therapy is being prescribed and what the benefits are likely to be, as well as empowering them to manage the garments/devices themselves are important for successful implementation.

Despite the recommendation that patients with lipoedema wear compression as much as possible, every day and during exercise (Fetzer & Wise, 2015; Hardy, 2015), Lipoedema UK's Big Survey 2014 revealed that only 55% of respondents did so 'most days' or 'every day' (Fetzer & Fetzer, 2016). The main barriers to

wearing compression were reported to be discomfort and difficulty putting it on (Fetzer & Fetzer, 2016).

A recent survey revealed that 50% of patients who used compression garments found them unhelpful, most often due to poor fit. Patients who received garments from specialist clinicians based in a lymphology clinic had a better experience (Fetzer & Wise, 2015). Individual preference, accurate fitting and the provision of advice or devices to aid donning and doffing should therefore be given high priority when planning a compression regimen.

Clinicians need to discuss options after ascertaining the patient's priorities. Devising personalised strategies that meet a patient's needs may require a creative and flexible approach, e.g.:

- Starting at low levels of compression and building gradually may improve tolerance

- Several options may need to be tried before finding the products and treatment regimens that best suit the patient.

Finding products that are acceptable to the patient and providing effective symptom relief is critical to a good long-term outcome and to ensure cost-efficiency of care (Williams & MacEwan, 2016).

## Compression garments and adjustable wraps

For patients with mild to moderate tissue enlargement and no obvious oedema, circular knit, ready-to-wear compression garments are usually the first choice. Where there is more significant tissue enlargement with soft skin, deep skin folds and fat lobes, circular knit garments are likely to cut in to tissues. For these patients, flat knit custom-made garments are more suitable because the fabric is more rigid and able to bridge skin folds without cutting in.

## Useful resources

- Selection and use of compression hosiery — see: Wounds UK *Best Practice Statement Compression Hosiery* (2nd edition). London, Wounds UK, 2015. Available from: [www.wounds-uk.com](http://www.wounds-uk.com)
- Use of compression therapy, including bandaging, in the management of lymphoedema — see: *Best Practice for the Management of Lymphoedema*. London, MEP Ltd, 2006. Available from: [www.woundsinternational.com](http://www.woundsinternational.com) and [www.lympho.org/publications/](http://www.lympho.org/publications/)

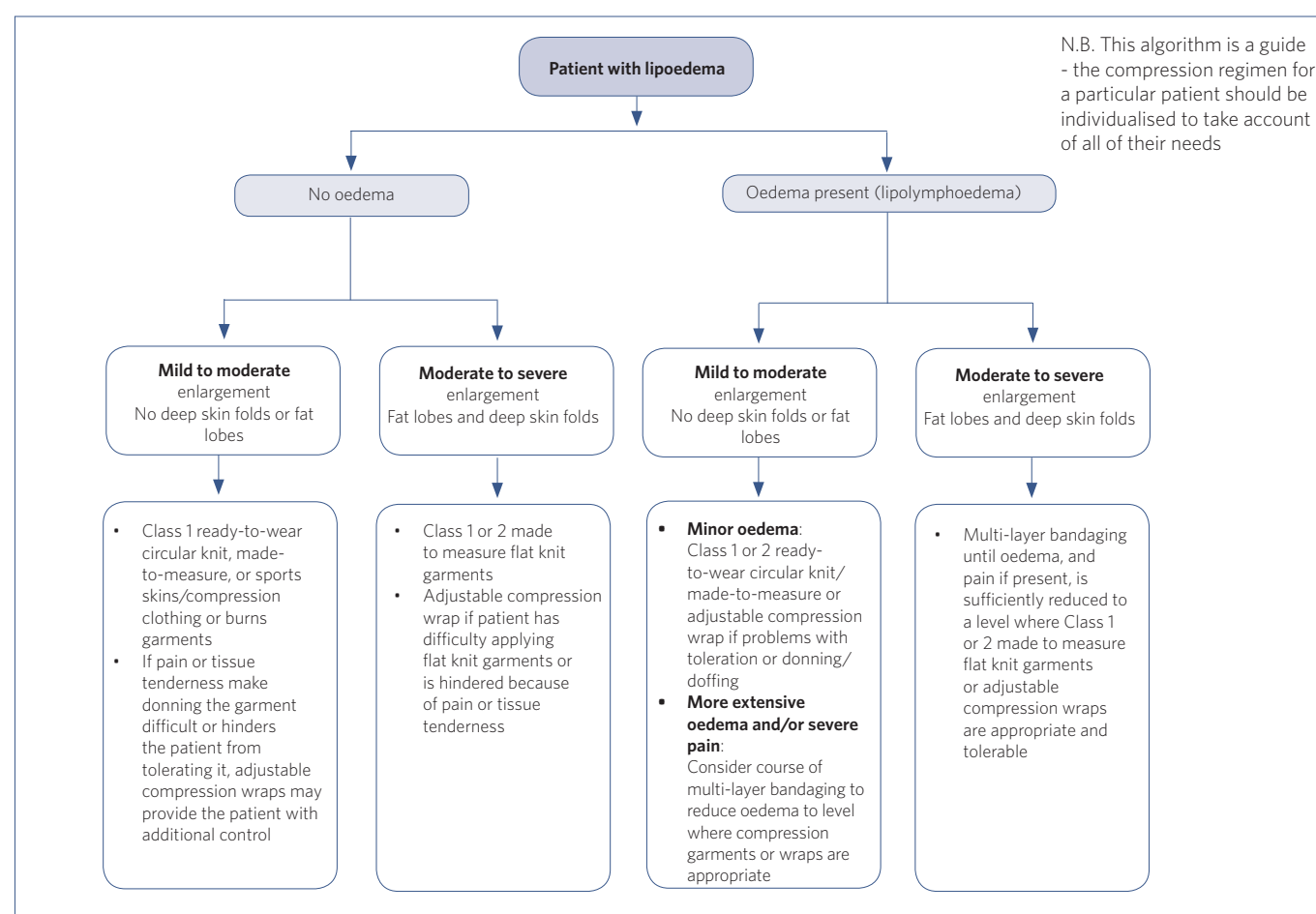


Figure 5: Compression therapy choice in lipoedema

Adjustable compression wraps may be easier and less painful to apply than compression garments. These also have the advantage of allowing the patient to adjust the degree of compression to some extent, and are less likely to cut in to soft tissues. Another option may be garments with hook and eye fastenings intended for wear after liposuction. Multi-layer bandaging may be helpful for particularly painful limbs as the level of pressure can be adjusted and additional padding added where needed.

Alternatives to prescribed compression garments for patients with mild lipoedema without significant shape distortion or oedema include sports skins/compression clothing or burns garments, both of which may be softer but provide support. However, such garments are not always available on prescription and advice from a specialist practitioner may be required.

### *Combining compression therapy types*

For some patients, it may be possible to combine types of compression therapy. For example, for a patient with severely enlarged thighs but lower legs that are affected to a lesser extent, an adjustable compression wrap for the thighs or compression shorts (Bermuda-style or longer Capri-style) could be combined with hosiery for the lower leg (Fetzer, 2016). Similar combinations may also be helpful for patients who find it difficult to put on full-length stockings.

It may also be appropriate for patients to have different types of compression therapy for different activities or situations, e.g. to use adjustable wraps when at home and circular knit garments when going out or exercising (Williams & MacEwan, 2016).

### *Compromise*

Despite these options, therapeutic compromise may sometimes be necessary to ensure that the patient is wearing at least a low level of compression and does not disengage with treatment (Hodson & Eaton, 2013). Patients who are very reluctant to wear compression therapy, could be encouraged to wear it during activities that are more likely to cause discomfort or additional swelling, such

as standing, walking, exercising or travelling (Fetzer & Wise, 2015).

### **Lipolymphoedema**

The type of compression therapy suitable for the treatment of lipolymphoedema is dependent on the degree of tissue enlargement and shape distortion and pain. For patients without significant shape distortion, compression garments may be sufficient to reduce and control oedema. However, for more severe oedema and in patients with significant shape distortion, multi-layer bandaging may be necessary to reduce oedema to the point where reassessment for compression garments is feasible.

Some patients with lipolymphoedema and moderate/severe oedema may need to receive therapy in line with best practice for the management of lymphoedema (Lymphoedema Framework, 2006; Fonder et al, 2007). This may include manual lymphatic drainage (MLD) (see page 28), multi-layer bandaging, skin care, and exercise and movement.

### **Measuring for and prescribing compression garments and adjustable compression devices**

Measuring for compression garments and adjustable compression devices is a complex task and should be undertaken by a clinician with specialist training and experience in selecting the most appropriate type, style, fastening type and compression strength of garment or device. Ease of application and removal should be given high priority in the decision process. Ensuring an accurate fit is essential to encouraging wear and to preventing problems.

Once a compression garment or adjustable compression device has been selected and is available, an experienced clinician will need to assess fit, teach the patient/carer how to don and doff, and explain care and the review/replacement process (Box 14, page 27). The need for garment/device renewal provides an opportunity to review progress and outcomes, and to check concordance, suitability of style and level of compression, fit and the patient's/carer's ability to put on and take off the garment.

**Box 14. Tips for the session for the first fitting of a compression garment or adjustable compression device (Hardy, 2015; Fetzer & Wise, 2015; Wounds UK, 2015)**

- Check that the garment fits well, e.g. does not dig into tissues, particularly at the ankles and knees
- Demonstrate to patients and carers how to don and doff the garment or device including how to spread the fabric evenly, and help them to practise doing the same
- Advise on application and removal aids — the use of an applicator, along with closely fitting rubber gloves and non-slip matting, is often invaluable; many application and removal aids are available on prescription
- Educate the patient and carers about when to remove the garment and who to contact if there are problems — signs that indicate compression should be removed immediately include increased pain, numbness, pins and needles or discoloured digits
- Manage expectations — explain that compression/containment is not a cure and, unless oedema is present will not reduce limb size, but may improve limb outline and improve symptoms
- Explain care of the garments — the manufacturer's recommendations should be followed: some garments/devices can be machine washed but others need to be hand washed; in general, harsh detergents and fabric softener should be avoided, and the garment/device should be air dried rather than tumble dried
- Discuss short-term and long-term review and renewal schedules — generally compression garments need to be replaced on average every 6 months.

**Intermittent pneumatic compression**

The main use of intermittent pneumatic compression (IPC) (Table 7, page 24) is as an adjunct to other forms of compression therapy to reduce oedema in patients with lipolymphoedema (Rapprich et al, 2015). IPC is thought to reduce swelling due to oedema in two ways: to reduce oedema formation by opposing capillary filtration and to encourage oedema resolution by increasing venous and lymph flow (Feldman et al, 2012).

IPC is also sometimes used as an alternative in patients with lipo-lymphoedema who

do not want to use compression garments or devices (Fetzer, 2016). Patients also report that IPC helps to reduce pain and discomfort even in the absence of oedema (Reich-Schupke et al, 2013).

Patients with lipolymphoedema who have pain and tenderness may find IPC difficult to tolerate unless used at very low pressures. Home use of IPC is possible after careful assessment: some clinics will loan IPC devices, alternatively individuals can also buy devices independently.

**Key points**

1. Compression therapy is used in lipoedema to reduce pain and support tissues. In lipolymphoedema it is also used to reduce swelling due to oedema
2. Compression therapy does not reverse adipose tissue enlargement
3. Patients being considered for compression therapy should undergo arterial assessment to exclude peripheral arterial disease
4. Choice of compression therapy depends on a wide range of factors, including individual choice and ability to manage
5. The main type of compression therapy used in lipoedema is compression garments
6. Most ready-to-wear garments are circular knit, which produces a thinner fabric that may be more prone to cutting into tissues
7. Most custom-made garments are flat knit, which produces a thicker more rigid fabric. These garments may be more suitable if there is considerable limb distortion
8. Adjustable compression wraps may be useful for patients who find applying garments difficult or painful, and can be used alongside compression garments applied to other body areas
9. Multi-layer bandaging may be useful in patients with lipolymphoedema as an initial step to reduce oedema and/or pain to a level where garments become manageable
10. Measurement and fitting of compression garments should be undertaken by appropriately trained and competent clinicians
11. Garments generally need to be replaced every 6 months
12. Intermittent pneumatic compression (IPC) may be used as an adjunct to compression therapy in patients with lipolymphoedema.

## SECTION 8: OTHER NON-SURGICAL APPROACHES

### Manual lymphatic drainage

Manual lymphatic drainage (MLD) is a very specific but gentle type of massage carried out by qualified specialists/practitioners (Box 15). MLD moves the skin in order to stimulate the activity of lymph vessels to remove interstitial fluid and relieve oedema.

Studies of the effect of MLD have found that the therapy reduced sympathetic nervous system activity (involved in 'fight or flight' reactions) and increased parasympathetic nervous system activity (involved in the 'rest and digest' state) (Kim et al, 2009; Kim, 2013). A review of evidence for the effect of MLD in lymphoedema concluded that there is some evidence that MLD reduces pain and discomfort and promotes physical and psychological functioning (Haesler, 2016).

MLD can be used as part of the treatment regimen for patients with lipoedema to help manage symptoms and when compression therapy does not control oedema sufficiently (Rapprich et al, 2015). However, patients with pure lipoedema report that MLD may also help to relieve pain and discomfort (Todd, 2016).

Although little research has been done specifically on the use of MLD in the treatment of lymphoedema and it is not generally available via the NHS (Langendoen et al, 2009), many patients feel that they derive psychological benefits from the opportunity to relax.

### Kinesiology taping

Kinesiology taping involves the application of a series of narrow strips of stretchable adhesive tape to the skin over the area to be treated. It was first developed in Japan and has been used most widely to treat sports and other soft tissue injuries (Kalron & Bar-Sela, 2013). It is thought that the tape moves and lifts the skin and subcutaneous tissues to improve blood circulation and lymph drainage (Wu et al, 2015), and may help to stabilise and realign tissues and joints (Kurt et al, 2016).

There is no definitive evidence that kinesiology taping provides benefits to patients with lipoedema as very little research has been conducted so far. However, there are anecdotal reports that kinesiology taping improves symptoms such as knee pain and plantar fasciitis. It may also help to streamline limb shape.

Patch testing is required to ensure no skin sensitivity to the tape. Once applied, the tape is left in place for up to 3–4 days. Patients can be taught the technique for use at home (Fetzer, 2016).

### Other treatment modalities

Two other approaches used by some in the management of lipoedema may act by stimulating lymphatic drainage. There are anecdotal reports that these methods help to manage symptoms and reduce oedema. However, no research is yet available to demonstrate benefit in patients with lipoedema.

- **Electrostatic massage therapy (Deep Oscillation®)** — a device is applied to the skin in a massaging movement by a therapist holding the device with a glove hand while the patient holds a linked electrode; this creates an electrostatic effect in the tissues that is suggested may reduce pain and inflammation (Teo et al, 2016)
- **Self-lymphatic drainage or dry skin brushing** — stroking movements using the hand or a very soft dry bristled brush may be used; to promote lymphatic drainage centrally, the patient may use breathing techniques or massage on the trunk followed by stroking movements on the limb, always moving towards the centre of the body. Care must be taken not to traumatise the skin (Fetzer, 2016; Williams & MacEwan, 2016).

#### Box 15. Finding an MLD practitioner

- Further details about MLD and a list of registered MLD practitioners can be found at: [www.mlduk.org.uk/therapists/](http://www.mlduk.org.uk/therapists/)
- The British Lymphology Society has a directory of lymphoedema treatment services, including MLD, at: [www.thebls.com/directory/](http://www.thebls.com/directory/)

### Key points

1. Manual lymphatic drainage (MLD) stimulates the activity of the lymphatic system and may be used in conjunction with compression therapy to reduce oedema and control symptoms such as pain in lipolymphoedema
2. Some patients with pure lipoedema find MLD helps to reduce pain and discomfort
3. Kinesiology taping may help to improve blood and lymph circulation and to stabilise and realign tissues and joints

## SECTION 9: SURGICAL MANAGEMENT

Surgical options that may be appropriate for some patients with lipoedema include liposuction (to treat the tissue enlargement) and bariatric surgery (to treat obesity) (Box 16). However, while both types of surgery may help with symptoms, neither has been shown to be curative of lipoedema itself.

### Liposuction

Liposuction (Box 16) should be carried out by a surgeon who is appropriately qualified to treat someone with lipoedema and who works as part of a multidisciplinary team.

Access to liposuction within the NHS is often limited and where available may be classified as a cosmetic procedure for which patients with lipoedema do not qualify. Advocacy groups such as Lipoedema UK are endeavouring to change the situation and to have liposuction recognised as an effective surgical treatment for patients with lipoedema.

Due to lack of NHS provision and/or long waiting lists, patients may decide to source liposuction privately in the UK or abroad. Patients should research clinics carefully to ensure an adequate standard of care and to establish that the procedures on offer are appropriate for their individual needs. They need to be aware that liposuction procedures are not without risk in the immediate post-operative period and may cause long-term complications (Stutz & Krah, 2009; Rapprich et al, 2015).

### *Effects of liposuction in lipoedema*

Overall, liposuction in patients with lipoedema reduces tissue bulk, pain and bruising, and improves mobility, functioning and quality of life (Reich-Schupke et al, 2012; Peled & Kappos, 2016).

A study of tumescent liposuction in 85 patients with lipoedema found that six months after surgery patients' scores for a wide range of symptoms, including pain, bruising, swelling and impaired mobility, were all significantly reduced in comparison

with pre-operative scores ( $p < 0.001$  for change in each item score) (Rapprich et al, 2015). Patients received a mean of 2.61 (range 1 to 6) sessions of liposuction.

There is also evidence of longer-term benefits of liposuction. A study sent a questionnaire to 112 patients who had undergone tumescent liposuction between 5 and 11 years previously and who had also been evaluated by questionnaire four years before (Schmeller et al, 2012; Baumgartner et al, 2016). Responses were received from 76% of patients. Changes over time in seven parameters (including pain, bruising, oedema, mobility and quality of life) that contributed to an overall impairment score were examined.

The significant reductions in pre-operative and post-operative scores for each item and for overall score (all  $p < 0.001$ ) noted at 4 years were also present after 8 years (Schmeller et al, 2012; Baumgartner et al, 2016). However, studies are awaited that present data for the longer-term outcomes that are relevant for this patient group.

### *Advising patients with lipoedema*

Patients with lipoedema considering liposuction should be advised and encouraged to undertake non-surgical treatment for at least 6-12 months as a first step. Box 17, page 30, lists factors that may be considered by surgeons when assessing a patient's suitability for liposuction.

Pre-operative counselling is very important to ensure that the patient has realistic expectations of what can be achieved, understands the procedure and the importance of post-operative care (including compression therapy), and comprehends that there is no evidence that liposuction is curative (Box 18, page 31). Provision of such advice is highly variable. Consequently, it may fall to lipoedema clinic staff to ensure that patients have had an opportunity to discuss these issues.

### Box 16. Definitions of liposuction and bariatric surgery (Shridharani et al, 2014; Albaugh et al, 2016)

- **Liposuction:** the removal of subcutaneous adipose deposits via a cannula attached to a suction device that is inserted through small incisions in the skin. Large volumes of fatty tissue can be removed from areas such as the legs, hips and arms. Liposuction is one of the most commonly performed cosmetic surgical procedures. There are numerous types of liposuction, including tumescent and water-jet assisted. The type used in the treatment of lipoedema may relate to the preferences of the individual surgeon.
- **Bariatric surgery:** gastrointestinal surgery used to treat obesity that aims to reduce intake of food or reduce absorption of food from the gut. A wide range of procedures is available including gastric banding, partial gastrectomy and gastric bypass.

## Box 17. Factors that may be considered by surgeons when assessing the suitability of a patient with lipoedema for liposuction

- **Lipoedema stage:** individual surgeons use different lipoedema stage criteria for indicating suitability for liposuction
- **Concomitant conditions:** patients should not have medical conditions that increase the risk of complications from anaesthesia
- **Weight management:** non-lipoedema fat should have been reduced as much as possible before surgery; patients need a good understanding of nutrition and the need to avoid weight gain post-operatively
- **Condition of the skin and tissues:** these will indicate how well the patient will heal and the likelihood of being left with excess skin
- **Presence of oedema:** a course of decongestive therapy and compression therapy may be required prior to surgery if oedema is present
- **Bleeding tendency:** the presence of varicose veins or bleeding disorders may indicate a high risk of bleeding during surgery; if the risk is severe surgery may be contraindicated
- **Ability and willingness to tolerate compression therapy:** compression therapy is very important post-operatively
- **Psychological status:** liposuction for lipoedema has a long recovery period and so psychological resilience and mental wellbeing are important to cope with and motivate self-care and ongoing wear of compression garments; patients with body dysmorphic disorder (BDD) may require psychological treatment before consideration for surgery
- **Pain management requirements:** patient's current pain levels and management strategies will help to inform discussions about post-operative requirements.

Care after liposuction beyond the immediate post-operative period may fall on lymphoedema clinics, and may prove challenging if the patient has been abroad and returns with little information about the procedure that has been performed and required aftercare.

Patients need to be advised to continue wearing the compression garments prescribed and may need advice on pain management, garment application and care, and who to contact if there are problems. Psychological support and encouragement may also be needed.

The post-operative swelling and pain take at least several months to resolve and may be perceived by the patient as signs of deterioration.

### Bariatric surgery

Bariatric surgery is not in itself a treatment for lipoedema, but as described previously weight reduction from areas of the body not affected by lipoedema or prevention of further weight gain in patients who are obese may be beneficial.

NICE has published guidelines on the criteria for considering bariatric surgery (NICE CG189, 2014). These include patients with BMI  $\geq 40$  kg/m<sup>2</sup> or 35-40 kg/m<sup>2</sup> with type 2 diabetes or hypertension who have tried all appropriate non-surgical measures to achieve weight loss.

**Box 18. Pre-operative counselling for patients with lipoedema undergoing liposuction**

- Liposuction aims to reduce fat tissue, improve limb/body shape and mobility, and reduce symptoms such as pain, but there is no guarantee that the condition will not deteriorate later
- A series of liposuction sessions may be necessary over several months, and plastic surgery may be required if large amounts of lax skin remain
- Liposuction can be carried out under general or local anaesthetic
- Risks of surgery include haemorrhage, infection, scarring, wound healing problems, altered sensation, deep vein thrombosis (DVT), pulmonary embolus, fat embolus and loose skin
- Liposuction is not a 'quick fix': pain, swelling and bruising will be marked for several months after surgery; it may take months for post-operative swelling and numbness to resolve fully; full recovery can take up to 12 months.
- Made-to-measure flat knit compression garments or adjustable wraps are applied immediately after surgery, and need to be worn for at least several months afterwards, if not on an ongoing basis ('for life')
- A patient choosing to have private surgery needs to understand that any pre- and post-operative care, e.g. MLD, bandaging and compression garments, may not be available in NHS lymphoedema clinics, and may need to be sourced privately
- Professional measurement for and fitting of compression garments is necessary to ensure correct fit and function; self-measurement is not likely to be accurate
- Weight management and physical activity, e.g. walking, continue to be very important post-operatively.

**Key points**

1. There is no evidence that liposuction cures lipoedema, but it may reduce limb bulk and so improve functioning and mobility
2. Patients should be advised to try at least 6-12 months' non-surgical treatment before undergoing liposuction
3. Pre-operative counselling is important to ensure patients understand the non-curative nature of liposuction, the long often painful post-operative course, and the need for ongoing wear of compression therapy
4. Bariatric surgery may be indicated for some patients with lipoedema who are also obese.

## APPENDIX 1: THE LYMPHATIC SYSTEM

### Function

The lymphatic system plays an essential role in fluid balance, the immune system and nutrition. Fluid leaks into tissue spaces from the blood in capillaries to provide cells with nutrients, oxygen and fluid. Formerly, it was thought that most of this interstitial fluid was reabsorbed into the venous end of the capillary. However, it is now known that most of the interstitial fluid is taken up into the lymphatic system and eventually drains back in to the venous circulation (Mortimer & Rockson, 2014).

The fluid in lymphatic vessels is known as lymph. In addition to interstitial fluid, it contains immune cells and proteins (Adamczyk et al, 2016). Lymph draining from the gut also contains fat (Lasinski, 2015).

Tissue oedema occurs when the amount of interstitial fluid formed exceeds the amount removed by the lymphatic system. This may be due to increased leakage from capillaries, e.g. as may occur in inflammation, and/or inadequate removal by the lymphatic system (Mortimer & Rockson, 2014).

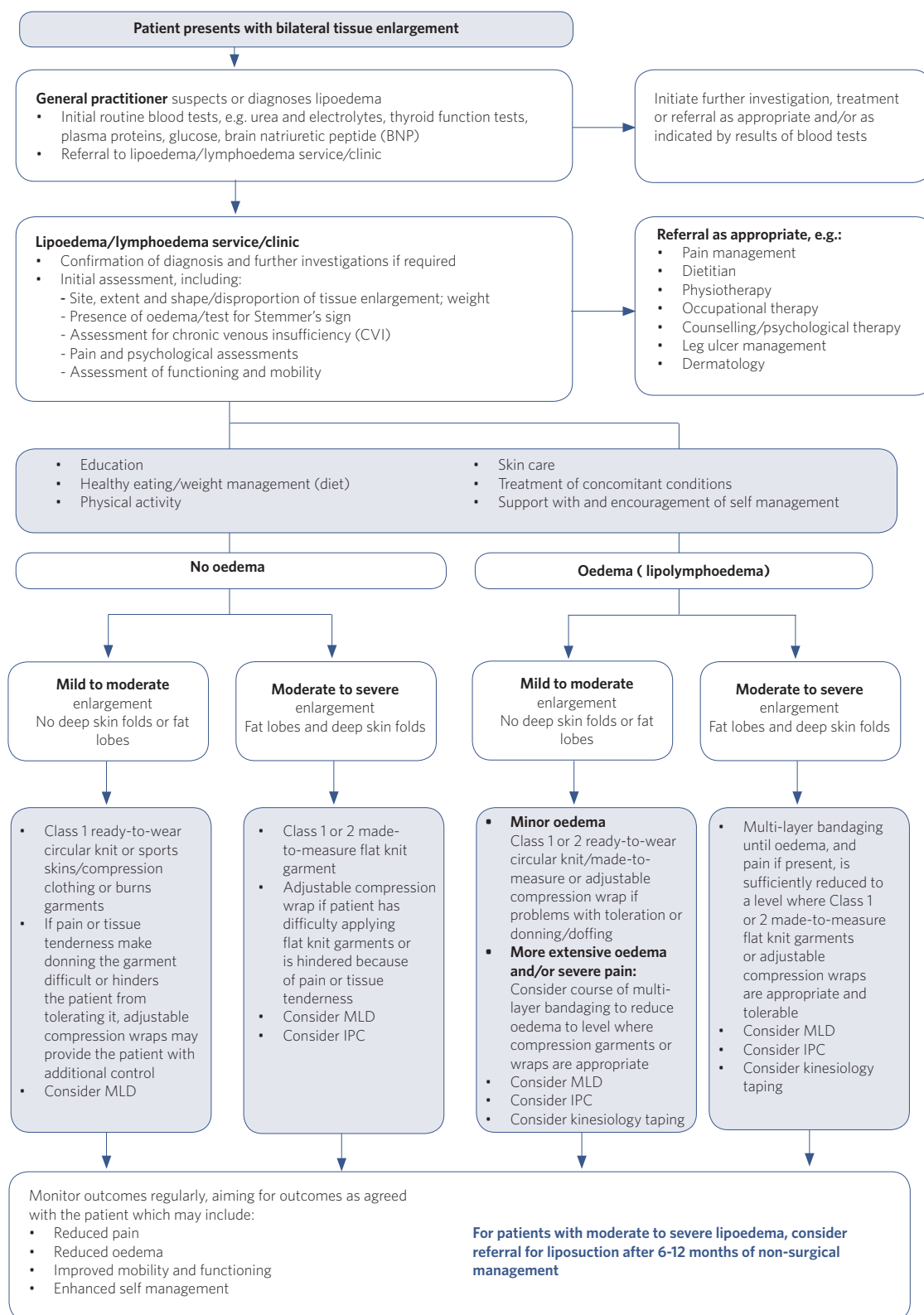
### Structure

Lymphatic vessels and lymph nodes form a network that returns lymph eventually to the blood circulatory system via the subclavian veins. There are three main types of lymphatic vessel:

- Initial lymphatics – blind-ended, non-contractile vessels that absorb lymph and drain into pre-collectors
- Pre-collector lymphatics – vessels that contain valves to prevent back flow of lymph and that drain into collector lymphatics
- Collector lymphatics – vessels that contain one-way valves and that can contract (Adamczyk et al, 2016).

Collector lymphatics contract rhythmically in response to distension to pump the lymph towards the venous system via the lymph nodes, the thoracic duct and right lymphatic trunk. Lymph flow is also assisted by pulsation of nearby arteries, skeletal muscle contraction and variations in intrathoracic pressure during breathing (Adamczyk et al, 2016).

## APPENDIX 2: PATIENT PATHWAY



N.B. This algorithm is a guide - the compression and treatment regimen for a particular patient should be individualised to take account of all of their needs  
IPC: intermittent pneumatic compression; MLD: manual lymphatic drainage

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